

# **Medical, Psychosocial and Vocational Aspects of Disability**

**Third Edition  
2009**

*Edited by*

**Martin G. Brodwin, Ph.D., C.R.C.**

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**John Howard, M.D., M.P.H.**

**Erin R. Brodwin, B.A.**

*Elliott & Fitzpatrick, Inc.*

**Athens, GA**

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# PREFACE

Our sincere thanks to the more than 90 universities and colleges in the United States and beyond that have selected the first and second editions of *Medical, Psychosocial, and Vocational Aspects of Disability* as a textbook for their rehabilitation programs. It has also been selected by continuing education programs, practitioners, and rehabilitation public agencies as a resource book. This third edition has three new topics: human body systems, complementary and alternative medicine, and pervasive development disorders (autism). Different authors for this third edition have written eight of the chapters that were in the second edition. Four chapters from the second edition have been combined into two chapters: hematological disorders (hemophilia and sickle cell disease) and diabetes mellitus and renal disease. All other chapters have been revised and updated.

## Background

The enactment of the Americans with Disabilities Act signaled a new era in America for persons with disabilities. These individuals, often excluded from mainstream society, have been able to participate in all phases of life to a far greater extent. Rehabilitation counselors, as well as educators, nurses, counselors in other settings, work evaluators, case managers, and family members have the responsibility to facilitate increasing progression into independent living, education, employment, and leisure-time activities for persons with disabilities. The Americans with Disabilities Act defined a “person with a disability” as someone who: “(1) has a physical or mental impairment that substantially limits one or more major life activities; (2) has a record of such an impairment; or (3) is regarded as having such an impairment.”

Rehabilitation involves counseling a person with a disability to assist that individual to acquire skills necessary for maximum functioning and greater independence. Professionals who have an understanding of medical aspects of disability and its relationship to employment are unique within the counseling profession. Counselors need to read, understand, and interpret medical reports and other medical information regarding clients for whom they provide rehabilitation services. Knowledge of chronic illness and disability is crucial for counselors working with persons with disabilities. With comprehension of this information, counselors are able to determine their clients’ functional limitations and potential for rehabilitation.

Knowledge of functional limitations allows the counselor and the individual with a disability to obtain information necessary for developing positive directions for rehabilitation. Rehabilitation services may include job modification, reasonable accommodation, educational pursuit, training or on-the-job training, supported employment, job placement, and independent living. The purpose of any rehabilitation counseling goal is to empower the individual with a disability to successfully participate in all phases of life.

## Overview

This book is a textbook for students and a reference book for practicing counselors and other helping professionals. The intended audience includes rehabilitation counselors in both public and private sectors, rehabilitation educators and their students (undergraduate and graduate programs), vocational experts, work evaluators, counselors in a variety of settings, and other helping professionals. Allied health professionals in related disciplines also will benefit from the information within this text.

The reader is encouraged to have a medical dictionary available when reading this book. A glossary was not included, as the editors have found glossaries to be inadequate, both in providing sufficient explanations and in defining most of the required vocabulary. Various words throughout the text are provided with a short definition in parentheses.

This edition of *Medical, Psychosocial, and Vocational Aspects of Disability* contains thirty-three chapters encompassing the common disabilities encountered in the field of rehabilitation. Most of the

chapters have case studies to help stimulate thinking and discussion about the particular topic. All the cases are hypothetical and do not relate to any particular individual. The book has a strong multicultural emphasis throughout the case studies. We believe the richness of cultural diversity adds an important dimension to the material.

We suggest the reader begin with Chapter 1, “A Case Study Approach: Rehabilitation, Intervention, and Assistive Technology” as this chapter describes the case study analysis approach used throughout the book. Subsequent chapters can be reviewed in the order that best suites the reader.

## **Acknowledgments**

The editors did their utmost to maintain the meaning and philosophy of each author and, at the same time, have the chapters conform to a similar style. We sincerely appreciate each author’s time and patience during the difficult writing and editing process. The authors of the chapters in this book each spent many hours writing and revising their manuscripts. We believe that through their efforts this text will provide the reader with an excellent resource of medical aspects of chronic illness and disabling conditions and their relationship to employment issues.

We also want to acknowledge the assistance of our Student Reader for this third edition, Ryan Kahanowitch, B.S. Ryan generously gave his time to assess the chapters submitted. We are grateful for his time and expertise.

# Chapter 1

## A CASE STUDY APPROACH, REHABILITATION INTERVENTION, AND ASSISTIVE TECHNOLOGY

*Martin G. Brodwin, Ph.D., C.R.C.*  
*Sandra K. Brodwin, M.S., C.R.C.*

### Introduction

Rehabilitation professionals need expertise concerning medical aspects of disabling conditions to work effectively with persons who have disabilities. Increasingly, rehabilitation counselors are expected to have greater knowledge and skills, and demonstrate case management expertise when working with employers to facilitate the return to work for individuals with chronic illnesses and disabilities. Counselors apply this expertise both when developing educational/vocational plans and determining functional limitations and rehabilitation potential. Knowledge of medical aspects of disability is a significant component when developing rehabilitation interventions. The “Vocational Profile” case study approach is one holistic method of evaluating a person’s vocational potential using analysis of age, education, work history, occupationally significant characteristics of work, and transferable skills.

Since the basic criteria for the acceptance to vocational rehabilitation is the existence of a disability which results in a handicap to employment, one of the attributes differentiating rehabilitation counselors from other counselors and mental health professionals is specialized knowledge and expertise related to disability. As no two people are alike, there are a variety of characteristics affecting each person’s potential to benefit from the provision of rehabilitation services. A similar medical diagnosis will have different effects, depending on one’s psychosocial circumstances, adaptation to functional limitations, vocational skills, education, and occupational history. Examples of components that affect rehabilitation potential for employment include (Brodwin & Falvo, 1998; Brodwin, Parker, & DeLaGarza, 2003):

- Severity of the disability (residual functional capacities and limitations).
- Age at onset of disability.
- Current age.
- Gender and ethnicity.
- Psychological characteristics.
- Pre-existing physical conditions.
- Social functioning.
- Educational and vocational training.
- Employment.
- Psychosocial adjustment to disability.
- Use of assistive technology.

Development of vocational and educational rehabilitation plans and programs relies heavily on the expertise of rehabilitation professionals. A realistic and practical objective for a vocational or educational plan must be formulated according to an individual's physical, intellectual, and emotional capacities. To establish a realistic and practical objective, the rehabilitation professional applies knowledge of the vocational profile, medical aspects of the disability, the individual's interaction with the environment, and the person's effort and persistence to accomplish goals.

This chapter addresses the functions of rehabilitation professionals providing services for a variety of rehabilitation systems. The authors believe the use of the "vocational profile" approach will enhance a thorough case analysis and improve the chances for a person's successful return to employment. This chapter focuses on the importance of the "holistic approach" to enhance the potential for successful and satisfying rehabilitation. Aspects of functional limitations, rehabilitation potential, case intervention, and assistive technology are discussed.

## **Vocational Profile**

The words, "Vocational Profile" are used specifically by Vocational Experts (VEs) for the Social Security Administration's (SSA) Office of Disability Adjudication and Review when testifying about disability-related issues. More broadly, however, the SSA vocational profile model is part of a nationally-mandated program that is uniform throughout the country. Vocational profiles are developed to include age, education, and work history categories as they relate to individuals, as well as occupationally significant characteristics and work skills transferability. The following categories - **age** (SSA Section # 404.1563), **education** (SSA Section # 404.1564), and **work experience** (SSA Section #s 404.1565, 404.1567, and 404.1568) - are taken from the guidelines of the Social Security Administration (Office of the Federal Register, 2001).

### ***Age Category***

Age is considered when evaluating the total person; as one ages, adaptation to new and unfamiliar situations becomes increasingly difficult. The importance of age as an evaluation factor varies with a person's work history. For example, if an individual has had only physically demanding jobs and developed few work skills, age can play a crucial role in rehabilitation. The following categories are taken from the guidelines of the Social Security Administration (Office of the Federal Register, 2001).

#### ***Younger person***

An individual is considered to be within this category if under the age of 50. Generally, if in this category, the person's age will not impact the ability to adapt to new work.

#### ***Person approaching advanced age***

An individual between 50-54 years of age is considered a "person approaching advanced age." A person in this age category who has a severe impairment and limited work experience may have major difficulty adjusting to many jobs within the labor market.

#### ***Person of advanced age***

This category is appropriate if a person is between 55-59 years of age. With advancing age, it becomes more difficult to obtain employment, especially with an unskilled work background, a history of physically arduous work, and minimal education. Skills that can be transferred to similar jobs and work activity (often less physically demanding) become more relevant as the individual ages.

#### ***Close to retirement age***

When individuals are 60 years of age or older, they are considered, by Social Security regulations to be "close to retirement age." Skills at this age need to be highly transferable and are pivotal to success in the rehabilitation process.

## ***Educational Level***

Educational level is the amount of formal schooling a person has achieved. Grade level is only one means of assessment. When evaluating rehabilitation potential, the counselor needs to consider how long ago a person attended school, the quality of the educational experience, additional informal or formal education, and any other training that occurred.

### ***Illiteracy***

"Illiteracy" means an inability to read or write. People are placed in this category if they cannot read or write simple messages, such as basic instructions, inventory lists, simple messages, and the like.

### ***Marginal education***

Generally, "marginal education" indicates a formal education at the 6<sup>th</sup> grade level or less. The person who has a "marginal education" is limited to basic reasoning, arithmetic, and language skills.

### ***Limited education***

A person of "limited education" has been formally educated between the 7<sup>th</sup> and 11<sup>th</sup> grades (without having attained a high school diploma or its equivalence).

### ***High school education and above***

"High school education and above" implies achieving a high school diploma or its equivalence. Attainment of a General Equivalency Diploma (GED) is at this level. People with additional education beyond high school are classified within this category. Individuals so classified have obtained reasoning, arithmetic, and language skills required for more complex work activity.

## ***Work History***

This section of the vocational profile is subdivided into several categories. These include skill requirements, physical exertion, occupationally significant characteristics, vocational skills, and transferability of skills.

## ***Skill Requirements***

### ***Unskilled Work***

Work that requires little or no judgment for the completion of simple duties and is learned on the job in a short time is unskilled work. Unskilled jobs require 30 days or less to learn. While they are quickly learned, these jobs frequently require considerable strength. Primary work duties for an unskilled occupation could, for example, involve stock or material handling, machine feeding, sorting, simple assembling, or machine tending. Little specific vocational preparation or judgment are thus required. **A person does not gain work skills (transferable skills) by working in unskilled jobs.** When a worker develops skills, the work is considered to have changed to semiskilled work.

### ***Semiskilled Work***

Semiskilled work requires some skills but not complex work activity. This category consists of activities that are less complex than skilled work but more complex than unskilled work and can involve dexterity tasks such as working with tools, equipment, and measuring devices. These jobs may involve alertness and close attention to observing machine processes or inspecting, testing, or quality control. Such aspects may include guarding equipment, materials, or persons against damage, loss, or injury. Typically, semiskilled work requires training from over 30 days to a maximum of two years.

### ***Skilled Work***

This category of work requires an individual to use judgment to determine fairly complex machine and manual operations performed to obtain the proper form, quality, and quantity of materials produced. Skilled work may require laying out work, estimating quality, determining suitability and needed quantities of material, making precise measurements, reading blueprints or other specifications, or making complex computations or mechanical adjustments to control or regulate the work. Other skilled jobs involve dealing

with people, facts or figures, or abstract ideas at a high level of complexity. Skilled work typically requires more than two years to learn.

### ***Physical Exertion Requirements***

When reviewing a client's past relevant work experience, the counselor evaluates the level of physical exertion required at work. The individual's residual functional capacity (RFC) in conjunction with age, education, and work experience, are considered to determine whether the individual can engage in any other substantial gainful work which exists in the national economy. Physical exertion requirements are summarized in the following categories. Each functional level is defined (in accordance with terms used by the United States Department of Labor) by the extent of its requirements in the primary strength activities of sitting, standing, walking, lifting, carrying, pushing, and pulling. If medical reports regarding the client's disability indicate return to work at one exertional level, the counselor and client need to investigate work situations requiring equal or lesser physical exertion.

"Occasionally" means occurring from very little up to one-third of the time. Typically, this is defined as ranging from 5 minutes up to 2.5 hours per day. A "frequent" activity occurs one-third to two-thirds of the time (a total of 2.5 to 5.0 hours). Since frequent lifting or carrying requires being on one's feet up to two-thirds of a workday, the full range of light work requires standing or walking, off and on, for a total of approximately 6 hours of an 8-hour workday. Sitting may occur intermittently during the remaining time. The lifting requirement for the majority of light jobs can be accomplished with occasional, rather than frequent, stooping. "Continuously" is defined as an activity which occurs for a total of more than 5.0 hours per day.

#### ***Sedentary Work***

The regulations define sedentary work as involving lifting no more than 10 pounds at a time and occasionally lifting or carrying items such as files and small tools. Although sitting is involved, a certain amount of walking and standing may be necessary in carrying out job duties. Jobs are sedentary if walking and standing are required occasionally or less and other sedentary criteria are met. By its very nature, work performed primarily in a seated position entails no significant stooping.

#### ***Light Work***

The regulations define light work as lifting no more than 20 pounds at a time with frequent lifting or carrying of objects weighing up to 10 pounds. Even though the weight lifted in a particular light job may be very little, a job is in this category when it requires a good deal of walking or standing – the primary difference between sedentary and most light jobs. A job is also in this category when it involves sitting most of the time but with some pushing and pulling of arm-hand or leg-foot controls, which require greater exertion than in sedentary work, such as mattress sewing machine operator, motor-grader operator, and road-roller operator (skilled and semiskilled jobs in these particular instances). Relatively few unskilled light jobs are performed in a seated position.

#### ***Medium Work***

The regulations define medium work as lifting no more than 50 pounds at a time with frequent lifting or carrying of objects weighing up to 25 pounds. A full range of medium work requires standing or walking, off and on, for a total of approximately 6 hours in an 8-hour workday in order to meet the requirements of frequent lifting or carrying objects weighing up to 25 pounds. As in light work, sitting may occur intermittently during the remaining time. Use of the arms and hands is necessary to grasp, hold, and turn objects, as opposed to the finer activities in much sedentary work, which require precision use of the fingers as well as use of the hands and arms. The lifting required for the full range of medium work usually requires frequent bending-stooping. (Stooping is a type of bending in which a person bends his or her body downward and forward by bending the spine at the waist. Crouching is bending both the legs and spine in order to bend the body downward and forward.) In most medium jobs, being on one's feet for most of the workday is crucial.



**Heavy Work**

Lifting a maximum of 100 pounds occasionally with frequent lifting or carrying up to 50 pounds defines heavy work.

**Very Heavy Work**

Very heavy work involves lifting more than 100 pounds occasionally with frequent lifting or carrying of 50 pounds or more.

The following table summarizes the above exertional categories of work.

**Table 1**  
**Exertional Categories of Work**

Category	<u>Lifting/Carrying</u>	
	Frequent	Occasional (Maximum)
Sedentary	2-5 pounds	10 pounds
Light	10 pounds	20 pounds
Medium	25 pounds	50 pounds
Heavy	50 pounds	100 pounds
Very Heavy	50 pounds or more	100 pounds or more

***Occupationally Significant Characteristics***

Occupationally significant characteristics are distinctive elements that contribute to the job, work environment, and work functions, but **do not involve skills or characteristics of a person**. Occupational characteristics exist independent of the worker and often can be modified, if necessary. Examples of occupationally significant characteristics include: eye-hand-foot coordination, visual perception, being around other people, exertional level of work (sedentary, light, medium, heavy, and very heavy), inside work, routine and repetitive job functions, activities requiring occasional bending and stooping, work involving fumes and irritants, among others.

An individual who has worked as a secretary, for example, may have some or all (depending on the particular job) of the following occupationally significant characteristics: indoor work, eye-hand coordination, sedentary work activity, work with other people, use of office equipment, clerical work, and work with information. In contrast, the work of a truck driver may include characteristics such as: manual dexterity, eye-hand-foot coordination, work within the transportation industry, medium exertion, working alone, and being in a variety of environmental conditions. **As can be seen from these examples, occupationally significant characteristics do not involve the acquisition or use of skills.**

***Vocational Skills and Transferability of Skills***

Skills involve abilities that are learned during work, training, or an educational program. They are distinct from occupationally significant characteristics because they require work experience and the acquisition of abilities. Skills involve expertise or knowledge specific to work functions, such as the ability to use personal judgment, work with specific tools and equipment, operate complex machinery, and/or work with people or ideas at a high level of intricacy.

The skills of a rehabilitation counselor include ability to communicate and organize, ability to counsel individuals regarding personal concerns, career development, and vocational pursuits; skills involved in helping persons secure employment; management and supervision capabilities; knowledge of medical aspects of chronic illness and disability, medical terminology, and medical treatments; and ability to work with troubled and distraught individuals with serious problems. In contrast, the skills of a secretary include: clerical skills; ability to operate various office machines; capability to use a word processor or

computer; compile, type, and file letters; aptitude to organize and maintain a record-keeping system; answer business telephones; communication capacities; and organizational skills required to maintain the clerical flow of an office.

The *Dictionary of Occupational Titles (D.O.T.)* (U. S. Department of Labor, 1991) and other resources can be used effectively when transferring skills from one job to another. Only skilled and semiskilled work provides transferable skills. Unskilled work, by definition, does not involve skills and, therefore, transferability of skills is not relevant to these positions. Skills, abilities, and knowledge found in skilled and semiskilled work can be transferred to a position requiring equal or lesser skills, but not to a position of greater skill requirements. An employee may be promoted to a more skilled position before acquiring the skills of that position and then learn skills on the new job.

An example of transferability from a skilled position is illustrated by the occupation of rehabilitation counselor. Duties of this job can transfer to other skilled and semiskilled positions. Skilled positions include manager of a human resource department, college counselor, rehabilitation director, academic advisor, supervisor, teacher, mental health clinician, parole and probation officer, and vocational evaluator. The skills of a rehabilitation counselor also transfer to semiskilled work activities such as personnel interviewer, job analyst, job placement specialist, research assistant, and work evaluation technician. A semiskilled occupation, such as secretary, has transferable skills to other semiskilled work such as office clerk, receptionist, file clerk, general office worker, and word processor. Since the work is semiskilled, there is no transferability to skilled work.

When analyzing rehabilitation potential, all job possibilities need careful evaluation. Initial exploration includes determining the skill requirements of previous jobs held by the client to indicate potential transferable skills. The rehabilitation counselor can then identify other jobs within the same industry that use these skills. Next, the research process expands to jobs within related industries using transferability of skills. Finally, if no positions exist within the client's physical exertional (or emotional) restrictions using transferable skills, the counselor investigates alternative rehabilitation options. While determining transferability of skills, the counselor needs to assess how long ago the job was performed and whether the skills are outdated or forgotten. Another aspect to consider is length of time the individual held the job.

If the client's skills are not immediately transferable to jobs within the current labor market (using direct job placement), the rehabilitation professional may consider on-the-job training, vocational training, and educational programs. The counselor and client use various criteria when selecting the type of plan best suited for the client.

The rehabilitation professional in a provider system applies expertise in medical aspects of chronic illness and disability to develop vocational and educational plans with practical, realistic, and obtainable objectives. The basis for the rehabilitation objective includes analysis of the client's age, education, work history, occupationally significant characteristics, transferable skills, capacities, and functional limitations (Brodwin, 2008; Brodwin & Falvo, 1998). Roessler & Rumrill (1995), in a discussion of reasonable accommodations and job retention, discussed the need for postemployment services. These services reduce or remove barriers to successful employment outcomes. Three factors are crucial to successful job retention: (a) identification of barriers to employment and job accommodation strategies, (b) initiation of requests for accommodation, and (c) implementation of accommodations with cooperation of the employer.

## **Functional Limitations**

By definition, a functional limitation is an inability to perform an action or set of actions, either physical or mental, because of a physical or emotional restriction (often referred to as a disability) (Rubin & Roessler, 2008). A clearly specified limitation of function helps the counselor understand the performance limitation(s) of the client. Restrictions can be analyzed with a clear understanding when medical conditions are stated in functional terms.



### **Examples**

**Client 1:** An individual has a below the knee (BK) amputation of the left lower extremity. In functional terms, this person may be limited to maximum ambulation of one hour during an eight-hour workday, no ambulation on rough or uneven surfaces, no stair climbing, and no continuous standing.

**Client 2:** A person has a low back injury and a problem with lifting and carrying. Functionally, the individual is limited to lifting a maximum of 20 pounds on an occasional basis with repetitive lifting and carrying not to exceed 10 pounds (light work).

**Client 3:** This person has a psychiatric diagnosis involving moderate depression and anxiety. In functional terms, this individual needs low stress work with an understanding female supervisor (because of a problem with dominant male authority figures).

**Client 4:** The individual has a diagnosis of schizophrenia, chronic undifferentiated type. In functional terms, this person needs work that involves simple, routine, and repetitive activities with minimal personal interaction and a structured work environment.

Medical conditions described in functional terms enable the counselor to more readily understand the client's limitations. First, the rehabilitation professional assesses the client's vocational profile. The second step is to review the medical file and make sure the limitations are understood in functional terms. The rehabilitation counselor then determines the potential for vocational rehabilitation.

## **Rehabilitation Potential**

For individuals to maximize their potential for rehabilitation, four factors need to be assessed: (a) the attainment of increased functioning in the areas of physical and emotional growth and development; (b) a sense of well being; (c) development of a personally satisfying level of independence; and (d) compatibility between the work and the person's capabilities and limitations. These four factors relate to work and psychosocial adjustment. One's attitude and adaptation to disability also impact rehabilitation potential and success in a vocational or educational program.

Rehabilitation systems (e.g., disability management, long-term disability, Social Security, state vocational rehabilitation, and independent living) define a client's rehabilitation potential differently. A person may have rehabilitation potential within an independent living program but not with the state department of rehabilitation. Another individual may be considered to have rehabilitation potential within a public agency but not in certain private agency settings (such as long-term disability).

Counselors need an understanding of the distinct requirements of the particular rehabilitation system that is providing services (Brodwin, 2008). Each system has its own advantages and limitations. Once the counselor determines the client's rehabilitation potential within the specific system, a rationale is developed and presented to support the decision as to what rehabilitation services will be provided. The counselor needs to clarify if the medical conditions are temporary or permanent, and whether they may be expected to improve, remain the same, or deteriorate.

Additionally, one needs to assess whether the particular rehabilitation system providing services influenced this determination (e.g., eligibility criteria). The counselor should inform the client of the decision and the basis for that decision. If denying services, the counselor may be able to refer the client to another system where appropriate and applicable services or benefits will be provided. If a client does not qualify for vocational rehabilitation services, the counselor may refer the person for Social Security benefits (SSDI, SSI). Each rehabilitation system evaluates rehabilitation potential, including length of time typically provided for rehabilitation services, medical expenses, rehabilitation costs, and the likelihood of return to gainful employment. As stated by Bolton, Bellini, and Brookings (2000), "the goal of the rehabilitation service delivery process is to maximize the probability of successful rehabilitation outcomes for people with disabilities" (p. 10).

## **Rehabilitation Intervention**

Through the provision of rehabilitation intervention the counselor may be able to modify environmental factors for clients with disabilities to empower them to succeed at productive activities.

Modifications of the environment include accommodation, job modification, and restructuring of job sites (Wright, 1980).

### ***Job Accommodation, Modification, and Restructuring***

“Reasonable accommodation” is a logical adjustment made to a job or work environment that enables a qualified person with a disability to perform the duties of the position (Berkeley Planning Associates, 1982). Reasonable accommodation recommendations must be considered on an individual basis for each employee and employer. Cooperation on the part of the employer is essential to successful accommodation on the job.

Section 503 of the Federal Rehabilitation Act of 1973 and the Americans with Disabilities Act (ADA) of 1990 mandated reasonable accommodation. Employers have an obligation to make reasonable accommodation for physical and mental limitations, unless the accommodation imposes an “undue hardship” on the employer. Undue hardship depends upon several factors including cost, financial resources of the company, overall size of the employer, employer’s operation (including composition and structure of the workplace), and nature of the proposed accommodation.

The ADA stated that modifications necessary under the “reasonable accommodation” provisions include (Rubin & Roessler, 2008; West, 1991): modifying the physical layout of a facility to make it accessible to employees who use wheelchairs or who have other impairments that make access difficult.

1. Restructuring a job to enable the person with a disability to perform the essential functions of the job.
2. Establishing a part-time or modified work schedule for employees with disabilities who are not capable of working a typical work day or work week.
3. Reassigning a person with a disability to a vacant position.
4. Acquiring or modifying equipment or devices (e.g., buying a hearing telephone amplifier for a person with a hearing impairment).
5. Adjusting or modifying examinations, training materials, or policies (e.g., giving an application examination orally to a person with dyslexia, or modifying a policy against dogs in the workplace for a person with a service dog).
6. Providing qualified readers or interpreters for people with vision or hearing impairments.
7. Providing an employee with a psychiatric illness a private place to work to alleviate stress.

Reasonable accommodation rarely involves considerable cost. Berkeley Planning Associates (1982) conducted a study for the U. S. Department of Labor on accommodation in private sector employment and noted the following results:

- About 50% of the reasonable accommodations surveyed cost less than \$50.
- Thirty percent cost between \$50-\$500.
- Ten percent cost between \$500-\$2,000.
- Ten percent cost in excess of \$2,000.

The Department of Labor study concluded that, based on the above figures, reasonable accommodation is “no big deal.” This study reported that the most expensive and extensive accommodations are usually provided to individuals with blindness and to persons who use wheelchairs. Frequently, reasonable accommodations that are expensive and extensive are provided by employers to current employees to maintain their ability to continue work. Assuming cooperation between employers and employees, most accommodations cost less than \$100 (Muther, 1996).

The public perceives accommodation efforts as helping persons with chronic illness and disabilities become employed or remain successful on the job. There appears to be no significant relationship between accommodation and upward mobility, either by providing an advantage to a specific employee or in limiting job potential. Highly skilled workers more often receive environmental adaptations of the workplace and special equipment, while lower skilled workers receive job redesign, retraining, and selective placement.

Large firms are most likely to hire and accommodate persons with disabilities for the following reasons (Berkeley Planning Associates, 1982; Rubin & Roessler, 2008):

1. The existence of affirmative action programs in larger firms.
2. The likelihood of encountering persons with disabilities because of the sheer numbers of workers employed.
3. The much greater diversity of job types in larger firms that lead to more possibilities of hiring and greater flexibility in job assignments.

Smaller firms are less likely to have knowledge of and information about the ADA and personnel to investigate accommodation.

### ***Types of Reasonable Accommodation***

#### **I. *Physical access accommodation***

- A. A change or modification of the physical structure
- B. Examples - Accessibility/ambulation solutions
  1. Situate job on first floor
  2. Situate job near employee parking lot
  3. Situate job near restroom

#### **II. *Resource accessibility accommodation***

- A. Providing an assistive person to enable the individual to do the job duties
- B. Examples
  1. Hearing impairment - Provide note taker or sign language interpreter
  2. Visual impairment - Provide reader or note taker
  3. Developmental disability (mental retardation) - Provide a job coach

#### **III. *Adaptive equipment accommodation***

- A. Provision of “low tech” and “high tech” assistive devices
- B. Examples
  1. Person with arthritis or carpal tunnel syndrome – Provide special pen/pencil holders
  2. Orthopedic problems
    - a. Provide desk or chair modifications
    - b. Provide speaker and earphone on telephone
  3. Reaching problems
    - a. Provide a turntable on a desk
    - b. Provide a special desk that has easy accessibility
  4. Neck problems - Provide a slant board on a desk
  5. Visual impairment - Provide a talking calculator or talking computer
  6. Hearing impairment
    - a. Provide a telephone amplifier
    - b. Provide a speaker telephone
  7. Quadriplegia - Provide a computer with specialized keyboard and an electronic wheelchair with assistive devices

#### **IV. *Job modification***

- A. Modifying the performance of job duties while maintaining the same job duties
- B. Examples

1. Energy or ambulation problems - Salesperson can do more telephone sales and decrease field appointments
2. Energy problem - Design drafter can work part-time and receive an adjusted salary
3. Orthopedic problems
  - a. Parking-lot attendant can use a chair for sitting instead of standing throughout the day
  - b. Warehouse worker can lift and carry lesser weights by making more frequent trips with lighter weights
4. Scheduling - Secretary can leave early for physical therapy appointments by coming to work earlier

#### **V. Job restructuring**

A. Changing some of the actual job duties performed (alter, eliminate, or replace job duties)

B. Examples

1. Ambulation problem - Rehabilitation counselor can be assigned more office work (labor market surveys, job development, job placement, initial interviews) and less field work (on-site job analysis, school visits, employer visits)
2. Emotional stress
  - a. Social worker can be assigned more case file analysis and paperwork and less interviewing and field visits
  - b. Attorney can be assigned more research, case preparation, and legal briefs and less time litigating in court
3. Lower extremity problems - Shipping and receiving clerk can be assigned more clerical work (typing reports, bills of lading, and expediting) and less lifting, carrying, standing, walking, and forklift driving

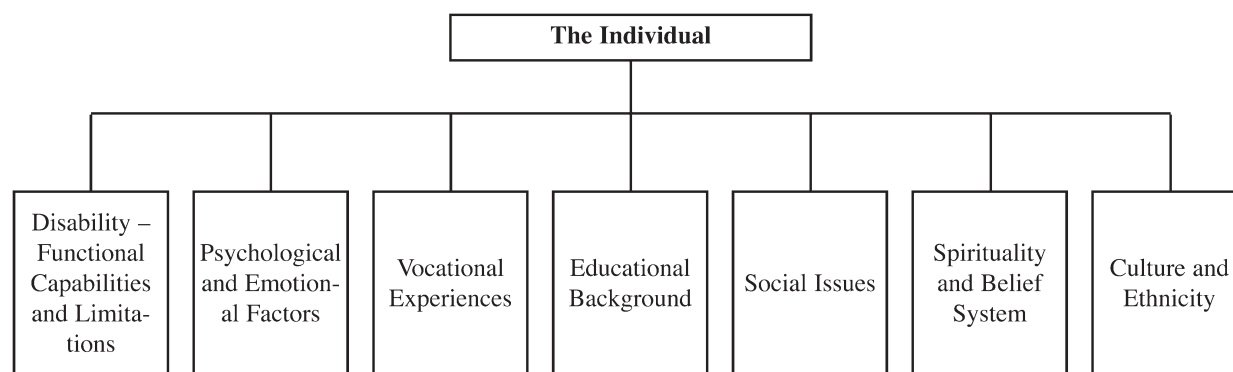
Job Accommodation Network (JAN) is a resource available for additional information about employer accommodations. They have an 800 toll-free telephone number. This telephone service is provided at no cost and allows the counselor, employer, and consumer access to information on restructuring possibilities and costs. Consultants are available Monday through Friday. Before calling JAN one needs the specific medical restrictions of the client, job duties which are precluded as a result of the restrictions, and general information about the industry in which the job is located.

## **A “Wholistic” Approach To Rehabilitation**

The present authors and Fernando A. Tellez, M. D. (personal communication, September 2001) believe “wholistic” is more appropriate than “holistic,” as the stem “whole” means entire or complete, whereas the stem “hole” indicates an aperture or opening. Wholistic rehabilitation stresses the importance of the person as a whole and the interdependence of the various facets of the individual.

An individual, with or without a disability, is not an isolated entity. There is continuous interaction between the person and the environment. Interventions in one area of the person’s environment have an influence on other areas. Seven areas most pertinent to a wholistic approach are the person’s disability, psychological status, vocational experiences, educational background, social issues, spirituality/belief system, and culture (see Figure 1). Disability can be expressed in functional limitations. Psychological status involves emotional factors that impede rehabilitation planning (Vash & Crewe, 2004). The more extensive the person’s vocational experiences and education, the greater the opportunities for success. Social functioning of the person involving interaction with family, friends, and associates is a factor to evaluate. Spirituality and one’s belief system can enhance rehabilitation outcomes (Joseph E. Havranek, Ed.D., personal communication, March 2000). The counselor needs an understanding, careful consideration, and acceptance of the client’s culture and ethnicity. Viewing rehabilitation from a wholistic perspective enhances the probability of a successful outcome.

**Figure 1**  
**A Wholistic View of the Individual**



## **Assistive Technology**

As all areas of our society continue to become more technologically oriented, individuals who have chronic illness and disabilities are benefiting by assistive technology and, hence, becoming more mainstreamed into society. Society benefits as people with disabilities become more independent in all areas of life and, therefore, less reliant on public services. The income for individuals with disabilities increases and therefore their purchasing power to buy goods, services, and products. The field of assistive technology (AT) is providing an effective means to help increase or restore functioning, enhance quality of life, and eliminate barriers, both physical and attitudinal.

### ***Definitions***

Assistive technology was defined in the “Technology-Related Assistance for Individuals with Disabilities Act of 1988 as “any item, piece of equipment, or product system, whether acquired commercially off the shelf, modified, or customized, that is used to increase, maintain, or improve functional capabilities of people with disabilities.” Technology service is “any service that directly assists an individual with a disability in the selection, acquisition, or use of an assistive technology device.” More than 20 million Americans with disabilities are using assistive devices, equipment, computers, and other apparatus. People benefit from assistive technology at work, in school, at home, and in leisure time activities.

### ***Use of Technology***

Through a greater understanding of technology as applied to people with chronic illness and disabilities, practitioners will be in a position to provide more meaningful, practical, and realistic clinical counseling and services for their clients. Technology is having a profound impact on improving the lives of people with disabilities. As noted by Scherer (2007), technology is radically changing the lives of individuals who have disabilities. Assistive technology has helped enhance the quality of life and extend the lifespan of persons with congenital and developmental disabilities, as well as those who have acquired disabilities and chronic, medical conditions. Devices and equipment may be low-tech (mechanical) or high-tech (electromechanical or computer-related) and can help compensate for sensory and functional loss. Assistive technology provides the means to move (e.g., adaptive equipment on vehicles, wheelchairs [including power wheelchairs], scooters, lifts), speak (e.g., augmentative and alternative communication devices), read (e.g., Braille input, voice recognition devices), hear (telecommunication devices for the deaf [TDD], hearing aides, audio loops), and manage self-care tasks (e.g., remote environmental control systems, prosthetic and orthotic devices, devices to aid a person in grooming, eating, and other activities of daily life).



## ***Examples of Technology***

### ***Robotics***

An example of technology includes the science of robotic controls. Robotics are able to accommodate for manipulative tasks, perform functions through prosthetic (e.g., artificial extremity) applications, render movement in spinal cord injuries (e.g., paralysis), and perform task operations in fixed workstation (at the work site) technologies (Cook, Pulgar, & Hussey, 2008). A robotic arm can be attached to a powered wheelchair and controlled by a direct selection and toggle interface. It provides assistance with such functions as drinking, removing items from shelves, desktop manipulation of objects, and recreation (e.g., games). Fixed workstation (desktop) robots assist with manipulation of objects. A desktop vocational robot assistant can perform activities of daily living tasks (e.g., meal preparation and feeding, writing, retrieving books, keyboard operations, and use of a telephone). It also assists with hygiene by washing and drying the operator's face, brushing teeth, combing hair, and shaving, as well as providing access to video and other games and craft (leisure time) activities.

### ***Sensory Technology***

Accommodations for people with visual impairments include both optical and non-optical devices for low vision. Examples of optical devices are magnifiers, specially coated lenses, and telescopes. Non-optical visual aides include talking clocks and watches, talking calculators, closed-circuit televisions that enlarge print electronically, and personal computers and peripherals with the capacity of print magnification, speech output, and optical scanning (Brodwin, Star, & Cardoso, 2007). Additionally, hearing aids, telecommunication devices for the deaf (TDD), cochlear implants, electronic ears, amplified telephones, and audio loops are helpful technology devices for persons who are deaf or hard-of-hearing.

### ***Eye-tracking Technology***

Eye-gaze technology can track the movement of one eye to activate a computer. The individual must have sufficient head control and the ability to gaze directly at a camera. Derived from military eye control systems, the camera is typically attached to the head; however, the weight of the camera requires the head to be positioned and supported. Current scientific research promises enhanced eyeglass systems. Researchers described innovative gaze technology that features virtual reality goggles with an integrated camera and viewing screen to provide communication through icon choice. The user chooses targets by gazing at the icons on the computer screen; the camera, integrated within the goggles, tracks eye gaze motion and relays information for instantaneous processing (Brodwin et al., 2007; Cook et al., 2008; Scherer, 2007).

## ***Benefits of Technology***

Upward social mobility is more realistic because of increased educational and employment opportunities and a greater sense of personal well-being (adjustment to disability). Technological devices, equipment, and services have allowed many people with disabilities to: (a) exert greater control over their own lives; (b) participate in and contribute more fully and readily to activities in their own homes, schools, employment, and communities; (c) interact to a far greater extent with non-disabled individuals; and (d) benefit from opportunities that are taken for granted by individuals who do not have disabilities (Brodwin et al., 2007; Scherer, 2007). The major reason cited for discontinued use of computers was a lack of 'relative advantage'. This concept encompasses the effectiveness of the product, its reliability, ease of use, comfort, and enhancement of the user's performance; the user needs to be involved throughout the process (Riemer-Reiss & Wacker, 2000). As many forms of assistive technology for people with disabilities are expensive, this needs to be carefully taken into account before recommending a device. This is not to say that most devices and equipment are costly as some are inexpensive and easy to find.

The goal of AT is to enhance functional independence for individuals who have disabilities. The focus, therefore, is not on the disability, but on the remaining functional abilities that people use to accomplish their chosen objectives. Expectations a person has concerning technology are individualized and influenced by the disability and the consequences of that particular disability. Through exploration of the individual's capabilities, functional limitations, and feelings about technological devices, practitioners can most effectively provide rehabilitation services.

## Case Study - A Vocational Profile Approach

The following case study is an example of those found in the remaining chapters in this book. The case study describes a person with presenting disabilities. After the case study, there is a series of questions. Answers to the questions are provided in this chapter to illustrate how to analyze the case study.

### Case Study

Mr. Samuel Williams is 60 years of age and is currently married with three grown children. He has a Bachelor of Arts Degree in fine arts, completed a real estate course, and holds a current real estate license. For the past seven years, Mr. Williams has been active and successful in residential real estate sales. Previous to this, he was both a salesperson and an assistant manager in a men's specialty clothing store. The sales position lasted for five years, at which time Samuel was promoted to assistant manager.

This client has hypertension (high blood pressure) and a heart condition. Six months ago, he suffered a myocardial infarction (heart attack). The treating physician reported that Mr. Williams has coronary artery disease and has restricted him to a maximum of light work not involving excessive emotional stress. It also was recommended he consider sedentary work, as it would be less physically demanding. No surgery is contemplated at this time.

The position of real estate sales agent, *Dictionary of Occupational Titles (D. O. T.) #250.357-018* (U. S. Department of Labor, 1991), involves renting, buying, and selling real estate property for clients. An agent is paid on a commission basis and does not earn a salary. Real estate sales agents are familiar with all state and local regulations relating to the purchase and sale of property. They review trade journals and other publications to keep current in the field and to be informed about marketing conditions and property values. A real estate sales agent holds a current license issued by the state. Agents interview prospective clients to solicit listings. They accompany clients to property sites, show properties, quote purchase prices, and describe features and conditions of sale or terms of lease. Agents draw up real estate contracts, such as deeds, leases, and mortgages, and negotiate loans on properties. Real estate agents typically are paid on a commission-only basis.

In addition to these functions, Mr. Williams served as an office manager, supervising clerical personnel in the real estate office. This was on a part-time basis for which he was paid a salary.

### Questions

1. Describe the client's vocational profile, including age, educational level, work history (skill and exertional levels), occupationally significant characteristics, and job skills.
2. How do the occupationally significant characteristics of the job impact Mr. Williams' disability?
3. What, if any, reasonable accommodations can be made for the client to return to his usual and customary occupation as a real estate sales agent?
4. What is this client's rehabilitation potential? Your rehabilitation supervisor is of the opinion this client may be too old for the provision of rehabilitation services. Comment on this issue.
5. What jobs can Mr. Williams perform using transferable skills?

### Answers

The vocational profile for this client is as follows:

Age:	60 years of age - Close to Retirement Age.
Education:	Bachelor of Arts degree and real estate license training course - High School Education and Above.
Work History:	2000-2008: Real Estate Sales Office Manager (part-time) and Agent – Skilled. Light exertion. 1993-2000: Real Estate Sales Agent – Skilled. Light exertion. 1983-1993: Retail Sales and Assistant Manager (men's clothing store) – Semiskilled. Light exertion.

**Occupationally Significant Characteristics:** Manual dexterity, eye-hand-foot coordination, attention to detail, visual perception and acuity, capacity to work with others, inside as well as outside work, varying work tasks, clerical functions, light exertion with frequent standing and walking, and activities involving emotional stress.

**Transferable Skills:** An ability to rent, buy, and sell property for clients on a commission basis; knowledge of property listings and ability to study real estate listings; capability to review trade journals to keep current on market conditions and property values; skills in interviewing clients; an ability to show property; capability to draw up real estate contracts; ability to negotiate; current real estate license issued by the state; ability to calculate costs, taxes, discounts, and other charges; arithmetic ability as applied to retail sales; capability to work with financing; capacity to present property in a positive light; ability to persuade, convince, and finalize sales; facility with words to clearly describe advantages of a particular product; capacity to use business diplomacy and tact when dealing with people; communication skills and organizational ability.

The work of a real estate agent and salesperson involve physical exertion at the light level. This is within Mr. Williams' physical limitations although the physician recommends that he consider sedentary work, as it would be less physically demanding. Work of a real estate agent is emotionally stressful, although the work of an office manager (real estate) or salesperson (not in real estate) are less emotionally demanding.

Mr. Williams can be assigned more office work, involving less standing and walking and decreased emotional stress. Working in the office and assuming full-time office management duties, the employer may be willing to pay him a higher salary than when he was a part-time office manager. Salaried work is less stressful than commission-only work. This client's perception of emotional stress as it relates to prior work activities needs to be explored by the counselor.

This client has good rehabilitation potential. He has a medical condition that is apparently under control with clearly specified functional limitations. There is at least a 25-year consistent work history at the skilled and semiskilled levels. During this time, he developed a variety of transferable skills. Age, in and of itself, cannot be used to deny the provision of rehabilitation services.

The following jobs use this client's transferable skills for light and sedentary jobs that are considered to involve low emotional stress.

- Light:**
- Real estate firm manager (real estate)
  - Title searcher (real estate)
  - Real estate appraiser (real estate)
  - Leasing or rental agent (real estate)
  - Public events facilities rental manager (business service)
  - Property manager (real estate)
  - Apartment house manager (real estate)
  - Condominium manager (real estate)
  - Salesperson (furniture, appliances, and other areas)
  - Insurance sales agent (insurance; real estate)
  - Counter clerk (clerical)
  - Sales representative (retail trade; wholesale trade)
  - File clerk (clerical; real estate)
  - Office clerk (clerical; real estate)
- Sedentary:**
- Real estate clerk (clerical)
  - Real estate assistant (real estate)
  - Risk and insurance manager (insurance)



Mortgage-closing clerk (clerical; real estate)  
Housing project manager (real estate)  
Receptionist (real estate)  
Credit-reference clerk (clerical)  
Credit clerk (clerical)  
Cashier (clerical)  
Credit-card clerk (retail trade)  
Check cashier (business service)  
Information clerk/assistant (real estate)

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# Chapter 2

## HUMAN BODY SYSTEMS

*Frances W. Siu, Ph.D.*  
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### Introduction

The human body is a complex network of some 63 trillion cells (McDowell & Windelspecht, 2004; Tortora & Derrickson, 2008). These cells combine to form tissues which in turn merge as organs. While each organ in the body performs a specific function those organs that perform related functions develop into body systems (Seeley, Stephens, & Tate, 2007). Together, these varying systems work to meet the daily needs of the human body. The physiological and psychological functions of body systems are known collectively as body functions. Body structures, accordingly, indicate the anatomical components of the organs and limbs. Knowledge of the structure and function of the human body builds the foundation for understanding illnesses and disabilities (Martini & Nath, 2008). Preceded by an introduction to the chemical and cellular levels of the human body, this chapter provides readers with an overview of the skeletal system, muscular system, nervous system, cardiovascular and lymphatic system, integumentary system, respiratory system, sensory system, digestive system, genitourinary system, and endocrine system. The visual and hearing systems are described in chapters in this book specific to those subjects.

### Chemical and Cellular Levels

In the human body, building blocks of matter called atoms interact and combine to form molecules such as sugar, fat, water, and protein. These molecules in turn form organelles, such as the plasma membrane and nucleus, which make up cells (Seeley et al., 2007). Cells are the basic units of the human body; they carry out the functions of protection and support while providing the body systems with a means of communication. Cells additionally metabolize and release energy, provide the basic materials for genetic inheritance, and make movement possible. Each cell is encased by an outer layer known as the plasma membrane which regulates what enters or leaves its boundaries. The hereditary material of the cell, Deoxyribonucleic Acid (DNA), is found inside the nucleus. DNA helps direct cell activities by utilizing Ribonucleic Acid (RNA) (Martini & Nath, 2008).

The cellular level of the human body, described above, is distinguished from the chemical levels. These levels rely on the basic components of oxygen, carbon dioxide, and water. Oxygen is necessary in the chemical reactions that extract energy from food (Seeley et al., 2007). Carbon dioxide and energy are then released during metabolism when the organic molecules from the energy source are broken down. Water is essential to the human body. Like oxygen and carbon dioxide, water not only helps stabilize body temperature but protects the body against friction and trauma, and directly participates in chemical reactions such as dehydration and hydrolysis (Martini & Nath, 2008). The human body relies on water; the body of a new born child has 90% water content, while that of an adult maintains water content of 70% to 85%.

### Tissue Level

Tissues are collections of similar cells and the substances surrounding them (Martini & Nath, 2008). There are four primary types of tissue:

- Epithelium forms the internal and external linings of various organs of the body. It is usually involved in activities such as diffusion, filtration, secretion, and absorption.

- Connective tissue connects and binds cells and other tissues together. Among the connective tissues, there are three types of specialized connective tissue (Seeley et al., 2007):
  - Cartilage is made up of collagen, proteoglycans, and water; its slightly elastic, gummy-like consistency allows body parts to spring back when compressed. The external parts of the ears and connecting tissues between the bones of the spine are examples;
  - Bone is a hard, calcified connective tissue composed of 35% organic substance, 45% inorganic substance, and 20% water that forms the internal framework of the body;
  - Blood contains cell fragments called platelets which play a key role in blood clotting. The cells and platelets are suspended in plasma. There are two types of blood cells. The *red blood cells* transport oxygen through the arteries and waste through the veins while the *white blood cells* fight infections and cancer.
- Nervous tissue, which responds to stimuli and transmits impulses throughout the body, is composed of neurons (conductive cells) and neuroglia (support cells) (Martini & Nath, 2008). Neurons have cell processes called dendrites that receive electric impulses, and axons that conduct them.
- Muscle tissue provides for bodily movement. They are grouped according to the presence or absence of striations (microscopic bands) in the muscle cells (Adams, 2004). Types of muscle tissue include: skeletal, cardiac, and smooth muscle. Skeletal muscle is classified as striated voluntary (usually consciously controlled) while cardiac muscle is striated involuntary (not normally consciously controlled). Smooth muscle is characterized by nonstriated involuntary tissue (Seeley et al., 2007).

## Organ Level

A body system is often defined as a group of organs that physiologically operate together to carry out bodily function (Seeley et al., 2007). Just as organs often act together to complete an internal task, some organs carry out a variety of functions. An example of such duality includes the hypothalamus which interacts with both the nervous system and endocrine system. The kidney performs filtering roles while producing hormones simultaneously.

The body systems that perform specific purposes in our daily lives also play a significant role in the process of homeostasis (Seeley et al., 2007). Homeostasis is a state of equilibrium (internal balance and constancy). When the body maintains homeostasis, good health and well-being follow. Conversely, when homeostasis is not maintained, sickness and even death can result. Environmental factors that negatively affect the body systems can upset the body's internal constancy and state of homeostasis (Martini & Nath, 2008).

## System Level

### *Musculoskeletal System*

As the name implies, the musculoskeletal system consists of the muscular system and the skeletal system.

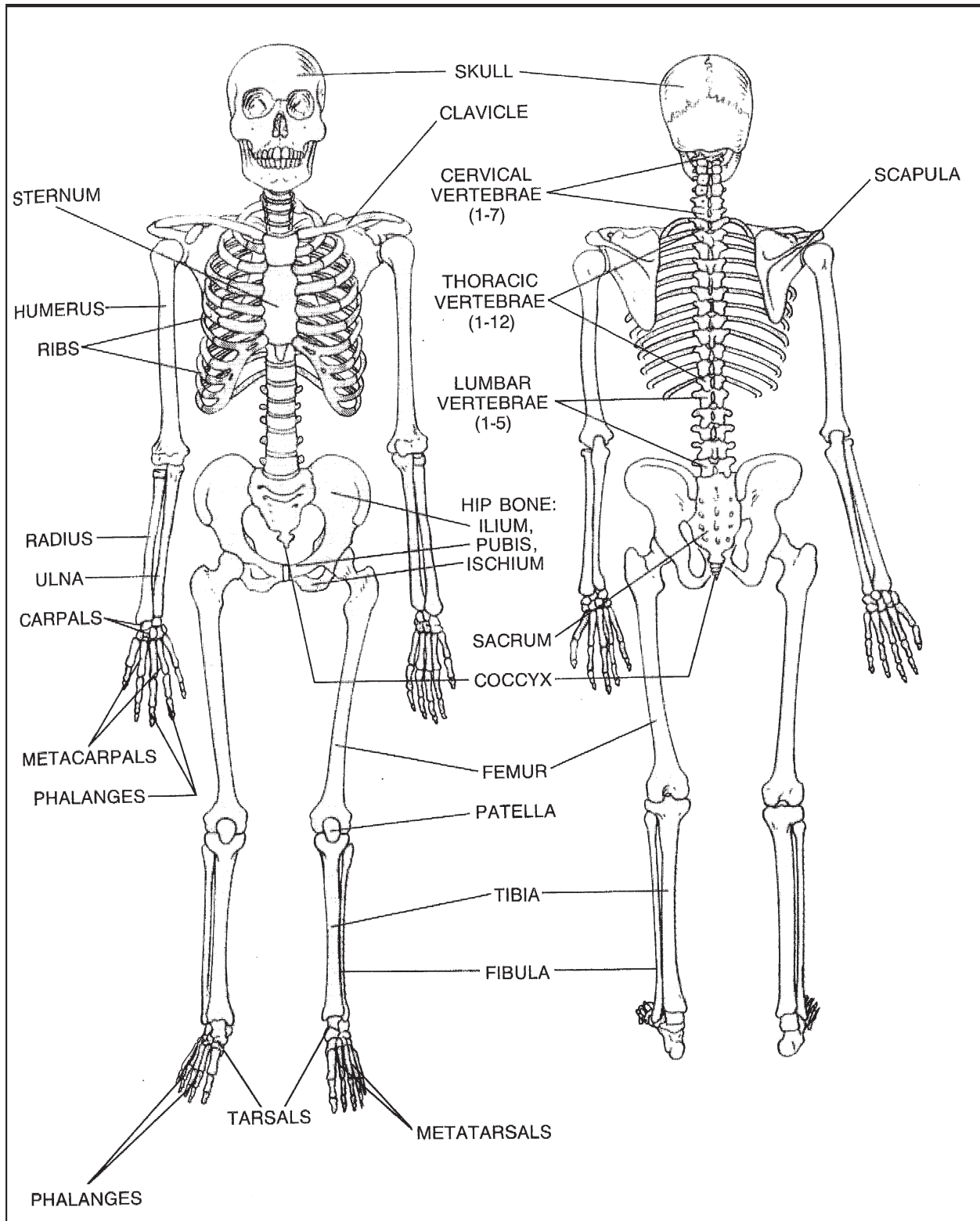
#### *Skeletal Structure*

The human skeleton consists of 206 bones which provide the body with its internal structural framework and functions to enable the maintenance of an upright posture and protect vital internal organs (Seeley et al., 2007). For purposes of classification, bones are grouped into the axial and appendicular skeletons. The axial skeleton, formed by the bones of the head and trunk, is distinct from the appendicular skeleton which includes the bones of the upper and lower limbs and the shoulder and hip bones.

#### *Skeletal Function*

The structure of the skeletal system described above allows it to carry out functions vital to the human body. These actions include movement, storage, and maintenance of chemical levels. The bones allow for movement in that they are a place for the skeletal muscles to attach. In this way, the bones act as

**Figure 1**  
**Skeletal System**



Source: F. A. Davis. (2009). *Taber's Cyclopedic Medical Dictionary* (21st ed.). Philadelphia, PA: F. A. Davis. Reprinted by permission of F. A. Davis.



levers to move body parts. Bones also store needed minerals; the most important of these include calcium and phosphate. By absorbing, storing, and releasing calcium as needed, bones help maintain a normal blood level (Martini & Nath, 2008). Additionally, bones store fats for cellular energy production.

Contrary to common classification, bones are living organs. Inside the bones, an entire living environment thrives. The outer shell of dense material encasing the marrow and its cells is the compact bone. The inner mass, known as spongy bone, is less dense than compact bone and has numerous small cavities. Within the bone marrow, red blood cells, white blood cells, and platelets exist. Connective tissues outside the compact bone attach skeletal muscles to the bone and house bone-forming cells which repair bone tissues.

Two sets of bones comprise the skull: cranial bones and facial bones. Cranial bones protect and enclose the brain and provide the site at which the head muscles attach. Facial bones provide the framework for the face and mouth. All bones comprising the skull are attached to each other via immovable joints except for the mandible, which is attached to the skull via a freely movable joint. Eight cranial bones make up the cranium. The cranial bones can be compared to an eggshell – they are thin and self-bracing. The forehead is formed by the frontal bone which also forms the upper half of the eye sockets. Two parietal bones make-up the bulk of the cranium and comprise the sides of the skull. Temporal bones lie inferior to the parietal bones on both sides of the skull. The occipital bone, also named for the part of the brain it covers, shapes the posterior of the skull. The sphenoid bone encloses the cranial cavity lying behind the eye sockets anterior to the temporal bones and posterior to the frontal bone (Tortora & Derrickson, 2008).

The spinal column (formed by 26 bones) connects the face and skull to the rest of the body. At the base of the flexible, curved spinal column is the sacrum, comprised of five fused vertebrae. Below the sacrum, the coccyx is composed of four fused vertebrae. The entire spinal column is divided into three parts. Cervical vertebrae encompass the first seven vertebrae below the skull while thoracic vertebrae refer to the next 12, and lumbar to the last five vertebrae (Tortora & Derrickson, 2008).

Anterior to the spinal column is the sternum and rib cage. While 12 pairs of ribs exist, only the first seven are attached directly to the sternum. The rest are known as “false ribs,” meaning that they attach to the sternum indirectly or not at all. The seven attached ribs are connected to the sternum by cartilage. At the inferior end of the sternum, the xiphoid serves as an attachment point for the diaphragm.

On each side of the skeleton are the scapulae, posterior to the rib cage, connecting the upper arm to the rest of the skeleton by a bone called the clavicle which, together with the scapulae comprise the shoulder girdles. The clavicle mainly acts as a brace holding the scapulae and the arms up and away from the ribs. It also enables us to raise our hands by allowing the scapula to slip forward and back (Tortora & Derrickson, 2008).

The humerus is the only bone in the upper arm. At the end of the humerus are two bones: the radius and ulna. These long bones allow twisting the hands from side to side. At the end of the radius and ulna lie the carpals, collectively referred to as the wrist. While most people consider their wrist to be located between the hand and lower arm, the true wrist, the carpus, exists at the base of the hand. The palm of the hand is supported by the metacarpals, while the three phalanges make up the fingers.

The rest of the body could not be connected to the lower extremities without the existence of one structure: the pelvis. The pelvis works to provide support for the body’s internal organs. Its flaring sides are called the ilium. The two ring-shaped portions located inferior to the hip sockets are called ischium while the femur refers to the bone in the thigh. The patella (kneecap) guards the knee joint against blows and improves leverage of thigh muscles which raise the lower leg. The tibia and fibula accordingly, form the lower leg. These tibiofibular joints, unlike the joints of the radius and ulna, do not allow for much movement. Of these bones, only the tibia bears weight – the fibula serves only as an attachment place for muscles.

The foot is much like the hand in the way the bones are grouped. The first group of bones, the tarsals, form half the foot. The calcaneus forms the heel of the foot, while the talus connects the foot with the tibia and fibula. The rest of the foot, excluding the toes, is composed of metatarsals. The toes, like the fingers, have three phalanges (Tortora & Derrickson, 2008).



The three types of muscles include skeletal, smooth, and cardiac (Adams, 2004).

- Skeletal muscle – Roughly 630 skeletal muscles are found in the body accounting for approximately 50% of male body weight and 40% of female body weight. Skeletal muscle tissue is the strongest and hardest working in the body and is also its longest form of muscle. Skeletal muscle tissue is known as striated muscle tissue because muscle tissue, which crosses over each fiber, causes an appearance of crisscrossing. Though fragile, skeletal muscle tissue is very strong because it is protected by a sheath. Interestingly, skeletal muscle tissue is also the only type of muscle tissue that is voluntary.
- Smooth muscle – Smooth muscle is involuntary and is commonly found in hollow organs such as the stomach, bladder, and respiratory passages. Its main function is to propel objects. These tissue are spindle-shaped and arranged in sheets or layers. Typically, two layers run together, alternately contracting to change the size of the organ.
- Cardiac muscle – Cardiac muscle tissue is found only in the heart; it is striated, involuntary, and responsible for pumping blood throughout the heart. Cardiac muscle tissue is protected by connective tissue.

### ***Joints, Tendons, and Ligaments***

Besides muscles and bones, joints play a pivotal role in body movement. These connecting structures of bones are classified either according to degree of movement or type of connective tissue that joins them. Fibrous joints are those ends of bones connected by fibrous tissue without joint cavity; they are capable of little or minimum movement. Cartilaginous joints are attached by hyaline cartilage and are slightly moveable. Synovial joints have joint cavities where the fibrous connective tissue hold the bones together. These joints are highly moveable, depending on their location. Synovial joints allow for various types of common movement such as gliding, angulations, rotation, and the combination of two or more movements. Elevation and depression, protraction and retraction, excursion, opposition and reposition, and inversion and eversion are some special movements also made possible by synovial joints. The term *range of movement* usually describes the amount and nature (active or passive) of movement occurring at the joint (Tortora & Derrickson, 2008).

Both tendons and ligaments are soft collagenous tissues. While tendons connect muscles to bone and carry the tension forces between them, ligaments connect bone to bone. Both tendons and ligaments play a significant role in musculoskeletal biomechanics.

As the foundation of the body, the skeleton functions alongside the muscular system, which provide the body with range of movement. The nervous system, integral to both the skeletal and muscular systems, provides the means by which all body systems function.

## ***The Nervous System***

The nervous system is the master controlling and communicating system of the body; it directs human thoughts, actions, and emotions. It is the most rapid-acting and complex system of the body, functioning via intra-cellular communication through electrical signals. The nervous system is but one single system, but typically it is discussed in two parts: the central nervous system (CNS) and the peripheral nervous system (PNS).

### ***Central Nervous System***

The CNS consists of the brain and spinal cord. As the command center of the nervous system, the CNS interprets incoming signals and responds to these signals based on past experiences, reflexes, and current conditions.

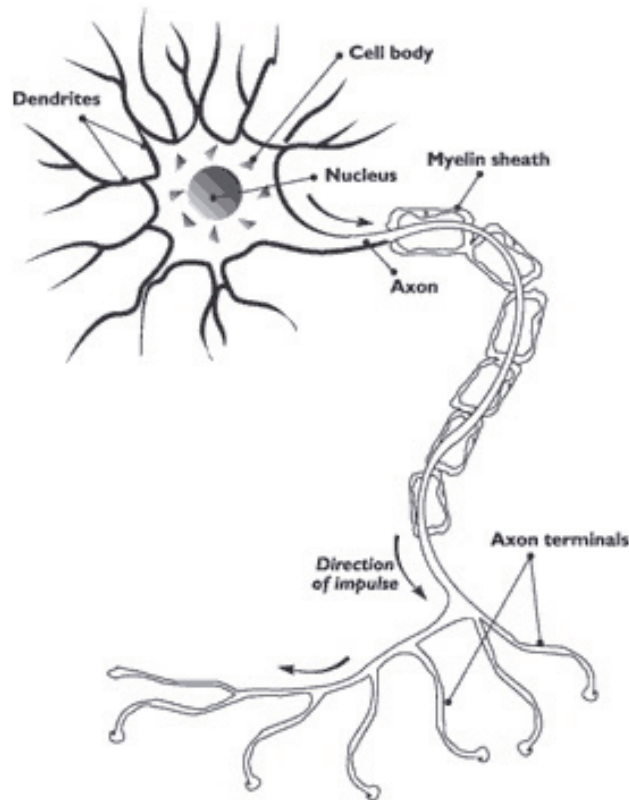
### ***Peripheral Nervous System***

The PNS is distinguished from the CNS, consisting of the nerves extending from the brain and spinal cord. These nerves, respectively called cranial nerves and spinal nerves serve as the communications link from the body to the CNS. The PNS is further divided into the following functional subdivisions:

- Sensory contains nerve fibers that carry impulses to the CNS from sensory receptors located throughout the body. There are two types of sensory fibers.



**Figure 3**  
**Neuron Diagram**



Source: F. A. Davis. (2009). *Taber's Cyclopedic Medical Dictionary* (21st ed.). Philadelphia, PA: F. A. Davis. Reprinted by permission of F. A. Davis.

- Somatic afferents convey information from the skin, skeletal muscles, and joints.
- Visceral afferents convey impulses from the visceral organs.
- Motor transports messages from the CNS to organs, muscles, and glands. The motor system is divided into two parts (Tortora & Derrickson, 2008).
  - *Somatic nervous system*, composed of motor nerve fibers that connect the CNS to the skeletal muscles, is often referred to as the *voluntary nervous system*. These motor neurons respond mainly to external stimuli. Reflexes, automatic reactions to a stimulus, are usually controlled by the spinal cord and brain stem.
  - *Autonomic nervous system*, consisting of nerve fibers that regulate the activity of smooth muscles, cardiac muscles, and glands, is not consciously controlled; therefore, it is generally referred to as the *involuntary nervous system*. The autonomic nervous system is subdivided into the sympathetic and parasympathetic nervous systems. These systems have opposing effects when they act upon the same organ. While the parasympathetic nervous system enhances activities and conserves energy, the sympathetic nervous system increases energy expenditures and prepares the body for action.

In the nervous system, there are two main types of cells – neurons and supporting cells. While neurons provide a pathway for messages traveling to and from the brain and spinal cord, supporting cells provide structural reinforcement, protection, insulation, and general assistance for neurons. As specialized cells, neurons provide for the transportation of signals. Although many different types of neurons exist, all

neurons share common characteristics. For example, all neurons share large cell bodies and have fiber-like extensions called processes. Processes are further divided into dendrites and axons.

Dendrites convey signals toward the cell body. These short, numerous cells branch extensively around the nucleus. Perpendicular to the body of the neuron, the axon conducts signals away from the cell body. Along the length of the axon, the Schwann cells form an insulating layer called the myelin sheath. Branches of axons terminate in hundreds or thousands of telodendria branlets which, in turn, end in synaptic knobs. Synaptic knobs relay messages to other cells via neurotransmitters. Gaps in the synaptic knobs are called synapses.

The brain and body are linked by the spinal cord, a long bundle of white nerve matter residing in the middle of the spinal column. The spinal cord also serves as the center from which reflexes stem.

The brain has three divisions: brain stem, cerebellum, and cerebrum. The brain stem is the core for most life systems commands - vital functions which are not consciously controlled. The cerebellum, the next brain division, controls balance and muscle coordination. After the cerebellum, the most advanced part of the brain is the cerebral cortex – conscious thought takes place in this region. While the cerebellum can be divided into the left and right hemispheres, it can also be divided into functional parts. The right hemisphere, generally, is the artistic side of the brain. People who are “right brained” (the right hemisphere is dominate over the left) are thought to excel at activities involving abstract thinking such as art and music. “Left brained” people are considered to be logical, allowing them to excel in math, physics, and other such logical thought processes. Although one sphere can be dominant, the two hemispheres are connected by the corpus callosum which allows the two hemispheres to communicate (Fix, 2008).

### ***The Cardiovascular System (including the Lymphatic System)***

#### ***The Cardiovascular System***

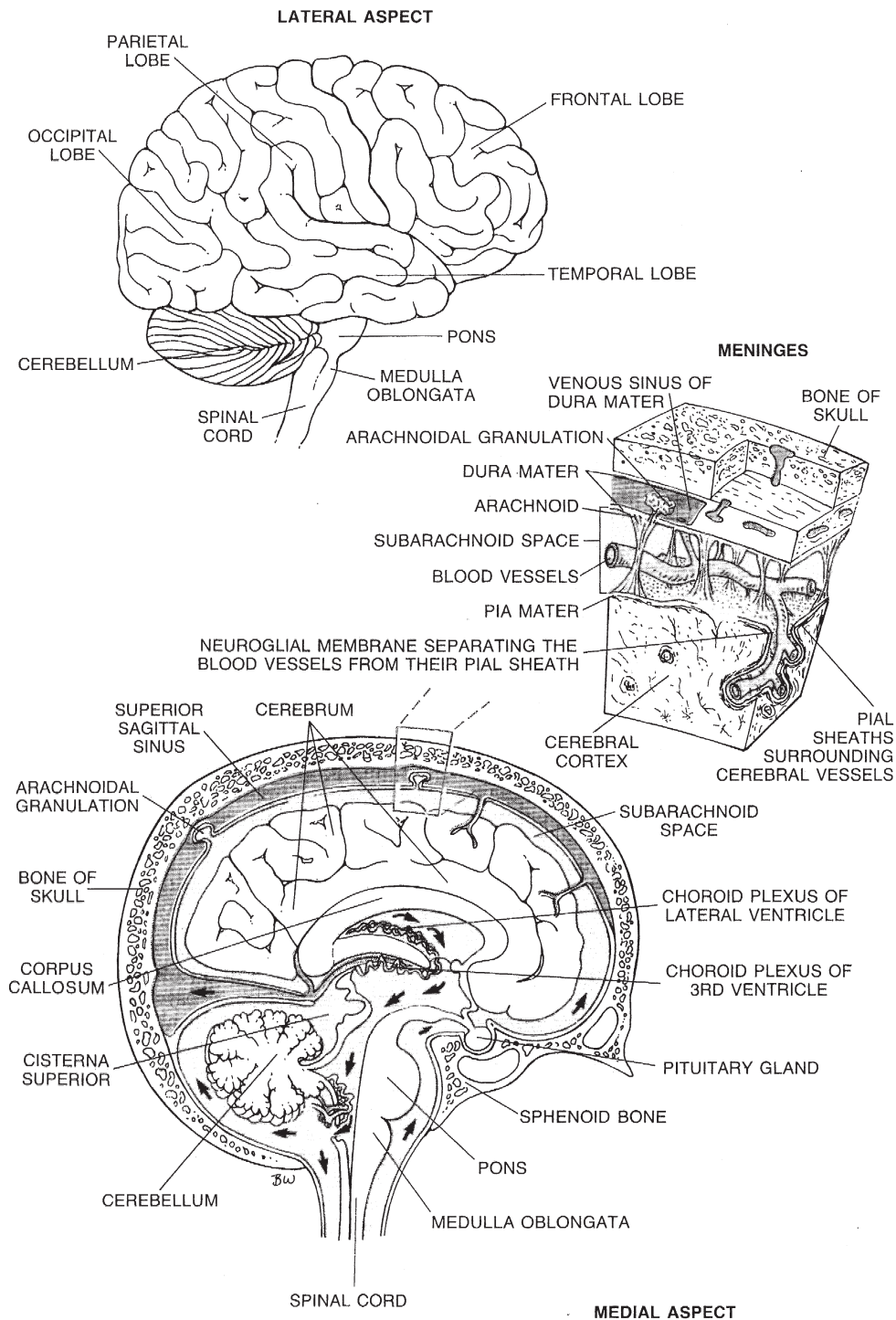
The cardiovascular system controls a singular vital function – blood circulation. The heart, the cardiovascular system’s central organ, is made up of cardiac tissue. It first pumps blood to the lungs to obtain oxygen, and then circulates the blood throughout the body through a series of arteries and veins. While the arteries carry oxygenated blood from the heart to the body, veins return the oxygen-depleted blood to the heart.

The heart, the size of a human fist, is centrally located in the upper body cavity and can be divided into four chambers: left and right atria and left and right ventricle. The atria consist of the upper half of the heart, while the ventricles compose the lower portion. The dividing wall between the left and right sides is the septum. Four valves of the heart control circulation (Tortora & Derrickson, 2008).

Circulation is necessary for the growth and development of cells. The blood is composed of red and white blood cells, platelets, lymph, plasma, and water. Forty-five percent of blood consists of platelets, red blood cells, and white blood cells. Of the white blood cells, *neutrophils* and *lymphocytes* are the most important. Fifty-five percent consists of plasma, the liquid part of blood. Red blood cells contain hemoglobin, the chemical compound that carries oxygen, while white blood cells are part of the immune system. Platelets emit the blot clotting substance, fibrinogen, into its active form, fibrin; lymph is the interstitial fluid in the blood. Plasma, the remaining portion of the blood, is the mixture of glucose and water in which the blood cells are suspended.

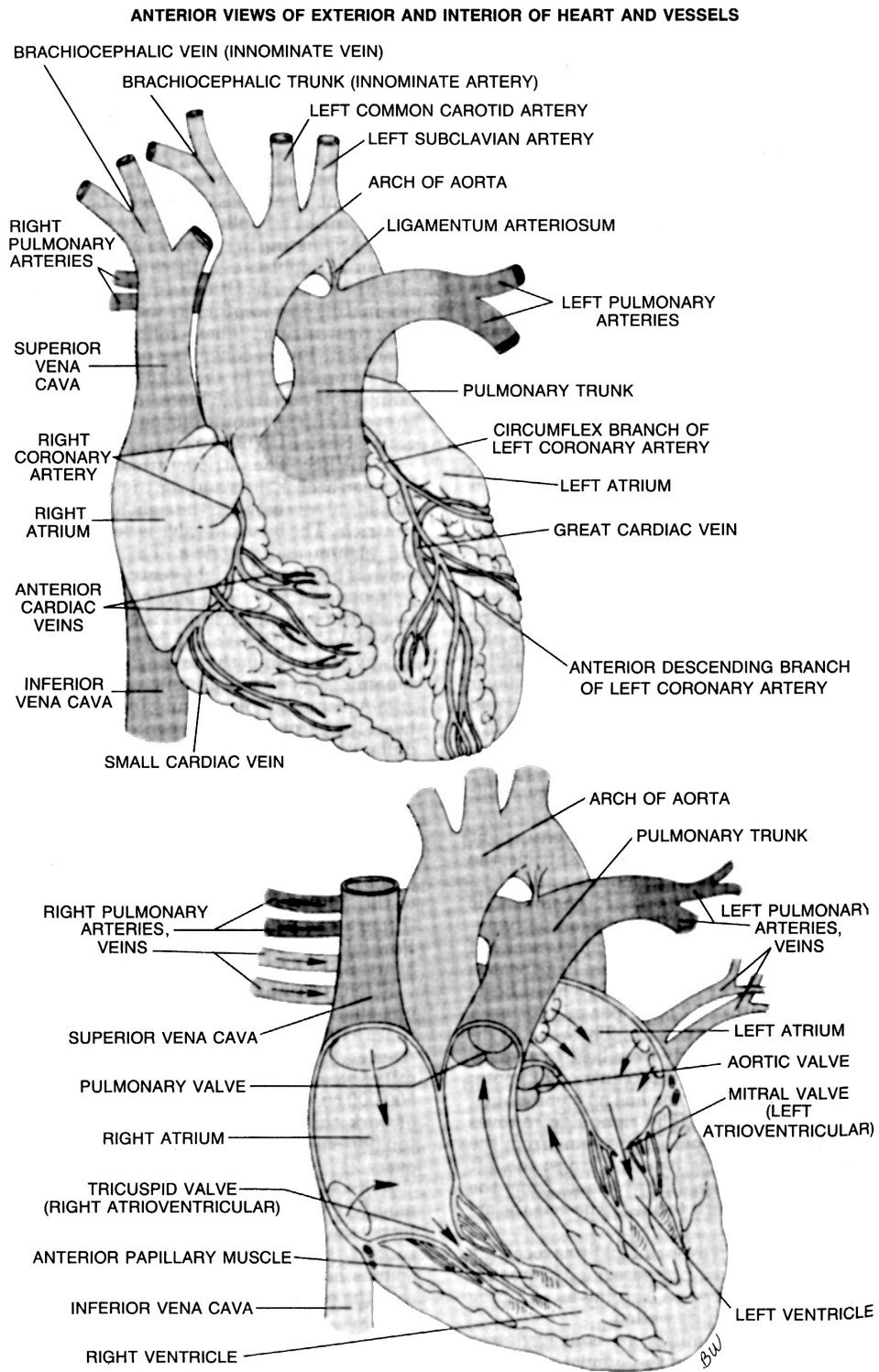
Blood enters the heart through the left atrium from the superior and inferior vena cava. The superior vena cava, the vein responsible for collecting the blood returning from the upper body differs from the inferior vena cava, which returns blood from the lower body. During the systolic phase of the heartbeat, the deoxygenated blood from the superior and inferior vena cava veins enters the heart through the right atrium. In this phase of the heartbeat, the atria begin to fill while the ventricles contract to pump the blood. Additionally, the pulmonary and aortic valves open allowing blood to exit the ventricles. During the diastole phase, the atria then contract, pumping blood into the ventricles. To allow blood to leave the atria and enter the ventricles, the tricuspid and mitral valves of the heart open.

**Figure 4**  
**The Brain**



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**Figure 5**  
**The Heart**



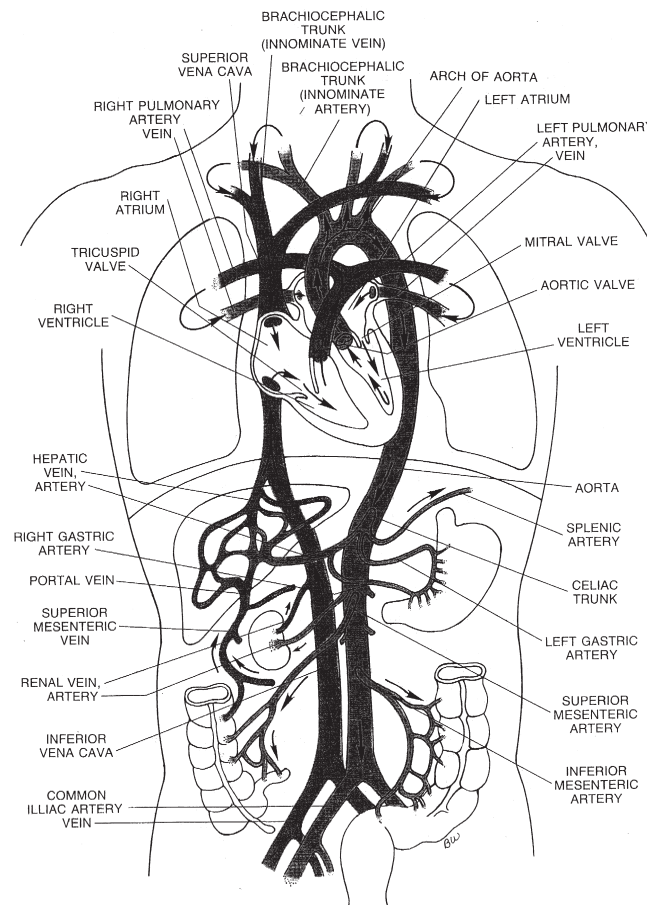
Source: F. A. Davis. (2009). *Taber's Cyclopedic Medical Dictionary* (21st ed.). Philadelphia, PA: F. A. Davis. Reprinted by permission of F. A. Davis.

New blood entering the right atrium is pumped by the tricuspid valve to the right ventricle. Once the pulmonary valve opens to the pulmonary artery, blood is carried to the lungs where it is oxygenated. The pulmonary artery is the only artery in the body that carries deoxygenated blood. When blood reaches the lungs, carbon dioxide (cellular waste) is diffused into the lungs. When blood is oxygenated within the lung tissue, the circulatory system and the respiratory system act.

The oxygen in the lungs, first diffused through the alveoli sacs, is next pushed through the wall of the lungs and into the bloodstream where the blood distributes oxygen throughout the body. To enable direct oxygenation of the cells, the arteries are branched into smaller arterioles. These separate further into capillaries, the smallest blood vessels whose walls are thin and elastic. When oxygen is released from the hemoglobin, it is diffused across the capillary wall, traveling to a nearby cell and entering through its membrane.

The carbon dioxide that returns to the blood travels back to the lungs via the capillaries. The capillaries fork into smaller venules, which divide further into veins. Carbon dioxide and blood are carried back to the heart by the veins, completing the circulatory cycle.

**Figure 6**  
**Human Circulatory System**



Source: F. A. Davis. (2009). *Taber's Cyclopedic Medical Dictionary* (21st ed.). Philadelphia, PA: F. A. Davis. Reprinted by permission of F. A. Davis.



### ***Lymphatic System***

A specialized component of the circulatory system is the lymphatic system, consisting of moving fluid (lymph/interstitial fluid); vessels (lymphatics); lymph nodes, and organs (adenoid, tonsils, thymus, thoracic duct, small intestine, appendix, liver, and spleen). By moving blood in and out of arteries and into veins and through the lymph nodes and into the lymph, the body is able to eliminate the products of cellular breakdown and bacterial invasion (McDowell & Windelspecht, 2004). The lymphatic system is a powerful network of small ducts, minor glands, specialized cells, and organs that are dispersed throughout the body. The lymphatic system functions to 1) absorb excess fluid, thus preventing tissues from swelling; 2) defend the body against microorganisms and harmful foreign particles; and 3) facilitate the absorption of fat. The lymph system is made up of T cells, B cells, antibodies, and platelets.

The skin plays a major role in preventing disease by protecting organs, blood vessels, and the lymph system. To combat bacteria, viruses, and disease, the skin contains lymph nodes. These nodes transport white blood cells (phagocytes) throughout the body via a network of vessels.

Mucous membranes, cells that line all openings of the body, provide a second line of defense against foreign bacteria. When dust particles enter the body, for example, they are trapped in the mucous membranes and then digested. Nose hair functions alongside the mucous membranes to protect the body; they act as an air filtration system.

Along with the lymph nodes, the lymphatic system includes lymph (interstitial fluid). Capillaries release excess water and plasma into intracellular spaces where they mix with *lymph*, a fluid containing proteins, fats, and a type of white blood cells called “lymphocytes,” the body’s first-line defense in the immune system. Lymph flows from small lymph capillaries into lymph vessels that are similar to veins in having valves that prevent backflow. The contraction of skeletal muscle propels lymph fluid through the valves. Lymph vessels connect to lymph nodes, lymph organs (bone marrow, liver, spleen, thymus), and the cardiovascular system.

Modern scientists believe that the immune system, throughout a lifetime, is capable of producing between one million to one billion different types of antibodies. A few hundred genes in our DNA are responsible for generating this diverse assortment of antibodies. DNA, a nucleic acid in the form of a double helix, contains genetic instructions or codes that control both the biological development of all cellular life as well as the production of many viruses (Seeley et al., 2007). Besides functioning as a line of defense for the human body, the lymphatic system functions as a separate circulatory system, working parallel to the cardiovascular system. The lymphatic system absorbs fats, returns approximately three liters of fluid from our body tissues to the circulatory system on a daily basis, and transports selected nutrients from the digestive system to the circulatory system. The next line of body defense includes the integumentary system.

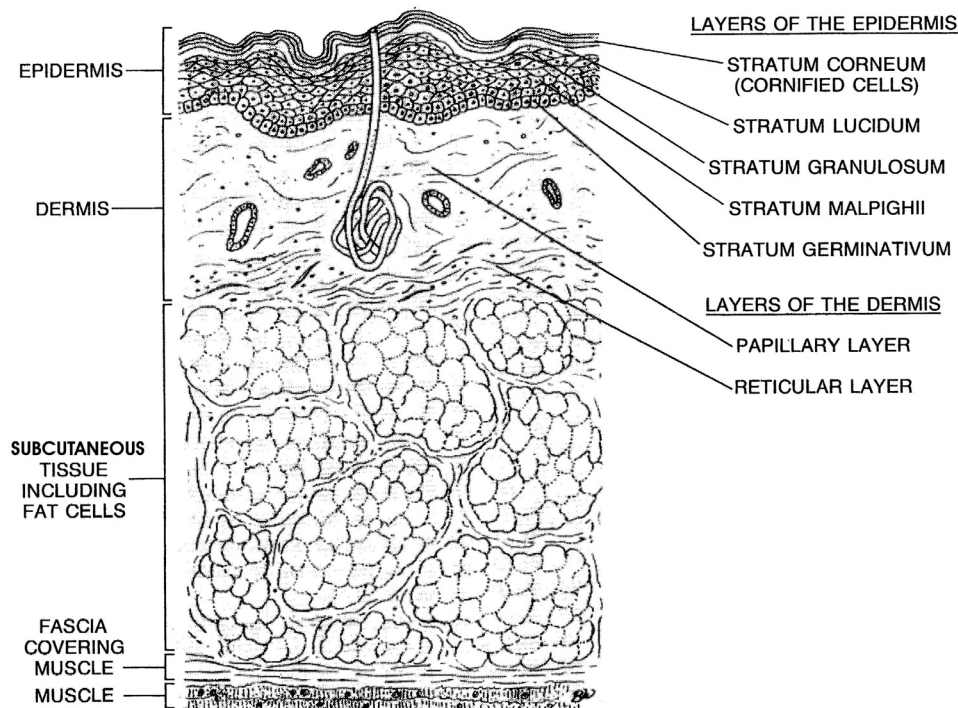
### ***The Integumentary System***

The integumentary system, commonly called the skin, encases the body. The skin is an ever-changing organ that contains many specialized cells and structures. It functions as a protective barrier against the environment and is involved in maintaining proper body temperature. The skin gathers sensory information from the environment and plays an active role in protecting the body from disease. The skin’s functioning is divided into three parts, representing three layers of skin - the epidermis, dermis, and subcutaneous tissue (Martini & Nath, 2008).

The epidermis, the outer layer of skin, varies in thickness. It is the thinnest on the eyelids and thickest on the palms of the hands and soles of the feet. The cells of the bottom layer are shaped like columns. In this layer, the cells divide and push other cells into higher layers. As the cells move into the higher layers, they flatten and eventually die. The top layer of the epidermis, the stratum corneum, is made of dead, flat skin cells that shed about every two weeks.

Dermis which connects the skin to the tissue underneath includes collagen, elastic tissue, and reticular fibers. It contains many specialized cells and structures such as hair follicles, sebaceous (oil) glands, and apocrine (scent) glands. This layer also contains eccrine (sweat) glands, unassociated with hair follicles. In addition, blood vessels and nerves course through this layer, transmitting sensations of pain,

**Figure 7**  
**Human Skin Diagram**



Source: F. A. Davis. (2009). *Taber's Cyclopedic Medical Dictionary* (21st ed.). Philadelphia, PA: F. A. Davis. Reprinted by permission of F. A. Davis.

itching, and temperature. Specialized nerve cells, in the dermis transmit sensations of touch and pressure (Martini & Nath, 2008).

Subcutaneous tissue is a layer of fat and connective tissue that houses larger blood vessels and nerves. This layer is vital in regulation of skin and body temperature; its size varies throughout the body and from person to person.

### ***The Respiratory System***

The respiratory system supplies the body with oxygen by circulating air throughout the body systems. After air is inhaled through the nose or mouth, it travels into the pharynx, passes through the larynx, and makes its way down the trachea. When air reaches the lungs, it is diffused into the blood via the alveoli (Martini & Nath, 2008).

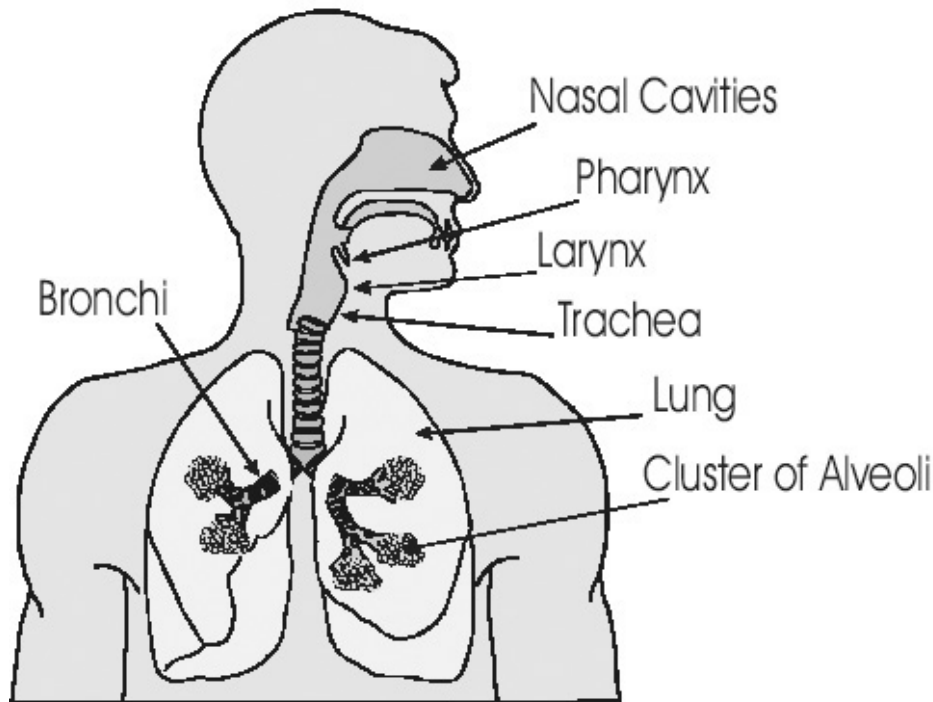
#### ***Nasal Cavity***

As we breathe, air enters the body through the nostrils. Inside the nose, the nasal cavity is divided by the septum and lined by the respiratory mucosa. In the roof of the nasal cavity, the mucosa house the olfactory receptors. This mucosa helps warm and moisten air entering the body while trapping invading pathogens. Three mucous covered membranes project into the nasal cavity. These projections help increase the area affected by the mucosa.

#### ***Sinuses***

The nasal cavity is surrounded by the paranasal sinuses located in the maxillary, ethmoid, sphenoid, and frontal bones. The sinus chambers lighten the weight of the skull, act as resonance chambers for vocalization, and secrete mucous for the nasal cavity.

**Figure 8**  
**Respiratory System**



Source: F. A. Davis. (2009). *Taber's Cyclopedic Medical Dictionary* (21st ed.). Philadelphia, PA: F. A. Davis. Reprinted by permission of F. A. Davis.

### ***Pharynx***

The pharynx (throat) is the muscular passageway which provides a means of transportation for air and food. Air enters the pharynx through its superior portion, the nasopharynx, and descends to the oropharynx and laryngopharynx. From the laryngopharynx, air moves into the larynx. Food mirrors the passage of air from the nasopharynx to the laryngopharynx. Instead of entering the larynx, as air does, however, food enters the esophagus. Since the auditory tubes from the ear open into the nasopharynx, the mucosa of each region are continuous.

### ***Larynx***

The larynx (voice box) routes food and air to its proper destination. This structure consists of eight hyaline cartilages and a flap of elastic cartilage, the epiglottis. It is the task of the epiglottis to prevent food from entering the superior opening of the larynx and traveling down the trachea. The epiglottis is opened by breathing, which allows air to pass freely to the lungs. Swallowing, conversely, causes the epiglottis to close. When the epiglottis is closed, food is forced down the esophagus. The mucous membranes of the larynx form the vocal folds. These folds are what give humans the ability to speak; when air is expelled, the folds vibrate, resulting in vocal sound.

### ***Trachea***

The trachea is divided into the right and left bronchi. Each of the bronchi enters the lungs. When air enters the bronchi, it is warm, humidified, and cleansed of all pathogens. The trachea (windpipe) contains walls covered with ciliated mucous. These walls beat constantly in the opposite direction of incoming air. Because its walls are made up of c-shaped cartilage rings, the trachea is quite rigid; however, the open part of the rings allow the esophagus to expand when one swallows a large amount of food while the opposite ends keep the trachea open.



**Lungs**

Lungs are the primary organs of the respiratory system and occupy the thoracic cavity. When the bronchi enter the lungs, they divide into smaller branches. After the continuous shortening of size, the primary bronchi become the bronchioles, which then divide to become the respiratory zone structures. In the respiratory zone, the transfusion of air to blood takes place. These structures eventually divide into alveoli. Within the lungs, millions of alveoli are responsible for the transfusion of oxygen into the blood. The remaining areas of the lungs are known as the conducting zone structures and contain all the passages to and from the respiratory zone.

## **The Sensory System**

The sensory system is vital for survival, growth, development, and the experience of bodily pleasure. Sense perception depends on sensory receptors that respond to various stimuli. When a stimulus triggers an impulse in a receptor, the action potentials travel to the cerebral cortex where they are processed and interpreted. Only after this occurs is a particular sensation perceived. Some senses, such as pain, touch, pressure, and proprioception, are widely distributed in the body. These are called general senses. Other senses, such as taste, smell, hearing, and sight, are called special senses because their receptors are localized in particular areas (Tortora & Derrickson, 2008).

**Sensory Components**

The two components of sensory experience are reception and perception. Sensory reception is the process of receiving data from the internal or external environment through the senses and includes: visual (seeing); auditory (hearing); olfactory (smell); gustatory (taste); and tactile (touch).

Sensory perception is the conscious process of selecting, organizing, and interpreting data from the senses and transforming this data into meaningful information. This process is influenced by intensity, size, and shape, as well as by past experiences, knowledge, and attitudes. In some situations, the way in which a particular sensation is perceived depends on where it is interpreted in the brain. Nearly everyone is familiar with sensory adaptation in the sense of smell. A particular odor becomes unnoticed after a short time even though the odor molecules are still present in the air because the system quickly adapts to the continued stimulation.

**General Senses**

General senses are found throughout the body. Visceral organs control these senses with the skin, muscles, and joints. General senses include touch, pressure, proprioception, temperature, and pain.

**Touch and Pressure.** As a group, the receptors for touch and pressure are widely distributed in the skin and are sensitive to forces that deform or displace tissues. Three of the receptors involved in touch and pressure are free nerve endings, Meissner's corpuscles, and pacinian corpuscles. These receptors are important in sensing objects in continuous contact with our skin. While Meissner's corpuscles lie just beneath the epidermis and sense light touch stimuli, Pacinian corpuscles are deeper in the dermis and are sensitive to heavy pressure.

**Temperature.** Temperature receptors lie directly under the skin and are widely dispersed throughout the body. The sense of temperature is stimulated by cold receptors (of which there are numerous) and heat receptors (of which there are fewer). Degree of stimulation depends on the number of each type of receptor stimulated. A person determines gradations in temperature by the degree of stimulation of each type of receptor. Extreme cold and extreme heat feel almost the same because the pain receptors are being stimulated. Thermoreceptors are strongly stimulated by abrupt changes in temperature and fade after a few seconds or minutes.

## **The Digestive System**

The digestive system which prepares food for use is one of the most complex systems of the body. When eaten, food cannot reach cells because it cannot pass through the intestinal walls to the bloodstream. The digestive system modifies food physically and chemically and disposes of unusable waste. Physical and chemical modification (digestion) depends on exocrine and endocrine secretions and the controlled

movement of food through the digestive tract. It provides the body with the means of transforming food to energy. Food first enters the digestive system through the mouth, goes through multiple organs, and is transformed into enzymes, glucose, and other nutrients (Martini & Nath, 2008).

### ***Mouth***

As mentioned, food enters the digestive system via the mucous membrane-lined oral cavity. The lips protect the mouth's outer opening, while the cheeks form its lateral walls. The hard and soft palates additionally form anterior or posterior roof. The floor of the mouth is characterized by the muscular tongue. Food is first processed by the teeth. The anterior incisors bite and tear food. Comestibles are then masticated by cheek teeth, premolars, and molars. This process is aided by saliva that enters the oral cavity through ducted salivary glands. The breakdown of food requires mechanical and chemical processes. Taste buds, while making food enjoyable, also alert the body of potential hazards, like toxins.

### ***Swallowing***

Once food leaves the mouth, it crosses the respiratory tract (trachea is anterior to esophagus) by a complicated mechanism known as swallowing. Swallowing empties the mouth and ensures that food does not enter the windpipe. This process involves coordinated activity of tongue, soft palate (pharynx), and esophagus. The first phase is voluntary, as food is forced into the pharynx by the tongue. After this phase, reflex controls swallowing. The tongue first blocks the mouth while the soft palate closes off the nose and the larynx rises, allowing the epiglottis to close the trachea. Food thus moves into the pharynx and onwards by peristalsis aided by gravity. The esophagus, the first part of the digestive tract, runs from the pharynx to the diaphragm and stomach. (Martini & Nath, 2008).

### ***Stomach***

The stomach stores and digests food. Two sphincters, circular valve-like muscles placed outside organs, surround the two openings. Cardioesophageal sphincter guards the entrance from the esophagus while the pyloric sphincter guards the outlet. The stomach secretes acid (to continue the digestive process) and mucus (for self protection). Food in the stomach is churned by three muscular layers to form chyme, a creamy substance voided via pyloric sphincter to the duodenum.

### ***Small Intestine***

The small intestine is the primary organ of the digestive tract. The first part of the small intestine, duodenum, curves around the pancreas, forming the entry of the common bile duct. Chemical degradation of food begins here as enzymes, secreted by the pancreas and duodenum, aided by the emulsification of bile. Most of food digestion occurs in the small intestine. In addition to digesting chyme, the small intestine is responsible for absorbing the processed food into the bloodstream so that energy can be derived from food. This absorption takes place through villi, valve-like structures, which allow solids, liquids, and acids to pass from one structure to another.

### ***Large Intestine***

The large intestine stretches from a valve resting between the bottom of the small intestine and the top of the large intestine to the anus. The main job of the large intestine is to digest any undigested chyme and rid the body of unneeded wastes. The large intestine has many subdivisions including cecum, appendix, colon, rectum, and canal.

### ***Accessory Organs***

In addition to the main digestive organs, several accessory organs include the salivary glands, pancreas, liver, and gallbladder. Salivary glands are placed in three categories: parotid, submandibular, sublingual. Parotid glands are located in the cheeks of the oral cavity, while the others open into the floor of the mouth. Saliva, a mixture of mucus and serous fluids, is produced in various glands. The pancreas includes the endocrine and exocrine glands. Exocrine produces enzymes while endocrine produces insulin. Bile is produced by the liver and secreted via the hepatic duct and cystic duct to the gallbladder for storage. Bile is released into the duodenum via the common bile duct. Bile salts, bile pigments, cholesterol, and phospholipids make up the bile while bile salts and phospholipids emulsify fats. The liver is a multifunctional

organ which receives fat and other nutrients from the small intestine via the hepatic portal system (Martini & Nath, 2008).

## **The Genitourinary System**

The organs involved in production, formation, and release of urine make up the genitourinary system. This system includes the kidneys, ureters, bladder, urethra, and the organs of reproduction - ovaries, uterus, fallopian tubes, vagina, and clitoris in women and the testes, seminal vesicles, prostate, seminal ducts, and penis in men (Kasper et al., 2005).

### ***Renal System***

The kidneys are regulatory organs which maintain volume and composition of body fluid by filtering blood and secreting filtered solutes. Kidneys take blood from the aorta via the renal arteries and return it to the inferior vena cava via the renal veins. Urine, the filtered product containing waste materials and water, is excreted from the kidneys and passes down the ureters, collecting in the bladder. The bladder distends to accept urine, allowing large volumes to be collected without damaging the renal system. When urine is passed, the urethral sphincter at the base of the bladder relaxes, the detrusor contracts, and urine is voided through the urethra (Martini & Nath, 2008).

### ***Male Reproductive System***

The organs of the male reproductive system are specialized for the following functions: production, maintenance, and transportation of sperm, protective fluid (semen), and male sex hormones. To best carry out these functions, the male reproductive anatomy includes internal and external structures.

Most of the male reproductive system is located outside the body. These include the penis, the scrotum, and the testicles.

The penis, the male sex organ, is comprised of three parts: the root, which attaches to the wall of the abdomen; the body, or shaft; and the glans, the cone-shaped end of the penis. The glans, which also is called the head, is covered with a loose layer of foreskin, sometimes removed through circumcision. The opening of the urethra, the tube that transports semen and urine, is at the tip of the glans penis. The body of the penis is cylindrical in shape and consists of three internal chambers made up of sponge-like erectile tissue. This tissue contains thousands of spaces that fill with blood during sexual arousal. As this process occurs, the penis becomes rigid and erect, allowing for penetration during sexual intercourse. The skin of the penis is loose and elastic to accommodate such changes. Semen, which contains sperm, is expelled (ejaculated) through the end of the penis when the man reaches sexual climax (orgasm). When the penis is erect, the flow of urine is blocked from the urethra, allowing only semen to be ejaculated at orgasm (Martini & Nath, 2008).

Scrotum is the loose pouch-like sac located behind the penis. It contains the testicles (also called testes), as well as many nerves and blood vessels. The scrotum functions as a climate control system to protect the testes; normal sperm development requires testes to maintain a temperature slightly cooler than that of the body. Special muscles in the wall of the scrotum allow it to contract and relax, moving the testicles closer to the body for warmth and protection or farther away from the body for cooling.

Testes, oval organs about the size of large olives, lie in the scrotum, secured at either end by a structure called the spermatic cord. The two testes are responsible for making testosterone (the primary male sex hormone) and for generating sperm. Within the testes, coiled masses of seminiferous tubules are responsible for producing the sperm cells through a process called spermatogenesis.

The internal organs of the male reproductive system, also called accessory organs, include the vas deferens, ejaculatory ducts, urethra, seminal vesicles, prostate, and Bulbourethral glands. The vas deferens, a long, muscular tube that travels from the epididymis into the pelvic cavity to just behind the bladder, transports mature sperm to the urethra in preparation for ejaculation. Ejaculatory ducts are formed by the fusion of the vas deferens and the seminal vesicles and empty into the urethra. Urethra, a tube that carries urine from the bladder to outside of the body also controls the ejaculation of semen.

The male reproductive system could not function without hormones, chemicals that stimulate or regulate the activity of cells or organs. The primary hormones involved in the functioning of the male

reproductive system include the follicle-stimulating hormone (FSH), the luteinizing hormone (LH), and testosterone.

### ***Female Reproductive System***

The female reproductive system, like that of the male reproductive system, is designed to carry out multiple functions. These functions include the production of female egg cells necessary for reproduction, called the ova or oocytes, transportation of the ova to the site of fertilization, and the production of female sex hormones. Conception, the fertilization of an egg by a sperm, normally occurs in the fallopian tubes. After conception, the uterus provides a safe environment for fetal development. In the absence of fertilization, the system carries out menstruation (monthly shedding of the uterine lining). During menopause, the female reproductive system gradually stops making the female hormones necessary for the reproductive cycle. When the body no longer produces these hormones a woman is considered to be menopausal.

The female reproductive anatomy includes internal and external structures. The function of the external female reproductive structures (the genitals) is twofold: to enable sperm to enter the body and to protect the internal genital organs from infectious organisms. The main external structures of the female reproductive system include the labia majora, labia minora, Bartholin's glands, and the clitoris. The labia majora, large and fleshy and comparable to the scrotum in males, enclose and protect the other external reproductive organs. Labia minora, lie just inside the labia majora and surround the openings to the vagina (the canal that joins the lower part of the uterus to the outside of the body) and urethra (the tube that carries urine from the bladder to the outside of the body). Bartholin's glands, located next to the vaginal opening, produce a fluid secretion. The two labia minora meet at the clitoris, a small, sensitive protrusion comparable to the penis in males. The clitoris is covered by a fold of skin called the prepuce, which is similar to the foreskin at the end of the penis. Like the penis, the clitoris is very sensitive to stimulation and can become erect.

The internal reproductive organs include the vagina, uterus, ovaries, and Fallopian tubes. The vaginal canal joins the cervix (the lower part of the uterus) to the outside of the body. The uterus (womb), a hollow pear-shaped organ, is home for a developing fetus. The uterus is divided into two parts: the cervix, which is the lower part that opens into the vagina, and the main body of the uterus, called the corpus. The corpus can easily expand to hold a developing baby. A channel through the cervix allows sperm to enter and menstrual blood to exit. The small, oval-shaped glands located on either side of the uterus are called ovaries which produce eggs and hormones. The Fallopian tubes are attached to the upper part of the uterus and allow the ova (egg cells) to travel from the ovaries to the uterus. Conception, the fertilization of an egg by a sperm, normally occurs in the fallopian tubes. The fertilized egg then moves to the uterus, where it implants in the uterine wall.

Females of reproductive age experience cycles of hormonal activity that repeat at about one-month intervals. *Menstru*, meaning "monthly," provides us with the term, menstrual cycle. Each such cycle prepares a woman's body for a potential pregnancy. The average menstrual cycle takes about 28 days and occurs in phases: the follicular phase, ovulatory phase (ovulation), and luteal phase. The term *menstruation* refers to the periodic shedding of the uterine lining. Four major hormones are involved in the menstrual cycle, including the follicle-stimulating hormone and luteinizing hormones, as well as estrogen and progesterone.

The Follicular phase starts on the first day of the cycle. During this phase, the follicle stimulating hormone (FSH) and luteinizing hormone (LH) are released from the brain and travel in the blood to the ovaries. These hormones stimulate the growth of about 15-20 eggs in the ovaries, each in its own "shell," called a follicle. As the follicular phase progresses, one follicle in one ovary becomes dominant and continues to mature. This dominant follicle suppresses other follicles, which then stop growing and die. The dominant follicle continues to produce estrogen.

The Ovulatory phase, which begins about 14 days after the start of the follicular phase, is the midpoint of the menstrual cycle. During this phase, a rise in estrogen from the dominant follicle triggers a surge in the amount of luteinizing hormone produced by the brain, causing the dominant follicle to release its egg from the ovary.

The Luteal phase begins immediately after ovulation. Once it releases its egg, an empty follicle develops into a new structure called the corpus luteum. Progesterone prepares the uterus for a fertilized egg to implant. If intercourse has taken place and a man's sperm has fertilized the egg (a process called conception), the fertilized egg (embryo) will travel through the fallopian tube to implant in the uterus, resulting in pregnancy. If the egg is not fertilized, it passes through the uterus, whose lining breaks down and sheds, starting the next menstrual period.

During fetal life, six to seven million eggs exist in the female baby (Collier, Longmore, Duncan Brown, & Collier, 2006). While no new eggs are produced, the vast majority of these eggs steadily die until they are depleted at menopause. At birth, a baby has approximately one million eggs, and by the time of puberty only about 300,000 remain. Of these, 300 to 400 will be ovulated during a woman's reproductive lifetime.

## **The Endocrine System**

The endocrine system is an integrated system of small organs that controls the release of hormones. This system is instrumental in the regulation of metabolism, growth, development and puberty, tissue function, and mood. (Martini & Nath, 2008).

The nervous system controls the sending of electrical messages which coordinate body movements. Conversely, the endocrine system uses chemicals, known as hormones, to communicate and coordinate functioning. Hormones, specific messenger molecules, are synthesized and secreted by a group of specialized cells called endocrine glands. These glands are ductless, meaning that their secretions (hormones) are released directly into the bloodstream to facilitate travel to target organs.

All hormones are characterized by specific shape, which is accordingly recognized by the corresponding target cells. The binding sites on the target cells are called hormone receptors. Many hormones come in antagonistic pairs that have opposite effects on the target organs. For example, insulin and glucagon have opposite effects on the liver's control of blood sugar level. Insulin lowers the blood sugar level by instructing the liver to take glucose out of circulation and store it, while glucagon instructs the liver to release some of its stored supply to raise the blood sugar level. Much hormonal regulation depends on feedback loops to maintain balance and homeostasis (Martini & Nath, 2008).

### ***Hypothalamus and Pituitary***

The major human endocrine glands include the hypothalamus and pituitary gland. The pituitary gland, called the "master gland," is under the control of the hypothalamus. Together, these glands control many other endocrine functions and secrete a number of hormones, especially several important to female cycles (Tortora & Derrickson, 2008).

A number of other hormones affect various target organs. One non-sex hormone secreted by the posterior pituitary is antidiuretic hormone or ADH. This hormone helps prevent excess water excretion by the kidneys. Another group of non-sex hormones include the endorphins which belong to a category of chemicals known as opiates and serve to deaden our pain receptors. Endorphins which are chemically related to morphine, are produced in response to pain. The natural response to rub an injured area, such as a pinched finger, helps to release endorphins in that area. People who exercise frequently or push their bodies "until it hurts" are consistently stimulating the production of endorphins.

### ***Thyroid Gland***

Thyroid hormones are responsible for the regulation of metabolism, body temperature, and weight. The thyroid gland requires iodine in order to manufacture hormones. If a person lacks dietary iodine, the thyroid cannot make its hormones, causing a deficiency.

### ***Pancreas***

The pancreas has two functions. First, it serves as a ducted gland, secreting digestive enzymes into the small intestine. Second, it functions using the islets of Langerhans to secrete insulin and glucagon to regulate blood sugar. By secreting glucagon, islets cells signal the liver to utilize carbohydrates and raise the



blood sugar level. These cells secrete insulin to command the liver to remove excess glucose from circulation to lower a blood sugar level that is too high.

### ***Adrenal Glands***

The adrenal glands sit on top of the kidneys and consist of the outer cortex and the inner medulla. While the medulla secretes epinephrine (adrenaline) and other similar hormones in response to stressors such as fright, anger, caffeine, or low blood sugar, the cortex secretes corticosteroids such as cortisone. Corticosteroids are anti-inflammatory regulators. Medicinal doses are typically higher than normal natural production.

### ***Gonads***

In addition to producing gametes, the female ovaries and male testes secrete hormones. These hormones (sex hormones) are secreted by the gonads, which in turn are controlled by pituitary gland hormones. While both sexes make some of each hormone, male testes secrete primarily androgens, including testosterone. Female ovaries make estrogen and progesterone in varying amounts depending on menstrual timing. In a pregnant woman, the baby's placenta also secretes hormones to maintain pregnancy.

### ***Pineal Gland***

The pineal gland is located near the center of the brain and is stimulated by nerves from the eyes. The pineal gland secretes melatonin at night when it is dark to promote sleep and depress activity of the gonads. Because melatonin production is affected by the amount of light to which a person is exposed, it additionally affects the circadian rhythm (having an activity cycle of about 24 hours), annual cycles, and biological clock functions (Martini & Nath, 2008).

## **Conclusion**

By the time a human reaches adulthood, the body consists of approximately 100 trillion cells. Each is part of an organ system designed to perform essential life functions. The body's organ systems include: the skeletal system, muscular system, nervous system, cardiovascular and lymphatic systems, integumentary system, respiratory system, sensory system, digestive system, genitourinary system, and endocrine system. The human body is a fascinating and fantastic machine. No one understands all of its many mysteries, and no single source can do justice to its many parts. We have provided an overview from the cellular level through ten body systems. The information learned in this chapter will be useful for the study of disabling conditions. As previously noted, the visual and hearing systems are discussed in chapters in this text specific to those areas.

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# Chapter 3

## MEDICAL TERMINOLOGY

*John Howard, M.D.*

### Introduction

Medical terminology is the specialized science-based vocabulary used to describe the structure and function of the human body as well as those diseases, conditions, and medical procedures used in medicine. The rehabilitation process involves interacting with an individual who has a disability that has been evaluated by one or more physicians. The physicians' reports are full of medical terminology that is often unfamiliar to the rehabilitation counselor. Ideally, medical reports should be immediately meaningful to the counselor without resorting to a medical dictionary. With time, the rehabilitation counselor will achieve that goal. However, rehabilitation counselors at the beginning of their careers cannot be expected to have a comprehensive knowledge of medical terminology.

The goal in mastering medical terminology is to achieve a level of common understanding with the evaluating physician that will permit a greater understanding of the client's physical and functional disorders (Cohen, 2004; Dorland, 2007; Gylys & Wedding, 2005; Marcovitch, 2005; Stedman, 2006; Thomas, 2005). Learning complex medical terminology will seem like a daunting task at first, but acquiring the ability to understand and use medical terminology will greatly aid the rehabilitation counselor's professional effectiveness (Anderson, Anderson, & Glanze, 2006; Applegate, Overton, & Pucillo, 1994). When discussing medical problems with a client, the counselor should never use technical terms unless the client clearly understands their meaning. Often, clients will be reticent to ask for an explanation of medical terminology because they do not want to be seen as ignorant. The use of overly technical medical terminology during an evaluation interview can frustrate real communication with the client.

This chapter provides the rehabilitation counselor with an initial understanding of the origins of medical terminology, how the most commonly used medical terms are linguistically structured, and knowledge of the standard medical terminology reference dictionaries. Inconsistencies, contradictions, and trends in medical terminology are described. The chapter also includes subscription and free internet resources on medical terminology.

### Medical Terms

Familiarizing oneself with strange technical terms is easier if one learns the meanings of the **parts** of words, rather than attempting merely to memorize a particular word of many syllables. For example, at first glance, the terms **sternocleidomastoid** or **hypertrophic pulmonary osteoarthropathy** can be overwhelming. Yet, when divided into their constituent parts, they become more understandable. In the first example, the prefix **stern** refers to the sternum or breastbone; **cleid** indicates the clavicle; and **mastoid** means the bony process behind the ear (the "o"s are merely connectors). The word itself is the name of the muscle that attaches to the three bony structures included in the full term.

The second lengthy technical term, **hypertrophic pulmonary osteoarthropathy**, is the scientific name for clubbing of the ends of the fingers that is seen occasionally as an accompaniment to certain lung disorders. **Hyper** means in excess or more than normal; **trophic** refers to nutrition; and the whole word denotes the enlargement or overgrowth of a body part. **Pulmonary** pertains to the lungs (from the Latin for lung, **pulmo**). **Osteo** signifies a relationship to a bone or the bones (from the Greek **esteon** or bone), while **arthr(o)** denotes a relationship to a joint or joints (the Greek word for joint is **arthron**). The ending **-pathy** (from the Greek **pathos**, meaning "to suffer") signifies a morbid condition or disease; the full word means

any disease of the joints and bones. When the words are used in sequence they indicate a disease overgrowth of the bones and joints (in this instance, the terminal phalanges or finger ends), secondary to a lung disorder. The term accurately describes the nature, location, and etiology (cause) of the disorder, in essence, consolidating a descriptive paragraph into a single term. These examples depict the manner in which medical terms, usually describing clinical disorders or diseases, become clear through a knowledge of the constituent words or parts of words.

### *Analysis of Terms*

Like many specialized vocabularies, medical terminology is built upon recurring components (Applegate et al., 1994; Haubrich, 2003; Layman, 2005). The more familiar the counselor is with these verbal building blocks, the more readily new terms can be understood. In the division of words, one identifies the constituent syllables. First, there is the core of the word, its **root** or **stem** (a term derived from a language such as Greek or Latin and usually describes the part of the human body). Second, there are modifying terms that precede (**prefix**) or follow (**suffix**) the root. As an example, in the word **dysmenorrhea**, the root is **-men-**, for month. The prefix **dys-** means difficult or painful, and the suffix, **rrhea**, denotes flow. With knowledge of the components, the meaning of dysmenorrhea is understood as “painful menstruation.”

The technical vocabulary of medicine is essentially a way of organizing medical information (Applegate et al., 1994; Magalini & Magalini, 1997). The vocabulary of medicine has developed over centuries, and, as a result, is not always clear and predictable. There are ambiguities and inconsistencies, duplications and contradictions. Medical terminology still embodies archaic terms generated by theories no longer valid, but it is also constantly adding new terms arising from such emerging fields like genomic medicine - a field of medicine made possible by the mapping of all the genes in the human genome (National Human Genome Research Institute, 2007). The hazards of language confusion can be avoided if one becomes familiar with the origins and creation of medical terms (Haubrich, 2003; Jaeger, 1978).

### *Origins of Medical Terms*

Most of the terms encountered in medicine today have their historical origin in Greek and Latin words, their initial formation having been somewhat capricious. Often, today’s vocabulary represents a combination of parts from both languages and others. For instance, many of the most common or lay terms for body parts are of Anglo-Saxon origin: eye (**eage**), heart (**heorta**), skull (**skulle** [a bowl]), hip (**hype**), and kidney (**kidenie**), for example (Haubrich, 2003). Many body parts underwent name changes through Middle English before taking today’s spellings, for example **heorta** or **herte** to **heart** (Jaeger, 1978). Other languages have contributed medical terms to English. From French have come such terms as jaundice (**jaune** [yellow]), **malaise**, **gout**, **curette**, **grand mal**, and **gavage**. German contributed **anlage**, Dutch gave us **sprue**, and from Italian we derived **influenza** and **petechia**. **Agar** (initially **agar agar**) is Singalese in origin, as is **beriberi**. Japan added **tatsugamuchi** fever, and from the Amazon Indians came **ipecac**. **Alkali**, **alcohol**, and **bezoar** descended from the Arabic language.

As interesting as tracing the etymology (the study of the origin of words) of a word can be, the current definition of a word can be different than its etymologic origin. For example, the term **mastoid** refers to the rounded prominence of the temporal bone behind the ear. In pre-antibiotic days, it was subject to inflammation and corrective surgery. It was believed to resemble a female breast, so the name was drawn from the Greek **mastos** (breast) and **eidos** (like). The anatomic designation should not be misinterpreted as being similar to such terms as **mastitis** or **mastectomy**. In most instances, however, the original meaning persists despite the lapse of centuries (Haubrich, 2003; Jaeger, 1978).

Several word roots occur in both Latin and Greek but have considerably different meanings. Perhaps the most frequently encountered pair is the root **ped(o)** -, which in Latin means **foot** (**pedicle**, **pedicure**), but in Greek means **child** (**pediatrics**). Other word roots can be confusing because they sound alike, but have different meanings: **hydr(o)** - for water (as in **hydrarthrosis**) and **hidr(o)** - for sweat (as in anhidrosis). Other difficulties arise when both Latin and Greek forms are inserted into contemporary language. For example, numbers derived from Latin and Greek are used in prefixes, yet each system is different. From Latin, **one** (1) provides the prefix **uni-**, as in unilateral (affecting one side only) or

**unidirectional** (flowing in one direction only). **One** in Greek provides the prefix **mono-**, as in **monochromatic** (existing in or having only one color) or **monocyte** (a leukocyte [white blood cell] having only one nucleus). Iron compounds usually carry the prefix **ferr(o)**, from the Latin **ferrum**, meaning iron, as in **ferrous** gluconate or **ferric** oxide. Still, there are many words that use the Greek **sider(o)**, from **sideros** (iron), as in **siderosis** (a lung disorder resulting from the inhalation of iron particles), or **sideropenia** (an iron deficiency in the body).

Not all medical terms emerge purely from their language roots. There are **eponyms** that are based on a person, usually the individual who first described the clinical entity or advanced a theory concerning the disorder (Sloane, 1997). Many diseases and diagnostic tests have such name origins, e.g., **Alzheimer's disease**, **Hansen's disease**, **Hodgkin's disease**, **Mantoux test**, and **Rubin's test** (Manser, 1996). Several operative procedures carry the originators' names as in **Matas' operation** or **Potts' operation**. Eponyms appear in the names of body parts such as **Hesselbach's triangle** and instruments such as **galvanometer** (named after Luigi Galvani) and **voltmeter** (named after Alexander Volta). Occasionally, a disease will derive its name from a geographic location such as **Bornholm disease** (a Danish Island in the Baltic Sea) or **Lyme disease** (Old Lyme, Connecticut), or from the name of the type of patients first affected such as **Legionnaire's disease** (bacterial pneumonia first seen in American Legion members).

The method in which medical terms are derived is changing and, as more is learned of an old perplexing disorder or as a new disease is identified, more descriptive names are given to the processes. For example, **Bornholm**, also is a disease called **epidemic pleurodynia**; others, like **tularemia** (named after the city of Tulare, California) continue to be identified by their old designations.

Certain disorders bear the designation **syndrome** (from the Greek meaning to run together). **Syndrome** means a set of characteristics, such as symptoms or specific diseases, which often occur together so that one characteristic leads the physician to look for the others. The term **syndrome** is most often used when the reason that the features occur together (the pathophysiology of the syndrome) has not yet been determined. For instance, 8,000 people died of severe acute respiratory syndrome, or **SARS**, between 2002 and 2003. SARS was later found to be caused by a coronavirus.

Physicians naming disorders may be from any one of many countries, adding more to the international flavor of medical nomenclature. From literature are the "**Alice in Wonderland**" **syndrome** (a delusional state) and the **Munchhausen syndrome** (a chronic disorder with physical symptoms that is not a natural abnormal condition of the body but has been "manufactured" by the patient). The former comes, of course, from Lewis Carroll's (Charles Dodgson's) wonderful story, while the latter is from the 18<sup>th</sup> Century baron, the teller of tall tales.

**Acronyms** are formed from the initials of lengthy names, such as **ACTH** (adrenocorticotropic hormone), **AIDS** (acquired immunodeficiency syndrome), **HIV** (human immunodeficiency virus), **LASER** (light amplification by stimulated emission of radiation), and **MRI** (magnetic resonance imaging) (Jablonski, 2005).

Preventive and therapeutic medications may have several different names. They can be named "chemically" (rarely referred to in medical reports), or by their generic name, or by a brand or trade name given to the agent by a particular manufacturer. For example, using the International Union of Pure and Applied Chemistry (IUPAC) nomenclature, the chemical descriptor of the most commonly used anti-anxiety drug is 7-chloro-1-methyl-5-phenyl-1,3-dihydro-2H-1,4-benzodiazepin-2-one, its generic name is **diazepam**, and the names under which it is branded and marketed in the United States are **Valium**, **Stesolid**, **Diazemuls**, **Seduxen**, **Bosaurin**, **Diapam**, **Antenex**, **Ducene** and **Apozepam**. Similarly, **paracetamol** is **acetaminophen**, and is **Tylenol**. There is no easy way to learn the names of various medications and in medical reports physicians sometimes refer to the same medication by different names. New medications are approved by the United States Food and Drug Administration (FDA) all the time; the rehabilitation counselor should consult with an authoritative pharmaceutical reference to learn more about new medications (Physicians' Desk Reference, 2009).

Finally, certain words are derived from an entity or action by a vocal imitation of the sound associated with it (onomatopoeic), as heard in **buzz** or **hiss**. Examples in medical terminology include **hiccup**, **tympany** (the sound produced by percussion over a cavity containing air), and **belch**. **Rale** (from

the French **r**âle meaning rattle) indicates an abnormal sound heard when listening to respiration with the stethoscope.

It is not within the scope of this chapter to provide a comprehensive glossary of medical terms. Medical dictionaries and other reference works cited at the end of this chapter are useful in the rehabilitation counselor's library. Many of the standard medical dictionaries are accessible electronically with powerful dictionary search engines. A number of internet resources for medical terms are available, but the user should exercise diligence when using internet definitions for medical terminology.

What follows are lists of prefixes, suffixes, anatomical landmarks, body areas, and medical abbreviations that are important components of medical terms needed by rehabilitation counselors in daily practice. These terms appear repeatedly in written medical reports and during counselors' discussions with physicians.

## Mastering Medical Terminology

### *Prefixes*

Word roots can be modified by prefixes that denote:

1. Position in time or space
  - ab - means **away from**  
**abnormal** – away from normal  
**abduction** – drawing away from the midline of the body by the arm or leg
  - circum – means **around**  
**circumcision** – a cutting around of the prepuce  
**circumarticular** – around a joint
2. Quantitative information
  - a or an – means **without**  
**anorexia** – without appetite  
**anoxia** – without enough oxygen
  - hemi – means **half**  
**hemiplegia** – paralysis of one lateral half of the body  
**heminephrectomy** – removal of a portion of a kidney
  - diplo – means **double**  
**diplopia** – double vision  
**diploscope** – an apparatus for study of binocular vision
  - quint – means **fifth**  
**quintipara** – a woman who has five pregnancies continued beyond the 20<sup>th</sup> week of gestation  
**quintuplet** – one of five offspring produced in one gestation period
3. Qualitative information
  - mal – means **bad** or **ill**  
**malfunction** – defective function  
**malocclusion** – faulty positioning of the upper or lower teeth in relation to the other
  - eu – means **good** or **healthy**  
**euphoria** – sense of well-being or condition of good health  
**euthanasia** – easy or painless death

- 
4. Sameness and difference
    - hetero – means **different**  
**heterogeneous** – differing in kinds or nature  
**heterosexual** – sexual orientation directed to the opposite sex
    - homo – means **same**  
**homogeneous** – of the same kind  
**homosexual** – sexual orientation directed to the same sex
  5. Physical attributes (size, shape, color)
    - micro – means **small**  
**microcephalia** – abnormal smallness of the head  
**micromelia** – abnormal smallness or shortness of the extremities
    - brachy – means **short**  
**brachycephalia** – having a short head  
**brachydactylia** – having abnormally short fingers and toes
    - oxy – means **pointed** or **sharp**  
**oxycephaly** – having a high and pointed head
    - albo – means **white**  
**albinism** – absence of pigmentation  
**albinuria** – passing of white or colorless urine

### *Suffixes*

Suffixes and significant word endings characteristically function in compound words to:

1. Form adjectives
  - -al means **pertaining to**  
**peritoneal** – pertaining to the peritoneum  
**arterial** – pertaining to an artery
  - -ible, or -able means **ability**  
**digestible** – capable of being digested  
**operable** – subject to being operated
2. Express diminutive size – a number of suffixes serve this purpose,
  - -cule, -icle, -ium, -ole, -ule, and variations  
**arteriole** means a small artery  
**granule** means a small grain
3. Indicate a surgical procedure
  - -ectomy means **removal** of an organ or part  
**appendectomy** – removal of the appendix
  - -lysis means **loosening**, usually of adhesions  
**cardiolysis** – freeing of the heart of pericardial adhesions
  - -ostomy means an operation in which an **artificial opening** is formed between two hollow organs or between one or more such viscera and the abdominal wall for discharge of intestinal content or urine.  
**colostomy** – the surgical creation of an opening between the colon and the exterior of the body

- -pexy means **fixation**  
nephrop**exy** – the surgical attachment of a floating kidney
  - -plasty means **plastic surgery**  
blephar**oplasty** – plastic surgery of the eyelids
  - -rrhaphy means **suture** or **operative repair**  
tenor**rrhaphy** – the suturing of a tendon
  - -scopy means **viewing** or **examining**, usually with an instrument  
endos**copy** – visual inspection of any body cavity by means of an endoscope
  - -tomy means **cutting** or **incision**  
lapar**tomy** – the surgical opening of the abdomen
  - -tripsy means the **intentional surgical crushing** of a structure  
litho**tripsy** – the disintegration of a kidney stone by a high-energy shockwave
4. Express conditions or changes related to pathological processes
- -mania means **excessive excitement** or **obsessive preoccupation**  
pyrom**ania** – an irrational compulsion to set fires
  - -sis means an **action, process, or condition**  
silicob**sis** – a fibrotic disorder of the lungs following inhalation of dust containing silicone dioxide  
thoracent**esis** – surgical puncture of the chest wall for removal of fluid
  - -itis means **inflammation**  
appendic**itis** – an inflammation of the appendix

Besides these loosely formulated classes of suffixes, there are a number of miscellaneous word endings, the most significant of which is included in the basic prefix/suffix vocabulary that follows. Rehabilitation counselors need to be familiar with this vocabulary.

### *Basic Prefix and Suffix Terms*

The following list of prefix and suffix terms aid in understanding medical terminology. This list will familiarize the reader with the flexibility of nomenclature. As new terms are encountered, the counselor can more readily recognize the components from which they are made.

<u>Prefix/Suffix</u>	<u>Definition</u>	<u>Example</u>
-algia	pain	neural <b>gia</b>
angio-	blood vessel	<b>angi</b> ogram
ante-	before	<b>ante</b> cubital
arth-	joint	<b>arth</b> roscopy
blephar-	eyelid	<b>blephar</b> ospasm
cardi-	heart	electro <b>cardi</b> ography
cele-	1) a swelling 2) a hernia	varico <b>cele</b> recto <b>cele</b>
cerebr-	cerebrum	<b>cerebral</b>
chole-	gall or bile	<b>cholec</b> ystectomy
chondr-	cartilage	<b>chondro</b> costal
contra-	opposed to	<b>contra</b> ception



cost-	rib	intercostals
cyst-	bladder	<b>cystitis</b>
-desis	fusion	<b>arthrodesis</b>
derm-	skin	<b>dermatology</b>
dys-	difficult, abnormal	<b>dyspnea</b>
ect-	outside	<b>ectomorph</b>
-ectomy	removal	<b>spenectomy</b>
-emia	condition of the blood	<b>polycythemia</b>
enceph-	brain	<b>encephalitis</b>
end-	within	<b>endocrine</b>
enter-	intestine	<b>enterostomy</b>
epi-	upon, above	<b>epithelium</b>
genic-	giving rise to	<b>psychogenic</b>
glyco-	sugar	<b>glycosuria</b>
hemi-	half	<b>hemiplegia</b>
hepat-	liver	<b>hepatitis</b>
hyper-	in excess	<b>hypertension</b>
hyster-	uterus	<b>hysterectomy</b>
-iosis	a pathologic condition	<b>amebiosis</b>
-itis	inflammation	<b>tonsillitis</b>
leuk-	white	<b>leukocyte</b>
lip-	fat	<b>hyperlipemia</b>
-lith	stone	<b>nephrolithiasis</b>
-megaly	a state of largeness	<b>hepatomegaly</b>
myel-	1) bone marrow 2) spinal cord	<b>myelocyte</b> <b>poliomyelitis</b>
myo-	muscle	<b>myocardium</b>
neph-	kidney	<b>nephrosis</b>
-oid	resembling	<b>thyroid</b>
-oma	tumor or swelling	<b>carcinoma</b>
-oscopy	visual examination	<b>laparoscopy</b>
-osis	disease process or condition	<b>necrosis</b>
osteo-	bone	<b>osteoarthritis</b>
-ostomy	creation of an artificial opening in an organ	<b>tracheostomy</b>
-otomy	incision	<b>craniotomy</b>
para-	near, alongside, beyond, outside	<b>paravertebral</b>
-pathy	disease of	<b>cardiomyopathy</b>
-penia	lack of	<b>leukopenia</b>

-plasty	reparative or reconstructive surgery	<b>rhinoplasty</b>
pneumo-	air	<b>pneumothorax</b>
pre-	before	<b>precancerous</b>
pro-	in front of, before	<b>prognathism</b>
proct-	rectum or anus	<b>proctology</b>
-ptosis	lowering of an organ or part	<b>nephroptosis</b>
pyel-	pelvis	<b>pyelogram</b>
pyo-	pus	<b>pyorrhea</b>
spondyl-	vertebra	<b>spondylolysis</b>
sub-	under	<b>submandibular</b>
super-	above, excessive	<b>supersensitivity</b>
supra-	above	<b>supraventricular</b>
syn-	together	<b>synarthrosis</b>
trans-	across	<b>transurethral</b>
tri-	three	<b>triceps</b>

### ***Anatomical Landmarks***

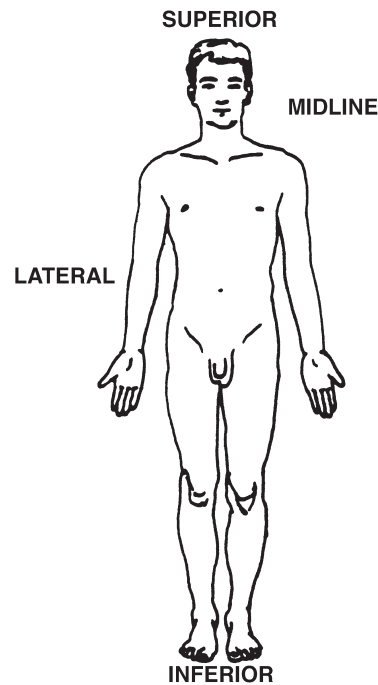
A rehabilitation counselor should be familiar not only with the names of anatomic structures and pathological conditions, but with the “landmark” terms that designate anatomic position and direction. These are described in relation to the patient who is standing in the erect position, looking forward, with arms to the side of the body, and hands with palms forward (see Figure 1). The most commonly used landmark terms are the following:

<b>lateral</b>	to the side (used in contradistinction to medial)
<b>medial</b>	to the center
<b>superior</b>	above (used in contradistinction to inferior)
<b>inferior</b>	below
<b>anterior</b>	before or in front (used in contradistinction to posterior)
<b>posterior</b>	behind or in back
<b>proximal</b>	nearest to the point of attachment or center of the body (used in contradistinction to distal)
<b>distal</b>	farthest from the center
<b>prone</b>	lying face downward (used in contradistinction to supine)
<b>supine</b>	lying face upward
<b>volar</b>	pertaining to the sole of the foot or the palm of the hand
<b>dorsal</b>	pertaining to the back (used in contradistinction to ventral)
<b>ventral</b>	pertaining to the front or abdominal surface
<b>palmar</b>	pertaining to the palm of the hand
<b>plantar</b>	pertaining to the sole of the foot

### ***Body Areas***

Specific sites of illness or injury are usually designated by terms derived from the adjacent anatomic structure. Among the commonly used terms to indicate body areas are the following:

**Figure 1**  
**Anatomical Position of the Human Body for Descriptive Purposes**



<b>abdominal</b>	pertaining to the stomach and intestinal area
<b>carpal</b>	pertaining to the wrist
<b>cervical</b>	pertaining to the seven vertebrae in the neck, or the neck region
<b>costal</b>	pertaining to the ribs
<b>cranial</b>	pertaining to the skull
<b>femoral</b>	pertaining to the thigh
<b>frontal</b>	pertaining to the forehead
<b>lumbar</b>	pertaining to the five vertebrae in the lower portion of the back, or that region
<b>pelvic</b>	pertaining to the pelvic girdle
<b>renal</b>	pertaining to the kidney area
<b>sacral</b>	pertaining to the four vertebrae in the lowest portion of the back, or that region
<b>sternal</b>	pertaining to the sternum or breastbone
<b>thoracic</b>	pertaining to the twelve vertebrae in the upper portion of the back, or that region

### *Medical Abbreviations*

Abbreviations denote instructions that are included in prescriptions for medications; some signify symptoms as reported by patients, and some refer to anatomic parts or body systems (Layman, 2005; Press, 1997; Sloane, 1997). Examples are noted:

<b>a.c.</b>	before meals (ante cibum)
<b>b.i.d.</b>	twice daily (bis in die)
<b>B.P.</b>	blood pressure
<b>C-1 through C-7</b>	cervical vertebrae by number

<b>CBC</b>	complete blood count
<b>C.C.</b>	chief complaint
<b>C.N.</b>	cranial nerve
<b>C.N.S.</b>	central nervous system
<b>CPR</b>	cardiopulmonary resuscitation
<b>CT</b>	computerized tomography
<b>CVA</b>	cerebrovascular accident
<b>DB</b>	decibel
<b>DX</b>	diagnosis
<b>ESR</b>	erythrocyte sedimentation rate
<b>F.H.</b>	family history
<b>FX</b>	fracture
<b>GI</b>	gastrointestinal
<b>Hg</b>	hemoglobin or mercury
<b>GU</b>	genitourinary
<b>HBV</b>	hepatitis B virus
<b>HDL</b>	high density lipoprotein
<b>h.s.</b>	at bedtime (hora somni)
<b>H &amp; P</b>	history and physical examination
<b>HX</b>	history
<b>L-1 through L-5</b>	lumbar vertebrae by number
<b>L.L.Q.</b>	left lower quadrant
<b>L.M.P.</b>	last menstrual period
<b>L.U.Q.</b>	left upper quadrant
<b>MRI</b>	magnetic resonance imaging
<b>OA</b>	osteoarthritis
<b>O.D.</b>	right eye (oculus dexter)
<b>O.S.</b>	left eye (oculus sinister)
<b>p.c.</b>	after meals (post cibum)
<b>P.H.</b>	past history
<b>P.I.</b>	present illness
<b>p.r.n.</b>	as needed (pro re nata)
<b>q.i.d.</b>	four times daily (quater in die)
<b>RA</b>	rheumatoid arthritis
<b>R.B.C.</b>	red blood count
<b>R.L.Q.</b>	right lower quadrant
<b>R/O</b>	rule out
<b>R.U.Q.</b>	right upper quadrant
<b>Rx</b>	prescription
<b>S-1 through S-5</b>	sacral vertebrae by number

<b>SOAP</b>	subjective findings, objective findings, assessment, plan
<b>T-1 through T-12</b>	thoracic vertebrae by number
<b>t.i.d.</b>	three times daily (ter in die)
<b>TX</b>	treatment
<b>W.B.C.</b>	white blood count

The abbreviations cited are examples of the hundreds, perhaps thousands, in use. Unlike medical nomenclature, abbreviations cannot be divided logically into components, nor is it feasible to try to memorize a list of abbreviations. With experience, one becomes increasingly competent in interpreting this medical shorthand. Available sources, such as those noted in the reference section of this chapter, will assist the counselor when reading or writing unfamiliar abbreviations (Jablonski, 2005; Press, 1997; Sloane, 1997; Stedman, 2008).

## Inconsistencies, Contradictions, and Trends

### *Inconsistencies and Contradictions*

Certain words within the medical vocabulary have multiple meanings. The word **sinus**, for instance, can be used in several contexts depending upon the specific meaning intended, e.g., nasal sinus, sinus node in the heart, anal sinus. The reader needs to be sensitive to the context in which it is used to determine which of various meanings is intended. Conversely, several names may apply to a single medical entity without necessarily conveying different meanings. **Brucellosis**, for example, is sometimes called undulant fever, sometimes Bruce's septicemia, and sometimes by a geographic name, Malta fever. Several disorders still carry the names of occupational worker groups who were primarily affected by the disease. Some examples follow: arc-welder's lung (**siderosis**), brown lung (**byssinosis**), white lung (**asbestosis**), and black lung (**coal workers' pneumoconiosis**).

### *Trends in Medical Nomenclature*

Much of medical terminology has remained fairly static over an impressive span of time, but the vocabulary of medicine, like the science itself, is ever-changing. A rehabilitation counselor must be alert to shifts in usage and be aware of additions being made. For example, the current trend is away from the use of eponyms, for such designations provide little information about the nature of a disease or the body organ system that is affected. Another recent trend is to use generic names rather than the brand or trademark name when specifying a pharmaceutical agent, e.g., diazepam instead of Valium (Drake & Drake, 2006; Haubrich, 2003; Magalini & Magalini, 1997; Sloan, 1997). Elements of medical slang and verbal shorthand may creep into reports where the wording is not defined in dictionaries (Segen, 1995). These citations can be confusing to the reader.

Technological changes often generate words for new processes and equipment; these innovations find a place within medical terminology, as did **LASER** when the device first appeared. Keeping current with medical terminology is a continuing assignment for both physician and rehabilitation counselor. The counselor needs a medical dictionary close by whenever reviewing medical reports.

Finally, the current interest in alternative and complementary medicine has brought into the medical arena an additional vocabulary comprising names of various herbal preparations, nutritional supplements, and alternative therapies. Dictionaries and texts are available to aid in defining this set of terms (*Physician's Desk Reference [PDR] for Herbal Medicines*, 2009; Segen, 1998; Leikin & Lipsky, 2003).

### *Aids in Learning*

Acquiring skill in the understanding and use of medical terminology is hastened by the use of one or more currently available learning aids obtainable in various media. A print reference is the standard tool for any rehabilitation counselor. Other learning formats are available on tape cassettes or CD-ROMs; some of these electronic devices are sold together with a standard print reference. Publications devoted to the explanation of medical nomenclature number in the hundreds. These publications may be loyal to the

terminology of medicine in general, to the language of certain specialties, or to the words and expressions common to a specific bodily organ system. Certain works are faithful to medical abbreviations, or acronyms, while others focus on the names of syndromes or medical eponyms. While the dictionaries cited in the list of references are large and heavy, and defy one's carrying them about; there are shorter publications that are portable, usually bearing the sub-titles of "medical desk dictionary," "concise dictionary" or "pocket reference library" (Sloane, 1997; Thomas, 2005). Both electronic medical dictionaries and medical spell checkers are available on the internet.

## Conclusion

Medical reports and records constitute the continuing bond between the rehabilitation counselor and physician. Terminology needs to be immediately meaningful to the counselor who works with individuals who have a wide range of disabilities and chronic medical conditions. With the general understanding of medical roots, prefixes, and suffixes provided by this chapter, together with the use of a print or electronic medical dictionary, the counselor can become reasonably adept at interpreting and comprehending medical terms as they are used in physicians' written reports. When consulting with a physician, it is necessary to have a clear understanding of the aspects of disabilities, as well as medical terminology, to establish realistic plans that help clients return to more satisfying and productive lives.

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## **Internet Resource Links**

A wide variety of medical terminology resources are available on the internet, but the user should exercise caution in accessing such resources. The National Library of Medicine has a useful site called Medline Plus which offers a dictionary of medical terms and is available at <http://www.nlm.nih.gov/medlineplus/mplusdictionary.html>. Martindale's Virtual Medical Center offers a listing of medical dictionaries, abbreviations, eponyms, medical procedures, and tests and is available at [http://www.martindalecenter.com/MedicalD\\_Dict.html](http://www.martindalecenter.com/MedicalD_Dict.html). Various medical schools have sites that offer medical terminology resources. An example is the medical reference guide offered by the University of Maryland Medical Center, available at <http://www.umm.edu/glossary/>.

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## Chapter 4

# CANCER

*Leo E. Orr, II., M.D.*

*Leo M. Orange, M.S.*

## Introduction

Cancer is a general term used to describe a group of tissues in which some cells in the human body grow uncontrollably, invade and destroy adjacent tissues, and, if malignant, spread from their organ of origin to other sites in the body. Additionally, cancer cells, unlike non-cancerous cells, are immortal, continuing to grow and destroy normal tissue. Cancer can be malignant (can metastasize distant from their organ of origin) or benign (grow only at their site of origin). The branch of medicine concerned with the study, diagnosis, treatment, and prevention of cancer is called oncology.

The human body contains billions of cells; each of these cells in turn contains 46 chromosomes that house DNA, the genetic blueprint of life. Within the chromosomes approximately 80,000 genes instruct each body organ regarding growth and function by initiating a vast number of messages (Tannock & Hill, 2005). Chromosomes reproduce themselves by multiple cell divisions, providing opportunities for error. When these errors or mutations occur, the altered genes send incorrect messages. When such an error is received, the cell begins to grow uncontrollably and often rapidly, transmitting the new genetic characteristics to its progeny. If the growth proceeds to the point that a lump forms, it is called a malignant tumor or cancer.

Cancer is not a modern disease. The word “cancer” is derived from the Greek word “karkinos,” meaning crab. In the past, women found to have breast cancer often experienced large, highly visible veins surrounding the tumor which had features resembling the claws of a crab. The earliest anthropological evidence of cancer was found in a one million year-old skeleton of a man unearthed in Java. References to cancer are found in early Hindu and Egyptian writings; mummies from as early as the 3<sup>rd</sup> millennium BC showed signs of bone cancer. Though characterized by ancient roots, cancer has been associated with fear and stigma until recently (Raghavan et al., 2006).

For many years, the causes and methods of transmission of cancer were unknown. At the beginning of the century, few means existed to diagnose cancer at an early stage. The majority of individuals were diagnosed only after long experiences of debilitation, pain, and possible disfigurement. The only treatment modality in existence, surgery, was radical and only sporadically successful in those patients with advanced cases of the disease (Rossi, Cady, & Martin, 2000). The public thus learned to interpret the diagnosis of cancer as a severe disability, ultimately causing death.

Cancer diagnoses were further confused with findings of syphilis, another common but untreatable illness that, like some forms of cancer, often produced unsightly lesions. As a result of its common confusion with syphilis, individuals with cancer felt the additional burdens of guilt and embarrassment as a result of having a sexual meaning applied to their disease. A diagnosis of cancer came to be equated not only with physical consequences such as severe pain, disfigurement, and disability, but with an emotional impact including dependence, isolation, and death. Even with the advent of modern methods of cancer treatment and substantial success rates, many fears and misconceptions once plaguing people diagnosed with cancer, continue to present a challenge to physicians and rehabilitation counselors.

Cancer currently affects one in four people in the United States. In 2006, over 1.4 million new cases were diagnosed; it is the second leading cause of death, following heart disease. Although cancer remains among the worst fears, it is becoming increasingly clear that cancer is not the “death sentence” it once was;

**Table 1**  
**Leading Sites of New Cancer Cases - 2008 Estimates**

<b>Male</b>	<b>Female</b>
Prostrate – 25%	Breast – 26%
Lung & bronchus – 15%	Lung & bronchus – 14%
Colon & rectum – 10%	Colon & rectum – 10%
Urinary bladder – 7%	Uterine corpus – 6%
Non-Hodgkin lymphoma – 5%	Non-Hodgkin lymphoma – 4%
Melanoma of the skin – 5%	Melanoma of the skin – 4%
Kidney & renal pelvis – 4%	Kidney & renal pelvis – 3%
Leukemia – 3%	Leukemia – 3%
Oral cavity & pharynx – 3%	Thyroid – 4%
Pancreas – 3%	Ovary – 3%
All other sites – 20%	All other sites – 23%

only one of six individuals diagnosed with cancer in 2006 will die from the disease. The leading sites of new cancer cases are shown in Table 1 (National Cancer Institute, 2008).

Although lung cancer incidence rates in women have recently stabilized, lung cancer remains the leading cause of cancer death in women. This highlights the need to reduce smoking prevalence and environmental tobacco smoke (ETS) exposure among all women, focusing especially on those populations whose tobacco use and ETS exposure remains high, such as women with lower levels of education.

Among major racial/ethnic groups, African-Americans have the highest rate of new cancers. Rates are relatively low among American Indians/Alaska Natives with regionally higher rates of some cancers. These disparities are not likely due to genetic differences. Rather, they are more likely due to social, cultural, behavioral, and environmental factors (National Cancer Institute, 2008).

## **Etiology**

### ***Gene Transformation***

A landmark scientific development was the discovery that normal genes may be transformed into the genes that promote the growth of cancer. Oncogenes, usually held in check by other genes called tumor suppressor genes, usually remain dormant within the human cell. However, when a stimulus or chemical agent turns on a “switch,” these oncogenes can be initiated to transform normal cells into cancer cells. Cancer is currently viewed as a multi-step process, with changes to the gene accumulating and interacting over time, leading to development of a malignant growth. When the triggers for such growth derive from chemical or foreign sources, they are termed carcinogens. While carcinogens serve as “initiators” for the cancer process, other factors may act as “promoters,” which accelerate the growth of abnormal clone cells (El-Deiry, 2003; Haskell & Berek, 2001).

### ***Environmental Factors***

Major carcinogens associated with an increased risk of cancer include tobacco (30-32%), diet (30-35%), and viruses and infections (10%). Tobacco use is linked to a number of malignancies, including those occurring in the lungs, larynx, pharynx, esophagus, bladder, pancreas, and cervix. Epidemiological studies have suggested that up to 37% of all cancers are due to the effects of smoking. Other causes include gender factors (7%), unknown causes (5%), industrial occupations (4%), heredity (2%), environmental pollution (2%), radiation - environmental, medical, and diagnostic (1%), sunlight - ultraviolet (1%), additives (1%), and alcohol (3-4%). Alcohol is associated with many cancers of the upper respiratory and

digestive tracts; both alcohol and tobacco serve as initiators and promoters and are synergistic rather than additive in cancer causation (El-Deiry, 2003; Tannock & Hill, 2005).

### ***Diet***

An individual's diet has a great effect on overall health, including the development of diseases like cancer. A high fat intake appears closely related to an increased risk of cancer in multiple areas of the body such as breast, colon, endometrium, pancreas, and possibly the prostate and ovaries (Jacobsen et al., 2004; Kleinsmith, 2006). The evidence of the link between diet and cancer rates is undeniable - migrant workers who take on the diet of the country to which they move, experience frequencies of cancer similar to the natives of that country. Additionally, changing rates of cancer for natives within countries is correlated with changes in dietary practices. Striking examples include the decline in both fat consumption and cancer mortality in Europe during World Wars I and II, and the parallel increase in breast cancer rates and fat consumption in Japan after the conclusion of these wars.

### ***Occupational Causes***

A minority of cancers are connected with occupational exposures (Cohen & Markman, 2008). Exposure to ultraviolet light, for example, is known to increase the risk of skin cancer of all types (basal cell, squamous cell, and melanoma), leading to a high incidence in people exposed to sunlight for prolonged periods of time. Exposure to ionizing radiation is a rare but long recognized cause of cancer. The development of malignancy following exposure to atomic bomb blasts and intense nuclear radiation can be quantified. Viruses cause several different malignancies including cervical cancer, Burkitt's lymphoma, a variety of lymphocytic leukemias, and AIDS-related neoplasms.

### ***Genetics***

Genetic factors are powerful determinants of certain cancers. Many families demonstrate increased risk of breast, ovary, colon, lung, and malignant melanomas. When identifiable abnormal oncogenes are found in family clusters with breast and ovarian cancer, family members have up to a 50% risk of developing that type of tumor. Although controversial, the discovery of a genetic predisposition in an unaffected individual may allow for increased cancer surveillance, possible early destruction of a new neoplasm, and other preventive treatment modalities (Kleinsmith, 2006).

### ***Symptoms***

Once a malignant tumor has grown in size, its presence is usually signaled in several of ways: (a) the tumor presses on nearby tissues producing pain; (b) it becomes so large that it is seen or felt; (c) the malignancy grows into nearby blood vessels and produces bleeding; and (d) it causes a change in the way some organs work, resulting in symptoms (Haskell & Berek, 2001). For example, persistent dysphagia (trouble swallowing) may indicate a tumor involving the esophagus. The symptoms of pressure, bleeding, a mass, unusual appearance, or interference with function, are reflected in the American Cancer Society's (2006) list of "seven early warning signs" (noted by the letters C-A-U-T-I-O-N):

Change in bowel or bladder habits.

A sore that does not heal.

Unusual bleeding or discharge.

Thickening or lump in the breast or elsewhere.

Indigestion or difficulty swallowing.

Obvious change in mole or skin nodule.

Nagging cough or hoarseness.

## Diagnosis

### *Physical Examination*

If a person identifies any of the above signs, an appointment with a physician for cancer screening is warranted. During cancer screening, the physician examines the body with emphasis on the parts of the body most prone to malignancy. Salient regions include the nose and throat, breasts in women (and, to a lesser extent, in men), and prostate in men. Pelvic examinations in women, including Pap smears, are essential to aid in detecting cancer of the cervix, uterus, and ovaries. The physician conducts a rectal examination; palpates the lymph nodes in the neck, axillae, and inguinal areas; and palpates the abdomen for enlargement of the spleen, liver, kidneys, or a mass arising in the abdomen. Examination includes questions about abnormal bodily functions and information about a family history of cancer.

### *Laboratory Testing*

**Nonspecific tests.** Blood tests that may reveal abnormalities in the blood indicating the presence of illness are analyzed; a complete blood count is obtained. These tests provide clues used to determine whether more definitive testing is needed. Anemia or abnormalities in the white blood cells or platelets suggests the need for more specific tests. Likewise, screening of serum chemistries is standard; this laboratory testing highlights function of the kidneys, liver, pancreas, and other internal organs. A chest x-ray may also be conducted during screening.

**Specific tests.** Specific tests are ordered if a physician suspects the presence of a malignancy. The most important blood tests indicating the presence of cancer are serologic tumor markers. Blood serology confirms a physician's diagnosis and is useful in monitoring a neoplasm following treatment or to confirm remission.

### *Imaging Studies*

Imaging studies include the use of x-ray, CT (computerized tomography) scans, MRIs (magnetic resonance imaging), PET (positron emission tomography) scans, radionuclide scans, and selective angiography (Cohen & Markman, 2008). These studies identify tumors in specific areas or certain organs, tumor size, and involvement of surrounding tissue. An ultrasound examination may also be used for cancer diagnosis.

## *Interventional Treatments*

At times, simple physical examinations are insufficient. Modern medical instruments allow for direct visualization inside body cavities. Thin, flexible telescopes are introduced into the lungs (bronchoscopy), bladder (cystoscopy), rectum and colon (colonoscopy), esophagus and stomach (esophagogastroduodenoscopy), and the nasopharynx (nasopharyngoscopy). Direct visualization of the bile and pancreatic ducts is termed Endoscopic Retrograde Cholangiopancreatography (ERCP); it is a method for obtaining x-ray pictures of the pancreatic and bile ducts. Instruments that allow for examination of the abdominal cavity are referred to as laparoscopic instruments. A thoroscopy involves the introduction of a visualizing scope into the thoracic cavity (Haskell & Berek, 2001, Raghavan et al., 2006).

## *Biopsy*

Ultimately, a definitive diagnosis of cancer depends on examination of a small amount of tissue which is submitted to a pathologist who performs microscopic evaluation. Special histochemical stains or immunologic markers further categorize a neoplasm. Biopsies are obtained with special needles used to sample tissue from bone marrow, skin, breast, pancreas, and other organs, from the fluid of body cavities (through aspiration), or from tissue removed during a surgical procedure. Pathologic examination is usually the final step in diagnosis prior to the formulation of a treatment plan by the physician. The pathologist's evaluation is crucial in determining origin of the cancer. The four broad cancer classifications are the following (American Cancer Society, 2006).

1. Sarcomas – cancer of the bones, muscles, and connective tissue.
2. Carcinomas – cancer of epithelial cells (such as skin), lining of the lungs, colon, and breasts.



3. Leukemias – cancer of the blood-forming organs.
4. Lymphomas – cancer of the infection-fighting organs.

### ***Staging Classifications***

Once cancer is diagnosed, the physician must determine the extent to which it has spread (metastasized). Staging is a method used to describe if the cancer is localized or has spread to other organs. The most modern staging classification includes the “TNM” system with subscript numbers 0 - 4 (0 = least; 4 = most) by the American Joint Commission on Cancer (American Cancer Society, 2006):

- T** = tumor size, location, and adjacent tissue involvement ( $T_1$  through  $T_4$ ).
- N** = regional lymph node involvement with tumor ( $N_0$  = no tumor involvement with lymph nodes;  $N_1 - N_4$  = increasing degrees of tumor involvement with lymph nodes).
- M** = absence ( $M_0$ ) or degree of spread (metastases) to distant organs ( $M_1 - M_4$ ).

Since “0” denotes no evidence of a tumor or cancer cells, for a cancer diagnosis, the “T” subscript must be at least “1.” The larger the tumor size respective to its site of origin, the higher the subscript number. Lymph nodes (the “N” subscript) draining the cancer site are subsequently examined and assigned a number from 0 to 4, with “0” denoting absence of any localized cancer spread to “4” indicating extensive lymph node involvement. The “M” refers to how numerous and pervasive the non-contiguous cancer growth(s) extend. Thus, a tumor staged  $T_2N_1M_0$  is less extensive and potentially more curable than one staged  $T_1N_2M_1$  (Haskell & Berek, 2001; Rossi, Cady, & Martin, 2000).

An older staging system is still used to a limited extent. In this system, Stage I refers to a state in which the tumor is confined to a single area, usually the tissue of origin. When involvement of adjacent or regional lymph nodes characterizes the case, the cancer is in Stage II. Stage III involves the local spread of cancer beyond the confines of the tissue or organ initially involved. In Stage IV, cancer has metastasized to distant sites of the body.

### ***Functional Capacity Categories***

The patient’s functional capacity is categorized using either the ECOG (Eastern Cooperative Oncology Group) or the Karnofsky scale. Because the ECOG is the most commonly used categorization method, the Karnofsky Scale is not described within this chapter.

ECOG performance scale:

- O** Normal activity without physical limitations.
- I** Symptomatic, but able to complete daily activities.
- II** Assistance needed at times, but can be out of bed more than 50% of the time.
- III** Needs skilled care; in bed more than 50% of the time.
- IV** In bed 100% of the time.

When determining the modality and extent of treatment possibilities, the patient’s age, degree of tumor-associated weight loss, nutritional level, and presence of comorbid conditions (i.e., heart disease, lung disease) are considered. For example, an elderly individual or a malnourished patient who has cancer is at high risk for aggressive surgery or chemotherapy. Individuals at high-risk for functional limitations may benefit from modification of the usual therapeutic approach (Kleinsmith, 2006).

## **Treatment**

Once a definitive diagnosis is obtained, the physician develops and recommends a treatment plan. Major modalities for cancer treatment are surgery, chemotherapy (pharmacological management), radiation, and biologic therapy. These methods are employed alone or in combination, depending on the nature of the malignancy and stage of the tumor. Bone marrow and stem cell transplantation are employed in selected circumstances. The treatment modality chosen depends primarily on the stage of the neoplasm, the degree to which the neoplasm impacts the patient’s health and ability to function, and overall health.

## ***Surgery***

Surgery is the oldest form (and the most effective) of cancer treatment. More cancers are cured with successful surgical interventions than with any other treatment modality. Surgery is often curative and is the primary treatment approach for several types of cancer. These include but are not limited to: stomach, colon, head and neck, prostate, sarcoma, skin, certain lung cancers, and thyroid cancer. Surgical procedures occur during the diagnosis, treatment, and post-treatment phases of cancer management and in the following specific circumstances:

- To diagnose or stage the disease, either with a biopsy or an open operation with removal of nonessential organ parts.
- To remove the primary tumor. This procedure may be curative or used to improve the effectiveness of other treatments by reducing tumor volume.
- To remove other tumors that have escaped prior treatment or are due to recurrent disease.
- To relieve symptoms. Used when the tumor blocks or compresses vital organ structures, such as occurs when a cancer obstructs the colon, stomach, or bile passages.
- To reconstruct or rehabilitate. Examples include reconstructive surgery following mastectomy, removal of cancer of the head and neck, and extensive limb surgery.
- To support radiation and chemotherapy. Catheters loaded with radiation implants are surgically placed in body cavities or organs to access deep-seated cancers when curative treatment with external beam radiation alone is not adequate (e.g., cervical and prostate cancer or to support other forms of cancer treatment). Treatment includes implantation of a venous access port to facilitate delivery of concentrated chemotherapy, such as placement of a hepatic artery catheter to allow for delivery of concentrated chemotherapy directly into the liver.

## ***Radiation Therapy***

About half of all cancer patients require radiation during treatment. This modality is used alone or in combination with chemotherapy, surgery, or both. The main goal is cure or relief of major symptoms.

Radiation therapy employs high-energy x-rays, electron beams, and radioactive isotopes to destroy cancer cells without exceeding safe doses to normal tissue. Malignant cells are destroyed through a process called ionization, which damages the DNA in the nuclei of the cells so that cell division is abrogated. There are several ways of delivering radiation, including:

### ***External beam radiation***

This treatment involves delivery of a source from outside the body; the delivery varies according to the photon energy of the machine used and types of beams produced (electrons, x-rays, or gamma rays). Generally, the equipment emits either low energy (orthovoltage) or high energy (megavoltage) beams. The “gamma knife” is a very high energy apparatus capable of focused, intense, but brief treatment to the target area using stereotactic techniques.

### ***Internal radiation***

Internal radiation involves use of interstitial radiation, radiation implants, or brachytherapy. Radioactive seeds are placed temporarily or permanently into cancer of the prostate, head and neck, and other organs. Intracavity radiation is used primarily for tumors of the cervix, uterus, and esophagus. Special hollow applicators are implanted within the body (where the applicator usually remains for two to three days) and are followed by insertion of a radioactive isotope of selected strength.

### ***Radioimmunocojugates (experimental)***

A radioisotope is bound to an antibody or glycoprotein targeting a specific receptor on the cancer cell. After the radioimmunocojugates is attached, dissolution of the cancer cells occur.

### ***Hyperthermia***

Used infrequently, hyperthermia is based on the knowledge that malignant tumors are more susceptible to destruction by heat than normal tissues.

Common side effects of radiation include nausea, fatigue, skin inflammation, and temporary lowering of blood counts. Specific side effects characterize particular cancer types. If radiation is given to the brain, for example, hair loss occurs. After treatment to the head and neck, the patient may experience dry mouth. Diarrhea is a side effect of rectal and bladder therapy, while frequent urination often occurs after prostate radiation. Neoplasms that are treated by radiation with a curative intent include prostate, head and neck, rectum, Hodgkin's lymphoma, and certain lung cancers. Most people who undergo radiation also receive palliative therapy (Tannock & Hill, 2005).

### ***Chemotherapy***

Chemotherapy is another form of treatment that involves the use of medications to interrupt cancer cell growth by attacking specific stages of cell division. These drugs are often administered in combination, enhancing the efficacy by promoting the interruption of cell division at multiple sites. A major obstacle for such treatments is the eventual development of drug resistance. Many cancers acquire the ability to overcome or evade the toxic effects of pharmaceuticals.

Delivery routes and methods for administration of chemotherapy include (a) intravenous (the most common route); (b) ambulatory infusion pump (employed for long-duration chemotherapy); (c) intra-arterial infusions (used for deep-seated localized tumors, found in liver cancer); and (d) intracavitary (used for some neoplasms of the chest cavity lining and ovaries). Since chemotherapeutic agents destroy rapidly growing abnormal tissues, rapidly growing normal tissue such as hair follicles, gut mucosae, and bone marrow are often affected. Common effects of chemotherapy include nausea, diarrhea, hair loss, sore mouth, and compromised blood count. The effects on normal tissue are often temporary.

Chemotherapy is used as a curative treatment for some malignancies and also for palliative reasons. Adjuvant treatment is increasingly employed and involves the use of drugs to eradicate microscopic or subclinical areas of cancer that may remain following surgery, or in high-risk patients who receive concomitant radiation, surgery, or both. Tumors for which a cure may be possible or when the patient's survival is substantially enhanced by chemotherapy include: (a) advanced diseases such as choriocarcinoma, acute lymphocytic leukemia, Hodgkin's lymphoma, aggressive non-Hodgkin's lymphoma, germ cell cancer, acute myelogenous leukemia, childhood cancers, small cell lung cancers, and ovarian cancer; and (b) adjuvant settings for breast cancer, sarcoma, colon cancer, and rectal cancer (Armitage & Antman, 2008).

### ***Biologic Therapy***

Biologic therapies are used to treat cancer by aiding the functioning of the immune system. They are also referred to as biologic agents, biological, biological response modifier therapy, BRMs, and immunotherapy. Biologic therapy works with the immune system, helping to fight cancer or controlling its side effects or those resulting from other cancer treatments by using the body's immune system to facilitate the destruction of cancer cells (Tannock & Hill, 2005, Raghavan et al., 2006).

While biologic therapy and chemotherapy may seem alike, these treatments work in different ways. Biologic therapy which helps the body's immune system fight cancer differs from chemotherapy which attacks the cancer cells directly. Researchers speculate that biologic therapy aids the immune system in its fight against cancer in the following ways:

- Stops or slows the growth of cancer cells.
- Makes it easier for the immune system to destroy, or get rid of, cancer cells.
- Keeps cancer from spreading to other parts of the body.

Biologic therapy is a particularly exciting and innovative area encompassing efforts to manipulate the human immune system to deter the progression of malignancy or to induce regression. By utilizing new technologies, such as recombinant DNA methodology, researchers have been able to produce large

quantities of biologic substances normally present in the body in only minute amounts. Most of these agents are present in lymphocytes or macrophages and include certain glycoproteins like interferon or cytokines such as tumor necrosis factor (Haskell & Berek, 2001).

Biologic response modifiers used for cancer treatment include interferon, interleukin-2, and monoclonal antibodies; fusion proteins have recently been developed (El-Deiry, 2003). Routes of delivery are subcutaneous and intravenous. Neoplasms currently managed with these agents include: interferon (kidney cancer, multiple myelomas, some lymphomas, Kaposi's sarcoma, chronic myelocytic leukemia, and hairy cell leukemia); interleukin-2 (kidney cancer); and monoclonal antibodies (breast cancer, small cell lymphoma, and peripheral T-cell lymphoma). Other neoplasms treated with biologic therapy include: cis retinoic acid (acute promyelocytic leukemia); cancer vaccines (trials ongoing for prostate, breast, colon, and skin cancer); and activated lymphocyte treatment (kidney cancer, melanomas, and brain cancer) (Tannock & Hill, 2005).

### ***Transplantation***

Bone marrow transplants are performed to treat illness in which the stem cells are defective or preclude the production of normal cells. Bone marrow contains immature (stem) cells capable of continuously producing new cells, including white and red cells and platelets. Bone marrow transplantation is recommended to replace a defective bone marrow or immune system. Many cancers cannot be cured with conventional doses of radiation, however, and receive high doses of chemical and pharmacological treatment. Although these treatments might be curative, they irreversibly damage bone marrow, the most sensitive of all tissues to chemotherapy and radiation.

#### ***Types of Transplants***

**Allogenic.** Allogenic transplants involve the transfer of marrow from one individual to another. Donors are matched immunologically as close as possible using a process known as HLA (human leukocyte antigen) typing. The closer the match, the less likely immune incompatibility, or "graft versus host disease," will occur.

**Autologous.** The patient's own marrow is removed and the stem cells are harvested and cryopreserved. Marrow or stem cells are then reintroduced after high-dose radiation or chemotherapy has ablated the old marrow. There is no risk of marrow rejection with this technique.

**Syngeneic.** Bone marrow is obtained from a person's identical twin. There is no risk of rejection.

#### ***Complications***

After receiving a bone marrow transplant, individuals are at risk for infection (Kleinsmith, 2006). The greatest window of susceptibility occurs between marrow ablation and repopulation of the bone marrow spaces by the transplanted marrow. Some people experience excessive bleeding after a transplant, while others develop pneumonia, usually caused by cytomegalovirus infection. Complications associated with marrow grafts occur when new marrow recognizes the host as foreign and begins attacking various tissues including the liver, skin, and intestines. Medication ameliorates this problem. Heart damage is an additional potential obstacle; delayed complications of bone marrow transplantation treatments include chronic graft versus host disease, cataracts, and sterility (Rossi et al., 2000).

### ***Current Treatment Rates***

America's youth have benefited the most from the nation's investment in cancer research. Twenty-five years ago, only one child in ten (10%) survived cancer. Today, seven out of ten children (70%) who develop cancer are living five years after diagnosis, and one of every 1,000 Americans between the ages of 20 and 40 is a survivor of childhood cancer.

In the past, testicular cancer (the leading cause of death due to cancer among men 15 to 35 years of age) almost always constituted a fatal diagnosis. Today, about 90% of men who are diagnosed with testicular cancer are cured. Likewise, the breast cancer death rate of women in the United States fell 6.3% between 1991 and 1995 and 9.3% in women under the age of 65. This drop in death rate is attributed to improvements in consumer awareness, modern testing methods, mammography, and modern treatments (National Cancer

Institute, 2008). The primary focus of cancer research for the past 30 years has been on cancer prevention, early detection, treatment, coping, and rehabilitation services.

### ***Negative Effects of Cancer Treatment***

While an individual is undergoing treatment, the focus intent is on improving one's health. Returning to "normal" means resuming relationships with family, friends, and co-workers. Underneath these adjustments linger many doubts (i.e., Was the treatment effective? What unexpected negative effects might show up after treatment? What if the cancer returns?).

Negative effects of cancer treatment vary for each individual; everyone experiences symptoms of varying intensity (El-Deiry, 2003). The most common physical changes and negative effects experienced from cancer treatment include: hair loss (including eyebrows and eyelashes), weight loss or gain, appetite loss or increase, fatigue, disfigurement from surgery, lack of concentration, nausea and vomiting, diarrhea, pale skin and changes in skin tone, sleep disruption, and sexual dysfunction.

For many people diagnosed with cancer, the most prominent and challenging negative effect is fatigue, an effect of chemotherapy that can be overwhelming and persistent. Chemotherapy can involve months of strong medications that cause this fatigue as the body tries to negotiate the healing process while regenerating the affected tissue. Family, friends, and co-workers of the individual may experience indirect stress and emotional involvement as a result of the individual's emotional changes, which in turn further impacts the person experiencing these changes (Raghavan et al., 2006).

## **Psychosocial Implications – Stress & Adjustment Reactions**

The diagnosis and treatment of cancer is a stressful event followed by a range of distressing symptoms including anxiety and depression. While these symptoms are likely to be transient, the experience of cancer is not a single, undifferentiated event. Rather, people with cancer encounter a series of stressful events and challenges over time, posing different demands and difficulties. A person's distress may become heightened at particular times, such as at the time of diagnosis or disease recurrence, before surgery, at the commencement of adjuvant therapy, during the advanced disease stage, or when presenting for medical surveillance (Mystakidou, 2006).

### ***Severe Emotional Distress***

Although the emotional distress experienced by people with cancer may be occasional and time-limited, some experience more severe emotional reactions. Stress and adjustment problems may include negative feelings lasting for one or more weeks. If such problems are not acknowledged, they may develop into more serious emotional states. Major psychological disorders resulting from emotional distress include major depressive episodes; anxiety disorders; post-traumatic stress disorders (PTSD); and negative emotional, behavioral, or cognitive states that are overwhelming and persistent (lasting more than two weeks) (Oeffinger & McCabe, 2006).

### ***Anxiety***

Some anxiety in response to a stressful life event is normal; severe cases can interfere with relationships, social or occupational functioning, and disrupt health-related behaviors. Symptoms associated with anxiety include heightened physical arousal, sleep disturbance, impaired concentration and decision-making, agitation, and anger. Avoidance of distressing issues and situations, and excessive reassurance-seeking, may indicate anxiety problems. These responses have a major impact on the person's functioning and that of the family. Severe anxiety problems include panic attacks, pervasive and generalized worry, treatment phobias, social anxiety, and post-traumatic stress reactions.

### ***Depression***

In addition to the personal suffering it causes, depression is a major risk factor for suicide. Depression undermines the capacity of the person to cope with illness, and is associated with increased severity of medical symptoms and additive impairment in social, educational, and vocational functioning (Livneh, 2000). It is therefore not surprising that depression is associated with increased health care costs in



those with medical illness. Depression may compromise the physical care of the person with cancer; in medical settings, patients with depression are three times more likely to be non-compliant with medical treatment recommendations. Depressed patients with breast cancer, for example, have been found to be less likely to accept adjuvant chemotherapy (Wimberly, Carver, & Laurenceau, 2005).

A diagnosis of a major depressive episode in patients is best evaluated by the severity of the depressed mood; loss of interest and pleasure; the degree of feelings of hopelessness, guilt, and worthlessness; and the presence of suicidal thoughts (Mystakidou, 2006). Recurrent tearfulness is often accompanied by social withdrawal and loss of motivation. Patients with depression may feel they are unable to control their negative feelings and as a result, these feelings begin to dominate one's life.

### ***Suicide***

Accurate figures of the incidence of suicide in patients with cancer are difficult to obtain. Studies undertaken in Sweden suggested that the incidence of suicide is higher in people with cancer than in the general population, and that suicide is more likely to occur in the first year after diagnosis (Jacobsen et al., 2004).

Risk factors for suicide include severe depression, a family history of suicide, past history of self-harm, alcohol or other substance abuse, poor social support, and a negative outlook for the future (Mystakidou, 2006). Assessment and exploration of suicidal thoughts is crucial to early recognition and treatment of psychological distress for the patient (Jacobsen et al., 2004).

### ***Post-traumatic Stress Disorder (PTSD)***

Diagnostic classifications of mental disorders have been revised to include diagnosis of a potentially life-threatening illness as a sufficient stressor for the precipitation of PTSD. There is limited but increasing research into traumatic symptomatology in patients with cancer, although most of this research has focused on patients with breast cancer (Oeffinger & McCabe, 2006).

### ***Sexuality Issues***

Cancer often causes changes in physical appearance as a result of surgery, chemotherapy, and radiation therapy, and the negative effects of various cancer treatments. In addition, psychosocial responses, including grief, depression, and anxiety occur frequently with a life-threatening diagnosis. Furthermore, challenges in communication arise around issues of life changes induced by the diagnosis and treatment of cancer, as well as the threat of its recurrence.

Sexuality encompasses body image, self-esteem, mood, support, intimacy, and a sense of emotional connection. The role of body image in sexuality is illustrated by the number of patients reporting sexual problems whose cancers do not directly affect sexual organs (Livneh, 2000). This finding evidences the need to address patients' issues of sexuality regardless of the cancer site.

As a topic in counseling, sexuality has received little scholarly attention. Professional training in sexual health is equally limited. Although the literature demonstrates the integral role sexuality plays in the life of patients, physicians rarely introduce the subject during clinical encounters (Houldin, 2000; Wimberly et al., 2005). Patients who have chronic diseases find that because of the complexity of their illnesses and medical treatments, concerns regarding sexuality are often left unaddressed as treatments are given priority. Without prompting on the part of the counselor or physician, patients are reluctant to voice sexual concerns (Huddart et al., 2005). Yet, it is often these patients with chronic illnesses who have difficulties with sexual functioning (Orange, 2002). By incorporating basic management strategies with an understanding of the impact of chronic illness on sexual functioning, health care professionals can more readily screen for and manage sexual dysfunction and thereby enhance quality of life.

### ***Emotional Adjustment***

The psychosocial aspects of cancer including dependence, stigma, discrimination, and personal self-concept, can have effects equal to the physical components of the disease. To achieve physical, mental, and emotional health, individuals need time to adjust. The acute stress of illness and treatment can result in distress and dysfunction in all areas of life.



The recurrence of physical symptoms signals the reactivation of those psychological associations that accompanied the initial diagnosis and treatment. As a form of negative information, these symptoms constitute the resurgence of feelings of stress, helplessness, fear, and pain sometimes leading to depression and withdrawal (Brodwin, Orange, & Chen, 2004). This can cause serious adjustment problems when left untreated. Depression is a reaction to the experience of loss, chronic frustration, and radical changes in life-style. Psychological and social supports function as stress buffers, helping a person modulate the somatic effects of stress (Livneh, 2000).

Rehabilitation researchers and counselors have long recognized the benefits of identifying and addressing the psychosocial needs of the client who currently has or is recovering from cancer (Karthaus & Ganser, 2000). Cancer is unpredictable; those with the disease experience symptoms which vary on a daily basis. To address and adapt to this unpredictability, individuals and health-care professionals should focus on the following psychosocial issues during the rehabilitation process: (a) feelings of loss of control during treatment; (b) difficulty coping with physical changes, attended by thought processes and moods resulting from the disease and medical treatment; (c) need for social support within the family and community; (d) impact and consequences of role changes within the family and need for both internal and external resources to aid adjustment to these role changes; (e) interpersonal issues associated with stress; and (f) psychosocial factors related to both unemployment and a potential return to work (Livneh, 2000).

## **Rehabilitation Potential and Employment Issues**

### ***Challenges***

Work is a vital aspect of one's conceptualization of individual self-worth and identity. While many persons who have had cancer view a return to work as the apex of achievement, those resuming employment face many challenges including discrimination, avoidance, financial concerns, reduced opportunities for advancement, and changes in physical (and possibly emotional) capacity.

Many people return to employment during or after experiencing illness and medical treatment. Cancer survivors benefit from the assurance that they are able to make contributions within the workplace. An essential component of rehabilitation is support from the employer, including managers, supervisors, and co-workers. Employers know it is good company policy to assist capable and valued employees (McDonough, 1992). Unfortunately, cancer survivors face problems when employers have misconceptions about the consequences of cancer and medical treatment.

Together, survivors of cancer and vocational rehabilitation counselors can dispel the following societal myths about the disease: (a) health insurance rates for employee coverage increase when individuals who have cancer are hired; (b) attendance by these workers is substandard and job turnover is high; (c) employees who have cancer are less flexible; and (d) cancer survivors are less able to perform essential job functions, thereby imposing more duties on other employees and entailing added costs to employers (Slivon, Hicken, & Marini, 1994). These myths continue to have a negative impact in society.

### ***To Disclose or Not to Disclose***

Negative attitudes, stereotypes, and fears about people with cancer form major hurdles for cancer survivors desiring full participation in the work place. Whether or not to disclose a disability or functional limitation to a potential employer is a difficult decision. Even with the influential federal legislation incorporated within the Americans with Disabilities Act (ADA) of 1990, it is not always clear if the individual should disclose the presence of a disability. The most appropriate response depends on personal choice and the functional limitations that occurred. If a disability affects a worker's performance on the job, or if accommodation is necessary to perform the duties of the position, disclosure is potentially beneficial. The ADA states that an employer is not entitled to information about a disability unless it directly relates to (a) ability to perform the job, and (b) a request for reasonable accommodation.

### ***Reasonable Accommodation and the Americans with Disabilities Act***

The ADA provides protection for individuals with a history of cancer, regardless of whether the cancer is cured, in remission, or being treated. According to the ADA, people with limitations due to past or

current disabilities are to be given the same opportunities as everyone else. Reasonable accommodations under the statute include: modifications or adjustments to a job or work environment that enable a qualified applicant with a disability to participate in the application process, perform essential job functions, and enjoy the rights and privileges available to other employees. Accommodations may be made by the employer to allow a qualified employee with cancer to perform the essential functions of the job. An employer is not required to consider providing accommodations under the ADA, however, unless the employee has disclosed that a disability exists.

### ***Empowerment and Employment***

Employment is a significant component of identity building and one's conception of self-worth. Individuals who have been diagnosed with a disease like cancer may place increased emphasis on work; it allows a focus on capabilities, rather than on illness and limitations. Work thus becomes a haven away from medical intervention, providing routine as well as psychosocial stability, familiarity, potential for reward, and a source of companionship. Being around people who are supportive is not only a source of great comfort but acts as an alternative to the isolating effects of cancer, and helps the individual surviving cancer maintain a sense of autonomy and control (Simon, 1999).

Counselors facilitate empowerment by encouraging and supporting a person's efforts to take an active role in the rehabilitation process. Determining the client's knowledge and awareness level with respect to disease state and prognoses helps clarify directions for inquiry. People who survive cancer and understand their disability and its implications are better able to actively participate in decision-making during all phases of medical treatment and rehabilitation (Brodwin, Orange, & Brodwin, 1994).

## **Multicultural Perspectives**

Americans with disabilities face varying societal beliefs, feelings, and behaviors. As noted by McDonough (1992), "Americans in society have a negative attitude about the disabled in general and those in the workplace in particular" (p. 58). Persons who possess attributes that are viewed negatively by the prevailing group in a society tend to be devalued by that society; individuals with disabilities often are evaluated and judged by the disability. Rehabilitation literature emphasizes the maintenance of a strong positive regard and advocacy for people who have disabilities (Brodwin et al., 1994).

Rehabilitation helps create positive behavioral changes in individuals with disabilities, allowing them to maximize their capabilities while minimizing limitations. As a strategy, rehabilitation works to maximize the potential for independent living. Rehabilitation philosophy emphasizes a multidisciplinary approach to guide people with disabilities to their pre-injury potentials of physical, mental, social, educational, vocational, and recreational status.

Competent counselors put their awareness and understanding of the complexities of the multicultural society in which they live into practice. If quality counseling services are to be provided to ethnic minority persons with disabilities, rehabilitation counselors and other human service professionals must examine the issues involved in interactions with culturally diverse individuals. They need an understanding and sensitivity to people of all cultures and beliefs.

To be effective, rehabilitation counselors address the issues, needs, and beliefs of a culturally diverse population and maintain respect and positive attitudes toward cultural and gender differences. Counselors need to rehabilitate the "whole person" and dedicate their efforts to the end goal of increasing the quality of life for all individuals, including survivors of cancer.

## **Case Study**

Cancer of the testes constitutes only 1% of male cancers but accounts for more deaths in males 15-34 years of age than any other cancer (Huddart et al., 2005). This case addresses this type of cancer.

Steve McPherson is married, 22 years of age, and the father of one child. Although he was a high school dropout, he attended adult education and successfully completed his G.E.D. (General [high school] Equivalency Diploma). Steve is a partner in a small, but highly successful company involved with telephone

and communication systems installation. By trade, he is an electrician and a member of the local electricians' trade union.

The *Dictionary of Occupational Titles* (U. S. Department of Labor, 1991) classifies Mr. McPherson's job as telephone electrician (telephone and telegraph) (*D.O.T.* # 822.281-018). Steve and his business partner install, test, and repair telephone and communication systems. They update and expand old equipment, install new computerized systems, and wire burglar alarm devices and related equipment. The work involves the use of hand tools and testing devices, an ability to read schematics, and knowledge of electrical and electronic principles. Lifting and carrying on the job involves a maximum of 50 pounds, with repetitive lifting up to 25 pounds.

Mr. McPherson felt in good physical and emotional health when he noticed a non-tender lump on his right scrotum. Two months later, he was diagnosed with embryonic cell carcinoma of testicular origin, well-differentiated, stage T<sub>2</sub>N<sub>0</sub>M<sub>0</sub>. Steve underwent a right orchiectomy (testicle removal) and lymph node dissection and received a complete course of chemotherapy. Three months postoperatively, he was assessed as being disease-free with an excellent prognosis. Steve returned to work, continued to have an active family life, and maintained sexual function.

Approximately one year later, a routine cancer follow-up visit (including a chest x-ray) revealed a solitary metastatic right lower lobe pulmonary lesion. His oncologist initiated chemotherapy using a combination of agents; the lesion disappeared. Again, the oncologist believed him disease-free or "in remission." After completion of chemotherapy, Steve returned to work and was fully independent. Fortunately, his business partner had managed to cover necessary work activities during his absences by employing other union members on a temporary basis.

One year later, Mr. McPherson was diagnosed with lung cancer. Within one month of open chest surgery, Steve developed left-sided brain seizures. After a new diagnosis which identified metastatic testicular disease of the brain, he underwent neurosurgery for removal of the brain tumor.

After undergoing the craniotomy procedure, Steve again developed left-sided weakness in the form of significant loss of strength in his left arm and leg. He not only required crutches for ambulation, but suffered from poor balance and was required to be in bed or in a chair most of the day.

Following radiation therapy, Steve received physical and occupational therapy. He maintained a 25% residual deficit in his left, nondominant upper extremity and could walk for short distances with a cane. The symptoms gradually subsided and he was able to return to work part-time at a functional capacity of sedentary work. His major responsibilities became administrative, including work site analysis, writing bids, and service contracts negotiation. Mr. McPherson remained asymptomatic and was able to gradually increase his work effort to full-time at the sedentary level of exertion.

### **Questions**

1. Discuss the various forms of cancer treatment.
2. Identify psychosocial implications of cancer.
3. Discuss the staging process and the use of the "TNM" subscripts. Early in diagnosis, Steve's cancer was thought to be at stage T<sub>2</sub>N<sub>0</sub>M<sub>0</sub>. From what you now know, was this accurate?
4. Why was radiation therapy not given to his lung nodules?
5. What were some precautions for him to take in returning to work?
6. Give a vocational profile including age, educational level, exertional and skill level of work activity, occupationally significant characteristics, and transferable skills (if any).
7. Was the return to modified work realistic? How were transferable skills used in this return to work? What other vocational possibilities are there using transferable skills or additional training?

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## Chapter 5

# DIABETES MELLITUS AND RENAL DISEASE

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## Introduction

Persons with diabetes mellitus (commonly called diabetes) account for nearly half of the cases of renal disease. It is for this reason we have combined the material on diabetes and renal failure. Individuals who do not have diabetes but have renal disease are also included within the section on renal disease. The first part of the chapter discusses diabetes. The second part of the chapter includes renal disease and renal failure.

## Diabetes Mellitus

Diabetes mellitus (diabetes), popularly referred to as “sugar diabetes,” is a health concern of worldwide proportion. It is estimated that 21 million children and adults in the United States have diabetes and a significant number of people have undiagnosed diabetes, making the disease one of the most common in the world. By the year 2025, it is estimated that 300 million people worldwide will be diagnosed with diabetes (King, Aubert, & Herman, 1998). Diabetes is recognized as the 6<sup>th</sup> leading cause of death in the United States.

A chronic disorder, diabetes is characterized by abnormal elevations in blood glucose (blood sugar) resulting from defects in insulin secretion, insulin action, or both (American Diabetes Association, 2006a). Along with abnormal blood glucose, diabetes is associated with a set of physical abnormalities collectively referred to as diabetes complications. It is these complications that are responsible for the mortality and morbidity associated with diabetes.

### *Risk Factors*

The four major risk factors for diabetes development are genetics, age, obesity, and race. A positive family history of the disease increases the potential risk of developing diabetes (American Diabetes Association, 2006a). Aging also increases one’s risk of developing diabetes, as glucose tolerance, the ability of the body to metabolize sugar, deteriorates with the aging process. Obesity induces insulin resistance; overweight individuals are more likely to develop diabetes. Several other uncontrollable factors contribute to the risk of disease development. Diabetes is more common among Hispanics than any other racial group, while its prevalence ranks second among African Americans; Caucasians have the least risk.

### *Diagnosis*

In the current American Diabetes Association criteria for diagnosis of diabetes, blood glucose values are a vital determining factor (American Diabetes Association, 2006b). A fasting blood glucose greater than 126 mg/dl (milligrams per deciliter) is considered abnormal. When blood glucose is greater than 200mg/dl and is concurrent with symptoms of diabetes, such as polyuria (increased urination), polydipsia (increased thirst), and unexplained weight loss, a physician may diagnose an individual with diabetes.

### ***Type I And Type II Diabetes***

There are two major forms of diabetes. Type I (Insulin-Dependent Diabetes Mellitus) diabetes results from autoimmune destruction of the insulin-producing cells of the pancreas ( $\beta$ -cells). This type is characterized by absolute insulin insufficiency. This was formerly referred to as juvenile-onset diabetes, brittle diabetes, or ketosis-prone diabetes. Type I diabetes accounts for 5-10% of the cases of diabetes and most patients manifest severe symptoms resulting from little or no insulin secretion. Even though primarily a disease with onset in childhood or adolescence, it can occur at any age.

Type II (Non-Insulin-Dependent Diabetes Mellitus) diabetes is the most common form of the disease (90-95% of the cases of diabetes) and is associated with defects in insulin secretion and/or insulin resistance at the tissue level. Type II diabetes, formerly referred to as maturity-onset diabetes or non-ketosis prone diabetes, is found especially in overweight individuals as obesity itself causes some degree of insulin resistance. Type II diabetes can remain undiagnosed over many years because insulin resistance and elevated blood sugar occurs gradually. Over time, Type II patients may require oral agents to lower their elevated blood sugar and ultimately insulin supplementation therapy.

Gestational diabetes (GDM) occurs when glucose intolerance occurs during pregnancy. The term is used regardless of whether insulin or only dietary modification is used for treatment (American Diabetes Association, 2006a). GDM complicates about 4% of all pregnancies in the United States.

### ***Complications***

Both types of diabetes develop similar complications, as discussed below.

#### ***Peripheral Neuropathy***

Patients with diabetes frequently exhibit abnormal nerve function, especially in the lower extremities. Common symptoms of this complication are painful sensations in the feet, known as paresthesias, such as feelings of burning or tingling. These symptoms typically worsen at night resulting in temporary numbness (loss of normal sensation). The individual may experience difficulty with muscular function at various locations within the body. Because of decreased feeling in the lower extremities, these areas of the body are more susceptible to trauma. Such trauma may lead to foot ulcers, infection, and ultimately lower extremity amputation. The cause of diabetic neuropathy is unknown; theories include decreased blood supply and abnormal chemical metabolism of the involved nerves (Adler, Boyko, Ahroni, & Smith, 1999; Marrero & Guare, 2005).

#### ***Peripheral Vascular Insufficiency (PVI)***

PVI involves narrowing and/or occlusions of arteries, especially in the lower extremities of patients with diabetes. When larger blood vessels are involved, the disorder is referred to as macro-vascular disease, while occlusion of the smaller blood vessels at the tissue level is called micro-vascular disease. In PVI, the patient experiences a significant decrease in blood flow, precluding the delivery of nutrients and oxygen beyond the areas of occlusion. Peripheral vascular insufficiency is a major contributing factor in development of foot ulcers, infection, gangrene, and subsequent amputation in patients with diabetes (Unger, 2007).

#### ***Combined Peripheral Vascular Insufficiency and Peripheral Neuropathy***

In combined peripheral vascular insufficiency and peripheral neuropathy, the lower extremities of the individual with diabetes become affected by both conditions. This potentially lethal coexistence causes the involved lower extremity to be particularly vulnerable to further problems (Izumi, Satterfield, Lee, & Harkless, 2006). The loss of protective sensations in a limb leads to trauma, resulting in foot ulcers and subsequent infection, as described previously. Peripheral vascular insufficiency slows healing of such lesions and makes therapy a difficult process. The end result is frequently amputation. In the United States, diabetes is the leading cause of lower extremity amputation. To help prevent such complications, major emphasis is currently being placed on patient education.

***Diabetic Retinopathy***

Diabetic retinopathy is the leading cause of new cases of blindness in adults annually (American Diabetes Association, 2006a). In diabetic retinopathy, the small arteries in the retina of the eye develop lesions known as micro-aneurysms which become weak and are subject to bleeding. These lesions often lead to hemorrhaging, which can severely interfere with vision. Some methods have been developed to limit the retinal damage and save vision.

***Diabetic Nephropathy/Renal Failure***

Diabetic nephropathy is a progressive kidney disease, usually occurring about 15 years after the onset of diabetes, and is the primary cause of kidney dialysis. It is caused by diabetes-induced abnormalities within the capillaries in the kidney glomeruli, which filters urine from the blood vessels into the collecting ducts of the kidneys. This abnormality interferes with normal kidney function and causes abnormal loss of protein in the urine (nephritic syndrome) and buildup of metabolic waste products in the blood. Diabetic nephropathy is usually progressive and may result in ultimate end-stage renal disease and renal failure. About 44% of new cases of renal disease are linked to diabetes (American Diabetes Association, 2006a). Dialysis and renal transplantation are therapy options; renal disease is described in greater detail later in this chapter.

***Arteriosclerosis***

Arteriosclerosis, also known as atherosclerosis, or “hardening of the arteries,” is not a specific complication of diabetes, but is often seen at an early age in patients with diabetes. This condition may develop throughout the vascular system while, in some cases, it may manifest only in specific areas, such as the lower extremities or heart.

***Cardiovascular Disease***

In addition to arteriosclerosis, the heart is subject to many complications in patients with diabetes. Although not a specific complication of diabetes, the high association between diabetes and development of cardiovascular disease is evident. The risk of cerebral vascular accidents (stroke) also is much greater. Cardiovascular events, such as myocardial infarction (heart attack) and strokes are recognized as the leading cause of death among patients with diabetes (McEwen et al., 2006). An estimated 73% of adults with diabetes have high blood pressure, hypotension, or hypertension. Diabetes is associated with abnormal elevations in blood lipids (fats), such as cholesterol and triglycerides. Physicians of patients with diabetes stress management of blood glucose, blood pressure, weight, and avoidance of smoking.

When complications develop in a person with diabetes, the rehabilitation counselor can provide services to help prevent such complications from interfering with a patient’s daily life. If a person with diabetes is able to manage blood glucose and is free of complications, it is unlikely that contact with a rehabilitation counselor will occur. Individuals who have difficulty controlling diabetes, or who have complications such as lower extremity amputation, visual loss, kidney failure, or severe peripheral neuropathy may seek the guidance of a rehabilitation counselor (Marrero & Guare, 2005). The counselor may help the patient learn to manage blood glucose levels, provide support, and aid patients with social and vocational adjustment issues.

***Lower Extremity Amputation***

Lower extremity amputation is, unfortunately, a frequent occurrence. Risk of amputation is twice as likely in individuals with diabetes as in persons without diabetes (American Diabetes Association, 2006a; Izumi et al., 2006). The frequent co-existence of peripheral neuropathy and peripheral vascular insufficiency increase vulnerability to trauma as numbness makes it difficult to feel pain. If the skin is broken during a trauma and not treated, severe infection may follow. Peripheral vascular insufficiency makes therapy of foot infections slow and difficult. Local spread of infection, which often occurs in soft tissue, is known as cellulites (inflammation of the cells of the skin). Osteomyelitis, infection in the underlying bone, may occur. Spread of infection via the blood, known as septicemia, can be life threatening. In such a scenario, amputation may be the only viable therapy as early medical intervention is necessary to prevent further complications, such as gangrene in an area without blood supply. Before serious infection develops, medical treatment is essential (Adler et al., 1999).

### ***Vision Loss***

A plethora of new cases of blindness in adults is recognized annually in this country (American Diabetes Association, 2006a). Diabetes is a major contributing factor to these cases. Diabetic retinopathy involves bleeding from micro-aneurysms at weakened bulges in the retinal arteries. These hemorrhages interfere with vision. Prompt laser therapy prevents extensive retinal damage. To decrease the frequency of vision loss, especially in the case of diabetic retinopathy, a patient needs careful management of blood glucose. Such management has been clinically shown to limit these complications.

### ***Blood Glucose Control and Development of Complications of Diabetes***

Prior to the discovery of insulin in 1922, patients with insulin-dependent diabetes had limited life spans. With the advent of insulin therapy, patients with diabetes enjoyed longer lives but now had to address the complications associated with diabetes. Recent medical literature has established the concept that blood sugar control and complication development are inter-related. Landmark clinical studies, such as the Diabetes Control and Complications Trial conclusively demonstrated this relationship in insulin dependent patients (Lasker, 1993). Results from these studies were later extrapolated to include non-insulin dependent patients and the United Kingdom Prospective Diabetes Trial confirmed that a relationship existed between blood glucose control and development of complications in Type II patients as well (American Diabetes Association, 1998). This study also showed that improved blood glucose control was associated with significant reduction in the risk of complication development. Education is clearly the most worthwhile method with which to treat, manage, and prevent or delay complications (American Diabetes Association, 2006c).

### ***Therapy of Diabetes***

Modern therapy offers various management strategies designed to control blood glucose levels and thus minimize development of long-term complications. Simple in theory but difficult in practice, diabetes requires four important variables to be carefully balanced. These variables are diet, exercise, medication, and careful management of blood sugar levels through blood sugar testing. The patient with diabetes is committed to continuous involvement in daily management of his or her condition. Patient education and knowledge is important because, aside from contact with a physician or other health care practitioner, health care of diabetes is primarily self-care (Falvo, 2009; Marrero & Guare, 2005).

#### ***Diet***

The first important variable is diet. Adherence to a healthy food plan is mandatory for all patients with diabetes and is a cornerstone of therapy. Yet, diet is also the single most difficult aspect of therapy to teach and learn. Dietary regulation may be all that is required for some Type II patients to control their diabetes, especially those who are significantly overweight. Each patient with diabetes should have a specific dietary program prescribed by a knowledgeable professional. The total number of calories and the proportions of carbohydrate, protein, and fat that make up those calories, along with their intake and distribution throughout the day, are all crucial factors.

#### ***Exercise***

Exercise can help lower blood glucose levels and may be of benefit in maintaining peripheral arterial circulation. However, variability of physical exercise when it is not maintained on a daily basis, may contribute to development of labile blood glucose and hypoglycemic episodes, especially in a patient using insulin. Patients with Type II diabetes, who tend to be older, may be limited in their exercise tolerance due to cardiovascular disease, peripheral vascular problems, or other medical conditions. All patients with diabetes, despite age or physical shape, should be encouraged to engage in regular physical activity, as tolerated.

#### ***Insulin***

Insulin, a necessary component of diabetes treatment, necessitates injection for administration. Insulin is required therapy for Type I individuals, and is needed for some Type II patients. Insulin, now manufactured by recombinant DNA technology, is identical to human insulin in chemical structure and has completely replaced animal source insulin. In recent years, biochemists have been able to alter the structure

of the basic insulin molecule by changing and substituting amino acids, producing insulin analogs with differing properties, onset, and duration of action. Currently, the majority of insulin-dependent patients require mixtures of different insulin types. Most patients use rapid acting insulin in conjunction with an intermediate or longer acting insulin, administered in multiple daily injections. Continuous subcutaneous insulin infusion is available for use via the insulin pump. This device continuously administers a variable basal insulin dose, which can be programmed to change during a 24-hour period, along with a patient determined pre-meal bolus via a small indwelling needle (American Diabetes Association, 2006c). Scientists are currently developing a “closed loop” insulin pump, consisting of a continuous glucose monitor that can vary the insulin infusion rate to maintain normal blood glucose, but such advanced technology is not yet available (Skyler, 2006).

### ***Oral Hypoglycemic Agents***

There are several different classes of oral medications, with varying modes of action, available for therapy of Type II patients who are unable to maintain adequate blood glucose control using diet and exercise therapy alone. The major mechanism of action of such drugs is stimulation of the pancreas to release stored insulin. These do not, however, cause the pancreas to produce more insulin. These drugs are not indicated for Type I diabetes since this form is characterized by absolute insulin insufficiency.

Patients with diabetes using the insulin therapies discussed previously are at risk of “insulin reactions,” hypoglycemia (low blood sugar). Such reactions may occur when the individual misses a meal, increases physical exercise, or inadvertently takes too much insulin. Insulin-dependent patients are educated to recognize and treat the symptoms of this common situation. If appropriately recognized, treatment is relatively easy, and includes immediate ingestion of carbohydrate (sugar) in a readily available form, such as eating a piece of candy or fruit. This ingestion will usually raise the blood sugar level and relieve a hypoglycemic situation in a few minutes. Some oral hypoglycemic agents may cause low blood glucose under certain conditions.

### ***Blood Sugar Testing***

The introduction of small, inexpensive, and accurate blood glucose sensors for home use has made the self-monitoring of blood glucose easily accessible. It is necessary for patients with diabetes to perform self-monitoring of blood glucose daily to gauge and maintain their levels of blood sugar. The procedure is simple and fast; a drop of blood, obtained from a finger or forearm via a small lancing device, is placed on a test strip, which is read by the blood glucose monitor in a few seconds. The result is displayed visually. Type II patients with diabetes should perform self-monitoring 1-4 times daily, while Type I patients should monitor blood glucose 4-6 times daily for ideal control.

### ***Functional Limitations***

If an individual maintains good blood glucose control and lacks complications, there will be few work restrictions. Persons taking insulin or oral hypoglycemic agents should try to avoid working irregular hours or rotating work shifts. It is desirable to have a work schedule that remains consistent for optimum control of blood glucose. Fluctuations in exercise cause variations in blood glucose levels. An occupation that requires consistent amounts of physical activity throughout the workday is preferred to one in which the physical demands vary greatly. Of course, jobs often require a variety of work duties with differing physical demands. A knowledgeable person with good fundamental diabetes control can adjust medication and diet to compensate for varying physical activity (Brodwin, Parker, & De La Garza, 2003).

People taking insulin need a readily available source of sugar to combat potential insulin reactions. All individuals who are insulin-dependent should carry candy or another source of sugar to alleviate insulin reactions immediately when they occur. Persons who are in good metabolic control manage occasional minor hypoglycemic episodes so that they go unnoticed by others at the workplace.

An individual with diminished vision may require job accommodation. The degree of visual loss and prognosis for further loss are crucial factors. The greater the amount of visual loss, the more the counselor needs to evaluate occupations requiring less reliance on vision for primary work functions.



Secondary job duties may be modified or eliminated through provision of reasonable accommodation (Brodwin et al., 2003).

Lower extremity complications require job modification involving less ambulating, lifting, carrying, and standing. The degree of lower extremity involvement determines the amount of work restrictions required. If a lower extremity amputation has occurred, sedentary work is realistic. The level of amputation and the individual's adaptation are considered before restricting him or her to sedentary work. Fortunately, above-the-knee amputations are rarely necessary for persons with diabetes (Unger, 2007).

Mild to moderate neuropathy of the lower extremities does not preclude heavier types of work, whereas moderately severe to severe neuropathy may preclude all but sedentary work activity (Falvo, 2009; Marrero & Guare, 2005). The worker, treating physician, and counselor can offer advice on the appropriateness of certain physical activities at the workplace and at home. Additional limitations of function may occur when there are skin problems at an amputation site. Minor skin problems can quickly develop into ulcerations, which impede proper fit of a prosthesis, limit ambulation, and require medical consultation.

Complications involving the circulatory system significantly limit physical activity. The counselor can be guided by the restriction of physical activity noted by the treating physician. Persons with renal failure usually have multiple complications. Depending on the multiplicity of complications and degree of renal failure, work activity may be restricted to part-time, at best. The counselor needs to evaluate the possibility of working at home for certain people.

Emotional factors play a role in control of diabetes from a physiological and psychological perspective; emotional instability complicates treatment. A stressful event may result in a temporary rise in blood glucose levels. If an occupation is stressful and affects good blood glucose control, intervention by a rehabilitation counselor is appropriate. When providing rehabilitation services, the counselor needs to assess the stressful components of work. Some people handle emotional stress well and almost "thrive on stress." For some, occupational stress is not a significant factor.

The longer diabetes has been present, the greater the potential complications. Yet, complications are at least partially dependent on the degree of control of blood glucose a person has maintained over the long term, how quickly a physician is consulted for treatment, and adherence to medical recommendations. Although some people remain relatively free of complications, rehabilitation professionals see those persons who have complications interfering with work functions.

### ***Rehabilitation Potential***

People with diabetes under good control and with no complications will not be referred for vocational rehabilitation. If diabetes is a secondary condition, the counselor needs to consider it when developing a rehabilitation plan. Attitude is crucial in this and other disabilities (Brodwin & Orange, 2008). The person with diabetes who has a positive attitude and outlook has much greater potential for successful rehabilitation.

For individuals with diabetes using insulin, proper therapy, and adaptation to medical recommendations increases the chances of successful rehabilitation planning. Poor control and failure to adhere to medical advice impedes chances for a successful outcome.

Visual loss impacts the ability to perform many job functions. This is dependent on the amount and type of visual acuity required to conduct various job duties. It may be possible to redefine work responsibilities through the provision of reasonable accommodation. Visual loss affects rehabilitation potential to a variable extent (Brodwin et al., 2003).

Many persons of working age with lower extremity amputation are good candidates for fitting and use of prostheses. The purposes of a prosthesis are to restore mobility and increase independence. Most amputations due to complications of diabetes are below the knee. For amputations that are above the knee, ambulation is more difficult and consumes additional energy. If the prosthesis fits well and is used properly and regularly, rehabilitation potential remains positive. In most cases, a person with a bilateral lower extremity amputation requires a wheelchair for mobility. On occasion, a person can learn to use two lower



extremity prostheses for ambulation, depending on age and general medical health (American Diabetes Association, 1998; Marrero & Guare, 2005).

Rehabilitation potential must be evaluated on an individualized basis. Factors to consider when assessing potential for rehabilitation services include physical limitations, control of daily blood sugar levels, motivation, attitude, psychosocial functioning, and coping strategies. Ninety-seven percent of 300 physicians surveyed by the Integrated Benefits Institute stated that “return-to-work is good medicine.” The American College of Occupational and Environmental Medicine Guidelines (2006) noted that safe, early return-to-work programs are in patients’ best interests and promote recovery. Prolonged absence from work makes adjustment back to the job more challenging and decreases the probability of the individual returning to work. There are four basic return-to-work options for individuals with diabetes:

1. Full time with no restrictions
2. Full time with restrictions
3. Part-time with no restrictions
4. Part-time with restrictions

Counselors knowledgeable in medical aspects of diabetes, ways to provide reasonable accommodation in business and industry, and a caring attitude can do much to assist clients with diabetes maximize their work potential and independent functioning.

## **Renal Disease**

Persons with diabetes mellitus account for nearly half of renal disease cases. In the following section, the reader will find information on renal disease and its connection to diabetes. Individuals who do not have diabetes but have renal disease also are included within this section.

Most adults go about their daily activities unaware of the various body systems continuously functioning to keep their bodies in a healthy state. The urological system is one such system. When in good health, the average person maintains only the cognizance to relieve the pressure resulting from urinary bladder distension through urination. Because of the general lack of public knowledge about body systems, internal malfunctions often require professional intervention. Despite advances in health care, some people are affected by disorders that permanently disrupt kidney function. This condition, known as end-stage renal (kidney) disease, is incompatible with normal life functioning, causing serious health problems and potentially resulting in early death.

### ***Kidney Functioning***

The role of the kidneys exemplifies the interdependence of various body systems. The urine excretory function of the kidneys plays a major role in maintaining the chemical balance within the body, and comprises one of the most effective filtering systems known to humankind (Andreoli, Carpenter, Goldman, & Ausiello, 2008). The two kidneys in the body function separately; to sustain life, only one healthy kidney is needed.

#### ***Physiology***

When filtering the blood, the kidneys have a remarkable ability to retain blood cells and certain proteins while passing on unwanted waste products. As the filtrate continues by way of the tubules within the kidneys, the process of re-absorption takes place. In this process, the tubules take back certain substances that have been filtered out and return them to the bloodstream, while allowing other products to be eliminated through the urine. This latter process is called the excretory function of the kidneys. This maintains the chemical composition and balance of the blood. Excreted substances form the urine which travels through the tubules into larger collecting tubes, finally being excreted through the right and left ureters. The ureter from each kidney conducts the urine from the kidneys downward to the urinary bladder to be eliminated from the body (Tanagho & McAninch, 2008). The processes of absorption and excretion give the kidneys the capacity to dilute or concentrate the urine in accordance with bodily needs (Brenner & Rector, 2008).

### ***Malfunctioning of the Kidneys***

When the kidneys begin to fail, they lose their ability to perform the functions of absorption and excretion; this disruption leads to a chemical imbalance with abnormal clinical consequences. In some diseases, for example, the kidneys retain sodium. When this happens, the body retains water, causing generalized edema. The person gains weight and appears to be bloated. As the severity of edema increases, the volume of fluid within the body increases. As a consequence, there is a rise in blood pressure; this places a strain on the heart as it must pump harder to circulate the blood.

During renal failure, unwanted by-products are retained within the body. Urea and creatinine, for example, are byproducts of cellular metabolism. When these chemicals build up in the body, they cause a toxic condition known as uremia, which can be life threatening. Failing kidneys not only retain but also excrete abnormal amounts of certain proteins, such as albumin and globulin, which are essential in maintaining the delicate fluid balance of the body.

### ***Other Essential Kidney Functions***

Additionally, the kidneys produce hormones. Hormones are chemicals produced in an organ and released into the bloodstream, affecting the cells of other organs and altering their functioning processes. Erythropoietin, a hormone produced by the kidneys, stimulates the production of red blood cells in the bone marrow. In failing kidneys, the decreased production of this hormone results in anemia (a deficiency of red blood cells).

Activated Vitamin D, produced within the kidneys, affects the absorption of calcium required for normal bone formation. Renin is also synthesized by the kidneys and plays an important role in the regulation of blood pressure. It activates the hormone, angiotensin, which triggers the release of aldosterone from the adrenal glands. Aldosterone causes the kidneys to retain sodium, expanding blood volume and increasing blood pressure (Tanagho & McAninch, 2008; Zipfel, 2006).

## ***Renal Failure***

The term “renal failure” refers to the malfunctioning of both kidneys to such an extent that renal function is below the level required to sustain life. The evaluation of renal failure takes into account many factors, including the individual’s subjective complaints and objective laboratory findings. The functional progress of acute or chronic renal failure is most often monitored by repeated laboratory measurement of urea and creatinine blood levels over time.

### ***Loss of Kidney Function***

Common causes of permanent loss of kidney functioning include (El Nahas, Harris, & Anderson, 2000; Tanagho & McAninch, 2008):

- Diabetes mellitus with nephropathy
- Hypertension
- Congenital defects
- Trauma due to accidents; in these cases, the individual usually does well since one kidney usually is functional
- Autoimmune reactions that damage the glomerulus
- Infection due to various types of bacteria that ascend from the bladder and infect the kidneys (pyelonephritis)
- Restricted circulation in the glomerular tuft, such as in arteriosclerosis (hardening of the arteries), thrombosis (blood clot), and hypertension (high blood pressure)

### ***Acute and Chronic Kidney Failure***

Kidney failure can be either acute or chronic. Acute kidney failure is characterized by a rapid decline in normal kidney function due to trauma, bleeding, shock, burns, and other causes. Irrespective of the cause, symptoms are related to the severity of failure and the extent of involvement of other body

systems (Thomas, 2004). Though many symptoms of acute kidney failure are treated successfully, some cases can result in chronic renal failure.

Chronic renal failure begins as a slow, progressive decline in kidney function that results in the build-up of metabolic waste products within the body. At the outset, chronic renal failure prevents the kidneys from adequately regulating the body's water content. As the condition evolves, laboratory tests indicate abnormal findings despite the fact that the person may present with only mild complaints. As the disease progresses, the individual begins to have back pain, experience frequency of and burning with urination, notices the presence of blood in the urine, and experiences nocturia (frequent urination at night). As toxic substances continue to build up within the body, the person becomes less alert and loses the ability to concentrate. Nausea and vomiting occur which, if left untreated, can lead to malnutrition, coma, and death (Rennke, 2007; Thomas, 2004). End-stage renal failure progresses gradually. It is categorized into three stages:

- Renal Impairment – Early-stage renal disease involving a 40%-70% loss of function
- Renal Insufficiency – Second stage renal disease involves a 75%-85% loss of function
- Renal Failure – End-stage renal disease where less than 15% of function remains

Human kidneys, like many other organs of the body, have a reserve capacity meaning that they are able to perform their normal work load with as little as 10% remaining function. Beyond this point, life is not sustainable. In cases where less than 10% of function remains, one must seek alternative or substitute kidney functioning. Two alternatives are dialysis and kidney transplantation (Rennke, 2007).

### ***Dialysis***

Dialysis makes use of the physical properties of a semi-permeable membrane. When two solutions, one more concentrated with small particles or molecules than the other, are separated in a container by a semi-permeable membrane, the concentrated substances move across the membrane until the two solutions become equal in concentration. Microscopic holes in the semi-permeable membrane allow for the diffusion of waste products but prevent the passage of proteins or cells, such as red and white blood cells, which would otherwise be lost (Nissenson & Fine, 2005). Dialysis removes excess fluids and impurities from the body and serves as a substitute for functioning kidneys. Two principal forms of dialysis are (a) hemodialysis and (b) peritoneal dialysis.

#### ***Hemodialysis***

This is a circular process wherein the person's arterial blood is transported outside the body to a dialysis machine where it is cleansed of waste products and returned to the circulatory system. In contrast to the transitory use of hemodialysis during acute kidney failure, where the condition is temporary, hemodialysis for chronic renal failure is a lifetime requirement. As a consequence of repeated and life-long needle puncture of the involved blood vessels, thick scar tissue eventually develops which makes the procedure difficult and painful. To avoid this complication, a surgical procedure known as an arteriovenous fistula is performed. Usually done in the forearm, it produces a large vein that can be entered safely and easily with large needles for the three times a week procedure (Nissenson & Fine, 2005; Thomas, 2004).

While dialysis is a life-saving procedure, it is not as efficient as normal kidney functioning. The patient with end-stage renal disease undergoes dialysis for three to five hours a day three days per week. In contrast to human kidneys that function 24 hours a day, dialysis results in the gradual build-up of waste products from one treatment to the next. Depending on the extent of this accumulation, the person may experience fatigue, loss of appetite, and have lapses in concentration.

#### ***Peritoneal Dialysis***

Peritoneum is the lining of the abdominal cavity. This tissue has the properties of a semi-permeable membrane, allowing the process of diffusion to take place within the abdominal cavity. A small incision is made through the anterior abdominal muscle wall; through this opening, a tube (catheter) is inserted into the abdominal space. Sterile dialysate (solution used to clear waste products) is introduced and allowed to remain in the cavity between four and six hours, or overnight. During this time, through the process of

diffusion, impurities pass through the peritoneum into the dialysate. The tube is then lowered and gravity induces a flow of the dialysate and its collected impurities out of the body (Hricik, Sedor, & Miller, 2003).

### ***Kidney Transplantation***

When possible, kidney transplantation is the best alternative for end-stage renal disease. One major complication of this procedure, however, is the negative reaction of the immune system to a foreign organ. In this scenario, a new donor kidney is not recognized as the body's own, but rather is rejected as an invader threatening the body.

To overcome this reaction to kidney transplantation, the recipient must undergo a medication regimen to suppress the immune system. Today, a national computerized kidney-harvesting program serves to locate donor kidneys and eligible recipients. Donor kidneys are ideally obtained from living relatives to avoid the recipient's immune system rejection of a new kidney. Potential donors are carefully evaluated for compatibility and freedom from disease (El Nahas et al., 2000; Nisseenson & Fine, 2005).

Cadaveric donor kidneys come from individuals who have recently died as a result of fatal brain trauma. In these cases, the compatibility procedures undergone mirror those performed for transplants from living donors. Surgical procedures for kidney transplantation are usually successful. In these procedures, a donor kidney is implanted in the front groin area of the body and the recipient's non-functioning kidneys are left intact as long as they do not seriously impact the patient's overall health.

In an effort to suppress the immune system and thus avoid donor kidney rejection, medications such as corticosteroids and cyclosporine are prescribed (Danovitch, 2005). Careful laboratory monitoring is essential as too low a concentration in the bloodstream exposes the kidney to rejection and high doses inhibit the body's capability to protect itself against infection. People who have had transplants remain on immune suppressors throughout their lives (Morris & Knechtle, 2008). As there are a great deal more people needing kidneys than those that become available, an individual may have to wait for several years before a donor kidney is obtainable.

In comparison with dialysis, kidney transplantation has advantages that positively affect the person's medical and vocational rehabilitation potential. The person with a kidney transplant does not spend three to five hours a day three times a week on a dialysis machine experiencing, in some cases, wide fluctuations in body chemistry. Furthermore, the individual is not exposed to the hazards of bleeding, clotting, and infection associated with constant use of needles and fluid exchanges.

### ***Emotional Issues***

Kidney failure causes severe emotional stress for both the individual undergoing treatment and his or her family members. Uncertainty of the future is a major stressor in all chronic conditions, including end-stage renal failure. Even after a successful kidney transplant uncertainty remains as the new kidney may be rejected at any time. People on dialysis experience a drastic shift in lifestyle, as the tri-weekly dialysis procedures take precedence over all other life activities. With effective treatment, however, improvements in quality of life occur. If a person receives a transplant, overall health and outlook on life improves substantially as one regains control over life. Some transplant recipients who are not employed return to work and function effectively. A return to work is more feasible for those who have undergone successful kidney transplantation than when receiving dialysis. Sedentary and light work activities are recommended for both transplant and dialysis patients to facilitate a return to the vocational environment. Emotional counseling and support groups further aid in the process of coping with the psychosocial aspects of this condition.

### ***Complicating Factors Affecting Employment***

#### ***Anemia***

All blood circulating in the body eventually passes through the kidneys to be purified of waste products. When the volume of oxygen in the blood reaches a minimal level, the kidneys sense a need for more red blood cells and produce a hormone called erythropoietin. This hormone stimulates bone marrow to produce red blood cells in greater numbers. When kidneys severely malfunction, such as in end-stage renal

disease, they do not produce erythropoietin, resulting in the development of anemia. The body responds to the paucity of red blood cells by increasing the breathing and heart rates to circulate fewer cells more rapidly and serve the same function. To help prevent anemia from occurring, human erythropoietin is prescribed, which is available by means of recombinant DNA.

### ***Infection***

For people on hemodialysis, the frequent introduction of needles into blood vessels and the repeated need to control blood leaks lowers resistance to infection. To avoid complications, individuals and paramedical team members must adhere to appropriate aseptic techniques (Teichman, 2001). As many individuals have lowered resistance, medical consultation is necessary at the first sign of fever or inflammation.

### ***Rigid Treatment Requirements***

When undergoing hemodialysis, most people require three to five hours of treatment each day three days a week. As a result, individuals on peritoneal dialysis must plan their home and work activities around treatment times, giving dialysis precedence over work and other life activities. Adherence to such rigid treatment requirements is essential for successful treatment (Danovitch, 2005). Many dialysis centers operate 24 hours a day, allowing recipients of dialysis to work regular daytime hours.

### ***Rehabilitation Potential***

Many persons who have left their jobs because of declining kidney function may be able to successfully return to work once they are on a consistent treatment regimen, such as a dialysis program, or after receiving a kidney transplant. For those beginning dialysis, adjustment time is required to arrange activities around dialysis. Dialysis and kidney transplant recipients should avoid jobs involving physically strenuous work activities. Maintenance of optimal health is essential for persons on dialysis since they are predisposed to anemia, have lowered resistance to infection, and experience fatigue. To help maintain a life-long program of well being, including a well-balanced diet and an appropriate exercise program, hospitals employ a team of professionals consisting of the treating physician, dietician, medical social worker, clinical psychologist, and vocational rehabilitation counselor to encourage, counsel, and assist the person (Nissenson & Fine, 2005). Ideally, the vocational rehabilitation counselor coordinates the services of all participants on the client's behalf in such a way as to bring about a successful return to independence, daily activities, and employment.

Despite passage of the Americans with Disabilities Act (ADA) (1990), job placement remains challenging for those who have had kidney transplantation or are currently undergoing dialysis. Employers have the perception that their overall medical insurance costs will increase and the individual may have too many days off work due to illness. The vocational rehabilitation counselor can help the person overcome obstacles like these by being persistent in following up with employers and medical insurance companies to assure the medical and employment stability of the client and, if needed, to explain the client's rights under the ADA.

## **Case Study**

Ms. Consuelo Mendoza is 44 years old and immigrated to the United States from Puerto Rico. Currently, she lives with her sister and the sister's 13 year-old son. While in her native country, she worked as a flight attendant, received a college education, and subsequently became employed as a language arts middle school teacher. Since arriving in this country, Ms. Mendoza has worked as a secretary, home health aide, and salesperson in a women's dress shop. On a part-time basis for many years, Consuelo taught ESL (English as a second language, [English Language Learners]) classes and tutored Spanish-speaking individuals wanting to learn or become proficient in English.

Since adolescence, Consuelo has had Type II diabetes mellitus. At age 40, she was diagnosed with kidney disease. As a result, she is restricted to a maximum of light work, although her treating physician would prefer that she work in a sedentary capacity. In end-stage renal failure for the past year, she has been put on a list to receive a kidney transplant. Ms. Mendoza requires dialysis three times a week for



approximately four hours each time, which she receives in the evenings and on weekends. For the year she has been on dialysis, she has worked only part-time as an ESL teacher and tutor.

Recently, Consuelo has sought vocational rehabilitation services because her part-time work does not provide her with sufficient income. There have been some months when she earns enough income to meet all her expenses and other times when she does not.

### **Questions**

1. Identify factors significant in assessing rehabilitation potential of this client.
2. Discuss Type I and II diabetes and complications that can occur. Relate this to this case. What probably caused this client to have renal failure?
3. Describe the two types of dialysis and kidney transplantation. Identify advantages of a transplant over dialysis.
4. Considering that Ms. Mendoza is on dialysis and may be receiving a kidney transplant within the year, what kinds of rehabilitation services will you provide at the present time and in the future? Are there any considerations needed for Consuelo's diabetes?
5. What issues related to dialysis and kidney transplant need to be considered in vocational planning?
6. Describe issues of reasonable accommodation for clients on dialysis.
7. Because of strong religious beliefs and her own fears concerning surgery, Ms. Mendoza is conflicted about receiving a transplant. As her rehabilitation counselor, how would you handle these mixed feelings she has shared with you?

### **Acknowledgment**

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# Chapter 6

## CARDIOVASCULAR DISEASE

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### Introduction

The purpose of this chapter is to assist rehabilitation counselors in comprehending medical evaluations that identify impairments of the cardiovascular system, and how such impairments relate to an individual's ability to function independently and become successful within the workforce. This chapter describes the New York Heart Association's classification system of heart disease, the different kinds of heart disease, diagnosis and treatment, therapeutic and diagnostic procedures, disability rating, functional limitations, and rehabilitation potential for individuals with cardiovascular impairment. The chapter concludes with a case study of an individual who has several disabilities, including coronary artery disease.

A principle direction of this chapter is the discussion of the causes, symptomatology, pathology, and evaluation of physical capabilities of persons with coronary disease. Increasingly, the direction of medical studies and treatment is on determining the principle course of coronary artery disease in the employed population. Modern research is directed toward preventing and decreasing progression of this disease, with the goal of increasing longevity and improving quality of life and productivity. The rehabilitation counselor's challenge is to assist this growing population to remain vocationally competitive and productive.

Heart disease is a leading cause of death in the United States. A heart attack, also known as a myocardial infarction, occurs when the supply of oxygen to a portion of the heart muscle is curtailed to such an extent that tissues die or sustain permanent damage. Generally, a heart attack is the result of a blood clot or thrombus that suddenly blocks one of the coronary arteries supplying the heart. Approximately 1.5 million heart attacks occur each year; only about 50% of these individuals survive. One-third of the 1.5 million individuals who experience heart attacks are under 65 years of age (Braunwald, 2005). The following seven major types of heart conditions are discussed within this chapter: (1) congenital heart disease in adult life; (2) valvular heart disease; (3) mitral valve prolapse; (4) cardiomyopathy; (5) hypertension and hypertensive heart disease; (6) coronary artery disease; and (7) congestive heart failure.

In the first five categories, heart strain or progressive heart muscle failure may eventually develop into congestive heart failure. In all groups, employability is dependent on a timely diagnosis, the potential for correcting or improving the disease process, anatomical abnormalities, and provision of rehabilitation services. In coronary artery disease, which represents the largest category, there is progressive inability of the heart muscle to function properly, primarily as a result of decreased blood flow to the heart muscle due to occluded coronary arteries. After a given quantity of muscle mass has been damaged or destroyed, pump failure or congestive heart failure, can result in the inability to pump sufficient blood to the rest of the body.

### Functional and Therapeutic Classifications of Heart Disease

The following discussion refers to the functional capacity of the heart and is adapted from criteria of the New York Heart Association (NYHA). Physicians and researchers accept this protocol as a standard for classification of cardiac conditions. Both classifications are evaluated to derive a cardiac status determination (Criteria Committee of the New York Heart Association, 1984).

### ***Functional Classification – NYHA***

“Functional Classification” is an estimate of a person’s symptoms. It is described in four classes, I through IV (see Figure 1). Class I represents a person with no symptoms, while Class IV is an individual who develops discomfort with any physical activity and has symptoms even at rest. Classes II and III describe symptoms between these extremes.

### ***Therapeutic Classification – NYHA***

“Therapeutic Classification” reflects the amount of physical activity recommended by the treating cardiologist considering all factors. There are five classes from Class A representing those individuals with no physical activity restriction to those in Class E whose physical activity is almost totally restricted.

It must be noted that the NYHA classification is dependent on the subjective reporting of the patient. Impairment ratings should preferably be done using objective guidelines, such as described in the section Disability Rating of this chapter.

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**Figure 1**  
**New York Heart Association Classifications of Patients with Heart Disease**

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#### *Functional Classes*

Class I	Patients with heart disease who have no symptoms of any kind. Ordinary physical activity does not cause fatigue, palpitation, dyspnea, or angina pain.
Class II	Patients who are comfortable at rest but have symptoms with ordinary physical activity.
Class III	Patients who are comfortable at rest but have symptoms with less than ordinary effort.
Class IV	Patients who have symptoms at rest.

#### *Therapeutic Classes*

Class A	Patients whose physical activity need not be restricted.
Class B	Patients whose ordinary activity need not be restricted, but who should be advised against severe activity.
Class C	Patients whose ordinary activity should be restricted.
Class D	Patients whose ordinary activity should be markedly restricted.
Class E	Patients who should be at complete rest in bed or a chair.

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## **Congenital Heart Disease in Adult Life**

Congenital heart disease is a condition caused by various malformations of the heart, which develop during gestation and become apparent in infancy or childhood. Many congenital heart problems require surgical or manipulative procedures early in life to improve cardiovascular function. Not all untreated individuals have residual problems; however, those who live into adulthood develop serious problems that require intervention to improve or correct abnormalities. Early adult heart failure is a major problem necessitating aggressive treatment and evaluation to determine functional limitations and work restrictions (Skala, Freeland, & Carney, 2005). Rehabilitation counselors must obtain medical information describing these limitations, based upon objective testing, before attempting to reemploy an individual with cardiovascular limitations (Topol, Califf, Prystowski, & Thomas, 2007).

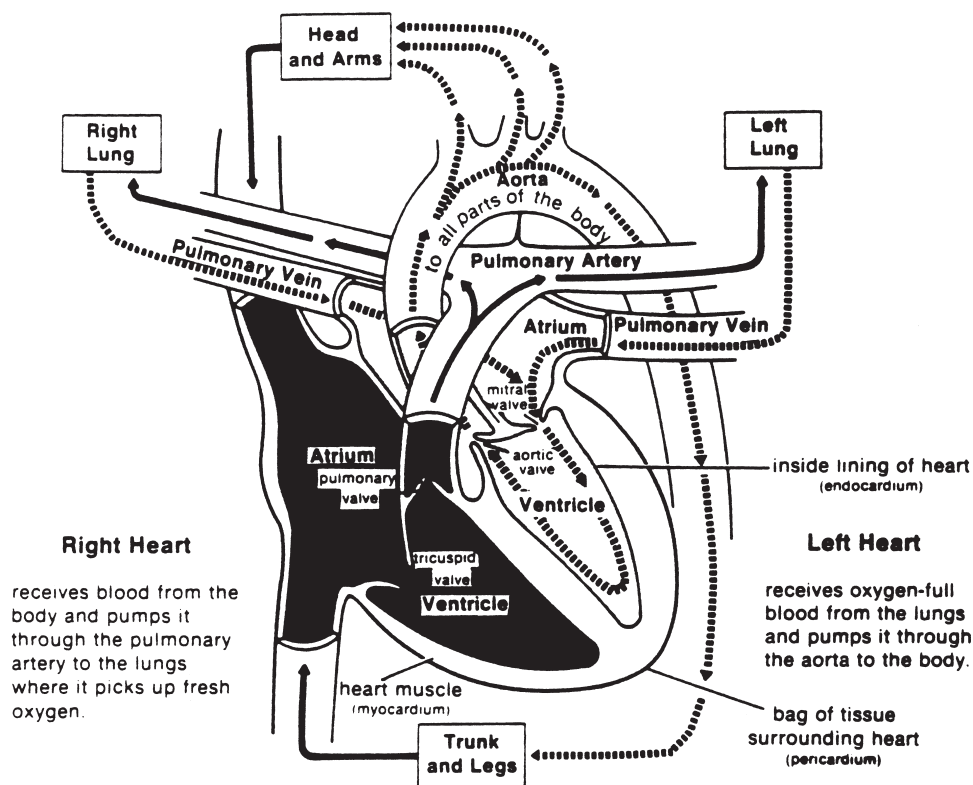
## Valvular Heart Disease

For adults under 65 years of age, valvular heart disease is usually a result of childhood rheumatic fever. It is an inflammatory condition that may damage the heart valves. If the damage eventually affects the performance of the heart, it is called rheumatic heart disease (Braunwald, 2005). In the United States, rheumatic fever was once a devastating disease, common in the early part of the Twentieth Century. Improved understanding of prevention and antibiotic treatment has reduced not only the incidence and recurrence rate of rheumatic fever, but its severity and frequency as well. Rheumatic heart disease as a cause of permanent disability has markedly diminished over the past 50 years (Moser & Riegel, 2008).

Rheumatic valvular disease, however, is still a major source of concern. One chief complication of this disease process is that a person may become symptomatic between the middle teens and mid-30s, depending on the severity of the original acute process. Acute rheumatic fever leads to acute cardiomyopathy (abnormal changes in the heart muscle). The disease causes damage to the heart valves, including the mitral valve, aortic valve, and tricuspid valve, in this order (see Figure 2). The mitral and aortic valves may become stenotic (i.e., constricted or narrowed), insufficient (i.e., the valve leafs do not come together, thus allowing leakage), or both. Although not subject to stenosis, the tricuspid valve can become insufficient. The greater the number of valves affected, the sooner the person develops heart failure, and the more severe the failure (McCarthy, & Young, 2007; Scodo, 1998).

Benefiting from emerging technology of prosthetic heart valve replacement, a growing number of people with rheumatic valvular disease can hold jobs not requiring heavy work or exposure to trauma. Individuals with valve replacement require medication to prevent abnormal blood clotting. A commonly prescribed preparation for this condition is Coumadin (warfarin sodium). People on this and similar

Figure 2  
The Human Heart



*Your heart and how it works* (1979). ©American Heart Association. Reproduced with permission.

medications bruise easily and should not work in areas where physical trauma could occur (Moser & Riegel, 2008). Today, there are an increasing variety of valve prostheses available. Surgery, when combined with proper medication and performed early in the disease process, can enable a person with valvular disease to lead a normal life. Cardiac testing helps determine one's work potential, both before and subsequent to surgery.

## **Mitral Valve Prolapse**

Mitral valve prolapse (mitral stenosis) is one of the most common valvular abnormalities. It involves a narrowing of the heart valve, or an incompetent valve which does not close completely (Skala et al., 2005). In recent years, there has been an increase in the diagnosis of mitral valve prolapse (MVP). The exact cause of the condition is unknown. Possible causes include an undetected congenital defect, viral infection, or most likely, myxomatous (degeneration of the heart valve). The most common symptoms experienced are anxiety, fatigue, palpitations, and chest pain. Frequently, the discovery of an asymptomatic extra heartbeat can alert a physician to a possible diagnosis of mitral valve prolapse. MVP may be accompanied by a degeneration of the valve musculature known as myxomatous degeneration. In some cases, this mucous degeneration affects the tricuspid valve. When both mitral and tricuspid valves are diseased, the person develops cardiac insufficiency, which may progress to heart failure.

A test commonly known as an echocardiogram (ECG, EKG) confirms a diagnosis of MVP, generally a benign condition. Progression to a severe condition is rare. However, when such progression does occur, it may lead to severe mitral regurgitation associated with heart failure. If this is the case, surgery is compulsory. If a Holter monitor (a device worn by a person during normal activity as an ECG) reveals significant arrhythmia and the person is symptomatic, beta-blocker medication is used for treatment (Moser & Riegel, 2008; Scodo, 1998).

## **Cardiomyopathy**

### ***Medical Implications***

This group of diseases, also known as myocardiopathy (cardiac muscle weakness), involves inflammation and resulting weakness within the heart muscle. Cardiomyopathy is a disease of the heart muscle making it weak and unable to function effectively. Myocardiopathies may be secondary to infectious processes or chemical toxic exposure, as well as part of the disease process of diabetes mellitus and alcohol abuse. Virus myocardiopathies are more frequently found in younger people. The virus causes inflammation and weakness of the heart muscle and sometimes interferes with normal nerve conduction throughout the heart. During the acute phase, individuals may require hospitalization and possible intensive care to become medically stabilized. People with myocardiopathy may additionally require short-term medication, or in the more serious case, a pacemaker for cardiac irregularities and life-long medication.

Some people experience arrhythmias in the form of extra heartbeats even after the acute phase, and require medication for control. Additional symptoms include weakness, fatigue, fluid accumulation in the extremities (sometimes in the lungs), difficulties in breathing, fainting, and nausea. Others need medication to support the myocardium (the muscle layer of the heart). In most cases, after the acute stage has passed, the person can continue in customary work requirements, with the exception of heavy lifting and arduous labor. Surgical correction of associated valve abnormalities may be recommended (Rey, 2005; Topol et al, 2007).

### ***Diabetes Mellitus***

When it is not optimally controlled, diabetes may cause complications resulting in poor function of one or more organs or systems of the body. Poorly controlled diabetes may cause cardiac muscle disease resulting in progressive, congestive heart failure. Thus, individuals with diabetes must maintain normal blood glucose, body weight, and appropriate diet to lessen such complications. The probability and severity of diabetic cardiomyopathy is diminished when proper medical care is attained; if diagnosis of cardiomyopathy is given, however, a patient is usually restricted to light or sedentary work (Moser & Riegel, 2008).



### ***Alcoholic Cardiomyopathy***

Toxic effects of alcohol and poor nutrition bring about alcoholic cardiomyopathy. Proper medical care for the alcoholic and complete abstinence from alcohol (and tobacco) reduces the severity of the myocarditis (inflammation of the myocardium). General improvement in health and overall functioning are the goals of alcohol rehabilitation programs, requiring total abstinence from alcohol, good nutrition, and treatment of complications. Every effort is made to provide and maintain an ongoing therapeutic prevention program, such as Alcoholics Anonymous (AA).

## **Hypertension and Hypertensive Heart Disease**

Hypertension (high blood pressure) occurs when the pressure of the blood against the walls of the vessels exceeds normal limits. About 50 million people in this country have hypertension. Many have no recognizable symptoms and are unaware of having the condition. An additional 45 million have pre-hypertension and are more susceptible to develop hypertension. Individuals with hypertension are at risk of having heart attacks, heart failure, and cerebrovascular accidents (Mertz, 2004).

During the normal heart cycle, action of the heart can be compared to a hydraulic pump. When the heart muscle contracts, it propels blood out of the heart chambers. This forceful action is called systole. Pressure is high during this expulsive phase, known as the systole. The phase immediately following, when the heart muscle is at rest, is called the diastole. Blood pressure, as expressed as a fraction, is written with the systolic pressure on top and diastolic pressure on the bottom (e.g., 120/80 = systolic/diastolic). Diastolic pressure thus represents blood pressure when the heart is at rest; elevated diastolic blood pressure is a serious condition. Systolic pressure greater than 140 mm Hg (mercury), and diastolic pressure greater than 90 mm Hg is considered hypertension.

### ***Causes of Hypertension***

Ninety-five percent of the cases of hypertension are of unknown cause, while 5% are related to kidney disease, endocrine abnormalities, increased cardiac output, and vascular abnormalities (Braunwald, 2005). Heredity is a factor in hypertension; therefore, there is a genetic predisposition to heart disease. Surgical intervention or specific medical regimens benefit many “secondary” cases of hypertension.

### ***Detection of Hypertension***

Symptoms of hypertension may surface suddenly without warning. Commonly encountered nonspecific complaints such as fatigue, headache, nosebleed, change in vision, or muscle weakness are typical. Complications include coronary artery disease, renal failure, cerebral vascular accident, and peripheral vascular disease. Since most individuals do not have symptoms, hypertension has been called the “silent killer.”

When hypertension is suspected, physical examination includes assessment for any abnormalities of the hypertensive person’s kidneys, heart, and cerebrovascular system. Additionally, chest x-ray, electrocardiogram, and echocardiogram are used to assess cardiac status. If hypertension is thought to be associated with coronary artery disease, a physician may conduct cardiac stress testing to clarify the diagnosis (Braunwald, 2005; Robinson, 2000).

### ***Complications***

Hypertension may cause significant renal damage leading to kidney failure; this damage contributes to further perpetuation of hypertensive problems. Frequently associated with hypertension, peripheral vascular disease is a major cause of disability. Unlike hypertension, this complication causes pain in the lower extremities, brought on by exercise, due to diminished blood flow in the legs. With a diagnosis of hypertension, the physician evaluates physical activity levels and suggests modifications, depending on the presence of complications (Rey, 2005). Physical limitations experienced with hypertension primarily result from coronary artery disease, stroke, and renal failure. Each of these conditions has specific guidelines as to physical disability and resulting levels of activity (Alpern, 1996). Through consultation with the treating

physician to identify functional limitations, the rehabilitation counselor is able to assist the individual through the vocational rehabilitation process.

Levels of disability for individuals with hypertension are related to the occurrence and severity of complications and presence of end-organ damage (Rey, 2005). Persons responsive to treatment may be able to continue their customary work activities. Individuals with hypertension who do not have major systemic complications but are unable to perform their current jobs may turn to vocational retraining for support and future success. Despite a diagnosis of hypertension, most people continue leading productive lives without serious limitations to work activity, recreation, and life style (Daida, Squires, Allison, Johnson, & Gau, 1996).

## **Coronary Artery Disease**

Coronary artery disease (also known as coronary heart disease) is a broad term describing many heart-related conditions, all involving narrowed coronary arteries affecting blood flow to the heart. In this country, more than 12 million people have this condition. Symptoms usually occur only when the disease is advanced, making it a particularly dangerous condition. Individuals reporting symptoms generally have one or more of the following: discomfort, dull ache, or sharp chest pain, especially with strenuous physical activities; discomfort in the shoulder, throat, or left arm; and shortness of breath (Mertz, 2004). These symptoms are referred to as angina pectoris. Extreme narrowing of these arteries can result in heart attack or heart failure.

Coronary artery disease is the leading cause of death in the United States. Approximately 1.5 million heart attacks occur annually in the United States, and more than 700,000 of these result in death. Many people have coronary artery disease not associated with myocardial infarction (heart attack). The conditions associated with all forms of coronary artery disease can lead to major problems and disability (Rey, 2005).

### ***Etiology***

Risk factors for coronary artery disease include high cholesterol levels, hypertension, diabetes, emotional stress, a diet rich in fats, obesity, lack of exercise, and cigarette smoking. Of the risk factors mentioned, the most significant are cholesterol, blood pressure elevation, and cigarette smoking. High cholesterol leads to accumulation of plaque in the coronary vessels and restricted blood flow. Nicotine from cigarette smoking diminishes blood flow by constricting vessel walls during smoking.

### ***Manifestations of Coronary Artery Disease***

Coronary artery disease leads to myocardial anoxia (insufficient oxygen to the heart muscle), due to diminished or absent blood flow. When this occurs, the person experiences chest pain (angina pectoris) and pressure that radiates from the left shoulder and arm, or sometimes from the heart to the abdomen. Other symptoms include: a feeling of high anxiety or impending doom, sweating, pale (cyanotic) face, labored breathing, and rapid pulse (Mertz, 2004). This disease affects the myocardium and nerve conducting pathways, causing abnormal nerve conduction. Irregular heartbeats (arrhythmias) or ventricular fibrillation (rapid, ineffective pulsations of the heart) occur. During this time, the heart muscle receives insufficient oxygen; if this condition persists, the affected myocardium dies (myocardial infarction). Persistent arrhythmias result in an inefficient pumping action of the heart.

Heart arrhythmias are heart rhythms that deviate from the normal pattern and pace of the heart. People are often unaware of the condition. Those who report having symptoms typically have one or more of the following: weakness, fatigue, dizziness, lightheadedness, and an irregular pulse (Mertz, 2004). When this happens, fluids in the body do not flow properly, leading to congestion. This condition, known as congestive heart failure, is a medical emergency and may lead to death if not treated (McCarthy & Young, 2007).

Four major clinical signs of coronary artery disease are angina pectoris, myocardial infarction, arrhythmias, and congestive heart failure. Angina pectoris is precipitated by physical exertion and relieved by rest or cessation of the specific physical activity. Discomfort may last from a few seconds to several

minutes, but can be a forerunner of a myocardial infarction (Mertz, 2004). Shortness of breath, a need to sit up in bed, and leg edema (swelling) are seen in congestive heart failure.

### ***Myocardial Infarction***

A heart attack (myocardial infarction) occurs when a coronary artery becomes partially or completely occluded by plaque, narrowing a portion of an artery. This clot can develop suddenly at the site of narrowing in the artery, and is usually associated with rupture of plaque, curtailing the supply of oxygen to such an extent that the tissue sustains permanent damage.

Symptoms of myocardial infarction usually begin with chest discomfort, similar to angina pectoris. This pain is more severe and lasts longer than in the case of angina. Associated symptoms include crushing chest pain, sweating, dizziness, nausea, vomiting, weakness, shortness of breath, abdominal pain, rapid heart beat, anxiety, and potentially, unconsciousness. Unfortunately, in some cases, there are no warning signs and sudden death may be the only sign of the presence of coronary artery disease (McCarthy & Young, 2007).

## **Congestive Heart Failure**

Congestive heart failure is the end stage of deteriorated heart function occurring as a result of cardiovascular disease. Because of impaired function, the heart must work harder to deliver a sufficient supply of blood to the body. The heart chambers enlarge (cardiac hypertrophy) and the muscle walls become thicker so that the heart may pump needed blood. Symptoms include dyspnea (shortness of breath), edema in the lower extremities, fatigue, weakness, and abdominal discomfort. This disorder occurs either suddenly (due to a heart attack) or over a period of years. If the underlying problem is treated, improvement occurs.

## **Diagnosis and Treatment**

Ideally, an individual with symptoms of myocardial infarction is immediately hospitalized. Paramedics initiate treatment with administration of pain relieving medication, oxygen, and other supportive measures. In the emergency room, a physician may administer a thrombolytic agent (Rosamond et al., 1998). These clot-dissolving medications are started as soon as possible after a heart attack is suspected, with the goal of improving blood flow in the affected artery. Alternatively, the patient may be referred to a catheterization laboratory for immediate evaluation and possible angioplasty and stent placement (Topol et al., 2007).

After stabilization of the acute event and observation of the patient in a critical care hospital unit, the individual begins a rehabilitation program involving gradual increases in physical activity. This program begins in the hospital and is continued on an outpatient basis, often in a cardiac rehabilitation program. The program provides information on coronary artery disease including dietary and weight management, proper exercise programs, information on medications, assistance with stress reduction, and cessation of smoking (Bates, 1996).

Symptoms and medical findings dictate ongoing treatment, which may involve a medical regimen and possible surgery. Medications are prescribed to control angina and arrhythmias. For example, nitroglycerine is a short-acting medicine that dilates the coronary arteries to relieve angina. The treating physician may prescribe other medications to relieve arrhythmias and help the body eliminate excess fluid.

### ***Diagnosis***

#### ***Echocardiogram (EKG, ECG)***

This is a non-invasive procedure that uses high frequency ultrasound, showing a functioning heart and all moving parts. An EKG displays the muscle activity measured from various directions. This display indicates overall rhythm of the heart and weaknesses in different parts of the heart muscle, to measure and diagnose abnormal rhythms of the heart. EKG can identify damaged heart muscle for an individual who had a heart attack.

### ***Holter Monitor***

A Holter monitor is a small, battery-powered device that is carried like a pager and records a continuous electrocardiogram for up to 24 hours. The individual is asked to keep a written record of the time of any special symptoms and physical exertion. Results are compared with the corresponding time on the EKG to denote abnormalities. If longer periods of time are required to monitor a rare event, an event monitor is attached to the skin that only runs when triggered by the patient with onset of the event.

### ***Cardiac Radionuclide Imaging***

Cardiac radionuclide imaging involves a procedure wherein a liquid radioactive substance (tracer) with an affinity to heart muscle is injected into a vein. A gamma camera scans the heart to detect that portion of heart muscle that has poor circulation and does not take up tracer material.

## ***Treatment***

### ***Surgical Procedures***

Several surgical procedures are used to correct the narrowing or blockage of coronary arteries associated with cardiovascular problems. Coronary angioplasty involves the surgical insertion of a balloon-type device (catheter) into a narrowed artery to flatten the occlusion and allow blood to flow more freely. Laser angioplasty uses lasers to clear plaque; catheters also can be used to install stents, tube-shaped metal devices that remain in place at the problem site to hold the artery open (Mertz, 2004).

Medical evaluation of coronary artery disease includes radioactive tracers or coronary arteriogram. Using a catheter (introduced in another body area and guided into the heart), the cardiologist injects the substance by means of the catheter directly into coronary vessels. The cardiologist is able to visualize blood flow restrictions, called stenosis, brought about by an accumulation of cholesterol plaques within coronary vessel walls. This condition is treatable through angioplasty or coronary bypass surgery. Angioplasty involves opening a blocked artery with a balloon catheter and inserting a stent to open the artery. While angioplasty is effective when the person has only one or two narrowed arteries, it is not practical when more arteries are obstructed. When this occurs or when angioplasty proves unsuccessful, the physician may recommend coronary artery bypass surgery. Instead of repairing the affected region, this procedure reroutes blood around it (Mertz, 2004).

### ***Cardiac Catheterization***

In cardiac catheterization, a thin catheter tube is inserted into an artery or vein and guided into the heart. Different types of catheters are designed to instill medications or dyes that appear on x-ray, allowing physicians to view abnormalities in heart structure, function, and circulation. Some catheters are used for treatment.

### ***Cardiac Transplantation***

The success of cardiac transplantation surgery has gradually improved. To become eligible for a heart transplant, a person needs to be in adequate general health; he or she must be able to withstand the surgical procedure itself, a prolonged recovery time, and a strict medical regimen. In addition to the operation, even being placed on a transplant list is emotionally stressful (McCarthy & Young, 2007). The individual does not know when the surgery will occur or even if it will happen, and must respond immediately to the call indicating a donor heart is ready. There is no warning period; the person's health may deteriorate while on the list for a donor heart.

When successful surgery is accomplished, the person is given immunosuppressants to prevent the body from rejecting the donor heart. Long-term exercise training after transplantation in a cardiac rehabilitation program increases exercise capacity. Three to six months is necessary to become sufficiently functional for work (Kobashigawa et al., 1999; Paris et al., 1997).

## **Functional Limitations**

Functional limitations of people with cardiovascular disorders depend on the severity of the condition and whether medical treatment alleviates the symptoms. Individuals with mild congestive heart

failure controlled by medication may be able to continue usual work with minimal modifications. Workers performing physically arduous jobs usually need changes in employment. Individuals with moderate heart failure who are limited to light or sedentary work may seek rehabilitation counseling services. Persons with severe conditions have extremely limited capacities for physical exertion and employment; some require cardiac transplantation, for which there is a lengthy waiting list. Exercise testing provides the level in METs where a patient becomes symptomatic with dyspnea; a poor heart rate response at that point suggests a negative prognosis.

### ***Disability Rating***

The treating physician determines the extent of disability resulting from coronary artery disease after carefully evaluating data obtained from a medical history, objective testing documentation, and response to treatment. Historical information is correlated with the NYHA classifications (see Figure 1) to determine symptomatic as well as therapeutic classification of the person's status, and in assessing ability to perform various activities (Alpern, 1996).

The NYHA classification system is one of the most helpful clinical determinants in establishing levels of cardiac disability. Physicians utilize these guidelines to classify patients on both functional and therapeutic levels (see Figure 1). Factors relating to disability are compared with objective evaluations, which are then correlated with the results of exercise testing on a treadmill. The cardiologist performing an exercise test estimates the energy expenditure of physical activity in terms of oxygen consumption required to accomplish certain tasks.

The basic metabolic unit for this estimation is the MET (metabolic equivalent). Studies have established MET requirements for various activities, both occupational and recreational. Workload is expressed in METS, which corresponds to the amount of oxygen required to perform a given activity. For example, a patient performs a Bruce Protocol exercise level of nine minutes (on a treadmill). This individual could be expected to perform up to 10 METs activity, indicating ability to perform mid-heavy to heavy activity by the NYHA, Functional Class 1 and Therapeutic Class A. The Bruce and Naughton treadmill tests are used to determine functional classification of disabilities. Test levels of performance have been correlated with levels of physical activity that relate to a variety of job performance (Alpern, 1996; Braunwald, 2005).

Information obtained from the patient's history and objective data from stress treadmill testing and other laboratory and physical findings are used to establish a classification of impairment and determine an optimal level of physical activity (Alpern, 1996). Goals of treatment are to decrease existing impairment and improve functional capacity to help the person establish a meaningful and productive life.

### ***Physical Limitations***

Most individuals, depending on the extent of cardiovascular disease, can perform at least light work. Some can engage in medium level work activity. Heavy lifting and carrying is usually discouraged, while moderate activities, such as standing and walking, are considered beneficial. The physician recommends physical exercise as part of the recovery process. Exercise testing reveals the exertion limit in METs so that a decision may be made as to possibility of employment and type of work. Undue emotional stress is contraindicated.

Chest pain (angina pectoris) occurs in some people with cardiac disease. This involves severe pain and a feeling of constriction around the heart area, including radiation of pain down the left arm. Angina is caused by an insufficient supply of blood to a portion of the heart and may occur with physical overexertion. Pain is usually transitory and relieved by medication (nitroglycerine), taken orally, which dilates the arteries. After angina pectoris, the person may resume work activity, usually in a few minutes, but is cautioned to avoid the activity that precipitated the event. A rehabilitation counselor needs to provide vocational guidance in accordance with the person's physical capacity. If angina is not relieved with medication, angioplasty or coronary bypass surgery may be recommended (Skala et al., 2005).

Some people experience arrhythmias and heart palpitations (throbbing sensations) in the chest area. These sensations may occur because of excessive physical activity. In all cases, the counselor needs to



inquire about client compliance with prescribed medications. The cardiovascular system is affected by environmental extremes of temperature. Poor air quality must be avoided by many with cardiovascular disease. A counselor's awareness of this and other factors in the work environment helps promote successful rehabilitation.

### ***Psychosocial Limitations***

The experience of hospitalization and separation from home and family produces emotional stress. Early intervention can decrease this stress by helping the person cope in a more realistic manner. Fear, anxiety, and depression are common emotions felt by persons with any chronic condition, including cardiovascular disease (Livneh & Cook, 2005). The impact these emotions produce on one's life ranges from minimal to devastating. Emotional support and counseling can help individuals cope with anxiety and depression and adjust to daily life with a disability. Denial, a normal psychological defense, is often part of the adjustment to life with a disability. If this denial causes a person to ignore symptoms, it may be dysfunctional and lead to selection of an inappropriate rehabilitation plan.

Today, treatment emphasizes early ambulation and involvement in a cardiac rehabilitation program as preventive measures to help avoid immobilizing reactions. With modern advances in technology and medical treatment along with rehabilitation intervention, one minimizes symptoms of psychological distress. As a result of modern treatment, most people regain strength and functional capacity to return to being productive and gainfully employed (Kobashigawa et al., 1999; Wadsworth, Smiling, & Lewis, 2002).

## **Rehabilitation Potential**

Multidisciplinary cardiac rehabilitation programs have assisted persons with cardiovascular problems in increasing performance during work, recreation, and leisure (McCarthy, & Young, 2007; Paris et al., 1997). Improved physical functioning and enhanced mental and social capacities aid in facilitating an active and productive life. A multidisciplinary treatment approach allows persons with cardiovascular problems to achieve increased awareness of their underlying conditions, and participate in exercise as a preventive and therapeutic measure. With the cardiac rehabilitation programs now available, individuals are able to increase strength, endurance, and improve overall functioning and psychological outlook. By educating the patient in the various manifestations of cardiovascular disease, the counselor can improve rehabilitation potential. Many persons with cardiac problems do not understand their condition, its severity, or their functional limitations. Some refuse to regularly and consistently take prescribed medications. Denial may play a part in this, along with the misconception that a lack of symptoms means medications are no longer necessary.

Rehabilitation potential for persons with Functional Class I and Therapeutic Classes A and B (see Figure 1) is excellent; these individuals have only slight restrictions in overall activities. Most do not require rehabilitation counseling services. Counselors see persons with Functional Classes II and III and Therapeutic Classes C and D, who need to modify their work activity to become reemployed. The amount of work adjustment necessary depends on the extent of cardiovascular disease and the physical and emotional components of the person's work activity. Individuals with Functional Class IV and Therapeutic Class E display such severe symptoms that their potential for rehabilitation is minimal.

Factors that may negatively influence return to work include: (a) duration on temporary disability, (b) perception of an inability to work, and (c) availability of disability income benefits. Rehabilitation potential is best for an individual who completes a cardiac rehabilitation program and feels positive about the results of therapy and the future (Paris et al., 1997). Development of appropriate educational and vocational goals consistent with physical and emotional restrictions is essential. Vocational rehabilitation ranges from returning to a former job with the same employer and minimal modifications, to establishing entirely new vocational objectives including a new occupation and occupational setting.

Investigation of job modification through reasonable accommodation helps both employee and employer maintain a productive work situation. One must consider both physical and emotional stress factors present in the employment environment, especially because emotional components are more difficult to define. Each person perceives emotional stress differently.



**Figure 3**  
**Approximate Energy Cost of Common Vocational and Recreational Activities**

METs	Functional Capacity	Activity
1.5 to 2.5	Sitting; light	Typing, driving a vehicle, using hand tools, light assembly, operating levers, machine sewing, crane operation, walking on a level surface at two mph, light carpentry, light automotive repair, washing dishes, desk work, carrying objects (15-30 pounds).
2.5 to 3.5	Standing; light to medium	Light assembly, use of hand tools, scrubbing, waxing, polishing, assembly, repair of machine parts, light welding, woodworking, interior carpentry, power sanding, stocking shelves, light to medium assembly-line work, cranking dollies, hitching trailers, wiring a house, bricklaying, plastering, house painting, paperhanging, raking, cleaning windows.
3.5 to 5	Walking	Attending vehicles at a gasoline station, carrying trays or dishes, dancing, gardening, making beds, mowing lawns.
5 to 7	Moderate	Climbing stairs slowly, exterior home building, shoveling dirt, using pneumatic tools, plumbing, using a wheelbarrow, carrying objects weighing 30-60 pounds.
7 to 9	Heavy	Sawing, shoveling, digging ditches, climbing stairs at a moderate speed, carrying objects weighing 60-90 pounds.
>9	Very heavy	Carrying loads upstairs, climbing stairs quickly, carrying objects weighing more than 90 pounds.

Source: Wackers, F. J. T. (1992). *Heart book*. New Haven, CT: Yale University, School of Medicine; adapted from Haskell, W. L. (1992), *Design and implementation of cardiac conditioning programs*. In N. K. Wenger, & H. K. Hellerstein (Eds.), *Rehabilitation recovery of the coronary patient* (3<sup>rd</sup> ed.). New York: Churchill Livingstone. Reprinted with permission.

A final dimension in the rehabilitation of persons with cardiovascular disease is employer discrimination (Brodwin, Parker, & DeLaGarza, 2003). Rehabilitation counselors can help educate employers and dispel negative stereotypes employers may possess toward persons with chronic disease. As stated by Yunker (1992), "employers value employees who have job skills, social skills, and dependability" (p. 17). Employers who have had past experiences with employees who have disabilities usually develop positive attitudes toward them. As more people with disabilities return to or enter the labor force and prove to be productive employees, we can expect further positive attitude change. Counselors play a crucial role in initiating and continuing this process.

## Case Study

Joyce Albert is a 52 year-old married Korean American woman with multiple disabilities, including coronary artery disease, hypertension, and diabetes. One year ago, after being diagnosed with coronary artery disease, she had successful heart bypass surgery, and does not currently require medication for her condition. Ms. Albert has never participated in a cardiac rehabilitation program. In addition to coronary disease, she has been successfully controlling diabetes with insulin for the past 10 years; she keeps her hypertension under control with medication. Additionally, Joyce lives with mild to moderate anxiety and depression.

For nine years before attending graduate school in social work, Ms. Albert worked as a self-employed photographer. This involved lifting and carrying up to 40 pounds of equipment, long hours, and frequent traveling to different locations. Before becoming self-employed, Ms. Albert worked for

various photographic studios, starting out as a photography assistant and working her way up to her current position. Joyce left each job within one year because she felt that none of her managers were satisfied with her work, though the managers noted satisfaction with her work on several occasions.

Joyce's first position following receipt of her M.S.W. was as a children's social worker. After two years, she left this position, because she believed her clients were never satisfied with her services. Her supervisors, however, consistently rated Joyce's work as excellent. She next obtained employment as an eligibility worker, but resigned after one year due to complaints from a client. The supervisor attempted to keep Ms. Albert as an employee by reassuring her that complaints were common among all eligibility workers; nevertheless, she left the job. Following these social work positions, Joyce decided to leave the field to secure employment as a photography assistant.

Ms. Albert was evaluated using an exercise study. She successfully accomplished five METS. There was no evidence of ischemia on a perfusion nuclear study at each exertional level. Her physician restricted Ms. Albert to a maximum of medium work with no undue emotional stress.

### **Questions**

1. Provide a vocational profile for Ms. Albert.
2. Mr. Alpert feels his wife should be seeing a Korean, female counselor. You are neither Korean nor female, and have been assigned the case. Although there is a Korean, female counselor in your agency, your supervisor wants you to keep the case. Discuss whether you will work with this client and the cultural and interpersonal implications involved with this situation.
3. What are Joyce's functional limitations that may effect employment?
4. Discuss the psychosocial implications of this case.
5. Outline three possible rehabilitation plans for this client.
6. Are there any additional services that may help this individual return to work? Explain.

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## Chapter 7

# CHRONIC OBSTRUCTIVE PULMONARY DISEASE AND NEUROMUSCULAR DISORDERS AFFECTING THE RESPIRATORY SYSTEM

*Ahmet Baydur, M.D., FACP, FCCP*

### Introduction

Chronic obstructive pulmonary disease (COPD) is defined as “a preventable and treatable disease with some significant extrapulmonary (systemic) effects that may contribute to the severity in individual patients. Its pulmonary component is characterized by airflow limitation that is not fully reversible. The airflow limitation is usually progressive and associated with an abnormal inflammatory response of the lung to noxious particles or gases” (Rabe et al., 2007, p. 532). In addition to airflow obstruction, individuals with COPD can exhibit cough, excessive mucus production and shortness of breath or dyspnea. In many cases, an individual patient with COPD can manifest symptoms of asthma (a reversible form of airway obstruction), bronchitis, and emphysema which is why these conditions are usually considered together. The American Thoracic Society/European Respiratory Society (ATS/ERS) guidelines specify that “asthma differs from COPD in its pathogenic and therapeutic response and should therefore be considered a different clinical entity” (Brusasco, Crapo, & Viegi, 2005; Kreider & Grippi, 2007).

COPD is the fourth leading cause of death in the United States. Estimates of the nationwide prevalence of COPD range from 14 to 20 million cases and continues to increase. COPD is second only to coronary heart disease in the number of individuals receiving Social Security disability payments for chronic disease.

Neuromuscular diseases, while less common in prevalence than asthma and COPD chronic obstructive airway disorders, comprise a significant cause of respiratory impairment and failure; these diseases account for a major source of disability and prolonged respiratory care in the United States. The basic premise of rehabilitation for patients with COPD is a return to as self-sufficient a role in society as feasible. By spreading awareness and highlighting the importance of appropriate selection, individuals with respiratory conditions can benefit greatly from vocational rehabilitation. Also important to the success of rehabilitation are the following factors: age, nature and stage of illness, overall health, presence of medical complications, possibilities for retraining, the current labor market, and return to work.

The purpose of this chapter is to provide an overview of asthma, COPD common chronic obstructive respiratory disorders, as well as respiratory impairments found in patients with neuromuscular diseases. Definitions, diagnostic features, physical limitations, treatment, and rehabilitation potential for these conditions are described.

## Chronic Obstructive Pulmonary Disease (COPD)

### *Introduction*

COPD is a general term for a group of diseases (i.e., asthma, chronic bronchitis, and emphysema) that can cause progressive damage to the lungs. This disorder is characterized by subnormal tests of expiratory flow from the lungs over several months of observation (American Thoracic Society, 1986). The

airflow limitation is usually progressive and associated with an abnormal inflammatory response of the lungs to noxious particles or gases. Although COPD affects the lungs, it also produces systemic problems throughout the body (Brusasco et al., 2005), including wasting, fatigue, and serious psychosocial consequences. Chronic bronchitis and emphysema are two major disorders usually included within the definition of COPD. Asthma is a type of COPD, characterized by reversible airway hyperactivity; with this condition, airflow improves following inhalation with a bronchodilator.

In asthma and COPD, inflammation, bronchoconstriction, and structural changes in the small airways (the major site of airflow limitation in COPD) and lung tissue are considered to be the most important contributors to airflow limitation and the rapid decline in lung function. Airway remodeling, a kind of scarring that occurs immediately underneath the mucus lining of the airways, may contribute to the development of these diseases. Though most cases of COPD are a direct result of cigarette smoking, recent studies indicate that inflammation persists in susceptible patients long after they have quit smoking (Hogg, Chu, & Utokaparch, 2004). Airway remodeling has been suggested as a target for treatment and prevention, although, to date, drugs used to treat airway diseases have little influence on airway structural changes. Research continues on medicines that may actually reverse or prevent airway remodeling (Bergeron & Boulet, 2006).

## ***Pathology and Symptomatology***

### ***Asthma***

Approximately 14 million adults in this country live with asthma, a condition characterized by an inflammation of the main air passages of the lungs and bronchial tubes (Petechuk, 2004). In patients with asthma, the muscles of the bronchial walls tighten and cells in the lungs produce extra mucus, further narrowing the airways. This can cause symptoms ranging from minor wheezing to severe difficulty breathing. Other symptoms include coughing, shortness of breath, chest tightness or pain, and increased need for bronchodilators (medicated inhalers that open airways by relaxing the surrounding muscles). In some cases, breathing may be so labored that a single asthma attack becomes life-threatening. Asthma is a chronic, but treatable condition (Bourke, 2003).

The direct cause of asthma is still unknown, but the disease is at least partially due to a combination of environmental and genetic factors. Exposure to various allergens and irritants may trigger symptoms of asthma, such as smoke, strong odors, cold air, respiratory infections, physical exertion, stress, and strong emotions. Certain medications, including beta blockers, aspirin, and other nonsteroidal anti-inflammatory drugs also trigger or worsen an asthma attack (Global Initiative for Obstructive Lung Disease [GOLD], 2007).

### ***Chronic Bronchitis***

While acute bronchitis originates mostly from viral infections, chronic bronchitis is primarily associated with cigarette smoking, which accounts for 80% – 90% of all cases of chronic bronchitis (Petechuk, 2004). The persistent inhaling of tobacco smoke damages cilia fibers which line the respiratory track. Damaged cilia lead to a decreased ability of the body to fight germs and infections that enter the respiratory track. Chronic bronchitis is a serious disease of prolonged inflammation or infection of the bronchial tubes that results in the swelling of mucous membranes lining the bronchi, increased mucus production, and impaired mucus transport by the cilia. Because the diameter of the bronchi is decreased, ventilation also is impaired (GOLD, 2007). Common signs and symptoms of chronic bronchitis are similar to those of an upper respiratory infection and include coughing, wheezing, sore throat, congestion, breathlessness, chest constriction, and fatigue.

### ***Emphysema***

Of the more than 16 million Americans with COPD, nearly three million have symptomatic emphysema. Millions more are in early stages of the disease long before signs and symptoms appear (Petechuk, 2004). Cigarette smoke is the single most common cause of emphysema. When a smoker inhales, the tobacco entering the lungs temporarily paralyzing the microscopic hairs (cilia) lining the bronchial tubes. The result is a loss of elasticity in the walls of the small air sacs in the lungs. Eventually, the walls stretch and break, creating larger, less efficient air sacs that cannot handle the normal exchange of oxygen



and carbon dioxide. In advanced cases of emphysema, breathing can consume up to 20 percent of a person's resting energy. Emphysema develops over many years until irreversible damage occurs. The main symptoms of emphysema are shortness of breath and a reduced capacity for physical activity, both of which are likely to worsen as the disease progresses. Other signs and symptoms of emphysema include enlarged chest size (barrel chest), chronic cough, fatigue, poor appetite, and weight loss (GOLD, 2007).

The causes of cachexia (wasting) in patients with COPD are many and include decreased oral intake, increased work of breathing due to abnormal respiratory mechanics, and chronic systemic inflammation and comorbidities (King, Cardova, & Scharf, 2008). It remains unknown whether long-term weight gain by using enhanced caloric intake, with or without using anabolic steroids or appetite stimulants, improves survival or provides other benefits to individuals with COPD. Many other conditions are associated with increased risk of, or share the same risk factors (smoking, obesity, and aging) for developing COPD, including cardiovascular diseases, lung cancer, osteoporosis, chronic renal failure, rheumatoid arthritis, psoriasis, and premature aging (Luppi, Franco, Begh , & Fabbri, 2008).

## ***Diagnostic Evaluation***

### ***Clinical Assessment***

Clinical symptoms of COPD include cough, sputum production, sneezing, and dyspnea (shortness of breath). Each symptom usually illustrates the presence of one or more resulting complications. Generally, chronic cough and sputum overproduction suggests the presence of bronchitis. Wheezing usually indicates the presence of bronchospasm and airway hyperreactivity and often suggests airways obstruction that is reversible (Bergeron & Boulet, 2006). Decreased breath sounds reflect reduced peripheral airflow due to lung tissue destruction (as occurs with emphysema). Tachypnea (rapid breathing), use of accessory respiratory muscles, and intercostal retraction indicate excessive breathing distress. These symptoms may appear alone or in conjunction with other symptoms, depending on the predominant type of abnormal physiology found in the patient.

### ***Assessment of Pulmonary Function***

Following the medical history and physical examination, the next step in diagnosis consists of obtaining objective measurements of airflow and lung volume. Spirometry, the most effective way to measure breathing capacity, is a test in which the patient blows into a machine (spirometer) with as much effort as possible. The machine measures both the expelled amount of air (called vital capacity, VC) and the speed of expiration of the air (forced expiratory volume in one second, FEV<sub>1</sub>). Graphs based on age, gender, and height provide the most ideal comparisons between the patient and the normal population, allowing a physician to quickly diagnose the condition. Traditionally, the severity of this disease is graded according to FEV<sub>1</sub> (GOLD, 2007). However, other factors including the body mass index (BMI), degree of dyspnea, and exercise capacity have been shown to independently predict outcome (Celli et al., 2004).

COPD is associated with hypoxemia, a condition characterized by insufficient oxygenation of the blood found in conjunction with a high count of red blood cells known as secondary erythrocytosis. Persons with these conditions should be considered for possible oxygen therapy. When considering the need for this therapy, an exercise test is performed to determine an individual's capability for a particular activity or occupation. Electrocardiographic (ECG, EKG) tests also show problems such as evidence of right ventricular enlargement of the heart muscle, which suggests the presence of pulmonary hypertension (Hlastala & Berger, 2001). In November of 2005, the American Thoracic and European Respiratory Societies jointly published a statement proposing a new interpretation scheme for pulmonary function tests. This new scheme leads to a diagnosis of obstructive pulmonary disease in a greater proportion of patients undergoing pulmonary function testing (Kreider & Grippi, 2007).

## ***Management of COPD***

### ***Prevention***

Of primary importance is avoidance of bronchial irritants, such as cigarette smoke and other environmental pollutants. Physicians recommend immunization against influenza and pneumococcus (a

common bacterium causative of exacerbation of symptoms) in people with chronic respiratory disorders, especially those over 60 years of age.

### ***Medical Treatment***

The principal treatment for patients with COPD involves the use of bronchodilators, medications that decrease airway reactivity, and reverse bronchospasm. Bronchodilators are given orally or by aerosol inhalation (GOLD, 2007). Corticosteroids are used in acute exacerbations of COPD, while inhaled corticosteroids in combination with long-acting bronchodilators improve outcomes, including survival (Calverley et al., 2007). Candidates for oxygen therapy include those who have low levels of oxygen, clinical or electrocardiographic evidence of pulmonary hypertension, cor pulmonale (right heart enlargement and failure), or increased hemoglobin concentration. With treatment such as oxygen therapy, prognosis for individuals with COPD and hypoxemia improves. Because the best results are achieved when oxygen is used 24 hours a day, ambulatory patients are often provided with portable oxygen delivery systems (Nocturnal Oxygen Therapy Trial Group, 1980).

### ***Lung Volume Reduction Surgery***

The surgical approach to treatment of emphysema is improving through the use of modern techniques and technology. Treating emphysema surgically includes the excision of large bullae (bullectomy) and resection of diffusely emphysematous lung. This treatment approach, commonly known as lung volume reduction surgery (LVRS), pneumonectomy, or reduction pneumoplasty, aims for a 20% to 30% reduction in lung volume and concomitant increase in vital capacity (VC), and is performed by surgical or laser resection, or a combination of both (Bourke, 2003). Despite the positive qualities of LVRS, questions remain regarding the duration of benefits, safety, and cost (Baydur, Gilgoff, Prentice, Carlson, & Fisher, 1990). LVRS is an alternative for individuals with severe upper lobe disease who remain symptomatic after optimal medical therapy (National Emphysema Treatment Trial Research Group, 2001).

### ***Lung Transplantation***

Damage to the structure of the lungs can affect the efficiency with which gases are exchanged. If an individual with poor lung function does not respond to medication or other methods of treatment or the severity of the condition is life-threatening, the physician may recommend a lung transplant. After enduring a long process of evaluation for eligibility and candidacy for transplant, if found eligible, the person's name is placed on a nationwide waiting list. Currently, about 3,500 people are waiting to receive lung transplants while the supply of donor organs is sufficient to provide only about 1,000 lung transplants annually (Bourke, 2003; Petechuk, 2004). Following a successful surgical transplant, the process still is not complete. The patient must undergo post-transplant treatment, a delicate balancing act between preventing rejection of the organ and managing unwanted side effects. Smoking is prohibited and alcohol use is strictly limited. The first year after the transplant is the most critical period; surgical complications, rejection, and infection pose the greatest threats during this time. For patients whose procedures are successful, lung transplantation improves many facets of life, from increased functioning to a significantly enhanced quality of life.

### ***Home Care for COPD Patients***

Health services may be provided to persons and their families in their homes to promote, maintain, and restore health, or to minimize the effects of illness and disability. Home care includes medical and dental treatment, nursing, respiratory care, physical therapy, speech therapy, occupational therapy, social work, nutrition, homemaker services, home health assistance (aides), transportation, laboratory services, and equipment and supplies (American Thoracic Society, 1995). The following are goals of home care:

1. Improve quality of life by allowing patients with advanced disease to remain in their environment and be with family and friends.
2. Minimize or prevent complications that require hospitalization.
3. Detect changes in physical and psychosocial status that indicate a need for modification in disease management.
4. Provide treatment for the person's primary diagnosis and foster adherence to the therapeutic program.
5. Foster a positive and independent attitude.

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### ***Home Mechanical Ventilation***

Patients with complex treatment programs, such as home ventilator care, require direct supervision by a home health care agency. Because care of tracheostomy-ventilator assisted patients in a hospital setting is extremely expensive, home management may be a safe alternative. Most patients prefer the home environment to living in a hospital. Presently, the principal role for mechanical ventilation in the home is management of patients with neuromuscular disease who have respiratory failure (Hlastala & Berger, 2001). Patients with severe COPD are rarely suitable candidates for home mechanical ventilation because complicated and frequently unstable medical problems often arise, and management of these patients in the home is usually found to be impractical and unsafe (Esau, Truwit, & Rochester, 1995).

### ***Respiratory Impairment and Disability***

In 1966, Gaensler and Wright published a widely accepted distinction between the terms “impairment” and “disability.” According to these authors, disability determination consists of assessing a person’s capacity to function in a specific manner compared with the required level for a specific performance. Evaluation of “capacity” in this sense requires consideration of factors other than health (i.e., education and age). Failure to recognize that “capacity” involves more than just the person’s medical impairment or physical abilities can lead to misunderstandings between rehabilitative physicians and administrative agencies.

By contrast, impairment implies that the capacity for a specific function has become less than the person previously possessed. Since the physician usually does not know the individual’s prior capacities, comparisons are made with normal statistical figures. Due to the wide variation of capacities for most specific functions, lung function testing has greater accuracy for depicting general capacity than for determining the presence or absence of a precise degree of impairment (Gaensler & Wright, 1966). While the rating of respiratory impairment is within the province of a physician’s expertise to quantify, the determination of disability is an administrative and medical decision that requires consideration of many non-medical and medical variables (American Thoracic Society, 1986).

### ***Rehabilitation of Patients with COPD***

Most studies investigating the effect of pulmonary rehabilitation on healthcare utilization have demonstrated a significant reduction in emergency room and physician office visits, as well as hospital inpatient days for patients with COPD. Generally, there has been a consistent reduction in patients’ hospital stays over the years following their initiation into rehabilitation programs (Tiep, 1997). Rehabilitation professionals are thus in a position to make a significant impact on not only the psychosocial and vocational aspects of pulmonary rehabilitation, but the well-being and daily life of people with COPD.

#### ***Comprehensive Care Program***

In chronic, progressive respiratory disorders, comprehensive care programs that use multidisciplinary treatment approaches for the various aspects of the disease achieve greater success when compared to treatment methods utilizing single disciplines. Ample evidence shows that patients who participate in multidisciplinary programs experience improved well-being and decreases in hospitalizations. Several authors have described the key components of comprehensive care programs as follows (Celli, 1995b; Ries et al., 2007; Tiep, 1997; Troosters, Casaburi, Grosselink, & Decramer, 2005):

1. Patient and family education.
2. Treatment of bronchospasm by bronchodilators or reduction in bronchial secretions.
3. Treatment of bronchial infections.
4. Right heart failure treatment.
5. Oxygen therapy.
6. Chest physical therapy, including breathing technique training.
7. Exercise reconditioning.
8. Psychosocial management, including vocational rehabilitation.

Breathing exercises improve respiratory muscle efficiency by increasing a person's control of expiratory flow and respiratory rate. Enhanced respiratory efficiency decreases shortness of breath and may improve tolerance for exercise. This regimen requires the individual to cooperate and remain motivated. When breathing exercises are performed regularly over several months, evidence of increased inspiratory muscle endurance becomes apparent.

The rehabilitation process includes a continuing program of education. For example, the patient must know the schedule of each medication and understand its purpose. Sharing information with patients and families allows for the formulation of constructive life plans. In this manner, the patient and family become active members of the health care team.

### ***Pulmonary Disability***

The antecedents of pulmonary disability due to COPD are based either on reversible corrective changes in the pulmonary system (as in asthma and chronic bronchitis) or on non-reversible, anatomic, structural changes (as in emphysema). The consequences of these alternatives are dependent on three basic interactions (Plummer, 1984):

1. The interaction between the patient and the physical environment.
2. Social interaction with significant others and treating personnel in contact with the patient.
3. The transactional results of the varied interactions involved in the disability and therapeutic interventions developed to help the patient and family cope.

### ***Vocational and Rehabilitation Aspects of COPD***

#### ***Team approach***

Successful vocational rehabilitation of patients with COPD can be achieved through effective participation of all members of a rehabilitation team. The team includes the rehabilitation program physician whose primary goal is to increase the activity level of the patient to an optimal level. Working along with the medical team members are the occupational therapist, social worker, rehabilitation counselor, and job placement specialist. The ideal time to begin vocational exploration is during psychosocial questioning when the patient may still be able to return to prior employment, with or without reasonable accommodation.

Unfortunately, some patients are subject to severe respiratory infections frequently requiring hospitalization. This may result in a patient's inability to cope with regular employment. It causes a disruption of socioeconomic status that imposes significant psychological impact on the individual and family, often leading to feelings of despair and uselessness (Mahler, 1998).

#### ***Vocational factors***

Information is obtained from the patient regarding personal-social background and educational-vocational history. The history, explored in depth by the counselor, needs to include all prior work, job duties, dates of employment, salaries, job satisfaction, reasons for termination of employment and a transferable skills analysis, when appropriate. Information about present means of financial support, educational background, as well as social and psychological factors is taken into consideration. Rehabilitation professionals, coordinators of the multidisciplinary team and liaisons with community agencies, make a significant impact on the psychosocial and vocational aspects of pulmonary rehabilitation.

Persons with COPD may be classified into four clinical and vocational rehabilitation groups (Hodgkin, 1981):

1. Those who can return to their previous work activities.
2. Those who should be retrained for more suitable work.
3. Those who are capable of work only in sheltered employment.
4. Those who can be trained for self-care only.

Proper evaluation and categorization of the patient's capacities are essential to successful educational and vocational rehabilitation. When selecting patients for vocational rehabilitation, counselors need to consider such factors as recent changes in lifestyle and evidence of rapid clinical deterioration, personality change, substance abuse, and psychological or social dysfunction (Brodwin, Parker, & DeLaGarza, 2003). Counselors must additionally assess whether a person's cardiorespiratory reserve will enable a return to a previous job on a full or part-time basis. If the individual's tolerance to work requires job modification, the rehabilitation counselor can help facilitate this process. Once the patient is physically rehabilitated, training in new vocational areas commensurate with educational level and decreased respiratory capacity may be needed.

### ***Sheltered or Home Employment***

Some people who have COPD can only function in locations where special respiratory equipment is available, such as in sheltered employment or at home. Occupational therapists, rehabilitation counselors, and job placement specialists work together to help locate settings for employment. The occupational therapist provides training in energy-saving methods for those who have a difficult time with self-care. This helps the individual perform tasks including housekeeping, shopping, and other activities of daily living with less dependence on others. Training methods taught by an occupational therapist include (Hodgkin, 1981):

1. Perform activities slowly.
2. Avoid noxious fumes.
3. Avoid areas of excessive heat, cold, and humidity.
4. Plan ahead to decrease ambulation and minimize body movement.
5. Change activities frequently.
6. Spread activities that promote fatigue or dyspnea throughout the day.
7. Transport heavy objects using carts or tables with wheels.
8. Perform most work activities while standing with the work surface (table) at hip level to minimize flexion and abduction.
9. Perform most work activities while standing with the work surface at a body level that minimizes trunk flexion and hyperextension.
10. Store utensils, tools, and similar devices in cabinets where height minimizes active trunk flexion, hyperextension, and active flexion of the shoulder girdle.
11. Use electric appliances (can opener, mixer, electric knife) to eliminate or minimize manual performance of daily activities.

Several factors impede vocational rehabilitation. These include psychosocial problems, severity of respiratory impairment, progression of disease, advancing age, limitation in skills and capabilities, and poor labor markets in certain fields.

## **Neuromuscular Disease Affecting the Respiratory System**

Pulmonary complications, including respiratory failure and aspiration pneumonia, occur commonly in individuals with neuromuscular diseases. A patient with end-stage neuromuscular disease is typically not able to adequately ventilate the lungs that have become vulnerable not only by weakened muscles of respiration, but also by recurrent respiratory complications. Neuromuscular diseases are generally described by the motor unit they affect and can be categorized as (Ringel & Carrol, 1980):

1. Diseases of the anterior horn cell (neuronal disorders), such as spinal muscular atrophies, amyotrophic lateral sclerosis (Lou Gehrig's Disease), and poliomyelitis.
2. Neuropathies due to metabolic, endocrine, toxic, hereditary, immunologic causes, and vitamin deficiencies.
3. Diseases of the neuromuscular junction, such as myasthenia gravis and botulism.



4. Muscle diseases (myopathies) such as muscular dystrophy, myotonic disorders, inflammatory myopathies (polymyositis, dermatomyositis), metabolic myopathies, and toxic myopathies (caused by alcohol and certain drugs).
5. Spinal cord injuries and related disorders are discussed elsewhere in this book.

### ***Pathophysiology of Respiratory Muscle Weakness***

After the severe poliomyelitis epidemics of the 1940s and 1950s, it became necessary to have objective criteria for starting treatment with a respiratory aid and to guide subsequent weaning from a respirator. In neuromuscular disease, vital capacity, maximal inspiratory and “sniff” pressures decrease with disease progression, and are key in assessing respiratory impairment and the need for assisted ventilation. Weakness of the respiratory muscles commonly occurs and is prevalent in critical illnesses which occur from a lack of oxygen, malnutrition, and a number of metabolic abnormalities (Baydur, 1988; Esau et al., 1995). In spinal cord injury, lung function is closely related to the level of injury (Baydur, Adkins, & Milic-Emili, 2001). Respiratory muscle weakness leads to impaired cough ability, retention of airway secretions, and increased risk for aspiration pneumonia, a common cause of morbidity and mortality in these individuals. Muscular weakness in chronic illness, for example, is related to malnutrition and cachexia from chronic infection and cancer. Kyphoscoliosis and chest wall restriction, often a consequence of chronic neuromuscular disease, not only impose mechanical disadvantage, but also predispose the person to respiratory impairment and failure. Pulmonary hypertension occurs as a result of progressive loss of lung volume and oxygen deficit, leading to right-sided heart failure.

### ***Therapeutic Approaches to Respiratory Failure in Patients with Chronic Neuromuscular Disease***

#### ***Resting the Respiratory Muscles: Assisted Ventilation***

Rehabilitation counselors need familiarity with home care ventilation products, as thousands of patients with chronic respiratory failure return to the community needing such services. Assisted ventilation therapy, popular during the poliomyelitis epidemics, is still the most effective method to allow respiratory muscles to return to normal functioning levels (Baydur, 1988; Baydur et al., 2000). While this is a logical approach during acute respiratory failure, intermittent rest therapy with ventilation for patients with chronic respiratory failure has only recently regenerated interest, especially for use in people with neuromuscular disorders. Mechanical ventilation dramatically reduces the oxygen cost of breathing in fatiguing muscles and dyspnea (Esau et al., 1995). Intermittent rest therapy in patients with chronic respiratory insufficiency has been successful when measured in terms of patient well-being, improving patients’ sleep, and eliminating the common problem of morning headaches. Patients using intermittent rest therapy report feeling more alert during the day and are able to complete daytime tasks with less fatigue (Bach, 1994; Baydur et al., 1990). The decline in lung function is often slowed or may even stop.

Researchers (Baydur, et al., 2000; Esau et al., 1995; Hess & Kacmarek, 2002) discussed the criteria for chronic respiratory muscle rest therapy and described types of ventilators used in this method of treatment. According to such research, individuals who use these modalities maintain a higher level of independence and an improved quality of life at home. Assisted ventilation is cost effective when compared to hospitalization, averaging about one-third the cost of a critical care unit.

Adjunct techniques designed to help eliminate airway secretions in neuromuscular patients include cough assist methods (both by manual and machine-assisted techniques), chest percussive devices, and glossopharyngeal breathing. When used consistently by patients and care givers at home, they prevent pneumonia and avoid the need for tracheostomy (Bach 1993; Baydur et al., 2000).

#### ***Ventilatory Muscle Training***

Inspiratory muscles can be trained to increase their endurance and strength. While promising, the success rate of ventilatory muscle training remains limited. First, there is a need for persistent patient compliance. Second, for any given disease, it is unclear whether respiratory muscle strength or endurance should be enhanced. Cessation of respiratory training programs results in a return to pretraining testing values within about one week. Physicians generally do not recommend exercise of respiratory muscles in



degenerative neuromuscular conditions such as polio, Guillain-Barré syndrome, and amyotrophic lateral sclerosis (Celli, 1995a; Faulkner, 1995).

### ***The Role of Nutrition in Respiratory Muscle Fatigue***

Increased work of breathing associated with respiratory failure increases caloric demands that cannot be met, leading to loss of muscle mass, strength, and endurance. While nutrition is a factor in recovery from respiratory failure and muscle fatigue, consideration must be given to proper ratios of carbohydrates, proteins, and fats. Several research studies suggested that appropriate nutritional support can successfully wean some patients from mechanical ventilation (Hess & Kacmarek, 2002; Mahler, 1998).

### ***Functional Limitations in Neuromuscular Disease***

There are two levels of concern regarding disability in neuromuscular disease. First is the functional limitation imposed by the physical impairment and resulting activity substitutions required. The second concern involves the gap between the person's functional ability and functional demands of work. Capacity to function requires consideration of multiple factors, including respiratory impairment, education, age, gender, economic circumstances, social environment, and energy requirements of a given occupation (American Thoracic Society, 1986, 1995; Gaensler, & Wright, 1966). Two people with identical respiratory impairments (based on lung function) are affected differently in their life situations, including ability to perform specific work tasks.

Those with little or no direct experience with long-term care of ventilator-dependent patients often raise the question of "quality of life" for these individuals. This problem is most often encountered with parents of the Duchenne muscular dystrophy patient who is approaching respiratory decompensation (Baydur et al., 1990; Gilgoff, Prentice, & Baydur, 1989). Yet, these young individuals adapt to a respirator and wheelchair more readily than older patients. By contrast, older patients with long-standing respiratory impairment, who are slowly developing respiratory failure and a need for assisted ventilation, require special sensitivity and tact. For example, post-polio survivors have managed their care over the past 50 years with independence from health professionals, except when in acute crises (Baydur et al., 2000). When their margin of reserve has decreased, the specter of a tracheostomy recalls the dark days of the 1940s and 1950s, when many of these people experienced emergency tracheostomies and iron lungs. Seeing many others around them dying, they resist the recommendation for a tracheostomy years later, a recommendation that causes them to relive the experience.

Nasally or orally applied intermittent positive pressure (noninvasive) ventilation is a common method used to provide a noninvasive means of assisted ventilation (i.e., without a tracheostomy) in those patients with marginal respiratory function (Bach, 1993; Bach, 1994; Hess & Kacmarek, 2002). Such modalities are helpful in those patients with associated sleep apnea syndrome. Sleep-disordered breathing often occurs in patients with symptoms of nocturnal insomnia, daytime somnolence, and elevated daytime arterial carbon dioxide tension levels. Sleep-disordered breathing can be associated with physical problems like high blood pressure and psychological disturbances, such as anxiety, irritability, loss of concentration, and depression.

### ***Vocational Limitations Resulting from Neuromuscular Disease***

Depending on the nature and extent of paralysis, respiratory and global muscle involvement results in a wide spectrum of vocational limitations. For patients with physical impairments, activities such as ambulation and employment present major challenges. Disability is reflected by fatigue from overuse of muscles, a noticeable limp, slowness of gait or inability to walk, and dyspnea (shortness of breath).

As with chronic obstructive pulmonary disease, shortness of breath is assessed first by taking a detailed history. Such a history includes documentation of specific activities leading to shortness of breath; intensity, type, and duration of exercise causing these episodes; and time required for recovery. The physician notes the influence of circumstances and emotions, posture, unusual sensations, variations of daily weather, and seasonal changes. Other influencing factors are recorded, such as degree to which shortness of breath interferes with employment or recreation, approximate date of onset, progression, or regression in severity since onset, and whether shortness of breath occurs even during rest (Gaensler & Wright, 1966).

The distress associated with work experienced by most individuals with severe pulmonary impairment, including those with neuromuscular disorders, is related to several activities. These include prolonged duration of low-level activity; no facilities for rest on the job; and the exertion, frustration, and discomfort experienced in travel to and from work. Other problems are motivation and attitude. People desiring rehabilitation tend to perform to the best of their abilities when asked to do graded exercise tests. By contrast, individuals claiming total disability and financial compensation are less motivated to exercise to maximum potentials.

Among groups studied, polio survivors have the highest rate of employment (Bourke, 2003). They are highly motivated to raise families and maintain full time employment. Many have attended college and received advanced degrees. Because of their personal experience, they often function as counselors, sometimes for other people with neuromuscular disease. Yet, because of the muscle overuse syndrome, many have curtailed daily activities and reduce employment hours or decrease exertional levels of work, much to their distress. The situation for individuals with more progressive neuromuscular disorders is more devastating. An almost complete loss of muscle function and various other disturbances results in severe limitations.

### ***Rehabilitation Potential for Those with Neuromuscular Disease***

Chest rehabilitation in neuromuscular disorders is basically geared to reduce or relieve hypoxemia and hypercapnia, and to cope effectively with respiratory infections. Individuals use their own resources as much as possible to achieve these goals. In all conditions, after hospital discharge, patients practice procedures and techniques learned in the hospital. They are prescribed thoracic expansion breathing exercises; deep breathing exercises encourage inspiration of air and lung expansion. People who are hyperventilating require breathing exercises, such as diaphragmatic breathing and pursed lip breathing to help them achieve slower and deeper breathing patterns. Since this breathing technique may increase the work of breathing, the goal is to balance improvement in lung expansion against discomfort.

Reconditioning exercises, while important for patients with spinal cord injury, may present a problem for patients with motor neuron diseases and myopathies. Many physicians feel that muscle fatigue exacerbates further muscle tissue destruction and should be avoided. Patients with progressively debilitating muscle disorders, such as poliomyelitis, need to limit their daily activities to what is most comfortable. They should not push themselves beyond physical limits when engaging in work, leisure activities, and home life. If they already receive assisted (usually noninvasive) ventilation, particularly at night, they need to add time on ventilation during the day (Hess & Kacmarek, 2002).

Buchanan, LaBarbera, Roelofs, and Olson (1979) found that 76% of families with children with Duchenne's muscular dystrophy identified psychological issues to be the major problem confronting them. Madorsky, Radford, and Neumann (1984) highlighted psychosocial issues surrounding death and dying in this population; they found discrepancies in perceived services and needs. Families and patients overwhelmingly desired more training, education, and support when confronting terminal issues. Importantly, while only 22% of patients and 17% of parents reported having been offered counseling services, 74% of clinic staff regarded counseling as readily available.

To address these issues, Gilgoff et al. (1989) suggested having a special clinic available to work with patients and families at the time of impending respiratory failure. This clinic combines the skills of a pulmonologist, pediatrician, and respiratory nurse specialist, with the support of physical, occupational, speech, and respiratory therapists, and a psychologist. This approach has three primary objectives:

1. Educate the patient and family.
2. Institute elective ventilation before onset of acute respiratory failure for those patients who select this option, and provide ongoing support for these patients and families.
3. Provide psychological support to those patients and family members when they do not wish to receive assisted ventilation.

For patients choosing mechanical assisted ventilation, sequential following of the vital capacity and carbon dioxide levels provides a useful guide in predicting the need for mechanical ventilation before acute respiratory failure occurs. This allows respiratory support to be started early, making the transition to a

respirator less stressful and preventing the occurrence of an emergency situation and lengthy hospitalization. Sharing information with patients and families allows them to formulate constructive life plans. In this way, the patient and family can become active members of the health care team, rather than solely relying on a medical bureaucracy.

## Conclusion

Individuals with respiratory conditions will have limitations in stamina due to shortness of breath and limited respiratory function. Shortness of breath occurs with or without exertion, depending on the extent of impairment. Environmental conditions such as pollutants, dust, extremes of temperature, and rapid temperature changes can trigger exacerbations of asthma and COPD and further limit stamina. Problems with stamina may involve specific activities or be present for a wide range of functions. If full time work is not possible, the person might consider part-time work or home employment.

The worker may be able to trade more physically demanding activities for less strenuous ones through reasonable accommodation. Motorized carts or power wheelchairs diminish carrying of objects and ambulation. If the worker has a rotating work shift, a change to a standard shift lessens energy expenditure. The counselor can perform a work site assessment to determine what factors of the job need modification for a worker to continue functioning effectively (Brodwin et al., 2003).

## Case Study

Mr. Robert Smith, a 58 year-old supervisor in an automobile parts plant, was referred because of a complaint of progressive exertional dyspnea. He and his wife have raised a son, 30 years of age, and a daughter, 28 years old. The son is an assistant hospital administrator and the daughter is an elementary school teacher for a local school district. Six years ago, Mr. Smith's medical condition started, following an episode of pneumonia which was treated with antibiotics. Subsequently, Mr. Smith noted exertional shortness of breath while walking on level ground. Robert smoked cigarettes (two packs a day for 30 years) until six months ago. Past medical history includes several previous episodes of bronchitis. He has no allergies or cardiac disease. For the past 25 years, he worked at his job at an automotive parts plant, starting initially on the assembly line, and steadily rising up through the ranks to attain a supervisory position in the production department.

Physical examination and pulmonary function testing showed moderately severe airway obstruction consistent with emphysema. An exercise test showed moderately severe exercise impairment during which the testing had to be stopped prematurely because of severe dyspnea.

## Questions

1. Does this person have an identifiable medical condition? Describe the condition.
2. Discuss possible causes and contributing factors to this condition.
3. Describe Mr. Smith's possible functional limitations as related to his job. Explain your response.
4. Should he attempt to continue working in his present occupation?
5. If your recommendation is to continue his employment, what advice would you give Mr. Smith and his employer regarding reasonable accommodation, if any?
6. What alternative rehabilitation possibilities can be considered in his case? Consider the use of transferable skills.

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## Chapter 8

# HEMATOLOGICAL DISEASES

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## Introduction

Hematology is the study of diseases of the blood – the body’s transport system. The solid component of blood contains three types of cells which are formed in the bone marrow: red blood cells (RBCs), white blood cells (WBCs), and thrombocytes or platelets. The fluid component of blood is called plasma. Plasma is mainly water, but also contains coagulation and immunological proteins, glucose, dissolved carbon dioxide, fatty acids, and electrolytes, such as sodium and potassium. RBCs are the most numerous types of cell circulating in blood; each RBC contains millions of molecules of hemoglobin, an iron-containing protein that is responsible for transporting oxygen from the lungs to the rest of the body. In addition to carrying oxygen within the RBCs, blood also carries proteins responsible for coagulation or the clotting of blood, one of the body’s most important methods of self-repair (Hoffman, Benz, Shattil, Furie, & Cohen, 2005). The spectrum of hematological disorders is quite broad; this chapter discusses two hematological disorders - hemophilia and sickle cell anemia. These disorders reflect the range of issues that a rehabilitation counselor is likely to encounter in providing services to people with hematological disorders.

## Hemophilia

Hemophilia refers to a collection of hereditary, chronic bleeding disorders. The most common types of hemophilia result from an inherited deficiency of coagulation protein factors VIII and IX. These factors are particularly crucial for generation of an effective fibrin clot, without which continued bleeding occurs after the body is traumatized. Deficiency of factor VIII results in the coagulation disorder known as hemophilia A or “classic hemophilia,” and deficiency of factor IX results in hemophilia B or “Christmas disease” (Hoffman et al., 2005). The frequency of occurrence of hemophilia A is one in 5,000 male births and that of hemophilia B is one in 30,000 male births (Greer, Foerster, Rodgers, Paraskevas, & Glader, 2008).

Survival beyond young adulthood of people with severe hemophilia was uncommon before 1960. By the mid-1980s, many countries with well-organized hemophilia treatment centers reported improvement in life expectancy that now approaches the average lifespan. Timely treatment of bleeding and judicious prophylaxis have dramatically reduced severity of joint disease, resulting in enhanced mobility and decreased disability (Pace, 2007). Increased numbers of persons with hemophilia marry and have greater involvement in social, educational, and work activities.

## Genetics

The genetic defects accounting for deficiency of factors VIII and IX both occur on the X chromosome. Males have one X and one Y chromosome (XY), whereas females have two X chromosomes (XX). A mother carrying the defect  $X^h$  on one of her X chromosomes ( $X^hX$ ) may transmit hemophilia to her son ( $X^hY$ ), or pass the defect to a daughter ( $X^hX$ ) who becomes a carrier for hemophilia. The mother may transmit her normal X chromosome to her son (XY) or daughter (XX), and no genetic transmission of the mutant gene then takes place. Sons of a carrier mother and a normal father have a 50% chance of having hemophilia and daughters have a 50% chance of being carriers. Since a father with hemophilia ( $X^hY$ )

contributes only his Y chromosome to his sons, none of his sons will have hemophilia, but all his daughters will receive his  $X^h$  chromosome and will be carriers ( $X^hX$ ). This explains why hemophilia is almost exclusively a disease of males. Usually, a male inherits his mutant gene ( $X^h$ ) from his carrier mother ( $X^hX$ ), but in about 30% of cases of hemophilia, there is no family history of the disease and it is believed to arise from a spontaneous mutation (Mannucci & Tuddenham, 2001).

### ***Disease Severity***

Critical as they are for coagulation, the amounts of factors VIII and IX present in normal plasma are extremely small, and can be measured only in sophisticated research laboratories. For clinical purposes, one measures the level of each of these factors by its functional activity. The normal functional level of both factors, based upon a pool of plasma samples from normal persons, is 1.0 unit/ml of plasma. This is expressed as 100% activity. Values for factor VIII and factor IX in normal persons range from 50-170%. The minimum amount of each factor necessary for adequate coagulation is approximately 30%. Physicians categorize severity of hemophilia into three forms. Mild hemophilia occurs when the plasma coagulation factor level is 6% to 30%. Moderate hemophilia occurs with plasma coagulation factor levels of 2% to 5%. Severe disease occurs when only 1% or less of normal plasma coagulation factor is detectable (Greer et al., 2008; Raphael, 2005).

### ***Clinical Manifestations***

The major clinical manifestations of hemophilia are bleeding into a joint (hemarthrosis) or a muscle (hematoma). Signs of hemarthrosis or hematoma include pain, swelling, increased warmth of the joint or muscle, and some degree of limitation of motion. Hemarthroses occur primarily within the knees, but also in the elbows, ankles, shoulders, hips, and wrists. In severe cases, despite similar levels of coagulation factor activity, frequency of hemarthrosis varies widely from patient to patient. Joint bleeds may occur as often as two to three times a week, especially in the younger years. As the person enters adult life, episodes typically become less frequent. Some severely affected persons may not have a significant hemarthrosis for months.

Bleeding into the joint capsule initiates development of hemophilic arthropathy (joint disease). The inner surface of the joint capsule is lined by tissue called synovial membrane. During a joint bleed, various blood components, together with enzymes released from the synovium cause inflammation of the membrane. The synovial membrane responds to repetitive bleeds by producing a fibrous, highly vascular tissue which replaces normal synovium. Chronic synovitis (inflammation of the synovial membrane) ensues. Frequency and severity of hemarthroses further increases (Rosendaal et al., 1998). This vicious cycle leads progressively to degeneration of cartilage, destruction of bone, and replacement of joint space with fibrous (scar) tissue. Bones may fuse (ankylosis) resulting in deformity and a severely impaired range of motion. The early stage of synovial proliferation and joint destruction resembles the disease process of rheumatoid arthritis, whereas end-stage hemophilic arthropathy is similar to severe osteoarthritis (McPhee, Papadakis, & Tierney, 2008).

In addition to joint bleeding, persons with severe hemophilia with a virtual absence of factor VIII or factor IX are more likely to bleed excessively into any traumatized organ. They are prone to bleed more readily into structures diseased in such a way as to favor bleeding (e.g., stomach ulcer, bladder tumor, inflamed pancreas). Intracranial bleeding accounts for about 25% of deaths in hemophilia and usually follows head trauma. The modestly increased incidence of seizure disorder in people with hemophilia probably stems from previous head injuries involving intracranial bleeding.

### ***Diagnosis***

When there is a known family history of hemophilia on the maternal side of a woman's family, testing for hemophilia is recommended to determine carrier status (Greer et al., 2008). If her father has hemophilia, she is a definitely a carrier. A female carrier of the hemophilia gene can choose not to have children or she can undergo pre-natal testing to establish the sex of the fetus early in pregnancy. If the fetus is a male, she may choose to terminate the pregnancy or continue with the pregnancy after being counseled that medical science has made great strides in managing hemophilia as a chronic condition.

At birth, diagnosis of hemophilia can be made by analysis of umbilical cord blood. When there is no family history of hemophilia, prolonged bleeding suggests the presence of severe hemophilia. A hematologist establishes the diagnosis of hemophilia, its type (deficiency of factor VIII or factor IX), and the degree of severity (percent level of factor activity); effective care of a person with hemophilia requires a multidisciplinary health care team of physicians, nurses, and social workers to address the problems as he progresses from childhood through adolescence and adult life. The professional team concept gave rise to development of a comprehensive network of hemophilia treatment centers throughout the nation and in many parts of the world (Kasper & Dietrich, 1985). Studies have confirmed the efficacy of the comprehensive care paradigm used in Hemophilia Treatment Centers (HTC) (Dietrich, 1991). Persons with hemophilia who receive care in HTCs have a 60% reduction in mortality compared to those not being seen at HTCs (Soucie et al., 2000).

### ***Treatment***

The mainstay of treatment for hemophilia A and B is periodic replacement of the deficient factor with human blood protein products (called “factor concentrates”) first extracted from pooled or multiple donor plasma beginning in the 1960s, now synthesized by newer and safer recombinant technology. By the 1970s, home-administered and self-administered factor concentrates led to prompt control of bleeds and reduced joint and muscle damage. Self infusion of concentrate is readily done by the individual in about 15 minutes. Studies have shown that a home-based program for self-infusion significantly reduces bleeding complications and absences from school or work (Soucie et al., 2001). Furthermore, prophylaxis (providing concentrate in anticipation of trauma, as opposed to treatment when a bleed occurs) reduces medical costs and minimizes loss of productivity (McPhee et al., 2008). Less than two decades after replacement therapy through self-infusions became available, data clearly has shown that the number and length of hospitalizations had decreased; visits to hospital emergency rooms and hemophilia clinics decreased in inverse relation to the number of infusions given at home, work, or school. The number of individuals with hemophilia who were employed had increased (Hoffman et al., 2005; Serjeant & Serjeant, 2001).

Concentrate has become increasingly safe from contamination by blood borne infectious agents because of donor deferral procedures put in place by the American Red Cross, as well as by viral inactivation techniques used in the laboratory preparation of concentrates from pooled donations (Hoffman et al., 2005). Both these procedures were begun in the 1980s after substantial numbers of men with hemophilia developed antibodies to the human immunodeficiency virus (HIV) and many developed AIDS. In addition, recombinant factors VIII and IX were licensed for the treatment of hemophilia and these preparations eliminated the risk of transmission of infectious agents (Mannucci & Tuddenham, 2001).

### ***Functional Limitations***

The rehabilitation counselor can expect to encounter widely different degrees of functional limitations among clients with hemophilia. The extent of limitation usually correlates inversely with the individual’s level of the deficient coagulation factor. Some older clients, or clients with severe disease who have not had the benefits of modern treatment, have severe, generalized arthropathy. A few need to use wheelchairs for ambulation. Most young men have minimal or no joint deformities. The ages in between younger and older include men with a wide spectrum of limitations. Some men who were previously severely limited due to deformed joints have had surgical correction of the deformities by insertion of endo-prostheses to restore function (Logan, 1995).

Individuals with hemophilia who grew up during the era of modern treatment have few vocational limitations. Counselors need to surmount any residual notions that these clients necessarily require sedentary jobs, must be in protected environments, cannot perform manual labor jobs, and frequently miss work because of disease. Many individuals work successfully in physically demanding jobs and also in light and sedentary white-collar and blue-collar vocations. Persons with severe hemophilia, especially those who bleed more frequently, need to be discouraged from pursuing jobs requiring maneuvers that are potentially traumatic to joints (e.g., repetitive clutch pedal operation in a man with chronic left knee arthropathy) and occupations clearly dangerous because of a particular person’s joint limitations. Because the person with hemophilia can normalize his coagulation mechanism within minutes, many potentially hazardous jobs

offer little more risk than to other workers. Counselors need to plan vocational rehabilitation efforts according to the specific limitations of the particular client.

### ***Intellectual Functioning***

Persons with hemophilia do not display any inherent intellectual limitations. In fact, some studies suggest higher than average intelligence. Data collected during the 1960s and early 1970s showed a notable gap between scores on intelligence tests and achievement tests, the lower achievement attributable to disrupted school attendance. The average educational level attained was considerably less than by men in the general population. Within two decades following the advent and implementation of factor replacement and self-supervised treatment, several countries reported the achievement by people with hemophilia were attaining educational levels at least equivalent to age-matched, unaffected men (Colegrove & Hurtzinger, 1994; Hoffman et al., 2005).

### ***Psychosocial Factors***

Psychosocial dynamics in men with hemophilia have provided focus for considerable investigation. As a group, these individuals display the same range of psychological variation present in non-affected persons. There is no general personality pattern for persons with hemophilia. Repeatedly, it is noted that many men with hemophilia are optimistic and exhibit strong ego strength. These men believe that coping with their disease has given them a “hardihood” they would not otherwise have developed. Despite these observations, the rehabilitation counselor needs to be aware of specific psychological limitations that sometimes occur (Falvo, 2009).

Particularly in previous years, when a boy with hemophilia was born, the mother not infrequently experienced varying degrees of guilt because of what she perceived as her “genetic responsibility.” Commonly, these mothers became inappropriately anxious and overprotective. Severe curtailment of her son’s activities coupled with excessive attention and indulgence sometimes led to development of an overly dependent, passive, anxious person who experiences feelings of hopelessness, pessimism, and social isolation. By contrast the boy sometimes responds with denial leading to reckless, “daredevil” activities and pathologic risk taking. HTCs have been able to do much to provide families with counseling and education. Better understanding of the disease and awareness of the dramatic effects of modern treatment have helped mitigate unhealthy parental concerns.

Employment remains a concern for people with hemophilia. Clearly, unemployment diminishes self-esteem and fosters passivity, pessimism, and depression. A client’s self-concept as being “unemployable” may be a key reason for reluctance to seek a job and failure to present oneself in the most positive light when seeking employment. Failure to find a job often reinforces initial self-doubts. If the rehabilitation counselor can help the client surmount reticence and negativism regarding employability, the client will be able to take a critical step forward (Raphael, 2005).

### ***Specific Functional Limitations***

Physical limitations vary with each person. Arthropathy of a lower extremity weight-bearing joint is most likely to be physically limiting. For some individuals, elbow or shoulder joint disease may pose the greatest limitation. The client and a member of the HTC team can provide reliable information about which joints are disabling and in what manner. A knee with almost no range of motion due to severe arthropathy, but with only rare hemarthroses is less disabling for many jobs than a right elbow with severe chronic synovitis and frequent bleeds, in a right-handed person.

### ***Occupational Limitations***

Viewed as a group, persons with hemophilia have few vocational limitations. On an individual basis, the counselor will see a variety of restrictions. Magnitude of the disease correlates closely with the presence of occupational limitations. There is a strong correlation between successful self-management of the disease and ability to function in the workplace. Clients with severe hemophilia who are on self-supervised treatment are best able to cancel the effects of an accident on the job that causes a bleed. Most clients with mild, and many clients with moderate disease, have almost no vocational limitations. A characteristic feature of severe disease, especially among those who bleed more frequently, is unpredictability. Joint bleeds and hematomas often appear to be spontaneous. Therefore, persons with



severe hemophilia may not always optimally perform certain jobs in which unbroken continuity of activity is essential. A 15-minute break is generally all the individual needs for self-infusion (Falvo, 2009).

The rehabilitation counselor must evaluate the client's vocational limitations individually (Brodwin, Parker, & DeLaGarza, 2003). Members of the comprehensive hemophilia treatment team can provide precise assessment of functionality. Most employees with hemophilia can perform equal to their peers who do not have hemophilia in a variety of jobs and professions. Inaccurately perceived limitations by prospective employers because of lack of understanding of this disorder far outweigh actual functional limitations.

### ***Rehabilitation Potential***

To be an effective advocate for individuals with hemophilia, the counselor needs to understand both the clinical behavior of the disorder and its impact on the person (Falvo, 2009). Contrary to popular misconception, persons with hemophilia are not fragile individuals only suitable for very specialized jobs located in protected environments. Often called "bleeders," many people erroneously believe individuals with hemophilia are vulnerable to rapid blood loss upon exposure to the slightest trauma. In reality, these people bleed no more rapidly or excessively from minor cuts and superficial abrasions than do persons who do not have hemophilia. This is because the earliest phases of clotting are unimpaired in hemophilia. Rather, the blood fails to clot and the person continues to bleed for a prolonged time. With modern treatment, individuals can work safely and productively in a variety of jobs.

Rehabilitation potential for individuals with hemophilia is excellent. Effective treatment of bleeding episodes, prevention or minimization of the disabling complications of joint bleeds, and ability to correct deformities through surgery has greatly enhanced employability. Because of modern treatment modalities, many persons have completed high school and, increasingly, have additional education.

Before the advent of modern medical therapy, some clients developed irreparable deformities. Others received inadequate medical treatment. Appropriate medical treatment can prevent further progression of existing disabilities and help prevent additional complications. The rehabilitation counselor may consider the initial status upon first seeing the client as being relatively stable and work jointly toward selecting a vocation in which the disability will not adversely affect functioning on the job (Taylor, 1976).

For clients who bleed frequently, the opportunity to self-infuse at the worksite effectively reduces morbidity and loss of work time. Capacity to store concentrate and necessary accessories (needles, syringes, and gauze pads) is needed.

Individuals self-infuse in their offices or in restrooms. Some self-infuse at a nearby gasoline station or restaurant to conceal their hemophilia. Many employers have a first aid room where employees can self-infuse at the worksite. Company (industrial) physicians and nurses should be encouraged to avail themselves of assistance and guidance through the local HTC. Rehabilitation counselors can educate employers by providing current information about the medical treatment of this disease. Persons with hemophilia frequently learn to compensate for their limitations and, therefore, often have fewer injuries than other workers in similar jobs. In many respects, people with hemophilia may be optimal clients for vocational rehabilitation because the disease itself can be effectively controlled (Brodwin et al., 2003). If unexpected deterioration of joint status or other unforeseen medical factors alters the client's physical abilities, he may again become a candidate for vocational guidance and rehabilitation.

Rehabilitation counselors need to be knowledgeable in modern methods of hemophilia treatment. Most employers are unfamiliar with recent medical advances that allow persons with hemophilia to live near-normal lives. Although common, employers typically do not understand self-supervised infusion. Counselors need to educate employers in this and other areas of treatment to maximize potential employment opportunities for these clients (Brodwin et al., 2003; Raphael, 2005).

## **Sickle Cell Anemia**

Sickle cell anemia is characterized by a single amino acid change in the globin (valine for glutamic acid in the 6th residue) that produces a hemoglobin molecule of reduced solubility, especially in the absence of oxygen. A red blood cell (RBC) containing sickle hemoglobin forms polymers inside the RBC, thereby

converting it from its normal biconcave shape to a crescent-shaped, or sickled, RBC. Sickled RBCs block small blood vessels and sometimes large ones, causing damage to them and eventually death to the cells supplied by the affected blood vessels (Platt, 2008). The sickled RBCs are responsible for producing all the clinical manifestations of disease in people who have sickle cell anemia. These present significant challenges for the person with sickle cell anemia, but medication has made much progress in treating sickle cell anemia so that now persons with sickle cell anemia live into their fifth and sixth decades (Pace, 2007).

Most cases of this disease occur in people of African or Mediterranean ancestry. In the United States, about 8% of African-Americans carry an abnormal beta globin gene for Hb S (sickle cell trait), about 2% carry an abnormal beta globin gene for Hb C, and another 4% carry an abnormal gene for beta thalassemia. Consequently, about one in every 500 African-American children inherits two abnormal beta globin genes and has sickle cell anemia, the most severe form of sickle cell disease. Considerable scientific evidence indicates that these abnormal Hb genes arose in Africa, the Mediterranean area, and some Southeast Asian countries as partial protection against malaria (Bunn, 1997; Serjeant & Serjeant 2001).

The most common and severe form of sickle cell disease is sickle cell anemia (Embury, Hebbel, Mohandas, & Steinberg, 1994; Serjeant & Serjeant, 2001). Hemolytic anemia is present in a child by the age of six months, although vascular occlusion symptoms may not appear until one year of age. Persons with sickle cell anemia have severe anemia. They exhibit frequent painful crises during childhood and loss of function of the spleen (an organ with multiple blood-cleaning functions), predisposing them to severe infections. Until recently, bacterial infections were the most common cause of death in children. Now, prophylactic treatment with penicillin has nearly eradicated this problem (Gaston et al., 1996; Platt, Brambilla, & Rossi, 1994). During the adolescent years, there is often a lessening in occurrences of crises as viral infections decline and individuals learn to avoid precipitating factors.

Children frequently have delayed growth and a delay in the onset of puberty. About half of persons with sickle cell anemia are tall and thin with abnormally long limbs. Delayed growth and sexual maturation appear to be related to nutritional factors, although the exact mechanism is unknown. Researchers have not determined a specific treatment for this. As an adult, there is wide variation in disease manifestations and severity. Approximately 10% of individuals have extremely benign disease, whereas 25% have a severe form requiring frequent hospitalizations for pain control. Most persons have symptoms requiring hospitalization once a year or less (Platt et al., 1991). The long-term effects of recurrent vascular occlusion, ischemia, infarction, and fibrosis may cause one or more types of chronic organ damage. As a result, the management of organ complications becomes a major issue and prognosis is uncertain (Platt et al., 1994).

### ***Clinical Manifestations***

Vaso-occlusions (blockage of blood vessels by sickled RBCs) account for most of the severe complications of sickle cell disease and can present as acute episodes (called “crises”) of severe pain in the chest, back, abdomen, or extremities. A painful vaso-occlusive crisis has a sudden onset, usually lasts five to six days, and may be localized to one area of the body or generalized (Francis & Johnson, 1991).

One type of vaso-occlusive crisis, acute chest syndrome, is the leading cause of death and hospitalization (Castro, Brambilla, & Thorington, 1994; Golden, Styles, & Vinchinsky, 1998). The acute chest syndrome is commonly caused by fat embolism and community-acquired pneumonia and can progress to respiratory failure if not treated aggressively with transfusions and bronchodilators to improve oxygenation.

Other organs of the body can be affected by vaso-occlusive complications, such as the kidneys which can lead to kidney failure. Leg ulcers occur due to infarcts in the skin around the ankles. Prolonged standing in one position, such as may occur with certain jobs, makes these ulcers worse. Vaso-occlusion of the bones and joints can lead to severe physical limitations. Bone infarcts (aseptic necrosis) in the lower extremities and the vertebrae of the spinal cord precipitate collapse of the involved bone and lead to degenerative osteoarthritis in older individuals. Serious limitations of function then result (Pace, 2007; Serjeant & Serjeant, 2001). Occlusion of the intracerebral arteries causes thrombosis and can result in a cerebrovascular accident (stroke). As with other diseases, both intellectual and physical disabilities can result from central nervous system complications in childhood, leading to limitations in adulthood and are challenging rehabilitation issues.



A major cause of illness and death in sickle cell anemia, especially in children, is bacterial infection. Septicemia (infection in the blood) and meningitis (infection of the membrane of the spinal cord or brain) are life-threatening and commonly occur in sickle cell disease. Prophylactic treatment with penicillin, and vaccination against *Streptococcus pneumoniae* of all children with sickle cell disease by age two, has substantially reduced infections and markedly decreased childhood mortality (Gaston et al., 1996). After age 10, occurrence of septicemia and meningitis decline, but high incidences of localized infections (i.e., pneumonia, pyelonephritis, and osteomyelitis) persist (Serjeant & Serjeant, 2001; Hoffman et al., 2005).

### ***Treatment***

In addition to supportive treatment during vaso-occlusive crises, or treatment for complications associated with sickle cell disease affecting specific organs, hydroxyurea has been used to try to shift hemoglobin production from sickle hemoglobin to fetal hemoglobin (which does not sickle as readily) by changing marrow-proliferation kinetics to favor fetal hemoglobin production. In 1995, a landmark study demonstrated the effectiveness of hydroxyurea in reducing the occurrence of painful crises from 4.5 to 2.5 per year (Charache et al., 1995). Clinical consensus is that persons with moderate-to-severe disease, those with three or more acute painful crises or episodes of acute chest syndrome in the previous year, should be given hydroxyurea. Information about the use of hydroxyurea in sickle cell disease has been published by the National Institutes of Health (National Institutes of Health, 2002). Hydroxyurea is not without side effects. Suppression of bone marrow function can occur, as well as adverse effects on reproductive function (Platt, 2008).

### ***Functional Limitations***

As in other chronic diseases with childhood onset, the individual with sickle cell disease has physical as well as psychosocial issues that play a role in the potential for vocational rehabilitation. A full range of medical, psychiatric, and social services are needed to maximize overall functioning for successful rehabilitation.

#### ***Physical Limitations***

Physical limitations are primarily the result of the fatigue and weakness associated with anemia and sickle cell crises (Raphael, 2005). Although there is great individual variation, sickle cell crises typically occur on an average of once or twice a year. Not all episodes are severe enough to require hospitalization. The crisis usually resolves within a week. Yet, there may be an additional 5-10 days during which the individual has residual symptoms. Some persons maintain work activity throughout sickle cell crises; others need to take time off work. This amount of time varies from one to several days and occasionally up to two weeks. This is dependent not only on the individual, but also on the exertional level of an employee's work activity. Therefore, counselors need to consider the exertional level of work when providing vocational rehabilitation (Brodwin et al, 2003).

Employees with sedentary or light jobs are able to continue working through sickle cell crises more often than persons who have physically demanding jobs. When providing vocational rehabilitation for individuals who are changing jobs or are entering the labor force for the first time, the counselor is advised to consider this with the client who has occasional sickle cell crises.

In the presence of lung disease, dyspnea (shortness of breath) becomes the limiting factor for exertional levels of work (Greer et al., 2008). Analysis of physical demands, both usual and occasional, for a particular vocation is useful. Job restructuring or accommodation is helpful when a position has occasional physical demands beyond the person's capacity.

One can find visual field defects and visual loss in one or both eyes in sickle cell anemia. Functional limitation is dependent on extent of visual loss and improvement that can occur with use of visual aids.

Leg ulcers produce pain and limitations of motion in the affected extremity. This interferes with activities such as standing, walking, climbing, and balancing. Prolonged standing is one possible cause of these ulcers. Counselors need to assess possible complications and resulting functional limitations in standing and walking (Falvo, 2009).

Bone and joint disease interfere with physical activity because of pain, deformity, loss of strength, and decreased endurance. The nature of the impairment depends upon the joints involved. Walking, standing, bending, climbing, and balancing may be affected. Some limitations are overcome by learning new techniques for physical activities through physical therapy. Prosthetic implant surgery has been a successful technique in this area.

### ***Environmental Factors***

Several environmental conditions exacerbate symptoms. Extreme cold or intense heat should be avoided. Jobs involving exposure to cold and damp conditions need to be avoided since these environmental factors precipitate sickling of RBCs. Exposure to employment environments with noxious fumes, high levels of dust, and poor ventilation are not tolerated well, especially for individuals with pulmonary problems. Hot and humid conditions add stress on the heart and precipitate dehydration (Francis & Johnson, 1991). On all jobs, persons with sickle cell disease need easy availability of water and other fluids to prevent potential dehydration.

### ***Psychosocial Issues***

Sickle cell disease is a chronic condition requiring careful attention throughout an individual's lifetime. Emotional stress can exacerbate symptoms and make management of the overall disease difficult. The disease itself, with its inherent uncertainty, causes emotional upheaval. An inability to predict or control symptoms leads to heightened anxiety and increased frustration. Recurrent painful episodes of sickle cell crises add to emotional distress (Brodwin et al., 2003). Understandably, emotional difficulties are common in persons with sickle cell disease and contribute to poor self-esteem, depression, and anxiety. Self-concept is formed early in childhood; with recurrent episodes of sickle cell crises and repeated hospitalizations, children with sickle cell disease often develop dependent personalities and passive-aggressive traits (Dell Orto & Power, 2007; Seeley, 1995).

Depression is the major psychological factor associated with this disease; it affects the ability to cope with the variable nature of the disease. Consequently, psychiatric consultation may be helpful when preparing a client for vocational rehabilitation.

## ***Rehabilitation Potential***

Most people with sickle cell disease will not need vocational rehabilitation services. Those with moderate or severe disease may have chronic complications and organ damage and become candidates for rehabilitation. Individuals with recurrent sickle cell crises that interrupt their work life may benefit from rehabilitation services. Counselors need to be able to interact with employers to explain sickle cell disease symptoms. An employee with an otherwise good employment record may be able to use accumulated sick leave for occasional sickle cell crises. Reasonable accommodation can facilitate a successful return to work for someone with moderate to severe sickle cell symptoms (Brodwin et al., 2003).

An example of reasonable accommodation for persons with sickle cell disease is an agreement with the employer that the individual will work through minor illnesses and save or accumulate sick days to be used for occasional sickle cell crises. Another accommodation is to have the worker put in non-paid overtime between crises to be used when work is missed during a crisis.

Onsite observation of the job helps the counselor modify physical aspects of the position. When a worker is having difficulty with physical exertion, the counselor can assist the client and employer in modifying the amount of lifting and carrying required (Falvo, 2009). One may be able to break down into lighter loads the amount lifted and carried; if this is not possible, the counselor may want to recommend job restructuring.

Mental stress present on the job and its effect on potential development of sickle cell crisis needs careful assessment. Modification of emotional stressors is more difficult than alteration of physical activities. Often, it is impossible to change emotional stress factors. At times, positive facilitation by the counselor is effective and results in decreasing emotionally stressful aspects of the job. Employer education creates a more thorough understanding of the disease, helping diminish work-related stressors.

When determining rehabilitation potential, the counselor evaluates physical complications the individual has and whether time off work may be necessary when sickle cell crises occur. With this information, in conjunction with appropriate medical consultation and an understanding of the provisions of the Americans with Disabilities Act, the rehabilitation counselor is equipped to provide rehabilitation for persons who have sickle cell disease.

## Case Study

Mr. Leo Jones is a 53 year-old college educated, self-employed machinist with sickle cell disease. He is married and has two young children. Mr. Jones developed bacterial septicemia (infection in the blood) and multiple complications of sickle cell disease requiring intermittent hospitalization over several months time. Both his marriage and business failed. His wife left him and moved to another state, taking their two children (ages 7 and 10) with her and moving in with her elderly parents. Mr. Jones' wife is Chinese and is bicultural, maintaining some of the beliefs of traditional Chinese culture. Her parents always wanted her to marry within their ethnicity and maintain a traditional Chinese home. This has caused emotional concern, especially when it comes to raising her children.

Marriage problems and the failed business contributed to Mr. Jones' anxiety and depression. Dr. Carla Williams, his treating physician, noted that Leo had severe depression and referred him for psychological counseling. As a result of his condition, Mr. Jones developed a fatalistic attitude and did not complete the recommended course of counseling. Consequently, his depression deepened.

Dr. Williams restricted Mr. Jones to light work. There are no limitations in the areas of standing and walking.

The daily work activities of a machinist (U. S. Department of Labor, *Dictionary of Occupational Titles* [DOT] # 600.280-022, 1991) include setting-up and operating machine tools and fitting and assembling parts to make or repair metal parts, mechanisms, tools, and equipment. One reads blueprints, diagrams, and mechanical drawings, uses precision-measuring tools, and understands a variety of machining procedures. Machinists use hand tools and power tools throughout the work shift. Mr. Jones had been self-employed for 30 years in this line of work.

Due to physical limitations related to bone destruction (avascular necrosis) in both hips, Mr. Jones was unable to return to his machinist business, which often required 10-hour days, six days a week. Beside himself, he had two helpers in the business who did general machining. Work in the machine shop involved occasional lifting up to 80 pounds when shipment orders arrived, and regular lifting of 45 pounds, with repetitive lifting of 20-25 pounds. Mr. Jones stood or walked 50% of the workday.

After the business failed and for the next 10 years, Leo filled his days with a variety of activities related to his church and various social clubs. By assuming a demanding schedule of responsibilities associated with these activities, he kept active. During this period, Dr. Williams needed to hospitalize him as often as six times a year for acute painful crises, and recurrent bone infections (osteomyelitis). Eventually, Mr. Jones was reunited with his children when they returned to the city to attend college. With these new family responsibilities, he gave up many of his outside activities and concentrated on assisting his children with their studies and making a home for them. Within a few months, Leo noticed a remarkable decrease in the frequency of hospital admissions, which he related to improved self-care. Proper rest, careful attention to fluid intake, and monitoring of physical activities has contributed to the improved clinical course of the disease.

Mr. Jones' depression cleared. Subsequently, he determined the need for a higher income and sought vocational rehabilitation services. A counselor from the state department of rehabilitation provided counseling, guidance, and job placement. These services were successful and Leo was placed as a reader and driver for people with visual impairments.

## Questions

1. Discuss physical, environmental, and psychosocial aspects of sickle cell disease.
2. Outline a vocational profile for Mr. Leo Jones including age category, educational level, work history (exertion and skill levels), occupationally significant characteristics, and transferable skills.

3. If Leo had decided to make further attempts to make his machinist business succeed, how would you as his rehabilitation counselor approach this decision? Provide supporting arguments.
4. Can you recommend use of transferable skills as another viable vocational rehabilitation option for Mr. Jones? If so, describe what skills are transferable to other skilled and semiskilled jobs.
5. Discuss the rehabilitation goal pursued in this case and explain why the counselor may have recommended it. Do you agree with this rehabilitation plan?
6. Speculate how Mr. Jones was able to achieve his vocational goal.
7. Evaluate this case from a multicultural prospective.

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## Chapter 9

# RHEUMATIC DISEASES

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## Introduction

Rheumatology is the scientific study and care of rheumatic diseases; a physician who specializes in rheumatic diseases is a rheumatologist. This specialty has developed within the last 75 years in the United States and Western Europe. All urban areas in the United States have practicing rheumatologists, and major urban centers have rheumatology training programs within medical schools and major hospitals. The American College of Rheumatology (<http://www.rheumatology.org/>) promotes research and education on arthritis and is the professional organization for rheumatologists; the Arthritis Foundation (<http://www.arthritis.org/>) is the organization that raises funds and conducts arthritis research and patient education throughout this country. Both these organizations have headquarters in Atlanta, Georgia and are excellent resources for rehabilitation counselors seeking information about rheumatic disorders (Harris et al., 2008; Lee & Abramson, 2005).

### *Definition*

Rheumatic diseases are a group of conditions affecting the supporting structures of the body, including the joints and periarticular tissues, connective tissues of the skin, bones, muscles, and diseases of the immune system. They include a group of inflammatory diseases affecting connective tissue that are insidious (develop in a subtle, gradual manner) in onset, characterized by exacerbations and remissions, and resistant to medical therapy (there are no cures, only palliative treatment). Included in this group are arthritis and diseases that cause pain, stiffness, and abnormalities of mobility. Fundamental features of rheumatic diseases are the signs of inflammation: warmth, swelling, redness, pain, and loss of motion. Individuals with rheumatic disease have complaints of pain, loss of energy, easy fatigability, stiffness, and limitations of joint motion. Work endurance and capacity are affected by these symptoms; with severe involvement, activities of daily living and mobility become impaired.

### *Arthritis and Myositis*

The common rheumatic diseases that affect the joints are called “arthritis,” those that affect the muscles are termed “myositis,” and “soft tissue rheumatism” is present when soft tissues (tendons, ligaments, bursa, and muscles) are involved accompanied by associated stiffness and pain. There are more than 100 rheumatic diseases that result in pain, stiffness, and functional impairment. Some of the more common diseases are osteoarthritis (the most common rheumatic disease), rheumatoid arthritis (the highest rate of disability), gout, bursitis, tendonitis, fibromyalgia, systemic lupus erythematosus, scleroderma, and ankylosing spondylitis. Less common types of rheumatic diseases include myositis (polymyositis and dermatomyositis), scleroderma, and arteritis (Harris et al., 2008; Klippel, Stone, Crofford, & White, 2008; McPhee, Papadakis, & Tierney, 2008).

## Arthritis and Rheumatic Disorders

Musculoskeletal diseases, which include arthritis and rheumatic disorders, are the leading causes of disability and absence from work (Harris et al., 2008). They are the second most common reason for which people see physicians, affecting 20% of the adult population. Arthritis and the other rheumatic diseases are difficult to diagnosis due to the subtlety of physical signs, the tendency for symptoms to overlap with other

medical problems, and the paucity of diagnostic clinical and laboratory tests. Symptoms tend to develop gradually, respond to treatment slowly, and require long-term care.

### ***Impact, Prevalence, and Cost***

The impact of arthritis in the United States is great. In 2003-2005, 50% of adults 65 years old and over reported a diagnosis of arthritis. It is estimated that 46 million people have arthritis. Of these, 294,000 are under age 18; this represents one in every 250 children (Hootman, Bolen, Helmick, & Langmaid, 2006; Hootman & Helmick, 2006).

Currently, over seven million Americans are disabled by arthritis. Annually, arthritis accounts for 500 million days of restricted activities and 68 million days of lost work. It is the leading cause of absenteeism in business and industry and the second leading cause of disability payments (following heart disease). In 2003, the total cost attributed to arthritis and other rheumatic conditions in the United States was \$128 billion, up from \$86 billion in 1997. Medical expenditures (direct costs) for arthritis and other rheumatic conditions in 2003 were \$47 billion, up from \$35 billion in 1997. Earning losses (indirect costs) for arthritis and other rheumatic conditions in 2003 were \$47 billion, compared to \$35 billion in 1997. Rheumatoid arthritis accounts for 2.2 million days of lost work and osteoarthritis accounts for 66 million days of lost work, annually. In the United States, one in every five people or 20% of the population has an arthritic condition (Hootman et al., 2006; Hootman & Helmick, 2006).

### ***Categories of Rheumatic Disease***

There are three categories of rheumatic diseases that the rehabilitation professional is most likely to encounter. First, are the **diffuse connective tissue diseases** (rheumatoid arthritis, systemic lupus erythematosus, scleroderma, and polymyositis). This category has the most severe and highest prevalence of disability. Second is **osteoarthritis**, the most common form of arthritis, accounting for the largest number of lost work days. The third category is **ankylosing spondylitis**, which causes severe back pain and often is confused with other causes of back pain.

## **Rheumatoid Arthritis**

### ***Explanation of Disease State***

Rheumatoid arthritis (RA) is a systemic disorder of the connective tissues of the body, characterized by exacerbations and remissions. Its primary targets are the joints and adjacent supporting structures. The lining of the joint (synovium) is the site of chronic inflammation. Synovial tissues proliferate and lymphocytes (white blood cells) and other chronic inflammatory cells invade its interstices (spaces or gaps in tissues). This inflammatory process produces pain, heat, swelling, and loss of joint motion. The process is locally invasive and rheumatoid tissue erodes cartilage, bone, and ligaments.

The etiology of rheumatoid arthritis is unknown, and was originally characterized as being of insidious onset and slow progression. However, there is substantial data that by the end of the second year from onset of symptoms, 90% of patients show radiographic evidence of damage.

### ***Incidence***

RA is a common disorder affecting approximately 1% of the adult population. It has a high disability rate estimated to be 21 per 1,000 people (Cooper, 2000). This disability rate is higher than the next three most common conditions: heart disease, back impairment, and hypertension. In addition, people diagnosed with rheumatoid arthritis utilize more medical services (physicians and hospitals) than patients with osteoarthritis, back pain, or tendonitis. Rheumatoid arthritis is three times more likely to affect women than men and has its peak onset between the ages of 20 and 30, although the incidence of RA has declined over the years. Because of its chronicity and severe consequences, it is a primary focus of arthritis research (Harris et al., 2008).

## ***Effects***

The primary effects of this type of arthritis are pain, swelling, loss of joint motion, joint deformities, and subsequent loss of function. The disease is one of exacerbations and remissions, but the overall course is progressive. Pincus et al. (1984) analyzed a group of individuals diagnosed with rheumatoid arthritis over a period of nine years and found a steady decline in physical functioning; 92% had significantly lower overall functioning. Of those under 65 years of age who were working at the commencement of the study, 85% demonstrated significant work disability. Meenan, Yelin, Nevitt, and Epstein (1981) studied the financial impact of rheumatoid arthritis on wage earning and found that people earned 50% of what was expected based on their earning potential, and average individual income dropped, accounting for a 32% decrease in family income. Most had medical coverage but not adequate disability insurance to cover loss of earnings. Of the 59% of people with insurance coverage for loss of earnings, the coverage provided income for a maximum of six months.

People with RA have a symmetrical arthritis affecting multiple joints. Initially, small joints of the body are affected, but eventually all joints can be impacted. The initial symptoms occur in the small joints of the hands and feet with symptoms of pain, swelling, and joint stiffness. Individuals commonly experience fatigue, morning stiffness, and may have weight loss. As the disease progresses, more joints are affected and the degree of disability increases. Development of characteristic deformities of the hands typically occurs with rheumatoid arthritis. These deformities limit performance of employment and leisure activities. Restriction of range of joint motion frequently accompanies the deformities. Additionally, individuals become self-conscious about the appearance of their hands and feet, restrict their activities, and tend to wear concealing clothing (Klippel et al., 2008; Pincus et al., 1984).

## ***Diagnosis***

Diagnosis is established through clinical features and laboratory testing. To establish a diagnosis, the arthritis must be polyarticular (i.e., involve more than one joint) and be at least six weeks in duration. Synovial joints are involved in a symmetrical manner. Initially affected are the small joints of the hands and feet, although any synovial joint may be involved; rheumatoid arthritis is a systemic (affecting the entire body) disease. Subcutaneous nodules develop at pressure points. These soft, non-tender nodules vary from a few millimeters to several centimeters in size and are most common at the elbow. Joints are swollen, tender, with increased synovial fluid. People with more severe disease may have inflammation of the lining of the lungs and heart, lung involvement with fibrosis (scar tissue formation) and nodules, and inflammatory vessel disease of the skin and other organs. Dryness of the eyes and mouth is common as are compression of peripheral nerves adjacent to joints (e.g., carpal tunnel syndrome).

Laboratory results are sparse but persons may have mild anemia, thrombocytosis, elevation of acute phase reactants, but normal urinalysis and uric acid levels, and have a serological abnormality, called rheumatoid factor. Rheumatoid factor, an autoantibody, is present in 75% of people with rheumatoid arthritis; it is associated with extra articular disease and a prognosis indicative of a more severe condition. In early disease, x-rays show loss of bone density (osteoporosis) and soft tissue swelling about the joints. X-rays of individuals with more advanced arthritis show loss of articular cartilage, erosion of bone adjacent to joints, and associated deformity (Harris et al., 2008).

## ***Treatment***

Currently, there is no cure for RA, although there are good palliative measures that can ameliorate symptoms or induce remission for extended periods. A three-pronged approach to treatment should be considered with 1) non-steroidal anti-inflammatory drugs (NSAIDs) and/or low dose oral and/or intra-articular steroids, 2) disease modifying antirheumatic drugs (DMARDs), and 3) biologic agents (Majithia & Geraci, 2007).

By decreasing inflammation, NSAIDs and steroids reduce pain and swelling and improve range of motion and strength. NSAIDs (e.g., ibuprofen) are generally preferred because they tend to have fewer side effects, although steroids are more potent. NSAIDs and steroids can be taken together.

DMARDS include methotrexate, leflunomide, hydroxychloroquine, sulfasalazine, and azathioprine. They are slow acting and may require up to six months to achieve a therapeutic response. By potentially reducing the inflammatory hyperplastic synovium and reversing erosions of bone and nodules, early use can slow progression of RA and improve overall prognosis (Majithia & Geraci, 2007).

The biological agents are the newest line of treatment and like DMARDS can take weeks to months to have discernible effects (McPhee et al., 2008). They have very diverse mechanisms of action. These biological agents are more effective than methotrexate at slowing radiographic progression. The biologic agents, however, do have significant potential side effects, such as increased susceptibility to serious infections (Weinblatt, Dremer, & Bankhurst, 1999).

Total joint replacement is a standard treatment for advanced joint disease which is progressive, symptomatic, and unresponsive to pharmacologic treatments. Through joint replacement, the diseased joint is surgically replaced with an artificial joint of highly polished metal, which articulates (forms a joint) with a plastic component. Surgery relieves pain and frequently improves mobility. Resumption of work and recreational activities may occur after joint replacement (Koopman & Moreland, 2005). Successful joint replacements have been achieved in the hands, wrists, shoulders, hips, and knees with the greatest level of satisfaction with hip and knee replacements.

### ***Functional Limitations***

Functional limitations for individuals with rheumatoid arthritis are of primary concern. As the disease progresses and the years pass, functional loss becomes progressive. The American College of Rheumatology established criteria that physicians use to describe functional status. These criteria are illustrated in Table 1 and include Functional Class I (normal function) through Functional Class IV (unable to perform self-care activities). People referred for vocational rehabilitation are usually in Functional Classes II and III. Functional limitations are stated relating to the stage of the disease and areas of the body involved (Hochberg et al., 1992).

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**Table 1**  
**Functional Status In Rheumatoid Arthritis\***

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CLASS I	Able to perform all usual activities of daily living.
CLASS II	Able to perform all usual self-care and vocational activities. Limited in avocational activities.
CLASS III	Able to perform all usual self-care activities. Limited in vocational and avocational activities.
CLASS IV	Unable to perform usual self-care, vocational, and avocational activities.

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\*American College of Rheumatology Criteria, Revised 1991 (Hochberg et al., 1992).

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### ***Chronic Pain***

All individuals diagnosed with rheumatoid arthritis have pain that is chronic in nature and exacerbated by movement of the affected joints. Additionally, many people have stiffness of the entire body, which is primarily a morning phenomenon and may last from minutes to hours. Specific functional disabilities occur in each individual, depending on the stage of disease and body areas affected. This disorder causes loss of motion and pain on motion; there is weakness, decreased strength, and diminished endurance. If the hands and upper extremities are involved, pain and loss of motion results in decreased capacity to perform grasping, fine dexterity hand activities, and overhead reaching. With lower extremity involvement, decreased capability to stand and walk for long distances occurs.

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### ***Emotional and Intellectual Factors***

People with rheumatoid arthritis have no emotional limitations except those imposed by a disorder associated with chronic illness, pain, and loss of motion (Harris et al., 2008). There is an increased incidence of depression; however, it is not greater than would be expected for any chronic disorder that is progressive in nature. Intellectual capacity is not affected and ability to interact on a social level is unimpaired. Occasionally, because of the deforming nature of the disease, embarrassment is experienced concerning deformities of the hands and feet, as well as use of ambulatory aids such as canes, walkers, and wheelchairs (Arango & Cano, 1998).

### ***Vocational Considerations***

If rheumatoid arthritis is not well controlled or of long duration, there may be chronic pain, stiffness, fatigue, and weakness, all of which influence an ability to work. Yet, during the first years of the disease, most people obtain good control with first and second line medications, and vocational limitations are a consequence of the specific area of the body affected. Because of a reduction in strength and involvement of articulations effecting movement, most individuals are unable to perform heavy or very heavy work activities. Employment in the exertional categories of light and sedentary is preferred. If a person, before acquiring rheumatoid arthritis, was performing medium work, it is likely that employment will need to be modified to light work (McPhee et al., 2008).

**Upper extremity involvement.** Hand and wrist involvement results in decreased power grip; most people with rheumatoid arthritis are assessed below the 5<sup>th</sup> percentile for grip strength (Cooper, 2000). Additionally, pinch, grasp, and manual dexterity are reduced. If there is involvement of the hands, wrists, elbows, and shoulders, activities involving reaching and rapid movements of the upper extremities are diminished.

**Lower extremity involvement.** Disease in the lower extremities results in impaired ability to rise from a sitting position, stand, and walk for long periods of time. Work activities should accommodate the specific deformities and symptoms of the individual. If the arthritis affects standing and walking, the person may be limited to sedentary work activity.

**Other factors.** Some people with rheumatoid arthritis require special considerations because of deformities, weakness, and loss of joint range of motion. Adaptations, such as higher chairs, raised toilet seats, and curb cuts accommodate lower extremity impairments. Occasionally, individuals have other health problems related to rheumatoid arthritis. For example, multisystem involvement from rheumatoid disease usually results in generalized fatigue with decreased strength and endurance.

### ***Rehabilitation Potential***

Rheumatoid arthritis does not affect intellectual capabilities. Life expectancy for a person with this disease is shortened by a few years. Therefore, if retraining is required, counselors may consider more extensive training and educational programs. The disease is progressive; however, it is slowly progressive and functional impairments increase at a gradual rate. Most people need accommodation or training for work that requires less exertion, perhaps one or two exertional levels less than prior work activities. Hence, heavy work activities prior to diagnosis will likely need to be restructured to activities involving medium or light work, and performance at the light level will need modification to sedentary work. No matter how severe the disability, the person with rheumatoid arthritis with work modifications should be able to do sedentary work activity. Many can work at the light level, and a few will be able to perform medium work.

Studies on return to work for people with arthritis indicated that work disability is related to age, number of involved joints, and a desire to remain gainfully employed. Control of pace and work activities was shown not to be a significant factor (Reisine, McQuillan, & Fifield, 1995). In a report from a state-federal rehabilitation agency, restoration of physical health, amount spent by rehabilitation agencies, and duration the case was open positively correlated with return to work (Straaton, Maisiak, Dortch, & Lopez-Mendez, 1990). Educational level, severity of disability, and financial status had no relationship to the outcome of vocational rehabilitation. As a result, rehabilitation counselors should persist with retraining, re-education, and vocational placement for people with rheumatoid arthritis unable to work at their current or prior positions.



Continued consultation with the physician, allied health professionals (i.e., occupational and physical therapists), and a work environmental specialist can help accommodate needs in the work environment. Prognosis for work is enhanced for individuals who have higher education levels because, in general, work environments for employees with higher levels of education require less physical exertion and mobility. Since intellectual functioning is not affected by RA and the expected lifespan is only a few years less than normal, rehabilitation counselors need to make every effort to return people diagnosed with this condition to the workforce.

## **Osteoarthritis**

### ***Explanation of Disease State***

Osteoarthritis (OA) is a degenerative disease affecting the articular cartilage lining the surfaces of joints. It is a localized disease and does not have systemic manifestations. Osteoarthritis is an ancient disease; there is anthropological evidence of OA involving the weight bearing joints in fossil skeletons of ancient humankind. This condition refers to a group of overlapping distinct diseases with different etiologies, but similar biologic and clinical outcomes. OA affects the entire joint, including the subcondral bone, ligaments, capsule, synovial membrane, and periarticular muscles (Brandt, Dieppe, & Radin, 2008). Pathophysiologically, there is degeneration of the articular cartilage with fibrillation, fissures, ulceration, and loss of joint surface (Kuettner & Goldberg, 1995).

### ***Effects***

Osteoarthritis is the most common arthritis in the United States, more often affecting women. Virtually everyone over 65 years of age will have x-ray evidence of osteoarthritis; yet, most people do not have symptoms. There is a characteristic wearing away of the surface cartilage at the joint, resulting in pain, swelling, stiffness (particularly with activities), and loss of motion.

The disorder primarily affects the joint cartilage; yet, secondary inflammation involving the joint lining is common. Surface cartilage is disorganized, lost, and adjacent bony growth at the joint margins occurs. Marginal bony spurs are characteristic of osteoarthritis. Diagnosis is primarily based upon symptoms and characteristic x-ray changes, which include loss of cartilage, increased bone density, and bone overgrowth at the joint margin (Lee & Abramson, 2005).

### ***Treatment***

Nonpharmacologic interventions such as exercise, weight loss, modifying activities of daily living, physiotherapy, braces, and use of a cane and orthotics benefit many people. Pharmacologic therapy includes topical agents and oral medications. Acetaminophen is the oral analgesic of choice (Hunter & Lo, 2008). Other patients require the use of narcotic analgesics for pain relief. Joint prostheses (total artificial joints) are effective for advanced OA when lesser treatments prove ineffective.

### ***Functional Limitations***

People with OA do not exhibit systemic features; no other organ systems are involved, and there are no emotional or intellectual consequences. The disorder is localized and may be symptomatic with severe functional problems; more commonly, it does not create major physical limitations. Most people with osteoarthritis are older; many are no longer in the workforce and are not candidates for vocational rehabilitation. Those who are in the workforce have functional limitations that are specific to the areas of the body involved.

If the small digits of the hands are affected, there will be stiffness and loss of motion that interfere with finger dexterity. Pinch and grip may be decreased. Other common areas affected by OA are the cervical and lumbar spine, resulting in pain and loss of motion with rotation of the body and with bending. The weight bearing joints of the lower extremities, especially the hips and knees, are commonly involved and have the highest incidence of disability. People with osteoarthritis of the hips or knees have pain particularly with standing and walking. Additionally, there is loss of range of motion and stiffness with the initiation of activities (Klippel et al., 2008).



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### ***Vocational Considerations***

Emotional, intellectual, organ systems, and other health factors do not limit the vocational process. For individuals that have OA of the lumbar spine, ability to bend and lift is reduced by one or more exertional levels. When the knees and hips are involved, tolerance for bending, squatting, ladder or stair climbing, long distance ambulation, and lengthy periods of standing are reduced. With OA of the hands, fine manual dexterity and strength are reduced.

### ***Rehabilitation Potential***

Persons with localized osteoarthritis and the lack of systemic features are good candidates for rehabilitation. Both short-term and long-term training and educational programs are realistic. Movement is affected, while strength is preserved. Communication skills, social skills, behavioral abilities, the capacity to learn, comprehend, and other intellectual skills are unchanged. Because of the lack of systemic features of this illness and involvement of only particular joints, rehabilitation potential usually is strong.

## **Systemic Lupus Erythematosus**

### ***Explanation of Disease State***

Systemic lupus erythematosus (SLE), commonly called lupus, is a disorder of the immune system, resulting in inflammation of the connective tissue. The disease is characterized by the presence of autoantibodies against the DNA and RNA/protein complexes. Some patients form pathogenic immune complexes (Oelke & Richardson, 2002).

### ***Incidence***

This disorder primarily affects women and has an increased incidence in African-Americans, Hispanics, and Asian Americans. In this country, the incidence of SLE is 3.5 per 100,000 per year in whites and 9.2 per 100,000 per year in African-Americans. The prevalence of SLE is 40-50 per 100,000 population with a total of 239,000 Americans affected. African-American females account for the most prevalent group. Eighty-six percent of people with lupus are women. The peak onset of this disease is during the most productive part of a woman's life, between the ages of 20 to 30 (Harris et al., 2008; Klippel et al., 2008).

### ***Effects***

Systemic features of fatigue, fever, weight loss, and joint pain characterize the disease. All individuals have joint pain as a predominant symptom; yet, major disability is not related to the joints, but rather to systemic involvement. The characteristic clinical course is one of exacerbation and remission; remissions may last for many years. Typically, SLE affects multiple organ systems but not all at the same time. Joint pain is the most common symptom with small joints being most affected. When compared to rheumatoid arthritis, there is no morning stiffness and joint deformity is rare. Some people have skin rashes, especially in sun-exposed areas of the body. There may be inflammation of the lining of the heart and lungs, and acute or chronic renal disease that can progress to renal failure. Central nervous system involvement manifests itself as stroke, psychosis, or depression for some individuals. Anemia is a common problem (Lee & Abramson, 2005; McPhee et al., 2008).

### ***Diagnosis***

The diagnosis of SLE requires at least four of the eleven criteria (at any point in time, not necessarily all at once) that the American College of Rheumatology has set forth. These criteria include the presence of a malar rash, discoid rash, photosensitivity (skin rash from the sun), oral ulcers, non-erosive arthritis, serositis, renal disorder, neurological disorder (seizures or psychosis), hematologic disorder (hemolytic anemia, leucopenia, lymphopenia, thrombocytopenia), immunologic disorder, and antinuclear antibody (Harris et al., 2008; Koopman & Moreland, 2005). Prognosis depends upon the organs involved and is most guarded when there is renal and central nervous system involvement.

### ***Treatment***

Treatment is a function of the organ systems involved. Musculoskeletal symptoms (arthralgias, arthritis, myalgias) are treated with NSAIDS. Corticosteroids are added when symptoms are refractory to NSAIDS, and are generally reserved as temporizing measures when other organ systems are affected (Dooley & Ginzler, 2006). When other organs are involved, immunomodulating drug therapy may be implemented (Harris et. al., 2008). Steroids are often taken concomitantly until other medications take effect. A small percentage of individuals develop chronic renal failure from the nephritis and require dialysis or kidney transplantation. Because of the variable nature and intensity of this disease, some people experience only minor symptoms while others have major symptoms with increased morbidity and even early death. Careful clinical follow-up is indicated for all patients with SLE (Klippel et al., 2008).

### ***Functional Limitations***

Fatigue is a major symptom of SLE and a prominent consideration in job placement. It is particularly apparent during periods of exacerbation, but not during remissions. In contrast to other forms of inflammatory arthritis (e.g., rheumatoid arthritis), there are no problems with mobility, motor strength, or control. Limitations are dependent upon the organ systems affected; pulmonary and cardiac involvement causing shortness of breath limit exertional activities. Persons who have central nervous system involvement may be limited by emotional problems and chronic depression. An individual who has had a stroke may have residual problems of hemiparesis (muscle weakness on one side of the body), which interferes with ambulation and motor skills in the involved extremities.

### ***Vocational Considerations***

Expected lifespan of people with SLE is good unless there is involvement of the central nervous system or the kidneys; hence, educational and training programs that are longer in duration can be considered. Special environmental factors involve avoidance of cold environments, as vasospasm of fingers and toes may occur when exposed to cold objects and environments. Sun intolerance (excessive ultraviolet light exposure) can worsen the illness; jobs with excessive exposure to sun are inadvisable. Most people are restricted to light or sedentary exertion because of fatigue (Straaton et al., 1990).

### ***Rehabilitation Potential***

When SLE is in remission, either naturally occurring or medication induced, the individual will have no physical or emotional impairments. This disorder may have years in which it remains in remission; persons diagnosed with lupus are good candidates for re-training and educational programs. Motor and physical limitations are not a common consequence of this disease.

## **Scleroderma**

### ***Explanation of Disease State***

Scleroderma is a chronic disorder of the connective tissue in which there is inflammation of multiple organs of the body resulting in increased deposits of collagen. Primary target organs are the skin, joints, lungs, gastrointestinal tract, and heart. The etiology is unknown.

### ***Effects***

There may be extensive fibrosis and tightening, particularly of the hands, face, chest, and feet. Inflammation may cause sclerosis (scarring) of the lungs, heart, and gastrointestinal tract. People with scleroderma have great intolerance to cold; cold temperatures cause spasm of the peripheral vessels that may result in gangrene in the tips of the fingers and even loss of entire digits. Deformities are not a common part of this disease. General fatigue, weakness, and loss of motion related to fibrosis about the joints and skin are common consequences.

### ***Treatment***

Treatment is only palliative and symptomatic; non-steroidal anti-inflammatory drugs are given for inflammation and pain. Second line drugs such as D-penicillamine have beneficial effects on interstitial lung disease and renal involvement if given over long periods of time. Other treatments are directed at symptoms associated with specific organs affected by this disease. Pulmonary artery hypertension is the major cause of death in patients with scleroderma (Chang, Wigley, White, & Wise, 2003).

### ***Functional Limitations and Vocational Considerations***

Scleroderma causes impairment of fine motor activities of the hands, as well as manipulations requiring hand strength. Grip and pinch are markedly reduced and the ranges of motion of the wrists, elbows, and shoulders may be mildly decreased. There is no intellectual impairment; mobility is usually not a problem. Individuals with pulmonary involvement have fatigue and restricted activity because of shortness of breath upon exertion. Because of this, jobs within the light and sedentary exertional categories are recommended. Preferred work environments for persons diagnosed with scleroderma are ones that are warm and not subject to extremes or changes in temperature. Job activities should not involve repeated trauma to the hands, since trauma and cold temperatures can result in vasospasm, gangrene, and even loss of fingers. Cognitive functioning is not affected by scleroderma.

### ***Rehabilitation Potential***

Many individuals with scleroderma have minimal limitations. In more severe cases, there are problems with manual dexterity, stamina, and working in cold environments. Other than these limitations and precautions, rehabilitation potential usually is good, with most workers being able to stay on their jobs with possible minor accommodations.

## **Polymyositis and Dermatomyositis**

### ***Explanation of Disease State***

Polymyositis and dermatomyositis are uncommon, having an incidence of 0.9 to 9.3 new cases a year per million people (about the same incidence as systemic sclerosis and 1/3 the incidence of SLE). This is an autoimmune disorder of skeletal muscle in which the muscles are inflamed and lose strength. The usual course of polymyositis is a sudden onset of severe weakness and then recovery. Remissions can last for many years with few exacerbations. There is a bimodal distribution of the onset of polymyositis – at ages 10 to 15 and then ages 45 to 60 (Harris et al., 2008). Dermatomyositis is a condition similar to polymyositis, but one in which a skin rash is present in addition to inflammation of the muscles.

### ***Effects***

The proximal muscles of the extremities are most involved; hence, patients have major involvement of the muscles about the shoulder and hip regions. Diagnostically, one sees elevation of serum levels of enzymes of skeletal muscle, and abnormalities on an electromyogram (EMG). Symptomatically, there occurs an inability to raise the arms above the head, arise from sitting, and climb stairs. For those who do not have full return of strength, there is partial return of strength that can be maintained for years. Only a small percentage of individuals have continued loss of strength.

### ***Treatment***

Most individuals diagnosed with polymyositis are treated with corticosteroids at doses of 30 to 60 milligrams per day until the muscle enzymes normalize. Steroids are tapered slowly over a six month period (Martino & Kagen, 2006). It is estimated that 90% will have a positive response, and 50-75% will go into remission. Those who do not respond well are treated with second line medications, such as azathioprine and methotrexate, either alone or in combination. Alternatively, intravenous immune globulin (IVIg) has been shown to be effective (Dalakas et al., 1993).

### ***Functional Limitations and Vocational Considerations***

When there is not a full recovery, there is either a minor degree of motor weakness (i.e., arising from a chair) or major motor problems (i.e., inability to stand or walk, and capacity to do only activities involving the musculature of the forearms and hands). Individuals in this latter group have functional capacities similar to persons with paraplegia (lower extremity involvement). Those with severe lower and upper extremity concerns have a functional capacity similar to people with quadriplegia. Fortunately, severe extremity complication occurs in only a small percentage of individuals who have this condition. People with polymyositis who have significant weakness of the lower or upper extremities require jobs with the lowest exertional categories, conducting primarily light and sedentary activities. These persons may require walking aids or wheelchairs; therefore, workstations need modification to accommodate these assistive devices. Once the disability is established, it is usually static, providing an opportunity for educational training and retraining programs. Intellect is not impaired; comprehension and learning are maintained.

### ***Rehabilitation Potential***

For the majority of persons with polymyositis, medication effectively controls symptoms. After treatment with various medications, most are able to return to work. Rehabilitation potential, therefore, is positive. For the few who do not respond well to medications, potential for rehabilitation may be poor; alternatively, the individual may have the capacity to do sedentary work activity.

## **Ankylosing Spondylitis**

### ***Explanation of Disease State***

Ankylosing spondylitis (AS) is a chronic inflammatory disorder of the synovial joints and entheses (points at which tendons insert into bones) of the spine. Primarily a disorder affecting men, the incidence is nine times greater for men than women. Initial onset occurs during the second and third decades of life and causes pain, stiffness, and loss of motion of the lower back. Frequently, the disorder is mistaken for low back pain of a mechanical or degenerative nature. AS is the most common inflammatory arthritis of the back. Prevalence is 197 per 100,000. Other related diseases include Reiter's syndrome, psoriatic arthritis, and enteropathic (i.e., bowel associated) arthritis. These disorders have similar symptomatology and disability patterns and are known collectively as spondyloarthropathies (Ritchlin, 2006).

### ***Effects***

This disorder is slowly progressive and, in its full expression, involves the entire spine from the skull to the pelvis. The sacroiliac joints (posterior pelvic joints) are initially involved. Also affected are the synovial joints of the back and ligamentous insertions that join the vertebral bodies. These areas become inflamed resulting in pain, swelling, and loss of motion. Approximately 20% of patients with ankylosing spondylitis have involvement of the peripheral joints, primarily the hips and shoulders. To a lesser extent, there is involvement of the more peripheral joints of the lower extremities and hands (Harris et al., 2008).

### ***Symptoms***

Pathology of AS involves inflammation of the lining of the posterior joints of the back and ligamentous connections between vertebral bodies. With progression of the disease, there is fibrous ankylosis (fusion) and later, bony ankylosis of the affected joints with consequent loss of all motion in the back. Pain is the main symptom, along with stiffness and gradual progressive loss of motion. When ankylosis is complete, the pain stops but loss of motion in the spine remains.

### ***Treatment***

Treatment of AS is symptomatic, primarily with NSAIDs. Exercise helps retain range of motion (Ritchlin, 2006). In some cases, sulfasalazine can be useful in reducing the length and severity of morning stiffness and pain. Physical therapy with maintenance of proper posture is indicated to prevent development of a bent fixed flexion posture of the spine (Klippel et al., 2008).

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### ***Functional Limitations***

Ankylosing spondylitis does not have systemic features; occasionally, patients have involvement of the aortic valve of the heart and pulmonary fibrosis with resultant symptoms. Systemic features are not correlated with loss of work capacity. In the majority of people, functional limitations are related to loss of motion of the spine. Bending, twisting, and rotational motions of the lumbar and cervical spine are impaired. If the hips and shoulders are affected there will be pain, loss of motion, and development of flexion contractures in those areas. Involvement of hips, shoulders, and peripheral joints commonly lead to loss of ability to work.

### ***Vocational Considerations***

Intellectual functioning is unimpaired. Aptitudes, interests, and communication skills are similar to the normal population and life expectancy is unchanged, unless there is cardiac or pulmonary involvement. There are no special environmental needs.

Early in the disease, most people with AS have chronic pain, but little impairment in range of motion in the cervical and lumbar spine. Anti-inflammatory medications and analgesics control this; hence, little modification of the work environment is needed. However, most individuals require reduction of exertion in work activities. As the disease progresses, the person becomes unable to perform work that is within the higher exertional categories because of pain and loss of mobility of the lumbar spine, including ability to bend forward. Individuals with the most severe form of ankylosing spondylitis lack motion of the cervical and lumbar spine and must bend from the hips; they need to move their entire body to see to the right or left. Life expectancy is not affected by ankylosing spondylitis and most people have full, productive lives. Social and behavioral skills are unimpaired, learning comprehension is unchanged and, with the exception of the few who have cardiac involvement, there are no other health problems associated with this condition (Harris et al., 2008; Koopman & Moreland, 2005).

### ***Rehabilitation Potential***

Rehabilitation potential for persons with this rheumatic disease generally is strong. With work accommodation for those individuals who have heavier jobs, most maintain productive employment for many years as long as they stay within the lifting and carrying restrictions recommended, usually within the light and sedentary levels. Persons with chronic back pain from ankylosing spondylitis may need jobs that do not involve undue emotional stress. With these factors in mind, the counselor can effectively rehabilitate the majority of people with this disorder.

## **Case Study**

Susan Valdez is 19 years of age and has had juvenile-onset rheumatoid arthritis for 14 years. During this time, she has had long periods of illness where she was confined to her home and, as a result, is still in her last year of high school. Her illness is currently well controlled and Susan has missed no school for the past six months. She takes oral medication three times daily, self-administers subcutaneous injections of etanercept twice a week, and visits her physician on a monthly basis to attempt to maintain her disease in remission.

To maintain ambulation, Ms. Valdez has required surgery for her arthritis. She has had two hip replacements and a knee replacement. Currently, she is able to walk about the house unassisted but uses a motorized wheelchair for long distances when out in the community. At the current time, Susan experiences morning stiffness and has arranged to take classes at school beginning at 10:00 A.M. or later. Occasionally, she has pain in multiple joints including her feet, shoulders, wrists, and the small joints of her hands. Her physician has prescribed pool exercise and Susan wears a wrist splint when she experiences pain in that area. In addition to her limited ambulation, she requires assistance from her mother to shampoo her hair, has a bilateral grip strength of 20 pounds, and pinch strength of five pounds.

Ms. Valdez is the fifth child of a second generation Hispanic family, and she is bilingual in Spanish and English. Her father passed away and general relief and contributions from the adult children, including Susan's Social Security Insurance (SSI) payments, currently support her mother. Susan's mother does not



speak English and has never worked outside the home; she sees her life's role as caretaker for her daughter. The relationship between Susan and her mother is close and has been criticized by the health care team as one that promotes dependence rather than independence.

Psychological testing has shown that the Ms. Valdez has high average intelligence, social immaturity, and the absence of depression. The health care team feels that Susan has been an underachiever in school and at home, relying on her mother and siblings rather than taking the initiative. Susan does not drive and has not started dating. Her primary source of emotional support is her family and one girlfriend from school.

The school principal referred Susan for vocational counseling. In six months, she will graduate from high school, and she has no plans after graduation. Ms. Valdez has done well in school and has particularly enjoyed the sciences, receiving a senior prize in biology. Unsure about attending college, Susan will be the second in her family to complete high school; there are no finances to support a college education. Her sister is currently the head cashier at a grocery store and has assured Susan that she can get her a cashiering job at that store. Susan's mother is against her working and feels that she should stay home and baby-sit for her nieces, nephews, and the neighborhood children.

### **Questions**

1. Is this individual a suitable candidate for vocational rehabilitation counseling services? What additional medical information is needed from her healthcare team?
2. If you feel Ms. Valdez is a suitable candidate for counseling and rehabilitation, what programs would give her the greatest opportunities for long-term employment? Is additional education indicated?
3. Discuss whether the job as grocery cashier with her sister is a good choice for employment.
4. How might her counselor handle Susan's inability to drive and her dependence upon her mother? Is reliance upon public transportation an option? Consider the impact of her motorized wheelchair on further education, training, and work.
5. How will Susan's current illness, physical limitations, and psychological status influence a decision about higher education? Is she a viable candidate for college, or would educational efforts be problematic? Should vocational training, higher education, or on-the-job training be considered?
6. What physical problems need to be considered if job placement is to be conducted? Indicate the impact upon Susan's ability to obtain and maintain employment.
7. List Ms. Valdez' strengths and limitations from a physical and psychosocial perspective that need to be evaluated when formulating vocational planning.
8. Discuss cultural implications regarding this case.

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## Chapter 10

# THE ACQUIRED IMMUNODEFICIENCY SYNDROME

*John J. Howard, M.D., M.P.H.*

### Introduction

This chapter presents a comprehensive overview of the scientific and medical aspects of infection with the human immunodeficiency virus (HIV) from the standpoint of the rehabilitation professional. First, the chapter discusses the modes of HIV transmission and the natural history of HIV infection. Next, the clinical manifestations of HIV disease are reviewed, including the most common infections and tumors which define the Acquired Immunodeficiency Syndrome (AIDS). The chapter then describes preventive vaccines as well as treatment strategies used against HIV itself and against the different types of HIV-related infections and tumors. Lastly, the chapter focuses on the functional and vocational limitations associated with HIV infection and AIDS and on the rehabilitation potential for persons affected by HIV.

In 1981, the first five cases of AIDS were described medically (Gottlieb, 1981). Since then, HIV has infected more than 65 million persons and more than 25 million have died of AIDS. Globally, there are an estimated 33 million people living with HIV (World Health Organization [WHO], 2008). The countries of sub-Saharan Africa have been significantly affected by the AIDS epidemic. In 2007, Sub-Saharan Africa accounted for 67% of all people living with HIV and for 75% of AIDS deaths (WHO, 2008). In the United States, more a million AIDS cases have been reported to the Centers for Disease Control and Prevention (CDC) and 500,000 deaths (CDC, 2006a). Using a new system designed to more accurately estimate the number of new HIV infections in any given year, 56,300 new infections occurred in the United States in 2006 (Hall et al., 2008).

Since its recognition as a new clinical syndrome in 1981, AIDS not only has had a profound impact on the medical world, but has also affected every social institution in American life. The problems caused by the AIDS epidemic have touched families, schools, communities, business, industry, courts, religious institutions, and government at all levels. AIDS has brought many controversial issues to the forefront of public discussion. These include discrimination against HIV-infected persons in housing, employment, and health care; the economic cost of providing care to persons with AIDS; the duty of government to protect its citizens from communicable diseases like AIDS; and the obligation of physicians and nurses to provide care for persons with AIDS.

The epidemic's most profound effect has been on those who have become infected with HIV. HIV causes progressive deterioration of the body's immune system, increasing an individual's susceptibility to life-threatening infections and tumors. Without treatment, the median period for transition from HIV infection to AIDS is 8-10 years (Brookmeyer & Gail, 1994).

Twenty-five years since the beginning of the AIDS epidemic, important medical advances in treating HIV infection and AIDS-related conditions are occurring, helping to improve the quality of life for persons with HIV infection and AIDS. Anti-retroviral therapy was one such medical advance introduced in 1996 (so-called highly active anti-retroviral therapy or "HAART"). Another is the development of more effective treatments for the specific infections and tumors that occur in persons with AIDS. Between 1994 and 1997, AIDS deaths decreased by 75% and the occurrence of AIDS-defining diseases decreased by 73% (Palella, Delaney, Moorman, & the HIV Outpatient Study Investigators, 1998).

Due to medical advances, many persons with HIV/AIDS now function at higher levels for longer periods of time than they were able to function when the epidemic first began in the 1980s (Dybul et al., 2002). Living with AIDS, however, still demands a tremendous expenditure of physical and emotional energy. Frequently, individuals with HIV infection and AIDS can no longer function at their previous levels of activity, and many stop working.

When AIDS was first recognized as a medical condition in 1981, few thought that persons with AIDS would ever have rehabilitation potential. At that time, AIDS was medically diagnosed only after the virus had already destroyed the immune system, and little could be done medically to prolong life. Persons with AIDS died within a short time following diagnosis. Before the advent of newer classes of anti-HIV drugs, 95% of persons with HIV infection died of AIDS within 2 to 15 years (Senechek, 1997). Medical progress has quickly transformed AIDS from a severely activity-limiting condition with a very short survival time, into a condition with significant rehabilitation potential.

In spite of medical advances, limitations caused by societal discrimination continue to be a challenge. Negative attitudes of the public and the medical community make rehabilitation of persons with HIV/AIDS difficult. Fortunately, such negative attitudes are gradually being replaced with more positive approaches to rehabilitative care due to the discovery of new medical treatments and rehabilitation approaches. Many HIV-affected persons can benefit from early recognition of rehabilitation potential, establishment of therapeutic goals, and prompt referral to a rehabilitation counselor.

## **Origins and Modes of HIV Transmission**

Much speculation has centered on the origin of HIV. Only now, some 25 years after its emergence as a pathogen of staggering proportions in the human population, is solid information emerging about HIV's origins. HIV-1, the more virulent strain of HIV, shows a close resemblance to a strain of a simian immunodeficiency virus (SIV) called SIVcpz which infected several geographically isolated chimpanzee communities in southern Cameroon (Taylor, Sobieszczyk, McCutchan, & Hanner, 2008). HIV-2 demonstrates a resemblance with an SIV that is found in the sooty mangabey, an animal indigenous to western Africa (Heeney, Dalgleish, & Weiss, 2006).

Although the precise mechanism of HIV-1 transfer from animal to human is unknown, speculation centers on butchering of chimpanzees and other animals for food and subsequent human contamination with SIV-infected blood (so-called "Hunter Theory"). Viral gene studies have shown that HIV actually originated in central Africa sometime around 1930 (Keele et al., 2006), spread among humans along the Congo River into Kinshasa, Zaire where the earliest documented case of HIV-1 infection in humans has been traced to a blood sample from 1959 (Zhu et al., 1998). The virus then moved from Africa to Haiti in 1966 and from there to the United States in 1969 (Gilbert et al., 2007).

HIV is a member of a family of Ribonucleic acid (RNA)-containing viruses, first discovered in the 1970s, called "retroviruses." Retroviruses are unique from other viruses in that retroviruses initially replicate in reverse (from RNA to DNA [Deoxyribonucleic acid] instead of DNA to RNA). Their ability to replicate in reverse is made possible by an enzyme unique to retroviruses called "reverse transcriptase." Once HIV converts itself from an RNA to a DNA form, the "translated" HIV viral DNA can then insert itself into the body's own genes (which are made of DNA), where it can remain dormant for long periods of time. The ability of HIV to integrate itself into human genes makes it difficult to completely eradicate from the body. Consequently, infection with HIV is a lifelong occurrence.

By the end of the 1970s, researchers had discovered that retroviruses cause disease in several animal species, but no immunosuppressive retroviruses were found to cause disease in human beings. In 1981, the first cases of an immunosuppressive disease began to appear in young gay males in the United States. The disease produced similar immunological effects in humans as those seen in animals infected with other types of retroviruses (Essex & Kanki, 1988). By the time an HIV antibody detection test was developed in 1984, most researchers had accepted that HIV was the causative agent of AIDS.

HIV infection is established in human beings through the introduction of blood cells or bodily fluids from an infected individual into the bloodstream of an uninfected person. This occurs either through sexual or blood-related contact.



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## ***Sexual Contact***

Sexual transmission is the most common mode of HIV transmission. Penetrative sexual intercourse involving the passage of HIV-containing white blood cells and bodily fluids, such as semen or vaginal secretions, can result in HIV transmission. The efficiency with which sexual contact transmits HIV varies with the particular type of sexual practice and gender of the sexual partners involved. Epidemiologic data points to anal intercourse between two males, especially for the receptive partner, as the most efficient means of sexual transmission. Despite being less efficient, penile-vaginal intercourse is the major means of sexual transmission in the world today (WHO, 2008).

## ***Blood Contact***

The second means of HIV transmission is blood-related contact, which can occur in five separate ways. The first two major types of blood contact involve transfusion of blood products. First, whole blood from an HIV-infected blood donor can be transfused to an uninfected person during medical emergency surgery. Second, a particular blood-clotting factor isolated from the whole blood of an HIV-infected blood donor can be transfused into an uninfected person who has hemophilia (a blood-clotting disorder). These two routes of HIV transmission have decreased in the United States due to mandatory HIV screening of all blood donations. Further, blood used for the preparation of clotting products for persons with hemophilia now undergoes a heat treatment process to inactivate HIV.

The third type of blood contact, intravenous drug use, accounts for most blood-related HIV transmissions. Intravenous drug users often share their “equipment” (e.g., needles or syringes) with other drug users. This equipment can contain minute amounts of HIV-infected blood and, if shared with an uninfected person, HIV transmission can occur.

HIV is also transmitted from an infected pregnant woman to her fetus. This route of HIV transmission, called maternal-fetal transmission or “vertical transmission” is preventable. This form of transmission accounts for most of the cases of pediatric AIDS. Vertical transmission can occur in one of two ways. The blood-borne, or intrauterine, route involves the fetus becoming HIV-infected because the mother was infected before giving birth. Mucosal, or intrapartum, transmission occurs when a neonate becomes infected during passage through the birth canal. Before the advent of more effective medical therapies, approximately half the infants born to HIV-infected mothers developed clinical AIDS. The use of newer therapies, however, dramatically decreased the proportion of babies that develop AIDS from HIV-infected mothers.

Lastly, HIV can be transmitted inadvertently during an accident in a health care setting. The routes of transmission that occur in these occupational settings are needlestick (sharp object injuries) and direct skin or mucous membrane contact with blood or other potentially infectious materials. For example, HIV can be transmitted when an uninfected health care worker is stuck with a needle or cut with a scalpel which contains blood from an HIV-infected patient (Marcus & CDC, 1988). HIV transmission can also occur when blood contaminated with HIV comes into direct contact with the broken skin or the mucous membrane of an uninfected person. The greatest risk in occupational exposure occurs when a healthcare worker sustains a needlestick injury with a large volume of blood from a patient whose blood contains a high HIV concentration (Daily, 1997). The mainstay of risk reduction for health care workers has focused on treating all blood as if it carries HIV (so-called “universal precautions”) and designing “safer” needle devices to prevent percutaneous exposure.

## ***Prevention of HIV Transmission***

The most effective means of preventing HIV infection is avoiding sexual or blood exposure. Much of AIDS public health education is directed at the interruption of the sexual transmission of HIV by encouraging people to engage in “safe sex” practices, e.g., using “condoms” during sexual intercourse and by avoiding shared needle behaviors.

If a healthcare worker sustains occupational exposure to blood or other body fluids that might contain HIV, post-exposure prophylaxis with anti-retroviral medications is recommended. In addition, for persons exposed within 72 hours to blood, genital secretions, or other potentially infectious body fluids of a

person known to be HIV infected, when that exposure represents a substantial risk for transmission, CDC recommends a 28-day course of HAART (Daily, 1997).

### ***Vaccine Development***

Historically, preventive vaccines have achieved enormous success against infectious diseases such as polio, smallpox, yellow fever, and measles; development of a vaccine against HIV has become a high priority. However, development of an HIV vaccine has proven to be a challenge for several reasons: (1) the natural immune response against HIV is inadequate and, once primary infection is established, fails to eradicate the virus; (2) HIV quickly integrates itself into the DNA of the host cell, where it can “hide,” remain latent, and essentially invisible to the immune system; (3) latency is established very early after infection, within days to weeks, making the opportunity to eradicate HIV through an immune response very short; and (4) the extraordinary ability of HIV to mutate makes it substantially more complex than any other human virus and potentially making any vaccine obsolete (Johnston & Fauci, 2008). HIV infects some of the same cells any vaccine would have to activate to be effective. If these cells are activated by a vaccine, the result may be unrestrained HIV growth. Despite these formidable obstacles, more than 20 AIDS vaccine formulations have been tested since 1988 with little success (Johnston & Fauci, 2007). Too little safety and efficacy information is currently available to suggest that an effective vaccine will be discovered any time soon. Yet, there is no doubt in anyone’s mind that HIV vaccines are the best hope to end this worldwide pandemic.

## **The Natural History of HIV Infection**

Most people infected with HIV are initially unaware of the infection. A few individuals experience symptoms of a condition called “acute retroviral syndrome,” but the majority remain asymptomatic. Up to a decade may pass before an individual infected with HIV experiences symptoms. This long latent period is considered the most dangerous, as infected individuals may unknowingly transmit HIV to others before they exhibit any symptoms.

### ***Acute Retroviral Syndrome***

The natural history of initial HIV infection is characterized by a very high level of HIV viremia (the presence of virus in the blood) and a vigorous immune response. During the peak of HIV viremia, the CD4+ helper T-lymphocyte cell count declines precipitously and the CD8 cytotoxic T-lymphocyte cell count increases to fight the invading HIV. Within 5 to 30 days of initial HIV infection, during the peak of HIV viremia and CD4+ and CD8 immunological activity, some individuals become acutely ill with symptoms of acute retroviral syndrome (Rosenberg, 1997).

Acute retroviral syndrome manifests itself as a flu-like illness characterized by fever, sore throat, fatigue, weight loss, and myalgias (muscle aches). Common physical findings include fever, lymphadenopathy (enlarged lymph nodes), and a diffuse body rash. The median duration of acute retroviral syndrome is 14 days. Many newly infected persons do not experience such an acute illness at all or may attribute their illness to the “flu.” Whether or not an individual experiences symptoms during the initial HIV infection, his or her levels of HIV are very high, and as a result, newly-infected individuals are quite contagious through intimate contact.

### ***HIV Antibody Formation***

A prolonged period of well-being follows recovery from acute retroviral syndrome, during which time the HIV viral load in the blood decreases, the number of CD4+ T-lymphocytes increases, and HIV antibodies become detectable. Such antibodies usually develop within 9 to 180 days after HIV transmission, but a minority of infected individuals can remain antibody negative for up to three years following infection (Brookmeyer & Gail, 1994).

For most individuals, realization of HIV infection does not occur until after the first time they undergo a blood test for the presence of HIV antibodies and are told they are “HIV positive” or “seropositive.” HIV antibody testing is usually performed in an outpatient clinic setting. “Home test collection systems” (where the individual collects the blood at home, sends the sample to a laboratory, and

then telephones an information line for the result) and “true home tests” (where the individual actually performs the test at home) are increasingly being utilized. In late 2006, the CDC issued recommendations that urged all health care providers in the United States to include HIV testing as routine part of their patients’ healthcare; in this way, more people can learn whether they are HIV-infected, allowing them to benefit from earlier access to treatment and reducing the risk of infecting their partners (CDC, 2006b).

Development of an antibody response to the presence of HIV in the blood is the body’s way of attempting to eliminate the invading virus and is called the host immune response. These antibodies are detectable throughout the course of HIV infection and decline only late in the course of HIV disease when the person has advanced HIV disease or AIDS. Unlike many of the body’s other antibodies that are protective against viruses, HIV antibodies do not protect the individual against progressive destruction of the body’s immune system. The presence of detectable HIV antibodies in the blood is the most common way a person is diagnosed as being HIV positive (CDC, 1992).

### ***Progressive Immunodeficiency***

Even though a person can remain asymptomatic for long periods after being infected with HIV, the virus begins to destroy the body’s immune system almost immediately after HIV infection occurs. The virus spreads throughout the body, infecting the entire immune system (including circulating blood cells like CD4+ T-lymphocytes and stationary tissues such as the lymph nodes) and many other cells of the body (Stebbing, Gazzard, & Douek, 2004).

The immunological destructive process begins on the cellular level when the virus attaches itself to a white blood cell, the CD4+ T-lymphocyte. Once inside the white blood cell, HIV replicates into hundreds of new HIV particles, a process which kills the CD4+ T-lymphocyte. Since this cell is responsible for coordinating the body’s immune response to certain invading microorganisms, the destruction of CD4+ cells leaves the person prone to infections and tumors. Destruction of the CD4+ cells which are crucial to the body’s immune defense is the major cause of the progressive immune dysfunction (immunodeficiency), the clinical and laboratory hallmark of HIV infection and disease. HIV-infected individuals are often categorized into three groups based on their number of CD4+ T-lymphocytes: Category 1 (greater than or equal to 500 cells); Category 2 (200 to 499 cells); and Category 3 (less than 200 cells) (CDC, 1992).

The destruction of CD4+ T-lymphocytes due to continuous low level viral replication is a constant feature of the infected state. At the time of initial HIV infection, there is a short but intense burst of HIV replication. The body responds by forming HIV antibodies and producing a surge in the number of CD8 lymphocytes. As a result, viral replication partially subsides. In fact, the laboratory discovery that HIV replication is very rapid during the clinical latent period (average daily production of  $10^{10}$  HIV copies) was the impetus for development of a new approach to treating infection, called “triple drug therapy” (Dybul et al., 2002). As a result of the knowledge gained about HIV viral dynamics that occur during the latent period, earlier and more aggressive antiretroviral treatment of HIV is now widely practiced.

Throughout the latent period, while infected individuals are without symptoms, HIV continues to replicate, damaging an increasing amount of CD4+ cells. The pace of this destructive process against the immune system varies. Even though the vast majority of HIV-infected individuals experience several years of relative immunological and clinical stability (median time period is about 10 years), a small percentage experience rapid immunological progression to advanced HIV disease and AIDS despite adequate anti-retroviral therapy (“rapid progressors”). Approximately 5% of HIV-infected persons maintain a normal CD4+ T-lymphocyte cell count and an undetectable viral load for long periods of time (“long-term nonprogressors”).

## **Clinical Manifestations**

### ***Asymptomatic HIV Infection***

After the acute retroviral syndrome subsides, an infected individual may continue to remain without symptoms for a prolonged period. This latent period, the period of time from the primary HIV infection to the development of symptoms (HIV disease), can range from 7 to 14 years in length. During this period, few clinical events occur to warn the individual that he or she is infected with HIV. The only condition which

occurs at a higher frequency in asymptotically infected individuals than in non-infected individuals is a painful skin eruption called herpes zoster (commonly called shingles), which represents a reactivation of the previously dormant chickenpox virus (Senechek, 1997). Otherwise, only laboratory tests, can reveal the effects of HIV infection.

During the long asymptomatic period following primary infection, the virus is not quiescent. HIV gradually depletes the body's supply of CD4+ T-lymphocytes, resulting in a progressively worsening immunodeficiency. Studies have shown that the viral load (measured in number of copies of HIV per milliliter of blood) correlates with the development of HIV disease and death, i.e., the higher the number of HIV copies present in the blood, the faster AIDS develops. The goal of high active anti-retroviral therapy or HAART is to achieve an "undetectable" level of HIV copies in the bloodstream (Dybul et al., 2002). To assess direct damage to the body's immune system, physicians also use a surrogate marker of viral activity, the quantitative CD4+ T-lymphocyte count. During the latent period, a physician can deduce the rate at which immune system destruction is occurring by serially measuring the number of CD4+ cells. A declining number indicates evidence of "progression" to a more advanced state of immunodeficiency and the development of early HIV disease.

### ***Early HIV Disease***

When the AIDS epidemic was initially identified, the CDC epidemiologically "defined" AIDS to facilitate tracking the number of persons who manifested the syndrome. According to the original 1981 CDC surveillance case definition, AIDS existed under two conditions. First, the individual had to be diagnosed with one of the qualifying types of infection or tumor, which was moderately indicative of the presence of a cellular immunodeficiency. Second, the individual could not have any known medical reason to have such an infection or tumor, such as receiving chemotherapy for cancer or receiving medications associated with the development of immunosuppression (CDC, 1981). In 1986, the AIDS surveillance case definition was refined to take account of the development, in 1984, of the HIV antibody test (CDC, 1987).

In 1992, the CDC recognized that HIV infection resulted in a spectrum of levels of disease from asymptomatic to early to advanced. The criteria for HIV infection for persons over 13 years of age include: (1) a positive HIV antibody screening and confirmatory test; and (2) direct identification of the virus in host tissues by isolation, HIV antigen detection, or a positive result by any other highly specific licensed test for HIV. In their 1992 revision, the CDC emphasized the importance of measuring the number or percentage of CD4+ T-lymphocytes and provided three categories: Category 1, which means greater or equal to 500 CD4+ lymphocytes per microliter; Category 2, which indicates 200 to 499 CD4+ cells per microliter; and Category 3, which means less than 200 CD4+ cells per microliter. Currently, an individual is said to have "AIDS" when they have laboratory confirmation of HIV infection (e.g., a positive HIV antibody test) and a CD4+ T-lymphocyte count of less than 200 CD4+ cells or a CD4+ T-lymphocyte percentage of total lymphocytes less than 14 percent (CDC, 1992).

In addition to laboratory testing, CDC has defined three clinical categories of HIV infection (CDC, 1992). Category A consists of asymptomatic HIV infection, persistent generalized swelling of the lymph nodes, or acute retroviral infection. Category B consists of a variety of symptomatic conditions that meet at least one of the following criteria: (a) the condition is attributed to HIV infection or is indicative of a defect in cell-mediated immunity; or (b) the condition is considered by a physician to have a clinical course, or to require management that is complicated by HIV infection. Examples of Category B conditions include: oral candidiasis (thrush), persistent vulvovaginal candidiasis, oral hairy leukoplakia, herpes zoster (shingles), and peripheral neuropathy.

In general, early symptomatic HIV disease (Category B) is characterized by relatively non-specific signs (what the physician notices) and symptoms (what the patient notices). These include swollen glands or lymph nodes, mild intermittent fever, and fatigue or low energy. In people with early symptomatic HIV disease, these signs and symptoms worsen while new symptoms appear. Left untreated, HIV-infected individuals begin to manifest daily fever, night sweats, fatigue and weakness, weight loss, intermittent diarrhea, oral thrush or candidiasis (yeast growing on the tongue and along the sides of the mouth), and experience various immunodeficiency-related skin conditions or rashes.



## ***Advanced HIV Disease and AIDS***

The CDC's Clinical Category C includes conditions that result in advanced HIV disease or AIDS and include: candidiasis of the lungs or esophagus; invasive cervical cancer; disseminated or extrapulmonary coccidiomycosis; extrapulmonary cryptococcosis; cytomegalovirus eye disease with loss of vision; encephalopathy; herpes simplex ulcers of over one month in duration or involving the lungs or esophagus; disseminated or extrapulmonary histoplasmosis; chronic intestinal isosporiasis; Kaposi's sarcoma; Burkitt's, immunoblastic or brain lymphoma; tuberculosis; disseminated or extrapulmonary mycobacterium avium complex; pneumocystis carinii pneumonia; recurrent pneumonia; progressive multifocal leukoencephalopathy; salmonella septicemia; toxoplasmosis of the brain; and wasting syndrome. Once a Category C condition has occurred, the person remains in Category C whether or not their general condition improves with treatment.

Several different bacterial, viral, fungal, or protozoal organisms in the aforementioned list can cause severe illness in persons with advanced HIV disease. These organisms share in common an inherently low level of virulence or ability to harm individuals with competent immune systems. Since these microorganisms rarely cause disease in immunocompetent individuals, they are called "opportunistic infections." Opportunistic infections take advantage of the "opportunity" of the body's defenseless posture caused by HIV to produce infection. Before the introduction of HAART in 1996, the most common "opportunistic" infectious disease that was seen in advanced HIV disease was *Pneumocystis carinii* pneumonia or "PCP" (Thomas & Limper, 2004). Recently, though, the number of patients with advanced HIV disease and AIDS who present with PCP has declined significantly and the most frequent complications of advanced HIV or AIDS involving the chest have become acute bronchitis, bacterial pneumonia, and, increasingly, hospitalizations for cardiac conditions (Grubb, Moorman, Baker, Masu, & the HOPS Investigators, 2006).

The most common tumor seen early on in HIV-infected persons is Kaposi's sarcoma. It most commonly affects the skin, resulting in a violet-colored skin tumor (in lightly pigmented individuals), but it can also occur in lymph nodes and in the lining of the gastrointestinal or respiratory tracts. Unlike opportunistic infections, the development of Kaposi's sarcoma is not necessarily related to the level of immunodeficiency present and HIV-infected persons with minimal immunodeficiency have developed Kaposi's sarcoma.

## **Treatment Strategies**

When researchers first discovered HIV as the causative agent of AIDS, many expressed doubt that a drug capable of successfully controlling HIV infection would be found. Despite this initial pessimism, a number of classes of antiretroviral medications have been discovered to be effective against HIV. In 1986, a nucleoside analog called zidovudine (AZT) was the first antiretroviral medication shown to prolong the lives of persons with AIDS. In the 1990s, significant knowledge about the life cycle of HIV was discovered, making it possible to design drugs that interrupt specific phases of the life cycle of HIV. As a result, several new classes of drugs (like protease inhibitors and non-nucleoside reverse transcriptase inhibitors) were introduced. In 1996, the widespread use of these newer medications, has markedly reduced the mortality rate associated with AIDS. The AIDS case-fatality rate has dropped from a high of 90% in 1981 to a low of 5% in 1997 (CDC, 1998).

All this success in the treatment of AIDS has given rise to the hope that HIV infection can be completely eradicated from an infected individual after the administration of a sufficiently lengthy period of antiretroviral suppressive therapy. Two obstacles, however, lie in the path of achieving complete HIV eradication. First, there are various HIV-infected tissue compartments, such as the germinal centers of lymph nodes, which cannot be reached by antiretroviral drugs. Unless HIV can be destroyed in these so-called "sanctuary sites," complete eradication of the disease is impossible. Second, even after an undetectable level of HIV is attained following a lengthy period of antiretroviral suppressive therapy, HIV is still capable of regrowth. This is possible because of the prolonged life span of some "latently infected cells," which, after undergoing cell division, can serve as a source of infectious HIV (Wong et al., 1997a, 1997b) As a result, the goal of complete viral eradication of HIV remains elusive.



Much of HIV treatment is experimental and therefore, subject to rapid medical changes. Treatment of HIV infection and AIDS is a rapidly evolving field. A particularly promising experimental therapy can turn out to lack effectiveness or be too toxic for use in humans. With these limitations in mind, a discussion of treatment strategies for HIV infection and AIDS can be divided into three conceptual categories: primary, secondary, and preventive treatment. Preventive treatment has already been discussed.

### ***Primary Treatment***

Primary treatment can be directed either at interrupting the life cycle of the virus itself (antiretroviral therapy) or aimed at positively modulating the body's immune response to the virus (immunomodulator therapy). Both types of primary treatment are designed to contain HIV growth, one by suppressing viral growth, the other by boosting the body's protective immune response against HIV. Anti-retroviral therapy and immunomodulator therapy both have the goal of preventing the development of secondary infections and tumors associated with advanced HIV disease or AIDS (see Category C infections and tumors listed above).

Anti-retroviral therapy can be categorized by which point in the life cycle of the virus-host cell interaction the particular class of medication intervenes. Currently, there are three major classes of antiretroviral medications in therapeutic use in HAART regimens: (1) nucleoside analogs ("NRITs"); (2) non-nucleoside reverse transcriptase inhibitors ("NNRTIs"); and (3) protease inhibitors ("PIs").

The majority of anti-retroviral agents currently in use, both clinically and experimentally, interfere with an enzyme called reverse transcriptase, essential to HIV replication after it enters the host cell (usually a CD4+ T-lymphocyte). The most prominent reverse transcriptase inhibitor is zidovudine (AZT). In 1987, the United States Food and Drug Administration (FDA) first approved AZT for use in adults with AIDS or in persons with symptomatic HIV infection who have fewer than 200 CD4+ lymphocytes per microliter (CDC CD4+ Lymphocyte Category 3). In early 1990, the FDA expanded the use of AZT to include asymptomatic HIV-infected persons with fewer than 500 CD4+ lymphocytes (Category 2) (Volberding et al., 1990). Its expanded use in the large population of asymptomatic individuals with HIV infection has been termed "early intervention therapy." Early intervention therapy was shown in 1990 to slow the progression from HIV infection to AIDS (Friedland, 1990). Since then, early intervention with a HAART regimen has become the standard primary treatment for HIV infection.

Nucleoside analogs are medications that block HIV replication in its early phase by binding to HIV reverse transcriptase. Nucleoside analogs are the oldest antiretroviral medications in use and include such drugs as: AZT (Retrovir), ddI (Videx), ddC (Hivid), 3TC (Epivir), and d4T (Zerit or Stavudine). Non-nucleoside reverse transcriptase inhibitors (NNRTIs) also block HIV replication in the early phase by binding to reverse transcriptase. Protease inhibitors block HIV replication in the late phase by binding to HIV protease which stops maturation of newly formed HIV virus, and as a result the capsid of the virus does not form properly (Stix, 2006).

Anti-retroviral medications of these three major classes are now used in combination. The most common approach, called "HAART" is the simultaneous administration of three to four effective antiretroviral medications. The therapeutic aim of combination therapy is complete suppression of HIV growth as measured by the viral load. An "undetectable" viral load serves as laboratory confirmation that HIV growth has been suppressed. HAART therapy has been credited for prolonging the life of persons living with advanced HIV disease or AIDS by an estimated 13 years (Walensky et al., 2006).

Newer classes of anti-retroviral drugs include fusion inhibitors which bind to the human chemokine receptor CCR5, blocking HIV from binding to the CD4+ T-lymphocyte, and thereby interfering with an essential early step in viral replication (Poveda, Briz, Quinones-Mateu, & Soriana, 2006). Integrase inhibitors are another new class of anti-retroviral drugs that inhibit the complex, multi-step process of integration of HIV provirus into the host genome and are being used clinically to combat drug-resistant HIV (Havlr, 2008).

HIV's inherent mutability can make it resistant to even the newest anti-retroviral medication or an entire class of medications. The phenomenon of viral resistance was demonstrated shortly after the introduction of the first anti-retroviral, AZT (Larder, Darby, & Richman, 1989). As many as half of HIV

infected persons under treatment have been found to be infected with viruses that are resistant to at least one drug in their current combination therapy regimen (Richman et al., 2004). When resistance is identified, the patient must have their drug regimen changed to continue viral suppression. Even under current HAART regimens, patients must continue taking medications, and constantly changing them because of residual replication-competent HIV hiding in viral reservoirs throughout the body. However, innovative approaches are being tried to purge HIV from these latent viral reservoirs which would greatly aid in long-term suppression of the virus (Marsden & Zack, 2009).

Even when viral suppression is achieved, anti-retroviral medications can produce serious side effects which often necessitate discontinuation of a medication. The most common side effects from nucleoside analogs like AZT include lowered red cell count (anemia), fatigue, headaches, sore tongue, nausea, vomiting, muscle aches, liver and pancreas inflammation, and numbness and pain in the arms and legs (peripheral neuropathy). As each new medication is introduced into practice, new side effects emerge. Since the introduction of protease inhibitors in 1996, for instance, several new potentially serious side effects have been noted in patients treated with these drugs. These side effects include: elevated blood sugar (diabetes mellitus), elevated fats in the blood (hypertriglyceridemia), and fat wasting of face and limbs with central obesity (peripheral lipodystrophy) (Dube & Sattler, 1998).

Primary treatment also can be directed at “boosting” the function of the body’s immune system (immunomodulation). Several drugs are available that do not interrupt the life cycle of HIV within the body, but improve the immune system’s ability to defend itself. For example, chemicals that are naturally produced by the cells of the body’s immune system to fight infections, such as interferons and interleukins, are now synthesized in the laboratory. They can be given to HIV-infected individuals to help their immune system overcome HIV infection. Another form of immunomodulation that shows promise in augmenting the body’s immune defenses against HIV involves the use of “therapeutic vaccines.” The goal of vaccine therapy in already infected individuals is to induce the immune system to better recognize and more effectively fight HIV than it does in response to natural HIV infection. Several therapeutic vaccine trials have shown that such vaccines are safe and immunogenic (produce an immune response), but little is currently known about the efficacy of HIV therapeutic vaccines as a primary treatment (Lu, Arraes, Ferreira, & Andrieu, 2004).

### ***Secondary Treatment***

Secondary treatment is directed not against HIV itself, but against the specific infections and tumors that are indirectly caused by the HIV infection. Secondary therapies are aimed at curing or controlling conditions like PCP, cryptococcal meningitis, cytomegaloviral retinitis, and tumors associated with AIDS such as Kaposi’s sarcoma and non-Hodgkin’s lymphoma. Individuals who are already receiving antiretroviral treatment nearly always receive secondary treatment.

Many specific preventive therapies exist for several AIDS-defining opportunistic infections. These strategies are designed to prevent a clinical occurrence or recurrence of specific infections. For example, a medication can be given to a person who is at risk of developing PCP or to someone who has already had an episode of the pneumonia to prevent its recurrence.

Steady improvement in secondary treatment modalities over the past 15 years has decreased mortality associated with AIDS-related secondary infections. The introduction of newer antiretroviral medications, such as protease inhibitors, has had a significant positive impact by reducing the occurrence of opportunistic infections and the need for life-long prophylaxis against disease reoccurrence. Rates of most major opportunistic infections have declined significantly since the introduction of protease inhibitors and it is expected that opportunistic infections in individuals infected with HIV will decrease in patients who respond favorably to anti-retroviral therapy (Solway, 1998). This has led to an interest in stopping primary prophylactic medications against specific opportunistic infections, e.g., PCP pneumonia, when a patient’s immune function improves on HAART. Data suggests that when the risk of PCP becomes small enough with successful antiretroviral treatment, primary prophylaxis may safely be stopped (Masur & Kaplan, 1999).

## **Functional Limitations**

### ***Physical***

HIV infection and AIDS cause disturbances in the normal functioning of many systems of the body. These disturbances often progress to both physical and mental functional impairments. Chief among the body's systems that can be physically impaired are the respiratory, cardiac, gastrointestinal, musculoskeletal, neurological, sensory, and metabolic systems.

#### ***Respiratory Disorders***

Energy-restricting disorders are the most common functional impairments resulting from HIV infection. The primary manifestation is fatigue, especially in individuals who have a low CD4+ T-lymphocyte cell count. As HIV-related immunodeficiency progresses, persons infected with HIV begin to experience decreased states of energy that restrict activities of daily living. Employed individuals with HIV experience frequent exhaustion, and often require midday rest periods. Persons with advanced HIV must rest after only a few hours of activity.

A major contributor to energy restriction in advanced HIV disease is the presence of respiratory infection. The single most common AIDS-defining disease is PCP which causes shortness of breath and fatigue during an acute episode and persists for weeks after recovery. Other potentially disabling respiratory conditions include respiratory tuberculosis, bacterial pneumonias, cytomegaloviral (CMV) pneumonia, and respiratory Kaposi's sarcoma. Since the introduction of protease inhibitors, energy levels in individuals infected with HIV have been increased.

#### ***Cardiac Disorders***

HIV can affect the heart, causing an inflammation of the heart muscle itself (myocarditis) or the thin lining around the heart (pericarditis). Both conditions restrict energy by causing shortness of breath, chest pain, and fatigue.

#### ***Gastrointestinal Disorders***

Individuals infected with HIV often have functional impairments caused by gastrointestinal disorders. Chief among these are infectious and non-infectious diarrheal syndromes that cause prolonged bouts of profuse diarrhea, impairing a person's ability to absorb vital nutrients from food. This malabsorption causes progressive weight loss, profound fatigue, and an aversion to food out of fear of triggering diarrhea. In addition, painful oral lesions are common in HIV infection and include recurrent and painful aphthous ulcers ("canker sores"), Kaposi's sarcoma, and infectious diseases like candidiasis, herpes simplex, and hairy leukoplakia, which may impair the ability to chew.

#### ***Liver Disease***

Hepatitis C virus (HCV) and hepatitis B virus (HBV) infections are common in HIV-infected individuals because of shared routes of viral transmission. Liver disease due to chronic HBV and HCV infection is becoming a leading cause of death among persons with HIV infection worldwide; risk of death related to liver disease is inversely related to the CD4 cell count (Koziel & Peters, 2007). An increase in the incidence of hepatocellular carcinoma and hepatotoxic effects associated with antiretroviral drugs in patients with HCV and HBV coinfection has been observed (Weber et al., 2006).

#### ***Musculoskeletal Disorders***

In addition to internal organs, HIV affects joint, muscle, and bone tissues. Prevalence of musculoskeletal disorders in individuals affected with HIV is high (up to 50%) and increases with disease progression. Arthralgias (painful joints) and frank arthritis (inflammation of the joints) are the most common forms of musculoskeletal disorders. The following three conditions involve the muscles: (a) AZT myopathy (found in patients on AZT and manifested by proximal muscle weakness, myalgias, and muscle wasting); (b) HIV-related polymyositis (muscle weakness in patients not on AZT); and (c) infectious myositis (tenderness and swelling of a muscle caused by staphylococcal bacteria) (Rodriguez, 1998).

Management of musculoskeletal pain can include the use of analgesic medications (some of which produce drowsiness and impaired judgment). The physician may prescribe physiotherapy, hypnosis, acupressure, or acupuncture. Sometimes musculoskeletal pain becomes chronic, further restricting activities. Musculoskeletal disorders associated with HIV can partially or totally impair ambulation because of pain or muscle weakness in one or both legs.

### ***Neurological disorders***

Neurological dysfunction occurs frequently from HIV infection and AIDS. Nearly 20% of patients with AIDS experience some neurological dysfunction. Often, neurological symptoms occur before other manifestations of HIV disease. Prior to the era of intensive anti-retroviral therapy, approximately 60% of persons who present with AIDS displayed neurological symptoms (Snider et al., 1983). Neurological disorders related to HIV are divided into two major types: (a) those that affect the central nervous system (brain and spinal cord), and (b) those affecting the peripheral nervous system (nerves to the arms and legs). Several neurological disorders affecting the peripheral nervous system are additionally seen in individuals infected with HIV. These include painful sensory neuropathies of the arms and legs that result in pain and weakness, both of which restrict an individual's activities.

Central nervous system impairment causes limitations ranging from serious (such as recurrent seizures due to an opportunistic infection or a tumor) to mild (such as mild dementia or mental slowness due to an HIV-related cognitive impairment). Opportunistic infections and tumors, which can affect the central nervous system, include toxoplasmosis (a protozoan infection causing seizures), cryptococcal meningitis (a fungal infection causing inflammation of the lining of the brain), and various types of brain tumors, such as lymphoma.

Sub-acute encephalitis (AIDS encephalopathy or "AIDS dementia complex") is the most insidious neurological problem and is primarily seen in advanced forms of the disease. Symptoms such as poor memory, inability to concentrate, verbal and motor slowing, affective and behavioral changes, and social apathy are the chief manifestations of AIDS dementia (CDC, 2006a). Individuals with AIDS dementia complex manifest a myriad of cognitive impairments, such as short-term memory deficits and a decrease in concentration ability. In addition, affective and behavioral disorders occur that range from social withdrawal and apathy to impatience, irritability, mania, and even psychosis. Any of these dementia-associated mental impairments can progress, like their physical counterparts, to a stage where the individual is not capable of living independently.

### ***Sensory Disorders***

Sensory organs are affected by HIV. The primary example of this type of impairment is visual loss due to infection of the retina either by HIV itself or by cytomegalovirus (CMV). CMV retinitis is the most common type of visual impairment, usually only seen in individuals with severe degrees of immunodeficiency. A person who loses any degree of vision faces physical challenges, such as loss of reading and writing, mobility limitations, and other daily living limitations. There are a variety of emotional and social problems that occur with visual loss.

### ***Metabolic Disorders***

A number of metabolic complications occur, such as hypothyroidism (deficiency of thyroid hormone), hypogonadism (deficiency of sex hormones), and adrenal hormone deficiencies. The most debilitating metabolic complication is called "HIV wasting syndrome" and its severity correlates with the level of immunodeficiency. The HIV wasting syndrome is manifested by weight loss (in excess of 10% from baseline), weakness, chronic diarrhea, and fever.

Wasting is thought to be caused by a combination of factors including: (a) the body's on-going immunologic response to HIV, (b) the number and severity of opportunistic infections that an individual has, (c) inadequate nutrition secondary to poor oral intake or malabsorption of nutrients due to diarrheal disorders, and (d) androgen deficiency. Successful treatment depends on anti-retroviral therapy, treatment of secondary infections and tumors, adequate nutrition, and measures to correct underlying androgen deficiency (Reiter, 1996).



AIDS therapies employing protease inhibitors have been associated with a range of metabolic disorders, such as diabetes mellitus and changes in fat metabolism. Disorders of fat metabolism include subcutaneous fat wasting (lipoatrophy) and abdominal obesity (lipomegaly), but the mechanisms by which antiretrovirals adversely affect fat metabolism are poorly understood (Lenhard et al., 2000). Loss of fat stored in the cheeks can be perceived by patients as “disfigurements” that may be socially limiting.

### ***Emotional***

The psychological reaction to impairments caused by HIV depends chiefly on: (a) severity and rate of progression of the particular impairment, (b) underlying personality of the affected individual, including self-image and coping style, (c) any neuropsychiatric changes caused by HIV itself, and (d) the reaction of those around the patient to the condition. The extent of the patient’s social support network and quality of psychological care available are crucial factors. Emotional disturbances are common, the most common of which are depression and anxiety. (Basu, Chwastiak, & Bruce, 2005).

Because HIV infection is a progressive condition, those affected by HIV become anxious about the future. Each measurement of their immune system’s function that reveals further deterioration result in increased feelings of anxiety, helplessness, and depression. Professional counseling is essential.

Psychological problems increase when a previously stable person infected with HIV develops an AIDS-defining disease. For young adults with AIDS, the losses associated with advancing disease are psychologically devastating. Loss of health, earning power, enjoyable activities, and sex life, coupled with having to experience premature old age and disability produce profound depression and suicidal ideation. The primary psychological effects of HIV infection and AIDS intensify a person’s physical impairments. Disturbances of sleep, appetite, and weight are only a few of the physical effects that can be a direct result of the emotional manifestations associated with HIV disease. Undiagnosed and untreated mental illnesses already present in an individual is compounded by the knowledge of having to face a life with HIV (Basu et al., 2005).

### ***Social***

Medical and rehabilitation professionals are aware of the influence that social factors exert on the course of illness. A person’s ethnic and cultural background, marital status, family support system, educational level, financial capability, and vocational background have an effect on the provision of rehabilitation services. The more problematic social factors are, the more likely recovery will be difficult. Understanding the social aspects that influence the daily life of a person with HIV/AIDS is important since HIV is transmitted mostly through sexual contact and shared needle behaviors, both of which are fundamentally social phenomena (Friedman, Kippax, Phaswana, Rossi, & Newman, 2006).

A great challenge facing rehabilitation for a person with HIV infection or disease is that of overcoming the social limitations associated with the names “HIV” and “AIDS.” A person with HIV infection or AIDS faces many social obstacles in the way of recovery. The HIV epidemic has elicited very strong responses from society as HIV transmission involves forms of behavior that are either illegal or socially taboo. The groups at greatest risk for becoming infected with HIV and developing AIDS, gay males and intravenous drug users, have traditionally been the subject of legal sanctions and social stigma. Further, AIDS affects a disproportionate number of African-Americans and Latinos, racial and ethnic groups already the subject of various social limitations and prejudices.

Because of the unique epidemiologic nature of HIV, individuals that are infected are frequently ostracized by family members and abandoned by society. They face discrimination in employment, housing, social services, and health care. Many persons with AIDS fear social interaction with family, friends, and society. Physical and social limitations combine to hinder meaningful social interactions. A sense of isolation, in some cases, can become so severe that they commit suicide. The situation is not entirely bleak, however. Many agencies, staffed largely by volunteers, have responded to the social isolation that persons experience by setting up organizations to meet the patient’s needs for companionship, shopping assistance, meal preparation, and other activities of daily living.



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## Vocational Limitations

Vocational rehabilitation services for persons with HIV infection and AIDS makes an individualized approach to vocational rehabilitation crucial. Although no single method of evaluation can be applied universally, three general issues can be stressed:

1. Is the individual able to return to his or her previous job or occupation?
2. Are the skills or abilities of the person transferable to a new job?
3. In conjunction with the medical and emotional factors, what type of rehabilitation training needs to be conducted to facilitate reemployment? (Matheson, 1984).

The age group most commonly affected by HIV infection is young adults, ranging in age from 20-40 years. Although they may later seek disability benefits, many people are employed at the time of diagnosis. While some individuals have well developed vocational skills, others are unemployed or employed on a part-time basis. After developing HIV, many persons cannot continue working at their prior levels of activity. They may have to quit their jobs because of functional impairments; some resign or are fired after the employer learns the nature of their condition. These persons are candidates for vocational assessment and rehabilitation.

During vocational evaluation, the physical, emotional, and social factors associated with HIV disease need to be kept in mind. Individuals have different levels of medical stability. Unlike impairments that reach a level of permanence before the process of vocational rehabilitation begins, impairments associated with HIV disease are progressively and inherently unstable. While some have conditions that are sufficiently stable and do not interfere with work, others have a disease state that makes occupations difficult or impossible.

A challenging factor in the vocational environment is the lack of physical endurance to work a full day. A rehabilitation counselor can assess physical tolerance for work by ascertaining daily energy patterns and energy requirements expended in the home. Assistance in home management activities may save energy that can be used for work.

Individuals can develop various cognitive impairments secondary to those which affect the central nervous system. Moderate to severe degrees of AIDS dementia complex manifested by memory deficits, lack of coordination, and poor concentration ability create difficulties in the work environment. Cognitive impairments may exist in any person infected with HIV and neuropsychological testing should be part of rehabilitation evaluation.

Emotional factors such as depression and anxiety interfere with a successful vocational rehabilitation outcome. Motivation to work is crucial. The level of emotional adjustment needs assessment by the rehabilitation counselor. Emotional adjustment to the disease must occur before a person with AIDS is vocationally placed.

The presence of skin lesions on the face or other exposed surfaces of the body from Kaposi's sarcoma are physical sources of embarrassment for patients which may impede rehabilitation. Changes in an individual's facial appearance due to fat redistribution secondary to antiretroviral medications can lead to reluctance to participate in the work environment. The application of cosmetics to cover such lesions may help a person overcome reluctance to enter the workforce.

Lastly, two social factors are particularly disabling for someone attempting to re-enter the vocational environment after being diagnosed with HIV. The most important factor relates to the incorrect belief that individuals infected with HIV pose a communicable disease risk in the workplace. The only possible risk of HIV transmission in the workplace is if an accident occurs resulting in blood from the individual infected with HIV coming into direct contact with the non-intact skin of a co-worker who is uninfected. Another social factor is the prejudicial treatment individuals with HIV infection face based on others' judgments or misconceptions about their sexual behavior or drug use. This prejudice may severely limit vocational rehabilitation.

## **Rehabilitation Potential**

Rehabilitation potential for persons affected by HIV depends, in large part on the level of their underlying immunodeficiency, the presence of any physical limitations arising from specific disorders, and complications from multi-drug treatments. Individuals with greater degrees of immunodeficiency, as measured by increases in their HIV viral load, or in reductions in their level of CD4+ T-lymphocytes, will have less residual capacity for rehabilitation. Severity of physical impairments frequently worsens with increased immunodeficiency. Many persons with HIV disease and severe immunodeficiency, however, manifest significant capabilities for rehabilitation. The development of effective antiretroviral therapies in the mid-1990s has greatly enhanced rehabilitation potential.

Most persons diagnosed with HIV disease are young. Since HIV primarily affects previously healthy young adults, these individuals maintain fairly high rehabilitation potential. A minority are in the category of limited rehabilitation potential, usually those who remain untreated or who have had the condition for many years. The vast majority of persons have sufficient physical, mental, emotional, and educational capabilities to respond favorably to vocational rehabilitation.

An additional positive factor contributing to rehabilitation potential is the absence of severely disabling neuromuscular impairments such as spinal cord damage, loss of appendages, and reduction in arm or leg mobility. Most ambulatory individuals with HIV have adequate residual neuromuscular capacity to engage in sedentary or higher exertional levels of work, and those with energy-restricting disorders usually maintain sufficient capacity to engage in light to medium work levels.

The rehabilitation counselor needs to conduct a comprehensive assessment of a person's strengths, capacities, and skills. As medical science improves the quality and length of life for those with HIV, rehabilitation professionals will be called upon more frequently to help restore the individual's ability to live and work successfully and to the fullest extent.

## **Case Study**

Mr. James Dexter is 29 years of age and works in an oil refinery. Mr. Dexter did not finish high school, completing most of the 12th grade. His job title of equipment mechanic (petroleum production), *Dictionary of Occupational Titles (D.O.T.)* # 629.381-014, puts him in charge of the installation, maintenance, and repair of oil well drilling machinery and equipment. He uses hand tools and power tools, and reads diagrams and schematics to repair equipment such as pumps, transmissions, and diesel engines. Occasionally, an equipment mechanic such as James also performs welding and soldering. All oil refinery mechanics run tests to insure that the equipment is fully functional. One to two years are required for proficiency in this field, which requires occasional lifting of 50 pounds and repetitive lifting and carrying of 25 pounds. Mr. Dexter worked as an oil-field equipment mechanic for six years.

Before this job, Mr. Dexter worked in the construction field as a construction Worker II (construction), *D.O.T.*# 869.687-026, a position which did not involve any skills but required lifting up to 120 pounds occasionally and 50 to 75 pounds repetitively. The job responsibilities included loading and unloading of building materials, tools, and supplies. Also involved were digging, spreading, and leveling dirt and gravel using a pick and shovel. Mr. Dexter held this job for four years.

In 2002, Mr. Dexter developed a painful tingling sensation on the left side of his chest, followed the next day by water blisters in the same area. He was diagnosed with shingles (herpes zoster). His physician advised him that herpes zoster was a sign of an impaired immune system. He thought no more about it; after four weeks, the shingles disappeared.

In late 2004, James began to develop feelings of fatigue at work, especially when climbing the "cracking" towers at the oil refinery. Gradually, his fatigue worsened and his work performance began to decline. James never felt rested. His boss told him to "shape up" or he would be fired. In January of 2005, his fatigue became severe and he lost 11 pounds. When he developed shortness of breath, a dry cough, and a fever of 102 degrees Fahrenheit, Mr. Dexter saw a physician, who diagnosed him with pneumonia. He was admitted to the hospital.

A thorough medical and social history revealed that, except for a broken leg in 1996, Mr. Dexter had been in good health all his life. James has been married for several years and has three young children but he and his wife have had some marital difficulties. During a period of about two years, when he and his wife were separated, Mr. Dexter had sexual contacts with other women.

Mr. Dexter's pneumonia turned out to be caused by *Pneumocystis carinii*. His doctors became suspicious that he was infected with HIV and ordered an HIV antibody test which returned positive. He was found to be severely immunodeficient with a CD4+ T-lymphocyte count of 127 (CDC CD4+ T-Lymphocyte Category 3) and an HIV viral load of 250,000 copies (normal is "undetectable"). His wife tested negative for the HIV antibody.

During his hospitalization, James experienced intermittent diarrhea with no diagnosed cause. He lost about 30 pounds and developed CMV retinitis in the left eye which left him with a 50% visual loss in that eye, even after treatment with medication; he was hospitalized for a total of six weeks. Upon discharge, James was sent home on three antiretroviral medications: AZT (Retrovir), Delavirdine (Rescriptor), and Saquinavir (SQV). After a tumultuous three weeks, his wife moved out with the children and filed for divorce. After his wife and children left, James considered suicide. His physician recommended emotional counseling, but Mr. Dexter turned it down.

After six months of convalescence, James began to feel better. His CD4+ T-lymphocyte count rose from 127 to 487, and his HIV viral load was undetectable. James decided to return to work. Occasionally, when at home he takes a brief nap during the day. He has been experiencing pain in the muscles of his upper legs, which his physician thinks may be a myopathy due to AZT. Despite his visual loss, occasional fatigue, and sore legs, James believes he can work an eight-hour shift. The physician has restricted him to a maximum of light work activity.

Mr. Dexter wants to return to his previous job because he feels confident and sufficiently physically fit to do the work. Concerned about his ability to support himself and his family, he desires to return to work as soon as possible. Recently, the oil company terminated his employment.

### Questions

1. What is the nature of James Dexter's HIV-related disease and what are the possible physical limitations associated with his medical condition that may influence his rehabilitation potential?
2. What emotional limitations does Mr. Dexter have that need to be addressed during the process of rehabilitation?
3. Discuss the social limitations of James' medical condition.
4. What is this client's rehabilitation potential? Discuss the possibility of him returning to his previous job.
5. Provide a vocational profile including age category, educational level, exertional level of previous work, occupationally significant characteristics, and work skills. Note if any of Mr. Dexter's skills are transferable and, if so, to what jobs.
6. Identify types of rehabilitation training that will facilitate his reemployment.

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The Internet hosts thousands of Web pages devoted to HIV infection and AIDS. Typically, these pages are more current than other sources of AIDS-related information. The United Nations' HIV/AIDS web site at [www.unaids.org/en](http://www.unaids.org/en) provides a current global perspective on HIV/AIDS. The U. S. Centers for Disease Control and Prevention offer the latest statistics about HIV/AIDS in the U. S. at <http://www.cdc.gov/hiv/topics/surveillance/basic.htm>. Current information about AIDS clinical drug trials is available at [www.hivatis.org](http://www.hivatis.org) or <http://www.amfar.org/cgi-bin/iowa/index.html>. The state of current vaccine research can be located at [www.iavi.org](http://www.iavi.org).

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## Chapter 11

# HEARING LOSS, DEAFNESS, AND RELATED VESTIBULAR DISORDERS

*Shawn P. Saladin, Ph.D., C.R.C.*  
*Sandra Hansmann, Ph.D.*

## Introduction

The ability to hear is a critical sensory function, yet hearing loss is the most common sensory disability. Although it can be challenging to accurately quantify the number of individuals who have hearing loss and other auditory disorders, one trend is clear—hearing loss among adults and children is increasing exponentially (National Research Council, 2005). Estimates suggest that by the year 2015, the number of people with hearing loss in the United States will reach nearly 13 million. The inability to hear has far reaching social and health related consequences and negatively impacts speech, language, academic, social, and vocational development.

This chapter reviews selected medical, psychosocial, and vocational aspects of major disorders of the auditory and vestibular systems that impact hearing and communication, specifically focusing on Deafness, hearing loss, and disorders associated with such hearing loss. A brief overview of the anatomy of the ear is provided, followed by a discussion of basic hearing evaluation and classification techniques. Next, common disorders of the auditory system are discussed, followed by a description of several related conditions of the vestibular system. Treatment, management, and aural rehabilitation options are reviewed. The typical functional limitations and related psychosocial and vocation implications of hearing loss and the rehabilitation potential of individuals who are Deaf or hard of hearing are explored.

## Anatomy of the Ear

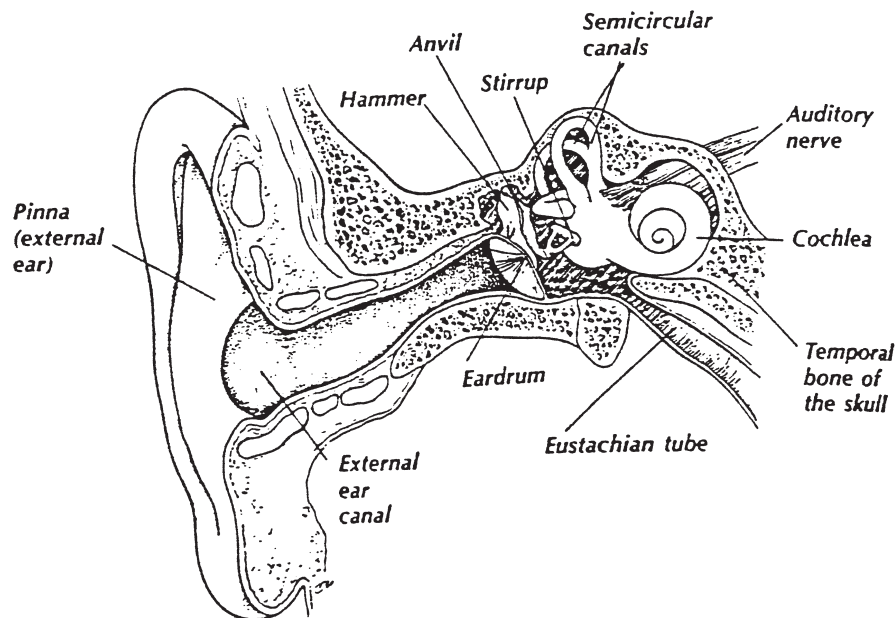
To understand hearing disorders, it is helpful to have a basic understanding of anatomy of the human ear and process through which sound waves are perceived as sound by the brain. The human ear includes two distinct but interrelated systems that provide important sensory functions — the auditory system (pertaining to the detection of sound) and the vestibular system (pertaining to balance, proprioception, and the detection of movement and acceleration).

### *The Auditory System*

The auditory system of the ear consists of three principle sections including the outer ear, middle ear, and inner ear (as shown in Figure 1). Sounds may consequently be conducted through the air into the outer, middle, and inner ear structures, or through the bones around the ear. Dysfunction in any one of the primary areas of the ear can result in difficulty hearing or the inability to hear.

**The outer ear** consists of the *pinna*, the externally visible part of the ear which is also called the *auricle*, and the ear canal up to the *tympanic membrane*, also known as the *eardrum*. Sound energy is collected by the pinna and then directed into the outer ear canal and onward to the tympanic membrane. The elastic cartilage of the pinna has an intricate funnel-like shape that facilitates the effective collection and transmission of sound energy into and through the ear canal (Koga, 2004). **Audition**, or hearing, begins when sound waves of various frequencies are created by vibrations in the environment. These vibrations travel through the atmosphere and reach the pinna.

Figure 1  
Ear



Source: Schwartz, S. (1990), *Choice in deafness – A parents guide to communications options* (2<sup>nd</sup> ed.). Bethesda, MD: Woodbine House. Reprinted with permission.

**The middle ear** functions to transfer and amplify vibratory energy deeper into the inner ear, where it can be perceived as sound. It is an air-containing space that includes the inside of the tympanic membrane and the **ossicular chain**, made up of three very small bones known collectively as **ossicles** and individually as the **malleus**, **incus**, and **stapes**. The ossicles are often referred to as the hammer, anvil, and stirrup. The tympanic membrane seals the middle ear. In hearing, sound waves vibrate the tympanic membrane which then presses against and vibrates the ossicles. The incus lies between the malleus (attached to the middle ear) and the stapes, articulating the two. The middle ear also connects to the back of the nose and throat through the **eustachian tube**, which functions to maintain equal pressure between the middle ear and the external environment.

**The inner ear** is a complex fluid-filled structure that converts vibratory sound energy into nerve impulses perceivable by the brain. The inner ear contains a membranous structure, the **labyrinth**, which is surrounded by bone and filled with perilymphatic fluid. The inner ear is involved in both auditory and vestibular functions, and its major structures include the **vestibule** and **semicircular canals**, and the **cochlea**. The stapes footplate in the middle ear is attached to the base of the **cochlea**, a critical structure of the inner ear. The cochlea is a small, spiral structure that includes the **organ of Corti** to convert sound from mechanical vibrations into electrical signals through a process known as **transduction**. Transduction is performed by specialized sensory cells inside the organ of Corti—the inner and outer **hair cells** which include **stereocilia**, cellular projections that look like fine hairs. The movement of the stapes vibrates the stereocilia above and is connected to the auditory nerve root. In addition, the inner ear is of little use without an intact and functional **cranial nerve VIII** or **auditory nerve** to pass signals to the brain stem and brain. The brain plays a key role in the identification, localization, and understanding of sound (Martin & Clark, 2006). The vestibule contains the **utricle** and **sacculle**, which help to facilitate balance.

## Hearing Assessment

Several considerations are relevant to assessment and categorization of hearing loss, including degree of severity, configuration, onset, and progression (Tye-Murry, 2004). Accordingly, the assessment

of hearing ability may encompass a variety of subjective, objective, and alternative methodologies. Severity is quantified through tests of hearing ability, which have become increasingly accurate and standardized through use of commercial audiometers, devices for measuring pure tone air and bone conduction of sound (Wilber, 1999). While a number of methods are available, pure tone audiometry, speech audiometry, and tympanometry are commonly encountered measures in rehabilitation and related settings.

### ***Pure Tone Audiometry***

***Pure-tone audiometry*** is an established standardized method for evaluating hearing loss (American National Standards Institute, 1996, 1997, 2003). Audiometers are used to assess hearing and are measured in ***decibels*** and ***hertz***. Decibels (dB) measure the intensity or loudness of a sound, while the tone or pitch of a sound is expressed as hertz (Hz). Using pure-tone audiometry, an ***audiologist*** uses an ***audiometer*** to generate and measure the softest pure tone sound a person can hear to establish an ***auditory sensitivity threshold*** (National Research Council, 2005). The American National Standards Institute defines threshold as “the lowest input level at which responses occur in at least 50% of a series of ascending trials” (American National Standards Institute, 1996, p. 10).

During a test, the individual responds to high and low pure tones delivered over a wide range of frequencies. The audiologist repeats each sound at increasing lower volume until no sound can be heard. This type of testing, termed ***air conduction testing***, relies on the transfer of sound waves to the inner ear through air. ***Bone conduction audiometry*** is an alternative procedure that uses a sound vibrator fitted to the bone behind each ear. Bone conduction testing covers a wide range of high and low sound frequencies repeated at decreasing volume (National Research Council, 2005; Plante & Beeson, 1999).

### ***Speech Audiometry and Tympanometry***

***Speech audiometry*** is conceptually similar to pure tone audiometry. It is used to determine the softest speech sounds a person can hear in each ear at approximately 50% accuracy, known as the speech awareness threshold (SAT) or speech reception threshold (SRT). Audiometric testing results are documented on an ***audiogram*** to provide a visual representation of severity of hearing loss in each ear. An important aspect of both audiograms and tympanograms is the information they provide about the configuration of an individual’s hearing loss. ***Tympanometry*** is a measure of the stiffness of the eardrum and is used to evaluate middle ear function. This test is helpful in detecting fluid in the middle ear, negative middle ear pressure, tympanic membrane perforations, and disruption of the ossicles.

### ***Configuration of Hearing Loss***

Hearing ***configuration*** refers to the specific location, consistency, and quality of an individual’s hearing difficulties. Determining the configuration of a person’s hearing loss provides a more holistic view of hearing ability. Initially, a person may be described as having hearing loss in either one ear, termed a ***unilateral hearing loss***, or as having a ***bilateral hearing loss***, when both ears are affected. If the degree or severity of hearing loss is the same in both ears, the person has a ***symmetrical*** loss; if the degree differs in each ear, the loss is ***asymmetrical*** (Nicolosi, Harryman, & Kresheck, 1996). Configuration also includes the amount of hearing loss present at low, mid, and high frequencies (Tye-Murray, 2004). For example, a person who has difficulty hearing low frequencies, but no difficulties at higher frequency levels has a low-frequency hearing loss.

## **Definitions of Hearing Loss and Deafness**

As with many health conditions, hearing loss severity may range from a very mild loss to profound Deafness. Before the field of audiology was developed, differences along the hearing loss continuum were imprecisely defined. Even today, precise definitions of deafness and hearing loss vary. Not all definitions of hearing loss rely exclusively on numeric data, using instead functional self-reports or observations of hearing ability. As a result, both medical and cultural definitions related to hearing loss exist.

### **Medical Definitions**

Most medical definitions of hearing loss rely on the average of audiometric test results at three specific frequencies, most often at 500, 1000, and 2000 Hz. Known as the pure tone average (PTA), this number is a basic indicator of how much hearing a person has in each ear independently without any type of amplification device (Hull, 2001). In general, six levels of hearing loss are calculated. Normal hearing ability is typified by sensitivity thresholds of 0 dB to 25 dB. *Mild* hearing loss is usually diagnosed when PTA results demonstrate a 26 dB to 40 dB threshold, while *mild-to-moderate* hearing loss is diagnosed in individuals who have a sensitivity threshold of 41 dB to 55 dB. An individual with a 56 dB to 70 dB threshold has a *moderate* hearing loss; with threshold ranges from 71 to 90 dB, a person has a *severe* hearing loss. Persons with a 90 decibel threshold or greater have a *profound* hearing loss (Tye-Murry, 2004). Diagnostic weight is given to the results of the better ear. People with hearing in the mild to moderate categories are considered “*hard of hearing*” while those with hearing loss assessed in the severe and profound categories are considered *Deaf* (Flexer, 1999).

### **Cultural Definitions**

Audiometric and functional definitions focus on severity and functional impact of hearing loss. Cultural definitions of Deafness, however, are grounded in personal choices about self-identification and language use. Thus, an individual may be identified as culturally Deaf, although he or she may only have a mild or moderate hearing loss from an audiological standpoint. The converse may also be true; a person may have an audiological profound hearing loss but not be identified with Deaf culture (Padden & Humphries, 2005). The term *Deaf* is capitalized when referring to people with hearing loss who identify themselves as part of the Deaf cultural group (Lane, 1992; Lane, Hoffmeister, & Bahan, 1996).

The cultural definition of Deafness represents a life and world view that is manifested by beliefs, values, and traditions unique to Deaf people and expressed through the use of American Sign Language (ASL). Within Deaf culture, the term *hearing impaired* is rejected because it reflects an illness-oriented medical paradigm that pathologizes Deaf people and negates the cultural component of Deafness. Deaf culture makes no distinction between person-first language and more Deaf-centric phrases such as *Deaf person*, finding either option acceptable (Lane et al., 1996). Throughout this chapter, the terminology and grammar preferred by the Deaf cultural community is used.

## **Onset and Progression of Hearing Loss**

Onset and progression are important dimensions of hearing loss classification. Onset encompasses both the rapidity of hearing loss, and the life stage at which onset occurs. Progression refers to the course of the hearing loss. If a person’s hearing loss occurs quickly and unexpectedly, it is considered a *sudden* hearing loss. If hearing status worsens slowly over time, an individual is said to have a *progressive* loss. A person may also have a *fluctuating* hearing loss, one that alternates in severity. When no such variability exists, the hearing loss is considered *stable* (Plante & Beeson, 1999).

Hearing loss occurs at any life stage. When a person is Deaf or hard of hearing from birth or in the first days of life, the person has a *congenital* hearing loss. This may include *pre-lingual* hearing loss which refers to hearing loss occurring prior to development of speech. Pre-lingual hearing loss occurs before the age of 2 or 3 years. If hearing problems develop later in childhood or adulthood, an individual has an *acquired* hearing loss that is *post-lingual*, because it has occurred after development of speech. Post-lingual hearing losses may be additionally classified as *prevocational*, occurring when an individual is school aged (Tye-Murray, 2004). *Late deafened* is a term used to describe people whose hearing loss occurs well into adulthood. Late deafened adults are the largest group within the total population of Deaf individuals in the United States (Vallaume et al., 1997).

## **Types of Hearing Loss**

Deafness and hearing loss occur due to a wide variety of genetic, obstructive, neural, age-related, and environmental factors, several of which are discussed later in this chapter. Regardless of exact etiology, hearing loss is described as *sensorineural*, *conductive*, or *mixed*. Hearing loss stemming from conditions of



the inner ear or auditory nerve is sensorineural. Most often, sensorineural hearing loss is due to damage to the pathway for sound impulses from the hair cells of the inner ear to the auditory nerve and the brain (Katz & White, 2001). Damage may occur as a result of noise exposure, viruses, head trauma, and other causes.

Conductive hearing loss most often originates in the outer and middle portions of the ear. Most conductive hearing problems result from some form of interference that results in the inability of sound waves to conduct from the pinna into the ear to vibrate the tympanic membrane and inner ear structures (Kiessling et al., 2003). Examples of conditions that may result in a conductive hearing loss include blockage by wax, a punctured eardrum, an ear infection, and congenital or acquired irregularities in the outer and middle ear. A mixed hearing loss simply results from a combination of both conductive and sensorineural losses.

## Specific Etiologies

The population of people with hearing loss and other auditory disorders is very diverse. Two accepted etiological categories of hearing loss are *acquired* and *genetic*. Acquired hearing loss is sometimes labeled *non-genetic hearing loss* and may be sensorineural, conductive, or mixed. Acquired hearing loss is related to a number of causes such as chronic infections, acoustical injury, occupation hearing loss, and aging. Other causes may include head injuries, inner ear disorders, and exposure to toxins (Robertson & Morton, 1999). Genetic hearing loss is caused by differences or changes in genes, may be inherited or occur as the result of an isolated gene mutation, and may be conductive, sensorineural, or mixed (Nance, 2003). This chapter section reviews several of the most common causes of acquired hearing loss, followed by an overview of key genetically linked causes of hearing loss.

### *Chronic Ear Infection*

Infections of the middle ear and middle ear structures are called *otitis media (OM)*. Causes of the condition include bacterial infection, injury to the eardrum, and presence of fluid in the ear. Symptoms include a sensation of pressure in the ear, dizziness, and discharge. Ear pain may be present and range from mild to severe (Anteunis, Engel, Hendriks, & Manni, 1999). Chronically occurring OM is defined as a perforation of the tympanic membrane that is permanent and occurs with or without permanent changes to the middle ear.

### *Injury*

Injuries to either the external or internal structures of the ear can result in significant hearing-related problems. The external ear may be damaged in accidents. If the pinna is no longer able to adequately gather sound energy, hearing ability can be significantly impacted. Internally, the tympanic membrane may be punctured by penetrating objects such as cotton swabs, pencils, or other small sharp objects. Sudden pressure changes, including pressure changes associated with explosions can rupture the eardrum, such as in swimming and diving accidents. When the tympanic membrane is punctured, dislocation of the ossicular chain can occur, and the stapes footplate may fracture. Sudden hearing loss usually accompanies perforation of the eardrum, along with dizziness and ringing sensations in the ear. Most perforations close completely without medical intervention (National Research Council, 2005).

*Noise-induced hearing loss* (noise exposure seriously injuring the inner ear) (NIHL) is a common yet preventable health concern (Koga, 2004). Occupational, recreational, and accidental causes of noise-related hearing loss may include sound exposure through either continuous or impulse noise exposure. Continuous exposure occurs when an individual is frequently in environments that include noisy on-going sounds such as industrial machinery or loud music. It is often associated with employment settings in adults and with recreational settings in adolescents (Scherich, 1996). Impulse exposure occurs when a person is exposed to sharp loud bursts of sound, such as gunshots or explosive blasts (Tambs, Hoffman, Borchgrevink, Holmen, & Engdahl, 2006). While NIHL has been closely linked to the volume, duration, and type of noise exposure, it is often due to a more complex interaction of factors, including age, psychological well-being, and lifestyle.

*Otosclerosis* is a common cause of hearing loss and can develop in late adolescence and early adulthood; it may develop rapidly during pregnancy. Primarily, otosclerosis results in a conductive hearing

loss, but sensorineural loss may occur (Plante & Beeson, 1999). As previously discussed, normal hearing results when sound enters the ear, vibrates the eardrum, and in turn, the malleus, incus, and stapes. Movement of the stapes vibrates fluids within the ear to stimulate the auditory nerve for hearing. This process is impeded by *otosclerosis*, involving hardening or fixation of the stapes.

*Presbycusis* refers to age-related hearing loss. It is actually a multidimensional disorder that spans not only hearing loss but also decreased auditory sensitivity. Presbycusis can result in discrimination and cause social, psychological, and communication problems (Hull, 2001).

As humans age, both the external and internal structures of the ear undergo changes. During the aging process, the external ear, or pinna, may become less firm and elastic, while skin may thin and wrinkle, and hair growth inside the ear may increase. These changes act simultaneously to impact the effectiveness of the ear as a sound collection mechanism. Middle ear structures, including the tympanic membrane and ossicular chain, become more rigid with age, thus decreasing vibratory response, while the hair cells and cochlea of the inner ear may also deteriorate (Hull, 2001; Villaume et al., 1997).

*Ototoxicity* refers to the negative effects some medications, solvents, and other substances have on the nerves and organs of the ear. Literally meaning “ear poisoning,” ototoxicity can result in Deafness. A number of industrial chemicals are classified as ototoxic, including mercury, styrene, and carbon disulfide. Most individuals do not come into contact with these substances unless they work in occupations where they are in direct contact with industrial chemicals. More typically, people are exposed through medications (National Research Council, 2005). A number of non-prescription medications have ototoxic effects, but those derived from salicylic acid are most common. Aspirin is a popular salicylate; an estimated 50 billion tons are consumed annually worldwide (Prepageran & Rutka, 2004). In most instances, the ototoxic effects of salicylates are the result of long-term, high dose use. Usually, these effects are temporary, while in rare circumstances permanent hearing loss and other complications result, including balance dysfunction.

Prescription medications used in treatment for cancer, severe infection, and following organ transplantation can have ototoxic properties, especially with prolonged use or at high dosages. Chemotherapeutic and immunosuppressant medications can damage the hair cells of the inner ear, resulting in sudden deafness or permanent high frequency hearing loss and tinnitus (Gratton & Smyth, 2004). Some classes of antibiotics, especially when administered intravenously, may result in profound, irreversible sensorineural hearing loss

### ***Genetic Hearing Loss***

Genetic hearing loss is the result of mutations in an individual’s genes. One in every 1,000 in the United States is born with severe hearing loss or Deafness, and about one-half of these impairments are classified as genetic (Luxon et al., 2003). Hereditary genetic hearing losses are generally classified by the pattern of inheritance. Approximately 77% of individuals who have severe to profound congenital hearing loss inherited it through an *autosomal recessive* pattern. This occurs when both parents carry a recessive gene for hearing loss and the trait is inherited by the child (Smith & Taggart, 2004). In *autosomal dominant* hearing loss, which accounts for about 22% of all genetic hearing loss, at least one parent carries a dominant gene for hearing loss.

*Nonsyndromic Deafness* (NSD) is associated with abnormalities in the auditory system; about 75% of genetically-linked Deafness is in this category (Nance, 2003). Although a variety of genes have been identified as related to genetic deafness, about half of all NSD are related to changes in the gap junction beta 2 (GJB2) gene, which is involved in the production of proteins critical to cochlear functioning (Steel, 1999).

*Syndromic hearing loss* or Deafness includes both a hearing loss and involvement of one or more other body systems, resulting in mental and physical differences. Most syndromes associated with Deafness cause mild hearing difficulties, while other syndromes have more significant manifestations. Pendred, Branchio-Oto-Renal (BOR), and Usher syndromes are causes of Deafness that have additional serious complications (Nance, 2003).

*Pendred syndrome* is an inherited, autosomal recessive disorder accounting for 4% to 10% of all hereditary hearing loss, characterized by thyroid dysfunction and profound sensorineural hearing loss occurring prelingually (Luxon et al., 2003). *Branchio-Oto-Renal (BOR) Syndrome* is hereditary as an

autosomal dominant trait related to a variety of genes. The manifestations of BOR syndrome differ according to the specific genes involved. Hearing loss often results from malformations of both the external and internal structures of the ear. BOR syndrome is closely associated with kidney and ocular abnormalities. ***Usher Syndrome***, an autosomal recessive disorder, is a leading cause of Deaf-blindness. Usher Syndrome is a group of disorders characterized by moderate to profound sensorineural hearing loss, vestibular dysfunctions, and vision loss due to retinitis pigmentosa (RP).

### ***Vestibular Disorders***

The vestibular system of the inner ear is responsible for orientation of sound location and balance. The system works in conjunction with visual images and head movement to send information to the brain regarding the physical orientation of the individual. Disturbances to this system, even when minor, may result in a variety of nonspecific sensations. ***Tinnitus*** is a condition characterized by the perception of sounds that originate in the head or ears rather than the environment itself (Sindhusake, Newall, Golding, Rochtchina, & Rubin, 2003). More severe disruptions of the vestibular system may result in ***vertigo*** — the sensation of movement, usually rotational, when no actual movement is occurring. ***Ménière's disease*** is a complex disorder that combines symptoms of tinnitus, vertigo, and progressive hearing loss.

#### ***Tinnitus***

Tinnitus is one of the most prevalent disorders of the vestibular system. It is estimated that 36 to 50 million people in the United States have experienced some form of this disorder. Tinnitus is caused by damage to the nerve endings in the inner ear and may be symptomatic of other problems of the ear, including elevated blood pressure, diabetes, thyroid problems, injuries, and infections (Sindhusake et al., 2003). Symptoms of tinnitus include ringing, buzzing, or whining sounds originating in the head or ears. The sounds may be intermittent or continuous; intensity may vary from a dull rumble to a high whine or buzz. Most people do not report being overly disturbed or distressed by these symptoms, and may only be able to perceive the sounds in very quiet environments. Rarely, people experience severe symptoms that interfere with all aspects of life.

#### ***Vertigo***

Vertigo is both a vestibular dysfunction and a symptom of other similar dysfunctions, including labyrinthitis or inner ear inflammation, tumors, brain hemorrhages or stroke, migraines, and multiple sclerosis. Imagined sensations of vertigo may be categorized as either subjective or objective. Abnormal eye movements and vision changes, hearing loss, nausea, tinnitus, imbalance, and gait disturbances are typical of the disorder. Symptoms may last just minutes or may linger for hours, be either intermittent or continuous, and range from mild to severe (Ruka, 2004).

### ***Ménière's Disease***

***Ménière's disease*** is a disorder related to volume and pressure changes in the inner ear fluids. The early course of the condition can be mild and episodic, but over time progresses to a more severe unilateral hearing loss with worsening vertigo and tinnitus. Generally, a diagnosis is made following two or more clear episodes of spontaneous vertigo lasting a minimum of 20 minutes with at least one episode of a sensation of fullness in the affected ear (Pullen, 2006).

## **Treatment Strategies**

Once the underlying causes of auditory or vestibular disturbances are addressed, treatment and management of residuals may be considered. Treatment and management strategies for hearing loss, Deafness, and related vestibular disorders are dependent on the type, severity, and etiology of the condition. In this section, key treatment and management devices and strategies for hearing loss and vestibular disorders are reviewed.

Treatment and management of hearing loss is vital. The most common hearing management method involves use of assistive listening devices. Assistive listening devices are beneficial in the social and emotional areas, and include ***hearing aids*** and ***cochlear implants***.

## ***Hearing Aids***

Hearing aids are the most common listening device used to treat and manage hearing disorders. Simply stated, a hearing aid is a self-contained system used to amplify sound. They use a microphone to detect sounds, an amplifier to increase the level of sounds, and several filters to modify sound to the needs of the user and reduce background noise. Hearing aids are generally prescribed for people whose hearing thresholds are 25 dB or greater in the better ear, and are usually not sought until the person experiences noticeable difficulties in daily living (National Research Council, 2005). These difficulties become more pronounced at approximately a 40 dB threshold, or the point at which background noise markedly impacts hearing ability.

***Air conduction or bone conduction hearing aids*** are available in a wide variety of styles and shapes, use air or bone conduction to transmit sound, and are either analog or digital (Martin & Clark, 2006). Air conduction hearing aids use a receiver to amplify sound and direct it into the ear canal using vibratory sound waves moving through air. Bone conduction hearing aids are suitable for individuals who have conductive loss related to the middle ear, but have inner ear functioning.

***Analog hearing aids*** generate signals that are analogous, or similar, to the sound energy they receive and then amplify those signals for use (Martin & Clark, 2006). Modern hearing aids operate by receiving and modifying sound signals (***signal processing***). Analog aids amplify all environmental sounds similarly and are thus appropriate for a variety of types of hearing loss. Programmable analog hearing aids use a microchip to store different settings; the person changes the level of amplification needed in different listening environments.

***Digital hearing aids*** use technology that converts sound waves into binary code to represent the frequency, intensity, and patterns of the signal, a function termed ***digitized sound processing***. The processing determines if the signal is useful sound or unwanted noise with extreme speed, providing an undistorted signal and improved filtering of background sounds. Many digital hearing aids are not completely digital, but rather combine aspects of analog and digital technology (Martin & Clark, 2006). Current trends, however, are focused increasingly on completely digital devices.

**Hearing Aid Selection and Use.** The primary goal of hearing aid use is to achieve the best auditory functioning possible. Binaural hearing aids often improve speech clarity and volume, improve perception of directional sound, and lessen the perception of background noise. Unilateral or ***monaural*** hearing aids are selected when hearing loss exists in only one ear.

Many health-related and psychosocial factors impact the selection of hearing aids, including the appearance, comfort, and financial considerations. Hearing aid models are tailored to different functional and aesthetic needs, and range from behind-the-ear devices to aids that fit completely inside the ear canal. Analog devices are the least costly type of hearing aid, a crucial advantage for many consumers. Digital and hybrid models are preferred by many consumers due to better performance in managing background noise, despite their significantly higher cost (Wood & Lutman, 2004).

## ***Cochlear Implants***

In the past, hearing aids have been the only available option for sound amplification for people who are Deaf or hard of hearing. Today however, rehabilitation options have dramatically changed with the advent of cochlear implant technology. A ***cochlear implant*** is a prosthetic device implanted in the inner ear that bypasses the external, middle, and inner ear hair cells, and uses electrical impulses to stimulate the auditory nerve directly through electrical impulses. Essentially, a cochlear implant performs the function of hair cells—stimulating the auditory nerves (Leake, Hradek, & Snyder, 1999).

Cochlear implant systems have both external and internal components and require at least four basic parts: an external microphone, signal processor, transmitter, and electrodes. The system receives sound through an external microphone, converts sound to magnetic impulses, and sends them to an audio processor connected to a transmitter beneath the skin. The transmitter converts the magnetic impulses to electrical impulses and sends them through the skin to a series of electrodes that have been implanted within the cochlea. The electrical signals stimulate the auditory nerves and are perceived as sound.



**Surgical Implantation Criteria.** Personal traits, including cultural and educational background, age, and preferred method of communication can play a more influential role in surgery than any physical factors.

**Outcomes.** The age at which an individual undergoes cochlear implantation, the length and severity of Deafness prior to implantation, and whether the person possesses useful speech reading skills are critical factors that influence cochlear implant outcomes. In general, individuals who are younger with shorter-term hearing loss and speech reading abilities have more successful outcomes than people with more adverse profiles, individuals who are prelingually Deaf, and those who have exclusively used manual communication.

### *Aural Rehabilitation*

Hearing aids and cochlear implants provide auditory feedback that may foster spoken language development in some consumers who are Deaf or hard of hearing due to improvement in speech perception (Panteleimidou, Herman, & Thomas, 2003). Aural rehabilitation plays a crucial role in the treatment and management of Deafness and hearing loss. For adults, aural rehabilitation focuses predominantly on the impact of hearing loss on communication and the procedures used to improve communication effectiveness (Hull, 2001).

Several methods of aural rehabilitation are centered on use of residual hearing and visual cueing with reinforcement for the development of auditory awareness and speech. *Auditory methods* use residual hearing abilities, with or without assistive devices, with special attention to development of listening skills. Techniques that focus on fostering awareness of the physical motion of speech and related sensations are considered *kinesthetic methods*. Increasingly, sophisticated computer-assisted systems are being used in kinesthesia (Panteleimidou et al., 2003). *Oral methods* combine assistive listening devices, speech reading, and kinesthesia to develop speaking abilities. *Speech reading* (inaccurately termed lip reading) allows an individual to comprehend speech by observing lip and facial movements along with body gestures to determine what is being spoken and in what context.

Aural rehabilitation includes other types of services, including training in *manual communication*. Manual methods of aural rehabilitation exist in contrast to oral strategies, embracing rather than discouraging the use of non-verbal communication. Manual communication training prepares the individual to communicate through non-verbal techniques such as finger spelling and signed languages including American Sign Language (ASL), and Signed English. *Total communication* is a highly inclusive rehabilitation option that involves selecting the most effective and appropriate strategies from all methods to maximize both expressive and receptive communication (Nicolosi et al., 1996).

## **Functional Limitations**

Functional limitations related to Deafness and hearing loss are largely dependent on pre-lingual versus postlingual occurrence, severity of hearing loss, presence or absence of other complicating factors such as a vestibular disorder or a secondary disability, and other situational factors. It is clear, however, that audition and communication difficulties are the primary functional limitation across all categories and configurations of hearing loss.

If an individual has a mild hearing loss, he or she will likely experience a slight difficulty in understanding speech when background noise is present. A person who has a mild-to-moderate loss will have some problems understanding conversation in group settings, and may need face-to-face communication to comprehend at least half of a conversation. A moderate hearing loss results in significant difficulty in group settings and in face-to-face verbal conversations. People who have a severe hearing loss are not likely to recognize voices or environmental noises, while individuals who have a profound hearing loss may only detect sounds through vibrations and are likely to use manual or visual methods of communication. Significant hearing limitations may result in marked difficulties in reading, writing, and comprehending English (Gallaudet Research Institute, 2003).

People who have a prelingual severe or profound hearing loss have considerable functional limitations in expressive and receptive language. For individuals with sensorineural hearing loss,



comprehension in noisy environments is more difficult than for people with conductive hearing loss. Conductive hearing losses do not usually result in functional deficits that are as significant as those associated with sensorineural hearing loss. People who also have severe tinnitus or vertigo may experience additional frustration, anxiety, and a notable disturbance of attention and ability to listen.

## **Psychosocial Implications**

Deaf or hard of hearing individuals often face significant isolation and social alienation due to functional limitations in audition and communication. Speech comprehension problems can result in a marked inability to participate in reciprocal conversations and social interactions (Villaume et al., 1997). The major psychosocial implications experienced by people who are Deaf or hard of hearing are social and familial isolation.

Feelings of isolation due to communication barriers within the family and society are prevalent. The impact of communication barriers on people who are Deaf and hard of hearing may begin at a very early age, especially if hearing loss occurred pre-lingually. The vast majority of parents and siblings of Deaf and hard of hearing children are hearing, resulting in incomplete communication within the family. Nearly a third of Deaf and hard of hearing children have difficulties with social interaction and behavior (Gallaudet Research Institute, 2003), probably as a result of communication barriers in the home.

Many people with hearing loss report negative experiences with the hearing world. Such experiences result in emotional and behavioral problems. Research indicates that nearly half of Deaf and hard of hearing people experience mental health conditions attributed to difficulties in communication.

## **Vocational Implications**

The broad vocational implications of functional limitations experienced by people who are Deaf or hard of hearing include unemployment and underemployment. Historically, jobs that require higher levels of reading, frequent interaction with coworkers, clients, and use of the telephone have not traditionally been open to people who are Deaf or hard of hearing. They tend to have trade jobs related to transportation, farming, and machine operation and are less likely to be in more lucrative professional areas. In some instances, Deaf individuals have faced significant bias from employers, and been shut out of the workforce completely. The cumulative effect of chronic unemployment and underemployment is markedly negative at the end of one's working years; the median net worth of people with hearing losses as they enter retirement is considerably less than reported for people without hearing loss (Brodwin, Parker, & DeLaGarza, 2003; Lucas, Schiller, & Benson, 2004). Limited abilities in detecting environmental sounds, difficulties in communicating with others, and occupational hearing requirements have resulted in limitations in job placement and have notable vocational implications.

### ***Environmental Sounds***

The ability to detect sounds such as warning bells, alarms, and door chimes, locate the source of sounds, and make distinctions among various sounds are critical in some work settings. Abilities to localize sounds and discriminate between sounds is especially crucial if an individual is working with or near machinery, so functional limitations in detecting environmental stimuli may result in work setting safety concerns (National Research Council, 2005). For example, the inability to hear or localize warning buzzers, alarms, or bells is a vocational limitation which could, if unnoticed, result in injury to the person or others in the work environment (Arlinger, 2003). Problems with balance, dizziness, and tinnitus impact a person's ability to function effectively in hazardous environments. General vocational precautions include avoidance of ladders, scaffolds, heights, dangerous machinery, and driving.

### ***Occupational Hearing Requirements***

Problems hearing can significantly impede employment in professions with specific job entry requirements. In some instances, individuals may be completely excluded from certain professions such as military service and law enforcement. While hearing may be important in many work settings, even severe or profound hearing loss does not prevent employment. Most young adults who are Deaf or significantly hard of hearing are employed at some level (Brodwin et al., 2003).

## **Accommodations**

Accommodations are those modifications, adjustments, auxiliary aids, and services used in home, work, and recreational settings to reduce barriers and diminish functional limitations. In the absence of accommodations, people who are Deaf or hard of hearing may be unintentionally excluded from almost all communicative interactions with the hearing world. A variety of available options for accommodations and assistive devices are available for use in activities of daily living and in work settings (Brodwin et al., 2003). Assistive communication technology, modifications of the environment, and use of communication strategies are particularly beneficial options for reasonable accommodation.

### ***Assistive Technology***

Bryant and Bryant (2003) have defined assistive technology (AT) as “the applications of science, engineering, and other disciplines that results in processes, methods, or inventions that support people with disabilities” (p. 2). Currently, a wide variety of AT is available. Technologies used by hearing people are useful to people with hearing loss, such as web-based chat, electronic mail, and text messaging. Additional communication technologies have been developed for people who are Deaf or hard of hearing including text-based telephone relay systems and video relay systems.

**Telecommunications Devices for the Deaf.** As early as 1874, telephone access for Deaf and hard of hearing users has been an issue. Unfortunately, early communication technologies were rarely efficient, increasing communication barriers for people who were Deaf or hard of hearing. To increase and facilitate communication between the Deaf and Hearing populations, the teletypewriter (TTY), also called telecommunications device for the Deaf (TDD), was developed, allowing people to type and receive messages over telephone lines. These systems continue to operate as a link between three parties. In such a system, the person with a hearing loss or speech impairment uses the teletypewriter to type out messages, while the hearing person uses a telephone and a relay operator conveys the messages to each through the appropriate voice or text format (Grossman, 2001).

**Video Relay Services (VRS).** This is an alternative to TTY/TDD relay service communication. VRS is a visual communication system requiring a minimum of three basic components: a video monitor such as a television or computer screen, a video camera, and a high-speed broadband line. Like the TTY/TDD system, VRS uses a third-party relay operator, but in the VRS system, the operator is also a sign language interpreter. People who communicate with sign language see and are seen by the interpreter through cameras linked to monitors. Using videophones, VRS is also used among people who use manual languages without a relay operator (Grossman; 2001; Nelson, 1996). Studies have shown that across all areas, participants demonstrated preference for the VRS over text-based services, despite the slightly higher cost of equipment. Participants noted that quality of the service was faster and more professional using the VRS system and reported greater positive impact on their quality of life.

### ***Environmental Modifications***

Across home, work, and leisure settings, individuals who are Deaf or hard of hearing may encounter situations in which a response to sounds in the environment is compulsory. Modifications to the environment that promote awareness of such environmental sounds and provide safety measures are valuable for Deaf and hard of hearing. Telephones, doorbells, and fire alarms may be linked to visual alerting devices such as flashing lights. Tactile alerting devices, such as vibrating pagers are also useful.

Noisy environments are often difficult. In many settings, individuals who are Deaf or hard of hearing benefit from noise reduction to promote use of residual hearing in communication. Quiet surroundings have less hearing interference due to background noise, and individuals may manage extraneous noises effectively through sound absorbing carpeting, upholstery, or panels. Well-lit surroundings are beneficial, as are surroundings free of objects or other barriers to visual sight lines, allowing for clearly visible manual communication or speech-reading (Geyer & Schroedel, 1999).

### ***Communication Strategies***

For successful communication between persons with hearing loss, it is necessary to establish and maintain attention and eye contact. It is acceptable to wave or gently tap individuals who are Deaf or hard of hearing to gain their attention (Foster, 1998). One should articulate clearly, although over-articulation distorts the shape of the speaker's mouth and hinders speech reading. Speaking quickly or too slowly may hinder speech reading and distort conversational cues. It is important not to look down or face away from a person who is Deaf or hard of hearing during communication, and speakers should avoid obstructing the mouth while talking (Luey, Glass, & Elliott, 1995).

Some people use sign language exclusively, while others may use spoken language or a combination of both. Those who communicate manually often have difficulty participating in situations where few or no other people use sign language, and a qualified sign language interpreter may be required to facilitate communication. Sign language training for family members and friends may significantly reduce isolation and improve home and community inclusion; sign language classes for employers and co-workers may serve as an accommodation (Geyer & Schroedel, 1999). When using interpreter services, it is appropriate for the hearing speaker to address the Deaf or hard of hearing person directly, and not look instead at the interpreter (Luey et al., 1995).

### ***Job Restructuring***

Job restructuring is a critical accommodation that relates to communication strategies, and typically involves redistribution or exchange of job duties or job assignments, redeveloping of job processes, and eliminating any non-essential job functions. For example, in work where speech communication is not essential, individuals may communicate through alternative means such as text messaging, electronic mail, or written memos (Geyer & Schroedel, 1999). Using support personnel to accomplish non-essential job functions, such as incidental telephone use, is an element of job restructuring (National Research Council, 2005). Changing the way meetings are conducted or providing transcripts of meetings are examples of changes to process. The selection of jobs and work environments not requiring an ability to hear is another possible job structuring strategy.

### ***Issues in Job Accommodations***

The Americans with Disabilities Act of 1990 (ADA) mandates collaboration among employees and employers in determining and providing reasonable job accommodations. A growing body of research suggests that people who are Deaf or hard of hearing face a number of challenges in obtaining accommodations. Employers exhibit greater willingness in providing job accommodations to workers in more prestigious positions, such as managers than to workers in lower status positions. Differences in hearing status may also account for differences among workplace accommodations provided or authorized by employers; in general, persons who are Deaf are more likely to request and receive accommodations than those who are hard of hearing (Geyer & Schroedel, 1999). Size of the company or organization seems to influence accommodation provision, with larger employers providing accommodations more readily than smaller companies.

## **Summary**

Understanding the unique needs of a person with hearing loss is a complex and sometimes challenging rehabilitation task. To serve the needs of these people, rehabilitation professionals must possess an understanding of critical dimensions of hearing, listening, and communication, and the impact these factors have on service provision. Although terms such as hearing, listening, and communicating are often used interchangeably, these concepts are actually unique. **Hearing** is best described as a passive function that involves the perception of environmental sound. Hearing includes having an awareness of sound, ability to locate sounds, as well as the capacity to discern the quality of sound as well as make distinctions related to pitch and volume. **Listening** is an active and purposeful process undertaken with the intention of experiencing sound. **Communication** is another process, one that requires bi-directional or multi-directional exchanges of information which includes hearing, listening, and comprehension (Kiessling et al., 2003).

The primary functional limitation of hearing loss is in the area of communication. Assistive listening devices, communication technologies, environmental modifications, and reasonable accommodation are available and may effectively address these limitations. Such options should be explored as part of comprehensive rehabilitation planning and service provision. Many people who are Deaf or hard of hearing have useful residual hearing abilities, and successfully use devices such as hearing aids or cochlear implants; some individuals are skilled in speech reading, while others are fluent in sign language. Each person has unique strengths on which to build. Despite the many challenges encountered by people who are Deaf or hard of hearing and by professionals who serve them, the opportunities for rehabilitation are abundant.

## Case Study

Alan Parchman is a 42 year-old married male with three children, ages twelve, fifteen, and sixteen. He has been working at a chemical plant as a kettle operator and tender (*O\*NET* code 51-9051.00) since graduating from high school. Mr. Parchman's job has required frequent exposure to both noise and ototoxic chemicals.

Three years ago, Mr. Parchman began to notice a decrease in hearing ability and developed intermittent severe tinnitus. He did not report the symptoms to his employer due to concerns about losing his position or being placed on unpaid medical leave. About six months ago, Mr. Parchman's symptoms worsened significantly, and at his wife's urging, he sought evaluation by his general practitioner.

His physician noted that Mr. Parchman had difficulty understanding speech in the office environment. She referred him to an audiologist for further evaluation. The audiological results demonstrated a pure tone average (PTA) in the right ear of 60 dB, and a PTA in the left ear of 55 dB. Mr. Parchman further demonstrated 82% word understanding in the right ear, and 78% word understanding in the left ear. The tympanogram was within normal limits. Mr. Parchman was diagnosed with sensorineural hearing loss due to noise and ototoxic chemical exposure and was scheduled for a hearing aid fitting.

### Questions

1. What level of hearing loss does the consumer have? What other characteristics of the hearing loss can be identified?
2. What transferable skills does this individual have; identify occupations using these skills?
3. If the consumer decided to remain in his current job, how would you as the rehabilitation counselor approach this decision? Provide supporting arguments.
4. What is this person's vocational profile?
5. Identify the consumer's likely functional limitations and recommend accommodations.
6. What are the implications of this type of hearing loss in regard to future functional limitations?

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## Chapter 12

# VISUAL DISABILITIES

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## Introduction

Vision is a complex and wonderful process of the human body. Although vision can be described as being analogous to a camera, it involves much more. The eyes are a matrix of millions of cells that convert light information into electrical signals which are processed in the brain. Two-thirds of the brain is involved in the complex process of vision (Crick & Khaw, 2003). The visual centers of the brain communicate with the motor, auditory, language, speech, and executive processing areas of the brain to influence reading, facial recognition, spatial awareness, and problem solving (Wolintz, 1976).

Vision allows us to gather information about the surrounding environment at distances well beyond the reach of the arms and fingers in a matter of milliseconds. The central visual system allows us to identify details and enables us to read, identify faces, and perform fine motor tasks. The peripheral visual system provides the brain with continuous information to assist with independent mobility in familiar and unfamiliar areas. Integration of both central and peripheral visual information allows adults with normal vision to perform job tasks efficiently and independently (Kanski, 2007).

Blindness and loss of vision are frightening and emotionally stressful. Vision impairment causes functional difficulties with reading, driving, walking, and working independently and may cause emotional, psychological, social, and financial difficulties. In the United States, over 70% of adults between the ages of 20 and 65 who are visually impaired are unemployed (Congdon et al., 2004; Mitchell, 2004). The absence of employment can exacerbate emotional, psychological, and financial difficulties of patients with vision impairment and contribute to the development of other problems. Rehabilitation counselors play a pivotal role in developing treatment plans that may involve ophthalmologists, optometrists, psychologists, orientation and mobility specialists, and teachers for the visually impaired to provide training and promote maximum productivity and independence at home and in the workplace. In this chapter, the many different aspects of vision are described, along with the ways diseases and disorders of the visual system impact the functional abilities of working age adults.

## Vision Impairment and Disability

The rehabilitation counselor must have a thorough understanding of the degree of vision impairment and functional vision of a client before developing a vocational plan. This can be challenging because the counselor must rely on medical reports, chart notes, and information provided by the client. Unfortunately, medical reports and chart records may describe the diagnosis without reflecting the client's visual strengths and limitations. Clarity of sight, peripheral vision, depth perception, eye-hand-foot coordination, color vision, day and night vision, and visual processing skills are some of the visual sub-skills that affect visual functions. The degree of function of each of these visual skills can range from non-functional to normal (Windsor & Windsor, 2001). Deficits in a specific skill can have significant ramifications on the client's ability to perform one task but may not affect another. It is crucial to define vision impairment as it relates to specific occupations or job tasks (Brodwin, Parker, & DeLaGarza, 2003).

In the United States, eye care professionals, the Social Security Administration, and other government agencies have adopted the definitions set forth by the World Health Organization (1994) to

provide a uniform definition of vision impairment and set standards to determine who qualifies for government services and benefits.

- *Functionally blind* describes individuals who are not able to perceive light.
- *Legal blindness* is visual ability with a best-corrected acuity of 20/200 or worse in the better eye or the widest diameter of peripheral vision measuring 20 degrees or less in the better eye.
- *Visually impaired or partially sighted* describes those with a best-corrected visual acuity of 20/70 to 20/180 in the better eye or a peripheral field of vision between 21 degrees and 140 degrees.
- *Fully sighted* describes people with a best-corrected visual acuity of 20/40 or better in the best eye.

There are 3.7 million adults between the ages of 20 and 65 who are visually impaired, despite wearing corrective lenses, while 337,000 are legally blind (Brodwin et al., 2003; Vitale, Cotch, & Sperduto, 2006). Although the definitions of functionally blind, legal blindness, and visually impaired or partially sighted are often used in medical reports, they do not completely describe a client's functional vision. These definitions only consider visual acuity and peripheral vision. Thus, the rehabilitation counselor should encourage the eye doctor to provide additional information regarding color vision, night vision, contrast sensitivity, depth perception, glare sensitivity, and other visual skills.

### ***Eye Care Professionals***

The counselor works with eye care professionals for information regarding the client's functional vision. In the United States, optometrists and ophthalmologists are licensed to diagnose and treat eye diseases and disorders of the visual system. Optometrists are often considered to be the primary eye care professionals in managed care health systems. If the patient requires surgery or additional medical treatment, the optometrist refers the patient to an ophthalmologist, while the ophthalmologist may refer the patient to a low vision specialist if the patient's vision cannot be fully corrected after medical and surgical treatments have been implemented (Kanski, 2007).

### ***The Eye Examination***

The general eye examination involves a case history, measurement of distance and near clarity of sight, screening of the neurological function of the visual system, inspection of the health of the eyes, and a refraction to determine the need for glasses or contact lenses (Crick & Khaw, 2003). The routine eye examination is usually performed by an optometrist, but may be performed by a general ophthalmologist. This section describes information obtained and the specific tests performed to quantify various visual skills and illustrates the functional implications.

#### ***Case History***

The case history is one of the most informative components of the eye examination. A medical history and results from prior eye examinations provide clues to explain the patient's symptoms and complaints. Questions regarding vision problems of family members help the examiner gain insight regarding the possible inheritance of genetic eye conditions. The case history provides the doctor with the opportunity to understand the patient's specific complaints, difficulties, goals, and psychosocial needs.

#### ***Visual Acuity Testing***

A Snellen eye chart is the most common test used to measure clarity of sight. It is typically placed 20 feet from the patient, who is then directed to identify letters of varying sizes with each eye. Each letter on the Snellen chart, called an optotype, has a specific size. The physician records the distance from the chart to the patient and then writes the smallest optotype the patient is able to identify in the form of a fraction. Thus, a patient who can identify the 20-size letter from 20 feet has 20/20 visual acuity, whereas the person who can only read the 200-size letter from 20 feet has 20/200 acuity. Visual acuity is measured for each eye with the patient's best correction of glasses and contact lenses to determine visual status. If the patient is not able to read the 200-size letter from 20 feet with either eye with the best corrected vision, he or she is considered to be legally blind. The patient with 20/200 acuity has difficulty identifying faces, reading street signs, and identifying text from a chalkboard (Kanski, 2007).

### ***Visual Field Testing***

A physician uses a visual field test to estimate level of functional vision. Defects in the central field affect reading, driving, eye-hand coordination, color vision, and facial recognition, while peripheral field defects cause difficulties with spatial orientation, mobility, and balance. Retinitis pigmentosa, glaucoma, and neurological visual disorders often cause loss of peripheral vision (Crick & Khaw, 2003).

The most sophisticated method of testing peripheral vision is through use of computerized perimetry. In this test, the patient is positioned in front of a large dome in which lights of varying sizes and intensities flash in different parts of the central and peripheral fields of vision. This form of perimetry detects a wide array of vision problems including brain tumors, neurological disorders, psychological blindness, and eye disease. Low vision specialists use the computerized perimeter to map out regions of the eye that have the strongest sensitivity and use lenses and prisms to deflect images onto the most usable fields of vision.

### ***Refraction***

After the doctor has inspected the health of the eyes, he or she performs refraction. This test determines the best lens prescription to improve visual acuity. Lenses focus light on the macula of the retina to achieve the sharpest image; the macula is responsible for detailed sight. When light focuses in front of or behind the macula, blurred sight occurs. Hyperopia (farsightedness) is the condition where light focuses behind the macula, reducing the clarity of sight. Patients with uncorrected hyperopia generally have difficulty reading small print but may also have blurred distance sight. The doctor will prescribe “plus (+)” lenses to converge the light to focus on the macula to maximize visual acuity. Conversely, myopia (nearsightedness) is a condition where light focuses in front of the macula and causes blurred distance sight and sometimes blurred near vision. “Minus (-)” spectacle lenses are prescribed to focus light on the macula to improve detailed sight (Hoyt, Miller, & Walsh, 2008).

Visual field defects due to eye disease do not necessarily affect functional vision. Cataracts, glaucoma, diabetic retinopathy, and other eye diseases initially cause areas of reduced sensitivity in one eye; the progression of the field loss may be slow (Congdon et al., 2004). The brain is able to “fill in” the dimmer areas and the patient may not have functional problems. However, as the disease progresses, areas of reduced sensitivity become absolute blind spots, called scotoma. Blind spots that affect the same field of vision in both eyes create significant functional difficulties.

## **Parts of the Eye**

### ***Orbit***

The orbit is the eye socket, housing and protecting the eyeball from direct trauma. It consists of several bones that are susceptible to fracture during trauma. Six ocular muscles of the eye have tendons that are attached to the orbit. This provides the foundation which allows eye movement. Brain tumors, thyroid disease, and trauma to the orbit itself can restrict the movement of the eyes, cause diplopia (double vision), and affect the positioning of the eyes.

### ***Eyelids***

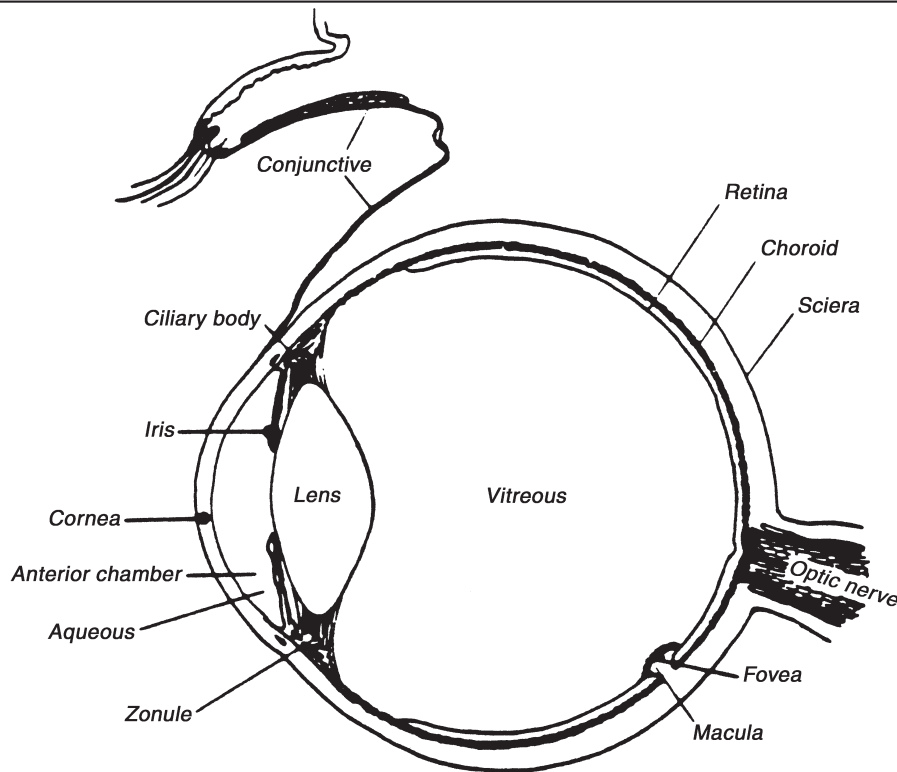
The eyelids, which allow for lubrication, protect the cornea and the front of the eye. A variety of glands produce tears to lubricate the eyes whenever an individual blinks. Trauma, tumors, and neurological disorders can affect the function of the eyelids. Incomplete closure of the eyelids causes dry eyes, blurred vision, sensitivity to light and glare, and eye pain. Abnormal turning-in of the eyelid is called entropion, while abnormal turning out of the eyelid is called ectropion. As with scarring, both conditions threaten ocular surface integrity.

### ***Cornea, Sclera, and Conjunctiva***

The cornea is the multi-layered transparent structure in the front of the eye. Vision of a patient with a corneal scar is analogous to a view through frosted glass; the patient will have blurred sight and extreme sensitivity to light. A patient with corneal disease may benefit from laser treatment, as well as corneal



**Figure 1**  
**The Eye**



Source: M.W. Leitman. (1994). *Manual for Eye Examination and Diagnosis* (4<sup>th</sup> ed.). Oradel, NJ: Medical Economic Books. Reprinted with permission of Blackwell Scientific Publications.

transplantation surgery to improve functional vision. Some patients with myopia and astigmatism benefit from refractive surgery which alters the shape of the cornea to eliminate the need for glasses (Kanski, 2007).

### ***Iris, Pupil, and Anterior Chamber***

The iris is the muscular tissue of the eye that provides eye color. In the center of the iris is a round opening called the pupil which regulates the amount of light entering the eye. In dim illumination, the pupils dilate, while they constrict in bright illumination. Eye diseases that affect the iris cause mild to moderate sensitivity to glare and bright light. Chronic inflammation of the iris (iritis) causes severe sensitivity to light, blurred vision, and disabling eye pain (Kanski, 2007).

### ***Crystalline Lens***

Behind the iris is a transparent focusable lens called the crystalline lens. The crystalline lens changes its shape when the ciliary muscle located on the iris contracts and relaxes, enabling the eye to focus at different distances. With aging, the lens loses its ability to change shape and the patient may require glasses for reading.

### ***Vitreous***

The vitreous, located behind the lens and iris, is filled with a gel-like substance called vitreous humor. Light must pass through the pupil, crystalline lens, and vitreous humor before it can focus on the retina. Scar tissue and blood can interfere with the manner that light focuses on the retina, causing blurred sight, reduced peripheral vision, poor color vision, and distorted vision in which straight lines appear wavy

(metamorphopsia). Hemorrhages in the vitreous due to diabetic retinopathy, trauma, and retinopathy of prematurity may develop scar tissue and retinal detachment (Kanski, 2007).

### ***Retina***

The retina is the light sensing tissue that consists of millions of rod and cone cells that absorb light and send electrical signals through the optic nerve to be processed by the brain. Cone cells are located in the centermost region of the retina. The macula is the area of the retina that consists entirely of cone cells and is responsible for detailed clarity of sight, color vision, high spatial contrast sensitivity, and ability to adapt to different lighting conditions. Rod cells are located on the periphery of the retina and provide peripheral vision, perception of movement, night vision, and low spatial contrast sensitivity, important for mobility.

Retinal hemorrhages occur secondary to diabetes, retinal tears and detachments, retinopathy, Stargardt's disease, retinitis pigmentosa, macular degeneration, and other retinal disorders. A patient with central retinal problems has blurred sight, poor color vision, reduced contrast perception, and sensitivity to light. Peripheral retinal problems cause difficulties with mobility, reduced night vision, and extreme sensitivity to light and glare. Retinal diseases, such as retinitis pigmentosa and complete retinal detachments, can result in total blindness (Panek, 2002).

### ***Optic Nerve***

The optic nerve is a bundle of nerve fibers, containing the rod and cone cells that send information to the visual cortex of the brain. Abnormalities of the optic nerve are diagnostic of eye disease such as glaucoma, optic nerve hypoplasia, optic nerve atrophy, and systemic illnesses, such as multiple sclerosis, brain tumors, and increased intra-cranial pressure. Eye injuries and trauma to the head are also causes of optic nerve damage. Optic nerve disorders lead to varying degrees of blurred sight, color blindness, peripheral and central visual field defects, and total blindness (Crick & Khaw, 2003).

## **Color Vision and Magnifiers**

Color vision is controlled by cones in the central retina. Some color vision deficits are more common in men than women, and may be partial, involving certain hues of color (such as red/green color blindness). Total color blindness is rare and associated with retinal pathology and a corresponding decrease in visual acuity (Panek, 2002). Colored filters and lenses enhance the perception of colors that are not perceived and allow the patient with color vision problems to be more functional.

### ***Eye Teaming and Binocular Vision Testing***

Binocular vision involves the ability to coordinate the eyes together. Eye coordination testing administered to a patient who complains of double vision, frequent loss of place when reading, and eye strain. Each eye has six extra-ocular muscles that control the movement of the eyes along with one internal muscle, which changes the shape of the internal crystalline lens to focus on near and distant objects. Abnormal coordination of these muscles results in a crossed or turned eye (strabismus), double vision, poor depth perception, blurred sight, headaches, and eyestrain. Trauma, tumors, amyotrophic lateral sclerosis (ALS), Parkinson's disease, diabetes, multiple sclerosis, and myasthenia gravis can affect eye movement skills and hinder a person's ability to perform daily activities (Falvo, 2009).

### ***Low Vision Rehabilitation***

Low vision rehabilitation has been shown to be extremely beneficial for adults with sub-normal vision (Crandell, Jr. & Robinson, 2007). Low vision specialists are doctors who design, customize, and prescribe visual aids and provide training to help patients with low vision perform their desired goals independently. Individuals are generally referred to low vision specialists by ophthalmologists when medical and surgical treatments no longer correct vision.

The low vision examination identifies visual strengths and limitations and offers possible solutions. Specialized glasses, filters, prisms, and technology are used to help a person read, write, use a computer, cross the street independently, and perform other activities.

### ***Customized Low Vision Aids***

Low vision specialists develop glasses and visual aids that will take advantage of the individual's visual strengths and compensate for their weaknesses (Crandell, Jr. & Robinson, 2007). Telescopes and high-powered lenses magnify images to allow a person to see small print, while prisms are used to reduce eyestrain or double vision caused by eye teaming problems. Filters and colored lenses maximize the perception of colors and increase contrast, as well as simulate lighting conditions that provide the best acuity. Additionally, prisms and eccentric viewing training can be incorporated to deflect the image onto the most usable areas of the retina, maximizing overall visual function.

### ***Bioptic Spectacles***

Bioptic spectacles are glasses that contain a small telescope to improve visual acuity, helping an individual see traffic signals, street signs, presentations at meetings and school, and read a computer screen. Bioptic glasses are one of the most useful low vision aids enabling people to read small print and perform specialized work tasks, such as reading the labels of items on shelves and the numbers on a cash register.

### ***High- powered Reading and Writing Glasses***

Glasses with specialized lenses magnify reading materials up to six times (Mitchell, 2004). They may be customized to reduce glare, increase contrast, and are available in bifocal forms to allow a person to focus both far and near. Accountants, bankers, electronic and computer technicians, and computer users benefit from high-powered reading glasses when performing work tasks.

### ***Visual Field Expanding Glasses***

Prisms, reverse telescopes, and amorphic lenses can be incorporated in glasses to help individuals with reduced peripheral vision due to glaucoma, retinitis pigmentosa, and traumatic head injury or stroke. Reverse telescopic glasses shrink the peripheral vision into the usable field of vision and allow the person to see an entire sidewalk to improve mobility, for example.

### ***Telescopes***

Hand held telescopes are helpful when a person has blurred distance sight. Monocular telescopes can focus on distant and near objects. Adults with low vision use telescopes to read bus numbers, street signs, and traffic signals to assist with independent mobility.

### ***Magnifiers***

Magnifiers are available with their own light source and are useful to read mail, prices at stores, and for spot reading. Unfortunately, magnifiers have many physical limitations in that one sacrifices field of view, contrast, and quality of the image as the level of magnification increases.

### ***Electronic Video Magnifiers***

Electronic video magnifiers offer more magnification, higher contrast, and a wider field of view than hand-held magnifiers. They consist of a small camera that displays the images on a large screen, allowing the user to have both hands free to write, draw, and manipulate objects. Electronic video magnifiers change the contrast level to allow the person to read faded print. They also change the color of the background and text to compensate for a patient's problems with glare and color vision.

## **Common Causes of Vision Impairment**

### ***Astigmatism***

Astigmatism is a condition in which the eye is not completely spherical, resulting in an uneven focus on images. A person with astigmatism who looks at a cross may perceive the vertical line of the cross as blurred while the horizontal line is seen as sharp (Hoyt et al., 2008). Eye doctors will prescribe cylindrical lenses for individuals with this condition.

## ***Diabetic Retinopathy***

Diabetic retinopathy is the leading cause of blindness among adults between the ages of 20 to 74 (Mitchell, 2004). It is a progressive condition in which the retina is damaged due to prolonged lack of oxygen. The onset of vision problems among those with diabetes is usually during adulthood (Unger, 2007).

The prevalence of diabetic retinopathy is directly related to the duration of the disease. For those with Type 1 (insulin dependent diabetes mellitus), more than 90% demonstrate evidence of diabetic retinopathy within 15 years. Sixty to 80% of individuals with Type II (non-insulin dependent diabetes) show signs of diabetic retinopathy after this same time period (American Diabetes Association, 2006).

## ***Optic Atrophy***

Optic atrophy is a condition in which some or all of the optic nerve fibers have been damaged, resulting in mild to profound loss of vision. This can be present at birth or develop from various conditions. Lack of oxygen (anoxia, hypoxia), trauma, inflammation, vitamin deficiency, drug toxicity, demyelinating diseases such as multiple sclerosis, and increased intra-cranial pressure cause optic atrophy (Congdon et al., 2004). Individuals may have blurred sight, red-green color blindness, reduced contrast sensitivity, limited depth perception, poor eye-hand-foot coordination, and large blind spots in the visual field. Visual losses related to optic atrophy cause difficulties with reading, walking, driving, and performing a wide variety of tasks. Optic atrophy is not treatable medically or surgically. The optic nerve cannot be transplanted because there are millions of nerves that must be precisely attached to the corresponding neurons of the optic tract (Crick & Khaw, 2003).

## ***Glaucoma***

In glaucoma, the intra-ocular pressure is too high, causing damage to optic nerve fibers. In a normal eye, aqueous humor is continually produced and drained through a structure called the trabecular meshwork, located in the anterior chamber angle. In open angle glaucoma, the trabecular meshwork does not drain aqueous fluid normally, resulting in an increase in intra-ocular pressure. Elevation of pressure is slow and generally is not noticeable in vision until significant loss has occurred. Small blind spots in the mid-peripheral field of vision initially develop and later progress to cause severe peripheral vision loss, reduced night vision, tunnel vision, and blindness. Advanced glaucoma creates difficulties with walking, reading, writing, and performing activities under dim illumination (Falvo, 2009).

Glaucoma is common among adults of specific sub-groups. It is more prevalent among adults over age 40, African-Americans, and those with a family history of glaucoma (Qulgle & Vitale, 1997).

Glaucoma is a chronic disease and daily medication to sustain the desired intra-ocular pressure is required. Compliance with therapy is frequently a problem because symptoms often are not immediate when medications are discontinued (Panek, 2002). Individuals with glaucoma need consultation with a low vision specialist to prescribe visual aids, including visual field expanders, high powered reading glasses, night vision aids, computer and assistive technology, and specific lighting for the workplace and home.

## ***Retinitis Pigmentosa***

Retinitis pigmentosa (RP) describes a variety of degenerative eye conditions that affect the cells of the retina causing night blindness, reduced peripheral vision, sensitivity to glare, and blurred sight (Hartong, Berson, & Dryja, 2006). Generally symptoms develop between the first and second decades of life. In severe forms of retinitis pigmentosa, individuals lose nearly all their peripheral vision and are left with a small central island of vision. They are considered legally blind due to the absence of peripheral vision. Some maintain 20/20 visual acuity, while others have blurred sight and a blue-yellow color vision deficiency.

Patients with RP tend to have many functional problems. Night blindness is the most common symptom. Adults with night blindness may have trouble finding their chairs in dimly lit restaurants or movie theatres, or may avoid going out at night altogether. As peripheral vision deteriorates, driving and walking become increasingly difficult. Color vision problems and blurred sight can also develop, making reading and identifying the color "blue" problematic.

Low vision aids are helpful for individuals with RP. Visual field expanding glasses increase field of vision two to three times, while night vision glasses enhance mobility at night. Low vision spectacles improve visual acuity for distance and reading, while specialized filters improve contrast vision and allow patients to adapt more readily to different lighting conditions. Desk lamps, track lighting, and full spectrum fluorescent light fixtures enhance illumination level. Orientation and mobility training is essential for independent mobility (Hartong et al., 2006).

### ***Macular Degeneration and Stargardt's Disease***

Macular degeneration, a disease of the central retina, is a leading cause of vision impairment. The macula (central retina) is a small region of the retina responsible for detailed clarity of sight, color, depth perception, and adaptation to light and glare. There are two main forms of macular degeneration. Dry (atrophic) macular degeneration is the most common form in which the cells of the macula atrophy due to age. It can occur in teenagers and young adults and is called Stargardt's disease. The second form is called wet (disciform) macular degeneration. In this form of macular degeneration, abnormal blood vessels develop under the retina forming a sub-retinal neovascular membrane that result in sub-retinal hemorrhages. Leakage of blood under the macula can result in a sudden loss of central vision (Mitchell, 2004). Symptoms of macular degeneration include blurred distant and near sight, sensitivity to glare and bright light, poor contrast vision, reduced stereoscopic depth perception, and color vision problems. People with this disability experience difficulty driving, reading, and performing tasks which require fine dexterity.

Presently, there are no known treatments to reverse the loss of vision due to dry macular degeneration and Stargardt's disease. Anti-oxidant vitamins and protection from the harmful rays of the sun have been reported to be beneficial for people with both types of macular degeneration while laser photocoagulation, photo dynamic therapy, and intra-ocular injections of anti-angiogenic medications help patients with wet macular degeneration (D'Amico, 2005).

### ***Head Injury, Neurological Vision Impairment, and Traumatic Eye Injuries***

Head injury, blunt trauma, sports, and employment-related injuries are common causes of vision loss among adults. There are approximately 2.5 million eye injuries in the United States yearly (Congdon et al., 2004). Penetrating injuries to the cornea and globe of the eye can result in total blindness due to retinal detachment, glaucoma, optic nerve damage, and ocular inflammation. In other cases, symptoms include blurred sight, distorted vision, severe sensitivity to glare, blind spots in the central and peripheral fields of vision, eye movement problems, and double vision. These eye problems interfere with depth perception, eye-hand-foot coordination, balance, and mobility.

Traumatic brain injuries (TBI) cause significant vision problems including double vision, poor depth perception, severe sensitivity to light, and profound visual processing difficulties (Panek, 2002). Patients have difficulty perceiving size, shape, depth, and have problems performing basic tasks, such as placing a cap on a water bottle. Some people with TBI have visual neglect where they ignore all objects on one side of their midline, including their own body; they may not comb half their hair, as well as leave food untouched on one side of the plate.

Cerebral vascular accidents (stroke) cause symptoms similar to traumatic head injury. When the stroke or injury affects the right side of the brain, there may be no peripheral vision on the left side with either eye and weakness on the left side of the body. Neurological involvement to the right occipital and parietal lobes of the brain often significantly impact visual perception; as a result, the individual would have poor depth perception, difficulty recognizing faces, and impaired visual-spatial perception skills. Conversely, strokes and injuries to the left occipital and parietal lobes of the brain cause a loss of peripheral vision on the right side of each eye, reduced motor function on the right side of the body, and affect language, speech, word recognition, and ability to remember sequences of information (Congdon et al., 2004).

### ***Cataracts***

A cataract occurs when the internal lens of the eye becomes clouded or opaque. Cataracts may be present at birth or develop at any age as a result of trauma, diabetes, steroid use, and aging (Mitchell, 2004).



Cataracts cause a wide variety of vision problems ranging from blurred sight, reduced color vision, glare sensitivity, reduced peripheral vision, double vision, and even total blindness. Dense cataracts that significantly impair vision may be surgically removed and an artificial lens implanted in the eye to restore its focusing power. When cataracts develop after the age of seven and there is no other eye pathology, individuals typically regain normal vision after cataract surgery.

Adults who are born with congenital cataracts may continue to have sub-normal vision even after cataract extraction surgery. Limitations include blurred vision, nystagmus (uncontrollable shaking of the eyes), strabismus (mis-aligned eyes), and severe sensitivity to glare. Visual acuity of these people can range from 20/40 to 20/200. Low vision aids and assistive technology enhance remaining vision.

## **Congenital Causes of Vision Impairment**

### ***Albinism***

Albinism is a congenital, inherited condition that affects the pigmentation and color of the eyes and may also affect the skin and hair (Crandell, Jr. & Robinson, 2007). Individuals with albinism have a very high degree of functional vision. Because the visual condition has been with them all their lives, they generally have learned to adapt to their vision impairments. These individuals usually have visual acuity ranging from 20/70 to 20/200; they have normal peripheral and color vision. People with albinism tend to have better vision at night and respond well to tinted contact lenses, specialized filters, and low vision aids to improve both their distance and near clarity of sight. Computer and assistive technology, orientation and mobility training, and independent living skills training are helpful for people with albinism who have severe sensitivity to light (Brodwin, Star, & Cordoso, 2007).

### ***Retinopathy of Prematurity***

Retinopathy of Prematurity (ROP) is a congenital condition that develops as a response to premature birth. It primarily affects the retina of newborn children born before 32 weeks of gestation and those weighing less than two pounds. Children with ROP may develop abnormal blood vessels in the retina which form scar tissue and can pull or detach the retina, causing blindness. The scar tissue in the eye often alters the shape of the eye causing it to be elongated which results in high myopia. ROP is related to the concentration of oxygen provided to a newborn child.

## **Functional Limitations**

### ***Emotional Adjustment***

While discovering new ways to accomplish tasks which formerly required sight, some clients experience a conflicting desire to deny that any change in method or lifestyle need occur. There are feelings of loss, sadness, and anger after learning of permanent visual impairment. Adjusting to vision loss is an emotional process and clients must find ways to express their feelings and adapt to this radical change (Kinash, 2006). Losing one's sight involves creating a new identity. Once the client begins to use the other senses and find alternative ways of doing activities, however, life can return to normal (Guide Dogs for the Blind, 1999).

Family and friends may find it difficult to accept a person's vision loss. They may become overprotective, thus limiting an individual's ability to function independently. The period of greatest emotional stress usually occurs at the onset of visual loss. While sudden loss of vision is extremely traumatic, it may be easier to cope than the uncertain, slow visual loss seen in many chronic ocular diseases.

Age plays an important part in the way the person reacts emotionally to loss of sight. Younger people are more resilient from the severe emotional sequelae generated by loss of sight. Middle-aged individuals are at greater risk of having severe psychological difficulty, perhaps because of other stresses associated with changes occurring during mid-life. Another variable is the overall degree of visual loss; the more profound the visual loss, the slower and more challenging the adjustment (Crandell, Jr. & Robinson, 2007).

The stages of grieving outlined by Dr. Elizabeth Kubler-Ross apply to vision loss as with most disabling conditions. Denial, anger, bargaining, depression, and acceptance are the major steps of reaction to visual loss. Not everyone goes through all the stages or in this order (Kubler-Ross & Kessler, 2005).

Carroll (1961) outlined secondary losses that occur with loss of sight, including diminished physical integrity, decreased visual contact with the environment, loss of a means of communication, and reduced mobility. With some loss of physical integrity, the individual may no longer feel like a “whole” person, resulting in feelings of isolation and withdrawal from one’s social network. The sighted person easily recognizes distant object characteristics including size, shape, position, speed, and direction; persons with visual impairment have lost part or all of this recognition ability. Unless other cues exist, the individual is limited in contact to the more immediate environment (Crandell, Jr. & Robinson, 2007). This limitation evokes fear and anxiety. Body language, facial expressions, and shadows suggesting time of day are missing when a person no longer has vision.

“Visual disability” refers to one’s inability to perform tasks visually. Both physical and psychological factors are important when defining functional status of the individual with visual impairment. A person with total visual loss but with newly acquired competencies may still achieve physical, emotional, and financial independence, as well as social integration (Kinash, 2006).

### ***Physical Adjustment***

Loss of sight is a major disruptive factor in daily living. Inability to perform essential tasks, such as reading, driving, and recognizing faces poses barriers to successful employment, along with fatigue, depression, and anxiety. As with other disabilities, visual loss negatively impacts most activities, and places stress on the individual’s daily living activities.

Use of one’s other senses frequently compensates for loss of visual acuity. Orientation and mobility specialists train clients to navigate with the assistance of white canes or guide dogs, stressing the importance of adaptive skills, such as listening to traffic flow to determine when it is safe to cross a street. Orientation and mobility play a vital role in an individual’s perception of personal freedom and independence. Factors that contribute to the degree of orientation and mobility include residual vision, age at onset of visual loss, posture and balance, intelligence, body image, space orientation, auditory tactile abilities, and personality (Panek, 2002). Substantial loss of peripheral field of vision is more debilitating than loss of central vision (Windsor & Windsor, 2001).

A white cane is the primary mobility aid used by persons who are blind or visually impaired. It is recommended that training begin following diagnosis; yet, clients with some progressive eye disorders may decide to wait until vision loss impacts mobility. Those in need of aid must first acknowledge loss of vision to be comfortable using a white cane, a clear identifier of a person with vision impairment both to the individual and others. Clients are taught to walk safely down the street by rhythmically touching the tip of the cane side-to-side in front of them to obtain advanced knowledge of what is ahead. Sounds and smells also provide information about one’s surroundings.

While some people prefer to use a red-tipped white cane for ambulation, many feel more comfortable relying on a guide dog (Guide Dogs for the Blind, 1999). Traveling with a guide dog differs from using a cane in that the dog navigates around obstacles, stops for stairs and other changes in elevation, and avoids overhead obstructions. The guide dog and owner work together as a team; the person needs to know the destination and how to get there to carefully instruct the dog.

### ***Vocational Adjustment***

As with daily living skills, vocational skills are a major component of successful independent functioning. The capability to perform job tasks well and provide for one’s self and family greatly enhances self-esteem, helps reduce financial concerns, and decreases family conflict. Components of adjustment to visual loss, which include actual physical vision, orientation mobility, daily living, and vocational skills all interconnect during complex life events. In addition to remaining visual capabilities, those with visual impairment can function independently by using physical dexterity, non-visual job skills, natural talents,

and refinement of the senses (Panek, 2002). These competencies, besides residual vision, help the patient enhance self-esteem and quality of life.

Advances in assistive technology play a major part in enabling persons with vision impairment to perform essential job tasks. The rehabilitation counselor plays a significant role in assessing assistive technology needs and potential accommodations that enable such clients to succeed in the workforce (Brodwin et al., 2007). He or she can also help dispel employer misconceptions about the capabilities of people with visual impairment.

## **Rehabilitation Potential**

Although the different types and degrees of vision impairment call for individualized approaches, the general goal of rehabilitation remains the same in each case - optimal physical functioning, economic independence, and social integration. Potential for rehabilitation depends on the physiology of the impairment (amount, type, and progression), conditions of the immediate environment, attitude and motivation, and use of residual vision. Since it is the auditory system that is the mainstay of mobility for clients with severe visual impairment, it is necessary for the counselor to determine the status of the person's auditory system. Duration and severity of disability, degree of personal adjustment, and positive or negative attributes of the family and the environment influence the vocational rehabilitation of people with visual impairment (Carroll, 1961).

Rehabilitation efforts focus on providing the best possible use of residual vision (vision enhancement), and assisting with development of non-visual skills that can substitute for lost visual function (vision substitution). Vision enhancement involves use of magnifiers, telescopic aids, colored filters, and computer and video-generated magnification screens. These devices generally present an enlarged image that is more readily seen (Windsor & Windsor, 2001). Braille, an example of vision substitution, is a tactile system developed in Paris by Louis Braille in 1829. Braille typewriters make it easier for people to take class notes, write, and read. Software is available that converts computerized text into Braille which can then be embossed on a Braille printer.

Residual vision can be aided by computers and assistive technology. Advances in technology allow patients with vision impairment to read, write, and perform job tasks and daily living activities independently (Brodwin et al., 2007). Thorough assessment of the client's need for assistive technology is beneficial. A vocational evaluator can assess the actual worksite, taking into consideration positioning of furniture and existing lighting to determine which aids promote optimum performance of essential job functions.

A most effective technology for clients with low vision is video magnifiers, also known as closed circuit televisions (CCTVs). This equipment provides up to 50 times magnification and offers a brighter, sharper, and wider field of view than magnifiers and glasses. Many models have special features in which the background and text colors are customized to maximize reading comfort. Other models can be interfaced with a computer and allow the user to share the same screen to see text both on the computer and on paper (Kinash, 2006).

Hand-held video magnifiers are useful for patients with low vision to read print when they do not have access to a desktop magnifier. These video magnifiers produce a bright and sharp image on an LCD screen. Distance and near focusing video magnifiers allow the user to see distant objects and reading material in a magnified view at work, school, and home (Crandell, Jr. & Robinson, 2007). This equipment is portable and is useful for clients who are attending conferences, lectures, and presentations.

Many clients with low vision who are re-entering the workplace need assistance in seeing the print on a computer screen. Specialized glasses maximize sight at the distance of a computer screen, while a larger computer monitor facilitates seeing. Windows XP and Macintosh OSX operating systems have built-in features that allow those with vision impairment to modify the computer settings.

Working age adults who require additional help to see a computer screen may benefit from special computer software for the visually impaired (Brodwin et al., 2003). Software magnification programs magnify all images on the computer screen and can also change the color of the background and text to help people with poor contrast vision and those with severe sensitivity to glare. These programs enable an

individual to access information from the computer, read stock charts, and perform various graphics programs.

Screen reading programs (SRPs) are available to help people who are functionally blind, as well as those who experience eye strain after reading for short periods of time (Windsor & Windsor, 2001). These programs can read text aloud from the screen thus allowing clients to access information from the Internet, type documents, and perform other software applications. Scanning software programs are available; they allow users to scan books, letters, and other documents. The text is then displayed in a magnified view on the computer screen and read aloud. Other technological advances include software programs that convert printed text into Braille which can be printed on paper or displayed on a portable keyboard, voice recognition software that allows users to speak into a microphone and have the dictated words typed into word processing, telephone voice-controlled Internet access, and talking PDAs (personal data assistants) (Crandell, Jr. & Robinson, 2007).

## **Conclusion**

Through careful interviewing and assessment, counselors enhance the rehabilitation potential of clients who have blindness or low vision. Counselors can enable their clients to choose realistic, attainable goals through the provision of rehabilitation services in a holistic manner. A variety of resources are available within the community. As noted by Panek (2002), these include Braille foundations, service organizations (e.g., Lions' Clubs), centers for the partially sighted, college programs and services, and public school programs which have specialists with knowledge of community services available for persons with visual impairments.

## **Case Study**

Ms. Alexa Bitari is a 51-year old Caucasian female diagnosed with (dry) macular degeneration. She has never been married and lives alone. Best corrected vision is reported as 20/40 (right eye) and 20/300 (left eye). Her central vision is compromised in both eyes due to macular degeneration. Ms. Bitari reports that her vision loss interferes with personal grooming and managing her finances, causing difficulties performing essential functions of her job. To be able to read, Alexa needs bright light, large print, and 3.5 magnification. Ms. Bitari reports no hospitalizations, takes no medication, and denies any history of emotional problems or substance abuse.

Ms. Bitari graduated from high school and earned a Bachelor's Degree in Communications. Subsequently, she obtained clerical skills training at a local business college and took classes related to advancement in the entertainment field. For the past 25 years within the entertainment industry, Alexa worked for a large firm as an administrative secretary. This position in the corporate graphics design department was eliminated due to downsizing; thus, Ms. Bitari is currently working within the clerical pool and will have to interview for a new position to remain with the employer. Despite ADA legislation, attitudes prevail throughout the entertainment industry that very few jobs can be performed by someone who is visually impaired. Ms. Bitari recognizes that the entertainment industry is populated by mostly young workers, and is concerned that she will be viewed as "washed up" if she discloses that she is coping with vision loss due to macular degeneration.

This client believes she will not be retained by the company in a permanent position because of her disability, despite her long tenure with the employer and excellent clerical skills. She enjoys the entertainment industry, especially the challenging, fast-paced, and at times, glamorous working environment. Because she has trouble using the computer and reading file labels, Ms. Bitari fears that her request for accommodation will be denied. Alexa is worried that she will begin making errors on the job if her eye condition worsens. Although she is still able to drive, Ms. Bitari realizes that she may need to consider alternatives as her vision deteriorates.

In her most recent position as an administrative secretary, Ms. Bitari was responsible for typing purchase orders and correspondence, answering telephones and making appointments, coordinating meetings, and performing other staff responsibilities. An administrative secretary lifts and carries a maximum of ten pounds, with frequent lifting of less than five pounds. Most of the work is conducted while



sitting. It takes approximately one year to acquire the skills needed for this position. Ms. Bitari has consulted with a rehabilitation counselor regarding the requirements and potential reasonable accommodation that will be needed for retaining a position with her current employer.

### Questions

1. Discuss possible limitations, both physical and psychosocial, for individuals with low vision and those with blindness.
2. Describe this client's job as an administrative secretary in terms of physical exertion and skill levels. Provide a vocational profile.
3. As Ms. Bitari's rehabilitation counselor, describe the steps you would take to identify appropriate rehabilitation technology services.
4. Given the size of Alexa's employer, discuss how you would propose she be accommodated through assistive technology.
5. If Ms. Bitari is unable to continue working for her present employer, what are the alternatives you will recommend as her counselor? Include short-term and long-term goals.

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## Chapter 13

# CHRONIC PAIN MANAGEMENT

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## Introduction

Pain management represents one of the most difficult healthcare issues facing the medical profession and the general population. Approximately 30% of the population in the United States experience chronic pain (Bonica, 1990). Over 75 million Americans live with disabling chronic pain; the annual expenditure is over \$125 billion dollars for hospital and medical treatment. About 50 million Americans are either partially or totally disabled by chronic pain which can last for weeks, months, or sometimes years. Twenty-six million adults yearly report back pain (mild, moderate, or severe); headaches are more prevalent than back pain, and the prevalence of undiagnosed pain (pain in several areas of the body) is reported by about 12% of the population (Block, Kremer, & Kremer, 2005). It has been ranked as the third greatest healthcare problem in this country, following heart disease and cancer (Banks & Mackrodt, 2005; Smith, 2007). Pain, acute or chronic, is associated with a variety of conditions and can affect any area of the body.

Though advances have been made in pain management, the above trend seems to be escalating. The rate of low back pain disability has increased by 14-fold. Frustration commonly results within the healthcare professions. An extensive problem confronts us with no easy answer forthcoming from current research.

What is needed is an acknowledgment of the complex nature of chronic pain. Learning the art involved in its successful management is the goal. This chapter summarizes the theoretical and practical aspects of treating chronic pain. Emphasis is placed on the multifactorial nature of pain and the need to treat each patient experiencing pain as an individual within a biopsychosocial environment.

The following topics are discussed in this chapter: pain theory, medical evaluation of chronic pain disorders, psychological assessment, medical management of chronic pain, vocational aspects and rehabilitation, and a case study of an individual with chronic pain.

## Pain Theory

Aristotle classified pain as an intensification of affect, Epictetus understood pain as a cognitive experience, and Descartes postulated that pain is a purely physiological process. Descartes' "specificity" theory was refined by the "summation" theory, which suggests that pain is produced by the sum of intense stimulation of nonspecific receptors. Central summation theory suggests that feedback loops become established following an injury, leading to development of chronic pain. Current theory integrates these historical tenets of pain and attributes pain to a "psychophysiological experience" (Miller & Kraus, 1990). The experience of both acute and chronic pain is viewed as being regulated by the interpretation of the sensation. Pain and injury are seen as having a variable rather than linear link (Banks & Mackrodt, 2005).

### *Definition of Pain*

Each individual defines pain differently. A widely accepted definition given by the International Association for the Study of Pain (IASP) (1986) is as follows: "Pain is an unpleasant sensory and emotional experience associated with actual or potential tissue damage, or described in terms of such damage. Pain is always subjective" (p. S217). This definition includes easily explained pain associated with known tissue trauma or active disease. Also, it encompasses more incompletely understood pain disorders, such as phantom-limb pain, central pain (e.g., post-stroke pain), atypical facial neuralgias (pain not corresponding

to a specific nerve disorder), fibromyalgia (non-specific aching pain, tenderness, and stiffness around joints and soft tissue), and enthesopathies (pain at the site of a tendon or muscle attachment). The IASP definition includes pain experienced in mental disorders that have no known noxious somatic source of the pain. Examples are pain as a symptom of mood disorders (depression and unresolved grief reactions), conversion disorders (e.g., hysterical neurosis) and delusional states (Pinsky, 1993).

Healthcare providers and insurance companies often incorrectly assume that pain is purely objective. Subjectivity of pain occurs during the processing of pain signals that enter the brain and are mediated by the physical trauma of an injury and a host of psychosocial factors, such as past pain episodes and emotional responses to injury and pain.

Differentiating between acute and chronic pain is required for the management of pain. Other than diagnosis, distinguishing between acute and chronic pain may be the most important factor influencing a treating clinician's options. Most researchers define acute pain as pain that lasts for less than six months (short-term pain), and chronic pain as pain that goes on for longer than six months (long-standing pain) (Banks & Mackrodt, 2005; Smith, 2007). The terms, acute and chronic refer to the period of time the pain has lasted, not the severity of the problem or intensity of the pain (Pinsky, 1993).

Chronic pain is complex, debilitating, and influenced by psychosocial factors. Rehabilitation often requires a team of healthcare providers as well as careful development and follow through of long-term strategies. Return to work frequently is impaired; emotional, social, and financial issues impede treatment interventions. Focus of care differs for patients experiencing acute versus chronic pain; treatment options for acute and chronic pain have significantly distinct interventions. Acute pain is typically managed with rest, immobilization of the particular joint(s), medication to reduce inflammation or provide analgesia, physical therapy, massage, injections, surgery, exercise, and other health strategies. Interventions are oriented towards resolution or return to baseline health and functioning.

Goals of acute and chronic pain treatment vary and consist of different interventions. While acute pain treatment strives for a cure, chronic pain treatment focuses on a longer-term solution aimed at diminishing pain while teaching the person techniques that focus on how to actively manage symptoms of pain. Interventions such as injections, adjunctive medications, implanted devices, cognitive and behavioral techniques, relaxation strategies, and exercise are some common methods used. Along with striving to lessen pain through procedural applications, patient needs have a more active role in managing pain. The healthcare provider reorients the individual to achieve quality of life despite pain, while pursuing long-term strategies for pain reduction. Unfortunately, chronic pain often becomes a central issue in the person's life. Individuals with chronic pain frequently have observable alterations in their behaviors, anxiety, depression, anger, and excessive use of medications and medical services.

## **Medical Evaluation of Chronic Pain Disorders**

### ***Assessment/Evaluation***

Assessment of chronic pain requires a comprehensive and multidisciplinary approach (Smith, 2007). Due to the complexity of chronic pain, the expertise required commonly spans multiple medical specialties and is guided by physicians whose sole practice involves management of chronic pain. Given that all treatment decisions are based on the diagnosis, no other factor carries more importance. History of an illness commonly gives the most clues as to the cause of pain. Specifying the location, frequency, severity, and radiation of pain forms the foundation of most medical assessments. How the pain began, what aggravates it, and what reduces it adds information to the diagnosis. The treating physician investigates coexisting diseases or symptoms that appear unrelated but, on further evaluation, point to previously undiscovered pathology (Clark & Treisman, 2004).

### ***Examples of Common Chronic Pain Disorders***

#### ***Complex Regional Pain Syndrome Type 1 And 2 (Reflexive Sympathetic Dystrophy)***

This syndrome has had many names including causalgia, shoulder-hand syndrome, and most often, reflexive sympathetic dystrophy. The following findings are commonly seen: pain and swelling in an

extremity, abnormal sweating patterns, temperature changes, hypersensitivity to touch and cold, and skin changes. Trauma to an extremity usually precedes the onset of symptoms. Treatment outcome improves with early recognition and an aggressive approach toward treating the sympathetic nervous system abnormality.

### ***Fibromyalgia Syndrome***

This disorder is marked by a presentation of chronic, diffuse muscular pain. Diagnosis is confirmed by the finding of specific, localized tender points. Insomnia, headaches, and irritable bowel syndrome are commonly associated problems. Fibromyalgia is resistant to a multitude of treatment modalities. It is a chronic condition in which minimal improvement is typical. People who have this condition need to remain as active as possible and avoid activities that tend to exacerbate symptoms.

### ***Arthropathies***

Arthritic conditions such as osteoarthritis and rheumatic arthritis are common chronic pain conditions. Other arthropathies include systemic lupus erythematosus and ankylosing spondylitis. Multiple joint involvement is typical, although one joint may be involved with a chronic, progressive course. Diagnostic tests are beneficial for categorizing the arthritis and enhancing treatment options. With family support and encouragement, depression tends to diminish.

### ***Phantom Limb Pain***

Typically following an amputation, a person has phantom limb sensation, a feeling that the amputated limb is still present. This is normal and diminishes over time. Phantom limb pain, however, occurs in some individuals and can involve moderate to severe chronic pain in the area where the limb had been present. The pain can reach a point where it becomes disabling. Medical treatment includes nerve blocks, surgical removal of neuromas, and chronic pain management (Clark & Treisman, 2004; Falvo, 2009).

### ***Neuropathies***

This general category of disorders often leads to chronic pain. Polyneuropathies, such as diabetic peripheral neuropathy, cause severe distal lower extremity pain that is resistant to treatment. A single traumatic nerve injury can be disabling if chronic extremity pain of high intensity becomes resistant to treatment. Carpal tunnel syndrome and cubital tunnel syndrome are neuropathies that are commonly the result of cumulative traumas.

### ***Lumbar Spine Disorders***

Several conditions have the primary symptom of chronic low back pain. Spinal stenosis, failed back syndrome, and chronic musculoligamentous strain are common examples. Successful formulation of a treatment plan and education of the patient with chronic low back pain necessitates a specific diagnosis. Diagnoses such as “chronic low back pain” or “lumbago” are confusing to patients and healthcare providers alike. Vague diagnoses promote inaccurate treatment approaches for a broad range of these disorders (Polatin, Kinney, Gatchel, Lillo, & Mayer, 1993).

### ***Headaches***

Headache is a common presenting pain complaint with a multitude of causes and frequent chronicity. The headache is frequently primary (as with migraine headaches); yet, it may be a secondary symptom of a cervical spine disorder. Treatment of neck disorders frequently resolves secondary headaches.

## **Psychosocial Assessment**

In preparation for treatment of acute and chronic pain, the clinician assesses the complete physical and psychological status of the individual. The skilled clinician relies on a multidisciplinary assessment and the most effective treatment strategies for the particular situation (Clark & Treisman, 2004; Smith, 2007). A three-part evaluation process begins with classification of the pain experience, followed by patient feedback, and concluding with development of treatment goals.

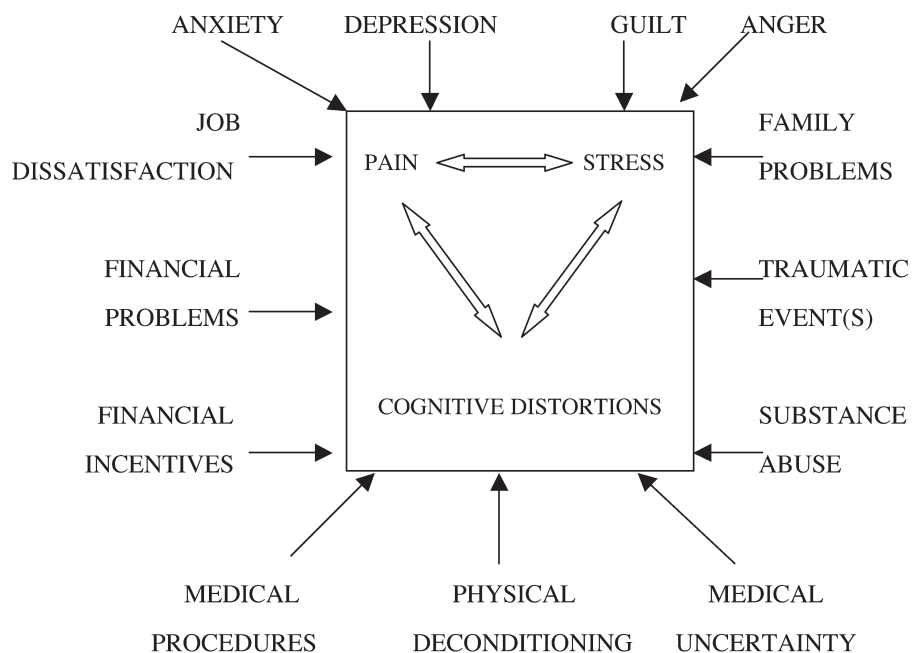
The focus of assessment is on the individual's current and historical reports of pain. Adequate understanding of the pain complaints includes an assessment of the duration (acute vs. chronic), location, cause, frequency, and intensity of pain. The McGill Pain Questionnaire (Melzack, 1975) is still the most widely used instrument addressing these factors. However, the resulting data lacks coherence without a detailed psychosocial history that places the pain complaints in the appropriate context. The following should be addressed during chronic pain evaluations (Getchel & Turk, 1996; Melzack):

1. Orientation and neurocognitive status.
2. Depression and the potential for suicide.
3. Anxiety and post-traumatic stress disorder.
4. Substance abuse.
5. Pain behaviors (assess systemic reinforcers and punishers).
6. Status of litigation.
7. Beliefs about pain.
8. Consequences of pain reduction.
9. Expectations of treatment.

Regardless of the specific syndrome, chronic pain is recognized as a biopsychosocial disorder. According to Block et al. (2005), "there is now fairly strong literature demonstrating that, at least for some medical interventions, psychosocial variables are more potent indicators of treatment outcome than medical variables" (p. 219). Biopsychosocial assessment is used to identify sources of feedback within the family, social interactions, and at work. As patients continue to experience chronic pain, they develop a constellation of beliefs. These attitudes become the psychological modulators of pain. Cognition and perceptions interact with affective, behavioral, and environmental factors in the interpretation of pain. As depicted in Figure 1, pain, stress, and their cognitive interpretation create affective, social, occupational, and physical disruption.

Given the interaction of biopsychosocial factors, it is easy to accept that 90% of individuals enrolled in chronic pain programs have Axis I disorders (Gatchel & Turk, 1996), while 40% have Axis II disorders.

**Figure 1**  
**The Cycle of Pain and Influences**





Axis I and Axis II disorders refer to the classification of mental illnesses as outlined in the *Diagnostic and Statistical Manual of Mental Disorders (DSM IV–Text Revision)*, the official classification of mental illnesses approved by the American Psychiatric Association (2000). Polatin et al. (1993) found that the majority of chronic pain patients had psychopathology prior to their current pain complaints. Seventy-seven percent of a sample of chronic low back pain patients had premorbid Axis I or II disorders, and 59% demonstrated current symptomatology for at least one diagnosis. Of those with lifetime disorders, 54% with major depression, 95% with anxiety disorders, and 94% with substance abuse experienced these disorders prior to onset of pain. These results suggest that psychopathology, or the precursors to psychopathology, predispose a person to chronic pain. Conversely, the co-morbidity of pain with psychopathology exacerbates already existing psychopathology.

## **Medical Management of Chronic Pain**

Treatment of chronic pain can be complicated because various attempts to control the pain may have been ineffective and the person may have become addicted to prior medications used in attempts to control the pain (Foley, 2003). There are many treatment options available. This overview of chronic pain treatment discusses those treatments that are most successful. Throughout this section, emphasis is on the need for a multidisciplinary team that blends a variety of treatments for each patient.

### ***Treatment Modalities***

#### ***Physical Therapy Modalities***

Therapists utilize a variety of physical therapy modalities as adjuncts to overall treatment plans. Application of heat and cold, electrical stimulation, and ultrasound relax underlying tissues; this assists injured tissues to become more amenable to other treatments. Physical therapy is considered the first approach during the acute phase of pain. If pain persists despite several weeks of physical therapy, further use of these modalities are prescribed for the person to administer at home. Individuals experiencing chronic pain commonly use heat or cold to alleviate symptoms of pain at critical times of the day. Electrical stimulation units (TENS) are used to treat chronic pain. By carrying a small battery-operated unit attached to a belt, this portable device can provide effective pain relief at the job site or while at home.

#### ***Manual Techniques***

Massage, manipulation, or manual adjustments may relieve common ailments that cause both acute and chronic pain. Myofascial pain syndrome benefits from specific massage techniques, which can be curative. During the acute phase of pain, these techniques attempt to correct the underlying problem. For chronic pain, symptom relief is more commonly the goal.

#### ***Medications***

Medication is the primary treatment of choice of many clinicians. Medications are used to treat the underlying condition and alleviate pain. Principles of pain management include strategic use of a few medications, conservative dosages, and frequent review of the necessity for continuing medications (Smith, 2007). The following is a review of the major medication classes used for chronic pain management.

#### ***Anti-inflammatory Medication***

Reducing inflammation is the goal of many pain treatment modalities. When injury occurs, inflammatory cells respond, causing pain. Medications to treat inflammation hopefully heal the tissues while concurrently alleviating pain. Nonsteroidal anti-inflammatory medications (NSAIDs) comprise the majority of pain relieving medications. They serve two roles: as a pain reliever and an anti-inflammatory agent.

#### ***Narcotic Analgesic Medications***

Narcotics are one of the most prescribed medications for back pain (Foley, 2003). Narcotic analgesics are divided into two categories: opiates (alkaloids found in opium like morphine) and opioids (fully synthetic and semi-synthetic derivatives of opiates like oxycodone and hydrocodone). They are used for acute pain, post-surgical pain, and pain related to malignancy. Controversy exists concerning the use of

long-term narcotic analgesics. Tolerance occurs over time and the patient requires additional medication for similar analgesic effects. Consequently, the duration of chronic pain corresponds with increased narcotic dosage and risk of side effects. Side effects of dependence include physical as well as cognitive deficits. Tapering of medications is required to change or terminate dosage.

### ***Non-narcotic Analgesic Medications***

To prevent drug dependence on narcotic medications, non-narcotic analgesic medications can be used for chronic pain management. The most commonly used non-narcotic analgesic is acetaminophen, which is sold without a prescription as acetaminophen or Tylenol. The daily maximum dose of acetaminophen is 4000 mg. Taking more than the maximum allowable dose or taking it for long duration can increase the risk of liver damage.

### ***Muscle Relaxants***

These medications are used during acute pain when muscle contractions are the predominant symptom. Side effects make long-term use difficult, especially cognitive deficits; therefore, muscle relaxants are frequently avoided for chronic pain management.

### ***Antidepressant Medication***

The nature of the chronic pain cycle promotes development of depression. Antidepressants neurochemically alleviate depression, directly relieve pain, and improve sleep (Clark & Treisman, 2004).

### ***Injections***

A multitude of injected medications exist for pain relief. In general, injected medications are provided for acute pain or as an initial component of a comprehensive chronic pain management program. Repetitive injections over an extensive time are not recommended. Chronic pain programs use injections strategically as an adjunct within a comprehensive program.

### ***Implanted Devices***

Morphine pumps and intraspinal drug infusion therapy are used for chronic spine pain that has been resistant to other treatment modalities (Banks & Mackrodt, 2005). An implanted catheter releases small amounts of morphine or other medications into the spine, allowing pain relief with small doses. The pumps do not cause the side effects observed with oral opioid administration. Spinal cord stimulators are used for chronic pain, including pain in the extremities, secondary to radiculopathy. Spinal stimulators are considered only when all other methods have been unsuccessful (Bonica, 1990).

### ***Exercise***

The central aspect of treatment is exercise. Strength, flexibility, and aerobic training are integrated and taught by an experienced physical therapist. The chronic pain cycle commonly deconditions a person, secondary to the sedentary lifestyle of bed rest and medications for the treatment of minor exacerbations. Pain increases as muscles atrophy, flexibility is lost, and endurance diminishes. Exercise is a method used to prevent this unnecessary slide into a deconditioned state of health (Smith, 2007). The act of exercise, however, is often difficult for the individual with chronic pain. Chronic pain programs assist with development of a comprehensive exercise routine.

## ***Biopsychosocial Treatment Management***

As noted previously, treatment choice is dependent upon diagnosis, biopsychosocial milieu of the individual, and treatment goals developed with the person's input. Pain interventions should be multidisciplinary and close to the time of injury (Smith, 2007). Whether acute or chronic, the integration of psychological approaches to pain management provides impressive gains even when the pain complaints are responsive to medications. The most effective chronic pain management programs provide medical intervention, physical therapy, occupational therapy, psychological treatment, and vocational therapy (Clark & Treisman, 2004; McCarberg & Passik, 2005). Treatment of chronic pain syndromes relies on two related modalities: cognitive-behavioral techniques and self-regulation.

### ***Operant Conditioning***

Many current psychological interventions in pain management are based on the conceptual understanding of operant pain (Fordyce et al., 1973). As “respondent pain” leads to altered accommodative behavior, expression of pain is shaped by behavioral reinforcement. Operant intervention occurs on two levels. Behavior is extinguished by withholding reinforcement, including switching to fixed medication intervals and eliminating social attention for the pain. Concurrently, “well behaviors” are reinforced and attention is given for behaviors that are incompatible with pain.

### ***Cognitive-Behavior Techniques***

Cognitive-behavioral techniques represent the development of a set of assumptions about behavior, thoughts, and emotions within a physiological and social environment. Patients with chronic pain usually have developed irrational interpretations of their environment. They often fear that steps toward progress will lead to reinjury, worsening pain, failure, and loss of financial support. While behaviors of chronic pain patients shape their environment, the reciprocal is also true; environment shapes behaviors. Thoughts, behaviors, emotions, and physiology affect one another. Changing thoughts and behaviors is generally easier than changing emotions. Four goals of cognitive-behavioral techniques are the following (Smith, 2007; Turk, Meichenbaum, & Genest, 1983):

1. **Act rather than react.** Assist patients to perceive problems as manageable by developing problem-solving skills instead of being consumed by emotional reactions.
2. **Monitor.** Increase self-monitoring of thoughts, emotions, and behaviors in response to changes in physiology or the environment.
3. **Just do it.** Change behaviors regardless of feelings or faith in the response.
4. **Develop flexibility.** Maintain and develop effective coping strategies and continually adapt them.

To accomplish these goals with the chronic pain patient, many specific cognitive-behavioral techniques have been adapted and developed. Cognitive restructuring and self-regulation techniques are part of a comprehensive treatment plan for individuals who have chronic pain.

### ***Self-Regulation Strategies***

Current development of self-regulation strategies is diverse and multimodal. They include any exercise aimed at teaching patients to regulate their physiological states and autonomic responses to the environment. Several forms of relaxation are used. Active strategies for regulation include relaxation, biofeedback, hypnosis, and guided imagery.

### ***Relaxation***

As with other psychological methods of pain management, self-regulation strategies (e.g., relaxation and biofeedback) are based upon operant conditioning theory. Relaxation exercises vary from simple deep breathing exercises to progressive muscle relaxation. These techniques, along with biofeedback or hypnotic induction, often form the foundation of treatment. Progressive muscle relaxation increases awareness of the level of tension or relaxation.

### ***Biofeedback***

Efficacy of biofeedback is dependent upon the person’s commitment to treatment, concomitant treatments, and the specific type of chronic pain disorder. While tension, cluster, and migraine headaches are treated effectively with biofeedback, other chronic pain syndromes need a multidimensional treatment approach (Gatchel & Turk, 1996).

### ***Hypnosis***

By far the most fascinating self-regulation technique is hypnotic analgesia. When considering hypnosis, it is important to assess the person’s attitudes and expectations. The clinician should inquire specifically about preconceived beliefs concerning the nature and practice of hypnosis. Direct induction procedure includes (a) eliciting the patient’s full cooperation, (b) focusing attention on the clinician’s voice, (c) turning attention inward, and (d) suggesting dissociation. By suggesting the client dissociate from environmental stimuli, the exercise proceeds from relaxation to hypnosis. Once dissociation has been

established, therapeutic suggestions are offered for a feeling or belief that is incompatible with present destructive thoughts or the pain experience (Barber, 1996).

### ***Guided Imagery***

Guided imagery incorporates relaxation techniques with either clinician-directed or self-directed “Ericksonian” suggestions (Araoz, 1985). It is an internally mediated sensory (typically visual) experience of focused attention (Burte & Acer, 1997). Association of pain and relaxation to positive images promotes control over an abstract concept along with dissociation from the experience of pain.

Essential components of guided imagery include (Barber, 1996; Burte & Acer, 1997): assessment and observation of the person’s behaviors and images; focusing the individual on his or her experience; transforming the experience; assessing the levels of relaxation, pain, and anxiety; and providing positive affirmation (Araoz, 1985).

### ***Patient Involvement in Treatment***

One of the most challenging aspects of pain management is patient compliance and involvement in treatment. From a psychological perspective, the problem is magnified. Prior to onset of psychological treatment, the patient typically has undergone multiple medical assessments and procedures. Some of these procedures are highly technical and invasive. Faced with the perceived relative simplicity of psychological techniques, the patient is often skeptical about the efficacy of treatment (Gordon & Dahl, 2004; Turk et al., 1983).

With the burgeoning emphasis on medically-based pain management, patients may ascribe the following to psychological techniques. They may believe that therapists are discounting medical procedures, suggesting that the pain is not real, but rather “all in your head.” Given this, even the individual who participates in treatment has difficulty succeeding with psychological techniques. Success indirectly confirms this belief, while failure provides assurance to the patient that the pain is real.

While enhancing the person’s motivation for treatment is difficult, there is a great imperative for patient involvement in medication, physical rehabilitation, and psychological treatment. Responsibility for treatment is shifted from the professional to the patient; the patient is given the lead despite possible lack of motivation to change.

The professional takes the role of “coach,” encouraging responsibility while assisting the person to develop a sense of efficacy through the use of empowering presentation styles, reframing, role playing, metaphors, paradoxes, and challenging irrational ideation (Turk et al., 1983). If the professional’s desire for improvement is greater than that of the patient, the treatment is likely to fail. When the individual is employed, there is often pressure to return to work by the workers’ compensation insurance company and the employer. Goals established by the clinician may be at variance with those of the patient and with the goals of the insurance company. While one person may wish to return to work quickly, another may not. All individuals need to be encouraged to develop realistic and practical goals to meet their specific needs and interests (Clark & Treisman, 2004).

## **Vocational Aspects and Rehabilitation**

Systemic factors influencing rehabilitation are as complex as they are powerful. Priorities of financially concerned insurance companies are often in conflict with those of patients. For the majority receiving benefits within workers’ compensation, financial concerns of the insurer may be in opposition to the provision of effective vocational rehabilitation services. A main priority of the insurance industry is to save finances by limiting services provided. Limiting care in the short-term reduces costs to the employer, removes non-eligible employees from the system, keeps financial settlements lower, and limits the responsibility of the company, impeding successful outcomes. Two other sources of financial compensation, Social Security and litigation, often reinforce limited success in treatment (Barber, 1996). For people in litigation, medical improvement must become more reinforcing than current disability status.

The employment situation may act in a number of ways, both to maintain pain behavior and as a disincentive to recovery and return to work. Early intervention in vocational rehabilitation increases

motivation of an injured worker. Employer interest and involvement, as well as an understanding of chronic pain, helps facilitate the employee's attempts to return to work. Reasonable accommodations for workers with chronic pain are variable and depend upon individualized assessment; each situation is unique (Brodwin, Parker, & DeLaGarza, 2003). Key factors to success include a motivated worker, an involved employer, and a successful pain management treatment program. By careful and comprehensive evaluation of a person's capabilities and limitations, analysis of the types of worksite accommodations needed, and assessment of potential assistive devices, the rehabilitation counselor can best improve employment opportunities for those who have chronic pain.

Early intervention enhances the motivation of an employee to remain on the job if currently working or to return to work if on disability. Through participation in a chronic pain program, identifying the causes of pain, understanding the side effects of medication, and assessing any pain factors at work, the client can more effectively work with the counselor toward successful rehabilitation. Side effects of medication may require modification of certain work duties; in some cases, a change in medication regimen may alleviate problematic side effects that affect work functions. Employers who comprehend the dynamics of chronic pain are more willing to consider reasonable accommodation and job modification. Accommodations and job modifications are as variable as chronic pain itself; each person needs an individualized approach taking into consideration the various concepts described within this chapter (Brodwin et al., 2003).

## **Conclusion**

The complex nature of chronic pain gives the healthcare practitioner no simple resolutions. Rather, an art form that takes into account all aspects of the individual is necessary. Directed multi-disciplinary treatment programs with a biopsychosocial emphasis are the approaches that have the greatest chances for a successful experience.

Outcome studies demonstrate the need for such an approach. Predicting success commonly requires an understanding of psychosocial aspects more than diagnosis or degree of pathology found. Williams et al. (1998) found that job satisfaction was a strong predictor of pain management success and eventual return to work. Workers who report high levels of dissatisfaction were two and a half times more likely to incur a job-related back injury than employees who reported work satisfaction. Individuals who blame their employers for the accident had poorer treatment outcomes (Block et al., 2005). Becker, Hojsted, Sjogren, and Eriksen (1998) reported that financial issues were a major factor for success in treatment. Though at times we would like to view pain as a purely mechanical problem that we need to "fix," the more successful approach involves widening one's scope to include the multifactorial nature of pain. Through proactive rehabilitation, the counselor can assist the employee to remain on the job, minimize consequences of impairment, and effectively work with the employer to understand the importance of accommodation in diminishing disability and increasing job productivity.

## **Case Study**

Mr. Thomas Malone is 29 years of age, married, with a child three years of age. His wife is a physical therapist, employed full time by a medical clinic. After completing high school, Tom attended a trade school for one year to become a respiratory therapist. During the final year in high school and during trade school, he was employed part-time as an orderly in a local community hospital. At times when the hospital was short staffed, he worked in the pharmacy as a pharmacy clerk, and also in the substance abuse clinic as a counselor aide.

The work of a respiratory therapist requires lifting up to 50 pounds occasionally and repetitive lifting and carrying of 10-20 pounds. It takes one to two years to learn proficiently. The job of orderly requires lifting up to 100 pounds and can be learned in three to six months. A pharmacy clerk lifts and carries small items up to a maximum of 20 pounds. The position takes one to three months to learn. Counselor aide is sedentary work involving brief on-the-job training.

Upon completion of the respiratory therapy training program, Mr. Malone secured employment as a respiratory therapist for the same community hospital. After working for two years, Tom injured his back



while helping lift a patient. For the next nine months, he received comprehensive multidisciplinary treatment first for acute, and then for chronic low back pain.

Presently, Mr. Malone's chronic pain has diminished from moderately severe to moderate. His treating physician restricts him to a maximum of light work activities.

### **Questions**

1. Provide a vocational profile for Mr. Malone, including age category, educational level, and work history; include skill and exertional level for each job.
2. What are the differences between acute and chronic pain?
3. Describe a comprehensive, multidisciplinary treatment program for chronic pain.
4. Explore the psychosocial factors involved in chronic pain and its relevance to this case.
5. How would you approach vocational rehabilitation for this client?
6. Discuss the various treatment modalities used for chronic pain.
7. Develop several vocational rehabilitation plans for Tom Malone.

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## Chapter 14

# COMPLEMENTARY AND ALTERNATIVE MEDICINE

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## Introduction

In 2002, the Medical Subject Headings Section staff of the National Library of Medicine classified alternative medicine under the term complementary therapies: therapeutic practices which are not currently considered an integral part of conventional medical practice or interventions not taught at United States medical schools or not available at U.S. hospitals. Therapies are termed as *complementary* when used in addition to conventional treatments and as *alternative* when used instead of conventional treatment (Cooper, 2004).

The National Center for Complementary and Alternative Medicine (NCCAM) is the Federal Government's lead agency within the United States Department of Health and Human Services for scientific research on Complementary and Alternative Medicine (CAM). As western medicine was confronted with more patients requesting CAM treatments, they needed evidence to support the CAM treatment modalities. The National Institute of Health (NIH) directed funding for this purpose and a panel was formed that defined CAM as "a broad domain of healing resources that encompasses all health systems, modalities, and practices and their accompanying theories and beliefs, other than those intrinsic to the politically dominant health system of a particular society or culture in a given historical period." Most CAM practices are guided by four common principles: prevention, natural healing, active learning, and holistic approaches (Mayo Clinic, 2007).

CAM includes, but is not limited to, the following disciplines: relaxation therapy, meditation, biofeedback, hypnosis, imagery, chiropractic, acupuncture, homeopathy, massage therapy, and aromatherapy (Cooper, 2004; Mayo Clinic, 2007). CAM is still a rather new concept in the United States; toward the end of the twentieth century, alternative therapies were reintroduced. According to Dr. Stanley Krippner, "The field of complementary and alternative medicine is past its infancy but has yet to enter adulthood. The field is in its adolescence and as with any promising but rambunctious teenager, it requires a strong hand to provide guidance" (Freeman, 2009). The concepts supporting CAM were first proposed by Chinese philosophers 3,000 years ago; *Huangdi Neijing*, an ancient medical text, has been used for two millenniums (Fein, 2009).

The goal of these treatment practices is to understand the potential effects of stress and conditioning on health outcomes and what can be done to alleviate the negative effects by addressing the mind, body, and spirit of a person as a whole, not by merely controlling the disease. Ader (1996) identified clearly delineated lines of evidence that demonstrate communication pathways exist between the central nervous system (CNS) and other body systems. Studies suggest CAM are sought by individuals that have acquired immunodeficiency syndromes, arthritis, cancer, back pain, and other medical conditions (Wessel, 2008).

It is not possible in one chapter to capture all the pertinent information on the total spectrum of Complementary and Alternative Medicine. Therefore, we have chosen the most commonly used of these therapies in the United States: Traditional Chinese Medicine-Acupuncture, Chiropractic Medicine, Biofeedback, and Homeopathy.

## **Traditional Chinese Medicine – Acupuncture**

Traditional Chinese Medicine (TCM) is one of the most ancient forms of medicine and embodies fine achievements accumulated over the millennia during the course of medical practice of Chinese physicians. This ancient form of medicine was introduced to the United States in the early 1970s after President Nixon visited China. In the last few decades, it has become an acceptable alternative and supports rehabilitation efforts (Normile, 2003). TCM includes acupuncture, Tuina manipulation, moxa, cupping, and other practices (Lao, Hamilton, Fu, & Berman, 2003). In this chapter, the focus is on the practice of acupuncture.

### ***Overview and History***

In 600 BCE, TCM doctors recognized that trauma could cause displacement and dislocation. Therefore, bone setting techniques were used which formed a system along with palpation, observation, and measurement. Hua Tou, a renowned medical expert in the Eastern Han Dynasty, created herbs used for anesthetic effects and performed orthopedic surgery. In the years 265 to 341 BCE, TCM practitioners used wood splints to stabilize fractures and trauma of the extremities to avoid surgery. With use of a splint, the patient could perform the extremity's function during the healing process which allowed the treatment and rehabilitation to occur simultaneously. During 112 to 206 BCE, functional exercises and rehabilitation were widely used in the therapy of trauma, bone, and joint diseases. Physicians asked patients to do exercises like other animals, such as the monkey, bear, bird, and rhino called the "five animal exercises." These exercises are used today after surgery to assist in the healing process (Wax & White, 2000).

TCM practices a complete system between treatment and rehabilitation (Beijing College of Traditional Chinese Medicine, 1990). The system is based on the theory of Qi and blood, Yin and Yang, internal organs, as well as the five elements and meridians in the body. In the 5th century BCE, the "Qi" concept was introduced in TCM. Body movement is the key for Qi circulation. If the body is injured and not able to move, the Qi will stagnate and cause problems. In 475 to 221 BCE, the first TCM book was written describing Qi as consisting of blood, brain, spinal cord, joints, and muscles.

When Qi or blood circulation is interrupted, injury or disease occurs. To protect the body with proper Qi and blood movement, TCM emphasizes prevention with acupuncture, herbs, and exercises. Generally speaking, the word Qi connotes both substance and function, two different but closely related concepts which cannot be entirely separated. Each function must be based on a certain substance, while a certain form of substance is bound to demonstrate a corresponding function (Beijing College of Traditional Chinese Medicine, 1990).

### ***Patterns of Use***

The theory of yin-yang is that every object or phenomenon in the universe consists of two opposite aspects: yin and yang, which are at once in conflict and in interdependence; further, this relationship between yin and yang is the universal law of the material world, the principle and source of existence, and the root cause for the flourishing and perishing of living things. These relationships between yin and yang are extensively used in TCM to explain the physiology and pathology of the human body and serve as a guide for diagnosis and treatment in clinical work (GU, 1989; Gu, 1994).

The theory of the five elements indicates that wood, fire, earth, metal, and water are basic materials constituting the material world. There exists among them an interdependence and inter-restraint which determines their state of constant motion and change. The theory of the five elements explains the inter-promoting, inter-acting, over-acting, and counter-acting relationship of yin-yang. Using the theory of the five elements and yin-yang, TCM classifies into different categories: natural phenomena, tissues and organs of the human body, and human emotions (GU, 1989; Gu, 1994).

The internal organs connect in the body through channels and collaterals with the various tissues and organs of the superficial portion of the body to create organic integrity (Teppone & Avakyan, 2007). In the network of channels and collaterals, the channels are the main trunks which pertain to the respective organs; the collaterals are their minor branches which are distributed throughout the body. Along each channel are points for applying acupuncture. The efficacy of acupuncture in prevention and treatment of



disease is due to regulating and strengthening the defensive function and helping restore relative balance within the human body as well as that between the body and its environment.

Seven emotional factors are classified in TCM: joy, anger, melancholy, meditation, grief, fear, and fright (Ots, 1990). These factors are reflections of “man’s” mental state as induced by various stimulations in the environment. They are physiological phenomena that do not cause disease under normal conditions. However, if the emotions are very intense and persistent or the individual is hypersensitive to the stimulations, they may result in drastic and long-standing change in emotion which leads to disease. Traumatic injuries can cause stagnant blood or phlegm to induce pain or deformity (Chen & Chen, 2009; Xinnong & Deng, 2004).

Inspection, auscultation and olfaction, inquiring, and palpation are known as the four diagnostic methods in TCM. Inspection includes expression, color, and appearance of the tongue. Auscultation and olfaction are listening and smelling. Inquiring is asking the patient about the disease condition to understand the pathological process. Palpation is a unique diagnostic tool in TCM because it involves feeling the pulse. The radial pulse represents different internal organs. The palpation of pulse means not only the speed, but also the quality such as thready, wiry, slippery, and so forth. (Gu, 1989; Zhiya, 2002).

Identification of a syndrome entails making further analysis and synthesis of the clinical data obtained by applying the four diagnostic methods to determine the stage at which the disease has developed, its location, and the degree of opposition between the body’s resistance and environmental factors. The differentiation of diseases can be derived from eight principles: exterior and interior, cold and heat, deficiency and excess, and yin and yang (Langevin, et al., 2004). Disorders of channels may affect the corresponding organs, and disorders of the organs are reflected at the corresponding channels. Channels connect with organs and exteriorly with the body surface where points are distributed. Points are thus the specific sites through which the Qi of the organs and channels is transported to the body surface. When the body is affected by a disease, treatment is possible by puncturing the corresponding points on the body surface.

Acupuncture is a method of treating interior diseases from the exterior. Various diseases are cured by the application of the methods of reinforcing the deficiency and reducing the excess to the points of the body surface. The basic principle for acupuncture treatment is to reinforce deficiency and reduce excess syndrome.

Generally speaking, there are many differences between Chinese medicine and conventional medicine. Conventional medicine tries to locate where the problem originates specifically such as organ level (heart, liver, kidney), cell level, or even molecular level. Rather, Chinese medicine focuses on the whole body and tries to balance the body with acupuncture and herbs. Sometimes TCM concentrates on the result and may not seek to understand what happened inside the body. Acupuncture helps induce the body’s own healing power and regain balance (Beijing College of Traditional Chinese Medicine, 1990; Gu, 1989).

Professionals working in the area of acupuncture receive formal training. The intensity and length of training varies including certificate or diploma programs and degree programs – bachelor’s, master’s, and doctorate degrees.

## **Chiropractic**

As with acupuncture practitioners, the goal of chiropractic is to support the natural ability of the body to heal itself. Chiropractic is a health care approach that focuses on the relationship between the body structure, mainly the musculoskeletal system, and its functioning. Although practitioners may use a variety of treatment approaches, they primarily perform manipulation (an ancient healing art) with the goal of correcting alignment. Chiropractors are the second largest group of primary care providers in the United States (Freeman, 2009).

### ***Overview and History***

The term “chiropractic” combines the Greek words *cheir* (hand) and *praxis* (action) to describe a treatment done by hand. Hands-on therapy—especially adjustment of the spine—is central to chiropractic care. No one culture has been identified as the originator of chiropractic. It has been found in the ancient

cultures of China, Japan, Polynesia, India, Egypt, Tibet, and in the Aztec, Inca, Maya, Sioux, and Winnebago cultures of the Americas. Through the 17<sup>th</sup> Century, there are medical records throughout the cultures of Europe detailing the use of traction applied to parts of the body while pressure was applied on a specific area of the spine. Hippocrates, considered to be the father of medicine, was a practitioner of manipulation. Practitioners of bonesetting during the Middle Ages and the Renaissance performed manipulation (Leach, 2004).

Therapeutic manipulation practice was founded in the late 19<sup>th</sup> Century. Three children and their mother died in that conventional medicine was not able to impact their illness of spinal meningitis. Dr. Andrew Still, watching his family die, became convinced that the body needed to heal itself, but had to be structurally sound to accomplish the healing. He decided that healing would take place when the “life force” could be accessed. The life force could be released when the spine was manipulated to relieve mechanical pressure on blood vessels and nerves. If the nerves were impinged in any way, blood flow was reduced resulting in pain, necrosis, and impingement of the healing life force (Freeman, 2009).

While some procedures associated with chiropractic care can be traced back to ancient times, the modern profession of chiropractic was founded by Daniel David Palmer in 1895 in Davenport, Iowa. Palmer, a self-taught healer, believed the body has a natural healing ability. Misalignments of the spine interfere with the flow of energy needed to support health, Palmer theorized, and the key to health is to normalize the function of the nervous system, especially the spinal cord. As with acupuncture providers, chiropractors perform diagnosis by visual inspection and palpation in addition to x-ray and other diagnostic methods.

### *Patterns of Use*

Chiropractic is based on these key concepts: The body has a powerful self-healing ability; the body’s structure (primarily the spine) and its function are closely related, and this relationship affects health; and therapy aims to normalize this relationship between structure and function and assist the body as it heals. Chiropractic has two main groups: “straights”, now the minority, emphasize vitalism, innate intelligence, and spinal adjustments, and consider subluxations to be the leading cause of all disease; “mixers” are more open to mainstream and alternative medical techniques such as exercise, massage, nutritional supplements, and acupuncture (Tetrault, 2004). This treatment modality is the most frequently used of the CAMs. However, many providers of chiropractic treatment do not associate it with CAM, but with western conventional medicine.

The theory of chiropractic is that misaligned spinal vertebrae interfere with nerve function, resulting in altered physiologic conditions which contribute to pain and disease. A common diagnosis to support this theory is subluxation – an impinged nerve as a result of misalignment in the spine, causing pain (Gatterman, 2005). Chiropractic practitioners believe that this misalignment also prevents body defenses from performing appropriately. By adjusting the spinal joints, subluxation is resolved, restoring normal nerve function and optimal health. Chiropractic has expanded to include improving joint mobility and alleviating restricted movement as a result of spinal fixation.

Adjustment procedures are the hallmark and defining technique of chiropractic treatment. Terms associated with chiropractic include: adjustment, manipulation, and mobilization. These terms relate to joint movement. Joint movement or adjustment is divided into active end range (how far the patient can, with muscular effort, move a joint in a particular direction), passive end range (at the end of the active end range, a clinician can move the joint further without injury or pain-physiologic joint space), anatomic end range (movement that results in rupture), and paraphysiologic joint space which exceeds the passive end range but not the anatomic end range. **Manipulation** is passive movement of the joint past the passive end range but not the anatomic end range. **Mobilization** is the clinician-assisted passive movement within the anatomic end range resulting in an increased overall range-of-joint motion. Over 150 chiropractic techniques are currently being used (Freeman, 2009).

The effects of manipulation are in two categories: mechanical and neurologic. Mechanical are defined in terms of the subluxation – spinal joint strain or sprain with local and referred pain and muscle spasm. Neurologic includes both direct and indirect effects on the function of the peripheral nervous system

(PNS) and CNS because of spinal dysfunction. Chiropractic emphasizes that it does not treat disease; rather it promotes the healing of the body by focusing on the body's innate homeostasis to heal itself.

People seek chiropractic care primarily for pain conditions such as back and neck pain, headache, musculoskeletal conditions, menstrual pain, and asthma. A 2002 national survey on CAM use found that about 20% of adults in the United States had received chiropractic care at some point during their lives. Those surveyed reported using chiropractic treatment for the following reasons: combining chiropractic services with conventional medical treatments would help (53%); conventional medicine would not help (40%); chiropractic would be interesting to try (32%); conventional medical professional suggested it (20%); and conventional medical treatments were too expensive (10%) (Ni, Simile, & Hardy, 2002).

During the initial visit, chiropractors take a health history and perform a physical examination, with a special emphasis on the spine. Other examinations or tests such as x-rays may be performed. If chiropractic treatment is considered appropriate, a treatment plan is developed. During follow-up visits, practitioners may perform one or more of the many different types of adjustments used in chiropractic care. Given mainly to the spine, a chiropractic adjustment (sometimes referred to as a manipulation) involves using the hands or a device to apply a controlled, sudden force to a joint, moving it beyond its passive range of motion. The goal is to increase range and quality of motion in the area being treated, and to aid in restoring health. Other hands-on therapies also are used (Ernst, Pittler, & Wider, 2006).

Chiropractors may combine the use of spinal adjustments with several other treatments and approaches, such as: heat and ice, electrical stimulation, rest, rehabilitative exercise, diet, weight loss, and other lifestyle factors, such as dietary supplements.

To practice chiropractic care in the United States, a practitioner must earn a Doctor of Chiropractic (D.C.) degree from a college accredited by the Council on Chiropractic Education (CCE, 2007). CCE is the agency certified by the U.S. Department of Education to accredit chiropractic colleges. Admission to a chiropractic college requires a minimum of 90 semester hour credits (approximately 3 years) of undergraduate study, mostly in the sciences. Chiropractic training is a 4-year academic program that includes both classroom work and direct experience caring for patients. Coursework typically includes instruction in the biomedical sciences, as well as in public health and research methods. Some chiropractors pursue a 2- to 3-year residency for training in specialized fields (Rondberg, 1996).

Chiropractic is regulated individually by each state and the District of Columbia. Board examinations are required for licensing and include a mock patient encounter. Most states require chiropractors to earn annual continuing education credits to maintain their licenses. Chiropractors' scope of practice varies by state in areas such as laboratory tests and diagnostic procedures, the dispensing or selling of dietary supplements, and the use of other CAM therapies, such as acupuncture and homeopathy.

Compared with other CAM therapies, insurance coverage for chiropractic services is extensive. Many HMOs (health maintenance organizations) and private health care plans cover chiropractic treatment, as do all state workers' compensation systems. Chiropractors can bill Medicare, and many states cover chiropractic treatment under Medicaid.

## **Biofeedback**

Biofeedback is a treatment technique in which people are trained to improve their health by using signals from their own bodies. Physical therapists use biofeedback to help stroke survivors regain movement in paralyzed muscles. Psychologists use it to help tense and anxious clients learn to relax. Specialists in different fields use biofeedback to help patients cope with pain. Clinicians rely on biofeedback machines that detect a person's internal bodily functions. This information is then used to gauge and direct the progress of treatment. The biofeedback therapist acts as a coach, helping the patient set goals and limits on what to expect and how to improve performance.

### ***Overview and History***

In the early part of the century in Germany, J. H. Schultz developed a technique called *Autogenic Training* in which verbal instructions are used to guide a person to a different, more relaxed and controlled physiological state. The technique is still used, but it may no longer be distinguished as *Autogenic Training*.

Edmund Jacobson developed the technique of *Progressive Relaxation* training in the 1930s in the United States. This was a series of muscle activities to teach people awareness of tension and relaxation. The effect was to reduce muscle tension and certain causes and effects of stress and other symptoms. In the 1960s and 1970s, there began to be an awareness in the Western world of the Eastern yogic traditions and the ability of some yogis and other masters to alter their physiology volitionally. The altered states that were being achieved by meditative means attracted the attention of a few key researchers. The gurus taught that in this state of relaxation and control they could change a number of variables that were thought to be autonomously regulated: blood pressure, heart rate, finger or hand temperature. These functions are managed by the autonomic nervous system (ANS), so named precisely because it was thought that such functions could not be altered voluntarily (Andrasik & Lords, 2009).

The ANS has two divisions—the *sympathetic* and the *parasympathetic*. The sympathetic nervous system regulates the flight/fight response. The parasympathetic nervous system calms and relaxes and manages body functions. The two work in tandem in a reciprocal relationship. It was Canon and Selye, researchers in the body response to stress, who increased general awareness of the role of stress in physical diseases and mental disorders (Becker, Breedlove, Crews, & McCarthy, 2002).

*Hatha yoga*, and other yogic traditions, became established in the United States as techniques for physical relaxation and enhancement of conscious control over physiology. Additionally, meditation techniques stimulated the elaboration of the relaxation and attention training techniques - influencing and controlling the body with the conscious mind to promote relaxation and manage pain and stress. A number of these techniques have been combined with biofeedback instrumentation to enhance learning physiological self-regulation (mind-body control).

### ***Patterns of Use***

The word “biofeedback” was coined in the late 1960s to describe laboratory procedures then being used to train experimental research subjects to alter brain activity, blood pressure, heart rate, and other bodily functions that normally are not controlled voluntarily. Research has demonstrated that biofeedback can help in the treatment of many diseases and painful conditions. It has shown that we have more control over so-called involuntary bodily function than we once thought possible. It has also shown that nature limits the extent of such control. Scientists are now trying to determine just how much voluntary control a person can exert (Andrasik & Lords, 2009).

At the highest level of organization, each life form may possess an innate biologic field (biofield) which is a complex, dynamic, weak energy field involved in maintaining the integrity of the whole organism – regulating the physiologic and biochemical responses for development, healing, and regeneration (Rubik, 2002). TCM and chiropractic are based on the concept of a vital force or life energy central to healing similar to the concept of biofield, although biofield rests on physical principles that can be measured. Both life force and biofield proponents agree that a form of life-giving energy flows through the body and illness arises as a result of blockages, excesses, or irregularities in this flow. There is no consensus of opinion for a definition of biofield.

The human body emits low-level light, heat, and acoustical energy, and has electrical and magnetic properties. Samples of commonly used measures of the biofield include electrocardiogram (ECG) and electroencephalogram (EEG) to assess the function of the heart and brain. Corresponding magnetic field measurements of the heart and brain have been discovered: magnetocardiogram (MCG) and the magnetoencephalogram (MEG). The MEG allows for localizing the activity of a region of the brain the size of a pea. Lie detectors and biofeedback use galvanic skin response (GSR) to measure the electrical conductance between electrodes on the skin. Thermography measures the emission of infrared radiation from the body. It is an accepted diagnostic procedure in conventional western medicine (Andrasik & Lords, 2009).

Patients are taught a form of relaxation exercise. Some learn to identify the circumstances that trigger their symptoms. They may be taught how to avoid or cope with stressful events. Most are encouraged to change their habits and are trained in special techniques for gaining such self-control. Biofeedback is a tool that reminds physicians that behavior, thoughts, and feelings profoundly influence physical health. It also reminds physicians that they must work together as a team with their patients. Biofeedback generally



refers to using measures derived from peripheral physiology rather than EEG results, measures such as skin temperature, muscle tension, skin conductance, heart rate, and breath. All of these are subject to management of the CNS and refer back to the brain. Neurofeedback and biofeedback both have the goal of improved self-regulation. However, each specialty area in biofeedback has its unique strengths (Evans & Abarbanel, 1999).

A biofeedback therapist is required when the problems that are experienced become greater than a person can handle. Perhaps medical help has been sought and symptoms still exist. Biofeedback becomes an education or a journey toward inner awareness and self-management using the tools of feedback. These may include instrumentation, dialogue with self and a therapist, and practice in various life situations, as well as in the biofeedback therapist's office. It requires dedication and time, and often a change in philosophy. Biofeedback techniques help the individual understand physiology and how to break a habit that may have existed for years before the symptoms appeared.

Despite a strong self-help component, it is prudent to do biofeedback under the care of a trained mental health professional, and only after medical evaluation has determined that a more serious condition will not be neglected by the training. The practitioner guides the trainee by designing an individualized program for improved health, encouraging regular practice, monitoring results, and supporting the training process in various ways, including advice regarding complementary interventions. Initially, the biofeedback therapist will do a history of past health and current symptoms. A psychophysiological profile is conducted with the biofeedback equipment, which may include EMG sensors, a temperature sensor to a fingertip, and a sensor to measure hand sweat. Occasionally, a measure of breathing skills is done with strain gauges. The client may be asked to do a variety of activities during which the monitor displays the physiological responses. These include reclining, sitting in a straight chair, standing, walking, talking of pleasurable things, relating stressful events, discussing symptoms, and relaxing during guided meditation.

Specialists who provide biofield services range from psychiatrists and psychologists to dentists, internists, nurses, and physical therapists. Most rely on many other techniques in addition to biofield practices. The most widely used biofield treatment is biofeedback. The Association for Applied Psychophysiology and Biofeedback (AAPB), founded in 1969, provides information regarding training.

## Homeopathy

Homeopathy (“home-ee-AH-pah-thy”) is derived from the Greek words *homios* (similar) and *pathos* (suffering) and teaches that stimulating the natural healing properties in the body cures disease. The practice of homeopathy is accomplished by introducing a substance into the body inducing symptoms identical to those caused by the disease. This action stimulates healing energy and results in a cure. It is based on the philosophy that “like cures like” (Yuan, & Bieber, 2003). The substances introduced are homeopathic remedies which are dilutions of natural substances from plants, minerals, and animals. These simillimum (most appropriate homeopathic remedy) are prescribed to match an individual patient's illness-symptoms. Homeopathy is an individualized treatment. Once the initial symptoms are resolved, the practitioner treats underlying symptoms (Freeman, 2009).

### *Overview and History*

Hippocrates taught the Law of Similars – like cures like. In the late 1700s, Samuel Hahnemann, a physician, chemist, and linguist in Germany, developed homeopathy as an approach to treating illness. This was at a time when the most common medical treatments were harsh, such as bloodletting, purging, blistering, and the use of sulfur and mercury. At the time, there were few effective medications for treating patients, and knowledge about their effects was limited.

Hahnemann was interested in developing a less-threatening approach to medicine. The first major step reportedly was when he was translating an herbal text and read about a treatment (cinchona bark) used to cure malaria. He took some cinchona bark and observed that, as a healthy person, he developed symptoms that were very similar to malaria symptoms. This led Hahnemann to consider that a substance may create symptoms that it can also relieve. Hahnemann tested single, pure substances on himself and, in more dilute forms, on healthy volunteers. He kept meticulous records of his experiments and participants' responses,



and he combined these observations with information from clinical practice, the known uses of herbs and other medicinal substances, and toxicology, eventually treating the sick and developing homeopathic clinical practice (Yuan & Bieber, 2003).

Hahnemann added two additional elements to homeopathy: “potentization” which holds that systematically diluting a substance with vigorous shaking at each step of dilution, makes the remedy more, not less, effective by extracting the vital essence of the substance. If dilution continues to a point where the substance’s molecules are gone, homeopathy holds that the “memory” of them – that is, the effects they exerted on the surrounding water molecules – may still be therapeutic and treatment should be selected based upon a total picture of an individual and his or her symptoms, not solely upon symptoms of a disease. Homeopaths evaluate not only a person’s physical symptoms but emotions, mental states, lifestyle, nutrition, and other aspects. In homeopathy, different people with the same symptoms may receive different homeopathic remedies (Yuan & Bieber, 2003).

Hans Burch Gram, a Boston-born doctor, studied homeopathy in Europe and introduced it into the United States in 1825. European immigrants trained in homeopathy made the treatment increasingly available in America. In 1835, the first homeopathic medical college was established in Allentown, Pennsylvania. By the turn of the 20th Century, 8% of all American medical practitioners were homeopaths; there were 20 homeopathic medical colleges and more than 100 homeopathic hospitals in the United States (Freeman, 2009).

In the late 19<sup>th</sup> and early 20<sup>th</sup> Centuries, numerous medical advances were made, such as the recognition of the mechanisms of disease, Pasteur’s germ theory, the development of antiseptic techniques, and the discovery of ether anesthesia. In addition, a report (the so-called “Flexner Report”) was released that led to major changes in American medical education. Homeopathy was among the disciplines negatively affected by these developments. Most homeopathic medical schools closed; by the 1930s others had converted to conventional medical schools (Haller, 2005).

In the 1960s, homeopathy’s popularity began to revive in the United States. According to a 1999 survey, over six million people in the United States had used homeopathy in the preceding 12 months. The World Health Organization noted, in 1994, that homeopathy had been integrated into the national health care systems of numerous countries, including Germany, the United Kingdom, India, Pakistan, Sri Lanka, and Mexico. Several schools of practice exist within homeopathy (Jonas, Kaptchuk, & Linde, 2003).

### ***Patterns of Use***

Persons using homeopathy do so to address a range of health concerns, from wellness and prevention to treatment of injuries, disease, and other conditions. Studies have found that many people who seek homeopathic care choose it for help with chronic medical conditions. Many users of homeopathy treat themselves with homeopathic products and do not consult professionals. Typically, in homeopathy, patients have a lengthy first visit, during which the provider takes an in-depth assessment of the patient. This is used to guide the selection of one or more homeopathic remedies. During follow-up visits, patients report how they are responding to the remedy or remedies to help the practitioner make decisions about further treatment (Eisenberg et al., 1998).

A homeopathic remedy is prepared by diluting a substance through a series of steps. Providers refer to the *Homeopathic Pharmacopoeia of the United States* to match symptom profiles of patients with the appropriate remedy. The U. S. Food and Drug Administration (FDA) recognizes homeopathic dilutions as official drugs and regulates the manufacturing, labeling, and dispensing of homeopathic medications. Homeopathy asserts that this process can maintain a substance’s healing properties regardless of how many times it has been diluted. Many homeopathic remedies are so highly diluted that none of the original natural substance remains (Ni, Simile, & Hardy, 2002).

Research has been conducted to address this dilemma. Researchers have found that homeopathic remedies give off electromagnetic signals; each homeopathic substance has a specific dominant frequency (del Giudici & Preparata, 1990). It is theorized that homeopathic remedies convey the electromagnetic message to the body that matches the frequency of the illness, stimulating the healing properties of the body:

one subtle energy (remedy) affects another subtle energy (human energy field). As in other CAMs, this subtle energy field in the body is called the *vital force*.

In the United States, training in homeopathy is offered through diploma programs, certificate programs, short courses, and correspondence courses. Also, homeopathic training is part of medical education in naturopathy, a medical system that originated in Europe which aims to support the ability of the body to heal itself through the use of dietary and lifestyle changes together with CAM therapies such as herbs, massage, and joint manipulation. Most homeopathy in the U.S. is conducted along with another health care practice for which the practitioner is licensed, such as conventional medicine, naturopathy, chiropractic, dentistry, or acupuncture.

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## Chapter 15

# TRAUMATIC BRAIN INJURY

*Stacey Hunter Schwartz, Ph.D.*

## Introduction

Traumatic brain injury (TBI) involves sudden physical damage to the brain. Males ages 14 to 24 years are at the highest risk of TBI, followed by infants and the elderly. Males are twice as likely as females to sustain TBI because of differences in risk exposure and lifestyle. Every year, about 235,000 Americans are hospitalized and 50,000 die as a result of TBI (Centers for Disease Control and Prevention [CDC], 2003). Approximately, 1.1 million Americans are treated and released from emergency rooms. Of these, 99,000 individuals sustain moderate to severe brain injuries resulting in lifelong disabling conditions (Dixon, Lyton, & Shaw, 2005). About 75% of TBIs that occur each year are concussions or other forms of mild injury (CDC, 2003). After one traumatic brain injury, the risk for a second injury is three times greater; after a second TBI, the risk for a third injury is eight times greater (Kraus & McArthur, 1999). CDC (1999) estimates that at least 5.3 million Americans currently have long-term or lifelong need for help to perform activities of daily living as a result of TBI.

A traumatic brain injury produces an injury to one's core sense of self. Persons with TBI undergo such dramatic cognitive and personality changes that they seem, both to themselves and to those closest to them, to be different people than they were pre-injury. Many persons with TBI regard their post-injury experiences as their second lives. At the same time, some have called TBI an "invisible disability" because the physical appearances of many persons with TBI do not change as a result of their head injury (Brown & Vandergoot, 1998). While persons with other disabilities may feel frustrated because they are treated as different, individuals with TBI often feel frustrated because others treat them as if they were the same as before the injury. This disjunction between appearance and functionality complicates adjustment of persons with TBI.

Rehabilitation is particularly challenging for other reasons as well. Counselors usually rely on their clients for information regarding their skills, abilities, and limitations. Like other individuals with new disabilities, people with TBI experience a state of psychological denial about the extent and ramifications of their deficits; additionally, they may have a neurologically induced failure to recognize the deficiencies. They may have become cognitively incapable of knowing their limitations. Others with TBI do not remember what they were like before their injuries, or have an idealized memory of their pre-injury capabilities. Consequently, some people with TBI insist on unrealistic goals for rehabilitation.

Other people are able to set realistic goals, but implementing those goals presents challenges. Deficits in attention or auditory processing interfere with successful educational and vocational planning (Rios, Periañez, & Munoz-Céspedes, 2004). Counseling may be unproductive because the client is unable to distinguish relevant from irrelevant information. Other clients show great insight and understanding while in counseling sessions but forget the content of the sessions soon after each session ends. For these reasons and others, rehabilitation presents unique challenges to rehabilitation counselors.

## Etiology

Brain injuries result from a variety of causes, including vehicle collisions, falls, stabbings, gunshots, infections, tumors, and strokes. Survivors with very different causes of injury may have similar symptoms. While the cognitive strategies taught or the vocational outcomes are similar for survivors of aneurysm or automobile accidents, the two could qualify for different assistance programs. In fact, although the labels

used by those who study or work with persons with TBI relate to the causes of injury, outcomes tend to be similar across causal groups based on severity of injury and location within the brain (Falvo, 2009; Reilly & Bullock, 2005).

Causes of brain injury can be divided into two categories. Traumatic injuries are those caused by something outside the body, such as by the impact of an automobile accident or by the penetration of a bullet. Atraumatic injuries are those that occur because of internal causes, such as a cerebrovascular accident (stroke) or an infection. Consequences of brain injury detailed in this chapter apply to survivors of traumatic as well as atraumatic brain injury.

## **Categories of Traumatic Brain Injury**

The Brain Injury Association (BIA), formerly the National Head Injury Foundation, defines traumatic brain injury as “an insult to the brain, not of degenerative or congenital nature, but caused by an external physical force that may produce a diminished or altered state of consciousness, which results in an impairment of cognitive abilities or physical functioning.” TBIs may be referred to as traumatic head injuries. Vehicle accidents are the leading cause of TBI, accounting for 50% of all injuries. Falls are the second leading cause, accounting for more than 20%. Alcohol is a significant factor in the occurrence of TBI; more than 50% of persons with brain injury were intoxicated at the time of injury (Kraus & McArthur, 1999; Kreutzer, Witol, & Marwitz, 1996).

### ***Closed Head Injury***

In a closed head injury, the skull is not penetrated. Damage usually occurs when the brain experiences rapid acceleration and deceleration because of a blow to the head, such as in a motor vehicle accident or a fall. The brain hits against the skull in an initial impact called the “coup.” Next, the brain stem turns and twists on its axis causing localized or widespread damage to the brain. The brain then rebounds and hits the opposite side of the skull (the “contre coup”). Consequently, in a closed head injury, damage often occurs to diffuse areas of the brain. Swelling and bleeding, problems that frequently occur with open head injuries, also cause injury to various areas of the brain.

Loss of consciousness may result, but in other instances, the survivor remains conscious and there are no outer signs of brain injury. The medical team concentrates on and provides treatment for more obvious wounds. Later on, the person with TBI and those who attempt to offer help may have a difficult time identifying and documenting that an injury occurred.

Severity of injury is a good predictor of eventual outcome, but many people who sustain what are considered mild injuries nonetheless suffer life-altering consequences (Trahan, Pépin, & Hopps, 2006). The consequences of mild TBIs are particularly pronounced among persons whose careers involve high-level cognitive demands. The American College of Rehabilitation (ACR) has developed a definition of such injuries. Although the definition adopts the somewhat understated term “mild” traumatic brain injury, it clearly communicates the idea that these non-coma type injuries are serious with permanent consequences. According to the ACR definition of mild traumatic brain injury, a permanent brain injury can occur under any of the following four conditions (Mild Traumatic Brain Injury Committee, 1993):

1. Any period of loss of consciousness.
2. A loss of memory for events immediately before or after the accident (amnesia).
3. An alteration in mental state at the time of the accident (e.g., feeling dazed, disoriented, or confused).
4. Focal neurological deficit(s).

### ***Open Head Injury***

In an open head injury, verification of TBI is not difficult. The person’s brain matter has been penetrated, such as by stabbing or gunshot. Skull fractures, visible brain matter, and obvious bleeding indicate the trauma. Often, the patient goes into a coma or a physician pharmacologically induces a coma immediately following the trauma. The coma may last minutes, weeks, or months. This type of head injury is more likely to damage a specific area of the brain. Additional risks exist as well; some patients develop infections or need to have their lost skull material replaced with artificial plates (Zink & McQuillan, 2005).

### ***Atraumatic Brain Injury***

A category of brain injury which receives less attention in the literature, but which some experts say is growing, is atraumatic causes of brain injury. Examples include arterial-venous malformations (blood vessels with weaknesses present at birth) and infections in the brain, both of which can result in cerebrovascular accident. In recent years, more people are benefiting from new surgical procedures which help increase survival of people with vascular injuries. Similarly, persons who have brain tumors survive more often today because of advances in oncology. Meningitis (inflammation of the brain and spinal cord) and cardiac arrest leading to anoxia (where the brain has been completely deprived of oxygen for a period of time) or hypoxia (where the brain has been partially deprived of oxygen) also cause serious diffuse brain damage (Thurman, Alverson, Dunn, Guerrero, & Sniezek, 1999).

## **Functional Limitations**

Because the brain is such a crucial organ that performs a wide variety of necessary functions, injury to the brain causes a host of deficits (Johnstone, Schopp, Harper, & Koscuilek, 1999). No one survivor is likely to exhibit all the functional limitations described below. As causal agents, severity of injury and location of injury vary as do resulting cognitive deficits. Even the number of locations of injury and severity at those locations are not necessarily determinative of functional outcome. Impairment of life processes related to deficits also depends on the client's personal lifestyle and career demands (Rios et al., 2004). The description of deficits (below) is helpful in understanding the potential consequences of TBI; each client must be assessed for the particular injuries and resulting deficits. Table 1 summarizes the types of sequelae and deficits that commonly follow TBI.

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**Table 1**  
**Various Categories of Deficits**

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Physical	Balance difficulties, fatigue, pain, hemiparesis, uneven gait, ataxia, apraxia, decreased motor speed, seizure disorders, sensory deficits
Cognitive	Impairments in attention/concentration, memory, visual or auditory perceptual processing, verbal reasoning, critical thinking/logic, language, awareness
Psychosocial	Personality changes, emotional lability, flat affect, depression, substance abuse, frustration intolerance, impulsivity/disinhibition, lack of initiative

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### ***Physical***

Traumatic events like automotive accidents and falls may result in physical impairments (such as immobility caused by bone fractures), which are unrelated to the brain. There also may be physical consequences resulting directly from the brain injury, as described below.

#### ***Balance***

Many people have difficulty with balance and coordination immediately following injury. In some, dizziness and unsteadiness are permanent conditions. Special retraining programs exist for these problems.

#### ***Fatigue***

For several reasons, extreme mental and physical fatigue is common after TBI, as are sleep disorders. Ouellet, Beaulieu-Bonneau, and Morin (2006) found that over half of patients with mild to severe reported insomnia symptoms, with almost 30% fulfilling the diagnostic criteria for an insomnia syndrome, remained untreated in almost 60% of cases. Tasks which were formerly easy may become far more mentally challenging and, consequently, physically exhausting. For example, four hours of concentrated activity may

necessitate several hours of rest. Some persons are poor at judging their physical limitations, and struggle to gradually ease into school and work patterns without taking on too many responsibilities.

### ***Pain***

Individuals with TBI frequently report persistent headaches. Those with concussions often have neck and back pain and require ergonomically correct workstations to reduce pain complaints. There may be increased sensitivity to pain.

### ***Hemiparesis***

Each side of the body is controlled by the opposite side of the brain. Therefore, injury to the motor strip of one hemisphere affects movements on the opposite side of the body. For example, the left side of the body may be paralyzed because the right hemisphere has been injured. Although a leg or arm may not itself be injured, the brain may not be able to control that leg or arm properly, resulting in dysfunctions of movement (Kraus & McArthur, 1999).

### ***Other Disorders of Movement***

Many persons have permanent impairment of movement caused by an inability of the brain to control the extremities. The individual experiences impaired gait, ataxia (jerky movements), apraxia (uncontrolled movements), muscle spasticity, tremors, and impaired small motor control.

### ***Decreased Motor Speed***

Many people feel as though they are moving in slow motion. Their brain signals travel at a slower pace; all tasks take longer. These individuals appear to have a few seconds delay between when they decide to move and when they carry out the planned action.

### ***Seizure Activity (Traumatic Epilepsy)***

Following TBI, recurrent seizures are possible (17% of all cases) (Reilly & Bullock, 2005). Most of those who experience seizures have their first seizure within 24 hours of injury. Usually, seizures are controllable with anti-convulsive medication. For the rare individual, however, continuing seizures may provide the greatest obstacle to rehabilitation. In some cases, cognitive skills progressively decline after each seizure.

## ***Cognitive***

Repeated mild TBIs occurring over an extended period (i.e., months and even years) can result in cumulative neurological and cognitive deficits while repeated mild TBIs occurring within a short period (i.e., hours, days, or weeks) can be catastrophic or fatal (CDC, 1999). People with TBI are likely to need assistance to perform activities of daily living. TBI can cause epilepsy, and increases the risk for conditions such as Alzheimer's disease, Parkinson's disease, and other disorders affecting the brain (National Institute of Neurological Disorders & Stroke, 2002). Cognitive deficits are divided into some common classification areas, although the distinction is somewhat artificial. In real-world experiences, it is difficult to find a task that draws exclusively upon a single, distinct skill. Almost every activity involves two or more of the cognitive skills described below.

### ***Attention/Concentration and Arousal***

Some persons with TBI find it difficult to focus attention on a task and maintain attention on that task (Rios et al., 2004). Persons may have difficulty becoming sufficiently aroused (i.e., alert) to focus on a specific activity, and they may have a shortened attention span for activities that capture their concentration.

Some may be unable to shift mental tasks (alternating attention) or follow multi-step directions. As an illustration, assume that Sam has TBI but is able, with effort, to focus on a particular work task. If the telephone rings, interrupting his attention for even a minute or two, Sam might be unable to return to the task on which he had been working. Many tasks involve divided attention, such as driving which requires a person be able to concentrate on steering while also obeying speed limits, monitoring other drivers, and perhaps navigating a new route, all while listening to the radio and having a conversation. Not surprisingly, driving is often impossible after TBI.

## ***Memory***

Television shows and movies about persons with TBI usually depict memory loss as the chief cognitive deficit of TBI (Rios et al., 2004). These media depictions are misleading both in their narrow focus on memory loss and their dramatizing the type of memory loss that occurs. While memory loss among persons with TBI is common, the type of loss depicted in the media (where the person awakens from a coma and needs to be informed about events that have occurred) is uncommon. Retrospective amnesia (long term memory loss), the inability to remember all that happened before the injury, seldom occurs.

Far more common is anterograde amnesia (short-term memory loss). For example, many persons cannot remember new information, such as what they had for breakfast that same morning. Individuals may have prospective amnesia (inability to remember future plans). Some persons cannot keep track of their appointments, and even struggle to use memory aids, such as day and week planning tools (e.g., Day Timer). Such tools, while helpful for persons with TBI, assume the user will remember to write down appointments, remember where to write the appointments, and remember to look at the planning guide with regular frequency. Intensive training may be necessary to establish what will become vital organizational habits.

## ***Perception/Visual Processing***

Some persons have problems with spatial orientation. They get lost in a parking lot, going home or to work, or even in their own home environment. These individuals can lose the ability to deal with smaller-scale spatial relations, as well. For example, they may find themselves unable to figure out which container is an appropriate size for storing leftovers or may have difficulty wrapping gifts. They struggle with visual field cuts, in which a large portion of their visual field is missing. (Imagine perceiving the left half of this page as blank.) Consequently, persons with TBI have to learn techniques to take in whole fields of information.

## ***Sensory Deficits***

Other sensory deficits include visual processing difficulties (e.g., neglect of one side of the visual field), photosensitivity (sensitivity to light), loss of hearing, and loss of the senses of smell or taste (Padula et al., 2001).

## ***Verbal Reasoning***

Many persons have problems understanding conversations. They cannot identify the main idea of written or spoken communication, distinguish relevant from irrelevant details, detect similarities and differences, or understand analogies. They struggle to organize ideas, important papers, and their lives. Many persons have difficulty paying bills, even if sufficient funds are available, because of organizational or comprehension difficulties.

## ***Critical Thinking/Logic***

One individual with TBI might declare an inability to return to work despite having received a medical clearance. On the contrary, a different person may attempt to return to work without recognizing the presence of severe cognitive deficits. Judgment and the ability to process information accurately with consistency and speed can be impaired. Inability to make decisions is an additional area of impairment (Rios et al., 2004).

## ***Language***

It is common for persons with TBI to have communicative disturbances (aphasia). Aphasia can take several forms. Individuals may have difficulty expressing thoughts (i.e., expressive aphasia), such as by having word-finding problems (Jane, 1990). For example, one might say, “that thing you write with” to communicate the word “pen.” Other speech problems stem from physical impairments including speech that is unclear when the person has decreased control of the muscles in the lips, tongue, and jaw or because he or she has developed poor breathing patterns. The individual may produce excess saliva that gets in the way of clear speech. Dysarthria, where motor function interferes with correct formation of sounds, produces slurred speech that is extremely difficult to understand. In addition, some persons have problems understanding others (i.e., receptive aphasia). They have difficulty understanding expressions or signs other people use or



find they are unable to understand meanings of words. For example, some are unable to discern humor or anger from a person's tone of voice.

### ***Unawareness***

Anosognosia is a failure to recognize one's own impairment. It is, of course, difficult to distinguish anosognosia from psychological denial, a normal defense mechanism. Giacino and Cicerone (1998) found evidence that three factors underlie lack of awareness of deficits following TBI: (a) diminished awareness of deficits secondary to impaired cognition, especially memory and reasoning deficits; (b) psychological reaction and denial of deficits; and (c) a relatively "pure" inability to recognize areas of impaired functioning as a direct consequence of brain injury. Self-awareness is most impaired for activities that have a significant cognitive and socioeconomic component, and is least impaired for basic activities of daily living (Fleming & Strong, 1999). Self-awareness improves significantly during the first year after injury.

Lack of awareness impedes rehabilitation but may also produce what could be called a benefit. Trahan et al. (2006) compared quality of life for persons with TBI, spinal cord injury (SCI), and for those with no disability. They found that unmet needs were stronger for the TBI group than for SCI and the group without disability. Interestingly, those with the most severe TBI (i.e., loss of consciousness for more than a month) rated their quality of life about the same as did people in the no disability group. Individuals who had experienced only brief losses of consciousness (less than 20 minutes), however, viewed their quality of life as significantly lower than did persons in the SCI group, no disability group, and persons in other TBI severity subgroups. This discrepancy probably stems from the neurological unawareness of some TBI survivors.

## ***Psychosocial***

Various psychological, behavioral, and emotional changes are common following TBI. The most frequent Axis I diagnoses are major depression and select anxiety disorders. Rates of resolution are similar for individuals regardless of their previous psychiatric histories. Major depression and substance abuse disorders are more likely to remit than anxiety disorders (Hibbard, Uysal, Kepler, Bogdany, & Silver, 1998). These conditions and others of a psychosocial nature are discussed below.

### ***Personality Changes***

TBI frequently exacerbates negative pre-injury personality characteristics. For example, someone who had a short temper now has angry emotional outbursts. One adult daughter said of her father after his TBI, "He's Dad, just more Dad." A man who was formerly self-confident is now annoyingly arrogant and critical of others. Conversely, family members often report that the shy have become outgoing, and vice versa. A common complaint of family members after TBI is that the survivor has become intensely egocentric. It may become impossible for a conversation to occur without the survivor turning it into a conversation about self.

### ***Emotional Lability***

Heightened emotional responses or reactions are common. Some feel unable to control mood swings from moment to moment. They laugh or cry for no apparent reason and at inappropriate times.

### ***Flat Affect***

Some survivors show a complete lack of emotions. For example, they report a sense of puzzlement that they neither feel elation at happy times, nor anxiety in stressful times. The individual may have a rational awareness that emotion is warranted in a given situation, yet be incapable of feeling or expressing emotions.

### ***Depression***

Major depression is present in about 40% of individuals who have been hospitalized for TBI (Jorge & Starkstein, 2005). TBI may be accompanied by loss of defining personality traits, career, status, income, relationships, or feelings of competency. It is understandable that depression is a risk for any person who has an awareness of all these losses. Persons with TBI have a high divorce rate, lose many of their friends, and suffer family estrangements. They discover that, after the initial burst of rapid rehabilitation gains following

their awakening from coma, progress levels off. Some deficits are temporary, while others are permanent. Consequences of TBI have emotional costs not only for the person who is injured, but also for the families (Rosenthal, Christiansen, & Ross, 1998). Persistent and high levels of emotional distress, including depression and anxiety, have been documented in family members (Seel & Kreutzer, 2003).

### ***Substance Abuse***

Hibbard et al. (1998) investigated TBI survivors eight years post-injury and found that a significant percent had substance abuse disorders prior to injury. Kreutzer et al. (1996) compared drinking rates after TBI with large-sample studies on non-TBI populations and found that young persons with TBI had pre-injury drinking patterns similar to those in the general population. There was evidence of a decline in alcohol use at initial follow-up, but pre-injury and second follow-up alcohol use patterns were similar. Post-injury illicit drug use rates remained relatively low, falling below 10% at both follow-up intervals. The findings suggested that men with moderate to heavy pre-injury alcohol use and those taking prescribed medications are at greatest risk for long-term alcohol use. Because the damaging effects of alcohol can be magnified after brain injury, it is important to provide education about the effects of alcohol for persons at risk.

### ***Decreased Frustration Tolerance***

After TBI, persons may become frustrated more easily than they did before their injuries (Falvo, 2009). For example, they may realize that particular tasks on which they are working seem elementary. They could competently perform the activities before injury, yet post-injury they are unable to perform them nor perform them with their former ease. They may have to depend on substantial compensatory strategies to do many activities they once performed readily. Under either circumstance, they behave inappropriately or aggressively. Some persons struggle with increased irritability and impatience, have reduced tolerance to stress, and are inflexible.

### ***Impulsivity/Disinhibition***

We all have many thoughts each day that we do not express, or urges on which we do not act. The frontal lobes within the brain keep us from acting on these ideas. Without this check system, people with TBI may ask inappropriate personal questions of co-workers, make inappropriate sexual advances, touch others inappropriately, act in an unsuitably aggressive manner, or make prejudicial and racist remarks.

### ***Lack of Initiative***

Some individuals with brain injury lack initiative, even if they once possessed it. This effect, like impulsivity/disinhibition, is common with frontal lobe injuries. Such persons may be content to watch television all day. They respond to most stimuli with apathy.

### ***Other Emotional Responses***

Sometimes TBI survivors experience sexual difficulties, denial of disability, anxiety, agitation, anger, isolation, and dependence. They may be unable or unwilling to assume responsibility for their actions.

## **Treatment and Rehabilitation Features**

An individual with signs or symptoms of moderate or severe TBI needs immediate medical attention. The priorities of treatment are to maintain the airway, breathing, and circulation. Medical professionals attempt to stabilize the person and focus on preventing further injury; concerns include ensuring the brain has sufficient blood supply, maintaining blood flow, and monitoring blood pressure. To determine a diagnosis and prognosis, the treating physicians order skull and neck X-rays to identify any bone fractures or instability in the spine. Imaging studies, such as Computerized Axial Tomography (CAT or CT) scan, is advisable to diagnose the extent of damage and brain areas affected (CDC, 2003).

Depending on the type and severity of injury, TBI rehabilitation typically includes physical therapy, speech therapy, occupational therapy, and cognitive therapy. These therapies are described below.

### ***Physical Therapy (PT)***

After a coma, many patients need to learn to walk again. It may take weeks, months, or years for a patient to progress from using a wheelchair, to a walker, to a cane and, finally to ambulate unassisted. Some persons never progress past wheelchair use. Nevertheless, because the physical impairments are most visible, survivors frequently put the bulk of their energy into PT, sometimes to the detriment of other therapies.

### ***Speech Therapy***

Beginning with relearning how to swallow (while in the hospital), TBI patients frequently receive a wide spectrum of therapy from speech and language pathologists. Depending on location of injury, a speech therapist helps the patient learn to speak again or may assist with developing word-finding skills or forming sounds. Many speech therapists teach cognitive remediation skills.

### ***Occupational Therapy (OT)***

Occupational therapists help with such skills as learning to button a shirt with one hand to compensate for hemiplegia, or increasing independence by structuring an environment using memory cues. OT may address learning to drive post-injury by teaching compensatory strategies for coping with physical and visual deficits.

### ***Cognitive Therapy/Retraining/Remediation***

Cognitive retraining principally occurs in four settings: hospitals (especially immediately after injury), neuropsychologists' offices, speech therapy clinics, and educational settings such as the New York University Rusk Institute in New York City and the Coastline Community College Acquired Brain Injury Program in Costa Mesa, California.

The cognitive retrainer begins by assessing the client's cognitive functioning and prescribing corresponding cognitive remediation. Such treatment includes tasks designed to help individuals utilize their cognitive strengths to compensate for areas of weakness. This involves teaching new strategies for common tasks. The cognitive retrainer seeks to stimulate progress for those activities with which the person is having difficulties. By stimulating these activities, the cognitive retrainer attempts to build new neural pathways to replace damaged areas, although it is currently felt that after childhood, no new brain cells develop. The dendrites or fibers on each neuron, however, can grow and be functional. From this, neuropsychologists have developed the theory of neural plasticity, which posits that a new neural pathway can be developed to take over a brain function that has been lost because of damage to the area (Reilly & Bullock, 2005).

Cognitive retraining is slow but rewarding. Survivors of TBI are taught new skills and develop compensatory techniques. They learn strategies to use laptop computers to compensate for memory deficits and cued memory techniques. Strategies are learned so the individual can use prosthetic devices effectively. For example, a person with TBI learns to program a wristwatch so that the watch beeps at designated times. These beeps are used as cues to look at a planner or calendar as a reminder of the activities that have been scheduled. Because procedural memory is often regained, persons with TBI are instructed on cognitive procedures to perform tasks on their own. They learn to break down the steps of thinking and organize their thoughts and materials better, allowing for increased productivity.

## **Rehabilitation Outcomes**

Cognitive retraining is a relatively new field, having first gained prominence in the 1980s. The field has yet to develop unassailable research documenting efficacy. Chestnut et al. (1999) conducted a systematic review of over 3,000 studies to evaluate evidence for the effectiveness of rehabilitation methods throughout the phases of recovery. They found significant practice variations occur across clinics and professionals. While some studies have demonstrated highly individualized treatment and assessment, others have shown that standardization of treatment is possible and results in positive outcomes. These researchers called for future studies with a commitment to population-based research, carefully controlled

research design, standardization of measures, adequate statistical analyses, and specification of health outcomes.

It is difficult to know when an individual has reached maximum recovery and improvement (O’Keeffe, Dockree, Moloney, Carton, & Robertson, 2007). Many physicians believe that the outer limits for continued functional improvement are between six months and one to two years following TBI. Yet, cognitive retraining has produced successes as long as 20 years post-injury. Corrigan, Smith-Knapp, & Granger (1998) studied the extent to which outcomes from TBI differed as a function of time. They concluded that outcomes over the first five years following discharge from inpatient rehabilitation programs were dynamic, with most improvement seen during the first two years.

## Factors Affecting Outcome

A variety of factors affect outcome. Severity of injury is the strongest predictor of recovery. Professionals working with TBI survivors measure severity of injury using several methods. First, TBI experts use length of coma to predict outcomes. Studies indicated that the longer the coma, the worse the outcome. Second, duration of post-traumatic amnesia is used to predict outcomes. The longer it takes to start remembering daily events, the more negative the outcome. Unfortunately, some people never regain ability to remember daily events (Zink & McQuillan, 2005).

Age correlates positively with outcomes. Among adults (18 to 89), the older a person is at time of injury, the worse the outcome (Rothweiler, Temkin, & Dikmen, 1998). Research has established a relationship between age and increased psychosocial limitations, especially in persons over age 60. Consequences of TBI worsen with age and severity of brain injury.

Researchers (Jorge & Starkstein, 2005; Kraus & McArthur, 1999; Sherer et al., 1998) have determined that higher pre-injury educational level correlates with better outcomes. Most persons with TBI have difficulty learning new information, but a good portion of pre-injury knowledge and skills can be left intact. Persons who were well educated before injury frequently need to learn new ways to organize and access their previous knowledge and cognitive skills.

A final predictor of good outcome is pre-injury work history. Those who have poor work histories before their injuries are likely to have even greater problems post-injury (Rubin & Roessler, 2008).

## Vocational Rehabilitation and Placement

There is wide variance in outcomes after TBI. Some people with TBI recover fully and return to their former routines. Others are able to learn compensatory strategies and to perform modified versions of their former routines; others retrain for new occupations. Still others require structured work settings or find volunteer positions. Unfortunately, some return home to nonproductive lives.

It seems clear for some persons with severe brain injuries that a reasonable rehabilitation long term goal is supported employment or volunteer work designed to increase life activity (Brodwin, Parker, & DeLaGarza, 2003). Other persons with TBI are able to return to some form of work. Briel (1996) and Sherer et al. (1998) addressed supported employment programs. Briel described an approach for promoting the effective use of compensatory strategies at the job site for individuals with TBI through supported employment programs. This approach has three main components: (a) assessment of residual skills, (b) identification of potentially effective compensatory strategies through situational assessment, and (c) incorporation of compensatory strategies into on-the-job training. Sherer et al. showed that self-awareness is essential for succeeding in employment. Characteristics of supported employment that can promote successful return to work include: community placement and integration (Sample & Langlois, 2005), competitive hiring and wages, zero exclusion policy, holistic assessment, emphasis on choice and job matching, intervention after placement, co-worker and employer education, long-term follow along, job completion guarantee, and intensive ongoing analysis of program outcomes.

Parenté and Stapleton (1996) advocated increasing work longevity using an intervention model which includes a comprehensive assessment of learning style, work sample evaluation, domain specific training, cognitive remediation that emphasizes learning functional skills, supported employment,



environmental engineering of the work site, and the use of prosthetic devices (e.g., electronic memory aides). This approach will not work if the person has “unawareness,” the cognitive condition that causes a lack of insight into one’s deficits (Trahan et al., 2006). Compensation strategies are challenging, time-consuming, and can be embarrassing. At the same time, persons with TBI must be aware of their deficits and the consequences to be able to implement the appropriate cognitive strategies.

Because the disability is not visible and often misunderstood, the employer and the TBI survivor must cooperate to work out appropriate accommodations. For example, the employee may need written instructions from the employer or may need to be assigned only one or a few tasks at a time. The employer must have knowledge of the employee’s strengths and limitations (Brodwin et al., 2003). For example, a mechanic may still be able to diagnose what is wrong with an automotive engine by listening to it and observing its functioning, even though the person is no longer able to reassemble the engine after diagnosing and fixing the problem part. Similarly, an attorney might be able to assist with legal strategizing, but be unable to remember what steps have been taken or which forms have been filed with the court. Persons who process information slowly take longer to perform assigned tasks. Some employers can work around this deficit, but not if impulsivity makes accuracy poor. Therefore, persons with TBI need to be trained to work more slowly when accuracy is essential.

Individuals whose TBI has caused them to lose their inhibitions often struggle to return to work successfully (Brodwin et al., 2003; O’Keeffe et al., 2007). Most jobs require working with others. Impulsive sexual remarks, racial slurs, and low frustration tolerance are not the characteristics of those who succeed in most employment contexts. While employers can usually work around many memory and other cognitive problems, they are rarely willing to deal with obnoxious behaviors. Job coaching helps in these situations. Some persons with TBI require this type of support permanently to retain employment; the coach serves as a “proxy frontal lobe” for the brain part that has been damaged.

## **Conclusion**

Each year, TBI changes the lives of over a million people. Those at highest risk are young males. In TBI, injury is classified as closed head injury, open head injury, as well as traumatic brain injury. A variety of deficits such as physical impairments, pain, fatigue, mobility disorders, seizures, cognitive dysfunctions, speech impairments, and changes in personality can be found. These can be devastating and challenging to the injured persons, family members, physicians, and helping professionals. Individuals may experience not only trauma to the brain but alterations in body image, changes in personality, age-related and role-related problems, and financial difficulties. Immediate attention to their physical and psychosocial needs, in addition to maintaining the focus on early rehabilitation and prevention of recurring injury or complications are imperative to the recoveries of people with TBI.

## **Case Study**

Mr. Randall Owens is 47 years of age, African-American, with an Associate of Science degree in the Building and Construction Trades from a trade and technical college. While on a construction site, he fell 25 feet from the roof of a house, landing on his right side. His wife was present and she reports that he was unable to speak for approximately five minutes and was moving in and out of a comatose state. Paramedics transported Mr. Owens to a local trauma center where a CT scan of his brain revealed cerebral contusions, intracranial air in the subarachnoid space of the frontal lobe, punctate hemorrhages in the frontal lobe, a right orbital fracture, a fracture at the base of the skull, right frontal skull fracture, right temporal skull fracture, and temporal bone fracture. Additionally, Randall suffered trauma to the left internal carotid artery, right rib fractures, and eccymosis of the right eye. Complications during hospitalization included pneumonia.

Discharged home after eight days of hospitalization, Mr. Owens received outpatient physical therapy for three weeks and speech therapy for six weeks. Eight months later, he initiated outpatient full-time day treatment at a comprehensive post-acute rehabilitation center where an evaluation indicated the following problems: minimally decreased visual processing speed, reduced binocular vision, diminished convergence skills, impaired depth perception, decreased strength in both upper extremities, word finding errors, decreased verbal fluency, diminished auditory and visual recall, and decreased abstract reasoning.



Through participation in a full-time day-treatment program for two weeks, Randall was able to transition to a part-time program at a rehabilitation center. Two weeks later, he was discharged.

Six months later (1½ years after injury), Mr. Owens was struggling to operate his own construction company, started years before the injury. The physical demand of working 12-hour days was difficult for Randall; there were problems remembering details of jobs, with sequencing the steps of the jobs, and communicating effectively with clients, vendors, and his two employees. He reported, “I want to get my life back. I want the motivation and the desires I used to have to work and enjoy life. I feel like I have lost control of my life.”

Mr. Owens recognized that he might need to work for someone else instead of operating his own business. He enrolled in a community college-based cognitive retraining program, which taught him compensatory strategies to use for memory, sequencing, and communication deficits. With the help of his instructors, peers, and a counselor, he learned to identify and accept his limitations, emphasize his strengths, and set realistic expectations for his performance. While still enrolled in the program, he became employed at a local store of a large home-improvement retail chain. He found he could utilize his past experience as a contractor by working in the window and door department helping customers and writing orders. Able to concentrate on a rather isolated portion of the building process, Randall was less overwhelmed and enjoyed great success, receiving a promotion to a department assistant manager. Being accustomed to being his own boss, he did encounter difficulties with following procedures he felt were illogical and in taking orders from others. Mr. Owens worked on the adjustment process with his counselor and was able to achieve success and personal satisfaction with his position.

### Questions

1. Identify possible functional limitations of traumatic brain injury, including physical, cognitive, and psychosocial.
2. Define emotional lability and its potential impact on job performance. How would this impact the case?
3. Some individuals with brain injury have problems with impulsivity and make inappropriate remarks (including sexual remarks and racial slurs). Discuss implications of this for vocational rehabilitation.
4. Evaluate cognitive retraining from the standpoint of this case.
5. Describe other possible vocational rehabilitation plans for this client.
6. Discuss supported employment for clients with severe head trauma and whether this is applicable to this case? If needed, refer to the chapter on mental retardation in this book for information on supported employment.

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## Chapter 16

# CEREBROVASCULAR DISEASE

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## Introduction

Cerebrovascular disease refers to a collection of conditions caused by abnormalities of the blood vessels supplying the brain. Stroke is a medical emergency characterized by the rapid loss of brain function due either to interruption of its blood supply (ischemic stroke) or bleeding into the brain from rupture of a blood vessel (hemorrhagic stroke). Ischemic strokes account for 80% and hemorrhagic strokes 20%. Both forms result in reduction of oxygen and nutrition to the brain which leads to the symptoms of stroke and possible brain damage (Mayo Clinic, 2005). The duration of brain function loss must be at least 24 hours in length or the episode is called a transient ischemic attack (TIA). Over 700,000 strokes (also known as cerebrovascular accidents [CVAs]) occur each year in the United States. They can result in temporary or permanent neurological dysfunctions; stroke is the third leading cause of death in this country (American Stroke Association, 2007a). Stroke occurs in people of all ages, but mainly among people over 60 years of age.

Approximately half of all cerebral vascular events are TIAs, a condition of temporary neurological deficit lasting less than 24 hours before full recovery of all functions (American Stroke Association, 2007a). Importantly, TIA symptoms mimic those of stroke and should be treated as an emergency. Symptoms which last longer than 24 hours are always defined as stroke and result in some permanent brain damage. Since 1995, an FDA approved medication has improved the treatment for ischemic stroke and been shown to reduce resulting disability (Mayo Clinic, 2005). Novel treatments and cutting-edge technology are being discovered and tested in clinical trials. Specialized emergency care and stroke centers have proven to be effective in further improving care. This chapter provides an overview of stroke as a disability. It begins with etiology, followed by anatomy of the brain, types of stroke, diagnosis, complications, and concludes with treatment, prevention, and rehabilitation potential of stroke survivors.

## Etiology

### *Risk Factors*

The two major categories of stroke risk factors are labeled modifiable and non-modifiable. Modifiable risk factors are under the control of the patient, while non-modifiable are beyond one's control. Modifiable risk factors for stroke include the following: hypertension, cigarette smoking, heart disease, Type II diabetes mellitus, excessive alcohol intake, high cholesterol, drug abuse (especially cocaine), and carotid artery atherosclerosis (cholesterol build-up causing blockage of an artery). The most important modifiable risk factor which contributes to the highest rate of stroke is hypertension, as many clinical studies have proven that blood pressure reduction leads to lower rates of stroke (American Stroke Association, 2007a; Mayo Clinic, 2005). Non-modifiable stroke risk factors include age, a history of prior stroke or transient ischemic attack, male gender, race, certain genetic disorders of blood clotting, and a family medical history of stroke or cardiovascular disease. Patients who have a history of TIAs are nine times more likely to have subsequent strokes than those without (American Stroke Association, 2007a; Oliveira, Nesbitt, & Murphy, 2006). Yearly, there are 500,000 recurring and 200,000 new cerebrovascular accidents in the United States. Additionally, African-Americans have higher rates of stroke, partly due to more cases of diabetes and high blood pressure within this population.



## ***Signs and Symptoms***

Stroke is defined by the sudden onset of neurological deficits. Although some patients may have warning signs that appear before a stroke, many occur without warning. Symptoms include sudden (a) numbness or weakness in the face, arm, or leg, which usually occurs on one side of the body, (b) severe headache with unknown cause, (c) vision impairment in one or both eyes, (d) confusion, such as having trouble understanding or speaking, and (e) trouble with walking or coordination (Goldman, 2002). As discussed above, when these symptoms dissipate rapidly and there are no permanent residuals, the patient may have had a TIA. If the symptoms persist for at least 24 hours from onset, the diagnosis is stroke.

## **Anatomy of the Brain**

Stroke occurs within the brain, the control center for the majority of body functions. The cerebral cortex, the largest part of the brain, is responsible for language abilities and personality. The brainstem and cerebellum control automatic functions such as heartbeat, breathing, and coordination. A wide range of symptoms and deficits results from damage to various areas of the brain.

Blood is transported to the brain from the heart via arteries, and is returned to the heart via veins. Stroke is primarily caused by interruptions in either the arterial circulation of the brain or, less commonly, abnormalities in the venous circulation. The four major arteries that supply the brain include two carotid arteries in the front of the neck and two vertebral arteries in the back of the neck supplying the posterior lobes of the brain and brain stem. Each artery supplies a specific area of the brain. When an artery is occluded, lack of oxygen (ischemia) leads to abnormalities of brain function and, if persistent, results in death of brain cells supplied by the involved artery. The brain can only tolerate four minutes of anoxia or lack of oxygen before brain cells begin to die.

The carotid artery splits into the middle cerebral arteries and anterior cerebral arteries. These arteries supply the parietal lobe and portions of the frontal lobe and anterior portion of the temporal lobe. If the dominant hemisphere middle cerebral arteries are blocked (usually the left brain), language and speech are affected. Blood from the anterior cerebral artery supplies the anterior frontal lobe. Small perforating arteries provide the blood supply to areas deep within the brain (e.g., the thalamus and ganglia). In the back of the brain, two vertebral arteries combine to form a basilar artery, which supplies blood to the brainstem and cerebellum. The terminal branches of the vertebrobasilar system supply the visual cortex. These small arteries are susceptible to rupture from long-standing hypertension. Collateral circulation generally does not occur in the blood supply of the central nervous system, even though it is present in most areas of the body. When collateral circulation does develop, arterial interconnections occur between major arterial systems – that is, when one artery is blocked, the area needing oxygen obtains blood from another major artery (American Stroke Association, 2007a).

## **Categories of Stroke**

The two main categories of stroke are (a) ischemic stroke due to blood vessel blockage and (b) intracranial hemorrhage stroke caused by blood vessel rupture. The etiology, diagnosis, and treatment differ for these two types of stroke. The following section reviews the different types of stroke in both categories.

### ***Ischemic Stroke***

Ischemic stroke is the most common type of stroke, accounting for 80% of all events (Interventional Management of Stroke [IMS]II Study Investigators, 2004). It results from blockage of an artery due to a blood clot or lodged plaque. The origination of the blood clot can be multifactorial, but the two major categories are cerebral embolus and cerebral thrombus.

#### ***Cerebral Embolus***

When a plaque or particle travels from the heart or a larger artery and lodges in a smaller artery, obstruction of blood flow causes an embolic stroke. If the obstruction persists for over a few minutes, brain cells known as neurons begin to die. The death of brain cells and tissue lead to functional limitations. However, if the arterial obstruction lasts for a brief period and permits full recovery, the condition is

described as a TIA. Sources of emboli are typically the heart (ascending aorta or aortic arch) and carotid arteries. Cerebral embolus is usually caused by atherosclerotic vascular disease (hardening of the arteries) in which a small portion of atherosclerotic plaque breaks off and travels to the blood vessels within the brain. Embolic stroke may also be caused by a blood clot originating in the heart. In people over 60 years of age, atherosclerotic vascular disease and cardiac disease are the most common causes of ischemic stroke. Cerebral embolus is rare in younger people.

### ***Cerebral Thrombus***

Another form of stroke, caused by a blood clot, develops from atherosclerotic plaque in the wall of the vessel. Years of uncontrolled hypertension, tobacco abuse, and high cholesterol may cause atherosclerosis or “hardening of the arteries.” The build-up of cholesterol plaque can result in a fracture causing a local clumping of platelets occluding a vessel. The resultant mix of platelets and fibrin, a thrombus, does not travel; rather it causes an arterial obstruction at the site of origin. Depending on the length and severity of the obstruction, resulting disability can vary from partial to full recovery.

## ***Intracranial Hemorrhage Stroke***

Intracranial hemorrhage, occurring in approximately 20% of strokes, is caused by a ruptured blood vessel (IMS Study Investigators, 2004). The focus of this chapter includes three main forms of intracerebral hemorrhage (ICH): (a) hypertensive hemorrhage due to uncontrolled high blood pressure, (b) subarachnoid hemorrhage due to aneurysm rupture (a congenital weakness and ballooning of the vessel wall), and (c) rupture of a vascular malformation (usually a congenital lesion).

### ***Cerebral Hypertensive Hemorrhage***

Long-standing uncontrolled hypertension can lead to damage in the arteries of the brain (and other organs, such as the kidneys and eyes). The resulting damage, called lipohyalinosis, affects the small blood vessels in the brain. Lipohyalinosis is a progressive process that weakens the wall of an artery and increases the risk of rupture. The most common sites of cerebral hypertensive hemorrhage occur in deep structures supplied by the small, penetrating arteries affected by lipohyalinosis. These structures include the basal ganglia and thalamus. In addition, the cerebellum (the portion of the brain responsible for coordination) is subject to hemorrhage. The morbidity and mortality for hypertensive hemorrhage is extremely high; one-third of these individuals die within a month. Unfortunately, treatment options for hypertensive hemorrhage are minimal.

### ***Aneurysmal Subarachnoid Hemorrhage***

An aneurysm is an out-pouching of a blood vessel due to thinning of the arterial wall. It is usually located at the branching points of cerebral blood vessels. Approximately 85% of cerebral aneurysms develop in the middle cerebral artery and the anterior and posterior communicating arteries. Roughly one-fifth of those who experience aneurysm have additional aneurysms in the future. Aneurysm formation is more likely to occur in individuals with long-standing high blood pressure. Weakened arterial walls can rupture, resulting in subarachnoid hemorrhage (SAH). Patients with aneurysmal SAH are typically younger than their ischemic stroke cohort (typically 40-60 year olds). Risk factors for aneurysmal rupture include: size of the aneurysm, excessive alcohol intake, tobacco abuse, and congenital abnormalities, such as fibromuscular dysplasia and polycystic kidney disease.

Patients who undergo aneurysmal ruptures usually present with severe headache and focal neurological deficits. Aneurysmal SAH is a catastrophic condition affecting 30,000 individuals in the United States yearly. Sixty percent of these individuals either die or suffer permanent disability; fifty percent of the survivors with favorable outcomes experience considerable neuropsychological dysfunction (Chyatte, 1996). Cerebral vasospasm (i.e., narrowing of proximal arterial segments) complicates 20-50% of cases and is the major cause of disability and death associated with aneurysmal SAH. The spasm of the vessel itself, which complicates SAH, can also lead to ischemic stroke.

### ***Cerebral Vascular Malformation***

Cerebral vascular malformation, also known as arterial venous malformation (AVM), is characterized by congenital abnormalities of blood vessels in the brain. AVM, tangled networks of blood

vessels, consist of arteries that directly empty into veins without the normal intervening capillaries that support brain tissue. These lesions, usually present at birth, tend to become apparent later in life when they leak or bleed. It is only half of all vascular malformations, however, which eventually become apparent in this manner. AVM rupture typically occurs abruptly without symptoms and results in neurological deficits, ranging in degree of severity depending on the size, area, and extent of hemorrhage in the brain.

## **Diagnosis**

Diagnosis of stroke is made by the development of acute neurological symptoms, abnormalities seen during a clinical neurological examination, aided by imaging techniques. Upon arrival at the hospital, all stroke patients receive a computerized tomography (CT) scan or a magnetic resonance image (MRI) of the brain, which evaluates for intracranial hemorrhage, establishes ischemic stroke from hemorrhagic, and eliminates other causes of the neurological symptoms (e.g., brain tumor). Specialized centers for the treatment of stroke can rapidly evaluate the cerebral arteries using either ultrasound or arteriography (injection of intravenous contrast dye to visualize the anatomy of the cerebral circulation). These studies assist the clinician with diagnosis, etiology, and treatment. Additional studies obtained during hospitalization include a cardiac evaluation, including electrocardiogram and echocardiogram. Once it is determined that the patient is having a stroke, appropriate management can be undertaken (Goldstein & Simel, 2005).

## ***Complications and Limitations***

Cerebrovascular diseases, such as stroke, follow a spectrum of severity. Frequently, small strokes occur resulting in mild or non-noticeable residual deficits. Contrastingly, a stroke classified as “severe” is a devastating occurrence. One-third of individuals with severe strokes die within two weeks. Mortality rates increase with advancing age and the presence of other comorbid diseases such as hypertension, heart disease, and diabetes. Approximately one-third of people with CVAs die, one-third have significant neurological deficits, while the remaining individuals have minor or no deficits. A stroke can affect any part of the brain and symptoms vary depending on size and location of the damage. When brain tissue is damaged, far reaching deficits occur including paralysis, cognitive decline, language difficulty, behavioral and mood changes, and psychological effects, such as depression. Those surviving a severe stroke are additionally at much higher risk for other medical complications, including pneumonia and blood clots in the legs, called deep venous thrombosis. Medical complications can result in death. Other long-term complications of stroke include the development of contractures, seizure disorders, bowel and bladder problems, and spasticity of muscles.

### ***Neurological Deficits***

In general, when the cerebrovascular accident occurs on one side of the brain, the opposite side of the body is affected. After a stroke, typical neurological deficits may range from paralysis, to hemiparesis, to minimal or unnoticeable deficits. Brainstem strokes caused by abnormalities of the posterior circulation (i.e., vertebral or basilar arteries) can be associated with display of abnormalities ipsilateral (affecting the same side of the accident) to the face, and contralateral (affecting the opposite side of the accident) to the body (Vander Worp & van Gijn, 2007). An overview of physiological effects and physical limitations related to post-stroke neurological deficits follow.

Typical clinical residual findings of middle cerebral artery (MCA) stroke include abnormalities of the motor and sensory cortex leading to spastic hemiparesis and sensory loss, respectively. If the stroke occurs in the right MCA territory, the patient may suffer from anosognosia (also known as neglect), a state in which the individual does not recognize the affected side of the body as belonging to himself or herself. In such cases, the person is unable to perceive objects on either the right or left side of the central field of vision and is unaware of the deficit. When the stroke occurs in the right side of the brain, visual-spatial deficits result. Individuals with visual-spatial problems may incorrectly interpret visual information and have difficulty orientating to the surrounding environment. A stroke on the left side of the brain may result in paralysis to the right side of the face, arm, and leg. Lesions of the left MCA may affect the language centers, causing speech and/or comprehension impairment. When the speech center is affected, the condition is known as aphasia. Aphasic patients have difficulty sounding words, understanding words, or both; the

different forms of aphasia are termed expressive, receptive, and global aphasia. Anosognosia and aphasia seriously impair vocational rehabilitation (Goldman, 2002). Dysphagia, a swallowing disorder, occurs in up to 65% of stroke survivors. If not identified and managed, this condition can lead to poor nutrition, pneumonia, and increased disability.

Posterior circulation strokes involving the vertebral and basilar arteries can be fatal. Basilar artery occlusion is known for its extreme mortality rates, up to 80%. These patients present with declining mental status, coordination difficulties (ataxia), double vision (diplopia), severe dysarthria (slurred speech), dysphagia (difficulty in swallowing), and trouble controlling saliva. Dysphagia can lead to an inability to maintain the clarity of the airway. Patients with posterior circulation strokes often require mechanical (artificial) ventilation with a breathing tube. Depending on the extent of the stroke, patients can have severe global weakness or even tetraplegia (quadriplegia) (Vander Worp & van Gijn, 2007).

### ***Paralysis***

The complete loss of function of one or more muscle groups due to damage to the brain is referred to as paralysis. Depending on the area and size of damage, post-stroke survivors may experience weakness, loss of muscle function, and loss of sensation.

### ***Paresis***

Paresis refers to weakness of muscle strength. Paresis can be localized, and often follows a pattern. For example, localized paresis of one side of the face may result due to damage of the brain tissues that leads to inflammation of the facial nerve affecting that side of the face.

### ***Hemiplegia***

This is a common consequence of stroke. It involves paralysis on one side of the body. A person is said to have right hemiplegia if the right side of the body is paralyzed. As mentioned earlier in this chapter, the lesion causing paralysis is typically in the opposite hemisphere of the brain. When the stroke occurs on the left side of the brain, paralysis limits functions of limbs on the right side of the body. This condition usually has more impact on the upper extremity than the lower extremity. The extent of paralysis is dependent on the site and size of the lesion. Visual-spatial deficits occur when the stroke takes place in the right brain. An individual with visual-spatial problems may incorrectly interpret visual information and have difficulty with environmental orientation. A stroke on the left side of the brain may result in paralysis to the right upper and lower extremities. Also, these individuals can have deficits in ability to solve problems. If the speech center is affected, aphasia (inability to produce/comprehend speech) may occur.

### ***Cognitive Deficits***

Cerebrovascular accidents cause a wide variety of cognitive deficits. It is common to find stroke survivors having difficulties in memory retention, especially that of newly acquired information. Complications of such accidents include slowed retention and disorganized processing of information. Intellectual deficits, behavioral changes, and problem-solving difficulties. Intellectual deficits can be minimized and others circumvented with relearning and through job accommodation. For example, patients with memory difficulty can use a notepad to record information. Specific intellectual tasks crucial to an individual vocation can be reassigned to other employees if unable to be accomplished by the person. If job modification cannot accommodate the extent of intellectual impairment, use of transferable skills or retraining needs assessment.

The most common psychological problem found among stroke survivors is depression; this condition may be related to the degree of functional disability and can impact a person's ability for rehabilitation (Thomas & Lincoln, 2006). Common symptoms of depression include (a) persistent sadness and anxiety, (b) feelings of hopelessness, (c) loss of interest or pleasure in daily activities, (d) fatigue and lack of energy, (e) insomnia or hypersomnia, (f) loss or increase in appetite and weight changes, and (g) difficulties in concentrating, remembering, and decision-making (American Stroke Association, 2007b). Effective treatment for depression ideally includes both consultation with a psychologist or psychiatrist and antidepressant medications. Treating depression improves the stroke survivor's mood and physical recovery. Treating depression enhances cognitive and intellectual improvement. In addition, social support from family, friends, stroke support groups, and rehabilitation professionals remains a crucial part of



recovery for stroke survivors dealing with depression. Emotional lability (partial loss of emotional control) may be experienced and expressed clinically as various emotional states, such as crying, sudden laughter, depression, and anger. Such emotions change rapidly and for no apparent reason; the individual may additionally express emotions considered inappropriate for particular situations. Persons with cerebrovascular accident may also show irritability, frustration, insecurity, hesitancy, anger, hostility, depression, flat affect, and a generalized indifference (Goldman, 2002).

Besides affecting the patient, stroke impacts family members and caregivers. The results of a stroke are physically, financially, and practically demanding, as well as emotionally distressing for all involved. Feelings of sadness, grief, exhaustion, fear, and anger are common reactions to increased dependence and vulnerability following a stroke. The disruption in family relationships that result from a stroke, in addition to resulting heightened conflicts and loss of supportive ties, can be minimized with help and support from available support groups (Palmer & Glass, 2003).

## **Treatment**

Emerging treatments for ischemic stroke and hemorrhagic stroke vary distinctly. Regardless of the type of stroke, timing is an important element of treatment. Emergency medical care must be provided at the onset of stroke symptoms. The sooner treatment is delivered, the less severe the resulting disability. Brain damage and other negative results are significantly reduced with prompt emergency care (Mayo Clinic, 2005).

### ***Medications***

In the case of ischemic stroke, pharmacologic thrombolytic therapy can be administered (within three hours) which aims to dissolve the clot causing the obstruction and unblock the artery. Tissue plasminogen activator (tPA) is the most commonly administered thrombolytic in use today. Delivered intravenously, tPA has been shown in a large clinical trial to improve functional outcomes by dissolving blood clots and restoring blood flow (National Institute of Neurological Disorders and Stroke rt-PA Stroke Study Group, 1995). Benefit was maintained at 12 months after stroke (Kwiatkowski et al., 1999). As expected, there was a risk of bleeding when stroke patients received tPA, but this risk was outweighed by its benefit; patients who received tPA were 30% more likely to have minimal or no neurological symptoms when compared to those who received the placebo. However, thrombolytic therapy is controversial. The American Academy of Emergency Medicine (2007) issued a position statement against the use of thrombolytic therapy. Research on this therapy is ongoing.

Other treatments for ischemic stroke are being investigated. For example, researchers are combining tPA with other blood thinners or delivering the tPA to the clot directly by placing a small tube through the groin and gaining access to the cerebral arteries (Interventional Management of Stroke II Study Investigators, 2004; Sugg, Pary, & Uchino, 2006). Other catheters which allow clot retrieval from inside the artery are being tested (Flint, Duckwiler, Budzik, Liebeskind, & Smith, 2007).

If a narrowed carotid artery in the neck proves to be the cause of either a TIA or an ischemic stroke, a patient can undergo surgery called carotid endarterectomy (CEA). This procedure removes plaque from the carotid artery, reopens the blocked artery, and improves blood flow. Performing the CEA reduces subsequent risk of ischemic stroke.

Treatment for hemorrhagic stroke is minimal. Researchers have failed to show that drugs which reduce bleeding lead to improved outcomes for these patients (American Academy of Emergency Medicine, 2007; Mayo Clinic, 2005; Mertz, 2004). Surgical decompression has failed to prevent disability or death. However, for subarachnoid hemorrhage, a neurosurgeon can perform aneurismal clipping around the base of the aneurysm to prevent future rupture and reduce the risk of subsequent stroke.

### ***Physical and Occupational Therapy***

For patients who suffer from post-stroke hemiplegia or hemiparesis, physical therapy provides strengthening exercises for both the upper and lower extremities. Occupational therapy provides range-of-motion activities and splints which assist in preventing contractures and spasticity. Programs can



be orientated toward recovery of occupational modalities and may utilize orthotic devices, such as leg braces, which help the person ambulate. Once the counselor and client have decided on a return to work objective, special orthotic devices can help the person perform specific job functions (Brodwin, Parker, & DeLaGarza, 2003).

### ***Medical Advances***

The brain was once thought to be a static, non-repairable organ. Although neurons do not regenerate, recent brain-imaging techniques have given researchers and physicians new understanding of the ability of the brain to adapt and regain function after a stroke. Brain-plasticity is the ability of one area of the brain to take over the function of a damaged region. Brain cells surrounding the damaged area assume the functions of the damaged cells. Recent rehabilitation advances include (a) constraint-induced therapy, restricting the use of the unaffected limb while intensively training the affected limb; (b) focused-use therapy that includes thousands of passive movement repetitions to help the injured brain relearn how to use the impaired limb; (c) electrical stimulation that uses low voltage electricity to stimulate muscles weakened or paralyzed by stroke; and (d) biofeedback which involves putting a sensor over a target muscle (Mayo Clinic, 2005). In biofeedback, when the target muscle moves, the electrical activity generated is sensed by the device, providing immediate feedback to the individual. Although the science is currently in the preliminary phase, research on stem cell implants has been initiated. It is hypothesized that delivery of stem cells to a stroke patient could stimulate other cells to grow in the brain and form new connections among cells to help restore motor function (Chu et al., 2008). If effective, stem cell implants may be used successfully alongside physical therapy to promote recovery from stroke.

## **Prevention**

Preventing a first-time stroke (primary prevention) and recurrent strokes (secondary prevention) remain the goals of stroke neurologists. Targets that provide protection include: blood pressure reduction, antiplatelet medications (e.g., aspirin), cholesterol reduction, diabetes control, smoking cessation, and blood thinners for heart arrhythmias. Because compliance with medication and resulting side effects is challenging for some individuals, vocational rehabilitation counselors are key in providing support and assistance, keeping in mind that people who have had stroke may be taking multiple types of prescribed medication. Familiarity with medications and their side effects, especially when considering return to work, is crucial to successful rehabilitation.

### ***Blood Pressure Control***

Blood pressure reduction remains the most crucial means of avoiding stroke. Long-term damage to the arteries that results from unmanaged hypertension increases the risks of both ischemic and hemorrhagic strokes. Numerous clinical studies have shown a reduction in stroke when blood pressure was reduced with antihypertensive medications (Dahlöf, Devereux, & Kjeldsen, 2002; Officers and Coordinators for the ALLHAT Collaborative Research Group, 2002; PROGRESS Collaborative Group, 2001). Aspirin and the combination of aspirin plus extended-release dipyridamole help prevent recurrent strokes (Diener et al., 1996; Interventional Management of Stroke II Study Investigators, 2004; International Stroke Trial Collaborative Group, 1997). Management of high cholesterol has been shown to decrease the risk of stroke and heart disease. To reduce the risk of stroke in patients with diabetes, control of blood glucose levels is crucial, as improved blood sugar control is associated with reduced stroke and heart disease. In patients with cardiac arrhythmias, blood thinners (anticoagulants) are effective in preventing clots originating in the heart.

### ***Diet and Exercise***

The maintenance of a healthy diet low in saturated fats and high in complex grains, fruit, and vegetables reduces the risk of stroke. Many metabolic changes occur during dietary restriction including (a) lower blood pressure; (b) reduced arterial stiffness, cholesterol points, and triglyceride level; (c) slower age-related decline in functional and cognitive capacities; (d) enhanced resistance to stress; and (e) improved ability to adapt to stress at the cellular level (Mayo Clinic, 2005). Along with a healthy diet, exercise is an important part of a healthy lifestyle. According to Mertz (2004), recommendations in 2002

were issued by the National Academies' Institute of Medicine to include at least an hour of "moderately intense" exercise daily, which may be distributed throughout the day. Methods of accumulating the 60-minute minimum exercise requirement include taking a brisk walk in the morning, climbing stairs instead of taking an elevator at work, and riding a stationary bike instead of sitting on the couch to watch television at night. Exercise is crucial for the individual's health and promotes proper functioning of blood vessels, improves delivery of oxygen to muscle tissues, and increases the good-to-bad cholesterol ratio. Stroke survivors should work closely with their health-care professionals to develop personalized exercise plans to meet individualized needs.

### ***Smoking and Drug Use***

Tobacco and illicit drugs should be avoided since they are addictive and have deleterious effects on blood vessels. Damage to the arteries can lead to accelerated atherosclerosis. Along with the medical community, the American Heart and Stroke Associations have bolstered their opposition to smoking and illicit drugs, as both are associated with the development of cerebrovascular disease.

## **Rehabilitation Potential**

Rehabilitation is a critical part of recovery and the outlook for stroke rehabilitation is increasingly optimistic. Approximately half of stroke survivors experience some permanent disability; the success of rehabilitation depends on several key factors. Such factors include (a) timing of treatment, (b) extent, location, and size of brain injury, (c) intensity and frequency of therapy, (d) availability of companionship and emotional support, and (e) the stroke survivor's personal outlook and attitude (Mayo Clinic, 2005). Maximum return of function after a stroke can require a six-month recovery period; counselors must take this into account before determining an individual's rehabilitation potential (Goldman, 2002; Heinemann, Crown, & McMahon, 2000). Early intervention is crucial to maintain and encourage the person's return to work, keeping in mind that maximum return of function does not occur immediately.

The goal of stroke rehabilitation is to help the survivor maximize independence. The level of independence reached after rehabilitation is different for each individual, depending on the size and extent of stroke and previous functioning. Physical, speech, and occupational therapy aid in recovery. Individuals work to improve independence in many areas such as mobility, communication, swallowing, cognitive and behavior modifications, and self-care. Below, are a few characteristics of rehabilitation unique to stroke survivors (Heinemann et al., 2000).

### ***Passive Exercise***

Manually-assisted movement serves two primary purposes. First, the repetitive movements help prevent spasticity and contractures which are limiting and painful for stroke patients. Development of contractures can lead to skin breakdown and infections. The inability to flex and extend muscles due to contractures inhibits caregivers from providing effective bowel and bladder care. Second, passive movement of the affected limbs may aid the brain in reorganization and recovery of function. This conventional forced-use therapy is performed in thousands of repetitions with the help of a physical therapist or caregiver (Mayo Clinic Clinic, 2005).

### ***Assistive Devices***

Canes, walkers, and motorized carts increase mobility after stroke. Hoists used at home and some worksites assist lifting and care of the patient. Mechanical devices aid pushing and pulling work activities. A variety of work-related equipment is adaptable for one-hand operations. Various types of orthotic devices improve hand functioning; use of a computer minimizes writing. Desk and file cabinets may be arranged to diminish limitations. Often, simple accommodations provide for access by wheelchair users. Various accommodations can be recommended, depending on the particular work site and functional needs of the individual (Goldman, 2002; Mayo Clinic, 2005).

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## ***Technology***

Science has enhanced the range and availability of adaptive devices for post-stroke survivors. For individuals with limited strength or hand dexterity, low-tech devices are available such as doorknob-turners which slip over a doorknob to allow the door to be opened with lever action. Fine motor skill impairment causes changes in the individual's daily routine. Assistive devices, such as those that allow buttons to hook, make the resumption of independence easier. High-tech assistive technology include computers with specialized programs that speak for people with severe aphasia or enhance vision. Such programs allow survivors to express their needs and wants with greater ease. Computerized robotic gait trainers allow patients to be mobile at early stages and are being studied in clinical trials (Brodwin et al., 2003). Funding for such technology can be assisted by programs like Medicaid, Medicare, Social Security Disability Insurance, state rehabilitation work incentive programs, and private insurance (American Stroke Association, 2007a).

## ***Increased Dependency on Caregivers***

As a result of physical and cognitive deficits, stroke survivors may need assistance from caregivers. These people are likely to be immediate family members. Because stroke is characterized by its sudden onset, change of roles and family dynamics occurs quickly and without preparation, causing emotional distress. Stroke survivors with high levels of emotional support demonstrate increased levels of recovery. Lack of perceived support is associated with the presence and severity of depression. Survivors and family members can experience positive effects after stroke, such as increased compassion and empathy for others, self-confidence, attention to relationships, spiritual growth, reassessment of personal values, and enhanced sense of the meaning of life (Palmer & Glass, 2003).

## ***Psychosocial Factors***

Studies of post-stroke outcomes focus on recovery of physical function and reduction of medical complications. Some studies cite psychosocial factors, such as motivation, as a key to successful vocational rehabilitation (Clark, Rubenach, & Winsor, 2003; Mayo Clinic, 2005). Socioeconomic issues also affect return to gainful employment. Workers with higher education levels and those with professional, managerial, and technical jobs return to work at a significantly higher rate than those with lower levels of education and unskilled manual job experience. Vocational programs that offer a multidisciplinary approach to stroke rehabilitation have greater success in rehabilitation and employment. A key to success is careful coordination of comprehensive services by a multidisciplinary team made up of rehabilitation counselors, physicians, therapists, psychologists, and social workers.

## **Conclusion**

As the most common cause of long-term disability, stroke impacts all aspects of a person's life and the lives of family and friends. Disability adjustments range from addressing medical and financial needs to physical and psychological adaptations. They also include loss of interpersonal relationships and employment status. Recovery from stroke is a challenging journey that can be life-long. Rehabilitation counselors, working together with a multi-disciplinary team of healthcare providers, can improve the quality of life of the individual and aid his or her family in coping with the complications of stroke.

## **Case Study**

La Shaun Jackson is a 59-year old African American widow living with an adopted son 15 years of age who has a record of substance use and juvenile delinquency. Two married daughters live nearby. She has worked as a claim processor for the Internal Revenue Service in Fresno, California for over five years. Prior to returning to school to earn her Associate of Arts degree in accounting, she held a secretarial position at a small import-export firm for over 15 years. Several long-term clerical and retail store sales positions were listed on her resume before and immediately after she moved from Illinois to California.

Ms. Jackson has had hypertension and chronic headaches for 13 years and has been struggling with obesity throughout her life. Since her husband died five years ago, her weight has continued to increase, as

well as her blood pressure. About a year ago, she experienced a mild hemorrhagic CVA. No surgery was contemplated. Her hypertensive condition has been controlled by medication. As a result of the hemorrhage, La Shaun has hemiplegia on the dominant side (right side) of her body and face; she has moderately slurred speech, and was unable to recognize or identify sensory stimuli, resulting in “neglect” of one side of her body. The trauma caused physical limitations and emotional problems.

A multidisciplinary rehabilitative program began almost immediately after La Shaun’s medical condition stabilized. Rehabilitative therapy started in the acute-care hospital within the first month following the stroke. After eight months of intensive care and physical therapy, Ms. Jackson regained almost 80% of her cognitive and physical functioning. Her physician released her to work in an environment with minimal stress. The position must be sedentary with some time allowed for physical therapy at a nearby clinic.

### **Questions**

1. Provide a vocational profile for Ms. La Shaun Jackson. Identify additional information you need to help this client.
2. How does La Shaun’s age affect your provision of rehabilitation services? Explain your response both for her and other older working individuals. Discuss age discrimination and other stereotypical factors important to this and other cases.
3. Outline the rehabilitation services you intend to provide including when to begin and the time frame for each step in the process.
4. How would you counsel Ms. Jackson if she has mixed feelings about returning to work?
5. Would you counsel her differently if she was 20 years younger?

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# Chapter 17

## EPILEPSY

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### Introduction

The word “epilepsy” derives from the Greek word meaning “to be seized” and is a generic term that refers to a wide variety of seizure conditions. As early as 150 A.D., Galen recognized seizures might originate in the brain and result from some underlying disease (Wanamaker, Brooker, Dreifuss, & Willmore, 1984). Several well-known individuals in history such as Socrates, Leonardo de Vinci, Charles Dickens, Thomas Edison, and Julius Caesar had active seizure conditions. A seizure involves a disruption of the normal activity of the brain through neuronal instability, firing in an abnormally rapid manner. This excessive electrical discharge results in a seizure. A seizure may be confined to one area of the brain (partial seizure) or take place throughout the entire brain (generalized seizure).

The extent to which seizures affect brain functioning depends on both the duration and location within the brain. Consequently, some seizures impair brain functioning slightly, while other seizures result in a complete cessation of normal activities. Risk factors include hypertension, substance abuse, depressive illness, and being a member of a lower socioeconomic group (Deray, Resnick, & Alvarez, 2004). Direct causes of epilepsy include traumatic brain injury, birth trauma, anoxia (insufficient oxygen), brain tumors, infectious diseases in a pregnant woman, parasitic infections, vascular diseases affecting the brain’s blood vessels, and substance abuse. Only a small portion of people with epilepsy (1-2%) have a diagnosable genetic etiology for their seizure occurrences (Anderson, 1988).

A seizure is a discrete event and is a symptom of brain dysfunction; it takes many seizures to establish a diagnosis of epilepsy. Epilepsy is a chronic disorder that involves recurrent and unprovoked seizures (Falvo, 2009; Fraser & Miller, 2005). The incidence of recurring (more than one) seizure or epilepsy in the general population is between 1-4% and varies with age.

### Symptomatology

Seizures are generally classified by review of clinical symptoms and supplemented by electroencephalogram (EEG) and sometimes videotape monitoring. There are two major types of seizures:

1. Generalized seizures that affect both cerebral hemispheres.
2. Partial (focal) seizures that affect a specific part of one cerebral hemisphere.
  - a. Simple partial seizures during which consciousness is not impaired.
  - b. Complex partial seizures, with one or more “complex” symptoms during which consciousness is impaired.
  - c. Partial seizures, which may evolve into secondary generalized seizures.

The classification proposed by the International League Against Epilepsy (ILAE) Commission on Classification and Terminology, published in 1981, is provided in Table 1. This Table gives a basic overview of the different types of seizure conditions.

**Table 1**  
**An Abbreviated Classification of Epileptic Seizures**

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**I. Generalized Seizures of Non-Focal Origin**

1. Tonic-clonic
2. Tonic
3. Clonic
4. Absence
5. Atonic/akinetic
6. Myoclonic

**II. Partial (Focal) Seizures**

1. Simple partial seizures with elementary symptomatology (consciousness is not impaired)
  - a. With motor symptoms (including Jacksonian, versive, and postural)
  - b. With sensory symptoms (including visual, somatosensory, auditory, olfactory, gustatory, and vertiginous)
  - c. With autonomic symptoms
  - d. With psychic symptoms (including dysphasia, dysmnnesia, hallucinatory, and affective changes)
  - e. Compound (i.e., mixed) forms
2. Complex partial seizures with complex symptomatology (consciousness is impaired)
  - f. Simple partial seizures followed by loss of consciousness
  - g. With impairment of consciousness at the outset
  - h. With automatisms
3. Partial seizures evolving into secondarily generalized seizures

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***Generalized Seizure Conditions***

These types of conditions tend to involve both cerebral hemispheres and several areas of the brain (cerebral cortex, thalamus, and brain stem structures). They are sub-categorized into two major types: (a) tonic-clonic seizures and (b) absence seizures.

***Tonic-Clonic Seizures***

The most common form of generalized seizure is the tonic-clonic convulsion (formerly known as grand mal seizure). Generalized tonic-clonic seizures occur in 10% or less of the population who have epilepsy. It is the stereotypic seizure with which most people are familiar. This type of seizure involves two stages. The tonic stage, during which the body becomes rigid, lasts for a few seconds. A clonic stage follows during which the person endures a series of convulsive and jerky movements. The entire seizure generally lasts about one to three minutes (Devinsky, 2002).

From a rehabilitation perspective, the severity of the seizure may leave some individuals disoriented and fatigued following cessation. Some clients can return to work immediately while others may require a full day of rest before returning to work activity. Tonic-clonic seizures, for some people, occur only in their sleep or early in the morning when they first awaken. Often, this does not interfere with the ability to arrive on time to the job site. Although most tonic-clonic seizures last for just a few minutes, an individual may enter an emergency state called “status epilepticus.” This involves a continuing, prolonged seizure, exceeding five to ten minutes, or recurring seizures within a brief period. This is rare but an emergency situation when it does occur, requiring immediate hospital care.

First aid for tonic-clonic seizures involves turning a person's head to one side and putting a soft article of clothing under the head. Clearing the area can be helpful, but it is unnecessary and even dangerous to try and insert something into the person's mouth.

### ***Absence Seizures***

The other commonly known type of generalized seizure is the simple absence seizure (traditionally known as petit mal). These seizures take only a few seconds and involve a brief disruption of consciousness (10-20 seconds). There are mild rhythmic movements of the eyelids and autonomic symptoms, such as pupil dilation, although some involve blank stares (Devinsky, 2002). Many patients with this type of epilepsy begin having these seizures before age 12. This category involves less than 5% of epilepsy cases (Hauser, 1997; Penry, 1986). They are often not identified for years and eventually may transition into generalized tonic-clonic seizure conditions. Early treatment, therefore, is essential. From a functional perspective, absence seizures may be minimally vocationally limiting, especially if there is no memory loss experienced. Exceptions occur when an individual is experiencing many seizures of this type throughout the day or has a physically high risk occupation, where even brief losses of consciousness affect job performance.

### ***Other Types***

Other types of generalized seizures include tonic or clonic seizures that are limited tonic-clonic seizures. In a tonic seizure, the person falls to the ground and experiences rigidity without convulsive movements. Conversely, in a clonic seizure, the person experiences convulsive activity without the earlier period of rigidity. Another type of generalized seizure is atonic akinetic seizures or brief drop attacks, which tend to affect children under five years of age. Adolescents and adults can experience generalized myoclonic jerks that are brief shock-like contractions and affect the entire body or only a part of the body.

## ***Partial (Focal) Seizures***

As represented in Table 1, partial seizures are divided into three categories: (a) simple partial seizures with unimpaired consciousness; (b) complex partial seizures with impaired consciousness; and (c) partial seizures evolving into secondarily generalized seizures. Approximately 60% of those with epilepsy have seizures classified in the partial seizure category (Deray, et al., 2004; Leppik, 1998). From a first aid perspective, observation is most important and blocking access to any area of possible danger. Each of these categories is discussed below.

### ***Simple Partial Seizures***

These seizures may be motor, sensory, autonomic, or involve a combination of symptoms without impaired consciousness. They are linked specifically to an affected area of the brain. Many simple partial seizures last less than 30 seconds and, due to unimpaired consciousness, are often not a significant problem in regard to simple job performance. Some individuals who experience partial seizures have time to pull over to the side of the road while driving a vehicle, and then proceed when the seizure ends. They function quite well in other life activities, including work.

### ***Complex Partial Seizures***

Impairment of consciousness accompanies complex partial seizures; these seizures are usually associated with temporal or frontal lobe foci. They often have an associated aura or warning that can involve a strange odor, aphasia, dizziness, nausea, headache, unusual stomach sensation, or a déjà vu experience. Common events include the individual having repetitive motor movements, fumbling with one's hands or clothing, lip-smacking, or aimless wandering. Complex partial seizures without motor components are less common. When they do occur, they can involve impaired consciousness and complex symptoms such as rapid sensory or emotional changes (some have been mistaken for psychiatric conditions).

From a functional perspective, complex partial seizures can be impairing due to loss of consciousness and the odd symptoms expressed by the person. It is fortunate when an individual has a brief aura or warning before a seizure which allows him or her to take safety precautions or alert others of the oncoming seizure.

### ***Partial seizures Evolving into secondary Generalized Seizures***

Some individuals have partial seizures, which, if not controlled, spread into generalized seizures. These persons are often not appropriately treated for their partial seizures, but only for the later observed generalized seizure activity (e.g., a tonic-clonic seizure).

### ***Psychogenic/Nonepileptic Seizures***

As opposed to seizures, these events (although often mimicking seizures) are panic attacks, conversion disorders, and dissociative events. The most definitive way to diagnose nonepileptic seizures is long-term video/EEG monitoring.

## **Diagnosis and Treatment**

### ***Diagnosis***

When individuals first experience seizures, they usually consult their personal or primary care physician. Diagnosis may involve hospitalization; however, if the seizure occurred several days previous, an outpatient evaluation may be adequate (Deray et al., 2004). The physician may begin a medical treatment program or refer the individual to a general neurologist. If the person does not achieve seizure control within three months, the personal or primary care physician should recommend a general neurological referral (National Association of Epilepsy Centers, 1990).

Neurological consultation includes a physical examination and patient history, metabolic studies, and other evaluations including EEG testing. The EEG provides the physician with a clearer definition of the nature of the abnormal neuronal discharging, often confirming a seizure diagnosis. When the diagnosis remains unclear, the physician uses more specialized and noninvasive techniques, such as computerized tomography (CT) scans or magnetic resonance imaging (MRI) of the head region. MRI is a technique useful for identifying small lesions or cerebral cortex abnormalities. It is generally superior to the CT scan. MRI scans provide better information in cases where bone interference hinders CT scans; they are more sensitive to certain tissue. Other scanning techniques such as single photon emission computed tomography (SPECT) or positive emission tomography (PET) can be helpful. Multi-channel magneto-encephalograph (MEG) is a new technique for measuring magnetic fields and is more definitive in locating seizure discharge sites.

As recommended by the National Association of Epilepsy Centers (1990), when the general neurologist does not achieve seizure control within nine months, referral to a tertiary or fourth-level epilepsy center should be made. These centers are staffed with neurologists who specialize in epilepsy treatment and are located within an allied health center that has specialized teams devoted to the medical and psychosocial needs of persons with epilepsy. The National Association of Epilepsy Centers recommends that referral to a center be made based on continuing indications of difficulty. These include pharmacological problems; possible psychogenic or pseudoseizures; the potential for epilepsy surgery; the need for invasive, intracranial video/EEG recording; and the need for complementing psychological or psychiatric expertise.

### ***Treatment Principles***

As indicated by the Epilepsy Foundation (EF) (Deray et al., 2004), anti-epileptic medications are selectively effective for one or more different types of seizures. The neurologist attempts to match the appropriate medication to the specific seizure type. Table 2 reviews the primary and secondary drugs most effective for different seizure categories. In the 1990s (called the “decade of the brain”), there were a number of new compounds approved by the FDA including felbamate and gabapentin in 1992, Lamotrigine in 1994, topiramate in 1995, tiagabine in 1997, and levetiracetam in 1999. Since then, zonisamide, oxcarbazepine, and pregabalin were approved (Devinsky, 2002). Although a number of these newer medications were developed as adjunctive or “add on” drugs, over time the majority have been established as monotherapeutic for generalized seizures.

It is desirable that individuals use one medication (monotherapy), whenever possible, to control symptoms. One drug is often the most effective, easier to manage, and has less potential toxicity. To achieve optimal daily life functioning for a person, the usual course of treatment is to prescribe the maximum effective tolerable dosage of one medication. Brodie and Kwon (2002) indicated that 47% of patients



**Table 2**  
**Seizure Types and Indicated Antiepileptic Drugs**

<u>Seizure Type</u>	<u>Effective Antiepileptic Drugs</u> (Listed alphabetically)
Simple partial or complex partial	<p><b>Primary Drugs</b>            Carbamazepine (Tegretol, Carbatrol)            Lamotrigine (Lamictal)            Levetiracetam (Keppra)            Oxcarbazepine (Trileptal)            Phenytoin (Dilantin)            Pregabalin (Lyrica)            Topiramate (Topamax)            Zonisamide (Zonegran)</p> <p><b>Secondary Drugs</b>            Clonazepam (Klonopin)            Felbamate (Felbatol)            Gabapentin (Neurontin)            Methsuximide (Celontin)            Phenobarbital (Luminal)            Primidone (Mysoline)            Tiagabine (Gabitxil)            Valproic acid (Depakene)</p>
Generalized tonic-clonic (primary or generalized secondary)	<p><b>Primary Drugs</b>            Carbamazepine            Lamotrigine            Levetiracetam            Oxcarbazepine            Phenytoin            Pregabalin            Valproic acid            Zonisamide</p> <p><b>Secondary Drugs</b>            Felbamate            Gabapentin            Phenobarbital            Primidone (Mysoline)</p>
Generalized absence	<p><b>Primary Drugs</b>            Ethosuximide (Zarontin)            Lamotrigine            Levetiracetam            Topiramate            Valproic acid            Zonisamide</p> <p><b>Secondary Drugs</b>            Acetazolamide (Diamox)            Clonazepam            Felbamate</p>
Myoclonic	<p><b>Primary Drugs</b>            Lamotrigine            Levetiracetam            Topiramate            Valproic acid            Zonisamide</p>

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responded to the first antiepileptic medication, 13% to the second, but only 1% to a third monotherapy choice. By adding a second medication, only 3% achieved control. A dosage should be established that maintains clinically effective concentration within the blood throughout the day. If people forget to take their medications at scheduled intervals, for some, it is better to take the dosage all at once (e.g., before bed). Serum levels are sometimes useful, and major recommended drugs have therapeutic ranges and toxicity levels. Table 2 overviews pharmacological data on the major antiepileptic drugs.

The time to reach steady-state, the appropriate drug-blood serum level, varies across medications relative to number of days used. The physician takes periodic blood level readings of the antiepileptic medication to ensure the person is maintaining an adequate dose for seizure control and has not reached an excessive concentration. “Toxic” levels of medication (excessive drug concentrations) can result in double vision, lethargy, cognitive impairment, coordination difficulties, weight gain, and other medical complications. Medication levels require periodic laboratory monitoring for assessment of appropriate ranges. Even within appropriate ranges, drug side effects may require medical intervention. Non-compliance with medication is a major issue and, in some communities, up to 90% of patients can be found with medication levels below therapeutic ranges (Leppik, 2006). Blood level monitoring, therefore, is important – particularly for those who have cognitive problems.

Another treatment approach which is used if medications or even epilepsy surgery are not effective, is the vagal nerve stimulator. The vagal nerve stimulator is implanted in the chest and stimulates the vagal nerve at an established cycle and at seizure onset. Approximately 50% of those using the stimulator achieve >50% seizure reduction, 5% to 8% are seizure free, and 30% have some benefit with seizure frequency reduced <50% (Devinsky, 2002). For some people, particularly those trying to work, this can make a difference.

### ***Surgery***

There are an increasing number of epilepsy centers and neurosurgical groups around the country that perform surgical operations for uncontrolled epilepsy. Surgery is considered only when thorough trials of appropriate antiepileptic medications failed in controlling an individual’s seizures. When seizures are impairing independent living, school performance, work capacities, or social activities in a substantial way, the physician may recommend a surgical consultation.

For surgery to occur, there must be a clear lesion (tumor or growth) on the surface of the brain cortex that can be surgically removed without producing substantial negative results. Although surgery can be helpful in reducing seizures, individuals may be left with significant language, memory, and other functional deficits. A series of diagnostic tests that include neuropsychological and continuous EEG/video monitoring are conducted so that the medical staff and the person have a clear understanding of the absolute need and prognosis for surgical intervention. Some individuals and families choose to accept potential minor memory or language deficits for possible seizure-free status achieved through surgery. As physicians choose patients more selectively for surgical procedures, cognitive deficit problems after the operation may be avoided or diminished. Individuals then may experience a symptom-free life.

Results of temporal lobe surgery throughout the country suggest that up to 80% of patients achieve a seizure-free or almost seizure-free outcome, with the remaining 20% having a moderate or less beneficial reduction (Engel, Wieser, & Spencer, 1997). Schaul (1987) estimated there might be up to 120,000 people within the United States who could benefit from this type of surgery. Other treatment alternatives exist, such as the ketogenic diet for children (Guillard, Shilden, Stafstrom, & Vinding, 1991), but this is not generally observed in adult treatment.

### ***Medical Prognosis for Patients with Epilepsy***

Annegers (1988) indicated that 10 years after epilepsy diagnosis, 65% of individuals seen are in seizure remission, and at 20 years, 76% (of the 65%) are still in remission. Long term follow-up of childhood epilepsy (Sillanpää & Schmidt, 2006) noted that 67% eventually achieved seizure control. The most important prognostic indicator for the eventual control of seizures is the duration of seizure occurrence. Other factors include seizure cause, seizure type, and age at onset. Seizure reoccurrence can be assessed in relation to established risk factors.

## Functional, Psychosocial, and Family Issues

### *Functional Issues*

Unless their seizures are due to stroke or other acquired brain impairment, clients with epilepsy generally have no physical limitations; they are fully independent in eating, dressing, walking, personal hygiene, and communication. At times, adverse effects of antiepileptic medication may impair the individual's physical and intellectual functioning. Even low-grade toxicity can produce awkwardness of gait, coordination problems, and eye-focusing difficulties. Other adverse effects include problems in physical functioning, memory, and attention span. These effects are resolved with alteration in drug regimen.

### *Psychosocial Issues*

Although epilepsy is a hidden disability, people with this disorder have many of the same insecurities and fears as with visible disabilities (Dell Orto & Power, 2007; Fraser & Miller, 2005). They may be dealing with one or more of the specific emotional problems that occur from having a seizure disorder. There are three prominent adjustment problems persons with epilepsy face:

1. Epileptic seizures are episodic; the person is functioning normally one minute and totally nonfunctional the next. This is not true for persons with orthopedic or audiovisual/speech disabilities, which may be more limiting than epilepsy, but allow affected individuals opportunities to develop adaptive reactions to their disorders. For the person with an active seizure condition, there is the possibility of an occurrence at any time. Because the time, place, and circumstances are unknown, the individual lives with a great degree of ambiguity, fear, and anxiety.
2. The epileptic seizure itself is often an alien, unusual, and frightening spectacle to observers (some partial seizures such as sensory will go unobserved). This is a reason epilepsy carries the stigma it does. A convulsion is a traumatic and unknown event which can terrify the observer.
3. The person experiencing the convulsion is not in control. Not only does the individual usually lose control during the seizure, but lacks control of the initiation of the seizure. All societies desire and respect predictability and regularity. This is particularly true in the United States, a society that stresses self-control and responsibility.

Many clients benefit from individual counseling. Group sessions are beneficial; however, cognitive level and emotional integration need to be considered when determining the appropriateness of group counseling. It can be emotionally helpful to discuss issues with others having the same disability and similar experiences (e.g., epilepsy self-help groups).

### *Family Response to Epilepsy*

Family response to the child or adolescent with epilepsy has a great impact on the young person's outlook. As part of the assessment process, it is helpful to evaluate an individual's developmental history. The following illustrates common family response styles:

#### *Overprotective*

The family is unwilling to risk any physical or emotional damage and is reluctant to set reasonable expectations for the child, expectations that might otherwise encourage a youngster to reach maximum social, educational, and vocational potential.

#### *Reality-Oriented Action*

This family accurately assesses the young person's abilities and potential, without having their fears and anxieties hamper growth.

#### *Spartan Orientation*

The spartan oriented family believes the child with epilepsy progresses further if strongly pressed to achieve.

### ***Rejecting***

The family believes itself to be defective because it produced a “deviant” child. The child can internalize the depression and anxiety the family feels.

The situation is different if seizures begin in adulthood. The individual may have developed a solid self-concept and benefited from a supportive home and work environment. Yet, emotional adjustment and denial of disability may occur for all people with epilepsy, as is true for any major disability (Saunders, Leahy, McGlynn, & Estrada-Hernandez, 2006). It can be more frequent in epilepsy as the disability is “invisible” most of the time.

## **Vocational Limitations and Rehabilitation Potential**

### ***Seizure Status***

A careful review of functional seizure-related considerations is helpful when determining employment potential. For those who do not achieve seizure control, employability is most affected by a combination of seizure type and related symptoms, duration, frequency, pattern of occurrence, as well as potential job. For example, a person can have generalized tonic-clonic seizures on a weekly basis, but have them nocturnally (at night) in a way that does not affect employment. The treating physician, at regular intervals, evaluates therapeutic blood levels of medication and assesses compliance. The review by Thorbecke and Fraser (in press) related to vocational needs and services in epilepsy indicates that although unemployment for adults with epilepsy is two to three times the general population, it increases with active seizure conditions. Underemployment, however, is a consistent issue worldwide.

### ***Neuropsychological and Psychosocial Assessment***

As discussed earlier in this chapter, seizures are not always the primary issue. Concerns about brain functioning and psychosocial adjustment are prevalent among those referred for specialized epilepsy medical and vocational assessment. Dodrill (1978) established a comprehensive battery of 16 neuropsychological measures directly related to brain impairment and epilepsy. Additionally, psychosocial adjustment difficulties are identified through use of the Washington Psychosocial Seizure Inventory as an initial screening device (Dodrill, Batzel, Queisser, & Temkin, 1980). This inventory is similar to the Minnesota Multiphasic Personality Inventory-II (MMPI-II) that is used with other tests to assess different areas of psychosocial maladjustment. Unlike the MMPI-II, it was developed and normed using an epilepsy population, and has been translated into more than 20 languages. Although a number of quality of life instruments have been developed, none are chiefly used for addressing points of psychosocial intervention. Standardized psychological or psychiatric interviews are helpful in clarifying mental health and psychosocial concerns.

Batzel, Dodrill, and Fraser (1980) established that the test battery differentiated very well between the unemployed (64% of tests outside normal limits), the underemployed (53% of the battery outside normal limits), and the employed (only 22%). Individuals with known brain injury, other neurological insult, or recurrent severe seizures over a number of years need periodic neuropsychological testing. Testing should be customized when possible to specific job goal demands (e.g., aspects of memory, problem solving, speed of information processing, motor functioning language). Different neuropsychologists at various centers will utilize different batteries.

### ***Work History and Prior Training***

The rehabilitation counselor carefully reviews employment history and determines reasons for any job changes or termination. At times, the individual had adequate abilities but lost employment due to discrimination because of occasional seizures at work. Conversely, some clients identify seizures as the reason for job termination when difficulties at work were related more to lack of abilities or interpersonal functioning. Information from family members, prior employers, and other third parties is helpful in clarifying job performance issues. For some clients with epilepsy and associated brain impairment, on-the-job training and supported employment are more effective avenues to placement than formal academic or technical training. Also, a person’s education may have ended early due to memory retention

deficits and similar problems. On-the-job training is effective for people who have had negative experiences in the school environment (Fraser, in press).

The review by Thorbecke and Fraser (in press) highlights that the amount of time working pre-entrance into a vocational program is a major indicator of employment success. Clients with little pre-program work experience often profit from using the U. S. Department of Labor (1993) waiver for doing unpaid situational assessments (up to 215 hours) prior to competitive placement.

### ***Contextual Considerations***

When beginning vocational rehabilitation services, the counselor should carefully evaluate the client's financial needs (Fraser, in press). Those on federal subsidy such as Supplementary Security Income (SSI), or Social Security Disability Income (SSDI), if facing significant seizure associated impairments, may need to seek part-time work as not to jeopardize their benefits. They may need to use either the Plan for Achieving Self-Support (PASS) or Impairment-Related Work Expenses (IRWE) work incentive programs to establish their capacity for self-support, not jeopardize living subsidies prematurely, and cover various work-related expenses (deducted from salary). It is helpful to determine how much financial support the family provides. Strong family financial support may curtail incentives for job seeking.

For people with epilepsy, ability to drive is often affected. In some states, this is at the discretion of the physician, based on review of seizure information, while in other states a specific seizure-free period must occur. If driving is not possible, access to public transportation is crucial. In some instances, employers are requested to make shift changes as a reasonable accommodation to better enable access to transportation.

### ***Considerations for the Rehabilitation Counselor***

Each client must be evaluated individually, with job skills and experience carefully assessed. Assets and limitations of clients vary widely. It is not so much the type of seizure that influences the person's functioning level as the frequency of seizures. Frequently occurring absence seizures can pose more problems than well-controlled tonic-clonic epilepsy. The counselor needs to ask these critical questions concerning seizure status (Bishop, 2004; Fraser & Miller, 2005):

- What type of seizures and specific symptoms does the client have? It is helpful to have a clear description of the seizure activity during and after the seizure. If there is a loss of consciousness, how much time is involved?
- How well controlled are the seizures? If the client has active seizures, at what times of day do they occur?
- Does the client have a warning or aura before a seizure? How consistent are the warnings and how much time do they provide the client?
- Are there certain precipitants (e.g., flickering lights, stress, fatigue) to seizure events?
- Is the client on the right type of medication for the seizure type? Many clients are not on optimal medication regimens. Is there consistent medication compliance? Does the client complain of side effects from medication; if so, what are they? How specialized was the medical evaluation? Did a general practitioner, neurologist, or epileptologist (neurologist specializing in epilepsy) perform the evaluation? Might alternative therapies (e.g., vagal nerve stimulator, epilepsy surgery) assist with seizure control?
- When was the last medical evaluation?
- What is involved in the recovery period following a seizure? What is a reasonable time in which a client can return to work?
- Does the client have any other physical, mental, or sensory disabilities? If so, have they been evaluated in the area of disability (e.g., cognitive concerns requiring neuropsychological evaluation)? These issues can be more impairing than the seizures.



The above information is helpful in establishing daily functional capacities. Motor performance can be slowed, primarily in cases involving repeated severe seizure activity over time, but also as a result of side effects or toxicity from medication and other acquired brain impairment.

## **Job Development and Placement**

### ***Service Methodology***

Interpretation of assessment data and individual counseling is helpful. The philosophy of the Epilepsy Foundation (EF) vocational programs is that of shared responsibility between employment services staff and clients. The philosophical emphasis is on personal empowerment and independence.

Most clients benefit from job-seeking skills training provided within a job club format. Job clubs follow the format of the highly successful program developed by Azrin, Flores, and Kaplan (1977) used nationally with hard-core unemployed and rehabilitation populations. The job club setting provides emotional support and encouragement in addition to teaching basic skills and ongoing organization of job search activities. Reinforcing information from individual counseling sessions within the group setting helps assure that clients retain information. Handouts and visual aids are useful, as well as weekly reminders about job club meetings and appointment schedules.

The issue of whether to disclose epilepsy to a potential employer is a topic for discussion within the job club setting. Although an individual may have a seizure condition that does not affect job performance, the majority of clients with epilepsy prefer to disclose their condition to prospective employers. Job seekers with seizure conditions have a responsibility to consider various work-related factors. These include type of seizure, frequency of occurrence, and risks associated with specific job duties. A related issue is whether the person experiences an aura or warning before a seizure occurs. For some with no significant safety issues in the workplace, disclosure is less of a concern (Bishop, 2004).

Recently, at one epilepsy center, a patient who was a medical assistant was terminated from two different medical clinics for failure to disclose epilepsy at the time of being hired. Since the job involved frequent taking of blood samples, the disclosure issue was related to the job. If epilepsy had been discussed at the time of hire, reasonable accommodation may have been possible by having another staff member take responsibility for taking blood samples. Job restructuring or modification may have allowed the person to retain the job.

### ***Disclosure and Accommodation***

People with frequent seizures occurring during the workday generally need to discuss their epilepsy with prospective employers. Some individuals have decided not to disclose their seizure condition on the job application but discuss it and have the information added to their application form during the interview process. This enables them to describe their skills and aptitudes in relation to the job before mentioning their seizure status. Other individuals decide to disclose epilepsy after being hired. There is no one strategy that is appropriate for most people with epilepsy (Fraser & Miller, 2005). The Americans with Disabilities Act (ADA) of 1990 states that employers cannot discriminate because of a person's disability. If the seizure disorder does not affect job performance, the job seeker can choose not to disclose.

Applicants should be able to discuss their seizure conditions clearly, positively, and succinctly, in terms that are understood by the layperson, keeping the information brief and job-related. For example, people who have absence seizures may describe periods lasting a few seconds in which they may look distracted. Following the seizure, they are able to resume work quickly. A job seeker with a seizure condition needs to practice disclosure to present the information comfortably and clearly to prospective employers (Saunders et al., 2006). A successful placement program includes active job development and a job bank from interested companies with integration of employers on an advisory board. Private sector firms and public agencies have enjoyed hiring from specialized vocational rehabilitation programs, but these are not readily available.

## ***Interacting with Employers***

Information that can be presented to the employer to affect positive hiring behaviors includes the following (Bishop, 2004; Fraser & Miller, 2005).

### ***Working Around Machinery***

In consideration of today's safety standards, it is rare that machinery will require special modifications. Plastic guards on hazardous machinery or rubber matting on a concrete floor are examples of inexpensive job-site modifications. In other cases, assigning driving tasks to a different worker produces simple job restructuring. Both job-site modifications and job restructuring are examples of reasonable accommodations.

### ***Attendance and Job Performance***

Most studies suggest attendance and performance records for those with epilepsy are equal to or better than the general working population (McLellan, 1987).

### ***Accident Rates***

Risch (1968) demonstrated the actual time lost due to seizures was approximately one hour for every 1,000 hours worked for individuals with active seizure conditions. Sands (1961), when comparing workers' compensation cases over a 13-year period in New York, indicated that accidents caused by sneezing or coughing on the job were twice as frequent as those related to seizure occurrences. Quattrini et al. (1999) indicated that there were no changes in industrial accident rates for 860 workers tracked 20 years from diagnosis.

### ***Insurance Rates***

Hiring a person with epilepsy does not increase industrial insurance rates. These rates are linked to hazards of specific occupational classifications. Health insurance providers generally link rates to age and gender in larger companies, while among smaller companies the providers usually pool claim experiences and no one employer is penalized. Epi-Hab, a sheltered work system for those with seizure disorders, reports receiving significant insurance premium reductions due to outstanding safety records. People with epilepsy generally refrain from drinking alcohol or using illegal drugs since these aggravate their conditions. Thus, they are often safer on the job than typical employees.

## ***Predictors of Successful Placement***

Factors that influence successful vocational rehabilitation can be considered across different categories: demographic, neuropsychological, and psychosocial. The national Epilepsy Foundation (EF) has found that individuals with less than one seizure per month, 12 years of education, no additional disabilities, and the capacity to drive, are more successful. Other factors associated with employment outcome include the recency and substantiveness of employment (months employed pre-program) before program entry, previous salary earned, and compliance with an anticonvulsant medicine regimen.

A University of Washington program compared early program dropouts to those people successfully securing jobs. Psychosocial variables (increased depression, anxiety, and financial distress) were the best predictors of failure in the program. Lack of adjustment to a seizure condition was also a strong indication of future lack of following through with successful job placement (Fraser & Miller, 2005). Job maintenance, however, was more related to cognitive variables such as visual-spatial problem solving and motor speed (Fraser, Clemmons, Dodrill, Trejo, & Freeloop, 1986).

## ***Existing Programs***

In 1976, the National Epilepsy Foundation established a national network of employment programs. This Department of Labor funded program developed from five original sites, to a network of 14 Epilepsy Foundation of America (EFA)-administered programs in local EFA affiliates. There were also 25 locally funded programs based on the same employment preparation model. The Training and Placement Services (TAPS) program had an overall successful placement rate of 65-75%, including either successful job placement or entrance into a training program. Other program model types include job centers that provide

clients with information about local services, training programs, and job leads; teen centers focusing on the needs of young job seekers transitioning from school to work; and a Skills Training and Employment Preparation Service (STEPS) program to meet the needs of clients with brain impairment and associated retardation who could not work effectively under the other models. This program had a supported employment emphasis. These EF programs no longer exist under a national mantle and only some of these EF affiliates still maintain an employment services presence.

The Rehabilitation Services Administration (RSA) has always advocated for and supported vocational rehabilitation on a national level for clients with epilepsy. Some state-funded vocational rehabilitation programs will assign clients with epilepsy to specialized counselors with particular expertise. Rehabilitation counselors seeking additional information can contact the national EFA as to local resources ([www.epilepsyfoundation.org](http://www.epilepsyfoundation.org)), or for legal advocacy assistance relating to employment. The RSA-sponsored Projects with Industry (PWI) are also a source of placement assistance (see [www.inabir.org/members](http://www.inabir.org/members)).

Intervention spans a wide range in epilepsy vocational rehabilitation. Some clients with clear goals, adequate training and experience, and well controlled seizures may need simple advisement, while others need vocational and neuropsychological evaluation, situational assessment, job placement, and even jobsite support.

## Case Study

Roger is 28 year-old Latino male with partial complex seizures. He is the only child in a strongly traditional Mexican-American family. The family response to his epilepsy tended toward “over-protectiveness.” Since his late teens, Roger’s general practitioner has been treating him with Phenobarbital; yet, adequate seizure control has never been achieved. He seeks vocational rehabilitation services while still suffering from third degree (deep partial-thickness burns) he sustained while working as a sous chef in a neighborhood restaurant. This is the second burn he has experienced while at work. Roger explains that his seizure condition is disconcerting, but he has “more or less” become accustomed to it. Seizures occur once every two to three months.

This individual is a high school graduate with one-half year of college. Results of aptitude testing suggest he has superior intelligence. He has an interest in engineering and completed several years of architectural drafting coursework while in high school. Due to lack of funding, Roger states he was unable to continue his college education.

Presently, Roger is unsure what to do. He is financially compromised and must leave the rental home he shares with roommates to move back with his family. There are no vocational plans and Roger describes minimal social involvement within his community. An avid user of his home computer and the internet, Roger believes he has developed expertise in this area.

## Questions

1. What do we need to know about Roger’s seizure type?
2. What are the issues related to his current medication and medical care? What should the referral sequence be to optimize his care?
3. Is Roger a possible candidate for epilepsy surgery? Explain.
4. Should Roger attempt to return to his job at the restaurant? Support your response.
5. What are the potential vocational rehabilitation counseling issues related to Roger’s situation?
6. Identify community resources that may be helpful to Roger psychosocially.
7. Describe cultural considerations relevant to this case and implications of the family’s overprotective approach to this individual’s epilepsy. Discuss the four “family responses to epilepsy.”

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## Chapter 18

# PSYCHIATRIC DISABILITIES

*George K. Hong, Ph.D.*

## Introduction

Psychiatric disabilities refer to disabilities caused by mental disorders that also are called emotional disorders and mental illness. Individuals classified as having psychiatric disabilities are those who have severe or chronic mental disorders, which typically cause impairments in social and/or occupational functioning. This chapter examines the psychiatric disabilities that are commonly experienced by clients seeking rehabilitation counseling. An overview of the prevalence of psychiatric disabilities, the classification system for these disorders, the assessment format of the *Diagnostic and Statistical Manual of Mental Disorders (DSM)* (American Psychiatric Association, 2000), and characteristics of specific psychiatric disabilities are described. Discussion focuses on schizophrenia, mood disorders, and anxiety disorders. Critical factors in rehabilitation counseling for clients with psychiatric disabilities, and a case example illustrating the issues conclude the chapter.

## Prevalence of Psychiatric Disabilities

According to the Surgeon General's Report on Mental Health (U. S. Department of Health and Human Services, 1999), mental illness is the second leading cause of disability and premature mortality in established economies such as the United States. The exact number of people with psychiatric disabilities is difficult to estimate. Operational definitions of mental disorders have been changing over the years, and various definitions have been used in different studies. This makes compilation of data difficult. In addition, criteria for determining which disorders are severe enough to constitute a disability change from time to time, often depending on social and public policy. The following statistics suffice to provide us with an idea of the prevalence of severe mental disorders. For example, schizophrenia, a chronic and disabling mental illness, has a lifetime prevalence (i.e., the proportion of people in the population who had the disorder at some time in their lives) estimated to be at 1% of any given adult population (Lauriello, Bustillo, & Keith, 2004). This translates into approximately two million adults in the United States, or roughly the population of the state of New Mexico. The lifetime prevalence of mood disorders and anxiety disorders is even higher. Based on the National Comorbidity Survey Replication Study (NCS-R), they are estimated at 20.8 % and 28.8 %, respectively (Kessler, Berglund, Demler, Jin, & Walters, 2005). These authors also estimated that the lifetime prevalence of having any mental disorder is a startling 46.4 %. These numbers underscore the importance for rehabilitation counselors to have a better understanding of psychiatric disorders and disabilities.

## Classification and Definitions of Mental Disorders

Psychiatric disorders have been defined and classified differently over the years. Quite often, the definitions are dependent on particular theories of psychopathology and psychotherapy that are prevalent at that time. Since the 1980s, the most commonly accepted classification system and definitions used by mental health professionals in the United States are those given in the *Diagnostic and Statistical Manual of Mental Disorders (DSM)*, which is presently in its fifth edition (Text-Revision) (DSM-IV-TR) (American Psychiatric Association, 2000). Instead of basing definitions of psychiatric disorders on particular theories, recent editions of the *DSM* have been following an empirical approach. Each psychiatric disorder, or mental disorder as it is called in this manual, is defined by a set of clinically observable criteria. A client who meets

the criteria of a disorder is diagnosed as having that disorder. This approach helped to standardize the labels and criteria used for psychiatric diagnoses.

## **The Multiaxial Assessment Format of the DSM**

In general, clients with mental illness who seek services from rehabilitation counselors have already been diagnosed as having psychiatric disorders and are receiving psychiatric treatment or psychotherapy. Many of them have made sufficient progress and are ready to return to work or participate in vocational training. Rehabilitation counselors need to be familiar with the DSM since mental health clinicians use it for diagnostic purposes.

The recent editions of the DSM use a multiaxial assessment format diagnostic system, which presents a psychiatric diagnosis in five component axes. These five axes can be viewed as five dimensions for describing a client's condition. The different editions of the DSM have modified specific information recorded in some of the axes. However, in general, there have not been major changes across the various editions. To avoid confusion, the rehabilitation counselor should note which edition of the DSM was used when the diagnosis was recorded. This is especially relevant for clients with long psychiatric histories that span the different editions of the DSM.

The five axes in the DSM-IV-TR (American Psychiatric Association, 2000) multiaxial assessment system are as follows:

- Axis I: Clinical disorders and other conditions that may be a focus of clinical attention.
- Axis II: Personality disorders and mental retardation.
- Axis III: General medical conditions.
- Axis IV: Psychosocial and environmental problems.
- Axis V: Global assessment of functioning.

The specific psychiatric disorder or disorders experienced by a client are specified in Axes I and II. If the client is suffering from any major physical illness or condition, this is specified in Axis III. Since the psychosocial and environmental stressors faced by a client are factors to consider during treatment planning, these problems are specified in Axis IV. They include interpersonal problems such as family discord or death of a family member, as well as other issues, including educational, occupational, housing, economic, healthcare, and legal issues. These are factors that the rehabilitation counselor considers during service planning. Finally, an overall description of the client's daily functioning is given in Axis V by means of a scale called the Global Assessment of Functioning (GAF) Scale. The scores on this scale range from 1 to 100, with "0" indicating inadequate information. A score in the 1 to 10 range indicates extremely low functioning, and the 90 to 100 range denotes superior functioning. This score is particularly useful for the rehabilitation counselor to consider during educational and vocational planning for the client. In sum, the five Axes of the DSM give a more comprehensive picture of a client's condition rather than simply stating the psychiatric disorder.

## **Psychiatric Disorders**

The DSM-IV-TR (American Psychiatric Association, 2000) classifies psychiatric disorders into 17 major categories:

1. Disorders usually first diagnosed in infancy, childhood, or adolescence.
2. Delirium, dementia, amnesic (causing amnesia), and other cognitive disorders.
3. Mental disorders due to a general medical condition not elsewhere classified.
4. Substance-related disorders.
5. Schizophrenia and other psychotic disorders.
6. Mood disorders.
7. Anxiety disorders.
8. Somatoform disorders.

**Table 1  
Global Assessment of Functioning (GAF) Scale**

Domain	1 - 10	11 - 20	21 - 30	31 - 40	41 - 50	51 - 60	61 - 70	71 - 80	81 - 90	91 - 100
<b>Symptom Severity</b>	Persistent danger of severely hurting self or others (e.g., recurrent violence) Or serious suicidal act with clear expectation of death.	Some danger of hurting self or others (e.g., suicide attempts without clear expectation of death; frequently violent; manic excitement) Or Gross impairment in communication (e.g., largely incoherent or mute)	Behavior is considerably influenced by delusions or hallucinations Or serious impairment in communication or judgment (e.g., sometimes incoherent, acts grossly inappropriately, suicidal preoccupation) Or	Some impairment in reality testing or communication (e.g., speech is at times illogical, obscure, or irrelevant) Or	Serious symptoms (e.g., suicidal ideation, severe obsessional rituals, frequent shoplifting) Or	Moderate symptoms (e.g., flat affect and circumstantial speech, occasional panic attacks) Or	Some mild symptoms (e.g., depressed mood and mild insomnia) Or	If symptoms are present, they are transient and expectable reactions to psychosocial stressors (e.g., difficulty concentrating after family argument).	Absent or minimal symptoms (e.g., mild anxiety before an exam),  Generally satisfied with life. No more than everyday problems or concerns (e.g., an occasional argument with family members).	No symptoms
<b>Level of Functioning</b>	Persistent inability to maintain minimal personal hygiene.	Occasionally fails to maintain minimal personal hygiene.	Inability to function in almost all areas (e.g., stays in bed all day, no job, home, or friends).	Major impairment in several areas, such as work or school, family relations, judgment, thinking, or mood (e.g., depressed man avoids friends, neglects family, and is unable to work; child frequently beats up younger children, is defiant at home and is failing in school).	Any serious impairment in social, occupational, or school functioning (e.g., no friends, unable to keep a job).	Moderate difficulty in social, occupational, or school functioning (e.g., few friends, conflicts with co-workers).	Some difficulty in social, occupational or school functioning (e.g., occasional truancy, or theft within the household), but generally functioning pretty well, has some meaningful interpersonal relationships.	No more than slight impairment in social, occupational, or school functioning (e.g., temporarily falling behind in school work).	Good functioning in all areas, interested and involved in a wide range of activities, socially effective.	Superior functioning in a wide range of activities, life's problems never seem to get out of hand. Is sought out by others because of his or her many positive qualities.

Source: DSM-IV, American Psychiatric Association, 2000

9. Factitious disorders.
10. Dissociative disorders.
11. Sexual and gender identity disorders.
12. Eating disorders.
13. Sleep disorders.
14. Impulse-control disorders not elsewhere classified.
15. Adjustment disorders.
16. Personality disorders.
17. Other conditions that may be a focus of clinical attention.

There are over 400 diagnostic labels and subtypes described under these general categories. Many of these mental disorders affect a person's social and occupational functioning. Since it is impractical to discuss all these disorders, this chapter focuses on those psychiatric conditions that rehabilitation counselors most frequently encounter. Counselors are advised to refer to the DSM whenever they come across unfamiliar psychiatric diagnoses, since all mental disorders are described in this resource. Additionally, some of the topics covered in other chapters of this book, such as drug abuse, alcoholism, mental retardation, learning disabilities, certain neurological conditions, and acquired brain injury are mental disorders defined in the DSM.

## **Schizophrenia**

Schizophrenia is one of the most common diagnoses of clients seeking rehabilitation services. Symptoms of schizophrenia fall into two main groups: positive symptoms (excess or distortion of normal functions) and negative symptoms (diminution or loss of normal functions) (DSM-IV-TR, American Psychiatric Association, 2000). The positive symptoms include hallucinations and delusions, which are conceptualized as belonging to the "psychotic dimension." They are usually the most evident symptoms observed in the person with schizophrenia. Hallucinations are sensory perceptions that are not based on external stimuli. As an example, the person may hear voices when there is clearly no one speaking. Delusions are false beliefs and can include the idea that an external force is remotely controlling one's actions or that one's thoughts are being monitored by some kind of transmitter. These ideas are often bizarre and clearly discerned to be without grounds by practically everyone else, including those from the same social cultural background as the client. Other positive symptoms belong to the "disorganized dimension," and include disorganized speech and behavior. Disorganized speech is demonstrated by incoherence or tangentiality. At times, it is impossible to understand what the client is attempting to say. Disorganized behavior may range from silliness to agitation and inappropriate behavior. Examples are when an individual acts in an eccentric manner or is catatonic.

There are three common negative symptoms: affective flattening (restricted range of emotions), alogia (restrictions in speech), and avolition (inability to initiate and persist in meaningful activities). For example, the person demonstrates a bland affect and speaks little when being interviewed, may not show any initiative in carrying out daily activities, and withdraws from social interactions. In general, someone with schizophrenia appears to be irrational and incoherent. The person either withdraws or engages in inappropriate behavior, such as dressing and grooming in an unusual or unkempt manner, including having a lack of personal hygiene. In severe cases, the affected individual is unable to take care of daily needs and may need hospitalization.

### ***Phases of Schizophrenia***

Schizophrenia is a chronic disorder that typically causes lifelong impairments (DSM-IV-TR, American Psychiatric Association, 2000). Onset usually occurs during young adulthood (i.e., the late teens and early 20s), and follows a particular course of development. In the prodromal phase, the person starts to deteriorate from previous levels of functioning, and begins to exhibit some symptoms of the disorder. In the active phase, the person exhibits the full symptoms of schizophrenia but is not likely to recognize that he or she has the disorder. The individual's behavior is usually so strange or disturbing that family members or law enforcement officers would bring him or her in for psychiatric help. Many patients require

hospitalization at this stage. In the residual phase symptoms disappear, but the individual still exhibits symptoms in a milder form. To be diagnosed as having schizophrenia, an individual must have exhibited symptoms continuously for at least six months. The course of schizophrenia differs from person to person. In general, its duration is chronic and complete recovery is uncommon (Lauriello et al., 2004; DSM-IV-TR, American Psychiatric Association, 2000), with most clients being unable to function at a self-sufficiency level.

### ***Types of Schizophrenia***

The DSM-IV-TR (American Psychiatric Association, 2000) describes five major types of schizophrenia: paranoid type, disorganized type, catatonic type, undifferentiated type, and residual type. They all share the common symptoms of schizophrenia. The first three types are characterized by their most prominent symptoms, namely paranoia, disorganization, and catatonic behavior, respectively. Undifferentiated type refers to a form of schizophrenia that currently does not display the specific criteria for any of these three dominant symptoms. Residual type is diagnosed in individuals who once had a full-blown case of schizophrenia but currently do not show any major positive symptoms. For some people, this is a transition period to full recovery; for others, the symptoms continue and last for years, sometimes with relapses to active schizophrenic episodes.

### ***Impairments and Treatment***

Schizophrenia is typically treated by medication, along with psychotherapy and life skills training. This may involve both inpatient and outpatient treatment. While medication is useful in controlling the symptoms of schizophrenia and preventing relapse, side effects are a major concern. The conventional antipsychotic medications, also called neuroleptics, often have neurological side effects such as involuntary movements of the tongue, jaw, and extremities; muscular rigidity; or Parkinsonian tremors. Newer medications, called atypical antipsychotics, are less likely to have these effects, but some have been associated with metabolic side effects such as weight gain or new-onset Type II diabetes mellitus (Buchanan & Carpenter, 2004).

In addition to possible side effects of medication, most clients with schizophrenia who are on medication may continue to exhibit residual symptoms, including inappropriate or “silly” affect in some clients, and flat affect in others. Some clients exhibit social withdrawal, occasional paranoid ideation or disorganization, as well as milder forms of other symptoms. Clients whose diagnoses have been changed to “residual” may relapse into active schizophrenic episodes, especially when they are under stress. Although there are occasional exceptions, individuals with schizophrenia are unlikely to fully return to their premorbid level of social and occupational functioning. Many have to work in sheltered workshops or supported employment where they can receive adequate supervision, support, and accommodation. Living with a person with schizophrenia can be very stressful and disruptive for family members. Some clients have to live in supervised housing units where their adaptive functioning and adherence with medication is monitored. Unless they have an adequate support network of family members and helping professionals, individuals with schizophrenia face chronic unemployment and financial problems. It has been reported that people with diagnoses of schizophrenia account for 30% of the homeless population in the United States (Buchanan & Carpenter, 2004; Hong & Ham, 2001).

## **Mood Disorders**

This section focuses on two major groups of diagnoses within the category of mood disorders: major depressive disorder and bipolar disorders. Major depressive disorders involve only depressive episodes. Bipolar disorders involve both manic and depressive episodes.

### ***Major Depressive Disorder***

The most prominent symptom of major depressive disorder is a depressed or dysphoric mood. A mood is described as a prolonged, pervasive emotion that colors a person’s perception of the world; it is not simply a transient emotion of sadness. During a depressive episode, the feeling goes beyond the intensity and duration of a normal reaction to an unpleasant external event. Sometimes, there may not be an



identifiable external event precipitating the feelings. The person with major depressive disorder is preoccupied with feelings of sadness, worthlessness, hopelessness, or guilt (DSM-IV-TR, American Psychiatric Association, 2000). The person often loses interest in practically all activities, including pleasurable events and hobbies, and is not able to concentrate or think clearly. Some people become easily tearful. There are changes in the person's physical functioning and overall condition, such as insomnia or hypersomnia, loss or increase in appetite, and a significant loss or gain in body weight. The individual may feel fatigue and a lack of energy, while others may feel irritable and agitated. Suicidal ideation or preoccupation with thoughts of death is a common symptom (Pine & McClure, 2004). By its very nature, major depressive disorder is a serious condition that causes impairment in social and occupational functioning.

To be diagnosed as having a major depressive disorder, an individual must have experienced symptoms for at least two weeks. A major depressive disorder can last for years, with recurring major depressive episodes occurring between periods of more normal functioning or less intense symptoms. As the DSM-IV-TR (American Psychiatric Association, 2000) noted, follow-up studies suggested that, one year after being diagnosed as having major depressive disorder, 40% of individuals still had severe symptoms, and 20% had residual symptoms. In severe cases of major depressive disorder, the person may demonstrate psychotic features, such as hallucinations or delusions.

### ***Bipolar Disorders***

Bipolar disorders, commonly known as manic-depressive illness, involve manic episodes, hypomanic episodes, mixed episodes, and depressive episodes (DSM-IV-TR, American Psychiatric Association, 2000). During a manic episode, a person demonstrates a distinct period of abnormally and persistently elevated, expansive, or irritable mood. Oftentimes, the person feels euphoric, has an inflated self-esteem, or becomes extremely gregarious, talkative, and overly active socially or occupationally. He or she may feel that their thoughts are racing, and become very distractible or impulsive. During the manic phase, the person is overly energetic and often has a decreased need for sleep. These symptoms are noticeable by other people who will find the person's behaviors to be disturbing. Symptoms of a hypomanic episode are similar to those of a manic episode, except less severe.

In a mixed episode, the person demonstrates both depressive and manic symptoms over a period of time (at least one week). A specific bipolar disorder is defined by the pattern of episodes experienced (Akiskal, 2004b). The diagnosis of Bipolar I disorder is given when the pattern includes a manic episode or mixed episode. For example, the person may have a single manic episode, a single mixed episode, or any of these episodes alternating with a major depressive episode. Individuals diagnosed as having Bipolar II disorders have only experienced hypomanic episodes alternating with major depressive episodes, but not manic or mixed episodes. Similar to major depressive disorders, individuals with severe cases of bipolar disorders may experience psychotic features, such as hallucinations and delusions.

### ***Impairments and Complications in Mood Disorders***

Both major depressive disorder and bipolar disorders cause serious impairment in a person's social and occupational functioning. The majority of individuals with these disorders do not seek psychiatric help by themselves. Oftentimes, the person experiencing major depressive symptoms is too dysphoric or lacking in energy to seek assistance. Many of them ask for help only at the insistence of family members. Depressed patients often seek help from general health care providers (i.e., general practitioners, instead of mental health professionals) (Akiskal, 2004a). Many of them consider their symptoms to be related to physical illness rather than psychiatric illness. Suicide risk is a major concern in patients with depression. For individuals diagnosed with the severe form of major depressive disorder, up to 15% die by suicide (DSM-IV-TR American Psychiatric Association, 2000). However, the actual prevalence of suicide is hard to establish since some incidents, whether successful or not, are treated as accidents rather than as suicide attempts.

Persons experiencing manic symptoms typically do not recognize that something is wrong. They may seek help only at the insistence of family members or employers who find their behaviors disturbing. They may be brought in by law enforcement officers for unruly or bizarre behaviors in public, or for

engaging in activities that are dangerous to themselves or others, such as reckless driving. Other features may include poor judgment, taking on multiple risky business ventures, disregard for ethical concerns, and engaging in antisocial behaviors (DSM-IV-TR, American Psychiatric Association, 2000).

There is a high prevalence of alcohol and substance abuse with individuals who have mood disorders (Akiskal, 2004b). Hence, it is common to encounter clients with dual diagnoses, such as being diagnosed as having both a mood disorder and a substance abuse disorder. One explanation is that persons with mood disorders may use alcohol or other substances as self-medication (i.e., for their sedative, disinhibiting, or stimulating effects). Substance abuse may be a direct result of the symptoms, such as when a person in a hypomanic state uses stimulants to maintain high-energy performance or enhance sexual behavior.

### ***Treatment***

Major depressive disorder and bipolar disorders are treated using medication, psychotherapy, or a combination of both (Hong & Ham, 2001; Merikangas, 2004). Individuals may seek the services of rehabilitation counselors when they are in partial remission, that is, when symptoms have decreased in intensity and have stabilized. A person in full remission has no symptoms. Since mood disorders can involve recurrent episodes, the individual may relapse into a depressive episode or a manic episode in the future, especially with emotional stress. This is a factor to be considered when providing rehabilitation counseling services for individuals with mood disorders.

## **Anxiety Disorders**

A number of disorders are categorized under the label anxiety disorders. This section focuses on three anxiety disorders: post-traumatic stress disorder, panic disorders, and generalized anxiety disorder.

### ***Post-Traumatic Stress Disorder***

Post-traumatic stress disorder (PTSD) is one of the most common anxiety disorders experienced by clients seeking rehabilitation counseling. This disorder gained significant attention in recent decades due to the considerable number of Vietnam War veterans demonstrating its symptoms. The more recent combats in Afghanistan and Iraq brought further attention to the problem. A study reported in the *New England Journal of Medicine* (Hoge et al., 2004) indicated that among the soldiers and marines deployed to Iraq, the prevalence of PTSD is 9.3% for those who had experienced one to two firefights, 12.7% for those with three to five firefights, and 19.3% for those with more than five firefights. In general, similar rates were found for those deployed to Afghanistan, with 8.2%, 8.3%, and 18.9%, respectively for the various firefight experience categories.

Aside from veterans, PTSD is common among those who came to the United States as refugees. For example, many Southeast Asian Americans developed this disorder as a result of their traumatic experience with socio-political unrest and war in their homeland, as well as their sojourn as refugees (Hong & Ham, 2001). In addition to war experiences, PTSD can also be experienced by people who have suffered personal traumas, such as serious life threatening incidents and major crimes of violence. Moreover, it may be caused by large scale upheavals that affect an entire community or region, including natural disasters like earthquakes, hurricanes, or floods and other distressing events, such as riots and acts of terrorism. Prevalence of PTSD in the general community ranges widely, with estimates between 2 to 15%, because exposure to trauma varies in different communities (Pine & McClure, 2004). A current example of PTSD occurring after a terrorist event is the occurrence of PTSD in responders to the World Trade Center disaster and New York City residents (Ganzel, Casey, Glover, Voss, & Temple, 2007).

According to the DSM-IV-TR (American Psychiatric Association, 2000), an individual suffering from PTSD has experienced or witnessed a major trauma or events threatening death or serious injury to self or others. Unlike the normal anxiety reaction to trauma that dissipates with time, a person with PTSD continues to have recurrent and intrusive distressing recollections of the event, such as flashbacks or nightmares. Any cues in the present environment that symbolize or resemble an aspect of the traumatic event arouse intense psychological distress in the person. For example, depending on the actual trauma encountered, one may become extremely anxious by a pat on the shoulder that may bring back experiences

of a physical assault, or by the sound of a jet flying overhead which brings on recollections of a wartime experience. These frequent recollections are disturbing to the individual who will attempt to avoid any stimuli that arouse the recollections, including conversation, activities, places, or people. The person may feel detached or estranged from others, and show markedly diminished interest or participation in daily activities. An individual with PTSD often demonstrates a restricted range of affect, such as an inability to have loving feelings, and a sense of a foreshortened future, such as not expecting to have a career or marriage. Other persistent symptoms include increased arousal, such as insomnia, irritability, emotional outbursts, difficulty concentrating, hypervigilance, and exaggerated startle responses (DSM-IV-TR, American Psychiatric Association, 2000).

To be diagnosed as having PTSD, the person must have experienced symptoms for one month or longer (DSM-IV-TR, American Psychiatric Association, 2000). Clients having the symptoms for over three months are diagnosed as chronic. For clients who have experienced the symptoms for less than one month, the diagnosis is acute stress disorder, which will be changed to PTSD if the symptoms persist. Clients with PTSD seeking rehabilitation services frequently continue to experience symptoms, resulting in impairments and limitations in their social, educational, and occupational functioning.

### ***Panic Disorders***

Panic disorders involve the presence of panic attacks that are very sudden and intense experiences of psychological fear and physiological arousal (DSM-IV-TR, American Psychiatric Association, 2000). A panic attack starts suddenly with a feeling of severe apprehension, fearfulness, or terror associated with feelings of impending doom. During an attack (which lasts only a few minutes), the individual experiences physiological symptoms like shortness of breath, rapid or skipped heart beats, chest pain or discomfort, and choking or smothering sensations. There is a fear of losing control or even dying. Some people may believe they are having a heart attack and call for emergency medical assistance. A diagnosis of a panic disorder is given if a person has recurrent panic attacks, and as a result of them, has a persistent concern of having more attacks, worries about the implications of the attacks, or significantly changes daily activities to deal with the possibility of future attacks. According to the National Comorbidity Survey Replication Study (NCS-R), the lifetime prevalence of panic disorders is estimated to be at 4.7% (Kessler et al., 2005).

There are two subtypes of panic disorder: (a) panic disorder with agoraphobia and (b) panic disorder without agoraphobia. Agoraphobia is defined in the DSM IV-TR (American Psychiatric Association, 2000) as anxiety about being in places or situations from which escape is difficult or embarrassing, or in which help will not be available in the event a panic attack or similar symptoms occur. The person avoids going to certain places or situations, and experiences significant distress or anxiety when these encounters are unavoidable. Agoraphobia also occurs in individuals who have no history of panic disorder. However, it is believed that the development of agoraphobia may be related to a fear of having panic attacks or panic-like symptoms (Pine & McClure, 2004).

### ***Generalized Anxiety Disorder***

Generalized anxiety disorder involves excessive anxiety or worry about minor daily problems. As defined by the DSM-IV-TR (2000), this anxiety is pervasive and chronic, lasting six months or longer. Most patients report they have had symptoms for many years before diagnosis and treatment (Pine & McClure, 2004), and many indicating that they have been anxious and nervous all their lives (DSM-IV-TR American Psychiatric Association, 2000). The lifetime prevalence of generalized anxiety disorders is estimated to be 5.7% (Kessler et al., 2005).

Unlike other forms of anxiety disorders in which there are clearly identifiable objects, situations, or events that the person finds fearful, a person with generalized anxiety disorder is anxious about a number of events or activities. Often, the source of the anxiety is diffuse and difficult to identify. The person may seem to be worried about practically everything, hence the term "generalized anxiety." The DSM-IV-TR describes a number of symptoms of this disorder. First, the person experiences anxiety and worry about a number of events that is far out of proportion to the impact of these dreaded events, and the person finds it difficult to control this excessive anxiety and worry. This fear is manifested by some of the following symptoms: restlessness, feeling on edge, being easily fatigued, concentration problems, irritability, muscle

tension, and difficulty with sleep. Chronic generalized anxiety causes significant impairment in a person's social and occupational functioning.

### ***Complications and Treatment of Anxiety Disorders***

Medication, psychotherapy, or a combination of both, are used to treat PTSD, panic disorders, and generalized anxiety disorder. Clients with anxiety disorders are likely to be continuing to experience some symptoms when referred for rehabilitation counseling. As such, stress in the work or educational setting is a factor that must be considered during program planning. There is a high comorbidity between anxiety disorders and alcoholism and other substance use (Merikangas, 2004). This may be a way in which some patients seek to alleviate their anxiety symptoms. Addiction to prescription medication is another concern.

## **Societal Barriers**

As indicated in the Surgeon General's Report on Mental Health (U. S. Department of Health and Human Services, 1999), while public understanding of mental illness has improved in recent years, stigma continues to be a major social barrier for people with mental illness. The public tends to view people with severe mental illness more negatively than those with physical illnesses (Penn & Martin, 1998). People with mental disorders are stigmatized and negatively stereotyped as unreliable, erratic, irrational, and violent. They are often considered to be undesirable as friends, co-workers, tenants, and employees (Corrigan, 1998; Hingsaw & Cicchetti, 2000).

Stigma and negative stereotypes associated with people with mental illness are deeply ingrained in society. In everyday language, people are called "sick" (i.e., mentally ill) whenever they have committed a heinous crime. This categorical equating of criminal behavior with mental illness is gravely unfair to people with psychiatric disorders. The mass media is another arena in which people with mental disorders are often misrepresented and demeaned (Corrigan, 1998; Hingsaw & Cicchetti, 2000). Negative images of people with mental illness abound, even though there are occasional positive portrayals. People with mental disorders are often the subject of jokes and comedies. Worse still, they may be depicted as violent and dangerous, as evidenced by the labels "psychotic killer" or "psychos" regularly found in publications, movies, and television shows. Although some patients with mental illness may be violent, the general likelihood of violence is low. There is very little risk of harm to a stranger from causal contact with a person with mental illness, and overall, mental illness has an exceptionally small contribution to the total level of violence in society (U. S. Department of Health and Human Services, 1999).

Stigma affects people with mental illness in a number of ways (Hingsaw & Cicchetti, 2000). On the personal front, it may discourage them from seeking help for their conditions or from employment. It affects their self-esteem as well as that of family members and significant others, and adds an additional burden of stress on everyone involved. In the social and occupational areas, stigma has an adverse impact on social relations and reduces employment and housing opportunities. On the public policy level, stigma negatively affects the public's willingness to provide funding for mental health treatment.

Rehabilitation counselors need sensitivity to the detrimental effects of the stigma attached to mental illness when helping clients seek employment (Hong, 1995). It is necessary to take a realistic assessment of the work setting to determine its suitability for a client. While the official policy of a company or agency may be supportive toward people with psychiatric disabilities, co-workers may view these employees with fear and mistrust. Some might fear that people with psychiatric disabilities could become disruptive or violent at work. Others might be concerned that if they accidentally say the "wrong thing," it will cause the person with a psychiatric disability to "break down." While these concerns are relevant in some cases, they are overly generalized to encompass all individuals with mental disorders. Ostracism by co-workers and occasional jokes or remarks by insensitive colleagues, whether intentional or unintentional, may create a hostile and stressful work environment that discourages people with psychiatric disabilities from remaining at work or seeking re-employment.

Rehabilitation counselors need to work with mental health professionals and client advocacy groups to promote an accurate and sensitive image of people with psychiatric disabilities. They should also address this issue on the individual level when working with clients. The client may or may not desire to disclose the



disability to an employer, depending on the particular job setting, degree of recovery, and personal preferences. Counselors can advise clients about the appropriateness of disclosing or not disclosing psychiatric disability. The decision of whether or not to disclose and how much information to disclose, is made on an individual basis. Disclosure depends on whether the client needs accommodation and on the benefits of disclosure. In some work settings, such as a cooperative job program designed for people with psychiatric disabilities, supervisors are aware of a client's psychiatric disability (Hong, 1995; Jacobson & Greenley, 2001). When placing a client in these settings, it is helpful for a counselor to provide specific information about the client's mental disorder to supervisors (with the client's consent), making sure to address the supervisor's stereotypes, fears, and concerns.

## **Social Functioning**

Social skills (i.e., ability to get along with others) are an important aspect of one's employability. Most people with psychiatric disabilities continue to experience and exhibit milder forms of symptoms when being served by rehabilitation counselors. For example, individuals with residual symptoms of schizophrenia may show some eccentricity in affect and behavior, or the appearance of apathy. People with residual symptoms of depressive disorders may be moody, pessimistic, or withdrawn. Some of these symptoms also can be true of people with PTSD and other anxiety disorders. These lingering symptoms may adversely affect the social relationships between people with psychiatric disabilities and other workers. Together with the stigma of mental illness, which may cause others to avoid them, individuals with psychiatric disorders may not be able to develop close or supportive relationships with co-workers. This may affect their morale and motivation to maintain employment.

In view of the importance of social skills in the context of employment, rehabilitation counselors need to evaluate the social functioning of clients with psychiatric disabilities. In some cases, counselors use a team approach and work with the client's other mental health providers (i.e., psychiatrists, psychologists, and clinical social workers), to help the client develop appropriate social skills required for a job setting. In other cases, the counselor helps the client locate other sources of support, such as a support group, to help maintain social skills and employment. Social rehabilitation along with psychological rehabilitation are essential parts of a therapeutic plan.

## **Vocational Functioning**

The mental disorders discussed in this chapter often cause major impairments in a person's occupational functioning. In general, the severity and chronicity of a client's mental disorder relate directly to vocational rehabilitation outcomes. Impairments caused by schizophrenia and its residual symptoms are self-evident. Low motivation associated with depressive symptoms, poor concentration, irritability, and physical symptoms such as fatigue and insomnia, which occur in mood and anxiety disorders, impact job functioning. Physical and emotional stress may precipitate or contribute to a relapse. Rehabilitation counselors need to match the functional level of a specific client with both the requirements of a job and the perceived stress level in the job setting. Counselors must pay particular attention to cyclical or seasonal changes in workload and pressure. For example, in certain industries during holiday periods, high sales activities may become particularly stressful for some individuals. A client may be able to handle the job at regular times when sales are stable, but finds the pressure at sales peaks to be overly stressful. In this regard, conducting a thorough job analysis may help facilitate successful vocational rehabilitation outcomes.

Like job placements for people with physical disabilities, people with psychiatric disabilities may be placed in different types of settings, including sheltered workshops, supported employment, mainstream settings, and other transitional programs aimed at easing the individual's return to full time employment. Major factors to consider are the client's length of absence from gainful employment, current symptoms, present level of adaptive functioning, as well as background factors such as education, prior job experience, and premorbid social occupational functioning. For clients who are high functioning and relatively stabilized (i.e., not at high risk for relapse), placement in mainstream settings can be considered. Some persons may be able to return to work simply with occasional support and guidance. For these clients, the crucial function of the rehabilitation counselor is to help them formulate realistic vocational goals and provide support and guidance during the job seeking process. For clients with major impairments or residual



symptoms, especially those with frequent relapses, sheltered workshops and supported employment may be appropriate choices, since both venues can provide the accommodation, support, and supervision needed.

Supported employment is a model aimed at moving clients to independence via competitive employment. It provides them with assistance such as job coaches, transportation, assistive technology, specialized job training, and individually tailored supervision (Metzel, Foley, & Butterworth, 2005; U. S. Department of Labor, 1993). Clients in supported employment may be placed as individuals or in a team with other clients. Another model involves a small crew of persons with disabilities working and operating as a self-contained business that generates employment, such as housekeeping for office buildings. A variation of this is the small business model where several employees with disabilities work along with others without disabilities.

When helping clients with job placement, rehabilitation counselors need to take into consideration the socio-cultural reality of the job market. Given the increasing cultural diversity of our society, some clients seeking rehabilitation services may need bilingual services in terms of job training and pairing with job coaches, and job settings where their lack of English proficiency will not be problematic. Can a monolingual client who is Chinese speaking or Spanish speaking be able to seek employment in a setting with mainstream English speaking customers? Or can the person work at a place where supervisors and colleagues are English speaking only? What are the realistic chances of being gainfully employed in a non-restrictive setting? Indeed, ethnic minorities with disabilities often face double jeopardy. They face discrimination and limitations as ethnic minorities, and they face discrimination and limitations (both in the mainstream community and the ethnic community) because of their disabilities (Hong, 1995). Those without English proficiency are further disadvantaged because of their need for bilingual services. As such, rehabilitation counselors must be ready to be change agents when necessary and advocate for institutional changes, such as the provision of bilingual-bicultural services for their clients.

When providing services for clients with psychiatric disabilities, rehabilitation counselors should be familiar with medications taken by their clients, and be ready to consult with mental health professionals when there are questions regarding the effects or side effects of medications. Successful vocational rehabilitation requires close collaboration among different providers, especially rehabilitation counselors and mental health professionals. All parties need to work together to monitor and promote the client's progress, to counteract the stressful, demoralizing and negative effects of stigma and social barriers associated with it, and to motivate and build the client's self-confidence in social and occupational functioning. In particular, counselors should focus on helping clients develop realistic and practical vocational plans rather than exposing them to repeated failures by placing them in jobs that are beyond their functional levels. Counselors should help families and significant others develop realistic expectations about client capabilities and not pressure them to "recover" and return to full employment before they are ready. From this perspective, a family-centered rehabilitation approach with active involvement of family members in service planning is a viable option (Roessler, Chung, & Rubin, 2006). At times, one may need to help clients and their families realize the chronic disabling nature of their mental disorders and address their frustration and discouragement.

In recent years, a recovery model is being advocated in the public mental health system (Anthony, 2000; Davidson, O'Connell, Tondora, Styron, & Kangas, 2006; Jacobson & Greenley, 2001). This is a comprehensive model with many elements, one of which is to help clients manage and live with their mental disorders, rather than expecting or waiting for them to be symptom free before returning to full participation in society. The model emphasizes self-determination and input from clients and their family members in the recovery process, and in the design and operation of the mental health service system. Rehabilitation counselors need to keep these factors in mind when working with clients with psychiatric disabilities.

In summary, people with mental disorders face many hurdles in their quest for gainful employment. These barriers can be grouped into three categories. The first includes societal barriers such as the stigma of mental illness and the stereotyping of people with mental disorders. A second relates to the impact of particular mental disorders on a person's social skills and functioning. The third category involves the impact of particular mental disorders on a person's occupational functioning. In general, the first category is primarily concerned with institutional and social environmental issues external to the client, while the

second and third categories pertain to rehabilitation potential. These three groups of barriers are interrelated and interact as impediments to employment of individuals with psychiatric disabilities.

## **Conclusion**

Individuals with psychiatric disabilities are typically living with severe chronic mental disorders which cause psychosocial, physical, and vocational limitations. Stigma and negative attitudes toward people with mental illness are deeply embedded in our society. There are multiple categories of psychiatric disorders; the most common diagnoses discussed in this chapter are schizophrenia, mood disorders, and anxiety disorders. Important rehabilitation factors include the client's social skills, probability of relapse, projected image, development of a practical vocational plan, and the match of functional level with not only work requirements but the perceived stress level of the job. Stigma and negative stereotypes are some of the barriers to successful social and occupational functioning for persons living with mental disorders. It is important to note that physical, vocational, and psychosocial rehabilitation are of equal importance. Professionals working together on an effective, multidisciplinary rehabilitation team to plan, monitor, and promote progress maximize the possibilities of success for individuals with psychiatric disabilities.

## **Case Study**

Mr. Robert Arsenije is a 23 year-old immigrant from Kosovo with chronic schizophrenia. He was first diagnosed with this disorder at age 18. His specific diagnosis is schizophrenia, paranoid type, episodic with interepisode residual symptoms. This indicates he has occasional relapses into episodes of active symptoms and has residual or less severe symptoms between these episodes.

Robert is the second of three children of an intact family. Prior to the onset of his mental disorder, he had a stable and impressive school performance record, including being a member of the high school golf team. When his problems first started, he was a 12<sup>th</sup> grade honor student at a prestigious high school. As the term progressed, he became increasingly reclusive and abruptly quit school in the middle of the school year. Robert reported paranoid ideation, believing that people were reading his mind for evil purposes; he began to neglect hygiene and self-care, and engaged in angry outbursts toward his family members when they confronted him about his poor school performance and issues of self care. When his family felt his condition became unbearable, they brought him in for psychiatric services; Robert was diagnosed with schizophrenia and hospitalized.

After two weeks in the hospital, Robert's condition stabilized and he was discharged for outpatient services, which included daily medication and weekly psychotherapy. While the intensity of his symptoms subsided, he continued to experience residual symptoms. Robert was particularly anxious in the presence of strangers. The idea that people were reading his mind, even though he acknowledged this to be impossible, continued to preoccupy him. After a few months of therapy, Robert re-enrolled in high school, but dropped out after one month. He was unable to concentrate in school, always feeling that someone was reading his mind. Feelings of depression and helplessness about his condition and being rejected by his classmates, who avoided him, predominated his thoughts. Most of his time was spent at home. During the following summer, he attended a GED (General Equivalency Diploma) program and obtained his high school diploma.

After obtaining his diploma, Robert began to feel motivated. With the encouragement of his therapist, he attempted various jobs during the following year, but quit each job after a month or two, indicating that they were boring. These jobs included short order cook, automobile detailer, landscape assistant at a golf course, and golf starter (takes golf reservations, collects fees, and assigns golf starting times). Robert decided he wanted to obtain a college education so that he could secure employment that was more fulfilling. A four-year college within commuting distance from his home accepted him. However, he dropped out before completing the first semester. He denied experiencing stress as a result of attending school full time. Instead, he claimed that he did not like the college or the courses he was taking. After a few months, he re-enrolled, this time in a junior college. During the next three years, Robert periodically took one or two classes. Most of his free time was spent at home, watching television or reading books; there was no one whom he considered a friend.

During this period, Robert had a few relapses into active symptoms, triggered by his refusal to take medication appropriately. On one occasion, Robert simply stopped taking medication, insisting that he had learned to “control” or “ignore” his paranoid thoughts. On other occasions, he stopped taking his medication after family arguments. Each time he stopped taking medication, his symptoms intensified. Fearful that evil people were reading his mind and coming to harm him, he would hide in his room, refusing to see anyone. His family had him involuntarily hospitalized. After resuming his medication, Robert’s condition stabilized. In a few days, he was discharged home and treated with outpatient services.

Through therapy, Robert accepted that his paranoid ideations were irrational, but was still distressed by them. Whether he was in college attending a lecture or at home watching television, he would suddenly feel that someone was reading his mind. These thoughts were frightening, even though he kept reminding himself that this could not happen. Recently, after an active episode followed by hospitalization, his family decided it was too stressful for them to have Robert at home. By mutual agreement, Robert was discharged to a supervised housing unit, where he could receive close monitoring of his medication and daily functioning. For a few months Robert worked in a sheltered workshop, although he did not really want to work there, feeling it was beneath his educational and functional level. He wanted to continue his classes at the junior college and was adamant about obtaining a college degree rather than going through a job training program. While discussing his career goals, Robert alternated between pursuing his high school dream of becoming a “famous scientist,” or majoring in business.

### Questions

1. Assign Robert a vocational profile including age category, educational level, and work history.
2. What effect might his culture have on the rehabilitation process and management?
3. What vocational alternatives can you recommend?
4. How should a rehabilitation counselor help Robert establish and achieve realistic educational goals?
5. How can a rehabilitation counselor better coordinate with mental health professionals to promote this client’s recovery?
6. What measures would you take to assure that Robert complies with his medication schedule?
7. Discuss prejudice and discrimination toward persons with mental illness and how these can be diminished in the workplace.

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## Chapter 19

# SEXUALITY AND DISABILITY

*Leo M. Orange, M. S.*

“After surviving a battle with cancer and an automobile accident which left me paralyzed with quadriplegia, I wondered if anyone would ever see me as a sexual human being and want to love me again” (L. M. Orange).

### Introduction

Sexuality refers to the expression of sexual sensation and related intimacy between human beings and the expression of identity through sex. Sexuality comprises a broad range of behaviors and processes, including biological, psychosocial, emotional, spiritual or religious, cultural, and political. Beginning with our relationship with ourselves, it extends to influence and encompass our relationships with others (Best, 1993). Our relationship with ourselves includes how we feel about who we are as people, as sexual beings, as men or women, and how we feel about our bodies, sexual activities, and behaviors. Relationships with others include friendships and emotional intimacies. Regardless of disability or illness, we are all sexual beings and share the right to experience healthy and satisfying lives.

Over the past 35 years, pivotal legislation has enabled people with disabilities more progress toward gaining their rightful place as equal members of American society. Together, the Rehabilitation Act of 1973, the Individuals with Disabilities Education Act (IDEA), and the Americans with Disabilities Act (ADA) of 1990 have provided added opportunities for the societal inclusion and integration of people of all abilities (Rubin & Roessler, 2008). With such integration, there are enhanced opportunities for sexual expression. Increased access to information and educational material is integral to affirming this progress by recognizing the sexuality of people of all capabilities and disabilities (physical, sensory, emotional, and intellectual), whether they be early or late onset.

Sexuality is both physiological and psychological. From a psychological perspective, sexuality involves expressions of intimacy, affection, caring, and love. When one acquires a disability, concerns and emotional disruptions relating to the disability cause anxiety that can prevent the person from expressing and perceiving these emotions. Injury and chronic disease also often produce decreased self-confidence and self-esteem making it difficult to see oneself as attractive. When a person loses respect for his or her own body, it becomes difficult to express and feel love and affection.

In present society, people with disabilities are often regarded as non-sexual beings (Brodwin, Orange, & Chen, 2004). If we accept sexual expression as a natural and significant part of human life, however, then perceptions that exclude sexuality for people with disabilities deny this basic right of expression (Best, 1993). Perceptions of people with disabilities as non-sexual beings prevents people with disabilities from gaining equal access to information and being accepted as sexual beings.

In recent years, the media has begun to represent people with disabilities as part of mainstream society. In film, television, commercials, magazines, and newspapers people with disabilities are beginning to be portrayed as legitimate people with desires, needs, and capabilities for success. Hahn (1991) illustrated these changes when he wrote that people with disabilities have had a variety of negative experiences (including isolation, alienation, and rejection) and yet have been able to partake in positive pursuits, increasing the quality of their lives and the lives of others. People with disabilities, like all individuals, have sexual desires and needs to fulfill; they are capable of enjoying the pleasures of marriage and other intimate relationships, including child rearing. Those who engage in vocational, educational, and social pursuits become active and productive members of the community and greater society.



## **Gender, Disability, and Sexuality**

Any discussion about people with disabilities having sex is guaranteed to raise a few eyebrows. An individual in a wheelchair is often viewed as an object of pity, not of desire. In our society, the existence of a physical, mental, or intellectual disability has implications of asexuality. For example, for many men, a spinal cord injury that results in impotence is seen as the end of their sexuality. Often, even less dramatic disabilities cause individuals to shut themselves off sexually due to feelings of inadequacy and a fear of rejection (Gill, 1996).

The implications of asexuality are equally devastating across the gender spectrum. While sexual problems may be less obvious for women, cultural demands for perfection can be overwhelming. A disability adds to the already impossible task of living up to unrealistic feminine ideals portrayed by the media (Alexander, 1997). Strong messages from the media teach us that people with disabilities are unattractive and unable to fulfill the role of partner in a relationship. People with disabilities do not fit the image of beauty in society and, therefore, are thought to be incapable of being supportive partners, fathers, or mothers. Statistics show that women with disabilities marry less often and those who acquire disability after marriage have higher rates of separation and divorce (Best, 1993; Brodwin & Chen, 2000).

Women with disabilities experience what is known as “double discrimination,” as they are socially judged based upon both their disability and gender. Traditionally, independence is seen as self-reliance, while disability implies the opposite. It is assumed that a woman with a disability cannot be independent. In societies where the need for personal assistance is translated into a need for “care” (meaning the need to be looked after), people with disabilities find it difficult to take charge of their lives (Danek, 1992).

The images that affect women also condition men. Culturally, the male is supposed to possess the ability to “provide for and protect his mate.” If he has a disability, this capability is questioned. The traditional image of masculinity simply does not apply to men who have disabilities.

A physical disability may be used as a cue to categorize a person as abnormal or different, and becomes a source of stigmatization when used to discredit an individual in a stereotypical manner (Goffman, 1963; Vash & Crewe, 2004). Unfortunately, the cultural stereotype that beauty and physical strength are good pervades society. Men and women are conditioned to desire an unrealistically perfect woman or man who does not have disabilities. Negative stereotypes and stigma are associated with persons who deviate from what is considered a normal physical appearance. Therefore, for each gender, disability can cause the individual to shut off sexually, either to avoid rejection and embarrassment or out of fear and misunderstanding.

## **Sexual Functioning**

Individuals with disabilities and chronic illnesses may have doubts as to whether they can still have children, if their sexual partner will stay with them, whether they will find a partner who finds them sexually desirable, and if they will ever enjoy sex again. Many factors influence the ability of an adult with a disability to be sexually functional and to value sexuality. According to Best (1993), the age of onset and the nature of the disability (e.g., progressive or static, congenital or acquired, long term or recent onset) impact the adjustment process. Previous sexual experience and disability-related characteristics (including mobility, assistive devices, medications, pain, strength, bodily function, and prosthetic devices) need consideration. Additionally, severity of disability, the presence of multiple disabilities, and the attitudes of the individual and others are factors requiring appropriate assessment. People with disabilities need to learn to focus on their strengths and capabilities, not their limitations. Stressors, depression, grieving over loss, and physical problems all play significant roles in the expression of one’s sexuality.

## ***Human Sexual Response Cycles***

Masters, Johnson, and Kolodny (1995) described sexual response as a result of two principal physiological changes: increasing blood flow to various parts of the body (vasocongestion) and increasing muscle tension (myotonia). Specific phases of the human sexual response cycle include excitement, plateau, orgasm, and resolution. These phases correspond to the levels of sexual arousal and describe typical physiological responses.

Immediate signs of sexual excitement include erections of the penis or clitoris and lubrication. As an individual becomes sexually aroused, the heart rate increases, blood pressure rises, nipple erection and an increase in muscle tension throughout the body occurs, and there is often a noticeable “sex flush” in the face, neck, and ears that is associated with increased blood flow or vasocongestion. As the excitement phase moves into the plateau phase, one sees significant increases in blood pressure, sex flush, breast size, respiration rate (heavy breathing), and muscular tension. At orgasm, all physiological responses peak and are followed by a rapid release of muscular tension and return to the pre-excitement physiological level.

With the changes in sexual functioning that may accompany disability and/or chronic illness, individuals may or may not experience erections or lubrication as signs of sexual excitement. A man, for example may not experience erection and ejaculation usually associated with orgasm, or may experience retrograde ejaculation (semen is forced into the bladder instead of through the penis) while a woman may or may not experience contractions in the uterus and around the vagina. An orgasm may or may not be experienced after injury or illness. However, if during the act of intercourse, both partners are attentive, all of the above responses may be experienced (Masters et al., 1995).

Sexual functioning and sensation is the result of complex interactions among the sensory nerves, autonomic nerves and muscles, brain, and to a lesser extent, the voluntary nerves and muscles. While people use voluntary nerves and muscles while making love to enhance their experience and express affection, these parts of the body are not in the mainstream of sexual sensation or response. There are voluntary muscles in the pelvic area in both sexes (around the vagina and at the base of the penis) that contract during orgasm in an involuntary manner. These muscles are controlled by the autonomic nervous system through signals from the voluntary nerves that directly control them.

### ***Arousal and Seduction***

Although sexual arousal is usually considered the beginning of sex, a diverse set of physical responses can initiate sexual intercourse. Often, it begins with a touch. Sensory nerve cells (receptors) anywhere in the body (especially in the erogenous zones) send signals to the brain and spinal cord indicating that sex is possible. Arousal can begin with sight, sound, smell, taste, and even thoughts. In this case, the brain starts the process of activating other parts of the nervous system. Seduction includes those things we do to entice someone into sexual activity (i.e., wearing perfume or cologne, dressing in attractive ways, using make-up, sending flowers and love letters, sharing feelings, and perhaps, just asking for sex). During this stage, we are open to sexual stimulation from all our senses. Sight, touch, sound, smell, taste, imagination, and fantasy all have the potential to create arousal. This potential is dependent upon how the person interprets the messages, and is partially based on prior learning and beliefs about what is sexually stimulating.

After the initial arousal event, one part of the autonomic nervous system, called the parasympathetic system, is activated. Cells of the parasympathetic nervous system, located within the spinal cord, send muscle-relaxing chemicals down their fibers. These chemicals dilate blood vessels in the genital region in both sexes, causing erection of the penis in men and clitoris in women. In women, the parasympathetic nervous system releases chemicals that cause the vaginal walls to secrete a lubricating fluid. All these events can be thought of as preparing the body for intercourse (Masters et al., 1995).

### ***Monitoring by the Brain***

The brain receives signals indicating that arousal and readiness are occurring. At this point, the brain either enhances or inhibits the process by the thoughts and images it receives and processes. Thoughts like “I should not be doing this” have a negative affect and begin reversing arousal. Thoughts like “I really enjoy his or her company” do the opposite.

### ***Orgasm and Surrender***

After a brief period of time (usually no more than a few minutes), arousal changes. The sympathetic nervous system gains control over the parasympathetic and begins to set in motion the process leading to orgasm. Cellular products of the sympathetic system that play a role in the flight or fight response increase blood pressure, send blood to the skeletal muscles, and inhibit digestion. Their function during the sex act is

a specialized one. In men, sympathetic nerve fibers carry chemicals that cause contractions in structures within the pelvic area, moving semen from these areas to the penis. In women, these cellular products stimulate the uterus and vagina, causing them to contract.

Usually, within minutes after sympathetic activation, muscles in the pelvic and genital areas in both sexes contract rhythmically for several seconds. These contractions, which are extremely pleasurable, move semen from the penis to outside the body in men. Their function in women, other than pleasure, is to move semen up the canal. Some of the muscles that contract during this phase are voluntary muscles triggered by the autonomic nervous system rather than by conscious intention in a process that resembles shivering. An example is when the body is very cold; the autonomic nervous system activates motor nerves that activate muscle movements thus increasing body temperature.

During this stage, desire and surrender are important components of achieving orgasm. Orgasm requires that the individual momentarily surrender and partially give up control. This requires trust in oneself and one's partner. Trust is the process of letting another be a part of your experience and is especially crucial when experiencing sex for the first time after acquiring a disability or chronic illness. The ability to communicate with one's partner about the physical and mental changes that are occurring during the sexual experience is paramount. A new sexual experience can be positive as well as negative, and how one feels needs to be expressed. Like everything else in life, this process is something that should be experienced and modified as needed. Though perfection may not be achieved the first time, the experience can be adapted if the desire exists for a fulfilling sexual life (Masters et al., 1995).

## **Psychological Perspectives**

Sexuality is a key component of human nature which can be difficult to express and communicate for people with disabilities. Reduced sexual functioning and feeling, body image concerns, and doubts as to how to negotiate the sexual act because of lack of knowledge or physical incapacity contribute to difficulty (Cash 2004). People with disabilities may suffer from reduced opportunities for sex for various reasons, including lack of privacy. Finding ways to express one's sexuality can thus be a vital part of rehabilitation in the case of physical disabilities acquired during adulthood.

Inability to move or perform in the same manner as before does not imply an inability to please or to receive pleasure; absence of sensation does not correspond with an absence of feeling. A person with a disability still feels desire even though their cognitive abilities, lower or upper body extremities, or genitals function differently. Ability to enjoy intimacy, passion, and closeness persists even with an inability to perform certain sexual activities. Although it may be different from the one shared prior to the disability or chronic illness, individuals with disabilities can still enjoy loving, close, and intimate relationships.

Physical changes occur as a result of disability and chronic illness. Psychological factors such as emotional stress, depression, and grief may diminish interest in sex and create performance anxiety. Pressures of not being able to work or feelings of being a burden affect one's feelings of sexuality. An individual may believe that the disability or illness has changed the way he or she looks and feels causing that person to be less independent with a restricted ability to play the "traditional" role in the relationship. For others, pain can make sexual response unpredictable and make uncomfortable what once felt stimulating. Bladder or bowel incontinence causes embarrassment and anxiety, and an indwelling catheter diminishes sexual interest. In addition, weakness, fatigue, and breathing difficulties become barriers to sexual activity (Thomas, 1992). To enjoy one's sexuality, an individual must have the opportunity to share and communicate feelings with others. The potential to experience and enjoy one's sexuality becomes broadened whether one desires to have or not have a sexual encounter.

People with disabilities learn new ways of viewing relationships. With the intervention of medicine and sexual aides such as injections, implants, and pharmaceuticals such as Viagra, a man with a disability has greater chances of having an erection and intercourse. Regardless of medical intervention, however, a person cannot enhance a relationship unless he or she surpasses the fears and insecurities that come with newly acquired physical limitations.

Men rarely discuss sexual difficulties with friends, acquaintances, or physicians. It is often the case that a man requests treatment from an urologist at the insistence of a partner, perhaps after years of repressed

depression, decreased self-esteem, and denial (Nusbaum & Hamilton, 2002). During classic denial, the pain of the perceived loss of “manhood” may be too great to face or express. Perhaps what is at issue is not the male sex drive or sexuality, but the fact that men are raised to believe that there are specific ways to express their emotions. Sexual intercourse may be one of the few permissible ways for a man to be close to someone. Due to cultural values and belief systems, for many men it is the only acceptable place to express feelings of love. This may be the result of deep cultural fears of women’s sexual passion, non-traditional roles, culture, and issues of power and status.

Women who have disabilities, on the other hand, are viewed as unfit to fulfill the traditional roles of mother, wife, homemaker, nurturer, and lover, and are seen as asexual (Brodwin & Frederick, 2006; Cash & Hrabosky, 2004; Fine & Asch, 1988). Although the traditional image of the mother as the sole caretaker and nurturer of her children is changing, it remains the predominant social conception. Since women with disabilities are seen as helpless, dependent, and in need of being taken care of, it is difficult for many to accept a woman with a disability as a potential mother and provider.

## **Social Components of Sexuality and Disability**

One of the major difficulties facing the study of sexuality and disability has been the tendency to view disability strictly from a clinical or biological rather than a social perspective. Although there has been much discussion about social issues in the literature on disability, most of these have been considered only within the context of particular physical impairments (Goffman, 1963). Since the concept of disability encompasses social as well as physical components, there is a need to devote increased attention to the social problems that may affect sexual relationships involving people with disabilities.

Despite the progress achieved in today’s society, the continued lack of recognition of sexuality as part of the lives of people with disabilities continues. Assuming that people with disabilities do not need to express themselves as sexual beings denies them the opportunity to develop healthy identities in relation to sexuality. Beauty and strength are perceived as integral, beneficial components of life in American culture (Vash & Crewe, 2004). At the interpersonal level, persons who are physically attractive are evaluated on the basis of their sexuality. A physical disability thus leads to the categorization of a person as abnormal. When it is used to discredit an individual in this stereotypical manner, it becomes a source of stigmatization (Arokiasamy, Rubin, & Roessler, 2008).

Pervasive social norms reinforce taboos against sexual contact and interaction with people who have disabilities (Brodwin & Orange, 2008). Often, physical attractiveness alone is considered paramount in the development of relationships. This emphasis may reflect traditional values that are attached to physical strength, power, beauty, and grace. There is a coexisting significance to the terms “the whole body,” or “the body beautiful,” and the narcissistic anxiety that is easily aroused in many people who are plagued by doubts about their own supposed physical flaws or defects.

## **Disability and Sexual Identity**

Interactions between people with and without disabilities are severely tested by attitudinal and architectural barriers. Many common social and recreational patterns that occur during dating involve physical skills or activities that are beyond the capacity of people with major disabilities. Both the person with a disability and the non-disabled individual may face intense resentment from others, including parents and friends who regard the relationship as inappropriate or inadvisable. As a result, dating relationships between people with disabilities and non-disabled individuals are subjected to unique and constant stressors (DeLoach, 1994). Those who are able to overcome the effect of adverse attitudes and taboos against physical contact between persons with and without disabilities must still confront the handicaps resulting from ambiguous social values and dating norms. Social customs, perhaps more than physical differences, are powerful deterrents to these relationships (Brodwin et al., 2004).

Many professionals are implicitly or explicitly aware of the social problems restricting the sexuality of people with disabilities; these problems increase vulnerability. Reduced prospects for forming sexual and marital relationships pose a serious threat to the natural instinct of all human beings to find love, acceptable mates, and establish families (Brodwin & Chen, 2000). The traits that limit opportunities for marital and



sexual relationships involve unalterable physical characteristics rather than mental and emotional attributes. They are the product of events or circumstances over which the person has no choice and little control. Hence, many individuals with disabilities experience anger and frustration in their personal lives. These feelings are also the result of widespread reluctance to discuss or examine the social factors that can inhibit the development of sexual relationships between those with disabilities and non-disabled individuals. Although professionals who work with people with disabilities may be cognizant of the reduced probability that their clients will achieve sexual and marital relationships, they often appear reluctant to acknowledge and discuss these subjects (Kroll & Klein, 1995).

## **Self-Esteem and Disability**

One of the most difficult accomplishments in life is being courageous enough to take chances with love, as it requires not only loving oneself but caring for another person. Self-esteem helps people with disabilities maintain a positive attitude. Research indicates that almost every aspect of our lives including personal happiness, success, relationships, achievements, creativity, and sexuality are dependent upon positive self-esteem (Davidson & Moore, 2005; Ivey, Ivey, & Gluckstern, 2006). With positive self-esteem, a person is more effective, productive, and responsive to others in healthy and affirmative ways.

Anxiety due to disability may cause an individual to withdraw and, as a result, lead to depression and loneliness (Trieschmann, 1988; Vash & Crewe, 2004). This loneliness or depression is often a symptom of frustration. If the frustrated individual is unable to socialize, he or she may become anxious and withdrawn. Increased loneliness, anxiety, and depression can make it problematic for people with disabilities to recognize their role in society.

## **Body-Image and Disability**

Body image encompasses perceptions and attitudes about one's own body, especially regarding physical appearance (Cash & Pruzinsky, 2002). The role of body-image and attitudes in human sexual functioning include one's appearance-related thoughts, emotions, and behaviors. Specifically, body dissatisfaction and excessive psychological investment in one's physical appearance may lead to physical self-consciousness and body exposure avoidance during sexual relations, which in turn may impair sexual desire, enjoyment, and performance.

Most body-image research has focused on eating disturbances among women (Cash, 2004). Clearly, body image has implications for other facets of psychosocial functioning in both sexes and can influence one's interest in and experiences during sexual activities. Wiederman (2002) has recently found that body dissatisfaction may inhibit sexual behavior and interfere with the quality of sexual experiences.

Western culture has firm ideas on the appearances of men and women. A person with a disability may feel unattractive, or even 'less worthy' of sexual partnership, because they cannot live up to the idealized image. If the disability was acquired later in life, the person may remember how they used to look and feel unattractive by comparison. Talking with others who have overcome their body image problems may be helpful (Kroll & Klein, 1995).

## **Sexual Abuse**

To express and enjoy one's sexuality, one must be able to grow in a healthy, nurturing, and loving environment. Sexual abuse of individuals with disabilities is a problem of epidemic proportions that is only beginning to attract the attention of researchers, service providers, and funding agencies. For different disability types, varying dynamics of abuse come into play. People with disabilities are more at risk of abuse and neglect than individuals without disabilities (Orange & Brodwin, 2005a). Sexual contact becomes assault when a person is unable to consent to an activity, does not consent, and/or when a service provider engages in sexual contact with a client. Adults with disabilities who have been sexually assaulted may have experienced the sexual assault or abuse as an adult or they may be adult survivors of childhood sexual abuse.

Factors that place these individuals at higher risk include those that place all people at risk of sexual abuse and maltreatment in addition to other risk factors that are more directly related to disabilities. These include: society's negative attitudes about disabilities; people's reactions to, and interactions with, children



with disabilities (including family members and non-family caregivers); factors that relate to the particular disability itself; and program policies and procedures governing the care of individuals with disabilities by others (Orange & Brodwin, 2005a).

## **Spirituality and Disability**

Spirituality is a basic construct that subsumes religious involvement. The highest level of our development is affected by our ability to appreciate the sacred in life and to find a sense of meaning and purpose. Spirituality invites people with disabilities to live fully in the present. Research (Havranek, 2003; Vash & Crewe, 2004) suggested that people, and in particular individuals with disabilities, depend on spirituality and religion as an important, if not primary, method of coping with physical health problems and life stressors. Spirituality has been shown to have a positive impact on enhancing people's health and ability to cope with disability or illness (Keonig, 2002). Ironically, however, spirituality is not addressed in the rehabilitation process because many believe that spirituality is equated with religion.

Although research about spirituality in the context of disability is sparse, many thoughtful writers have considered religion and spirituality to be crucial factors in adjustment to disability (Havranek, 2003; Koenig, 2002; Vash & Crewe, 2004). According to Vash and Crewe, high rates of belief in God indicate that spirituality cannot be ignored as a source of power within the individual. This spirituality can help overcome the adversity of living with a disability or chronic illness.

## **Multicultural Perspectives**

A multicultural perspective is not exclusive to minority groups. Culture is a part of all our lives, though at times it is viewed as something that other people possess. It encompasses the socially transmitted behavior patterns characteristic of a community or population (Davidson & Moore, 2005; Pedersen & Ivey, 1993). People with disabilities have developed a culture as a result of characteristics that are part of the environment in which they live (Orange, 1995). Counselors may attempt to understand these characteristics, which are often viewed as peculiarities, without seeing their own cultural biases. This is true of sexuality and disability. With regard to sexuality, people with disabilities are seen as needing to be corrected to fit into the dominant, non-disabled culture. Professional services always reflect the dominant culture.

To provide appropriate services when counseling ethnic minorities about issues related to sexuality and disability, counselors need to understand both the meaning of disability in their lives and the cultural context within which they live. Ideas about sexuality are part of a larger culturally-based belief system. All cultures have shared ideas of what makes people sexy, appealing, and helps them maintain health through time (Orange & Brodwin, 2005b). These beliefs help people understand their sexuality and make sense of the world around them. All cultures have beliefs about the "appropriate" type of sexuality; cultural and ethnic beliefs describe how people view what is sexually desirable. Defining sexuality from a multicultural perspective is particularly important for people with disabilities. Counselors need the understanding that we are all sexual beings, young or old, married or single, heterosexual or homosexual. As people change, their sexuality changes which remains a lifelong process no matter what the disability, culture, or ethnic background.

Heterosexual relationships are not the sole form of interpersonal attraction. As more individuals feel comfortable acknowledging their sexual orientation, gay and lesbian relationships have gained wider acceptance. Despite the gradual subsidence of homophobia over the last several decades, individuals with disabilities with a same sex gender orientation are intimidated from expressing their true sexuality.

## **Conclusion**

Sexuality is a form of communication, a way of expressing part of one's personality to another. The way people present themselves largely depends on how they see and feel about themselves. Sexuality is a very intimate area of an individual's personal life and is usually shared among a few people who care and respect one another. Relationships are developed between people sharing their experience and insights concerning their sexuality and learning to appreciate each other for who they truly are as unique human beings.

People with disabilities need to learn to express and enjoy their sexuality. Learning to appreciate one's sexuality is a lifelong process that takes courage and understanding, with or without a disability. The most important task faced by individuals with disabilities is to develop a value system independent of the often negative interactions with others in society. Only then can they reach the point where their sexuality is truly appreciated and learn to love others as well as themselves.

"I feel extremely privileged to wake up each morning knowing that I have the ability to love and be loved. My beautiful wife, Marie, and two sons, Leo Jr. age 15, and Brandon age 11, fill each day and moment of my life with joy, happiness, and a passion for living. My professional life is filled with many rewarding challenges as Coordinator of Disabled Student Services at Oxnard College in Oxnard, California. One must never lose sight of the ability and opportunity to live and love" (L. M. Orange).

## Case Study

Larry Young is a 26-year-old male who sustained an incomplete spinal cord injury at the C-5 level three years ago. While on vacation from work with his fiancée, he was injured in a swimming accident. He was flown to a nearby trauma center where he received six weeks of neurosurgical care before being transferred to a rehabilitation center in his home community. During the next two months, he received interdisciplinary rehabilitation and was then transferred to his parents' home where he has lived until this time. There is permanent and significant decreased function in all four extremities, and Larry must use a wheelchair to ambulate. He has about a 50% loss of function in both upper extremities, with a capability for some gross and fine hand dexterity. Since leaving the rehabilitation center, Mr. Young has spent the majority of his time in his room watching television and reading while his mother provides his personal care. His fiancée and friends have been visiting infrequently, and although he used to be socially active and well known in his community, he is now alone much of the time. Recently, Larry has become dissatisfied with his life and is seeking rehabilitation counseling services to help him plan for a possible return to college and a career. Prior to his accident, Larry obtained an Associate of Arts degree in liberal arts with an emphasis in library services. Before the accident, Mr. Young had held various positions in his parents' computer retail store, involving sales, computer repair, stock clerk, and cashier. He had planned to marry his fiancée and take over the family business someday, but he feels that this is no longer possible due to his physical limitations.

## Questions

1. What are some issues that may need to be addressed in regard to Larry's sexuality?
2. How would you try to learn about Mr. Young's psychosocial issues and how he feels about his body, sexual activity, and social interaction? If Larry wants counseling in this area, will you provide it? Discuss.
3. What kind of adaptive equipment and devices would you recommend to increase Mr. Young's independence?
4. Does Larry possess transferable skills? Explain.
5. As his counselor (either at the college or a rehabilitation agency) discuss educational and vocational counseling services to be provided.

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## **About the Author**

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## Chapter 20

# NEUROLOGICAL CONDITIONS

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## Introduction

Neurology is the specialty of medicine that is concerned with the study and treatment of the nervous system and its disorders. The practice of neurology requires specialized knowledge of anatomy and physiology of the nervous system, and familiarity with many diseases encountered in general medicine. Neurologists may partner with neuropsychologists who are expert in evaluating brain-behavior relationships. Through a wide array of assessment techniques, neuropsychologists elucidate the brain's miraculous processes of thought, memory, judgment, and motor function (Lezak, Howieson & Loring, 2004). This chapter focuses primarily on the practice of neurology; some references from neuropsychology are also employed. The complexities of the neurological system challenge the expertise of the most experienced health professionals. While there are many aspects of brain functioning that are not fully understood, considerable advances occurred in recent years, and there is a growing confidence among neuroscientists that “a real understanding is beginning to emerge” (Beaumont, 2008, p. 3).

This chapter familiarizes rehabilitation counselors with basic aspects of neuroanatomy, the neurological examination, and frequently diagnosed neurological illnesses and conditions. It is not intended to be an all-inclusive discussion of neurology. Standard textbooks are referenced for in-depth inquiry (Aminoff, Greenberg, & Simon, 2005; Beaumont, 2008; Brust, 2007; Hauser, Kasper, Braunwald, Fauci, & Longo, 2006; Lezak, Howieson, & Loring, 2004; Pliszka, 2003; Victor & Ropper, 2005).

## Anatomy and Physiology

The nervous system consists of the central nervous system (CNS), peripheral nervous system (PNS), and autonomic nervous system (ANS). The ANS is formed from components of both the CNS and PNS (Beaumont, 2008). We begin with the CNS, which is composed of the brain and spinal cord.

### *Central Nervous System (CNS)*

The brain itself can be divided into four regions: the brain stem (medulla oblongata, pons, and midbrain or mesencephalon), the cerebellum, the interbrain (diencephalon and the limbic system or rhinencephalon), and the cerebral cortex (telencephalon). From the diencephalon upward, the brain is divided into left and right hemispheres, resulting in pairs of structures that typically function contralaterally; the left hemisphere involves the right side of the body, and vice versa. A few structures control the side on which they are located.

The cerebral cortex is the part of the brain most familiar to us because it is most visible in texts and media, as perhaps is the cerebellum, or the cauliflower-shaped region that parallels the brain stem. As we discuss the different functions of the brain, it is important to note that there is not complete agreement regarding the precise purpose of the different regions of the brain. Older paradigms of focal or specific functions of a brain region have given way to integrated or network models of brain function (Beaumont, 2008; Lezak, et al., 2004), including consideration of complex interactions between the brain and its somatic environment, or the body in its context (Damasio, 2006).



**The Brain Stem**

As the spinal cord enters the skull it becomes the brain stem, which controls particular senses, vital life processes, visceral and somatic functions. The brain stem enters the skull first as the medulla oblongata, followed by the pons, and then as the midbrain. The cerebellum is situated parallel to these structures, interconnected by innumerable pathways at the base of the cerebral cortex; it coordinates the timing of muscle activity at a subconscious level. The brainstem structures control functions such as body temperature, blood pressure, respiration, heart beat, and balance. Low level visual and auditory functions are processed in the midbrain. Interconnected throughout the brainstem is the reticular formation, influencing arousal of the brain (e.g., wakefulness and attention) (Beaumont, 2008). Twelve pairs of cranial nerves enter the brain without passing through the spinal cord, and impulses from these nerves influence brain stem functioning. They are part of the peripheral nervous system (discussed below), and their related functions are illustrated in Table 1 (Felton, Perkins, & Lewin, 1966, p. 60).

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**Table 1**  
**The Cranial Nerves and Their Related Function**

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<b>Nerve and Number:</b>	<b>Related Functionally to:</b>
Olfactory	Sense of smell.
Optic	Vision.
Oculomotor	Movement of eye muscles.
Trochlear	Movement of one eye muscle.
Trigeminal	Sensation over skin of face, lining of mouth and nose, tongue, and muscles used in chewing.
Abducens	Movement of one eye muscle.
Facial	Muscles of the face, salivary glands, and sense of taste in anterior part of tongue.
Auditory (with a vestibular branch)	Hearing, sense of balance, and sense of rotation.
Glossopharyngeal	Salivary glands, muscles of the pharynx, swallowing, and sense of taste in pharynx.
Vagus	Heart action, peristalsis of small intestine, speech, swallowing, secretion of gastric glands, and respiratory rhythm of lungs.
Spinal Accessory	Muscles of the shoulder and neck.
Hypoglossal	Musculature of the tongue.

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**The Brain**

The subcortical regions of the brain, including the diencephalon and the limbic system (rhinencephalon), control sensation, movement, states of awareness, and instinctive physical behavior. These regions also influence motivation, emotion, and the ANS. The cerebral cortex (telencephalon) supports high-level brain functions.

At the subcortical level, the diencephalon can be divided into three structures: the thalamus, hypothalamus, and basal ganglia. The thalamus is a primary control center for registering sensation and controlling movement. The hypothalamus has been called “the center of the brain” and influences eating, drinking, sleeping, walking, sexual behavior, response to reward/punishment, and the fight-or-flight response. The limbic system, located on the borders of the diencephalon, directly connects to the cerebral cortex, and has been labeled the visceral brain for its involvement with emotion and behavior. Two

important structures within the limbic system include the amygdala, involved with rage response, and the hippocampus, central to memory functioning.

Structurally, the cerebral cortex is divided into four lobes by fissures or sulci: frontal, temporal (behind the temples), parietal (across the top of the brain), and occipital (back of the head). Research has suggested that the frontal lobes are associated with executive functioning and language. Temporal lobes are associated with hearing and language. Parietal lobes are associated with sensation and perception. The occipital region is associated with vision. Actual brain-behavior relationships are much more complex than this very basic explanation.

Functionally, the cerebral cortex is divided into three zones. The primary cortex initiates voluntary movement and related sensation, located in the occipital, temporal, and parietal regions of the brain. Because primary cortices receive projections directly from sensory receptors located throughout the body (e.g., skin, retina, inner ear) and initiate voluntary movements, they are considered the first stage of processing within the brain. The secondary cortex integrates sensory and motor behavior and related perception, and is located adjacent to the primary cortex. Secondary cortices perform higher level processing on incoming information via direct connections with the primary cortices, and are considered second order processing. The tertiary or association cortex, though not thoroughly understood, appears to make up the “thinking brain,” in that it analyzes information received from multiple primary and secondary cortices. These brain areas are involved in attention, planning, problem solving, decision-making, and complex abstraction (Beaumont, 2008).

There are specialized regions of the cortex that deal with language, located in the left hemisphere and named after the scientists who originally described them. Broca’s (French scientist, Pierre Paul Broca, 1824-1880) area, within the left frontal lobe, involves expressive communication, and is connected by means of nerve pathways to Wernicke’s (German neurologist, Karl Wernicke, 1848-1905) area in the left temporal lobe, which involves receptive communication.

The brain uses about 1/5<sup>th</sup> of the blood pumped from the heart, and must receive a constant supply of oxygen and glucose to meet its incessant metabolic needs (Felton et al., 1966). If because of a vascular or metabolic disorder, this supply is interrupted, the brain will metabolize its own proteins and fats, and rapidly destroy itself. Without oxygen, brain cells die within five to six minutes. Historically, it has been suggested that the CNS is not capable of regenerating itself, but there is an emerging body of evidence to suggest that neurogenesis does occur. Common clinical practice suggests that there is no significant degree of regrowth possible. To some extent, other uninjured areas may assume the functions of an injured area (neuroplasticity) (Beaumont, 2008).

### ***Peripheral Nervous System (PNS)***

The cranial nerves noted previously, 31 pairs of spinal nerves, and nerves of the ANS make up the PNS. Unlike the CNS, when the PNS is damaged, parts of it can grow back, but damage can be permanent. The purpose of the PNS is to bring information from receptors throughout the body into the CNS, and to carry instructions outward to effectors in muscles and glands; it is a link between the spinal cord and the body’s periphery. Some neurological functioning is organized entirely within the spinal cord itself in the form of reflexes that occur without the initiation of the higher CNS; however, the CNS can influence the speed or sensitivity of the reflex (Beaumont, 2008).

The PNS begins in the spinal cord at which point spinal nerves form from anterior (motor function) and posterior (sensory function) roots which are distributed at various levels to all parts of the body. There are 31 pairs of spinal nerves, numbered according to four regions of spinal cord vertebrae: the cervical (C1-C7) region has eight pairs, thoracic (T1-T12) has 12 pairs, lumbar (L1-L5) has five pairs, the sacral (S1-S4) has five pairs, and the coccyx has one pair. Each nerve and nerve root serves a particular cutaneous area of the skin and a specific set of muscles (Drake, Vogl, Mitchell, Tibbitts, & Richardson, 2007).

Neurologists must be well versed with the many patterns of neuropathy (abnormality) associated with each nerve and nerve root. For instance, root involvement at the fifth lumbar level (L5) causes weakness in ability to lift the ankle and toe from the floor. Forearm involvement of the ulnar nerve (C7-T1) produces weakness in the intrinsic muscles of the hand and an inability to move the hand outward (see Spinal Cord Injury chapter for more detailed information).

### ***Autonomic Nervous System (ANS)***

Nerves of the ANS are concerned with activation of neurological systems, responding to emergency situations, and with emotion. The ANS controls vital involuntary or autonomic body functions, such as respiration, heart rate, digestion, defecation, urination, and sweating. The ANS is made up of sympathetic and parasympathetic components; one acts to neutralize or antagonize the other. For example, a sympathetic nerve increases heart rate, while a parasympathetic nerve decreases heart rate (Drake, et al., 2007).

## **Neurological Examination**

Individuals with complaints suggesting disease of a neurological origin (e.g., headaches, motor weakness, pain, sensory disturbance, vertigo, and difficulties in cognition and speech) seek help from their treating physicians. After a preliminary evaluation, the family practitioner may refer the patient to a neurologist for further evaluation. The neurologist attempts to determine the specific anatomical location of the abnormality within the nervous system and assesses possible etiology (cause). Causes or origins of the problem may include trauma, tumor, immunological response, infection, hereditary predisposition, among other diseases and disorders. Based on the evaluation, history of the illness, and special testing, the neurologist establishes a specific diagnosis (DeMyer, 2004; McPhee, Papdakis, & Tierney, 2008).

The neurologist begins the examination by obtaining a neurologically-oriented history. He or she then performs a neurological evaluation, including a systematic examination of the following: mental status, cranial nerves, gait and station, cerebellum, and motor and sensory function (explored below). Based upon these data, the neurologist may order laboratory tests, imaging procedures, or neuropsychological testing. All data are analyzed and synthesized into preliminary diagnoses.

### ***Mental Status***

The mental status examination evaluates the ability to reason, follow commands, speak, and solve problems. It reveals gross abnormalities of mental functioning and difficulties in speech. When dysfunction exists, return to typical functioning is dependent on the cause and may worsen, improve, or remain unchanged over time. During acute, subacute, and post acute recovery, neuropsychological testing is used to monitor treatment progress; once the condition is stable it can be used to estimate the person's remaining functional capacity (Lezak, Howieson, & Loring, 2004).

### ***Cranial Nerves***

Examination of the cranial nerves involves evaluation of the nerves that serve the areas of the face, including sense of smell, vision, eye movement, face movement, and sensation. Of particular importance are visual disturbances and disturbances in speech formation (dysarthria).

A frequently encountered neurological finding from an examination of the eyes is homonymous hemianopia. In this condition, there is blindness of the nasal half of the field of vision of one eye and the temporal half of the other, or right-sided or left-sided blindness of the corresponding sides in both eyes. In either situation, there is blindness in one half of each eye with vision unaffected in the other half of each eye (Brust, 2007). In addition to the cranial nerve as a cause, this condition may also result from damage to the primary occipital cortex.

With abnormalities of the facial nerve, there are disturbances of facial movements that may result in pronunciation difficulties or slurred speech. There is a functional limitation in ability to produce understandable speech (dysarthria).

### ***Gait and Station***

Disorders of gait and station include various disturbances of walking, such as ataxia (gross incoordination of muscle movements), spasticity, and steppage gait (high-stepping gait so that the toe clears the ground). Gait disturbance causes limitations in ability to walk, particularly for long distances. If a significant gait disturbance is present, vocational rehabilitation needs to be directed toward work involving limited ambulation (e.g., sedentary work).

### ***Cerebellar Function***

The cerebellum controls movement and coordination. Damage to the cerebellum may manifest as jerky movements, intention tremors (tremors only during deliberate movement), and loss of balance and proprioception (position sense), especially with eyes closed (Beaumont, 2008).

### ***Motor and Sensory Function***

During the motor and sensory function evaluation, the examiner systematically tests reflexes and muscle tone in each group of muscles. For example, the examiner uses the prick of a pin or a vibrating instrument to examine sensory response. Touch proprioception, or the ability to tell where a finger or toe is in relation to the rest of the body, is also queried. The motor system consists of upper motor neurons (nerve connections from the central nervous system to the spinal cord) and lower motor neurons (nerve connections from the spinal cord to muscle fibers in the periphery). Upper motor neuron dysfunction occurs in illnesses involving the cortex and brain stem. This often produces abnormalities involving at least one limb or one-half of the body. There is a characteristic loss of strength, increase in reflexes (hyperflexia), and spasticity associated with these disorders (Aminoff et al., 2005).

Lower motor neuron diseases involve the nerve roots, peripheral nerves, or both. They cause a smaller area of deficit that affects a specific group of muscles, resulting in abnormal muscle functioning. Characteristically, there is muscle weakness, lack of muscular tone (hypotonicity), and loss of reflexes (areflexia). The sensory system has CNS and PNS components. In PNS disease, a small area of the skin may show decreased sensation corresponding with abnormality of the specific nerve or nerve root that supplies that given skin area (Brust, 2007).

## ***Electrophysiology***

### ***Electroencephalogram (EEG)***

Neurologists and physicians can request an electrophysiological test called the EEG, which generates graphs of electrical activity in the brain that are used to explore suspected pathological processes, particularly seizure disorders. An EEG involves the use of electrodes that provide amplification and summation of regional brain activity measured at the scalp level.

### ***Electromyography (EMG)***

Another electrodiagnostic test is **electromyography**, which involves inserting needles into various muscles to amplify the electrical activity of muscles. Electromyographic examination reveals abnormalities suggestive of muscle or nerve disease. This procedure aids in diagnosis and pinpointing a disturbance of a particular nerve, nerve root (radiculopathy), or muscle (myopathy). A **nerve conduction study** is a test that delivers small electric shocks via electrodes placed on the skin. Electrical stimulation is then followed as it travels along the nerve. Motor and sensory nerve fibers are tested. This test is most useful for identifying mononeuropathy (e.g., carpal tunnel syndrome) and peripheral neuropathy.

## ***Neuroimaging Techniques***

Advances in neuroimaging technology have allowed physicians to construct detailed visualizations of brain structures and vasculature, blood flow and related metabolic activity, and basic neurochemical composition can be detected. Neuropsychological assessment is used in combination with this technology to help identify impairment and residual functioning (Bigler, 2000).

### ***Computerized Tomography (CT)***

Useful images of the brain and spinal cord are created with neuroimaging techniques like CT, or in the past, computerized axial tomography (CAT) scans. CT uses X-rays directed from different angles through the object of interest to the X-ray film on the other side. Many images are combined by computational methods to depict “slices” through the body. Relatively inexpensive and widely available, it has limited ability to image abnormalities, and has lower definition, but tends to be a first-line of inquiry before the more advanced and expensive imaging techniques (Kasper et al., 2005; Drake et al., 2007).

### ***Magnetic Resonance Imaging (MRI)***

An MRI creates a more precise image of the brain by placing the head in a powerful magnetic field where radio waves are used to align the nuclear magnetization of hydrogen atoms. This process allows for greater contrast between different soft tissues of the body than CT. MRI provides a high resolution, three-dimensional image of the brain.

### ***Functional MRI (fMRI)***

The fMRI goes a step beyond showing clear structures of the brain to illustrating brain function. The fMRI detects brain neuron activity through the “haemodynamic response,” which is associated with the concentration of oxygen in the blood in areas where brain neurons are most active. Images taken seconds apart can suggest dynamic brain function in association with different cognitive processes. Brain images taken during various stages of a cognitive process are later compared to suggest areas of the brain associated with specific tasks or functions.

### ***Positron Emission Tomography (PET)***

PET scans can also dynamically image brain function, but at a much slower pace than the fMRI. This technique uses a positron-emitting radionuclide (tracer) to detect regions of greater activity. However, the process is very expensive and takes 30 seconds for a single image, impractical for some areas of inquiry. A related technique that allows the construction of three-dimensional images from two-dimensional images based on gamma ray technology is the single-photon emission computed tomography (SPECT).

### ***Developing Technologies***

Magnetoencephalography (MEG) uses a technique similar to that of an EEG except that it detects magnetic signals associated with brain activity. The MEG is purportedly able to detect areas of brain activity as small as two millimeters, outperforming any existing techniques; however, the equipment is very expensive and not widely available. Fiber tractography (FT), combined with an adaptation of the MRI signal into diffusion-tensor imaging (DTI) creates striking 3-D images of the connections between brain areas (white matter) that promise to shed greater light on the remarkable complexity of the brain (Beaumont, 2008).

## **Abnormalities of the Brain**

Clinical data suggest specific behaviors associated with areas of the brain; our understanding of these relationships is evolving with advances in neuroscience (Beaumont, 2008; Lezak et al., 2004). Historically, there has been an inherent difficulty in building a sound scientific basis in this area of inquiry, as it depended on incidental clinical cases. Lesions could not ethically be introduced into the human brain; researchers had to make the best use of the clinical data available to them in the normal course of healthcare. Recent advances in functional neuroimaging have allowed neuroscientists to test hypotheses in the healthy brain regarding the role of discrete function areas and their associated networks across a number of cognitive domains. Neuroscientists have begun to use a “virtual lesion” technique known as Transcranial Magnetic Stimulation (TMS), whereby an innocuous magnetic field is applied to the scalp temporarily disrupting function of the underlying brain area. Together, clinical cases and data from advanced neuroscientific methods provide converging evidence to elucidate the function of many brain areas (Hallett, 2000).

### ***Subcortical Structures***

Much of what is known about subcortical brain function has been learned from animal research. Clinical studies of humans with damage to these structures is difficult, as it causes major functional impairments of consciousness and basic drives. In fact, damage to the brain stem, mesencephalon, and the diencephalon can be catastrophic and is often fatal. A notable exception is research examining the role of the thalamus and basal ganglia; together with frontal and parietal cortex, these areas form a number of interconnected cortical-subcortical circuits involved in such diverse cognitive operations as response selection, learning, attention, and the execution of routine motor operations (Alexander, DeLong, & Strick, 1986).



## ***Frontal Lobes***

Functioning associated with the frontal lobes includes a broad variety of behaviors. The frontal lobes have been associated with the ability to think abstractly: the ability to form and change mental sets; consider different aspects of a situation, take apart and put together (synthesize) the elements of some object, to think symbolically, and to plan ahead. These functions are essential to effective independent living and work; damage to the frontal lobes can impair these abilities with dramatic consequences. Severe frontal lobe damage can cause dull emotional affect and lack of initiative and spontaneity. The archaic frontal lobotomy procedure was used with people with psychiatric diagnoses who were difficult to manage to bring about these effects and control their behavior. Other frontal lobe-related abnormalities include poor self-monitoring (e.g., inattention to personal hygiene) and socially inappropriate and impulsive behavior. Interventions in this case are very challenging as it is difficult to improve a problem behavior if one has trouble tracking its significance. These symptoms have been associated with a frontal lobe syndrome that is quite debilitating and may require substantial and long-term care (Beaumont, 2008).

Four frontal lobe sections are associated with different functional consequences, beginning from the top of the head and moving down toward the orbits of the eyes. These are the motor and pre-motor cortex, prefrontal cortex, Broca's area, and orbital cortex. Damage to the motor cortex may result in loss of voluntary control over specific parts of the body and possibly fine-motor control (e.g., movements of the hands, fingers, face). Lesions in the pre-motor cortex result in difficulty with coordinating gross body reflexes and movements, verbal and design fluency, and spelling. Lesions in the prefrontal cortex may affect planning and programming of sequences of behavior, problem-solving, perceptual judgment, memory, and attention. The relationship among these regions parallels the primary, secondary, and tertiary cortex relationships described earlier (Beaumont, 2008).

Located in the left frontal lobe, Broca's area, when damaged, can result in impairment of expressive communication (see also receptive speech as it relates to the temporal lobe discussion below). Finally, the orbital cortex region of the frontal lobes relates to aspects of personality and social behavior. Motor vehicle accidents damaging the orbital cortex bilaterally can result in dramatic personality changes, problems with anger management, and inappropriate social behavior.

## ***Parietal Lobes***

Abnormalities of the parietal lobes result in a wide-range of symptoms, from misperception of somatosensory events, loss of or deficits related to spatial functioning, and problems with navigation. Lesions in the parietal lobes can result in contralateral or ipsilateral (same-side) loss or alteration of sensation for parts of the body. The loss or change may be total or it may only involve a single feature of sensation, such as touch, pressure, temperature, or pain. In addition to sensations perceived by the skin, damage to the parietal lobes may also affect ability to perceive information about body position and movement. Parietal lesions may cause apraxia (loss of intentional movement) (Beaumont, 2008).

Diseases of the left parietal lobe produce problems with language functions, including reading (dyslexia) and writing (alexia). It can cause difficulty with recognizing objects both visually and through the sense of touch (visual or tactile agnosia). Disease of the right parietal lobe cause difficulty with mathematical calculations (acalculia) as spatial representation is helpful in solving more complex math problems.

Spatial neglect is a parietal lobe condition associated with a failure to pay attention to a particular area of space, typically the half of space opposite the parietal lesion. Most often persistent symptoms of neglect occur following damage to the right parietal lobe. Patients who have spatial neglect may present with scratching and bruising on the side of the body opposite the lesion due to bumping into things on that side. They may shave only one side of the face or forget to completely dress one side of the body.

## ***Temporal Lobes***

Damage to the temporal lobes impact all aspects of auditory perception; traumatic brain injuries that lead to skull fracture and associated damage in the temporal region lead to hearing loss or deafness. The temporal lobes impact some aspects of visual perception (given its connections with the occipital lobes).

Reception and comprehension of language is associated with Wernike's area, in the left hemisphere, at the junction of the temporal, parietal, and occipital lobes; damage to Wernike's area produce receptive or Wernicke's aphasia, compromising the ability to receive and comprehend written and spoken language. Lesions in the right temporal lobe cause difficulty with musical ability. The temporal lobes also influence affective, emotional, and personal experience, having close physical connections with the limbic system described earlier. Damage to these structures produce personality changes, psychosis, and changes in sexual behavior (Beaumont, 2008).

The temporal lobes, along with some subcortical structures (hippocampus and some other limbic structures) are involved in encoding and retrieving long-term memory. Typically, lesions in the left temporal lobe impact verbal memory, while lesions in the right temporal lobe impact spatial memory. Korsakoff's disease and Wernike-Korsakoff syndrome are bilateral diseases of the temporal lobes (along with some of the subcortical structures) that create difficulty with retaining new information, or anterograde amnesia. Korsakoff's disease is caused by thiamine deficiency often brought about by chronic alcoholism or infection. Wernike-Korsakoff syndrome is caused by a diffuse disease of the brain called Wernike's encephalopathy (Beaumont, 2008).

### ***Occipital Lobes***

Abnormalities of the occipital lobes affect elementary aspects of visual sensation and perception. For example, homonymous hemianopsia, visual distortions, and blind sight result from lesions in either occipital lobe. Disease of both lobes cause cortical blindness where the pupils remain reactive, but the individual cannot see or react to visual stimuli, part of Anton's syndrome, which may also include a lack of awareness of the deficit. Smaller areas of damage cause gaps in vision (scotomas). Sometimes the entire visual field is affected except for the very center or macular region of vision (Beaumont, 2008).

## **Disorders of the Central Nervous System**

### ***Dementias***

Dementias generally speaking are the decline of cognition in adulthood. They can be degenerative or nondegenerative in nature.

#### ***Nondegenerative Dementias***

Nondegenerative dementias have a number of causes, including infection, chronic drug or alcohol abuse, cerebrovascular accidents (CVAs or strokes), and autoimmune disease (such as lupus). Various types of infection affect the CNS. Patterns of infection include a focal inflammation site (cerebritis) or abscess, most frequently bacterial in origin. Bacterial and viral inflammations of the meninges (membranes enveloping the brain and spinal cord) cause meningitis. Meningoencephalitis, usually viral in origin, affects the brain and meninges. Most CNS infections result in residual deficits, such as personality change, sleep disturbance, decreased cognition, and seizures. When complications occur, damage to cranial nerves and brain tissue result in permanent deficits.

Hardening of the arteries (arteriosclerosis) and hypertension (high blood pressure) are disorders which predispose an individual to arterial vessel abnormalities that may result in a rupture causing a CVA. Functional impairment resulting from CVA depends upon the location and size of the area affected. Depending on the type of vascular compromise, strokes can produce either focal or diffuse brain damage. Depending on severity, CVAs result in loss or diminished use of one side of the body due to paralysis (hemiplegia). Speech disturbance often occurs with damage to the dominant hemisphere of the brain (see Aphasias). Even in mild cases, many individuals have loss of stamina and incoordination on the affected side (hemiparesis). Most improvement after stroke occurs over three to six months, but the window of recovery has widened with ongoing research. Once stabilized, individuals with CVA may benefit from rehabilitation evaluation, beginning with an evaluation of functional limitations, both physical and psychological, as well as the person's capabilities.

Finally, a variety of chemicals and drugs affect the CNS. These substances cause a multiplicity of side effects and reactions. Abnormalities include cognitive and behavioral deficits, seizures, and peripheral

neuropathies. Predictable side effects related to drug (both legal and illegal) intoxication and idiosyncratic reactions to medication are two areas related to drug and chemical effects on the nervous system. Predictable side effects are linked to drug dosage. Idiosyncratic reactions are peculiar to an individual and are unpredictable and unrelated to the amount of drug taken. Cocaine, for instance, causes a CNS vasculitis (inflammation of the vessels) in a few individuals that leads to small strokes; this is not a predictable side effect of the drug. These people generally have diffuse brain involvement, and may have decreased cognition and deteriorated intellectual functioning as the principle finding. Chronic alcohol abuse causes several specific neurological problems such as peripheral neuropathy, cerebellar degeneration, and alcohol-related dementia. With abstinence, improved nutrition, and rehabilitation, these are partially reversible (Victor & Ropper, 2005).

### ***Degenerative Dementias***

Degenerative dementias are caused by the death of neurons intrinsic to the CNS. Alzheimer's disease (AD) is a disease that is classified within this category. The hippocampus, a structure critical to memory, is often the first impacted by AD, which undergo marked atrophy. Upon autopsy, neurofibrillary tangles are the diagnostic feature of AD. Found first in the hippocampus, they spread throughout the brain as the disease progresses. Senile plaques are also associated with AD, but their correlation with cognitive impairment is less strong than with the neurofibrillary tangles.

There is an early onset form of AD (5-10% of cases) affecting people ages 28 to 60 years. People with Down Syndrome are more susceptible to AD due to their common relationship with chromosome 21; AD has been linked to this specific chromosome. People with Down Syndrome have an extra chromosome 21. The dementias are not specifically treatable but medication may slow progression.

The presence of a brain tumor often denotes a poor prognosis with probable progressive deterioration. Brain tumors include primary tumors (arising directly from brain cells) and metastatic tumors (spreading from a tumor elsewhere in the body). The brain is affected in approximately 20% of individuals with cancer. In most cases, once the brain is affected, prognosis is extremely poor and death follows rapidly, often within several months of diagnosis. Metastatic disease comprises the largest percentage of brain tumors; primary brain tumors are less common. Generally, brain tumors grow and cause local destruction within the brain. The rate of growth is variable, at times very rapidly with death ensuing after several months, or sometimes more slowly with the person living several years. For individuals with brain tumors, attempts at continuing current employment may be appropriate, depending on the symptoms, deficits, and rate of tumor growth. Retraining is probably inappropriate. Occasionally, very slow growing tumors are found in younger people. In these cases, the counselor should obtain information from the treating physician regarding prognosis (Drake et al., 2007; Drake, Vogl, & Mitchell, 2004; Kasper et al., 2005).

### ***Aphasias***

Language function can be associated with focal areas of the cortex, and while there continues to be much debate, advances in neuroscience suggest that language functioning spreads across a large area of the cortex (Beaumont, 2008). For right-handed people, a significant portion of language functioning is in the left hemisphere. In the case of left-handed people, very few actually have right (contralateral) hemisphere dominance in language; most have the same brain functioning as right-handed people, and a smaller percentage have bilateral representation of language functioning (Carter, Hohenegger, & Satz, 1980). In either case, disorders of the language system are called aphasias. Classification of aphasias is "one of the most hotly contested issues in the history of neuropsychology, and there is yet no firm agreement" (Beaumont, 2008, p. 136). The most widely accepted paradigm divides aphasias into several categories. *Global aphasia* involves massive and severe disturbance in language functioning across a number of the six categories. Some of these also involve *alexia* (a specific disorder of reading) and *agraphia* (a specific disorder of writing). In almost all aphasias, singing, producing simple automatic phrases, and swearing are often preserved.

**Broca's aphasia** manifests as impaired ability to express oneself verbally, and is therefore also called expressive aphasia. Speech may omit articles, adverbs, and adjectives, or in more extreme cases muteness occurs. Prompting with sounds or contexts assist people with less severe forms of the disorder; some do not realize their problem. In any case, difficulties occur while language comprehension is

unimpaired. People understand what is being said and read as well as ever. Broca's area was described above as one of the language centers of the brain, located typically in the left rear frontal lobe.

**Wernicke's aphasia** is characterized by a severe deficit in auditory comprehension and is, therefore, also referred to as receptive aphasia. Impaired reading and writing often accompany it. While most individuals are capable of expressive and often voluminous speech, their speech may contain substituted but related words or sounds, resulting in a "word salad" that is not always intelligible. Wernicke's area was described earlier as located in the left temporal lobe.

With **conduction aphasia**, people comprehend both speech and writing and produce nearly-normal speech, but at times substitute one sound for another (*phonemic paraphasia*). For example, one might substitute "fable" for "table." Severe impairment may be noted when the person is asked to repeat a phrase that is spoken or while reading aloud. It has been hypothesized that this type of aphasia results from problems with communication between Broca's and Wernicke's areas.

The most common form of aphasia is **anomic aphasia**, which may exist in residual form while one recovers from other types of aphasia. The key element is difficulty with name finding, especially for nouns. Those recovering from it may substitute related words as a means of coping with the difficulty, so it may go undetected in spontaneous speech.

The last two types of aphasia reviewed here are **transcortical motor** and **sensory aphasias**. In the worse case of combined motor and sensory aphasia (which is very rare), the person cannot understand speech or read, is totally nonfluent in speech, speaks only when spoken to, and usually cannot write. Strangely enough, the person can repeat what is heard, also referred to as *echolalic aphasia* (the person echoes back whatever is said). In *transcortical motor aphasia*, most evident is the impairment of speech output. In *transcortical sensory aphasia*, language reception is usually severely affected, while the ability to repeat is preserved, even though little is understood of what is repeated or read. These aphasias are referred to as transcortical, because it is assumed that intact cortical language functions are not connecting with the underlying supporting structures (Beaumont, 2008).

### ***Traumatic Brain Injury (TBI)***

In younger individuals, TBI is the most common disorder of the nervous system. Brain trauma includes closed and open head trauma. Closed head trauma implies that the skull remains intact; open head trauma involves a breach of the integrity of the scalp and skull. Closed head trauma may involve contusion, intracerebral hemorrhage, and edema (swelling). Both closed and open head trauma can be severe and result in significant deficits and possibly death. Speech deficit, difficulty with spatial relations, incoordination, personality change, sleep disturbance, and irritability are common residual symptoms from this type of injury. Neuropsychological evaluation is helpful in determining rehabilitation capabilities and potentials.

Persons with head trauma have a constellation of symptoms called post concussion syndrome. After even minor head trauma (trauma with no apparent neurological deficit at the onset), the patient may develop headaches, nausea, vomiting, dizziness, sleep disturbance, and irritability. There can be forgetfulness and decreased eye-hand-foot coordination. In about 5% of individuals, post concussion syndrome symptomatology is chronic and persists beyond six months. These individuals may continue to have headaches and related behavioral problems that negatively affect personality. They experience forgetfulness, irritability, and decreased stamina; at times, these individuals require medication to maintain daily activities. A major factor in post concussion syndrome is loss of stamina. These deficits have significant educational and vocational implications.

### ***Spinal Cord Disorders***

In younger individuals, especially males, spinal cord injury is frequently the result of acute trauma. Residual muscular and sensory defects are dependent on the level of injury in the spinal cord and on the number of nerve fibers injured within the cord. Trauma to the spinal cord may result in paralysis of the lower extremities (paraplegia) or both the upper and lower extremities (quadriplegia, also known as tetraplegia), depending on location of injury. Damage to the thoracic spine may produce abnormalities involving the legs and cause incontinence (see Spinal Cord Injury chapter for more information). Trauma to the cervical spine



typically produces abnormalities involving at least some of the function of the arms and hands, as well as incontinence. Individuals who have spinal cord disorders require light or sedentary types of work and may have fatigue, loss of stamina, and incoordination. All these are factors the rehabilitation counselor evaluates on an individual basis.

### ***Other Degenerative Diseases of the Central Nervous System***

#### ***Multiple Sclerosis***

Multiple sclerosis (MS) is an inflammatory disease of the CNS that affects the myelin sheath (the lining covering the axons), causing diffuse lesions almost anywhere in the nervous system. Some lesion patterns affect the spinal cord and related motor functioning; others affect the brain resulting in cognitive impairment. MS is currently diagnosed by these typical patterns of lesions detected in an MRI scan. Only 10% affected will need wheelchair support. There are two typical presentations of MS. Relapse-remitting type is characterized by remissions and exacerbations (temporary improvement and worsening). After each exacerbation, new neurological deficits occur which remain stable until the next exacerbation; the frequency of exacerbation is unpredictable. Chronic-progressive type of MS presents a steady decline in function; some begin demonstrating the decline in the early stages of MS.

The cause of MS is thought to be an auto-immune dysfunction; there is currently no cure. Symptoms include spasticity, weakness, incoordination, incontinence, speech problems, and visual disturbances. Psychological impairment accompanies this disease, as well as reduced speed of mental information processing and impaired attention span. Forty to 60% have disturbances in mood; suicide risk may be as high as 15%. Early stages of MS can be incorrectly attributed to psychiatric problems. Beta interferon treatment may slow the progression of MS in the early stages. Cognitive rehabilitation and supportive counseling are very helpful as well (Beaumont, 2008; Freedman, 2005; Goldman & Ausiello, 2004).

#### ***Parkinson's Disease***

Parkinson's disease (PD) is a degenerative disorder that results in the loss of neurons in subcortical structures that serve involuntary movement (e.g., basal ganglia, brain stem). Its major effects include loss of motor control and problems with initiation of motor operations (freezing). Medication and environmental toxins can produce Parkinsonism with symptoms similar to PD, but due to other factors. Symptoms of both involve tremor, slowness (bradykinesia) or rigidity in movement, a shuffling gait, loss of associated movements (e.g., arms moving as you walk), and loss of emotional expression. With PD, some have specific or general psychological impairment, including difficulties with executive functioning, memory, visual spatial skills, while others may have no such impairment. While the cause is not certain, treatment with dopaminergic drug therapy has been helpful, suggesting a lack of dopamine as a culprit (Beaumont, 2008; Brust, 2007; Pfeiffer & Bodis-Wollner, 2004).

#### ***Huntington's Disease***

Sometimes referred to as a subcortical dementia, Huntington's disease (HD) affects the integrity of the basal ganglia. Persons having either parent with HD have a 50% chance of inheriting it. HD is a neurodegenerative disorder that begins midlife with the onset of involuntary body movements. Dysarthria, difficulty with upper limb movements, abnormal gait, and progressive loss of mental functioning occurs, including disinhibition, depression, personality change, and even schizophrenia-like symptoms. HD is a particularly difficult disease in that most people who have it also had personal experience with observing the certain decline ahead. Ultimately, problems with physical functioning lead to death within 15 to 20 years. There is no cure for HD; medications are used to treat symptoms as they arise (Bates, Harper, & Jones, 2002; Beaumont, 2008).

#### ***Motor Neuron Disease***

Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig's disease (a famous New York Yankees' baseball player) occurs on one side of the body and involves one arm and leg. It is a disease of the alpha motor neurons involving specific cells in the brain stem and spinal cord. This disease is degenerative, of unknown etiology, and rapidly progressive. There is loss of strength with hyperflexia and fasciculation (involuntary small movement of muscles). ALS is progressive and individuals eventually require



wheelchairs for mobility. At times, the downhill progression is such that the person cannot eat or swallow within a year of initial onset (McPhee et al., 2008).

Two other forms of motor neuron disease include the bulbar form, where abnormalities begin in the brain stem and affect the face, tongue, and ability to swallow. Progressive muscular atrophy begins in both hands and progresses. Medications may decrease the rate of progression of motor neuron diseases.

### ***Developmental Diseases***

These diseases are genetic in origin or occur as a result of maternal or fetal events prenatally, perinatally, or immediately subsequent to birth. Genetic diseases are typically progressive over time. If the causative factor is a specific event, such as intrauterine hypoxia or viral infection, the disease results in a specific and permanent deficit.

#### ***Cerebral Palsy***

Cerebral palsy results from injury to the brain occurring during development. The usual cause is hypoxia (a deficiency of oxygen). Typically, weakness and spasticity are present in the arms and legs. Spastic diplegia is a congenital spastic stiffness of the limbs.

#### ***Mental Retardation***

Mental retardation (MR) implies markedly below average intellectual ability. It may or may not be detected in infancy, and is typically identified as a child enters the education system. MR affects all aspects of cognitive functioning, with related problems in adaptive functioning.

#### ***Epilepsy***

Epilepsy is caused by congenital abnormalities and often results in damage to the brain over time if uncontrolled. This condition involves transient electrical disturbances in the brain resulting in various types of seizures. Seizures range from mild to severe, occur rarely or often, and are partial (in a specific area of the brain) or generalized (affecting both hemispheres of the brain). Trauma, tumors, high fevers, and certain drugs cause them. Counselors need an awareness of the sedative effects of medications that are prescribed to control seizures (eg., Dilantin, Tegretol). Medication controls most seizures, although some individuals have seizures that are resistant to medication, and surgical intervention becomes the only alternative. Persons with seizures should avoid situations where they would injure themselves or others if a seizure occurs (Brust, 2007).

## **Diseases of the Peripheral Nervous System**

Peripheral nervous system disease involves the nerves, nerve roots, and associated muscles. These diseases may be progressive or nonprogressive.

### ***Diseases of the Muscle***

#### ***Muscular Dystrophies***

Muscular dystrophies are inherited or genetically determined illnesses involving a progressive weakening of muscles. Particular types are described based on the muscle groups involved; etiology is unknown. These diseases usually begin in childhood or in the teen years and are slowly progressive. Individuals with muscular dystrophy usually require sedentary work, either because of current limitations or future anticipated impairments. Because muscular dystrophy progresses, short-term goals need to be established; periodic reevaluation and modification of these goals is necessary. The more sedentary the work activity, the longer the individual will be physically able to maintain a particular employment situation. Prescribed exercise is recommended as prolonged inactivity leads to worsening of the disease state. Maintaining activities and possible physical therapy during non-work hours is beneficial for most persons.

#### ***Primary Myopathies***

These are a rare group of diseases in which there are histological abnormalities involving muscle fiber or muscle cell mitochondria. They produce mild diffuse weakness and a decrease in stamina. The abnormalities are slowly progressive, although life expectancy is not significantly affected. No specific

treatment is available. Individuals with primary myopathies do best in light and sedentary types of employment.

### ***Cranial Neuropathies***

The 12 cranial nerves manifest their own specific symptomatology when involved in a disease process (see Table 1). Abnormalities involving eye and face movements present significant barriers to employment. Cranial nerves three, four, and six control eye movements. Abnormalities of these cranial nerves cause variations of eye movement, including diplopia (double vision). Often, the physician prescribes an eye patch for this condition, leaving the person monocular as long as the patch is worn. Seventh nerve (the facial nerve) abnormalities cause difficulty moving the facial muscles and closing the eyes. Individuals with this problem need protective glasses and must avoid environments where particles of dust can irritate the eyes. Additionally, they may have some degree of dysarthria (difficult and defective speech). Eighth nerve or auditory nerve abnormalities are associated with hearing deficits and vertigo.

### ***Radiculopathy***

Radiculopathy refers to an abnormality of a nerve root that produces pain in the affected region of the spine. This is accompanied by limited motion of the spine, muscle spasm, and nerve root pain radiating down an extremity. Sensory and motor losses occur in the affected area. There are various causes for radiculopathy, including degenerative changes in the spine with the formation of bony osteophytes (outgrowths). A ruptured or bulging disc is also a frequent cause. Pain generally is experienced below the knee in lumbar radiculopathy.

Individuals with radiculopathies may require surgery to help alleviate symptoms, depending on pathology and severity of the condition. Work limitations for individuals with radiculopathy frequently involve limitation to light or sedentary work. Since many sedentary jobs involve extended sitting, workers may need accommodation allowing them to alternate positioning between sitting and standing. Frequency of change in position is dependent on an individual's tolerance for discomfort. Generally, it is beneficial to avoid either prolonged sitting or standing.

Back and neck pain are major employment problems in most industrialized countries. Workers frequently have chronic back and neck pain complaints without objective findings of disc disease or radiculopathy. Often, there is an initial injury, possibly a strain, with ongoing complaints of severe pain. For rehabilitation purposes, one must distinguish between actual physical limitations and pain complaints. A thorough medication history should be obtained with particular attention to addicting or sedating drugs (Brust, 2007).

### ***Peripheral Neuropathy***

Peripheral neuropathy is a disease that can affect any of the nerves within the body. Frequently, peripheral neuropathy affects lower extremity nerves more than the nerves of the upper extremities. Weakness, sensory loss, and incoordination will be noted. There are multiple causes of peripheral neuropathy. Approximately one-third is due to systemic disease, such as diabetes. Another third of the cases are due to autoimmune inflammation. The final third have multiple causes, both inherited and degenerative.

Despite medical treatment, some mild residual symptoms may persist, with symptoms such as mild incoordination, weakness, diminished stamina, and decreased sensation in the extremities. These individuals require lighter types of employment that do not involve frequent use of fine coordination and manual dexterity. At the time of referral for rehabilitation, the level of dysfunction should be stable and not anticipated to change, thereby permitting the counselor to provide vocational rehabilitation services based on current limitations (Aminoff et al., 2005).

### ***Mononeuropathy***

Mononeuropathy involves injury or disease of a single nerve. This usually results from compression of a nerve in a specific area of the body (segmental mononeuropathy). The most frequently seen mononeuropathies involve the median nerve of the wrist (carpal tunnel syndrome), ulnar nerve at the elbow (cubital tunnel syndrome), and peroneal nerve at the head of the fibula.

**Carpal tunnel syndrome.** This condition involves compression of the median nerve in the carpal tunnel of the wrist. It causes numbness and pain in the thumb, palm, and second through fourth fingers. The person may experience radiating pain and pain during the night. Repetitive wrist motion, sustained grip and pinch activities, continuous wrist angulation, poor posturing, repetitive vibration, and physical injury cause carpal tunnel syndrome. There is weakness in the affected hand and a loss of grip strength. Individuals with carpal tunnel syndrome need to avoid frequent pronation, supination, and extension of the wrist. Repetitive activities, such as using a screwdriver for long periods of time, operating a typewriter or computer terminal, need to be modified or avoided.

**Ulnar neuropathy.** Individuals with ulnar neuropathy have decreased strength in the muscles of the hand. There is weakness of grasp and frequently a decreased coordination of the hand. Sensory disturbance occurs in the fourth and fifth fingers. Usually, the nerve is injured at the elbow level; the individual should avoid activities where the elbow rests on a firm surface or in which minor trauma to the elbow occurs repetitively.

**Peroneal neuropathy.** In the lower extremity, peroneal neuropathy causes a foot drop. Thus, the person is not able to lift the foot against gravity and walks with a peculiar gait termed a steppage gait. Often a brace is used to maintain a 90° angle of the foot with the ankle. Individuals with peroneal neuropathies have difficulty walking and climbing stairs; they require sedentary work. Sensory disturbance, if seen, is usually minimal.

Rehabilitation potential for persons with mononeuropathies is generally good. Functional disability is limited to one specific area of the body. These conditions are often stable and improve with treatment; they typically do not worsen over time. Yet, temporary exacerbation frequently is experienced. Rehabilitation counselors working with people who have these conditions need to consider job modification with the current employer, whenever possible (Victor & Ropper, 2005).

## Rehabilitation Case Planning

Knowledge of the history, onset, and course of the disease process is significant in rehabilitation. To maintain employment or return a person to work, the neurological disorder must be stabilized or slowed in its progression. If a patient has had an acute episode that has run its course and reached maximum improvement, the counselor can formulate a rehabilitation plan based on the knowledge that the deficit will not deteriorate further. When an individual is under optimal medical control, but continues to have debilitating neurological symptoms, frequency and intensity of these events help determine the feasibility of providing vocational rehabilitation services (Aminoff, et al., 2005; Goldman & Ausiello, 2004).

## Case Study

Ms. Nancy Smith is 48-years of age, African-American, and married. The Smith's have three children, two living at home and dependent on their parents' support, and one married and living in the same community. Mr. Ralph Smith works full-time as a computerized numerical control (CNC) machinist. After graduating from high school, Ms. Smith attended a community college and received an Associate of Science degree in nursing. Ms. Smith has been a grocery checker for the past 12 years. The *Dictionary of Occupational Titles* (U.S. Department of Labor, 1991) classified the work as GROCERY CHECKER (retail trade), *D.O.T.* # 211.462-014. It is work involving lifting and carrying a maximum of 20 pounds with repetitive lifting between 5 and 10 pounds. The *D.O.T.* provides the following job description:

211.462-014 CASHIER CHECKER (retail trade) Operates cash register to itemize and total customers' purchases in groceries, department stores, and other retail stores. Reviews price sheets to note price changes and sale items. Records prices and departments, subtotals taxable items, and totals purchases on cash register. Collects cash, checks, and charge payments from customers and makes change for cash transactions. Stocks shelves and marks prices on items. Counts money in cash drawer at beginning and end of work shift. May record daily transaction amounts from cash register to balance cash drawer. May weigh items, bag merchandise, issue trading stamps, and redeem food stamps and promotional coupons. May cash checks. May use electronic scanner to record price. May be

designated according to items checked as Grocery Checker (retail trade) (U. S. Department of Labor, 1991, p. 183).

Before this job, Ms. Smith worked for three years as a nursing assistant in a convalescent home, and for four years as a pharmacy technician in a retail drugstore. Her first experience in employment was as a house cleaner, which Nancy did for two years. The work of a nursing assistant involves lifting of 25 pounds repetitively and up to 50 pounds occasionally. A pharmacy technician lifts up to 15 pounds and stands or walks one half of the work shift. Ms. Smith's job as a house cleaner required lifting and carrying up to 40 pounds on an occasional basis with repetitive lifting and carrying of 20 pounds or less.

For the past year, Ms. Smith has had complaints of pain and numbness in her left nondominant hand, primarily the second through fourth fingers. She has night pain and pain radiating into the left shoulder. There is hand weakness and a slight loss of grip strength. Dr. Elizabeth Kim stated that Ms. Smith has a mononeuropathy involving compression of the median nerve in the wrist. Recently, Nancy has complained of low back pain, and feels she is unable to lift and carry more than 25 pounds without experiencing mild pain.

Currently, Ms. Smith receives physical therapy three times a week at 5:00 p.m. on Monday, Wednesday, and Friday. Work hours for a grocery checker on her shift are 8:00 a.m. to 5:00 p.m. Nancy leaves work early at 4:30 p.m. to attend physical therapy. Her employer has concerns about her missing 1½ hours of work each week.

The physical therapist suggested to Ms. Smith that she see a rehabilitation counselor for advice. She has followed through with this suggestion. You have been assigned this case.

### Questions

1. Assign Ms. Nancy Smith a vocational profile, including age category, educational level, and work history (skill and exertional levels).
2. What medical conditions do Nancy's symptoms indicate she may have?
3. Describe Ms. Smith's functional limitations in regard to her job.
4. Should she attempt to continue working as a grocery checker for her current employer? Explain.
5. If you recommend she continue her employment, what advice would you give Nancy and the employer regarding reasonable accommodation?
6. Describe the occupationally significant characteristics (worker traits) and possible transferable skills of Ms. Smith.
7. Suggest rehabilitation possibilities, including use of transferable skills from previous work and possible training programs if Nancy does not continue working for this employer. Remember, this client has a college degree.

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## Chapter 21

# SPINAL CORD INJURY

*Nancy M. Crewe, Ph.D.*

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## Introduction

Compared with many other disabling conditions, spinal cord injury (SCI) affects a relatively small number of people; each year about 11,000 people are added to the current United States total of approximately 230,000 people with this condition. Yet it commands intense interest among rehabilitation counselors and other health care professionals because within a moment in time, SCI causes profound changes in virtually all physical systems and functional abilities. Further, the majority of people who sustain new injuries are young adults with a lifetime of experiences ahead of them. Over half these individuals are in the 16-30 year age group; the average age at injury is 31.5 years (National Spinal Cord Injury Statistical Center, 2005). They require multidisciplinary medical and rehabilitation services to rebuild their lives and contemplate the opportunity for an independent and productive future. This chapter provides essential information on physiological, functional, and psychosocial characteristics pertinent to people with SCI along with important factors contributing to their rehabilitation needs and potentials.

The public views the inability to walk as the primary consequence of SCI. Although this ability is undeniably important, SCI also affects such areas as arm and hand strength and dexterity, bowel and bladder control, sexual function, temperature regulation, susceptibility to infections, and even the ability to breathe. To understand these consequences, it is necessary to become familiar with the anatomy and physiology of the central nervous system.

## Anatomy and Physiology

### *Spinal Cord Structure*

Voluntary motion takes place when nerve impulses travel from the brain down the spinal cord and out to the body through peripheral nerves. Sensory stimuli are carried from the peripheral nerves through the spinal cord to the brain. The full circuit enables tactile perception and coordinated movements. As a result, damage to the spinal cord can result in both loss of voluntary movement (paralysis) and loss of sensation.

The spinal cord extends from the brain stem to a point in the lower back called the conus medularis. Beyond that point, nerve fibers known as the cauda equina (horse's tail) fan out. The spinal cord is encased in a protective canal that is formed by the spinal vertebrae. At each vertebral junction, a pair of spinal nerves exit from the spinal cord and innervate specific muscles, and sensory nerve filaments enter the spinal cord. The vertebrae and nerves are classified into several sections beginning at the neck with seven cervical vertebrae, seven pairs of nerve roots that exit above each of those vertebrae, and an eighth pair that exits below the seventh vertebra. Below the cervical vertebrae are 12 thoracic vertebrae and 12 pairs of spinal nerves. In the lower back there are five lumbar vertebrae and nerve roots and five fused sacral vertebrae with five nerve roots. The lowest part of the spinal column is a single bone called the coccyx. The vertebrae and nerves are numbered from the top with a letter that corresponds to the spinal section. For example, the first vertebra below the skull is C-1, whereas T-1 is the first vertebra in the thoracic section.

Nerves for the voluntary motor system originate in the motor cortex of the brain and extend down through the basal ganglia to the brain stem. Here they cross over to the opposite side and continue to descend in the spinal cord until they synapse at the point where they are about to exit from the spinal cord. The nerves

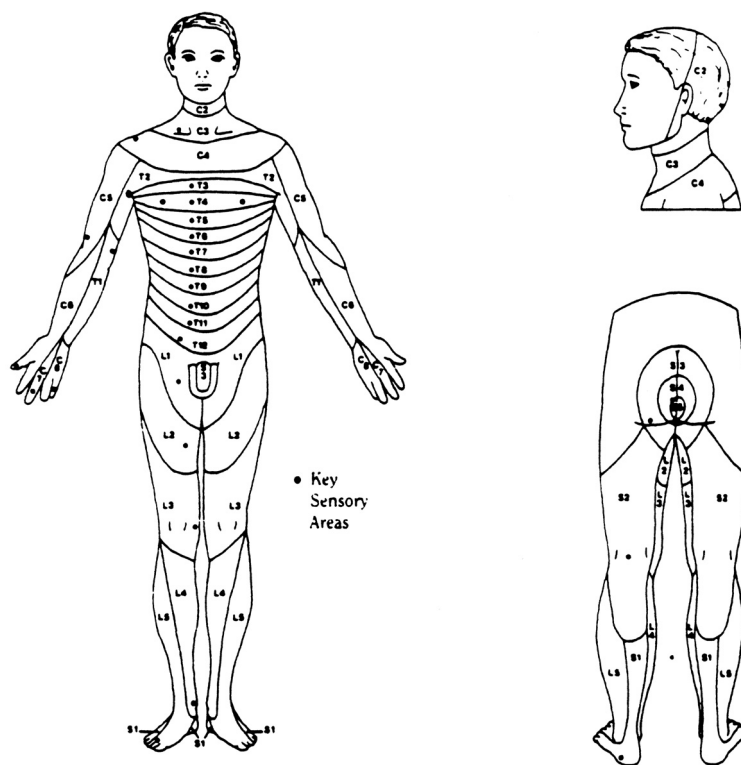
## Spinal Cord Injury

that originate in the motor cortex of the brain's left hemisphere cross over to innervate the right side of the body, and those from the right hemisphere cross over to the left side. These are known as "upper motor neurons." Beyond the synapse, the "lower motor neuron" exits the cord and extends to its particular muscle destination. Any point in the body, then, is connected to the controlling center in the brain by only two neurons (This does not hold true for sensory neurons).

When the spinal cord is damaged, communication is disrupted between the brain and parts of the body that are innervated at or below the lesion. The lesion may be complete (no nerve fibers are functioning below the level of injury) or incomplete (one or more nerve fibers is secure). The cord need not be completely severed to result in a complete injury; the nerve cells may be destroyed as a result of pressure, bruising, or loss of blood supply, and if they die they do not have the ability to regenerate. The amount of functional loss depends upon the level of injury (the higher the damage occurs, the more of the body that is affected) and on the neurological completeness of the injury. Individuals with neurologically complete injuries have more severe and more predictable patterns of functional impairment.

The level of SCI can be determined in two ways. The first method specifies the level of bony damage as indicated by x-rays. The more useful approach, however, indicates the level of neurological damage, measured by careful testing of an individual's ability to perceive pinprick. Skin surface has been mapped into segments called dermatomes (see Figure 1); each dermatome is known to be innervated by sensory nerves at a particular spinal level. Testing the skin, therefore, can reveal the level at which the spinal cord has been damaged.

**Figure 1**  
**Dermatome Patterns of Spinal Cord Injury**



This information was obtained from *Publication Standards to Neurologic Classification of Spinal Cord Patients*, American Spinal Injury Association, pp. 6-7. Reprinted with permission.

## ***Spinal Cord Function***

Individuals who sustain damage at the cervical level will have impaired function in both their upper and lower extremities, a condition known as tetraplegia (or quadriplegia, a term that is now going out of favor). Slightly more than half of injuries result in tetraplegia (National Spinal Cord Injury Statistical Center, 2005). Those who are injured at or below the thoracic level will have paraplegia, with function maintained in their upper extremities but some degree of impairment in the trunk and lower extremities.

The American Spinal Injury Association (ASIA) developed a system for describing the severity of injury that is widely used in the medical community using letters that pertain to the extent of injury (usually A through D) (Young, 2006). ASIA A injuries are complete, with no motor or sensory function preserved below the neurological level of injury, including the sacral segments S4-S5. ASIA B injuries are incomplete, with sensory but no motor function preserved below the neurological level of injury. ASIA C and D classifications refer to incomplete injuries with increasing degrees of motor function preserved below the neurological level of injury.

Certain incomplete spinal cord injuries produce unusual patterns of deficits, depending upon which tracts within the cord are affected. If the damage occurs within the central part of the cervical cord, leaving the outer ring of fibers intact, the individual will have greater weakness in the upper limbs than in the lower limbs, and sacral sensation may be spared. Brown-Sequard syndrome is a lesion that affects only one side of the cord. This causes paralysis on the same side of the body as the lesion, and loss of pain and temperature sensation on the opposite side of the body.

## **Acute Medical and Rehabilitation Care**

Nearly half of spinal cord injuries are the result of motor vehicle crashes; the other major causes include falls, violence, and sports accidents (National Spinal Cord Injury Statistical Center, 2005). Recent reports suggest that the number of new injuries due to violence peaked at 24.8% during the 1990s and has declined to some degree since then. Acts of violence remain the primary cause of SCI among some minority populations. The number stemming from motor vehicle crashes has diminished over a longer period, probably as a result of air bags and other improvements in auto safety.

Emergency evacuation procedures executed at the scene of an injury have improved over the years, with careful stabilization of the neck and spine at the injury scene, the availability of emergency transport, and an increasing tendency to utilize specialized trauma hospitals rather than small local facilities for acute neurosurgical care. Initial damage to the spinal cord aside from the trauma caused by the injury, is due to bleeding, swelling, and oxygen deprivation. The damage is quickly compounded by the body's release of free radicals and other toxic substances, which leads to inflammation and scarring. Several neuroprotective agents, most frequently methylprednisolone, are often administered soon after injury in an attempt to disrupt this cascade of events and prevent further cell death. Research has shown mixed effects of these medications; other drugs are being researched (Klebine, 2005).

During the acute period of hospitalization, physicians may determine that the spinal column is unstable and further neurological damage could ensue. In this case, surgery may be recommended to fuse the spine at the point of injury or otherwise stabilize it with rods or other surgical hardware. The individual may be fitted with a halo or body cast to enable him or her to maintain immobility of the fracture site without being confined to bed for excessive periods of time.

Along with the SCI, a person may have an array of other complications including fractures, internal injuries, and brain injuries, all of which require treatment. When the need for acute medical services has passed, the individual is usually transferred to a rehabilitation unit for multidisciplinary services to help build strength, redevelop skills in activities of daily living, identify and obtain adaptive equipment, and prepare the individual and the family for return to home and community. An SCI rehabilitation team typically includes one or more physicians, nurses, physical therapists, psychologists, occupational therapists, rehabilitation counselors, and social workers. A network of comprehensive Model SCI Centers has been established across the country, providing excellent medical services as well as research on medical and psychosocial aspects of SCI. Follow-along services are highly important, and linking medical

rehabilitation with community-based independent living programs is essential to achieving optimal outcomes (Tate, Maynard, & Forchheimer, 1993).

Changes in the health care system have greatly reduced the length of time needed to complete rehabilitation. Three decades ago, it was not unusual for a person with a new SCI to be in a hospital and rehabilitation center for six months or more before returning home. The average combined length of stay in 1974 was 140 days, while in 2003 it had dropped to 64 days (National Spinal Cord Injury Statistical Center, 2005).

**Table 1**  
**Muscles Supplied and Functions Served by Spinal Nerve Motor Roots**

<b>Root Segment</b>	<b>Representative Muscles</b>	<b>Functions Served</b>
<b><i>Cervical</i></b>		
C1 & C2	High neck muscles	Aid in head control
C3 & C4	Diaphragm	Inspiration (breathing in)
C5 & C6	Deltoid Biceps	Shoulder flexion, abduction (arms forward, out to side) Elbow flexion (elbow bent)
C6 & C7	Extensor Carpi Radialis Pronator Teres	Wrist dorsiflexion (back of hand up) Wrist pronation (palm down)
C7 & C8	Triceps Extensor Digitorum Communis	Elbow extension (elbow straight) Finger extension ("knuckles" straight)
C8 & T1	Flexor Digitorum Superficialis Opponens Pollicis Interossei (intrinsic)	Finger flexion (fist clenched) Thumb opposition (thumb brought to little finger) Spreading and closing the fingers
<b><i>Thoracic</i></b>		
T2 – T6	Intercostals	Forced inspiration (breathing in) Expiration (breathing out, coughing)
T6 – T12	Intercostals – Abdominals	Forced inspiration (breathing in); Aid in expiration (coughing); Aid in trunk flexion (sitting up)
<b><i>Lumbar</i></b>		
L1, L2, L3	Iliopsoas Adductors	Hip flexion (thigh to chest); Hip adduction (thigh to midline, legs together)
L3 & L4	Quadriceps	Knee extension (knee straight)
L4, L5, S1	Gluteus Medius Tibialis Anterior	Hip abduction (thigh out to side, legs apart) Foot dorsiflexion (foot up, walk on heels)
L5, S1, S2	Gluteus Maximus Gastrocnemius	Hip extension (thigh in line with trunk, hips straight, e.g., standing); Foot plantar flexion (foot down, walk on toes)
<b><i>Sacral</i></b>		
S2, S3, S4	Anal Sphincter Urethral Sphincter	Bowel function (fecal continence) Bladder control (urinary continence)

Source: W.C. Stolov and M.R. Clowers (1981), *Handbook of Severe Disability*. Washington, DC: U. S. Government Printing Office, p. 67. Reprinted by permission.

## Levels of Injury and Functional Expectations

Table 1 summarizes the functions that are made possible by the nerves at various spinal levels. For individuals with complete lesions, patterns of functional loss and preservation are fairly consistent from person to person. The most dramatic changes in function are apparent between adjacent neurological levels in the cervical area. For example, nerves that innervate the diaphragm are at cervical level 3-4, so many persons with injuries at or above C-3 need ventilator assistance to breathe. Most individuals with C-4 injuries regain breathing capacity, but do not have usable function in their arms and hands. As a result, they need assistance with virtually all activities of daily living (ADLs), including feeding and dressing.

Individuals with injuries at the C-5 level usually have function in the deltoid and bicep muscles, so they will be able to bend the arm at the elbow. Some have the ability to develop a weak pinch by using an automatic motion known as tenodesis (when the wrist is extended, the thumb and index finger come together). This makes it possible to hold a light object (such as a washcloth) and carry out some self-care activities. Most people with C-5 injuries use power wheelchairs for mobility.

Many individuals with C-6 injuries use manual wheelchairs, particularly if the chairs are equipped with plastic wheel rims and projection knobs. Most people with C-6 tetraplegia can move independently with the use of appropriate equipment. Assistive devices (e.g., tableware with built-up handles) make many activities of daily living possible.

People who have C-7 and C-8 levels of injury are nearly independent in their daily lives. They have the use of tricep muscles (which allow them to straighten and lock their arms for transfers from wheelchair to automobile or chair) and finger extensors (which allow them to open their hands). Usually, they manage to use regular automobiles rather than needing expensive modified vans.

At the thoracic levels, the nerves innervate muscles that provide control of the trunk, so injuries below this level allow for some balance and trunk stability. Control over hip muscles is maintained at the lumbar level, so individuals with injuries below T-12 are sometimes able to ambulate using crutches and braces. This takes a great amount of effort, however, and most people choose to use wheelchairs for mobility, particularly when they are covering anything except short distances within a home or workplace.

Injuries at the lumbar level often result in paraplegia. Persons with L1-L2 injury may be capable of standing and walking with braces. However, this involves a great amount of energy even for short time periods; a wheelchair will be needed for mobility of any significant distance. Those with an L3-L4 injury also may be able to walk with orthotic devices.

Innervation for bowel, bladder, and sexual functioning occur at the sacral level. Persons who have injuries at the cervical or thoracic level have sustained damage to all of the upper motor neurons that innervate the body at or below the point of injury. This results in loss of voluntary control of bowel and bladder functioning. Fortunately, an intact lower motor neuron reflex arc may be retained from these muscles to the synapse in the spinal cord and back. This enables the possibility of “retraining” the body to respond to direct stimulation (for example, tapping on the lower abdomen to trigger voiding) and provides some degree of control over bowel functions. Individuals who have sacral injuries, on the other hand, may have direct injury to the lower motor neurons in this area. Thus, they may not have the ability to develop these reflexive responses.

## Selected Common Sequelae of SCI

### *Bowel and Bladder Dysfunction*

Following SCI, most individuals lose voluntary control of their bowel and bladder and need to develop ways to manage these functions. The bowel can be trained to empty at regular intervals (say, every day or two) in response to a suppository and/or digital stimulation. For people who are working, the time that is required to complete the bowel program (up to two hours) can affect productivity. People need to carefully monitor the foods they eat and avoid things that cause irregularity or intestinal irritation. Prospects of an involuntary bowel accident in public may lead some individuals to avoid activities, including work that they might otherwise enjoy. Bowel regulation becomes a greater problem as people age, as intestinal function



changes and tissues suffer from years of external stimulation. If bowel management becomes highly problematic, physicians may recommend a colostomy (surgical procedure that provides for emptying fecal contents through an abdominal stoma). This procedure may reduce the amount of time required for bowel care and serve to prevent accidents.

Bladder and kidney functioning is a complex issue following SCI. Persons who retain reflexive function in the bladder may be able to initiate voiding, but those who have a flaccid bladder need catheterization or external pressure to void. Years ago, it was common for people to use indwelling catheters that were continuously in place. Now, intermittent catheterization, several times each day is possible (National Spinal Cord Injury Statistical Center, 2005)

Many people have recurrent episodes of urinary tract infections that cause fever and other symptoms. Infections develop because the bladder does not empty often enough or completely, allowing bacteria to multiply. Without sensation, a person does not receive automatic signals indicating the bladder needs to be emptied. If the bladder becomes overly full, urine backs up into the kidneys (a condition known as reflux) and causes damage. For many years, renal problems were the major cause of premature death among persons with SCI (DeVivo, Black, & Stover, 1993), but antibiotics and better preventive care have reduced the danger. Yet, some people face new dangers from years of antibiotics to control infections; they may develop allergic reactions or resistance to all antibiotics.

### ***Sexual Dysfunction***

Research has been done in recent years with the goal of diminishing the impact of SCI on sexual functioning. This is an area that merits attention and counseling because of its importance within marriage and as a component of personal identity and esteem. As with the bladder, the nerves that innervate genitalia are almost invariably affected by SCI. Sensation may be altered or eliminated. The connection between sexual arousal, which occurs in the brain and the genital organs is usually broken, making it impossible for most men to attain an erection or women to produce lubrication in response to psychological arousal. On the other hand, if the lower motor neuron reflex arc is intact, a physical response may be produced by direct stimulation. For men with SCI between T6 and L5, sildenafil (Viagra) has been shown to significantly improve the quality of erections and lead to higher levels of satisfaction (Derry et al., 1998). Other techniques have also been developed to enable intercourse including penile implants and injections.

A woman who has an SCI frequently does not have a menstrual period for several months after the trauma. If a woman is pregnant upon injury, however, fertility is ultimately unimpaired. Many women with SCI remain fertile and can become pregnant. A woman will require prenatal and delivery care from a knowledgeable physician to avoid complications resulting from the pressure that a growing fetus places on her lungs, bladder, skin, and circulatory system. Special issues may arise during childbirth, in part because she may not experience the typical sensations that signal the onset of labor. Care must be taken to manage any occurrence of autonomic hyperreflexia during labor. A woman with tetraplegia needs to plan the way in which she will be able to care for a baby – perhaps with help from a personal care assistant.

For men, fertility has often been a more difficult challenge. Ejaculation typically does not occur after SCI, or semen may be deposited into the bladder rather than being discharged externally. Sperm motility may be so low that pregnancy is unlikely to occur even with ejaculation. In recent years, clinics have successfully used techniques such as electrical stimulation to induce ejaculation, and a number of couples have borne children as a result. Many couples use artificial insemination to start families.

### ***Autonomic Hyperreflexia***

This is potentially life-threatening for persons with injuries above the level of T-6 (Staas et al., 1993). In response to some kind of noxious stimulus (e.g., a urinary tract infection, a blocked catheter, or even wrinkled clothing that is irritating the skin), blood pressure shoots up, producing symptoms of a crashing headache, dizziness, and sweating. Unless treatment is quickly obtained to lower blood pressure, the individual is at risk of a stroke. Persons whose injuries place them at risk of hyperreflexia (also called dysreflexia) need to be aware of the symptoms and treatment.

### ***Spasticity***

Immediately after SCI, the muscles that would be innervated by neurons below the lesion are usually flaccid. The upper motor neurons that were destroyed do not recover, so voluntary motor function will not return. The lower motor neurons some distance below the lesion may not have been directly injured; however, as the initial shock passes, they may begin to conduct signals in a reflex arc from the peripheral nerves to the synapse in the spinal cord and then back to the stimulated muscle. In an intact nervous system, the signal goes to the brain and the brain modulates the body's response. A lesion blocks that pathway and sets up, in effect, a reverberating circuit. Resulting spasticity can be quite disruptive and may cause pain and possible embarrassment.

A positive side of spasticity is that it can serve to help maintain tone in paralyzed muscles and may be functionally useful in some instances such as in facilitating wheelchair transfers. At one time, Valium was routinely prescribed to control spasticity, but it often affected mental clarity, making studying and working difficult. Fortunately, less psychoactive medications are now available. Other procedures such as motor point blocks (an injection of chemicals into the muscle to block nerve impulses) and surgery are tried when other means of control are ineffective. Nevertheless, using prescription medications to treat spasticity and other complications (pain, sleep loss, depression) are associated with greater risk of additional injuries due to events that occur in the years and decades after the initial injury (Krause, 2004).

### ***Contractures***

Joints need to be moved through their full range of motion regularly; if they are not, the muscles shorten into permanently flexed positions known as contractures. The fingers of many individuals with quadriplegia are bent and some people develop permanent contractures in their hips, knees, and elbows. Hygiene is made difficult by this permanent contracture and any functional capacities that the individual may possess are compromised. Rehabilitation nurses and physical therapists make a concerted effort to avoid development of contractures, and range of motion exercises need to be a continuous daily routine.

### ***Pressure Sores***

Decubitus ulcers, also known as pressure sores, are a frequent and costly problem for individuals with impaired sensation. Whereas people with intact nervous systems shift their positions at frequent intervals due to signals of discomfort that may be barely conscious, those with SCI need to train themselves to shift their weight regularly. Krause (1998) surveyed 1,000 people with SCI and found that nearly half of them reported having at least one skin sore within the past two years. Studies have been done in an effort to identify the variables that contribute to pressure sore development, and the results have been variable (Garber, Rintala, Hart, & Fuhrer, 2000; Krause & Broderick, 2004). Pressure sores are frequently preventable, but people who are depressed or angry may neglect hygiene and self-care, therefore contributing to increased vulnerability (Lindsey, Klebine, & Oberheu, 2000). Social problem-solving ability has been shown to predict the occurrence of pressure sores in people with SCI (Elliott, Bush & Chen, 2006). Physical variables, including age, health of the circulatory system, number and type of transfers made in a typical day, could expose one to scrapes or bruises and therefore play a part. Quality of care and personal assistance is another consideration because checking daily for signs of redness on the skin is necessary for preventing development of pressure sores.

### ***Subsequent Injuries***

Subsequent injuries are those that occur in the years and decades after the initial SCI. It is rather surprising that there has been so little attention to this area, since it is predictable that the same pattern of behaviors that led to the initial SCI will elevate risk for additional event-related injuries. In the primary study in this area, 19% of individuals in a sample of 1,391 participants reported at least one other injury during the previous 12 months severe enough to require medical attention (Krause, 2004). Of those with one or more injuries, 27% reported at least one injury-related hospitalization during the same timeframe. Sensation seeking, heavy drinking, prescription medication used for pain, spasticity, depression, and lack of sleep were all associated with an elevated risk of subsequent injuries (consistent with the risk factors for the initial SCI). Because those with incomplete motor function injuries were also at greater risk, Brotherton, Krause,

and Neitert (2006) surveyed 119 participants with ambulatory SCI. Seventy-five percent reported at least one fall the previous year. Of those who reported at least one fall, 18% had a fracture and 45% reported decreased participation in community activities as the result of a fall.

### ***Bone Changes***

Almost immediately after SCI, osteoporosis begins to develop. Part of the reason for bone loss is the period of bed rest and disuse that follows trauma, but additional unknown factors are almost certainly involved (Garland et al., 1992). The loss occurs throughout the body, but initially directly affects areas below the pelvis, especially the knees. Decreased bone density makes a person vulnerable to fractures.

Another sequela of trauma, such as SCI or head injury, may be heterotopic ossification, a condition in which the body begins to develop pieces of bone in soft tissue. This phenomenon has been reported in 16% to 53% of people with SCI; reasons for its occurrence are not well understood (Stover, 1997). Besides being painful, this phenomenon interferes with functioning, sometimes to the point where joints fuse. Surgery is used to treat severe cases (Staas et al., 1993).

### ***Respiratory Problems***

Individuals with complete SCI above the level of C-4 are likely to need ventilators to enable them to breathe. Survival after so high an injury was rare until recently, but with advances in emergency rescue procedures, many individuals are able to reach hospitals and receive the kind of care that enables survival. Questions have been raised about quality of life experienced by people with tetraplegia who are “ventilator dependent.” Interestingly, studies have shown that individuals with such conditions quite consistently rate their lives as more satisfying than health care professionals predicted (Bach & Tilton, 1994). With sufficient support and assistive technology, these people may be capable of working, socializing, and living rewarding lives.

Respiratory function is often a matter of concern for people with SCI who are not placed on ventilators. Because of weakened chest muscles, people with tetraplegia and those with high thoracic injuries may not be able to cough with sufficient force to clear their lungs of irritants and mucus, and are prone to pneumonia. This is especially dangerous if they have developed immunity to several antibiotics because of frequent use in combating other infections. Pneumonia is the most frequent cause of death among people with SCI (Klebine & Lindsey, 2001). Pulmonary embolism is another serious condition that occurs at greatly increased rates for this population.

### ***Cardiac Problems***

Cardiac disease is a frequent cause of death for the general population, as well as among people with long-term SCI (Coll, Frankel, Charlifue, & Whiteneck, 1998). Among people with SCI, however, heart problems appear at earlier ages. Furthermore, cardiac insufficiency may contribute to reduced energy and strength and can lead directly or indirectly to the development of other secondary disorders such as pressure ulcers. Paralysis poses formidable barriers to exercise and the maintenance of cardiovascular fitness.

### ***Pain***

The majority of people with SCI experience chronic pain that affects their functioning and quality of life (Rintala, Hart, & Priebe, 2004). They may report a sharp, stabbing pain below the level of injury that occurs intermittently, or a dull, aching pain that persists for long periods of time and becomes worse with activity. The cause for pain is not well understood and there may be no means of relief available. Individuals may strive to manage the pain with relaxation or cognitive management techniques. Use of narcotic analgesics is not recommended with chronic pain conditions.

Another source of pain comes from overuse of shoulders and upper extremities needed to propel a wheelchair and transfer from one surface to another. One study of 450 individuals with SCI found that over 70% of participants reported some degree of chronic wrist and shoulder pain (Subbarao, Klopstein, & Turpin, 1995). The pain did not respond to ordinary therapies; authors of this study recommended that alternative methods for transferring and wheelchair propulsion need to be developed to help prevent these

problems. As more persons with SCI age, chronic pain from overuse becomes an increasingly common problem.

## **Psychosocial Aspects of SCI**

Adequately describing the psychological and social consequences of SCI is complex. People experiencing objectively similar levels of trauma respond in entirely different ways as a result of their differing personalities, social support systems, cultural and ethnic backgrounds, available resources, age, education, and intellect. Every aspect of a person is affected by the injury; thus, the whole person is instrumental in shaping response to disability.

There are some characteristics of SCI that play a role in the adjustment process for most people, including:

- SCI usually involves sudden and unexpected onset. In a moment's time, life is irretrievably altered. With no opportunity for preparation, the entire adjustment process takes place after onset. In past years, hospital stays of six months to a year often allowed time for gradual assimilation of change with the support of a diverse rehabilitation team. Currently, much shorter stays lead to discharge of individuals who are just beginning to comprehend the impact of injury and plan for the future.
- The person with SCI usually experiences losses in many life areas, including the inability to participate in valued activities, financial losses related to interruption or discontinuance of work, and the burden of new expenses. Eventually, most people perceive gains stemming from the injury (e.g., deepening of family relationships, personal growth, and new educational or vocational opportunities), but awareness of losses occurs first.
- One of the most profound changes following SCI is the loss of independence in daily living. The individual with tetraplegia needs to rely on another person to provide the most basic and personal care including toileting, bathing, and feeding. The psychological stresses are different when the care provider is a loved one (a parent or spouse) compared with a stranger, but both situations are challenging. Unless the individual has personal resources to pay for a personal care assistant (PCA) privately, he or she may need to rely on inadequate public subsidies to recruit a helper from a pool of marginal workers who are willing to apply for minimum wage positions. Many individuals have reported harrowing experiences stemming from theft, abuse, and neglect by PCAs.
- SCI affects mobility, so individuals must be continually alert to accessibility. The Americans with Disabilities Act (ADA) and previous legislation have been helpful in increasing the number of public places that are wheelchair accessible. Many places are only partially compliant, and private structures such as homes are often inaccessible. Each new experience involves uncertainty.
- The bodies of persons with SCI are a further source of insecurity because of the loss of voluntary control of functions. A bowel or bladder accident or even a bout of spasticity may bring unwanted attention and embarrassment. Some people choose to stay at home rather than risk such occurrences.
- SCI is a visible disability, so the individual is subject to the attitudinal barriers of people who are uncomfortable with or biased against people with disability. Although the stigma of physical disability is apparently not as profound as that attending other conditions, it may still compromise social interaction and job opportunities. Many persons have reported that pre-injury friends drift away and new acquaintances must be made to avoid isolation. The incidence of marriage is lower and that of divorce is somewhat higher among individuals with SCI compared with the general population, but many do sustain satisfying intimate relationships (Crewe & Krause, 1992; DeVivo, Hawkins, Richards, & Go, 1995).
- Some individuals with SCI must cope with additional barriers, such as substance abuse, which involves additional treatment. Up to half of the people admitted to emergency rooms with severe trauma, including SCI, were intoxicated at the time of injury (Heinemann, Doll, Armstrong, Schnoll, & Yarkony, 1991). Thus, people who abuse substances are more likely to engage in risky behavior that leads to injury. Furthermore, substance abuse may be a coping strategy subsequent to injury, and such



behavior may be enabled by family and friends who feel the individual deserves whatever happiness he or she is able to obtain.

- Depression is a significant problem for 20% to 40% of individuals with SCI (Kemp, Kahan, & Krause, 1999; Weingardt, Hsu, & Dunn, 2001). The rate of depressive disorder is approximately three times higher than in the general population. Depression can interfere with health, psychosocial functioning, and lead to suicide. The Consortium for Spinal Cord Medicine (1998) published guidelines for treatment of depression. While it is addressed to primary care physicians, the information is applicable to rehabilitation counselors and other service providers.
- People are remarkably resilient in their abilities to cope with the challenges of SCI. Psychological adjustment (as measured by satisfaction with life and involvement in work and social activities) continues to improve for many years after injury. The degree to which adjustment improves over time is impacted by the extent to which the general environment is favorable to life with SCI. A 30-year longitudinal research study identified stability and increased satisfaction with different areas of life over the first 15 years, followed by some noteworthy declines, particularly in areas related to social life, sex life, and health (Crewe & Krause, 1991; Krause & Coker, 2006). In contrast, self-rated adjustment improved, suggesting that individuals evaluate favorably their ability to adjust to the increasing obstacles introduced by aging. Without denying that SCI is a tragic event, most survivors rate their quality of life as good to excellent years after the event; the capacity to maintain a satisfying life appears to be intricately related to longevity (Krause, Sternberg, Maides & Lottes, 1997).

## **The Search for a Cure**

Writings from ancient Egypt described SCI as a condition that was untreatable. Survival prospects for those injured did not improve significantly until the discovery of antibiotics in the early 1940s. Cells within the central nervous system do not regenerate once they have been destroyed so medical treatment and rehabilitation have always focused on preventing complications and maximizing residual functional capacities. Today, researchers have been working with increased intensity to find ways to stimulate regrowth of nerve cells, to block the processes that serve to inhibit growth, and to promote connections across the site of injury (Jones, Anderson, & Galvin, 2003). Repair through transplantation of nerve cells is being studied and many believe that a breakthrough may be achieved in the foreseeable future. When this occurs, the most likely beneficiaries will be people with new injuries whose bodies have not experienced the muscle atrophy and other effects of years living with SCI. In the meantime, it is important to remember that thousands of people are experiencing fulfilling and productive lives with severe disability.

## **Employment**

The most prominent benefits of employment are financial. However, employment is also related to other favorable outcomes. For example, individuals who are employed after SCI consistently reported greater satisfaction with nearly all areas of life (Krause, 2003). Of even greater importance, return to gainful employment was associated with a significantly greater likelihood of surviving SCI over an extended period (Krause, 1991; Krause et al., 1997).

### ***Employment Rates***

Without question, SCI adversely impacts employment outcomes for many persons. Historically, efforts have been made to identify the overall employment rate after SCI. For example, Trieschmann (1988) summarized employment rates in various studies, finding that the rates ranged from 13% to 48%. Employment rates vary dramatically depending upon characteristics of the group under investigation, including race, age, severity of injury, education, and employment history.

The most favorable employment situation is when the individual is able to return to a pre-injury job, although this is infrequent and most likely among those with less severe injuries (Krause & Anson, 1996). Among those who returned to work after SCI, it took an average of about five years to return to the first post-injury job and just over six years to return to the first full time job (Krause, 2003). Not surprising, those with non-cervical injuries returned to work about two years before those with cervical injuries.



## ***Return-to-Work Approaches***

There are two separate tracks to employment—a fast track, defined by return to the pre-injury job or having worked as a professional prior to injury, and a slow track, reflecting the absence of these characteristics. Those in the fast track took substantially less time to return to both their first job (2.1 years compared to 5.5 years) and their first full-time job (3.3 years compared to 7.1 years). It is interesting to note that differences between those with cervical and non-cervical injuries were observed only for the slow track. These findings suggest there is a window of opportunity shortly after SCI onset where there is greater likelihood of return to work if the preinjury employer is willing to rehire or if preinjury education and work skills can be applied to a new position. However, once this window of opportunity passes, it will likely take years to return to work, and only after considerable education and training, particularly for those with more severe injuries. It is likely that some individuals who do not return to work quickly may never return to work. An aggressive approach may maximize employment opportunities when compared with the frequent practice of delaying discussion of employment issues until after the person returns to his or her community and has had time to “adapt” to the injury (Krause, 2003).

## **Employment Outcomes**

It is important to understand the types of characteristics associated with differential employment outcomes. Primary factors associated with post-injury employment were race-ethnicity, age, severity of injury, etiology, duration of SCI, pre-injury employment status, and years of education. The highest employment rates were obtained by participants who had the following characteristics: Caucasian, 25 and younger at injury, ASIA D severity, not injured as the result of violence, had been injured 20 years earlier, were working at time of injury, and had completed 16 or more years of education.

Some more interesting findings related to combinations of factors. For instance, there was an interaction between race-ethnicity and gender, as men reported higher employment rates for all groups, except African-Americans where the opposite trend was observed. Similarly, being employed at the time of injury was associated with a greater probability of employment, but only during the first few years following injury, after which time this factor was unrelated to employment status. The employment rate of those working at injury ranged from 18-31% at one, two, and five years post-injury, compared with a range of 6-16% over the same timeframe for those unemployed at injury. However, by 10 years post injury the trends had reversed (33% compared with 40%).

## **Severity of Disability**

Although severity of injury and functional limitations play a role in return to work, the relationship is not simple. First, there is a greater likelihood of individuals with less severe injuries returning to their preinjury job (Krause, Sternberg, Maides, & Lottes, 1998). For example, if jobs require manual dexterity, but not lower extremity function, a person with paraplegia is more likely than someone with tetraplegia to return to this position. Another reason why people with tetraplegia find it more difficult to return to work is that they are likely to have higher medical costs, including medications, supplies, and especially for personal care assistance. Medicaid often covers these costs, and eligibility for Medicaid generally is lost when an individual begins earning more than about \$500 per month. Enactment of Ticket to Work legislation in 1999 represented an effort to change public policy to remove this disincentive to employment. Individuals with SCI cannot survive without medical services, and many jobs fail to provide health insurance or offer insurance that excludes pre-existing conditions.

## **Education**

By far the most important factor in return to work is education. It is rare for a person with less than 12 years of education to return to work after SCI, and even more uncommon for them to retain employment. For example, in one study, the percentage of participants with less than 12 years of education who were currently working was only 11% (Krause, 2003). The percentage increased with each successive milestone passed, peaking among people with more than 16 years of education. In fact, 74% of individuals with more

than 16 years of education were working at the time of the study, and fully 89% had worked at some time since injury.

The significance of education is best demonstrated in conjunction with injury. Education mediates the extent to which severity of injury is associated with diminished labor force participation (Krause et al, 1999). Among those with 12 years of education, only 8% of participants with C1-C4, ASIA A-C were currently working, compared to 33% for those with motor function injuries (i.e., ASIA D, all levels). However, the differences disappeared among those at the Master's-Ph.D. level. The benefits of further education were most dramatic for those with the most severe SCI as the employment rate for the C1-C4 group was nearly eight times greater between high school and Master's-Ph.D. level education compared with less than two times the increase for those with ASIA D. In fact, the employment rate for those with ASIA D injuries flattened, starting at an Associate's degree, but continued to increase for those with C1-C4 ASIA A-C, all the way through the Master's-Ph.D. (increasing from 36% with a 4-year degree to 61% with a master's or higher degree). In summary, education is the great equalizer of employment opportunities for people with SCI.

## **Conclusion**

A spinal cord injury usually begins with a sudden, traumatic impact on the spine that fractures or dislocates vertebrae. An injury to the spinal cord can damage a few to almost all its axons. There will be almost total recovery from some injuries. Other injuries result in complete paralysis. Improved emergency care for people with spinal cord injuries and aggressive treatment and rehabilitation can minimize damage to the nervous system and even restore limited abilities. Rehabilitation programs combine physical therapies with skill-building activities and counseling to provide social and emotional support. Spinal cord injuries are classified as either complete or incomplete. People with incomplete injuries retain some motor or sensory function below the injury. A complete injury is indicated by a total lack of sensory and motor function below the level of injury. People who survive a spinal cord injury will most likely have medical complications such as chronic pain and bladder and bowel dysfunction along with increased susceptibility to respiratory and heart problems. Successful recovery depends upon how well these chronic conditions are handled on a daily basis. Severity of disability has a profound impact on resuming employment. Education is the greatest equalizer for employment opportunities.

## **Case Study**

Charles Kopecki is 34 years of age. He sustained a C-6 level SCI a year ago in a motorcycle crash. He was returning from a friend's home on a rainy evening when he struck a patch of gravel and failed to negotiate a curve in the road, striking a tree. A passing motorist telephoned for help, and he was airlifted to a regional trauma center where he underwent surgery to stabilize his spinal column. After three weeks on the neurosurgical service, he was transferred to the rehabilitation unit where he received six weeks of comprehensive services prior to returning home. At the time of injury, Charles had been employed for nine years as a long distance truck driver for an agricultural processing company. After dropping out of high school in the 11<sup>th</sup> grade, Charles held a number of unskilled jobs in construction and food services. The Kopeckis live in a town of about 10,000 people, which is approximately 25 miles from a small city. Charles and his wife have two children, ages 9 and 10. His wife has been providing personal care services for Charles since his injury, including bathing and bowel and bladder hygiene. She works at a filling station/convenience store to supplement the public assistance income which Charles receives. Between these responsibilities, housework, and parenting, she is feeling increasingly stressed. At her encouragement, Charles scheduled an appointment with a rehabilitation counselor to discuss gainful employment. He enjoyed being a truck driver, but he does not see a future in that career.

## **Questions**

1. Provide a vocational profile for Mr. Kopecki, including age category, educational level, and work history (skill and exertional levels).
2. Does Charles possess transferable skills? If so, what are they and what vocational alternatives might provide an opportunity to use them?

3. What kinds of adaptive equipment and devices would you recommend to increase Charles' independence?
4. How would you learn more about Mr. Kopecki's psychological adjustment to his disability? What services would be appropriate for him?
5. Would you involve the family in Charles' rehabilitation counseling? Discuss.
6. Outline a comprehensive rehabilitation plan for this client.

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## Chapter 22

# CEREBRAL PALSY

*Sherwood J. Best, Ph.D.*

## Introduction

Most people recognize the words “cerebral palsy” but few are able to describe this heterogeneous and complex disorder. The first descriptions of cerebral palsy were made by Dr. William John Little (1810-1894), who described “spastic rigidity” in a group of children who had suffered brain damage at the time of their births. Called “Little’s Disease” for decades, the term cerebral palsy was first used in 1937. Each individual with cerebral palsy is unique with clinical, educational, rehabilitative, and vocational needs and outcomes that are not easily predictable.

Cerebral palsy (CP) cannot be cured by medical or educational interventions. As children with cerebral palsy grow into adulthood, they are faced with a variety of challenges related to physical and emotional maturation. Fortunately, the advent of assistive technology has done much to improve the quality of life of persons with cerebral palsy. The physical limitations should no longer define their educational or career aspirations and outcomes.

## Definition and Classification

### *Definition*

Cerebral palsy is defined as “the term for a range of non-progressive syndromes of posture and motor impairment that result from an insult to the developing central nervous system” (Koman, Smith, & Shilt, 2004, p. 1619). The components of this definition contain the following features: (1) aberrant control of movement and/or posture; (2) early onset; and (3) no recognized underlying progressive pathology (Taylor & Kopriva, 2002). Cerebral palsy is not a disease but the result of damage to the developing brain. The disorders of movement and posture are due to damage to areas of the brain that control motor function. The damage is referred to as “static;” it does not worsen. However, the dysfunction that results from muscle imbalance over time, combined with secondary characteristics of CP, contributes to more severe disability as the person ages (Benda, McGibbon, & Grant, 2003). Persons may develop scoliosis (curvature of the spine), hip dislocation, uneven bone growth, and chronic pain. These all contribute to reduced functionality and quality of life.

In addition to posture and motor abnormalities, individuals may have secondary outcomes of brain damage, including epilepsy, mental retardation, sensory and cognitive impairments, and orthopedic complications (Best & Bigge, 2005; Odding, Roebroek, & Stam, 2006). Disorders of speech, such as dysarthria (slurred speech due to muscle tightness, weakness, or incoordination) and aphasia (impairment in the ability to communicate through speech or writing) are frequent impairments (Taylor & Kopriva, 2002). However, not everyone will experience each of these impairments.

In the past three decades, the prevalence of CP has risen to over 2.0 per 1000 live births (Odding, et al., 2006). Increases in both incidence and prevalence of cerebral palsy have been attributed to improved documentation, advances in obstetrics, and medical interventions that save the lives of infants and children while failing to prevent central nervous system damage. A recent phenomenon has been the occurrence of multiple births resulting from assisted reproduction (Blickstein, 2003). Children who are the products of multiple births, or who are very premature, are more likely to have CP than children who do not experience these intrauterine and birth conditions (Sutcliffe & Derom, 2006).

### Classification

Cerebral palsy is generally classified in one of several ways: (1) area of brain damage (neuroanatomical); (2) type of movement disorder (spastic, dyskinetic, ataxic, and mixed); (3) limb involvement (topographical); and (4) function. Neuroanatomical classification describes the location of brain damage and is associated with the type of movement disorder. If damage occurs in the cerebral cortex and pyramidal tracts (nerve fibers that originate in the nerve cells in the cerebral cortex and descend to the limbs to provide voluntary control of muscles), the outcome is a movement disorder called *spasticity*. In spastic CP, limb muscles contract (tighten) abnormally, resulting in movement that is stiff and jerky. Over time, spastic muscles become shorter and exert differential pull around joints. The result is skeletal deformity, as the limbs, pelvis, and spine become misaligned.

When damage occurs in the extrapyramidal tracts (cells in the deep structures of the brain called the basal ganglia), the outcome is a movement disorder called *dyskinesia*. In dyskinetic CP, purposeful movement is distorted and muscles move randomly and involuntarily, especially in the arms, hands, and face. Movement may range from writhing to jerking to tremor depending on the type of dyskinesia. Muscle tone may be more normal when the individual is asleep.

Finally, if damage occurs in the cerebellum (tissue at the base of the brain that controls balance and coordination), the outcome is a movement disorder called ataxia. Persons with *ataxia* may have great difficulty stabilizing their gait, and walk with feet wide apart while holding the arms out for balance. Rarely do these types of cerebral palsy exist in “pure” forms. Frequently, for example, persons have both spasticity and dyskinesia, referred to as mixed CP. In addition to classification by neuroanatomy and type of movement disorder, CP also is classified by the location of limb involvement, as follows:

**Figure 1**  
**Classification of Cerebral Palsy by Location (Topography)**

<b>Limb Involvement</b>	<b>Description</b>
Monoplegia	Single limb involvement. Uncommon in cerebral palsy.
Paraplegia	Legs only are involved.
Hemiplegia	Limbs on one side of the body are involved. The arm is usually more involved than the leg.
Triplegia	Three limbs are involved, usually both legs and one arm.
Quadriplegia	All four limbs are involved. The trunk is often involved.
Diplegia	Greater involvement of the lower limbs than the upper limbs.
Double Hemiplegia	More involvement in the upper limbs than the lower limbs. One side of the body may be more involved than the other.

Classifying CP by location of damage and quality of movement may not help practitioners to understand the capabilities of persons with cerebral palsy, which is more apparent when using functional classification. Descriptions for mild, moderate, and severe levels of functional limitation are provided in Figure 2.

Adding functional limitation descriptions to other types of classification systems provides a clearer picture of the person and assists practitioners in communicating about levels of support needed. For example, a diagnosis of “severe spastic quadriplegia” suggests that the individual has tight, contracted muscles in all four limbs, probably uses a wheelchair or some other type of mobility device, and may have additional needs in performing daily living activities. However, even if the description reflects severe impairment, this is no indicator of intellect or learning potential (Best & Bigge, 2005).

**Figure 2**  
**Classification of Cerebral Palsy by Function**

<b>Mild Functional Impairment</b>	<b>Moderate Functional Impairment</b>	<b>Severe Functional Impairment</b>
Ambulation and speech are present	Impairments affect ambulation and speech	Impairments are incapacitating
Head and neck control are present	Head and neck control are affected	Head and neck control are absent or severely limited
Limitation of activity is unimpaired to slight	Limitation of activity is moderate to severe Activity of daily living and other physical activities are limited without assistive technology	Physical deformities and contractures are present Individuals are unable to complete activities of daily living or other activities without assistive technology

Adapted from Heller, K. W., Alberto, P. A., Forney, P. E., & Schwartzman, M. N. (Eds.). (1996). *Understanding physical, sensory, and health impairments: Characteristics and educational implications*. Pacific Grove, CA: Brooks/Cole.

## Etiology

Many factors contribute to a diagnosis of cerebral palsy. The neurological damage can occur before birth, during delivery, or as a result of insult to the brain before the age of two (Koman et al., 2004). The range of factors including the cause, location, time, and severity of damage helps explain why this condition is so complex and varied.

### *Causes of Cerebral Palsy*

Approximately 5,000 infants are born (*prenatal*) with CP annually in the United States (Heller et al., 1996). Brain malformation, genetic syndromes, maternal infection, and anoxia (lack of oxygen) have been identified as causes during the prenatal (pre-birth) period. Recent advances in neuroimaging have improved understanding of prenatal causes of cerebral palsy (Hoon, 2005). Prenatal brain malformation resulting in CP may be caused by genetic syndromes, acquired injuries from infections such as cytomegalovirus, or mutations. Premature infants whose birthweights are very low are at risk of injury to the white matter in the brain. As the brain matures, it is less susceptible to white matter injury, but may be more vulnerable to focal injury. Asphyxia (loss of oxygen) during the later prenatal period may damage deep structures in the brain, leading to dyskinetic cerebral palsy. Finally, multiple births place stress on the uterine environment and heighten the possibility of preterm birth, with increased risk for cerebral palsy (Sutcliffe & Derom, 2006). Blickstein (2003) reported an 800% increase in high-order multiple pregnancies (triplets and above) since the 1970s.

The birth process itself (*perinatal*) can result in cerebral palsy. Risk factors such as maternal bleeding, problems with the placenta, maternal infection (such as herpes), and obstetrical complications such as prolonged labor, use of forceps, prolapsed cord, and abnormal presentation of the infant can all result in brain injury either from direct insult to the brain or from anoxia. Although modern obstetrics have done much to eliminate birth injuries, parents must still seek proper prenatal care to support their child's development.

After birth (*postnatal*), the brain can be damaged through direct injury due to lack of oxygen (Best & Bigge, 2005). Infections to the central nervous system from encephalitis, poison, near drowning, suffocation, electrocution, and other traumatic events can result in CP. One entirely preventable cause of postnatal CP is physical abuse. Infants and young children can suffer severe head injury from shaking and

hitting, resulting in CP and necessitating special educational and vocational services (Karandar, Coles, Jayawant, & Kemp, 2004). Another preventable cause of brain injury occurs when infants and children are not properly secured in automobiles. Safety and supervision are important in avoiding preventable accidents that can result in brain damage.

### ***Diagnosis***

Due to the complexity of causes and risk factors associated with this condition, many infants may not be diagnosed for several years. To make a diagnosis, physicians rely on clinical judgment and comparison of infant function to their knowledge of typical infant development (Hamilton, 2006). Most scales of early development are heavily focused on motor skills, which are almost always absent, delayed, or distorted in infants with cerebral palsy. Physicians and other professionals look for the presence of certain reflexive motor behaviors to assist them in a diagnosis. These reflexive behaviors, common to all infants, appear and disappear at predictable times during the course of development. They are involuntary and largely mediated through the brain stem. As infants mature, these motor behaviors are subsumed by higher order brain development and become controlled by voluntary movement. Since human development is logical and predictable, the presence of these reflexive motor behaviors past the time when they should no longer be predominant is a strong indicator of brain damage and possible CP.

It is almost impossible to predict developmental outcomes in young children with CP. Physicians and other professionals need to work collaboratively with parents for information that will assist them in providing optimal treatment. Because there are many treatment options, it is easy to feel confused and even overwhelmed when making treatment decisions. A thorough understanding of available treatments and their efficacy for treating symptoms is necessary.

### **Treatment**

Persons with cerebral palsy have been the recipients of a variety of medical and therapeutic treatments. These treatments do not “cure” this condition, but are employed to manage symptoms. Because these symptoms range in severity, location, and form among different persons, treatments may be more useful for some than for others. Close communication with health providers, as well as patient and caregiver attention to treatments, ensure the best outcomes.

#### ***Medications***

Medications are used to treat associated conditions, such as epilepsy or more direct treatment of muscle spasticity. Two of the more recent medications are intramuscular botulinum A toxin (Botox) and intrathecal baclofen (Lioresal). Both medications have been used successfully with children and adults (Reading, 2004). As with all medications, caution is needed for correct use; expectations for positive results should be appropriate.

Botulinum A Toxin (BTA) is injected into spastic muscles to release tension in the feet, upper limbs, pelvis, and back, which assists in more functional walking and sitting (Koman, Paterson Smith, & Balkrishnan, 2003). It has also been effective in reducing back pain and muscle spasms in adults with cerebral palsy. Although BTA acts to relax muscles, the effect is not permanent.

Baclofen acts to inhibit spinal reflexes and is administered orally or via a pump that is worn externally or inserted under the skin. Oral Baclofen can cause drowsiness and is more efficiently delivered from the pump through a catheter to the cerebral spinal fluid in the lower back. The pump regulates the amount of Baclofen needed to reduce spasticity and also bypasses the need for repeated injections (Best & Bigge, 2005). Baclofen has been successful in reducing spasticity in the legs and also improving intelligible speech (Awaad et al., 2003; Leary et al., 2006).

#### ***Surgical Treatments***

The goals of surgical intervention are to correct defects, reduce deformity, and increase functionality. A variety of surgical procedures can be used to treat symptoms. “These may include neurosurgery to reduce hydrocephalus (fluid collecting in the brain), remove benign tumors, and eliminate intractable seizures from epilepsy” (Taylor & Kopriva, 2002, p. 393). A recent neurosurgical breakthrough



in treating spasticity is selective dorsal rhizotomy. The best candidates for rhizotomy are young persons with spastic diplegia who have ambulation before surgery. Following surgery, the patient receives physical therapy to maintain the reduced spasticity and improved function (Hagglund, 2005). Fine motor skill improvement has been observed after rhizotomy (Sandeep et al., 2002). It appears that selective dorsal rhizotomy is most successful when it is performed early in life, is followed up with intensive physical therapy, and is restricted to persons with mild to moderate spasticity.

Orthopedic surgery is complementary to surgical treatment of underlying causes of CP, such as selective rhizotomy or intrathecal Baclofen. Orthopedic surgery is frequently performed to “release” contracted muscles and improve standing and walking, as well as reduce pain, stabilize joints, enhance comfort, and ease caregiver issues (Gormley, 2001). Three fairly common orthopedic procedures illustrate the range of problems addressed by orthopedic surgeons. All are done under general anesthesia and usually require a hospital stay of one to five days.

### ***Tendon Lengthening***

Tendon lengthening involves exposing the tendon and dividing it lengthwise into two halves. The two ends of the cut tendon are then rejoined to create a longer, single tendon. The lengthened tendons reduce the tension of the muscle thus reducing muscle tightness.

### ***Tendon Transfer***

In this procedure, the tendon is removed from its point of attachment to the bone and is secured to a new site.

### ***Osteotomy***

This procedure is performed on the femur (thigh bone) to correct alignment. A metal plate and screws are inserted to hold the realigned bone in its new position until it heals. Casting after surgery may or may not be necessary. Once the bone is fully healed - usually nine to 12 months later - the metal plate and screws are removed through the original incision, usually as an outpatient operation. Children are able to walk within two days after plate removal and return to their regular activity level.

### ***Multiple Lower-Extremity Procedures***

Typically, four to seven procedures are performed at one or more locations in the legs (hips, knees, ankles, and feet) and may include any of the above procedures. Surgical management of symptoms of cerebral palsy is complex and has short and long-term effects. Many treatments are complimentary, must be goal-oriented and functional, and should be reasonable, based on such factors as family support and severity of disability (Gormley, 2001). Surgical management is supported with a variety of ongoing therapies to optimize functional outcomes.

## ***Occupational and Physical Therapy***

Occupational and physical therapists work with physicians to facilitate motor development and management. *Physical therapists* (PT) work to normalize the quality of patient movement through program planning for posture and balance, deformity prevention, and gross motor function, including walking. They work “to align the spine, legs, and feet, provide postoperative rehabilitation, assist with physical management at home, and are responsible for fitting and monitoring positioning equipment, braces, prostheses, and casts” (Best & Bigge, 2005, p. 93). *Occupational therapists* (OT) focus on development of functional skills for performance of activities in daily life. They work on eye-hand coordination skills; use of hands and arms for functional activities such as keyboarding, feeding, and writing; assess and remediate perceptual skills; evaluate the ability to organize and respond to incoming sensory information; and perform prevocational assessments (Best & Bigge). While physical therapists are more focused on motor abilities in the lower extremities, occupational therapists are focused on abilities in the upper extremities; the disciplines are meant to complement each other.

Many systems of treatment have developed in physical and occupational therapy. Two of the most widely recognized approaches are neurodevelopment treatment (NDT) and sensory motor integration (SMI). Neurodevelopment treatment is based on the work of Karel and Bertha Bobath from the 1940s. It features the use of positioning to inhibit inappropriate reflexive movements and enhance postural control. In

this way, movement potential is maximized while musculoskeletal complications are reduced. Therapy is applied within the context of functional activities that promote motivation and participation.

Sensory integration “refers to both a theory, originally developed by Ayres and a neurological process that enables the individual to take in, interpret, integrate, and use the spatial-temporal aspects of sensory information from the body and the environment to plan and produce organized motor behaviour” (Bumin & Kayihan, 2001, p. 394). Children may have dysfunction of sensory-motor integration because they lack normal motor control and therefore do not experience what normal movement “feels like.” Occupational therapists engage children in activities such as climbing, puzzle completion, ball-related games, and many others that supply sensory input, enhance body awareness, stimulate the vestibular system, and support perceptual and fine motor skills (Bumin & Kayihan).

A critical aspect of therapy support is assessment and use of orthotic devices, which include custom-made braces, splints, and other appliances. Orthotic devices are prescribed after surgery to hold muscles in correct positions and avoid contractures. They also support weak muscles and aid the person in walking. Children are taught to care for their orthotic devices as early as possible. Modern orthotic devices are lightweight plastic and can often be worn with regular shoes, a distinct improvement over the heavy, metal and leather “Forrest Gump” braces that many people associate with CP.

In addition to assisting with orthotic devices, physical and occupational therapists assess and recommend equipment for positioning and mobility (Best & Bigge, 2005). This equipment is selected for function, comfort, durability, safety, as well as acceptance by individuals with cerebral palsy and their families. Equipment to assist with sitting, standing, and lying on the side may be prescribed. Physical therapists assess and recommend mobility equipment, including wheelchairs, walkers, canes, and crutches. Wheelchairs are customized individually and include specialized wheelchairs for street mobility, road racing, and beach use.

Exercise is an area of physical activity that is often limited for persons with CP. Exercise is beneficial because it strengthens muscles, supports physical fitness, and provides opportunities for social interaction. Strength training and aquatic exercise are good forms of exercise. Strength training has enhanced outcomes for walking, running, and jumping (Morton, Brownlee, & McFadyen, 2005). Aquatic exercise is especially appealing because water helps eliminate pressure on joints and reduces the influence of gravity. The resistance offered by water promotes aerobic and strengthening outcomes (Kelly & Darrah, 2005).

### ***Other Therapies***

Many therapies have been used to treat symptoms of CP. Some are not commonly used in the United States, but have gained popularity in other countries (Rosenbaum, 2003). These are categorized as examples of complimentary and alternative medicine (CAM), defined as “a group of diverse medical and health care systems, practices, and products that are not presently considered to be part of conventional medicine” (Liptak, 2005, p. 156). Complimentary therapies add a therapeutic component to activities that children would otherwise normally perform, like riding a horse, swimming, and skiing. Alternative therapies are separate from traditional therapies and are frequently based on different and “unusual ideas of the biology of the condition to which they are being applied” (Rosenbaum, p. s91). Among others, these treatments include patterning, conductive education, the Adeli suit, equine therapy, therapeutic electrical stimulation, conductive education, and hyperbaric oxygen therapy.

#### ***Patterning***

The premise of patterning is to put the patient through a series of repetitive motor sequences, repeated daily. No studies have confirmed the claims of patterning experts regarding the long-term changes in motor ability (Liptak, 2005).

#### ***Conductive Education***

The goal of this therapeutic modality is more independent functioning through repetitive movements supported by specific verbal guidance by a “conductor.” The combinations of motor repetitions and active cognitive participation, coupled with simple adaptive equipment, makes conductive education an appealing therapy (Liptak, 2005).

***Adeli Suit***

The Adeli suit was originally developed from 1960s Russian technology for maintaining physical fitness in a weightless environment (Liptak, 2005; Rosenbaum, 2003). The tight-fitting Adeli body suit provides complete trunk stability (postural support) to allow better use of the legs, arms, and hands. Deep pressure provided by the suit on limb joints is alleged to promote coordination. The suit is uncomfortable and expensive, the therapy that accompanies its use is extensive and fatiguing, and there is no conclusive evidence that it provides lasting change in muscle tension and coordination.

***Equine Therapy***

Hippotherapy (horse-assisted therapy) works to improve balance, posture, and coordination in persons with CP as they adjust to the movement of the horse beneath them. Riders must keep the head and trunk controlled while astride the horse, and horseback riding provides incentive for these tasks.

***Therapeutic Electrical Stimulation***

This procedure involves applying subthreshold electrical stimulation on top of or through the skin to affected muscles. The stimulation is purported to increase blood flow to the affected muscle, which helps it increase in bulk (Liptak, 2005). Studies found lack of improvement in range of motion, muscle growth, degree of spasticity, and motor function (Dali, Hansen, & Pedersen, 2002).

***Hyperbaric Oxygen Therapy***

While not categorized as a CAM, hyperbaric oxygen therapy is another therapeutic approach for persons with CP. The assumption of this therapy is that areas of the brain near damaged areas can be “re-awakened” with oxygen added in increased concentrations via a hyperbaric chamber. Evaluations of this therapy have not resulted in any lasting results other than the positive effect of being involved in clinical trials (Rosenbaum, 2003).

## **Educational and Psychosocial Implications**

### ***Early Intervention***

Whatever medication, surgical, and therapeutic experiences the child encounters is secondary to the fact that the typical experiences of childhood are necessary for growth and development. Early intervention provides many opportunities for development of the young child with cerebral palsy. Supported by federal law since 1986, early intervention is available for families of infants and young children. It provides opportunities for the child to develop in the domains of gross/fine motor skills, self-care, social/emotional, communication, and cognition while receiving services in the home, at a center for children with special needs, or with typically developing peers. In addition to specific structured activities, many skills in early intervention programs are promoted through play.

Play is a crucial activity that enhances learning in children. Many fine and gross motor skills are practiced and refined during play. Children engage in symbolic play when they “practice” adult activities such as cooking and fantasy play. Play fulfills social needs as children learn to take turns, explore roles, and learn to cooperate. Because it is intrinsically motivating, play is a pleasant activity that is undertaken for its own value rather than for a particular outcome.

Many children with CP cannot play in typical ways. Their motor impairments prevent them from grasping and manipulating toys. If they have visual or auditory deficits, their interactions with toys is further limited. The intensity of their motor needs may prevent others from engaging them in play activities because there seems to be no way to do so. The outcomes of reduced play experiences affect not only the child’s motor development but also increases dependency and diminishes motivation, self-esteem, and self-competence (Miller & Reid, 2003).

### ***Assistive Technology in Elementary and Secondary School Programs***

In recent years, assistive technology (AT) has opened new worlds of exploration and interactions for children and youth with CP. AT is defined as any item, piece of equipment, or product system, whether acquired commercially off the shelf, modified, or customized, that is used to increase, maintain, or improve

the functional capabilities of a child with a disability (Individuals with Disabilities Education Act [IDEA], 2000). This means that AT can be as simple as a pencil with a thicker handle for easier grasp, a book podium to adjust height and angle of reading materials, or as complex as a computer and its peripheral elements.

Children can learn to operate computer software using a switch interface to replace the hand-controlled mouse. A “head mouse” is available that is mounted on spectacles. Head movement controls the cursor, and maintaining the head in a steady position acts to “click” the mouse. Computer programs are available that are voice controlled for the individual whose speech is adequate but hand use is not.

An exciting variation of environmental control for older children and adolescents is virtual reality play (Reid, 2004). Virtual reality technology immerses the child in an experience and allows levels of interaction otherwise prevented by the disability. For example, a child engaged in a virtual reality artistic experience can draw, play paintball or sports, or even play a musical instrument. Miller and Reid (2003) stated that “Virtual reality applications have the potential of improving life skills, social participation, mobility, and cognitive abilities, while creating a motivating experience for children with disabilities” (p. 624). In their research, children and adolescents who engaged in virtual reality experiences in sports/games, artistic activities, and technology/computer assisted activities reported increased belief in their ability, creativity, and competence.

In research on affective quality of school life, Best (1995) reported that children with visible physical disabilities (including cerebral palsy) evaluated themselves more negatively than peers without disabilities in areas of social competence, perception of acceptance by others, school attitude, and self-concept. This dynamic was more pronounced for children in mainstream settings than for children attending special day class programs. While these findings should not be interpreted to mean that children with CP should receive their education in separate environments from their peers, efforts must be made to assist them to manage social interactions, cope with teasing, and develop social competence skills.

## **Rehabilitation and Vocational Outcomes**

### ***Rehabilitation***

Persons with cerebral palsy do not acquire their disability as a result of disease, accident, or other experience later in life. Cerebral palsy is a developmental disability, a condition that begins before, during, or shortly after birth or during early childhood. Therefore, the concept of rehabilitation is actually one of “habilitation,” with professionals, family members, and the person with CP all focused on maximizing the individual’s physical, intellectual, emotional, and social potential from the time of diagnosis forward. Previous discussion of medical and therapeutic treatments indicates the need for early intensive intervention. Although beneficial for physical outcomes, medical and therapeutic interventions can complicate attainment of an appropriate and complete educational experience. Recuperation from surgeries, therapy sessions, and other necessities such as orthotics evaluations and medication regimens all contribute to lost time from school.

In the educational setting, continuation of occupational and physical therapy services, with additional services such as speech/language therapy, may distract the student from academic achievement. Lack of adequate school-based vocational preparation represents an additional hurdle to successful employment. Poor preparation of teachers to adequately address the needs of students with CP results in underestimation of academic potential and poor application of critical supports, such as augmentative and alternative communication (AAC). Family members who have shouldered the majority of caregiving may find themselves exhausted and unmotivated to act as advocates for their children. If they have spent considerable effort struggling for services and have lost trust in the professional institutions that provide services for their children, their involvement may be further reduced (Best, 2005). For conditions as complex as CP, communication, cooperation, and mutual respect among all parties is necessary to achieve a balanced and successful outcome.



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## ***Vocational Potential and Outcomes***

The vocational potential for individuals with cerebral palsy is broad and as varied as the disabilities that are associated with this condition (Taylor & Kopriva, 2002). Becoming employed and staying employed is a strong indicator of successful rehabilitation and adult achievement (Mitchell, Adkins, & Kemp, 2006). Unfortunately, proportionately fewer adults with CP are employed than their counterparts who do not have disability. When they are employed, the rate of employment drops when people with disabilities are in their 40s. Changes in health status associated with age, increased fatigue, need for medical services, and pain may account for workplace loss. Persons without a college education had the lowest employment rates throughout their life spans. Unfortunately, disability entitlements (Social Security, Medicare) received by non-working persons with disabilities may eclipse the wage rate for unskilled employment. In addition, lower paying jobs are more physical in nature and often not possible for persons with CP. Maintaining people in the workplace could include knowledge about age-related physical changes, anticipation of changes with appropriate accommodations, and improved workplace and job efficiency to reduce such factors as fatigue (Mitchell et al.).

Unfortunately, an individual with cerebral palsy may be judged more on physical appearance and motor limitations than either intellectual potential or vocational motivation. Although cognitive limitations frequently accompany CP, they are not synonymous conditions. Severity of CP is not a predictor of either intellectual ability or limitation. Unfortunately, while physical barriers can be eliminated through legislation and its enforcement, attitude is more difficult to manage (Darrah, Magil-Evans, & Adkins, 2002). McNaughten, Light, and Gulla (2003) noted that tangible benefits can be derived from hiring a person with CP that far outweigh the effort to provide accommodations. The key lies in believing that the individual can make a valuable workplace contribution.

An even more important re-adjustment in attitude is one that places responsibility for making accommodations within the community, rather than viewing the person with CP as someone who needs to “meet the challenge” of disability. Incorporating concepts such as “universal design” into living and work spaces is one example of adaptation that meets the physical needs of all individuals, including those with cerebral palsy. When all people benefit from physical barrier reduction, such as incorporation of ramps, elevators, and other devices into physical structures, or less tangible accommodations such as flexible work schedules and cyber-commuting, the partition between persons with disability and those without disability is removed (Darrah et al., 2002). Research on disability and employment strongly suggested that higher education can do much to mediate the factors that result in early employment loss (Mitchell et al, 2006). However, not all persons with cerebral palsy will be successful in college. For this reason, the implementation of supported employment programs and cognitive impairment is a positive addition to school programs. Social skill development needs to be part of a “life skills” or formal vocational preparation program.

Interactions among educational personnel and adult service delivery systems can assist with transition to work and other aspects of adult living. These interactions often occur in formal meetings such as those to evaluate the Individual Education Program (IEP) and Individual Transition Program (ITP) while the individual is still in school. Connections to Departments of Rehabilitation, post-secondary vocational training programs, and job placements can be made. Carryover of services that aid in daily living, such as transportation, equipment provision and maintenance, leisure activities, and health care, must be maintained even when the person shifts from school to community and state agency support (Ko & McEnery, 2004). In all these service areas, adolescents and their family state that caring and supportive professionals, clear communication and readily available information, reduced struggle for services, and awareness of the needs created by disability were strong indicators of satisfaction with service delivery.

## **Assistive Technology**

The role of AT in providing educational and vocational access for persons with cerebral palsy is clear. Within the broad arena of AT is a sub-category known as augmentative and alternative communication (AAC). AAC attempts to compensate, either temporarily or permanently, for the impairment and disability patterns of individuals with severe expressive communication disorders. It can



augment speech that is present but difficult to understand or be an alternative for speech that is absent. Not every individual requires AAC, but it is a revolutionary breakthrough for persons who previously were unable to express their thoughts and feelings. AAC systems can be simple, such as two-dimensional picture-based communication boards that are accessed by pointing. They can be complex electronic, dedicated communication devices that store and retrieve entire messages and output them through synthesized voice. These high tech devices frequently interface with computers, Personal Digital Assistants (PDAs [e.g., “palm pilot”]), and other systems so that messages can be downloaded and placed into word processing documents. The current communication possibilities with AAC are almost limitless (Best, Reed, & Bigge, 2005).

Persons who use AAC face unique issues of employment and co-worker interaction. Discussion with a group of AAC users who were fully employed in competitive job situations included three themes: (1) barriers to employment; (2) necessary supports for employment; and (3) recommendations for improving employment outcomes for persons who use AAC (McNaughton et al., 2002). Barriers to employment activities included negative attitudes of others, poor educational attainment, lack of technological training, policy and funding shortfalls, inadequate personal care/support services, and problems with transportation. Supports for employment include the user’s personal characteristics, education, work experience, family assistance for transportation and personal care, ability to use technology, the presence of workplace mentors, and legislative supports, such as the Americans with Disabilities Act of 1990 (ADA). Recommendations for improving employment outcomes included strengthened vocational training and job search skills in educational systems, personal persistence and use of available resources, changes in AAC systems such as better portability, enforcement of zero tolerance workplace discrimination, more stringent enforcement of the ADA, and tax relief for consumers in the areas of attendant care and technology equipment and services.

Both AT and AAC are specifically legislated rights for children through the Technology-Related Assistance for Individuals with Disabilities Act (also called the Tech Act Amendments) and the Individual with Disabilities Education Act of 2000. The ADA and Section 504 of the Rehabilitation Act of 1973 can be used to support AT and AAC equipment and services for adults. Many avenues of funding exist for the assessment and purchase of AAC systems. Children may be eligible through their school, therapy providers, or private insurance, while adults can work with state rehabilitation agencies.

## **Quality of Life**

Quality of life is difficult to define and yet desired by all. It encompasses the opportunity to be an active part of one’s community and engage in interactions critical to personal well-being (Best, 2005). In addition to objective factors such as physical health and material security, it includes subjective factors such as personal satisfaction and feelings of competence and empowerment.

Like anyone else, persons with cerebral palsy have interests in life fulfillment beyond good education and employment satisfaction. Attainment and maintenance of physical and material well-being, social relations, participation in community and civic activities, personal development and fulfillment, and recreation all indicate good life quality (Best, 2005). These more inclusive outcomes should be the goal for educators, rehabilitation counselors, or other professionals who assist persons with cerebral palsy to reach their potential.

## **Case Study**

Justin is a 20 year-old male who graduated high school last year. At two years of age, he was diagnosed with cerebral palsy. The cerebral palsy involvement was later diagnosed as quadriplegia (involving all four limbs), and of Class III degree of severity (moderate to great limitation of activity). In his early years of school, Justin was enrolled in a special class for students with orthopedic impairments. During the four years from kindergarten through third grade, Justin was frequently hospitalized for respiratory problems and other illnesses associated with his physical impairments. This led to his falling increasingly further behind in school. While he was not unhappy in the environment, his parents felt that Justin would be

challenged to excel at a higher level, both academically and socially, if he were transferred to general education classes with his non-disabled peers. His teachers agreed.

From the third grade (which he repeated) until graduation from high school, Justin was included in age-appropriate general education classes with support from special education. Primarily, Justin's special education support involved the provision of physical therapy to assist him in increasing mobility, and speech and language therapy using augmentative communication devices to help in developing communication skills. Special education teachers provided assistance in developing his computer skills and helping him learn to use special switches, as well as other assistive technology to access his computer and thus, the academic curriculum.

Despite undergoing numerous surgical procedures (thereby missing school) as part of the orthopedic management of his contractures and deformities related to cerebral palsy, Justin maintained average achievement from elementary through high school. Since elementary school, Justin has been non-ambulatory and has used a motorized wheelchair. The wheelchair has a special tray designed to carry a lap top computer. With support from a paraprofessional assistant who helps him with personal care and eating, Justin is able to perform all the learning activities required in school.

Justin is sure he will be able to succeed in the world of work. His goal is to obtain a job in the computer field and work while attending university classes part-time until he graduates with a degree in computer science. Using several different assistive technology devices, Justin has developed a high level of proficiency with the computer. Throughout school, he was a member of a group of young men and women who frequently met during high school to play computer games. Justin not only excelled at these games but was able to alter many of them to make them more challenging and entertaining.

In the past year, one of Justin's friends from the group found a position with a computer game manufacturer as a programmer in research and development. This friend assured Justin that the company would be interested in hiring him also. The pay is excellent and includes benefits; Justin feels that he is more knowledgeable than his friend in this particular area. He could not think of a single reason not to seek employment with this company – until he spoke to his parents.

His parents explained that, while they wanted to join him in his excitement, they were very concerned that Justin complete college before becoming employed. They reminded him that technology is developing at a phenomenal rate and many technology companies are unable to keep pace and have layoffs or declare bankruptcy. Without a college degree, Justin could find himself out of work with no prospects for finding another job. They asked him if he knew the expectations of the company for work output and whether they expect employees to be able to work at a certain pace.

This individual knows what he wants to do. The Americans with Disabilities Act provides some workplace protections against discrimination based on disability and Justin is aware of this. Although he respects his parents and knows they have his best interests at heart, he believes this job is an opportunity of a lifetime. Justin reaches a compromise with his parents by agreeing to speak to his rehabilitation counselor at the Department of Rehabilitation and seek the counselor's advice. Both Justin and his parents agreed to seriously consider the counselor's ideas.

### ***Questions***

1. The vocational rehabilitation counselor needs to have basic information about the client to recommend a realistic vocational goal. What is this information?
2. Where does the counselor obtain this information?
3. As the counselor, identify how you will handle the dilemma regarding Justin's potential job and his completion of college.
4. Are the career goals of Justin and those of his parents realistic and compatible with his capabilities and limitations?
5. Identify other possibilities open for Justin as a client of the Department of Rehabilitation.
6. What obstacles to employment may Justin encounter? How limiting is his disability and society's perception of him, including potential employers? Include a discussion of the concept of a "disabling environment."

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## About the Author

Dr. Sherwood J. Best is a Professor and Coordinator of the program for Physical and Health Impairments at California State University, Los Angeles. In 1995, Dr. Best received her Ph.D. from the University of California, Riverside, focusing her research in the area of psychosocial aspects of physical impairments. Dr. Best has presented at many local, state, and national professional conferences and is the Past-President of the National Division for Physical and Health Disabilities (DPHD) of the Council for Exceptional Children and the California Association for Physical and Health Impairments (CAPH). In 2003

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## Chapter 23

# MULTIPLE SCLEROSIS

*Barbara Barton, Ph.D., M.S.W.*

## Introduction

Multiple sclerosis (MS) is a chronic, variable, and often progressively disabling autoimmune disease of the brain and spinal cord. Some people experience periods of disease stability (remissions) followed by a relapse in symptoms (exacerbations), others experience a steady progression of their disease course, and for a few, the disease remains relatively benign over the course of a lifetime. Because of the variability of MS and the broad range of symptoms, it is important that rehabilitation counselors understand the scope of MS and its impact on vocational rehabilitation from an individualized perspective. This chapter discusses the etiology, symptoms, treatment options, psychosocial considerations, and vocational implications of multiple sclerosis.

## About the Disease

According to the National Multiple Sclerosis Society, an estimated 400,000 people live with MS, and each week 200 people are newly diagnosed with the disease (Brill, 2008; Shannon, 2007). MS is an inflammatory disease of the body's T cells that target the myelin that surrounds nerves in the brain and spinal column, causing sensory and motor problems that produce symptoms such as fatigue, weakness, muscle stiffness, poor coordination and balance, tingling, numbness, slurred speech, cognition problems, and changes in bowel/bladder function (Bren, 2005). Not every individual with MS experiences all these symptoms and the degree to which they occur varies distinctly from individual to individual. Symptoms are identifiable when areas of the white matter of the central nervous system (CNS) are inflamed (plaques), leaving multiple scarring (sclerosis) and possible permanent damage through destruction of the myelin sheath that covers and protects axons (Rodriguez, 2008).

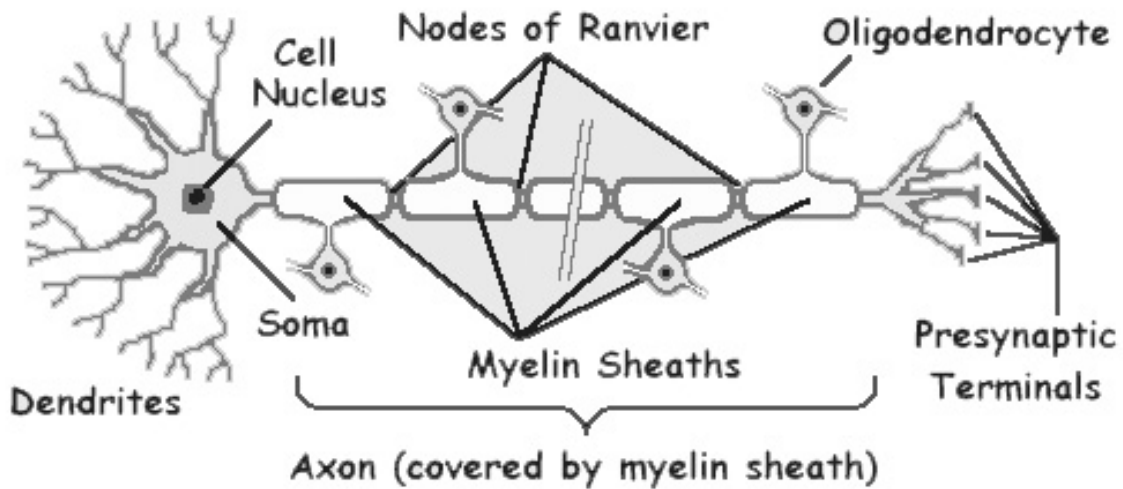
The etiology of MS is as perplexing as the disease itself. Scientists believe that the disease is caused by more than one factor, such as individual genetic susceptibility to the disease and an unknown environmental trigger that targets the immune system to attack the central nervous system (MS Active Source, 2006).

## Diagnosis and Progression

It often takes several years to confirm a diagnosis of MS due to the fluctuating nature of the disease. Diagnosis is often made through a process of elimination of other possible conditions. In 2005, revised diagnostic criteria were published, called the McDonald Diagnostic Criteria for MS (Rodriguez, 2008; Shannon, 2007).

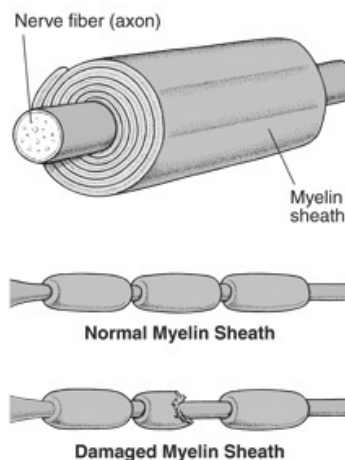
- At least two attacks with objective clinical evidence of at least two lesions.
- At least two attacks with objective clinical evidence of one lesion plus dissemination in space shown on MRI, or two or more Magnetic Resonance Imaging (MRI) lesions consistent with MS plus positive cerebrospinal fluid (CSF) finding or second clinical attack.
- One attack with objective clinical evidence of at least two lesions plus dissemination in time on a second MRI or second clinical attack.

Figure 1  
Neuron



Source: <http://www.mult-sclerosis.org/howms.html>

Figure 2  
Insulation of a Nerve Fiber



Source: <http://www.merck.com/mmhe/sec06/ch092/ch092a.html>

- One attack with objective clinical evidence of one lesion, plus dissemination in space shown on MRI, or two or more MRI lesions consistent with MS, plus positive CSF finding and dissemination in time shown on MRI, or second clinical attack.
- Insidious neurologic progression suggestive of MS plus one year of disease progression determined retrospectively or prospectively and two of the following: positive brain MRI result (nine T2 lesions, or at least four T2 lesions with a positive Visual Evoked Potential), positive spinal cord MRI result with two focal T2 lesions, and positive CSF findings.

There are five internationally recognized forms of MS: (a) relapsing-remitting, (b) secondary-progressive, (c) primary-progressive, (d) progressive-relapsing, and (e) benign. **Relapsing-remitting** MS is

marked by periods of symptom flare-ups followed by disease stability. The individual may return to full functioning or sustain some disability as a result of the exacerbation. Approximately 55% of people have relapsing-remitting disease. Over time, however, it is not unusual for these individuals to develop the second form of MS, **secondary-progressive**, which occurs in approximately 50% of individuals with relapsing-remitting disease within ten years, and in nearly 90% of persons with relapsing-remitting disease after 30 years. In secondary-progressive disease, there is incomplete recovery of function between exacerbations, resulting in progressive disability. Approximately 30% of individuals are diagnosed with secondary-progressive MS; approximately 90% of these people were initially diagnosed with relapsing-remitting MS.

The third form of multiple sclerosis, **primary-progressive**, is marked by a steady progression of symptoms without remission, resulting in almost complete functional disability. Approximately 10-15% of all people with MS experience this form of the disease. The fourth form of MS, which many neurologists consider a variant of primary-progressive, is referred to as **progressive-relapsing** and affects approximately 1% of the MS population. This type of MS begins as a progressive form, but has occasional superimposed exacerbations. Although it is called progressive-relapsing, it may be a variant of either secondary-progressive or primary-progressive. There are currently treatments available to slow the progression of the disease for those with relapsing-remitting and secondary-progressive, but no treatment has been found effective against primary-progressive and progressive-relapsing MS. About 10% of persons exhibit a clinical course characterized by relatively few and mild exacerbations; this is called **benign** MS. It is not possible to predict which individuals will exhibit a particular course. The type of disease can only be identified retrospectively (Brandes & Willmott, 2002).

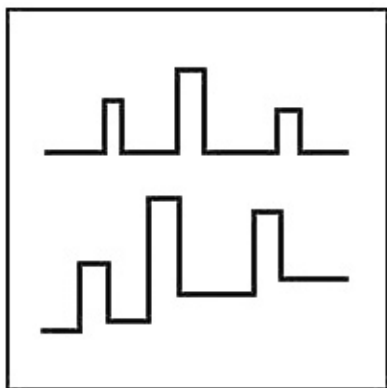
Diagrammatically, the progression of disability for each form of MS appears as follows in Figures 3-6 (diagrams retrieved from [www.mult-sclerosis.org](http://www.mult-sclerosis.org)).

Multiple sclerosis most often occurs during an individual's prime of life. Although rare, it can affect children and older adults, but MS is diagnosed most frequently in people between the ages of 20 and 50, and more frequently in women in this age category by a 2:1 ratio when compared to men. It is not contagious, and except when complications arise, is not considered life-threatening (Blackstone, 2007).

## Treatment

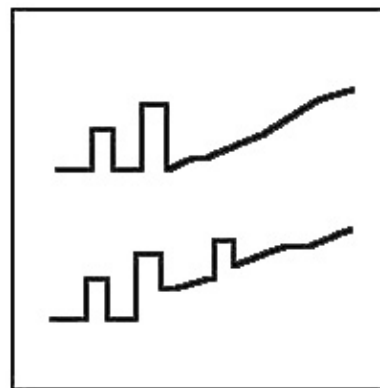
There is currently no cure, but several treatments are available to slow the progression of the disease and disability. The most recently approved treatment (Bren, 2005; Rodriguez, 2008), the monoclonal antibody Tysabri, was first used in combination therapy with the interferon Avonex, but was taken off the market due to complications. The Food and Drug Administration (FDA) has approved Tysabri as a stand-alone treatment for relapsing-remitting MS.

Figure 3



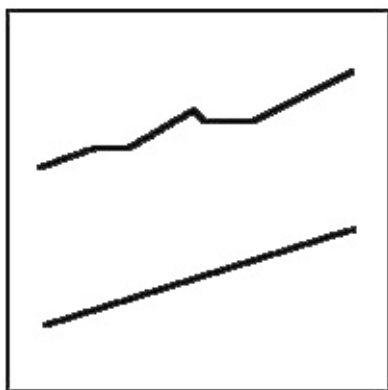
Relapsing-remitting MS and progression of disability, as evidenced by spikes. Baseline, or remission, status is evidenced by flat lines.

Figure 4



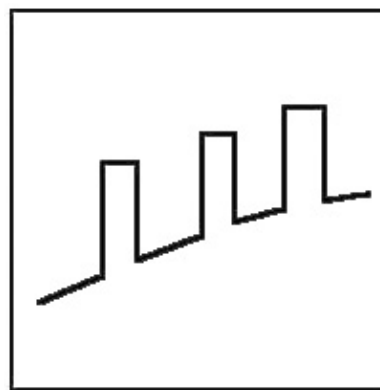
Secondary-progressive MS and progression of disability.

Figure 5



Primary-progressive MS and progression of disability.

Figure 6



Progressive-relapsing MS and progression of disability.

Each of the four injectable interferon drugs can be self-administered at home. Side effects include flu-like symptoms and potential irritation at the injection site. Copaxone injections may result in a short-term reaction that includes heart palpitations, chest pain, flushing, and shortness of breath. The National Multiple Sclerosis Society recommends patients begin one of these treatments as soon as possible following diagnosis to decrease damage to the nervous system (Shannon, 2007).

Acute exacerbations of the disease are frequently managed with high doses of intravenous (IV) methylprednisolone (Solu-Medrol) which reduces the severity and duration of exacerbations (Brandes & Wilmott, 2002). This steroid, like many others, has multiple effects, including increased blood pressure, hyperexcitability, increased blood glucose levels, and weight gain. Long-term use can lead to loss of bone

**Table 1**  
**FDA-Approved Drugs for the Treatment of Multiple Sclerosis**

Brand Name	Generic Name	Manufacturer/ Distributor, Year of FDA Approval	Indication (From FDA-Approved Labeling)	Frequency of Injection
Betaseron	interferon beta-1b	Berlex Laboratories Inc., 1993	Relapsing forms of MS	Every other day
umAvonex	interferon beta-1a	Biogen Idec, 1996	Relapsing forms of MS	Once a week
Copaxone	glatiramer acetate	TEVA Neuroscience Inc., 1996	Relapsing forms of MS	Every day
Novantrone	mitoxantrone	Serono Inc., 2000	Worsening relapsing-remitting MS and progressive-relapsing or secondary-progressive MS	Four times a year; lifetime limit of 8,12 doses
Rebif	interferon beta-1a	Serono Inc., 2002	Relapsing forms of MS	Three times a week
Tysabri	natalizumab	Biogen Idec and Elan, 2004; marketing suspended Feb. 28, 2005, due to reports of two serious adverse events	Relapsing forms of MS	Every four weeks

Source: National Multiple Sclerosis Society

mass (osteoporosis). Unfortunately, Solu-Medrol is the only known effective treatment for the management of acute relapses. Anecdotal reports indicate that complementary medical interventions such as massage, Reiki, acupuncture, a balanced diet, and nutritional supplements help promote relaxation and relieve stress that accompanies the disease. Scientific data on the benefit of these interventions is sparse, but they may be useful for the overall well-being of the person (Rosner & Ross, 2008).

Contemporary research on MS is focused on further understanding the etiology of disease and identifying safe and effective treatments for all forms. Halting the attacks on the myelin structures, repairing nervous system damage, and identifying effective treatment agents are other focal areas of research (Blackstone, 2007; Rodriguez, 2008).

## Symptoms

Symptoms of multiple sclerosis arise when an area of the CNS becomes inflamed during the early course of an exacerbation. These symptoms may persist once scarring develops within the nervous system. If the affected areas are axonal nerves, damage can be permanent (Rodriguez, 2008).

MS symptoms are unpredictable. A person may have visible neurological issues, such as gait disturbances, visual problems, or upper extremity weaknesses one day, but return to normal functioning within days, weeks, or months.

In relapsing-remitting MS, an exacerbation can last for a day or up to several months. Recovery rate and degree of recovery is as unpredictable as the disease course itself. In the secondary-progressive form of the disease, complete resumption of normal neurological status is rare. With the primary-progressive form of the disease, functional neurological status steadily declines.

A discussion of the various symptoms of multiple sclerosis follow which include heat sensitivity and overwhelming fatigue. Prior to the existence of scientifically-based diagnostic tests for MS, a “Hot Bath” test was used with those thought to have MS (Brill, 2008). In the Hot Bath, patients were immersed in a tub of hot water. If neurologic symptoms appeared or worsened, the person was diagnosed with multiple sclerosis. However, many people experience a temporary worsening of their symptoms when the weather turns hot and humid, when they run a fever, become overheated from exercise, or take very hot showers or baths. Symptoms are generally reversible when the heat-producing source is removed and the individual is cooled. The effects produced during exposure to heat, however, do not mean that the MS symptoms are becoming worse or that nervous system damage is advancing (Shannon, 2007).

The National Multiple Sclerosis Society recommends several strategies for easing the effects of heat (Shannon, 2007):

- Stay in an air-conditioned environment during periods of extreme heat and humidity. An air-conditioned working environment may be considered an accommodation, and a personal air conditioner may be tax deductible with a prescription from a physician.
- Use cooling products (personal fans, cooling vests, neck wraps, bandanas) during periods of exercise or outdoor activity.
- Wear lightweight, loose, breathable clothing.
- Icy drinks such as “slurpees” or popsicles can provide temporary relief.
- Use an oscillating fan during indoor exercise.
- Exercise in a cool pool (<85 degrees).

Some people also experience cold sensitivity, especially related to symptoms of spasticity. It is generally recommended that persons with MS who are sensitive to temperature variations avoid exposure to extreme temperature changes.

Fatigue related to employment is defined as a state of reduced capacity for work following a period of mental or physical activity (Schwid, Covington, Segal & Goodman, 2002). Fatigue is the most common and difficult symptom of MS and occurs, in varying degree, in about 80% of cases. It is more severe than



ordinary fatigue experienced following vigorous exertion and may be similar to the severe fatigue associated with some viral infections (Brandes & Willmott, 2002). With some individuals there is such a high degree of fatigue where accomplishing any physical or mental task becomes impossible. While objective scales to measure fatigue are available, subjective diaries are used to monitor a person's fatigue triggers and patterns (Krupp, LaRocca, Muir-Nash, & Steinberg, 1989).

In addition to fatigue, approximately half the people with MS develop some degree of cognitive dysfunction, which affects ability to think, reason, concentrate, and remember (Blackstone, 2007). Cognitive functions most likely to be affected include memory, speed of information processing, executive functioning (planning and prioritizing), visual spatial functions (visual perception and constructional abilities), abstract reasoning and problem-solving, word-finding (aphasia), and attention and concentration. However, only 5%-10% develop problems that are severe enough to interfere significantly with daily life. MRI scans have indicated that the extent of demyelination within the brain is associated with indirect cognitive problems, including depression, anxiety, stress, and fatigue (Rodriguez, 2008).

Additionally, neuropsychological examinations can identify areas of deficit so that strategies can be specifically tailored for an individual. Compensatory strategies for dealing with cognitive dysfunction include the consistent use of daily planners or personal data manager devices, limiting multi-tasking, using computers, filing systems, and periodic rest breaks.

## **Other Potential Symptoms of MS**

MS has the potential to affect nearly every body system. Symptoms are frequently related to the focal area of demyelination or inflammation within the central nervous system. Possible symptom areas include the following (Brill, 2008; Shannon, 2007):

- Bladder problems - urinary retention problems, and stress and urge incontinence
- Bowel dysfunction - constipation or incontinence
- Dizziness and vertigo
- Depression, emotional changes, and lability
- Gait problems, such as spasticity, poor balance, endurance and coordination, and ataxia
- Numbness and tingling (parasthesia)
- Pain
- Sexual dysfunction - In men, erectile problems; in women, vaginismus (painful intercourse), lubrication difficulties, and anorgasmia
- Vision problems - diplopia (double vision), optic neuritis (blurred vision), nystagmus (rapid pupil movement)
- Sensory problems - intense itching, burning, or "rubber band" sensations of the skin
- Tremors
- Speech (dysarthria), word-finding (aphasia), and difficulty swallowing (dysphagia)
- Lhermitte's Phenomena - an electric shock-like sensation down the spine to the toes when a person tucks the head to the chest

## **Managing Mood Symptoms**

Research has shown the high prevalence rate of depression among people with MS (Chwastiak, Gibbons, & Sullivan, 2002; Livneh & Antonak, 2005). Contributing factors that induce such depression include focal lesions in the brain, the individual's acute response to an exacerbation, and a general fear of the unknown. Treatment with therapy and psychotropic medication provides support to patients experiencing mood instability.

The rehabilitation counselor can positively impact an individual's mood and self-esteem by sharing her or his perspective and providing encouragement, as well as understanding the value changes occurring in the person with a disability. Emotional support from the counselor provides a hopeful perspective, including (Wright, 1960):

1. *Enlargement of the scope of values* – assist the individual in identifying fully what is important in her/his life, which may have changed or expanded since the diagnosis.
2. *Subordination of the physique* – since MS is often diagnosed when a person is in the most active period of life, concerns may arise such as: “How will I be able to play with my children?” “Will I have to give up tennis?” “Can I still enjoy hiking with my significant other?” One can provide reassurance while acknowledging that some modifications of physical activity are necessary and, at times, vital to the individual's well-being. A counselor may also explore hobbies, skills, and interests with the client that do not require extensive physical stamina.
3. *Containment of disability effects* – reassuring the individual that although s/he is experiencing a chronic illness, it does not mean that one is less of a parent, partner, or friend. The character traits of caring, compassion, and socialization abilities can still be intact despite the illness.
4. *Shift from comparative to asset values* – a counselor can help identify the client's unique potential and characteristics, rather than comparing her or him with those seen in the media.

Another important framework to embrace when working with clients is the psychosocial adaptation cycle to disability, as explored by Livneh and Antonak (2005). The first stage represents *shock*, a short-term reaction to the diagnosis. The second stage, *anxiety*, is characterized by a panic-like feeling when an individual recognizes the magnitude of the disabling condition. Stage three, *denial*, is regarded as a defense mechanism used to negate the potential severity of the condition. The fourth, *depression*, is a common observance in people diagnosed with chronic illness as they begin to integrate the complexity, severity, and potential future implications of the disease. In the fifth stage, *internalized anger*, clients have feelings and behaviors of resentment, bitterness, deviousness, and self-blame. In contrast, in stage six, *externalized hostility*, an individual puts the blame for the disease or treatment failure on external sources such as friends, family, and the medical profession. The final stage, *adjustment*, occurs when an individual begins to integrate the cognitive, affective, and behavioral components of the diagnosis, and can successfully negotiate disease-related obstacles and positively pursue personal, social, and vocational goals. Just as the disease is experienced differently by each individual, so are the emotional reactions. This cycle of stages is simply an outline of possible common reactions to a diagnosis of multiple sclerosis. Every new exacerbation after a period of remission can trigger these psychosocial responses. The counselor needs an awareness of the diverse nature of the disease to avoid categorizing and stereotyping the person's physical and emotional sequelae.

## **Multiple Sclerosis and Employment**

The average age of onset of MS is 28, often occurring at the time of the height of a career path. Approximately 75% of people with MS are unemployed five years after diagnosis (O'Day, 1998). Some retire prematurely or shift to less-demanding careers. Unfortunately, this shift can lead to a decreased quality of life, social supports, and financial stability (Johnson, Amtmann, Yorkston, Klasner, & Kuehn, 2004). As a result, nearly 40% of individuals with MS desire to return to work (LaRocca, Kalb, Scheinberg, & Kendall, 1985).

### ***Vocational Potential***

An important consideration in reviewing vocational literature on MS is the date and sampling design of the study. Studies conducted prior to the advent of the immunomodulating treatments showed more severe disease along with hopeless prognoses (Johnson et al., 2004). Medications, such as the interferons which modify both the rate and severity of relapses, have helped normalize life and lead to successful employment stability. Rehabilitation counselors, with the input of other professionals such as psychiatrists and neuropsychologists, can develop employment plans which are both realistic and accommodating to individual strengths and challenges. For example, if fatigue is an issue, the counselor can assess what part of the day the person's fatigue level is at its peak. Accommodations for such fatigue could

include breaks during this time, a work schedule modified around the onset of fatigue, or telecommuting. Heat sensitivity can be addressed by utilizing indoor work sites, adaptive cooling devices, and employment in an air-conditioned environment.

Accommodations for cognitive problems which affect work performance and social relationships with peers are dependent upon severity. For some, accommodations are unnecessary. For others, however, cognitive demands cause issues such as overwhelming fatigue. A neuropsychological examination can help locate and identify in which areas workplace accommodations will be needed (Rumrill, Hennessey, & Nissen, 2008).

Taking into consideration the recent positive impact of treatments for MS, the contribution to a higher quality of life that employment provides, variability of symptoms, and the relatively young age of those affected, rehabilitation counselors need to be creative, innovative, and persistent in crafting a job with an employer from a perspective of strengths, while respecting the limitations caused by the disease.

Roessler, Turner, Robertson, and Rumrill (2005) identified six areas to be addressed by counselors, consumers, and employers to promote positive employment rates of individuals with multiple sclerosis. These areas are (a) employer support, (b) program knowledge, (c) external support, (d) service provision, (e) work potential, and (f) health care.

### ***General Attitudinal Barriers***

Positive employer relations are crucial to the work atmosphere for people with MS. Title I of the Americans with Disabilities Act (ADA) offers some protection to employees with disabilities; the National Multiple Sclerosis Society has several free resources on employment and the issue of disclosure. The “win-win” approach for employers and employees that has been suggested by researchers states that an employee should request an appointment with the supervisor or manager and disclose only what is necessary about the disease state, including any needed accommodations (Roessler & Rumrill, 1998). Barriers such as negative attitudes toward people with MS and lack of knowledge about the disease must also be addressed among fellow employees.

Public opinion about individuals with MS has not kept pace with the disease-modifying treatments that often slow the functional progression of disability. MS is still viewed by many as a disease that quickly leads to wheelchair use, communication issues, and cognitive decline. For the majority of people with MS, sporadic exacerbations and limited functional loss of ability are a more realistic appraisal of health. In many, the “invisibility” of the disease is the most frustrating factor since their external appearance to others appears normal. People find it difficult to defend fatigue, cognitive problems, bowel and bladder issues, and so forth when their disease is not visible.

The societal response to disability is often a function of the beliefs of the general population toward the disability and individuals with disabilities, in general. These beliefs toward limitations are often negative and affect the interpersonal relationships between persons with disabilities and those without disability, as well as society’s attempt to rehabilitate persons with disabilities, making rehabilitation a more challenging process (Dell Orto & Power, 2007; Rumrill et al., 2008).

## **Conclusion**

Multiple sclerosis is a challenging disease, both for the person and the rehabilitation counselor. Management of symptoms is crucial for the person with MS to decrease the feelings of hopelessness and allow for normal personal, social, educational, and vocational activities during the course of the disease. Encouraging mood stability and a positive outlook can promote resiliency and appropriate coping. Counselors need to address the impact on the family, role shifts and changes, lack of education about the disease, and pre-conceived negative notions of the future, which can strain even the strongest interpersonal relationships.

An excellent resource for professionals and people with MS is the National Multiple Sclerosis Society (NMSS). This organization maintains chapters in each state. Services such as information, referral to community resources, free literature, support and education groups, and free equipment loans provide an additional support framework.

## Case Study

This person is a 29 year-old, single, African-American female who lives alone in a suburb on the outskirts of a major Midwestern city. Tracey has been employed for four years as a fund raiser for a statewide non-profit organization which operates federal food banks. As a fund raiser, she meets with individual and corporate donors and coordinates special events throughout the state, setting up agency displays at conferences and events. This occupation requires long hours, considerable travel, and heavy physical labor, but Tracy loves it. Since graduating with her college degree in communications, she has been working for this organization.

Six months ago, Tracy began experiencing episodes of weakness in both arms and dropped a box of brochures on her way to a conference. There were also balance problems and Tracey had to stop wearing high-heeled shoes. She blamed these symptoms on overwork and being tired, so she did not see her physician. The symptoms disappeared after six weeks.

Recently, Tracy noticed numbness in her right leg; she kept ‘stubbing her toe’ as she walked. In addition, she was exhausted, as if recovering from the flu. This time she decided to see her physician. Tracy told him about her current symptoms, and those she had experienced six months earlier. The physician ordered a complete MRI of her brain and spinal cord, performed a lumbar puncture, and arranged for a visual evoked potential test. The tests confirmed a diagnosis of multiple sclerosis. Tracy had two plaques on her brain and other plaques at C-5 (the 5<sup>th</sup> cervical vertebrae) and T-11 (the 11<sup>th</sup> thoracic vertebrae). Her physician immediately referred the case to a neurologist who prescribed one of the immunomodulating MS injection treatments, which have caused additional fatigue and muscle aches.

Between the tests, her symptoms, and the physician appointments, Tracy has had to miss several days of work, which has put her behind on several important projects. In the near future, she has a golf tournament and reports that she is anxious about the long drive (2-1/2 hours), as she has numbness in her right leg and difficulty feeling the car pedals. Also, there is significant carrying and walking during the event and she is concerned that she will not be able to physically complete the tasks.

Her boss is very supportive, but travels extensively and does not micro-manage her staff; therefore, she does not have the ability to help with the event. Additionally, Tracy has not disclosed her diagnosis to anyone at the organization, including her boss.

Tracy is concerned about losing her job and wonders if she will require use of a wheelchair for mobility. She is starting to feel anxious as she reads more about the symptoms of MS. Tracy’s family, except for a brother, lives out of state and she has no close friends. Concern regarding the stability of her job is increasing and she is extremely anxious about her employment options. This individual is a bright, independent person who is having a difficult time adjusting to MS. A nurse gave her the number of the local vocational services office to explore vocational rehabilitation possibilities with a counselor.

### Questions

1. As Tracy’s counselor, where would you begin?
2. Discuss the positive and negative aspects of Tracy disclosing her MS to her employer.
3. What are some potential accommodations you would recommend to this client?
4. Identify mental health issues Tracy is experiencing and how the two of you would develop ways to manage them?
5. Explore several vocational options for this client.

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## Chapter 24

# MUSCULAR DYSTROPHY

*Roy K. Chen, Ph.D., C.R.C.*

### Introduction

Muscular Dystrophy (MD) is a group of over 40 forms of neuromuscular diseases and related myopathies. Persons differ in age of onset, etiology, severity of progression, locations of muscles affected, and mode of inheritance (Mathews, 2003; Rando, 2002; Shannon, 2004). One common characteristic present in the wide array of neuromuscular disorders is the gradual weakening of the skeletal muscles that control voluntary movements. Despite recent advances in the molecular diagnosis of defective genes and the continuing breakthroughs in curative treatments over the last decade, MD still remains an incurable disease (Fowler, 2002). This disabling illness impacts the quality of life for people affected by this condition, along with their families. In addition to learning to cope with the functional limitations associated with the progressively deteriorating physical condition, these individuals encounter attitudinal and architectural barriers to living independently and obtaining gainful employment. Collectively, they experience higher unemployment and underemployment rates as well as a lower acceptance rate for vocational rehabilitation counseling services than those with other disabilities (Fowler et al., 1997). There is an urgent need to improve the effectiveness of rehabilitation services provided for these clients.

The purpose of this chapter is to review clinical and psychosocial aspects involving MD and to offer suggestions for developing and implementing rehabilitation plans for adults with MD from a holistic perspective. This holistic approach includes addressing the disability (functional limitations and capabilities), psychological and emotional factors, education, vocational history, social resources, and belief system. Through the use of a holistic approach, counselors can help clients maximize their rehabilitation potential.

### Prevalence of Muscular Dystrophy

Although no exact incidence figures for persons with MD are available, based on the 2005 world population (6 billion, 451 million people) and the prevalence of neuromuscular disorders, it is estimated that there are at least 1.26 million children and adults with MD worldwide. In the United States, more than 250,000 Americans have been diagnosed with one of the 40 types of MD (Shannon, 2004). Unfortunately, about two-thirds of them are children (Siegel, 1999). Premature death among children with Duchenne MD (DMD) is common. These individuals rarely live past their second decade of life due to irreversible and continuous weakening of involuntary muscles affecting various body systems and organs such as the cardiovascular system, pulmonary system, and gastrointestinal system.

The American public tends to view MD as a disease affecting only children, in part due to the huge success of the annual Muscular Dystrophy Association (MDA) Jerry Lewis Telethon on Labor Day. While it is commendable of Jerry's poster boys and girls and their parents to come forth on national television in an effort to share their courageous personal stories and to plead to television viewers for generous financial support, adults often go unnoticed. There are several reasons why adults as a group fail to generate strong public interest. The debilitating physical condition impedes many people from actively participating in and organizing grass roots support groups. Mobility and transportation difficulties present major challenges for persons who do not drive. Children experience a reduced life expectancy, consequently, the number of persons with MD declines as age advances.

Based on the figure estimated by Shannon (2004), the total number of adult Americans with MD is about 83,000 – a number far fewer than the number of people with AIDS, visual impairments, or psychiatric disabilities. Additionally, due to early onset of disability, the education of children with MD is often interrupted with frequent hospitalizations and a decline in general health. Consequently, people with MD do not always have the necessary education and vocational training to assume leadership positions in professional fields. There are no prominent public figures or celebrities with MD to advocate for the cause and rights of this community. Michael J. Fox, Janet Reno, and Mohammed Ali are high profile spokespersons for Parkinson Disease. Christopher Reeves was the charismatic ambassador for people with spinal cord injuries. Elton John is a steadfast ally for AIDS/HIV. United States Senators Tom Harkin and Ted Kennedy are long-time crusaders for people with developmental disabilities. In comparison with other interest groups, there is an absence of strong political representation for MD to advocate for more funding for medical research (Sirotkin-Roses, 1991).

## **Major Types of Muscular Dystrophy**

The three most common procedures used by physicians to confirm the presence of MD are blood testing, electromyography (EMG), and biopsy (Alexianu & Gooch, 2004). The first two procedures are non-invasive while the third is an invasive procedure. MD is a taxonomy of several types of hereditary, mostly X chromosome-linked, progressive muscular disorders characterized by gradual degeneration and wasting away of the muscle cells and fibers (Siegel, 1999). The subsequent discussion examines some of the most commonly seen types.

### ***Duchenne Muscular Dystrophy (DMD)***

DMD, named after French physician Guillaume Duchenne de Boulogne, is by far the most common and severe form, with an incidence of 63 per million people (Shannon, 2004). The average onset of the disability is between two and five years of age. Most children diagnosed with DMD lose their ambulatory function during elementary school. Improper sitting positions in a wheelchair for prolonged periods of time may cause scoliosis (curvature of the spine). Early signs of the disease include frequent falls and difficulty in running, trouble changing from a squatting or sitting position, and problems climbing stairs. Initial weakness occurs in proximal muscles – particularly in the hip and shoulder. In contrast to the atrophy of muscles, enlargement of calf muscles, a sign of pseudohypertrophy, may occur. A significant number of children with DMD do not survive into adulthood (age 20) due to failure of respiratory and cardiac muscles. Like the wide range of intelligence levels exhibited in the general population, people with MD vary in their cognitive abilities. However, researchers have no explanation as to why people with DMD tend to have lower intelligence scores when compared to people with other types of MD (McDonald et al., 1995a). Reading disability due to poor simultaneous and sequential information processing has been observed in children with DMD (Hendriksen & Vles, 2006).

### ***Becker Muscular Dystrophy (BMD)***

In 1956, Peter Emil Becker, a German physician, was the first person to recognize this mild, variant form of DMD. The clinical presentations of BMD are similar to that of DMD. However, the time of initial diagnosis usually appears much later (as late as 25 years) and has a slower rate of muscle deterioration. The prevalence figure of BMD is about 24 per million people (Shannon, 2004). The absence of a vital protein surrounding muscle fiber called dystrophin distinguishes DMD from BMD. Dystrophin is needed to allow muscle cells to work properly. Dystrophin produced in people with BMD is often inadequate and of poor quality. As with people with DMD, the body simply fails to manufacture this protein. Muscle weakness begins in the lower extremities and gradually includes the upper body. Due to the late onset of the disease, individuals with BMD often ambulate until about age 30, after which they may need to use a wheelchair. The major risk in BMD is from cardiac, as opposed to pulmonary complications (McDonald et al., 1995b). Death can occur in the 20s or 30s, yet some patients live beyond this age range. Persons with BMD have longer life expectancies than those who have DMD.

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### ***Facioscapulohumeral Muscular Dystrophy (FSHMD)***

Face, shoulders, and upper arms are the three groups of musculature affected by this progressive myopathy. FSHMD is characterized by persons having difficulty raising the arms, closing the eyelids, sucking from a straw, and swallowing food. Speech may become indistinct if facial muscles continue to weaken. FSHMD affects both sexes and its usual onset is between the ages of 20 and 30. Yet, persons with FSHMD are expected to have near normal life expectancies. The prevalence of FSHMD is estimated at 10 to 20 per million (Shannon, 2004). As one of the least threatening forms of MD, this disease does not affect pulmonary and cardiac functions. Because the severity is mild in comparison to DMD and LGMD, most people do not develop pelvic muscle problems and retain an ability to walk throughout their lifetimes (Chen, 2006).

### ***Limb-Girdle Muscular Dystrophy (LGMD)***

Frequency of LGMD is estimated to be 20 to 40 per million people (Shannon, 2004). Shoulder and pelvic girdle muscles are the primary areas affected. LGMD occurs in both sexes. These individuals have weakness of proximal muscles with a moderate rate of deterioration and onset of disability is in late adolescence or early adulthood. The typical pattern of muscle deterioration begins in the hip girdle and moves toward the arms and shoulders. Weakness in the legs advances more rapidly than weakness in the arms because lower extremities bear more stress from their constant involvement in maintaining posture. In all cases, people who contract LGMD will lose their ability to ambulate and require use of a wheelchair. Life expectancy is somewhat reduced.

Autosomal diseases like LGMD are characterized by gene mutations in any of the first 22 pairs of non-sex-determining autosomal chromosomes (Mathews, 2003). The diagnosis is generally classified as either autosomal dominant form (LGMD 1) or autosomal recessive form (LGMD 2). Researchers have identified five autosomal dominant and ten autosomal recessive gene defects linked to this group of myopathies (Wicklund & Hilton-Jones, 2003). A person with an autosomal dominant disorder usually has one parent who carries mutated genes. Someone with an autosomal recessive disorder has two unaffected parents each carrying defective genes.

### ***Myotonic Muscular Dystrophy (MMD)***

The most prominent difference between MMD and other types of MD is that the distal muscles instead of the proximal muscles are the first to be affected. A major characteristic symptom is the delay in timed motor performance (Johnson et al., 1995). Persons with MMD lack strong small muscles of the hands and have difficulty relaxing a handgrip. Due to stiffness in their hands, it takes longer to let go of an object. The onset of disability ranges from early childhood to adulthood. MMD has an estimated prevalence of 1 in 8,000 people, similarly affecting males and females (Yu, Blackenship, Yang, & Lee, 2006).

Noticeable muscle deterioration in the neck and face, forearms and lower legs, cardiac dysrhythmias, endocrine problems, frontal baldness, and cataracts of the eyes are common complications (Johnson et al., 1995). The severity of this condition increases from one generation to the next (Emery, 1994). Children with MMD who inherit genetic defects show far more serious conditions than their affected parents.

## **Functional Limitations**

Physical impairments caused by MD have an adverse effect on a person's ability to function in the overall environment. Rehabilitation counselors may encounter difficulty pinpointing functional limitations associated with MD because there is no uniform symptom presentation and severity experienced (Chen, 2006). Depending on the type, its effect on quality of life varies. Counselors need to evaluate each client individually, as these muscular diseases vary greatly in symptoms, complications, limitations, and progression of the disease. Swenson (2000) suggested examining a client's functional capacity in the following areas: cognitive, physical, social, emotional, and vocational.

### ***Cognitive Functioning***

There is lack of consensus among researchers regarding cognitive deficits in people with DMD. Studies have shown that lower intelligence is one of the primary manifestations (McDonald et al., 1995a; Ogasawara, 1989). However, some investigators have argued that the correlation between low intelligence levels and presence of DMD is a consequence of experiential conditions (insufficient early education due to illness) rather than being an inherent nature of the disorder. Mean intelligence scores of people with LGMD and FSHMD measured by full scale IQ were not significantly different than the general population (Kilmer et al., 1995; McDonald et al., 1995c).

### ***Social Functioning***

Quality of social interaction suggests a person's capability in interacting with the environment. People with pleasing physiques often have higher self-esteem and egos because they have been receiving praise since early age. Freud (1963) believed that body image is formed in conjunction with the development of the ego, and interpersonal relationships are reflections of body attitude. Because people have an inborn urge to establish kinship with others, rejection can inflict major consequences toward feelings of worthiness. Disability alters not only physical functioning but also reciprocity of human actions and reactions. Walkers, wheelchairs, respiratory ventilators, and other assistive devices often evoke discomfort and fear in others. Not wanting to cause embarrassment or not knowing how to respond to the person's altered physical status, friends and associates may be hesitant as to how to interact appropriately (Brodwin & Chen, 2000). Opportunities for casual friendly gatherings and dating are further diminished due to architectural barriers and unavailability of transportation (Chen, 2006).

### ***Physical/Vocational Functioning***

Limited range of motion in the upper and lower extremities is a common complication in clients with LGMD, FSHMD, and MMD. Occurrence of muscle contractures in the legs and heels, because of shortening of muscle fibers and fibrosis of connective tissues, limit ambulatory function (Emery, 1994). In most cases, hand function is maintained long after leg function ceases. Raising the arms above shoulder level and lifting the feet off the ground becomes difficult. Buttoning shirts, twisting jar tops, tying neckties, and gripping pens pose difficulties as a result of loss of gross and fine motor movements. As muscles grow weaker, personal attendant care may be needed for personal hygiene and dressing. Lifting, carrying, and walking on uneven surfaces may not be possible and can require accommodation at the worksite. Clients with dystrophy affecting their hips and joints tend to find squatting and kneeling positions difficult or impossible. Fatigue and physical exhaustion occasionally interrupt workflow for brief moments, as people with MD often require short rest breaks to replenish energy levels.

### ***Emotional Functioning***

To revert from a self-reliant person to a dependent person needing assistance is traumatic and emotionally painful. Role reversal inflicts emotional distress. Furthermore, physical inadequacy produces self-doubts as adults with late onset MD come to terms with the reality that functional losses are inevitable, progressive, and irreversible. A study conducted by Ville, Ravaud, Marchal, Paicheler, and Fardeau (1992) found that having MD greatly impacts self-identity. It is not unusual for someone who has been performing a particular job for years to lose the identity associated with that profession when no longer able to perform the work. The self-perception of people with MD varies in accordance with the status of their ever-changing physical condition. Positive and negative moods oscillate back and forth as clients struggle to deal with stresses and attempt to develop new self-identities.

## **Employment Profiles for Persons with Muscular Dystrophy**

Of the 54 million Americans with disabilities, 26 million of them have severe disabilities (Roessler & Rubin, 2006). Evidence has shown that disability has a negative effect on employment prospects and earnings among persons in the "disabled" community. In 2000, the employment rate for the working aged (21 to 58) individuals with disabilities was 46% lower than individuals without disabilities (Hotchkiss,



2004). Findings also revealed that the median monthly earnings for nondisabled male workers between the ages of 35 to 54 were \$2,566, compared to \$1,568 among those with severe disabilities. Not surprisingly, people with neuromuscular diseases frequently are near the bottom level of income (Wevers, Brouwer, Padberg, & Nijboer, 1993). There are several reasons for the disproportionate numbers for people with disabilities and why they are unrepresented in the workforce. In the ensuing paragraph, the author will discuss those reasons from the perspectives of both client and rehabilitation provider.

Education has long been regarded as a necessary requisite for entry into the labor force. Chronic medical conditions often create insurmountable obstacles for receiving a quality education needed in today's labor market. The increasing emphasis on strong communication skills and computer literacy will close windows of opportunity for those who are not prepared. Inadequate job preparation due to insufficient education is a great concern to rehabilitation counselors. About 40% of people with disabilities never finish high school and 69% never attend college (Harris, cited in Mackelprang & Salsgiver, 1999). Another factor that affects the employment level of people with MD is the type. In a study of employment status of rehabilitation clients with MD, Fowler and his colleagues (1997) found a significant difference in employment across type. Specifically, 49% of individuals with BMD and FSHD were employed at the time of the survey, compared to 26% of LGMD and 31% of MMD.

Lack of interest in finding employment is one common reason reported by those who choose not to work. Perceived inability to perform tasks by people with MD engenders a sense of low self-esteem. Consequently, such persons may develop unfounded fears regarding failure and either not seek employment or become easily discouraged during job search. The Supreme Court's rulings in favor of employers in job discrimination trials have discouraged many individuals with disabilities and their advocates from searching for work. Such pessimistic views were corroborated by Chen's (2006) study in which he found people with neuromuscular and neurological diseases had little confidence in what legal protection the Americans with Disabilities Act provides to defend their rights to employment.

The unfamiliarity with MD among rehabilitation service providers decreases the referral and acceptance rates for vocational rehabilitation services. Rehabilitation counselors at state agencies generally do not possess sufficient working knowledge regarding MD, which in turn has led to a higher denial rate for services (Fowler et al., 1997). Of those polled, 95% of rehabilitation counselors did not know the nearest MDA chapter in which to refer clients. Due to the unpredictability of the pace of deterioration of this condition, counselors may see little vocational potential in those affected. Negative attitudes toward MD by rehabilitation counselors inevitably produce unsatisfactory rehabilitation outcomes. It is, therefore, understandable that people with MD underutilize rehabilitation.

## **Holistic Interventions**

To increase rehabilitation efficacy, counselors must distinguish the type of MD when developing vocational plans with their clients. It is a great disservice to clients if rehabilitation counselors lay out generic plans without regard to the nature of MD. Due to shortened life expectancy and progressive deterioration, mapping out a long-term career plan for clients with DMD will not always be realistic, practical, or appropriate. The complex nature of the condition makes it difficult for rehabilitation counselors to understand the general well-being of these clients. A multidisciplinary team approach draws on expertise from different professionals such as occupational therapists, psychologists, neurologists, physical therapists, rehabilitation counselors, and orthopedists. Bartalos (1990) identified key areas of quality of life that are of great concern to people with neuromuscular disorders: physical symptoms, medical complications, functional limitations, emotional symptoms, interpersonal relations, work adjustment, leisure activities, and sexual function. The following section will suggest several interventions to ameliorate the debilitating effects of MD in various areas.

### ***Psychosocial Counseling Considerations***

Clinicians have long believed that persons with MD go through stages of emotional distress similar to persons faced with dying, as described by Dr. Elizabeth Kubler-Ross. These stages include denial, anger, bargaining, depression, and eventual acceptance (Bradley, 1987; Emery, 1994). When considering stage theories, one needs to keep in mind that no two people react in the same way to the diagnosis of a severe



disabling condition, nor is progression through stages necessarily linear. Adaptation to disability is not time-limited. Rather, it is an ongoing lifelong developmental process.

Rehabilitation clients with MD face loss of mobility, reduced opportunities for employment, changes in living arrangements, and diminished social interaction. Clients with these conditions are at considerably higher risk for mental health issues; depression is a common problem among people with neuromuscular diseases (Fowler et al., 1997). In addition to social stigma, environmental restrictions such as architectural barriers, unfriendly terrain, and lack of availability of suitable means of transportation further impede persons with MD from establishing meaningful relationships with others (Chen, 2006).

To improve self-esteem and self-concept for people with disabilities, rehabilitation counselors must listen to their clients' problems with empathy and provide encouragement and support. In a multicultural society like the United States, people practice different religions and beliefs to draw upon sources of power to sustain psychological growth. Research has shown that spiritual well being is positively correlated with life satisfaction (Chen, 2006). Rehabilitation counselors need to show sensitivity toward religious beliefs that are unfamiliar to them. Because the impact of disability influences the lives of clients and their families, counseling outcomes will be more positive if the healing process invites nondisabled family members to share their own views and expectations.

### ***Vocational Counseling***

Formulating feasible vocational objectives requires collaborative efforts between clients and rehabilitation counselors. In addition to teaching job search and interviewing skills, counselors need a thorough understanding of their clients' existing functional capabilities. Without knowing the functional limitations, it is difficult to match clients' physical exertion levels with jobs they can successfully perform. For example, LGMD clients with mobility deficits may find sedentary work more suitable than work that requires walking, lifting, and carrying. Additionally, counselors can take a proactive approach to educating prospective employers about neuromuscular diseases and recommend necessary accommodations. Hicks (1998) suggested several adaptive techniques and mobility devices to help people with MD increase their functional capacities. For example, using an elevated seat cushion or a high stool can help an LBMD client get on a chair more easily rather than having to sit on the edge of a chair and rock back and forth, or pushing up on their thighs or chair arms to stand.

American society measures success by one's earning power and job status. Holding gainful employment is a way to earn peer respect and command social acceptance and high regard. The myths regarding people with disabilities and unwarranted fears of increases in medical insurance costs can dissuade prospective employers from considering hiring qualified job candidates with MD. Self-employment empowers clients by providing financial independence. Another advantage of being one's own boss is the flexibility of arranging convenient work hours. While self-employment presents a viable alternative for some rehabilitation clients to secure financial security and independence, it is by no means a decision to be made casually in light of the high business failure rate of new entrants. Counselors can consult with their local Small Business Administration (SBA) to evaluate a client's prior work experience, level of educational attainment, complexity of business, technical training, and availability of start-up business financing capital before contemplating this challenging and complex vocational route.

As the business world moves in the direction of telecommunication and computer technology, the traditional work setting in an office environment is no longer the only option. Electronic transmission of data via the Internet may soon make daily commuting to work by some employees with MD unnecessary. For those clients who prefer to use personal transportation or for whom telecommunication is not feasible, rehabilitation counselors can direct their clients to contact special driving schools for individuals with disabilities to locate engineers who perform vehicle modifications. In addition, major automobile manufacturers provide financial assistance for clients with disabilities to modify new vehicles. Having accessible transportation is a crucial step toward making people with MD more employable.

### ***Medical Counseling Considerations***

Obesity is an issue that tends to be overlooked by many people, including clients with MD. Slow metabolism coincides with low levels of physical activity. Due to diminished mobility, people with MD do

not always engage in active cardiovascular tasks, which consume a large quantity of carbohydrate, fat, and glucose intake. A nutritionist can help clients maintain their ideal weight. Excessive weight makes it more difficult to perform physical activities, already compounded by the functional limitations imposed by this disease.

Proper physical therapy and suitable orthopedic footwear can effectively prolong the duration of a client's ability to walk. Being able to ambulate independently serves as a strong psychological boost for someone with a prognosis for eventual use of a wheelchair. To extend productivity in life, clients need to anticipate the eventuality of the disease and find ways to enrich the quality of life as physical functioning diminishes (Chen, 2006). Although clients may initially find assistive devices to be awkward and embarrassing, rehabilitation counselors nevertheless should elucidate the advantages of using a walker, wheelchair, and scooter.

Decline in daily physical activities as a result of muscle deficiency is a substantial problem in the MD population. People with sedentary lifestyles are prone to developing adverse medical conditions including coronary artery disease, osteoporosis, obesity, anxiety, depression, and musculoskeletal impairments (Matthews, 2003). Healthy bodies need regular exercise to maintain toned and conditioned muscles. Paradoxically, people with MD are at greater risk to damage muscle fibers when undergoing regimens of exercise because the dystrophin protein cannot function properly to repair injured muscles. Hence, it is advised that patients consult neurologists and physiotherapists familiar with MD before beginning an exercise regimen.

Improved pain management serves to enhance quality of life among people with muscular disease. BMD and DMD are known to cause uncomfortable muscle cramps and painful stiffness. Ruptured and swollen connective tissues set off prolonged painful and irritating spasms that may persist during sleep. Lethargy in the daytime attributed to poor quality of sleep seriously affects concentration and productivity at work. Weakening in pulmonary muscles in people with neuromuscular failure requires respiratory support to regulate the inhalation of oxygen and exhalation of carbon dioxide (Yates, Festa, Gills, Waters, & North, 2004). To minimize sleep disruption, suitable ventilators must be chosen to meet the unique needs of individuals with MD to ensure that respiratory mechanics function to maximum capacity (Fanfulla, Delmastro, Berardinelli, Lupo, & Nava, 2005).

### ***Leisure and Recreational Issues***

Exercise for persons with MD has been somewhat controversial due to dystrophin deficiency and decreased muscle contractile properties (Hicks, 1998). Nonetheless, an appropriate amount and intensity of exercise is recommended, depending on the progression of the disease. While there is a benefit in conditioning muscles, precautionary measures need to be taken to avoid excessive muscular tissue damage that will accelerate the deterioration process. Aquatic therapy and hippotherapy (horseback riding) are ideal exercises for persons with neuromuscular diseases. Resistance in water allows the body to harden its flex and reflex movements with minimal risk of injuring muscles. For persons who have weak spinal, thoracic, cervical, and lumbar muscles, hippotherapy helps strengthen and maintain the correct postures of those areas. Furthermore, animals can be great companions for riders who have disabilities and help build self-confidence.

Clients should be encouraged to take trips to visit different places. Due to mobility, toileting, and respiratory difficulties, clients with DMD, BMD, and LGMD may hesitate to travel. Transportation of battery-powered wheelchairs and ventilators may be viewed as a nuisance to someone who is not trained to take care of passengers with special needs traveling by airplane, train, bus, or cruise ship. Careful planning before taking a trip can alleviate many of these concerns.

### ***Sexual and Reproductive Concerns***

For many people with disabilities, rehabilitation counselors are often the first persons they confide in about their anxiety over sexual issues (Vash & Crewe, 2004). Prior to providing counseling on sexual issues, counselors must first examine their own attitudes. If the counselor feels apprehensive about discussing intimate and sensitive questions with clients, appropriate referrals should be made. Rehabilitation professionals can serve as vital information sources to alleviate clients' anxieties and doubts about their

sexuality (Brodwin & Chen, 2000). The onset of LGMD and MMD often occurs at the time when young boys and girls are entering puberty. Severity of disability and its subsequent impact on social interaction deprive young people the chance to experience sexual intimacy. Dating and courtship can be quite stressful and elusive. To project a warm and amicable image, clients need to learn about proper social etiquette and maintaining positive attitudes toward other people.

Inheritable disorders are a legitimate concern when planning a family. Testing of DNA and analysis of cell mutations provide information to predict the likelihood of passing on inherited diseases to offspring (Krajewski & Shy, 2004). The desire to have children can be carefully weighed against the possibility of passing mutant genes to offspring.

The purpose of genetic counseling is to provide a person with MD and his or her spouse with knowledge of the nature of the genetic disorder, its transmission, and the risk of occurrence in potential offspring. Depending on the type of disorder, the probabilities of genetic risks range from 1 in 2 people (with a dominant gene) and 1 in 4 people (with a recessive gene) (Gustavson, 1999). An ethical issue that arises is the acceptance of human diversity and disability. While the responsibility of explaining family planning rests on rehabilitation and medical professionals, clients with MD and their families are responsible for the decision as to whether or not to have children.

## **Conclusion**

MD is a group of neuromuscular diseases that gradually lead to deterioration and wasting away of muscles. Although there is no cure, people with this condition do not have to hold a bleak view of their future. When preparing a rehabilitation program or plan, it is imperative for rehabilitation counselors to acquire both knowledge of the client's capacity to handle tasks and knowledge of what the client is unable to manage. Because of the wide range of differences, each client needs careful, individualized evaluation. Fear and uncertainty about the future may induce clients to believe they are living on "borrowed time" because they have no control over the progressive weakening process (Chen, 2006).

When helping clients implement planning, rehabilitation counselors are wise to consider holistic interventions, i.e., psychological counseling considerations, vocational counseling advice, medical counseling issues, leisure and recreational issues, and sexual and reproductive concerns. Compassion is an imperative aspect in building a good and mutually respectful rapport between client and counselor. However, rehabilitation counselors should neither communicate an overly pessimistic outlook nor raise false hopes. By formulating realistic and practical goals with input from clients with MD, counselors will attain better rehabilitation outcomes.

## **Case Study**

Ming Lin is a 20-year-old immigrant who came to the United States from Hong Kong one year ago. She lives with her parents and one older sister in an apartment adjacent to Chinatown in Los Angeles, California. Before arriving in the United States, Ming finished the 11<sup>th</sup> grade in her home country, learned a basic command of conversational English, and worked for two years as an apprentice tailor. Ming is a referral to the Department of Rehabilitation from a Muscular Dystrophy Association support group. Ms. Lin has been working at a clothing factory as a children's apparel seamstress since she arrived in this country.

At age 19, Ms. Lin was diagnosed with fascioscapulohumeral muscular dystrophy (FSHMD) and was treated at a public hospital in Hong Kong. Despite the presence of FSHMD, Ming has been able to care for herself. Since her arrival in the United States, Ming has been working for the same employer. Job evaluations indicate she is a reliable employee and gets along well with co-workers and supervisors. Four months ago, Ming noticed her productivity steadily decreasing. Dust from cutting and sewing textiles irritated her eyes. She had difficulty sewing with precision and maintaining productivity. Her arms quickly became sore and tired after working on the sewing machine for a short time. To meet the daily quota, Ms. Lin took fewer breaks and a shorter lunch. Ming often could not finish her lunch in the designated time due to swallowing difficulties; she does not have this problem when she eats slowly.

This client feels she is on the verge of a nervous breakdown. The fast pace of garment factory work is difficult for her, both mentally and physically. She does not want to be a burden to her family and desires

to continue working. Ming is experiencing mild depression because of social isolation. Typically, she spends her time watching television and cooking for her family. Occasionally, they have dinner out. Presently, Ming is living with her parents and has doubts about dating due to her physical condition. In addition, her father does not approve of interracial dating. At a co-worker's invitation, Ming attended a local Chinese-speaking church several times. However, her Buddhist parents prefer that Ming not attend this particular church.

Before taking a few weeks of sick leave because of muscular dystrophy symptoms, Ming was considered as the top candidate for promotion to floor supervisor. Additionally, the company is in the process of computerizing the production lines. Ms. Lin has expressed a strong interest in learning the new computer-aided fashion design software to sketch clothing designs, a position available at her employer. However, she is pessimistic about the likelihood of being promoted to a designer position due to her lack of education, limited computer experience, and because her employer now is aware that she has MD. Except for a brief period of sick leave, she has been able to continue working full time. Additional interests of Ms. Lin include child care, counseling, and working with people who have disabilities. Due to a family business failure, Ming had to leave high school at the beginning of the 12<sup>th</sup> grade; she immediately secured employment in the clothing industry and never returned to school.

### Questions

1. Identify some problems persons with MD encounter in society and in seeking and maintaining employment.
2. As Ms. Lin's counselor, what possible physical and emotional limitations would you take into consideration when planning vocational rehabilitation services?
3. If you recommend continued employment with her employer, what advice would you give concerning reasonable accommodation? Would you recommend one of the two positions available, one as a floor supervisor and the other as a computer-aided fashion designer?
4. How would you suggest Ming expand her social activities without creating tension with her parents? Provide your rationale within a cultural context.
5. As a career counselor, provide a career path for this individual. Remember that Los Angeles is a garment-manufacturing center. Take into consideration her particular type of muscular dystrophy.

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## Chapter 25

# MENTAL RETARDATION

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## Introduction

Evidence of mental retardation can be traced as far back as 1552 B.C.E., during the time of the therapeutic papyri of Thebes (Luxor), Egypt (Ainsworth & Baker, 2004). Through the ages, persons with mental retardation have faced challenges relative to the customs and beliefs of their culture. In ancient Greece, children with cognitive or physical disabilities were considered nonfunctional and were subsequently eliminated. During the Middle Ages, society found a function for these children as slaves or for use as entertainment. It was not until John Locke (1690) published *An Essay Concerning Human Understanding* that the public began to realize that persons with mental illness still maintain a capacity to learn. He proposed the idea that humans born with a blank slate (*tabula rasa*) learn through experience. Society, in his work, was shown as a possible cause of developmental retardation, rather than mental illness. This turning point in the conceptual world of mental retardation inspired the development of new treatment approaches and ultimately improved cultural perceptions of mental retardation.

Changes in policies, services, expectations, and personal values over the past several decades have significantly affected the quality of life for persons with mental retardation. This chapter discusses programs, services, and support systems available for these individuals. The framework of the chapter includes the definition, prevalence, etiology, specific learning characteristics, functional limitations, and rehabilitation potential generally observed in people with mental retardation. While living in the community, employment has been demonstrated to have a positive impact on the lives of those with intellectual disabilities (Stephens, Collins, & Dodder, 2003). However, there continues to be challenges associated with employment (Wehmeyer et al., 2003). We encourage rehabilitation professionals to consider the information presented in the hope that they may develop better quality programs, services, and support for their clients.

## Definition of Mental Retardation

Definitions of mental retardation are usually based on the individual's mental and functional limitations. The American Association of Mental Deficiency (AAMD - now called AAMR, American Association of Mental Retardation) has developed the following widely accepted definition:

Mental retardation is a disability characterized by significant limitations both in intellectual functioning and in adaptive behavior as expressed in conceptual, social, and practical adaptive skills. This disability originates before the age of 18 (American Association of Mental Retardation, 2002, p. 1).

Significant limitations in the area of adaptive behavior are identified by sub-average performance in at least two of the following categories: communication, self-care, home living, social skills, community use, self-direction, health and safety, functional academics, leisure, and work. Through time, there has been a movement to change the definition of mental retardation from a person's limitations to the importance of identifying that person's needs. This newer way of thinking places the burden of responsibility on society to meet those needs (Falvey, 1995, 2005). Specifically, Marc Gold (1980), who developed the "Try Another Way" system, defined mental retardation by noting that one should consider the following:

“The level of power needed in the training process required for the individual to learn, and not by limitations in what he or she can learn. The height of a person’s level of functioning is determined by the availability of training technology and the amount of resources society is willing to allocate and not by significant limitations in biological potential” (p. 5).

After Gold’s work was published, the AAMR updated their definition of mental retardation to address the interactions between disability and the environment.

“A complete and accurate understanding of mental retardation involves realizing that mental retardation refers to a particular state of functioning that begins in childhood, has many dimensions, and is affected positively by individualized supports. As a model of functioning, it includes the contexts and environment within which the person functions and interacts and requires a multidimensional and ecological approach that reflects the interaction of the individual with the environment, and the outcomes of that interaction with regards to independence, relationships, societal contributions, participation in school and community, and personal well-being” (AAMR, 2002).

Although mental retardation is always manifested before age 18, people with mental retardation are not a homogeneous group. The degree of developmental delay (mental retardation) ranges from mild to moderate, from severe to profound, though the criteria for these categories continue to be debated; intelligence quotient (IQ) scores remain the primary decisive factor. A person with mild mental retardation has an IQ score between 50-75, while a person with moderate mental retardation has an IQ of 35-49. An individual with severe mental retardation has an IQ of 20-34; a person with profound mental retardation has an IQ of below 20 and often has other accompanying physical or sensory disabilities (see Table 1).

The degree of developmental delay manifested is related to the assistance, support, and instructional opportunities available to people who have mental retardation. These individuals possess interests and strengths that are as varied as persons without disabilities, and display equally diverse personalities and characteristics.

## **Prevalence of Mental Retardation**

Approximately 3% of the school-age population and 1-2% of pre-school and post-school age populations are identified as having mental retardation (Brimer, 1990; AAMR, 2002). The variations of these percentages are directly related to the labeling and classifying procedures used in school programs. When mental retardation is classified within the mild to moderate range, pre-school children and adults may not be labeled or in any way classified as inferior or different than the general population. Traditionally, schools have grouped and classified school-age students into distinct ability levels, resulting in the labeling of students with mental retardation. This classification explains the higher percentage identified within the school-age population.

In the past, race and economic status have been used to dictate prevalence. Major litigation and legislation, however, has ruled that the identification of mental retardation among minority groups based on racial factors is unjust and incorrect (Larry P. v. Riles and Diana v. State Board of Education, Public Law 94-142, 1975). A disproportionate number of school-age students identified as mentally retarded from certain minority groups remains, however, especially among African Americans and Hispanics or Latinos (U. S. Census Bureau, 2000). Though the primary method of identification has been the use of IQ tests, professionals have criticized these examinations as being culturally and linguistically biased in favor of members of the non-Hispanic White, American, middle-class. Thus, researchers consider the results of IQ tests invalid when given to children who are not members of the White middle class community (Bishop & Falvey, 1989).

## **Etiology of Mental Retardation**

Although up to 50% of cases of mental retardation have no known or recognizable etiology (Daily et al., 2000), there are many known causes of the condition. When physicians rule out specific syndromes and causes, they often cite other prenatal (before), perinatal (during), and postnatal (after) pregnancy conditions

**Table 1**  
**Developmental Characteristics Related to Level of Mental Retardation (DSM-IV Criteria)**

<b>Mild retardation</b>	<b>Moderate retardation</b>	<b>Severe retardation</b>	<b>Profound retardation</b>
85% of all cases of retardation	10% of all cases of retardation	3% to 4% of all cases of retardation	1% to 2% of all cases of retardation
Function at one half to two thirds of CA (IQ: 50 to 75)	Function at one third to one half of CA (IQ: 35 to 49)	Function at one fifth to one third of CA (IQ: 20 to 34)	Function at < one fifth of CA (IQ: < 20)
Slight delays in all areas	Noticeable delays, especially in speech	Marked and obvious delays; may walk late	Marked delays in all areas
May have no unusual physical signs	May have some unusual physical signs	Little or no communication skills, but may have some understanding of speech and show some response	Congenital abnormalities often present, hindering speech and physical growth
Can acquire practical skills	Can develop communication skills, especially with augmentative assistive devices	May be taught daily routines and repetitive but meaningful activities	Need close supervision
Useful reading and math skills up to grades 3 to 6 level; can complete most adaptive skills independently	Can learn elementary health and safety habits; can participate in simple activities and self-care	May be taught simple self-care skills	Often need attendant care
Can conform socially and develop peer relationships with support	With assistance, may learn how to respond to common social cues and basic social rules	Need direction and supervision throughout the majority of the day	May respond to regular physical activity and social stimulation
Can acquire vocational skills for self-maintenance	Can perform tasks in supported conditions	Can perform tasks in supported conditions	Not capable of self-care
Integrated into general society	Can travel alone to familiar places	Need supervision and accommodations during travel to and from places	Need supervision and accommodations during travel to and from places

Note: Additional problems with vision, hearing, or speech, congenital abnormalities, seizures, emotional problems, or cerebral palsy may be present.

DSM IV = Diagnostic and Statistical Manual of Mental Disorders (4th ed.) (American Psychiatric Association, 2000); CA = chronological age; IQ = intelligence quotient.

Adapted from D. K. Daily, H. H. Ardinger, & G. E. Holmes (2000). Identification and evaluation of mental retardation. *American Family Physician*, 61(4).



which may result in mental retardation. Mental retardation can occur before birth (prenatal), during birth (perinatal), or after the birth (postnatal). These causes are described in the following paragraphs.

### ***Prenatal Causes***

Down's Syndrome is the most frequently observed genetic cause. Down's Syndrome, identified in 1866 by Doctor Langdon Down, is commonly known as Trisomy XXI. Persons with Trisomy 21 have an extra 21<sup>st</sup> chromosome. Factors influencing the prevalence of Down's Syndrome include maternal age. The older the mother, particularly over age 35, the higher the possibility the baby may have Down's Syndrome. Other developmental problems, such as respiratory and heart abnormalities often accompany Down's Syndrome, and the degree of mental retardation observed in persons with Down's Syndrome ranges from mild to significant.

In 1934, Dr. Asbjourn Folling, a Norwegian physician, discovered phenylketonuria (PKU) as a cause of mental retardation. PKU is a sign of deficiency in the production of phenylalanine hydroxylase, an enzyme necessary in the metabolism of phenylalanine, an essential amino acid. Brain damage occurs with the buildup of phenylalanine in the blood. In 1959, Dr. Robert Guthrie developed a screening test for newborns that physicians routinely use today to detect PKU. When a physician identifies PKU in a newborn, a phenylalanine-restricted diet is started. The degree of mental retardation is significantly less severe for persons with treated versus untreated PKU; dietary restrictions during the primary years of life are essential to lessening the severity of the condition.

Other biological conditions, such as disease, can result in mental retardation. Tay-Sachs is one such illness. This disease, a lipid metabolic disorder, is most often found in persons of Jewish descent. Besides mental retardation, it causes a progressive deterioration of nerve tissue and generally results in death at an early age.

There are several physical factors other than genetic ones that influence the presence and severity of mental retardation. Beginning with the prenatal stages of development, the mother's general eating habits and physical health are crucial. Chronic maternal illnesses, such as diabetes, may impair fetal development. Maternal use of alcohol, drugs, tobacco, and exposure to environmental pollution during pregnancy along with maternal infections and viruses such as rubella, perinatal cytomegalovirus infection (CMV), meningoencephalitis, syphilis, and toxoplasmosis increase the probability of mental retardation and other disorders. Fetal exposure to radiation during development can also cause genetic mutations that eventually result in mental retardation.

### ***Perinatal Causes***

During childbirth, several factors can increase the possibility of mental retardation (Ainsworth & Baker, 2004). Cephalopelvic disproportion, for example, a condition in which the size of the birth canal is too narrow for the presenting infant's head, can result in brain damage to the newborn. Other factors include extreme prematurity, accidental physical trauma, asphyxia, hypoglycemia, infection, blood cell or blood type diseases, and Rh factor blood incompatibilities between mother and baby.

### ***Postnatal Causes***

Following birth, several other factors can increase the probability of mental retardation. These include malnutrition, acquired traumatic brain injury, meningitis, encephalitis, other infections, and chemical substances (e.g., pesticides, drug abuse, and metal [lead] poisoning). Additional causes are demyelinating and degenerative disorders, consequences of seizure disorders, toxic-metabolic disorders, and environmental deprivation. Hyperthyroidism, whooping cough, chickenpox, measles, and Hip disease (a bacterial infection) may cause mental retardation if they are not treated adequately. Brain injury can be caused by a blow or a violent shake to the head and result in mental retardation in children.

## **Normalization**

During the nineteenth and early part of the twentieth centuries, institutions for persons with mental retardation were created. In the last four decades, organized coalitions of parents and the "normalization"

principle played a significant role in developing community-based services as an alternative to institutionalization.

In the 1950s, parents of children with mental retardation formed an organization called the National Association for Retarded Children (renamed the National Association for Retarded Citizens). These parents rejected institutionalization for their sons and daughters. They organized alternative programs and services for children with mental retardation. Until the 1970s, many public schools, along with state and local rehabilitation agencies, excluded individuals with mental retardation. The parents had to raise money independently for their children to receive community services.

### ***Services and Support – The Principle of Normalization***

In 1959, the director of the Danish Mental Retardation Service, Bank-Mikkelsen, helped develop Danish law reflecting the principle of “normalization.” This principle states that persons with mental retardation must be able to “. . . obtain an existence as close to normal as possible” (Wolfensberger, 1980, p. 7). The first systematic statement of normalization in world literature was published by Nirje (1969). In 1972, Wolfensberger applied this normalization principle to American society. As people with mental retardation move into the community and obtain employment and housing, a reconceptualization of service delivery models has evolved. Instead of social service agency personnel being responsible for supporting a person with mental retardation, the individual takes charge, utilizing support from family, friends, neighbors, and co-workers.

### ***Deinstitutionalization***

The normalization principle has been a strong driving force in establishing the deinstitutionalization movement. This movement involves integrating institutionalized persons with mental retardation into the community. Biklen and Knoll (1987) identified the major problems with institutions and other large congregate facilities, as well as the primary reasons for deinstitutionalizing people with mental retardation:

- Institutions are the most expensive way of providing residential or any other services to people with mental retardation.
- Institutions do not offer the warm, homelike, individualized environments necessary for full human development.
- Institutions do not provide opportunities for interaction with members of the community.
- Institutions do not allow for positive community living experiences or for development of skills needed for community life.
- Institutions perpetuate and enforce the image that people with mental retardation are oddities.
- People residing in institutions do not have opportunities to learn adaptive behavior from functioning non-institutionalized people.
- Institutionalized people model their behavior after other people living in the institution; these behaviors are often maladaptive.
- Institutions provide a minimum of social and recreational activities and interaction; when they do, it is often demeaning and age inappropriate.

Along with the above drawbacks, institutional living does not offer opportunities for vocational development. Typical jobs are not performed within an institutional environment, and opportunities to learn through observation are absent. Most, if not all, people living in institutions are unemployed, as few have ever had the opportunity to become employed. Discussion of work is nonexistent; normal vocational development does not occur for institutionalized individuals.

### ***Homelike Settings***

As an alternative, state and local communities and parent organizations developed group homes, board and care homes, and foster homes to create homelike settings for persons with mental retardation. These settings are less restrictive than institutional residences. During the later part of the 1980s, and with

increasing success since the 1990s, people with mental retardation began living in their own homes, with supports from social service agencies (Falvey, 2005).

### ***Self-Advocacy***

A movement toward self-advocacy or “people with disabilities asserting their sworn rights and interests, usually without others’ help” developed as a result of the normalization principle (Turnbull, Turnbull, Bronicki, Summers, & Roeder-Gordon, 1989, p. 314). Two self-advocacy international networks, *People First* and *United Together*, are becoming increasingly influential in the areas of employment advocacy, fair working conditions, and financial compensation.

In recent years, self-advocacy also has been referred to as self-determination. Self-determination is a vital part of the education and vocational rehabilitation of persons with cognitive disabilities. Kilsby and Beyer (2002) found that enhancing self-determination in job matching increased the success of persons with cognitive disabilities. Participants in the study increased their accuracy of recalling daily experiences and demonstrated a decrease in the need for job coach assistance.

### ***Legislation and Normalization***

*Rehabilitation Act of 1973.* Professionals often refer to Section 504 of the 1973 Rehabilitation Act as the “civil rights” bill for individuals with disabilities. This legislation requires public and private employers, educators, and service providers to use nondiscriminatory and affirmative action practices.

*Education of All Handicapped Children Act of 1975.* This act and its amendments ensure a free and appropriate education in the least restrictive environment for people between the ages of 3-21. The least restrictive environment mandates education of students in the presence of their peers without disabilities and within the general education program to the “maximum extent appropriate.” Research has consistently supported maximum integration of all students in “regular” education programs (Falvey, 2005; Lipsky & Gardner, 1997).

*The 1987 Developmental Disabilities Act.* The stated goals for the 1987 Developmental Disabilities Act include:

[to] assure that persons with developmental disabilities receive the care, treatment, and other services necessary to enable them to achieve their maximum potential through increased independence, productivity, and integration into the community,

and

[to] establish and operate a system that coordinates, monitors, plans, and evaluates services which ensure the protection of the legal and human rights of persons with developmental disabilities.

*Social Security Act Amendments.* In 1987, Congress amended the Social Security Act to provide a work incentive program within the Supplemental Security Income (SSI) program. Before these amendments, regulations encouraged dependency. Medicare coverage (medical insurance) and financial support were removed when individuals acquired jobs. The new program allows the SSI recipient to continue to receive Medicare benefits if an employer does not provide equivalent comprehensive medical coverage. Most persons with mental retardation qualify for SSI benefits and have been more likely to obtain employment since these changes in the Social Security Act.

*Americans with Disabilities Act.* In 1990, Congress passed the Americans with Disabilities Act (ADA). This act has broad implications concerning employment, transportation, public accommodations, state and local government, and telecommunications. The most significant area for persons with mental retardation is employment. Employers may not discriminate against individuals with disabilities in hiring or promotion; they need to provide reasonable accommodation to the known physical or mental limitations of employees with disabilities, including job modification and job restructuring. All large employers with 15 or more employees need to comply. Enactment of the ADA has helped rehabilitation counselors facilitate obtaining competitive employment for their clients.

## Functional Limitations

The most salient learning characteristics of persons with mental retardation are slower rates of learning than their typical peers, and difficulty generalizing or transferring information learned from one situation to another. Though the rate of learning is slower in persons with mental retardation, these individuals maintain the ability to learn and acquire new skills. However, they must be taught specific skills in environments in which they will perform these skills. For example, teaching public transportation skills needs to include using the public transportation system in the person's community; one needs to teach job-related skills in "real" work environments (Salkever, 2000).

Other functional limitations include behavioral challenges, lack of self-management skills, and inappropriate social skills. Behavioral challenges can be expressed through short attention span, impulsivity, and difficulty in making judgments. Often, persons with developmental disabilities need to be directly taught self-management skills. Although persons with developmental disabilities may be sociable, they are often unaware of social pragmatics or other rules governing social behavior (Bielecki & Swender, 2004). In the workplace, persons with mental retardation will inevitably build interpersonal relationships with their co-workers. With limited ability to process pragmatic cues in social situations, employees with mental retardation may inadvertently ostracize themselves.

Along with these challenges, persons with mental retardation face another hurdle – communication. Communication impairment is often associated with mental retardation. When designing a vocational rehabilitation program, an individual's reading ability, slower rates of information processing (especially instructions given only verbally), and slower rates of information output (vocal expression or performance) must be considered. Some individuals have limited vocal expressive skills (e.g., speech impediment, sing-song rhythm of speech, and difficulty regulating volume). For the rehabilitation professional, it is important to consider using visual tools (e.g., visual schedules and instructions) and/or augmentative adaptive communication tools.

At times, persons with mental retardation have been characterized as no longer capable of learning or as having reached a "plateau" (Falvey, 1989). When a person's learning has reached such a plateau, it may simply be an indication that different services and support are necessary (Libal, 2004). The learning possibilities and potential of persons with mental retardation are directly dependent upon the commitment of services and support society is willing to provide, as opposed to limitations inherent in the disability itself.

### *Learned Helplessness*

A phenomenon repeatedly observed in institutions and segregated schools, workshops, and activity centers is "learned helplessness." This phenomenon involves a pattern of submissiveness developing when individuals repeatedly discover that their actions are of no consequence and that outcomes are beyond their control (Payne & Patton, 1981). Individuals learn to not demonstrate skill proficiency, independence, or an awareness of their surroundings. This phenomenon has a similar effect to that of the self-fulfilling prophecy which states that most people's opinions and expectations about themselves are influenced in significant ways by those around them. If teachers, social workers, rehabilitation counselors, and job coaches believe an individual student or client is unable to learn, that person will be discouraged from learning. If the same group of supportive professionals expect the individual to learn and be successful, the chances for a successful outcome are greatly enhanced (Falvey, 2005).

### *Environmental Factors*

The environment plays a significant role in influencing the success and employability of individuals with mental retardation. The traditional model provided by adult service agencies has been vocational training in a "train and place" model, such as that which occurs in a sheltered workshop. In such a situation, a client is trained in a simulated job setting and then employed. Because clients with mental retardation do not generalize well across different settings, this model generally does not result in successful employment in integrated community jobs. The "place and train" model, a reverse method of employment, has resulted in a significantly greater number of employed clients within integrated community job settings (Moore, Flowers, & Taylor, 2000). These two models are discussed in greater detail later in this chapter.

## ***Societal Discrimination***

Over the years, society has discriminated against people with mental retardation, primarily due to ignorance, stereotyping, and incorrect information. Until recently, most employers had no prior interaction with persons with mental retardation (O'Brien, Revell, & West, 2003). Much work is currently being done to change the attitudes and expectations of employers by allowing them to observe other employers who have hired individuals with mental retardation. This observation often results in raised expectations of what a person is capable of performing and more positive attitudes toward these individuals. Although ignorance and discrimination in the workforce still exists, these attitudes are slowly changing.

## **Rehabilitation Potential**

### ***The Supported Employment Model***

Research over the past several decades has influenced changes in the delivery of services. In 1972, Gold demonstrated that persons with severe mental retardation who were deaf and blind could assemble complex electronic circuit boards. Hunter and Bellamy (1977) taught persons with profound mental retardation to assemble harnesses. Such discoveries led to the realization that people with mental retardation could successfully learn work skills for competitive, gainful employment. This form of employment, in which an employee receives support or assistance, is called supported employment. Rehabilitation counselors throughout the country are currently using the principles of supported employment. The concept and implementation of supported employment have been expanded through research and legislation (Wehman, 2001, 2006).

The traditional service options available to most adults with mental retardation before 1980 were developmental centers, work activity centers, and sheltered workshops. Professionals have criticized these settings because they generally do not provide "real" work (work in actual work environments). The purpose of sheltered work environments is to prepare people for employment in the real world. However, after a national survey was conducted by Bellamy in 1985 (Bishop & Falvey, 1989), it was concluded that too much time was spent training people for competitive employment. According to his research, people with mental retardation spent an average of 37 years in adult developmental centers, 10 years in work activity centers, and 9 years in sheltered workshops. Since most persons with mental retardation "graduate" from school at approximately age 21, a person would be, on the average, 77 years old before entering the work force. According to Bellamy, the supported employment model has been extremely successful in placing persons with severe disabilities in gainful work environments. There are several anti-habilitative characteristics of sheltered workshops that do not exist in supported employment (Falvey, 2005):

- Work performed often requires minimal learning on the part of the workers.
- Sheltered workshops often retain the best workers for more demanding work, instead of training them for non-sheltered vocational environments.
- Adequate time is not available for training workers in non-sheltered environments.
- Personnel often lack training in critical areas, such as effective teaching methods, production, and providing real job training.
- "Down time" or "dead time" is frequent.
- Practitioners often base formal vocational evaluations on inferences, not on actual observations of clients in real community jobs.
- All the students/clients receiving training have disabilities and, therefore, there is no contact with workers without disabilities.
- Inappropriate assumptions are made that persons in sheltered workshops and activity centers "enjoy" or are more competent at tasks that involve sitting or standing at tables and using predominantly fine motor and coordination skills.
- Tasks vary according to the contracts secured, not according to the client's training needs.



- Equipment is often outdated and in poor condition, resulting in worthless training.
- Frequently, the workshop does not follow normal workdays or workweeks.
- Consequently, persons in workshops or activity centers do not have sufficient opportunities to build endurance. Historically, workshops and work activity centers have interrupted work periods, particularly at holiday times, with events such as dances that feature age-inappropriate music.
- Workshop personnel do not develop systematic strategies to assist clients in making the transition to less restrictive work settings.
- Irrelevant exit requirements or prerequisite skill requirements are often greater than requirements in real community jobs.

### ***Characteristics of Supported Employment***

Supported employment is an alternative to sheltered workshop training. The characteristics of supported employment services are as follows:

#### ***Place and Train***

Refers to developing services that reverse the traditional method of “train and place.” “Place and train” involves placing a person on the job, regardless of job readiness, and immediately training that person to perform the job. To ensure success, a job coach is available to support the person in the job (Moore, Feist-Price, & Alston, 2002).

#### ***Integrated Job Settings***

Job settings used for training are the same as used for employees without disabilities (Wehman, 2006). In addition, these integrated job settings should not create a disproportionate number of employees with disabilities. Integrated job settings should reflect the natural proportion of persons who do not have disabilities within the community. The rehabilitation counselor needs to make every effort to encourage healthy interactions and relationships among the trainee with mental retardation and co-workers (Turnball, Turnball, Shank, & Leal, 1999).

#### ***Self-Determination and Individual Placements***

Successful supported employment models foster self-determination by empowering employees with disabilities to make their own choices and to take control of their career paths (Wehman, Revell, & Brooke, 2003). Job placements should be based on accessibility along with, most importantly, the trainees’ preferences and strengths.

#### ***Meaningful Work***

This characteristic implies that rehabilitation counselors need to avoid providing makeshift work or otherwise made-up jobs that persons who do not have disabilities would not perform. Wehman (2006) explained that indicators of meaningful employment occurs when an employee with a disability is hired, supervised, and paid directly by the business where the job setting is located and receives wages/benefits commensurate with coworkers without disabilities.

#### ***Job Development and Marketing***

This service involves systematically accessing existing jobs, modifying existing jobs, and creating new ones. Job development and marketing are essential to the delivery of supported employment and should be provided by individuals who have knowledge of the local job market and have credibility with employers. Interactions with potential employers must be conducted in a business-like manner (Falvey, 2005; Moore et al., 2000).

#### ***Job Coaching***

Provides instruction on the job and teaching job-related skills, including social skills. In addition, job coaches provide job-related skills instruction in the client’s home and community setting. These skills include preparing for work, using public transportation, depositing and budgeting paychecks, and developing social networks and friendships within the community. The job coach provides salient yet subtle

instruction to allow the client to successfully develop skills while not embarrassing or discouraging the person. If the client is not performing correctly, the job coach provides additional instruction. If the individual fails to arrive for work, the job coach will act as a substitute worker and perform the necessary tasks of the absentee. Also, the job coach facilitates natural supports from co-workers and supervisors within the work environment.

Providing the support needed to access and maintain employment, housing, and other community services and activities enhances rehabilitation potential (Salkever, 2000). Self-determination and autonomy are positively influenced in a supported employment model while a segregated vocational day program is generally less successful. Supported employment services across the United States and within other countries (e.g., Canada and Italy) have successfully employed millions of people with mental retardation. Nationwide, however, individuals with mild mental retardation are more likely to be placed in competitive supported employment than individuals with more severe levels (Moore, Harley, & Gamble, 2004). Overall, many persons have been provided with the support needed to live where they wish and with whom they want while accessing their neighborhoods and communities, while simultaneously receiving sufficient support to ensure success.

Attitudes of people in the community have changed over the years, leading to greater acceptance of their neighbors with mental retardation (Hampton & Xiao, 2007). Technology also has influenced successful integration within the community. Simple adaptations such as calculators and other technological devices, including voice synthesizers and other electronic communication aides, have greatly enhanced the participation of persons with mental retardation.

## **Conclusion**

This chapter provides the rehabilitation counselor with an overview of the issues related to mental retardation. The most critical concept is that the degree of participation and potential of persons with mental retardation is dependent upon society's commitment and willingness to support people who have mental retardation in work, housing, and within the community. The supported employment model has proven to be the most effective service delivery system.

## **Case Study**

James is a 23 year-old African-American who has spent the last 10 years of his life in an institution. While in the institution, his day consisted of being lifted from bed, having his clothes changed, being fed pureed foods, and sitting in front of a television set for hours. Five days a week, he attended a classroom on the institutional grounds that was referred to as "school," where he listened to pre-school level music and played games like placing pegs into a board. There was little interaction with others, unless he was receiving directions or instructions. James never left the institution and had no friends as a result of his limited interaction with others. One day, a social worker came to his room and asked if he wanted to share an apartment with two other men in the neighboring town about 10 miles away. Although James had very little reaction to anything that went on around him, he smiled and made direct eye contact with the social worker. A month later, James moved into the apartment and began to learn about living and working in the community.

This individual has multiple disabilities, including severe mental retardation, cerebral palsy, and significant vision loss. He uses a wheelchair with the support of an attendant for transportation. The attendant assists him with basic self-care skills. After James lived within the community for awhile, the social worker referred him to a supported employment agency in the area. A job developer and job coach visited him in his apartment to begin preparation for possible employment. James used a new picture communication aid which was provided through rehabilitation services to interact with the job developer and job coach.

The job coach and job developer spent an hour in the apartment observing James participate in various tasks with the support of his attendant. They went into the neighborhood with James to observe his reactions to various stimuli and determine potential job possibilities within a reasonable distance from his apartment. Two blocks away, a hardware store supervisor indicated they needed someone to sort the

incoming inventory into bins and place the bins on shelves. The job developer suggested to the supervisor that he consider employing two people with developmental disabilities who have complementing strengths to do the job. Together, with the support of a job coach, they could be very successful on the job. The supervisor was reluctant because he lacked experience hiring and working with people with developmental disabilities. The job developer gave him the names and telephone numbers of other employers who had hired employees with developmental disabilities and encouraged him to contact them.

A week later, the hardware store supervisor contacted the job developer and stated that he would like to hire James and Rick (the other person identified in the original conversation). The supported employment agency provided a job coach to support both men.

A year later, both James and Rick were still working at the hardware store; the supervisor was very enthusiastic about their employment. With the support of a job coach, James and Rick share the job tasks of sorting and organizing the inventory of the hardware store. Additionally, they have become friends and have developed good relationships with their co-workers, both at work and away from the job. The quality of life for James has been greatly enhanced. He shows greater interest in life and his surroundings; James smiles and laughs often.

### Questions

1. What are the possible causes of James's mental retardation?
2. Identify the major problems with institutional living and the primary reasons for deinstitutionalizing people such as James.
3. Describe the advantages of supported employment over sheltered workshop employment.
4. Discuss the roles and functions of the job coach in supported employment. If James and Rick were not performing the work correctly or did not show up for work, how would the job coach proceed?
5. Outline the six characteristics of the supported employment services model.

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## Chapter 26

# LEARNING DISABILITIES

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## Introduction

This chapter focuses on the various issues related to the successful habilitation and rehabilitation of persons with specific learning disabilities (LD). Unlike many other disabilities, LD is considered an invisible disability in that it is not initially evident. There is a common misconception that LD is only a “mild” disability and therefore has little impact on quality of life (Sabornie & deBettencourt, 2004). “Issues of stigma and a lack of awareness and understanding of LD by the general public have contributed to the confusion about their existence and impact” (Cramer & Ellis, 1996, p. xxvii).

Upon completion of this chapter, the reader will have (a) an understanding of the various dynamics and life implications that people with LD face, and (b) the ability to design effective vocational rehabilitation plans for persons with learning disabilities. Topics covered include: prevalence, definition of LD, diagnosis and evaluation, characteristics of LD, reasonable accommodation, rehabilitation potential, and a case study.

## Prevalence

LD affects approximately 15% of the United States population; additionally, many people who have learning disabilities are never diagnosed. In 2004, the federal government estimated that there were 2.8 million students with learning disabilities in school. Getzel and Gugerty (2001) noted that it is estimated that 35% of youth with learning disabilities drop out of high school. Of these students who complete high school, 62% are unemployed a year following graduation. Approximately 56% of students with LD who drop out of high school eventually are arrested.

Learning disabilities are not a homogeneous set of disabilities (Heward, 2006). Generally speaking, the term learning disabilities refers to a variety of disorders in the domains of listening, speaking, reading and writing, mathematics, and reasoning. These disabilities interfere with an individual’s ability to store, process, and produce information and are often hidden by the individual’s general level of functioning. Learning disabilities may occur with or be complicated by problems involving inadequate attention and inappropriate social skills (Cramer & Ellis, 1996).

As of yet, no single current database exists to determine the prevalence of learning disabilities in the adult population. Although the actual number of adults with LD in this country has not been determined, the 2004 national estimate of about 2.8 million students in schools, suggests the number of adults with LD is likewise substantial (Rubin & Roessler, 2008). Estimated prevalence discussed below has been derived from various national data sources.

The United States Employment and Training Administration (1991) reported that between 15% and 23% of Job Training Partnership Act (JTPA) trainees have learning disabilities, while 50% to 80% of adults reading below the 7<sup>th</sup> grade level have learning disabilities. Additionally, between 10% and 50% of Adult Basic Education students likely have learning disabilities (Adult Education Amendments, 1988).

Regarding gender-related prevalence, it is a common observation that the school-identified LD ratio of males to females is 4:1. In research and self-identified samples, gender ratio was found to be almost equivalent (1:1), very similar to the self-reported numbers described in the National Adult Literacy Survey (NALS) results (Kirsch, Jungeblut, Jenkins, & Kolstad, 1993; Lyon, 1994). Several researchers (Heward,

2006; Sabornie & deBettencourt, 2004) have suggested that the discrepancy between school-identified and self-reported LD gender ratios is related to referral bias in school samples and higher prevalence of attention deficit hyperactivity disorders (ADHD) in males, thus increasing the likelihood of identification of males in school-age special education programs.

## **Definitions of Learning Disabilities**

The earliest and most widely used definition of LD was integrated into federal legislation as the Education for All Handicapped Children Act of 1975. Each subsequent version was incorporated into the Individuals with Disabilities Education Act (IDEA) of 1990 and the Individuals with Disabilities Education Improvement Act of 2004. The earlier definitions included only children. The 2004 definition includes adults with LD, and is written as follows:

The term “specific learning disability” (SLD) means a disorder in one or more of the basic psychological processes involved in understanding or using language, spoken or written, which disorder may manifest itself in an imperfect ability to listen, think, speak, read, write, spell, or do mathematical calculations. SLD includes such conditions as perceptual disabilities, brain injury, minimal brain dysfunction, dyslexia, and developmental aphasia. It does not include a learning problem that is primarily the result of visual, hearing, or motor disabilities; mental retardation; emotional disturbance; and environmental, cultural, or economic disadvantage (p. 182).

Adults with LD have legal protection from discrimination under federal statutes, including the Rehabilitation Act of 1973, specifically Section 504; the Americans with Disabilities Act (ADA) of 1990; IDEA (1990); and the Individuals with Disabilities Education Improvement Act of 2004. These statutes acknowledge the lifelong nature of specific learning disabilities. While Section 504 has its greatest impact on educational environments, the ADA concerns employment. These acts protect persons with disabilities from discrimination, provide for equal access to education and employment, and recommend the provision of reasonable accommodation (Heward, 2006; Rubin & Roessler, 2008).

## **Diagnosis and Evaluation**

### ***Diagnostic Criteria***

Given the complexity and heterogeneity of learning disabilities, the creation of a comprehensive diagnostic study to ensure data-based vocational and service decision-making for individuals with LD is essential (Heward, 2006). Despite the fact that this is the fastest growing population of people with disabilities in state-federal vocational rehabilitation programs, adults with LD remain largely underserved (Dowdy & Smith, 1994).

The diagnosis of LD for vocational rehabilitation purposes should directly relate to the criteria established in the Rehabilitation Services Administration (RSA) definition. Major definitional criteria include evidence of a central nervous system processing difficulty and manifest deficits in one or more of the following areas: attention, reasoning, processing, memory, communication, reading, spelling, writing, calculations, coordination, social competence, and emotional maturity. Psychological evaluation must be preceded by a thorough medical examination; the physician needs familiarity with specific learning disabilities to rule out any systemic organic abnormality that may be causal to the symptoms of a learning disability.

Hawks et al. (1990) developed a research-based diagnostic test battery for adults with LD in vocational rehabilitation that provides an exemplary model for vocational rehabilitation purposes. Suggested deficit areas and sub areas of the diagnostic battery include:

- Cognitive processing (perception, memory, attention, and reasoning).
- Academics (reading, mathematics, and written expression).
- Speech and language (receptive language and expressive language).
- Psychological adjustment (social skills, self-control/impulsivity, self-direction/initiative, and self-esteem).

- Balance and coordination (motor skills).
- Vocational skills (job-seeking skills, vocational interests, coordination, fine motor dexterity, and applied visual motor skills).
- Work methods.

A diagnostic test battery should include neuropsychiatric, psychological, and educational data conducted by a multidisciplinary team consisting of a physician, psychologist, and vocational evaluator. The vocational evaluator assesses interests, aptitudes, abilities, and work behaviors through inventories, observation, and work samples. Other professionals, including neuropsychologists, speech and language therapists, audiologists, occupational therapists, and medical specialists may serve on the team, depending on the nature of the person's learning disability.

### ***Assessment Practices***

Overall, the diagnosis needs to focus on identification of any discrepancy existing between a person's potential achievement and actual achievement. Having an invisible disability, a person with LD may be labeled as bright but bored by his or her parents, "lazy" or "dumb" by teachers, "strange" by peers, and incapable of work by employers (Fletcher, 2007).

Hawks et al. (1990) suggested that to appropriately assess areas of potential deficit or need, four essential sources of information need to be used, including standardized tests, a record review, behavioral observations, and in-depth clinical interviews. The record review, including the person's school and vocational history, previous assessments and results, and the transition plan from high school (if available) must be comprehensive to yield a complete case history specifying previously identified deficits, past interventions, and treatments. Behavioral observations should focus on the following considerations: the person's appearance, punctuality, social skills, activity level, attention span, impulse control, reaction to praise, memory, emotionality, language, and persistence. Clinical interviews include gathering information regarding an individual's self-perception of educational, social, vocational, and cognitive processing abilities and limitations, as well as information on family and personal adjustment issues.

## **Characteristics of Learning Disabilities**

As stated previously, the population of adults with learning disabilities is heterogeneous (Fletcher, 2007). The nature of this disability and its impact on a person's life is influenced by various personal characteristics and external contextual variables including: (a) the unique individual strengths and needs of the individual; (b) cognitive capacity; (c) the precise and unique nature of the specific learning disability; (d) educational history; (e) current contextual cognitive and psychosocial demands; and (f) the historical pattern of successes, frustrations, and failures. The following section discusses the medical aspects of disability, learning characteristics, social skills/adaptive behavior, and job-relevant characteristics.

### ***Medical Aspects***

A basic understanding of how the central nervous system (CNS) operates is crucial to understanding the underlying physiological dynamics of LD. The CNS is the information processing mechanism of the body. Sensory input is obtained through visual, auditory, and tactile stimuli, thus allowing a person to learn through seeing, listening, touching, and moving the body (Carter, Lane, Pierson, & Glaeser, 2006). Individual learning style includes one or more of the person's preferred input modalities. New information is sent to the brain for processing (i.e., analysis, integration, and storage). To assimilate the information, a person must pay close attention to relevant details, classifying new information efficiently for later retrieval. The brain creates *schemata*, or cognitive organizational files, by which to classify and store information. Persons with LD have difficulty recognizing crucial details and associating new information efficiently with what has previously been learned. This results in an inefficient memory system, making later retrieval of information difficult. Persons with learning disabilities appear unorganized, especially in terms of their time management skills (Levy, 2005). Because of the apparent disorganization, a person is often misperceived as lazy, unmotivated, or forgetful.

A learning disability is not synonymous with mental retardation, as often believed. Learning disabilities occur in individuals irrespective of intelligence level. The majority of persons with LD have difficulties related to reading. Between 15% and 20% of school age individuals are affected by specific reading disabilities (Lyon, 1994), ranging from mild to severe. Adults who read below a fourth grade level are considered to have a significant reading learning disability (SRLD).

Individuals with reading disabilities have a language-based, auditory processing difficulty that persists even after learning to read and comprehend with accuracy. For such people, rapid and accurate decoding of unfamiliar, multisyllabic, and foreign language words remains inaccurate and the rate of decoding is slow. Although people with LD read slowly, with appropriate intervention some become proficient at reading comprehension, especially in their fields of expertise (Fletcher, 2007; Sabornie & deBettencourt, 2004).

Vogel (1998) found that adults with SRLD are employed more frequently in part-time jobs, earn significantly lower wages, and have lower occupational status. These statistics have major implications for vocational rehabilitation. First, young adults with learning disabilities would benefit from staying in school and achieving a diploma. Second, upon graduation, students with LD need to complete appropriate postsecondary education or vocational training. To facilitate this process, education and rehabilitation professionals need to: (a) create more meaningful secondary school programs for youth with LD; (b) improve the implementation and procedures involved when transitioning youth from high school to postsecondary training and employment; and (c) increase the number of adults with LD who enroll in and complete a variety of adult secondary education programs.

### ***Learning Characteristics***

Most adults with LD experience reading and other language-related disabilities as major barriers to learning and life functioning. Vogel (1985) conducted a major review of the literature to synthesize the reported manifestations of LD in adults attending college. A brief summary of the most significant limitations follows.

#### ***Language***

Adults with LD often have difficulty perceiving sounds, recognizing syllables within words, and understanding rapid conversation. Frequently, they cannot find the “right” word or mispronounce words with several syllables. Adults often experience various reading disorders, including inaccurate decoding of words and various kinds of comprehension difficulties. In written language, they may have great difficulty with legible penmanship, grammar, spelling, and organizing and developing ideas on paper.

#### ***Mathematics***

People with LD may exhibit limitations related to mathematics. They have difficulty mastering basic operations of mathematics, remembering the sequence of steps in algorithms, and solving multistep mathematical problems.

#### ***Study skills and Attention***

Adults with LD frequently have trouble organizing their time, note taking, focusing on critical components of any task (i.e., becoming distracted readily), and completing tasks.

These various limitations noted above impact effectiveness of daily functioning. Appropriate accommodations help alleviate these obstacles and promote success.

### ***Social Skills Issues and Adaptive Behavior***

Friendlessness, social ineptitude, and loneliness are social development patterns which can result from LD. There is a significant impact on psychological well being, social functioning, emotional adjustment, achievement, motivation, and self-esteem (Fletcher, 2007; Palmer & Roessler, 2000). Persons with LD often exhibit ineffective adaptive behavior across varying life contexts. If individuals have experienced a history of failure and frustration, they may display patterns of procrastination and lack of initiative, attributing failure to external causes and success to luck. Mithaug and Horiuchi (1983) found that adults with LD are often socially inactive as a result of fear of intimacy. This fear leads to social isolation and



loneliness. Loneliness and mild depression may lead to the overuse of prescription mood-altering drugs, alcohol, caffeine, and illegal substances. Medical consultation, individual or group therapy, and support groups are helpful in minimizing such psychosocial symptoms.

Young adults with LD are at greater risk for depression, anxiety, and even potential suicide. A study of social skills problems and issues of adults with LD indicated the following (Lehtinen-Rogan & Hartman, cited in Rubin & Roessler, 2008):

- They feel responsible to form themselves into likeable and successful people but find it difficult to do so.
- They find social relationships trying. They want and need others but lack the confidence that people will like or respect them.
- There is a substantial tendency to move between despondency and euphoria in relationships because of an underlying level of depression which is lifted by social contact.
- They are overly sensitive and easily hurt while, at the same time, being tense and anxious.

### ***Job-relevant Characteristics***

Non-academic problems, like those described above, often relate directly to employment (Carter & Lunsford, 2005). Adult employees with LD experience challenges, such as visual-perceptual, auditory processing, and temporal-perceptual problems, as well as short-term memory difficulties. Directional confusion also presents job-related challenges (Fletcher, 2007).

As previously stated, many individuals experience significant difficulties in the areas of psychosocial functioning and social adaptability. Deficits in social skills are the most debilitating aspect within the workplace (Interagency Committee on Learning Disabilities, 1987). Adults with LD may find it difficult to recognize subtle social cues and misinterpret interpersonal dynamics; they may be “socially incompetent.”

## **Reasonable Accommodation**

The following list was adapted from Vogel (1998, pp. 10-11) describing aspects of reasonable accommodation. Counselors and educators apply reasonable accommodation in schools and employment when assisting individuals who have learning disabilities.

1. **Reasonable Accommodations** in educational environments include equal opportunity for acceptance, providing or modifying equipment, and modifying instructional methodology and examination procedures to ensure that individuals with disabilities have equal opportunities to learn and to be evaluated on the basis of knowledge and skills, rather than on the basis of disability.
2. **Reasonable** refers to the idea that the suggested accommodations will not create an “undue hardship” on an institution. Undue hardship is defined by the ADA as any action that creates “significant difficulty or expense” for an employer given “the size of the employer, the resources available, and the nature of the operation.”
3. A person with a **disability** is an individual who has a “physical or mental impairment which substantially limits one or more major life activities, has a record of such impairment, or is regarded as having such impairment” (ADA, 1990).
4. **Physical or mental impairments** include physical disabilities and mental disabilities (e.g., mental retardation, organic brain syndrome, emotional or mental illness, and specific learning disabilities [ADA, 1990]).
5. **Learning disabilities** are protected under Section 504 of the Rehabilitation Act of 1973, but the learning disability must be significant enough to substantially limit one or more major life activities.
6. A **substantial limitation** occurs when a person is unable to perform a task or is significantly restricted because of the disability.
7. The concept of an **otherwise qualified person** pertains to adults with LD across various settings, such as literacy environments, job training, adult education, adult secondary education classes, and

postsecondary institutions meeting the criteria for admission and participation in education programs and activities.

8. A **program or activity** refers to all education programs that receive federal subsidies or grants. This includes all public elementary and secondary schools; adult educational environments, such as adult education programs; postsecondary vocational, trade, and military schools; and public (and most private) colleges and universities.

Requests for accommodation must be documented and establish that the identified disability interferes with the person's ability to perform the relevant tasks in a manner similar to persons without disabilities. It is incumbent upon the instructor, employer, or accrediting organization to establish whether or not the proposed "reasonable" accommodation will modify the essential requirements, standards, or functions of the task. If it is found that the accommodation does in fact modify those essential functions, the request for accommodation can be denied.

## **Rehabilitation Potential**

### ***Characteristics for Success***

Despite the unique challenges facing adults with learning disabilities, many experience success in employment and other life circumstances. A study conducted by Ginsberg, Gerber, and Reiff (1994) investigated 70 adults with LD who were successful in their professions. This study and previous research by Silver & Hagin (1985) suggested that higher socioeconomic status, greater educational levels, and appropriate intervention maximize the possibilities of successful employment.

Ginsberg et al. (1994) indicated that the overriding factor leading to success with persons who have learning disabilities was *control*. Taking control was categorized into several themes related to: (a) internal decisions and (b) external manifestations. Internal decisions included the characteristics of desire, goal orientation, and reframing. External manifestations (i.e., adaptability) were characterized as persistence, goodness-of-fit, learned creativity, and social ecologies. All these characteristics were found to interact with one another to either optimize or reduce a person's potential for success.

### ***Internal Decisions***

While desire and motivation to succeed relate to general success, they play an even more vital role in the success of adults with learning disabilities. Through goal orientation, practical and obtainable aspirations can be established. Reframing involves a reinterpretation of the LD experience from negative and dysfunctional to positive and functional, allowing one to recognize that obstacles can be overcome (Trainor, 2005). Steps in the process of reframing include: (a) recognition of the disability; (b) acceptance of the negative and positive ramifications of having LD; (c) understanding the disability and its implications, as well as one's own strengths and weaknesses; and (d) taking productive action.

### ***External Manifestations/Adaptability***

Various aspects of external manifestations/adaptability are key to success. *Persistence*, for example, involves consistently working hard, being willing to sacrifice, and delaying personal gratification. *Goodness-of-fit* involves personal adaptation to surroundings and environments which encourage success. *Learned creativity* incorporates various strategies, techniques, and other mechanisms devised to enhance one's ability to perform. *Social ecology* includes ability to utilize supportive and helpful people, and develop and implement a variety of self-improvement processes. By identifying unique and personal ways to accommodate limitations to accomplish tasks, anticipating possible difficulties, and devising various options for problem-solving, adults are provided with a variety of alternative coping strategies (Heward, 2006; Fletcher, 2007).

### ***Interventions and Accommodations***

#### ***Functional literacy***

Various adult basic education and postsecondary education programs exist to ameliorate functional literacy deficits. Whatever program is chosen, instructional interventions should be individualized to meet

area-specific functional literacy needs. Crawford (1998) provided a thorough discussion of the process for developing abilities-based literacy and employment preparation services for adults with learning disabilities.

### ***Assistive Technology (AT)***

Another tool utilized to maximize functional literacy skills is assistive technology. Various applications and types of AT afford powerful accommodation/compensatory means to ensure the ability to perform competently and successfully despite reading, writing, and organizational problems. For difficulties in written language, assistive devices include word processors, spell checkers, proofreading programs, speech synthesizers, and speech recognition systems. For reading, optical character recognition systems, screen review systems, and tape recorders are options. With organizational or memory compensation deficits, personal data managers and free-form databases are effective. Difficulties in mathematical computations are compensated with use of calculators and simple spreadsheets. An appropriate and useable fit may determine if a person will continue to use the AT.

The importance of AT for people with disabilities cannot be overemphasized. The effective use of AT provides opportunities for enhanced self-sufficiency, increased independence, and greater potential for gainful employment. AT extends into the areas of daily living, social functioning, recreation, and work opportunities, thus diminishing a person's functional limitations and helping to "level the playing field" (Brodwin, Star, & Cardoso, 2007).

AT is an effective and powerful tool to increase functioning, enhance quality of life, and remove physical and attitudinal barriers for persons with LD. With increased access to the environment and enhanced social integration, individuals can more effectively and readily interact in all phases of society. Through use of AT, people with LD can achieve greater productivity within both education and employment, leading to enhanced self-sufficiency, independence, freedom of choice, and social integration (Brodwin, Swett, Lane, & Star, 2005).

### ***Job Relevant Skills***

There are several skills related to employment that affect adults with LD (Rubin & Roessler, 2008). Adults often lack adequate job acquisition skills. Because of reading and auditory difficulties, there are often significant problems acquiring employment. Individuals with LD have difficulty accessing information about jobs through newspapers, the Internet, radio, and television. They have difficulty completing employment applications due to lack of skills in handwriting, reading, spelling, grammar and punctuation, organization, and comprehension.

The high level of credentialing and entrance examinations required by vocational and professional jobs function as a barrier for adults with LD. These exist despite the ADA mandate for reasonable testing accommodations. Adults often are hesitant to request such testing accommodations prior to employment for fear of discrimination by prospective employers.

The interview process is a challenge for many adults with learning disabilities because a positive interview involves the utilization of complex social skills, and the ability to decide whether or not to disclose information pertinent to the learning disability. Job coaches and job preparation workshops can assist people in successfully navigating these job acquisition skills (Trainor, 2005). Through these programs, vocational strategies are devised to optimize work-related assets and minimize disability-related problems (i.e., development of possible reasonable accommodations for a job).

Krishnaswami (1984) identified the following 15 employability factors needed for persons with severe LD to enter the labor market:

1. Hygiene, grooming, and appropriate dress.
2. Ability to relate to supervisors and co-workers.
3. Skills in communication.
4. Increased frustration tolerance.
5. Appropriate responses to criticism.

6. Proper social behavior at work.
7. Punctuality and attendance.
8. Initiative in carrying out work tasks.
9. Awareness of safety and work rules.
10. Increasing stamina/energy levels for work.
11. Organizational skills with job tasks.
12. Consistency in the various aspects of work.
13. Improved speed performance on the job.
14. Enhanced independent functioning with work assignments.
15. Attending to and following up with directions.

Rubin and Roessler (2008) summarized job adjustment problems for individuals with LD. These included: slow work pace, arriving late at work, problems with coworkers and supervisors, inefficiency, high error rates, proneness to accidents, deficient academic skills, problems in learning sequences of tasks, and deficiency of social skills.

### ***Social Competency and Adaptive Behavior***

Job skills training should involve careful development and refinement of needed job-related social skills. Adequate social competence is necessary for all jobs, especially as the workplace becomes more collaborative. Classes in social skills, support groups, and individual sessions with a rehabilitation counselor are various ways of enhancing social competence (Palmer & Roessler, 2000).

Colleagues can provide social assistance help through on-the-job mentoring (Carter & Lunsford, 2005). Mentors can explain the dynamics of inappropriate behavior, provide tools for overcoming specific problems, and explain unspoken cultural mores. Social skills are inherent for job advancement in higher-level positions, and involve managerial responsibilities that include the ability to work closely with a variety of personnel.

### ***Self-efficacy***

Adults with LD develop self-efficacy as they experience various problem-solving strategies. These perspectives allow individuals to increase means, techniques, and strategies to overcome the challenges presented by learning disabilities (Trainor, 2005). To learn effective coping strategies, individuals must cultivate a personal understanding, a process aided by perseverance, creativity, and support. By learning and using effective employment techniques, adults with LD optimize their potential.

## **Case Study**

Dennis Hoover is a 34 year-old Anglo male with a well-documented learning disability; he is currently unemployed. Because Mr. Hoover has been unable to maintain employment for more than seven months at a time, his job counselor at the employment office has referred him to the Department of Rehabilitation for further job training.

As the youngest of six children, Dennis grew up in a working class family in a small rural town. As an infant, he experienced several serious pneumonia infections and spent much of his first two years of life in and out of the hospital. Eventually, his pediatrician discovered that Dennis had a genetic autoimmune disorder. Medical records obtained from the family physician supported this information.

During elementary school, Dennis experienced serious difficulties socially as well as academically. He had trouble learning to read and write. As a result, he failed first grade and reports that his parents were unsuccessful in having school officials provide additional academic help. Finally, in fourth grade, Dennis was referred, evaluated, and identified as having a specific learning disability and was placed in a self-contained classroom for learning disabilities. He remembers being teased by the neighborhood children and his older brother, occasionally being referred to as “retard.” Throughout his school experience, Dennis worked hard to learn to read, write legibly, and master basic mathematics.

Upon entering high school, Dennis was fully mainstreamed. His mother and siblings helped him complete reading assignments and written homework. Since his grades were insufficient to qualify for a vocational program at the high school, Dennis was placed on a non-vocational track. Although proud of Dennis' participation in high school football and wrestling, the football coach often complained that Dennis was "clumsy" and "lacked coordination." Standing 6 foot 4 inches and weighing 220 pounds, his nickname became "the Hulk."

After graduating high school, Dennis obtained a job as a cook in an upscale restaurant and bar in his hometown. Though he enjoys cooking and reported considering going to "chef school," Dennis changed his mind after discovering that the nearest program required passing a competency test involving proficient reading. Through intervention by the Department of Rehabilitation counselor, Dennis was able to maintain his job as a cook. However, after one year, he quit because of the irregular and long hours, without notifying his counselor.

Since that time, Mr. Hoover has had several unskilled, general labor jobs. These include a position as a factory worker in the canned food industry, a seasonal laborer, a non-union construction worker, steel worker, dock worker, and dishwasher. Keeping a job has been difficult; at times, Dennis was fired for arriving at work late or leaving before the end of his shift. Other times, he quit a job because he felt the pay to be insufficient. Dennis also had disagreements with fellow workers and supervisors. Currently, Dennis states he wants to "get his act together" and is thinking about acquiring a career which will help him obtain financial stability.

Mr. Hoover's poor finances and its affect on his family have motivated him to request vocational rehabilitation. As Dennis stated, he is tired of feeling overwhelmed and unable to provide for his family's financial stability. During the past three years, he has been living with a woman and her three teenagers, children from a prior marriage who are frequently in trouble at school. A local social service agency has threatened to take the 13 year-old girl away unless they are able to provide a more stable home environment. He and his girlfriend spend money excessively and drink alcohol heavily on weekends. Dennis wants to be a good provider and father figure, but does not know how to change his current situation.

### **Questions**

1. What kinds of services are available and relevant to Mr. Hoover's needs?
2. Describe how you, as the rehabilitation counselor, will proceed with this case?
3. Develop two appropriate and feasible vocational goals for Dennis. Outline two vocational plans.
4. Are there additional assessments that need to be conducted to better understand the work-related strengths and needs of this client?
5. What kinds of work-related accommodations would you suggest?
6. Identify other human services you would recommend for Mr. Hoover to assist him in stabilizing his life.

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## Chapter 27

# PERVASIVE DEVELOPMENTAL DISORDERS (AUTISM)

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## Introduction

In the area of developmental disabilities, two terms are used interchangeably: pervasive developmental disorders (PDD) and autism spectrum disorders (ASD). For the purposes of this chapter, the term PDD is used to present the information. PDD has been a diagnosis since the 1940s (National Research Council, 2001). In recent years, the diagnosis has become more prevalent in the United States. It is estimated that two to six per 1,000 children have a PDD (National Institute of Mental Health [NIMH], 2007).

PDD includes five different categories: autistic disorder, Asperger syndrome, pervasive developmental disorder, child disintegrative disorder, and Rett's syndrome (American Psychiatric Association [APA], 2000). PDD ranges from a severe form, called autistic disorder, to a milder form, Asperger syndrome (National Research Council, 2001). Debates regarding Asperger syndrome include whether it is a part of PDD, or should be classified as its own distinct disorder. The primary feature of all PDDs is impairment of social reciprocity, such as limitations in social motivation and emotional recognition. In addition, impairments in communication and restricted behaviors like repetitive behaviors (e.g., hand flapping), narrow interests, and rituals or stereotypes are usually present (APA, 2000).

The following sections will focus on (a) types of PDD categories, (b) characteristics of adolescents and adults with PDD, (c) prevalence, (d) etiology of PDD and, (e) rehabilitation potential of individuals with PDD. Functional limitations found in adolescents and adults with PDD are highlighted.

## Types of Pervasive Developmental Disorders

### *Autistic Disorder*

Among the PDDs, autistic disorder is the most widely known (National Research Council, 2001). This condition has its onset in infancy with manifestations varying greatly depending on the developmental level and chronological age of the person. Autistic disorder is marked by a significant presence of abnormal or impaired development in social interaction and communication, and a high degree of restricted, repetitive, and stereotypical patterns of behavior, interests, and activities. Individuals with this disorder have a range of behavioral symptoms, which include hyperactivity, short attention span, impulsivity, aggressiveness, self-injurious behaviors, and temper tantrums, especially in young children (APA, 2000).

Autistic disorders are linked to other conditions such as mild to profound mental retardation, fragile X syndrome, Turner's syndrome, tuberous sclerosis, and Tourette syndrome. Occasionally, individuals with autistic disorders are misdiagnosed as having psychiatric illness, such as schizophrenia (Volkmar & Klin, 2005).

### *Asperger's Syndrome*

This differs from autistic disorder in that there are no significant delays or deviance in speaking, receptive language, or cognitive development. Individuals with Asperger's syndrome have significantly stronger verbal skills. Other than social interaction, there are no marked delays in adaptive behavior,

age-appropriate self-help skills, and curiosity about the environment in childhood. Psychological disorders associated with Asperger's syndrome, but not caused by it, include mild mental retardation, compulsive disorders (e.g., Tourette syndrome and obsessive compulsive disorder), dyslexia, dyscalculia, bipolar disorder, depression, anxiety, panic disorder, Attention Deficit Hypertensive Disorders (Corbett & Stanczak, 1999), overactivity, eating disorder, and sleep problems (Attwood, 2007).

### ***Pervasive Developmental Disorder-Not Otherwise Specified (PPD-NOS)***

PPD-NOS has been a source of confusion for clinicians and parents. Unlike autistic disorder and Asperger's syndrome, PDD-NOS has been a necessary but vague designation that does not have specific criteria (Strock, 2004). In the *Diagnostic and Statistical Manual of Mental Disorders-Fourth Edition Text Revision* (DSM-IV-TR) the PDD-NOS category refers to the diagnosis that does not meet the specific criteria for either autistic disorder or Asperger's syndrome. There is, however, a severe and pervasive impairment in the development of reciprocal social interaction linked with impairment in either verbal or nonverbal communication skills or with the presence of stereotyped behavior, interests, and activities (APA, 2000).

### ***Other PDD Categories***

These include Childhood Disintegrative disorder and Rett's syndrome both of which are rare. Childhood Disintegrative disorder has a strong male preponderance. Symptoms may appear by age two, but the average onset is between three and four years. Until this age, the child has age-appropriate skills in communication and social relationships, before the dramatic and extensive loss in motor, language, and social skills. Childhood Disintegrative disorder is also accompanied by loss of bowel and bladder control, seizures, and low IQ (Strock, 2004). Rett's syndrome is relatively rare, affecting only females. After a period of normal development, between 6-18 months, autism-like symptoms begin to appear, such as loss of social contact, expressive and receptive language, as well as motor coordination problems. These individuals may develop seizures and ataxia (Towbin, Mauk, & Batshaw, 2002).

## **Characteristics**

### ***Adolescents and Adults***

The life course of individuals with PDDs is varied; some individuals lose skills over time, others reach a plateau in adolescence, while others display patterns of continued development into adulthood (LeBlanc, Riley, & Goldsmith, 2008). Developmental changes occurring during adolescence and adulthood may explain the changes in key PDD symptoms. The core symptoms of autism often decrease during adolescence and young adulthood, particularly for individuals without intellectual disabilities. As a result, only 55% meet the criteria for autism in adulthood (compared to 100% in childhood) with basic language being the greatest improvement, and friendship development having the least symptom improvement (Seltzer, Shattuck, Abbeduto, & Greenberg, 2004).

Some persons experience improvements that are limited to certain core features of PDD with variable timing of improvements across behaviors. Shattuck et al. (2007) found that adult participants had fewer maladaptive behaviors (i.e., self-injury, noncompliance, aggression) and experienced more improvement in these behaviors over time when compared to adolescents. Table 1 provides the characteristics of individuals with PDD.

### ***Social Interaction***

Individuals with PDD, especially adults with autistic disorder, may be eccentric intellectuals who speak their mind and make insensitive remarks in social situations. Some adults with autistic disorder have delayed use and understanding of speech (Shea & Mesibov, 2005). These individuals tend to have long-standing difficulties in non-verbal expression, avoid mutual gaze, lack gestures, and have diminished facial expression. They may misunderstand the reactions of other people and lack empathy (Tantam, 2000). There is a lack of spontaneous desire to share enjoyment, interests, and achievements with other people, preferring solitary activities.



**Table 1**  
**Summary of Characteristics of Individuals with PDD**

Disorder	Childhood	Adolescents and Adults
Austistic Disorder	<ul style="list-style-type: none"> <li>• Impairments in reciprocal social interaction</li> <li>• Impairments in communication and language</li> <li>• Repetitive, stereotyped behaviors</li> <li>• Oversensitivity to sensory stimuli</li> <li>• Feeding and eating problems</li> <li>• Aggression and temper tantrums</li> <li>• Self-injurious behaviors</li> <li>• Short attention span</li> <li>• Insomnia</li> </ul>	<ul style="list-style-type: none"> <li>• Problems in friendship development</li> <li>• Eccentric intellectuals who speak their mind</li> <li>• Make insensitive remarks in social situations</li> </ul>
Asperger's syndrome	<ul style="list-style-type: none"> <li>• Impairments in social interaction and reciprocity</li> <li>• Language appears typical but have semantic and pragmatic language impairments</li> <li>• Have severe impairments in recognizing and expressing nonverbal aspects of communication (e.g., eye contact and body position)</li> <li>• Have restricted interests and repetitive behaviors</li> <li>• Exhibit weakness in motor coordination skills</li> <li>• Poor in concept formation, flexible problem solving, and complex verbal or reading comprehension</li> <li>• Strong aptitude for rote learning, spelling, and vocabulary</li> </ul>	<ul style="list-style-type: none"> <li>• Failure to develop peer relationships</li> <li>• Lack of social or emotional reciprocity</li> <li>• Impairment in use of multiple nonverbal behaviors</li> <li>• Unable to conform with social demands</li> <li>• Restricted interests and repetitive motor behaviors</li> <li>• Depression and generalized anxiety</li> <li>• Mood problems and other disorder problems</li> <li>• Inflexible adherence to routine</li> </ul>
Childhood Disintegrative Disorder	<ul style="list-style-type: none"> <li>• Dramatic and profound loss of any two areas below: <ul style="list-style-type: none"> <li>- Expressive or receptive language</li> <li>- Social skills/social interaction</li> <li>- Play and motor skills</li> <li>- Bowel/bladder control</li> </ul> </li> </ul>	

Similarly, individuals with Asperger's syndrome exhibit problems with social interactions. Some isolate themselves from others, taking refuge in a special world of idiosyncratic interests, routines, and private preoccupations. For those who cope in this way, close friendships and sexual relationships hold little attraction (Attwood, 2007). Other people with Asperger's syndrome are painfully aware of their desire for closeness, and may succeed in making friendships with people of their own gender, and who often have some social difficulties themselves (Tantam, 2000).

Generally, even when persons with PDDs develop friendships, there is often more reported loneliness compared to their typically developing counterparts. Although high functioning individuals with PDDs are more likely to report having friendships than those with less developed skills, their relationships often focus on common restricted interests rather than social interactions. Besides a low prevalence of friendships, there is minimal presence of participation in social and recreational activities. In addition, there is little planning or direction about the future, and a reluctance to accept responsibility (Leblanc et al., 2008).

### ***Victimization***

Throughout their lives, individuals with PDD are at risk for victimization, with the form and type of victimization changing across the lifespan. For adolescents with PDD, bullying and peer victimization is

common, besides gang attacks, while robbery is linked with adulthood. Adolescents are at a high risk for emotional bullying by siblings or peers (Leblanc et al., 2008).

People with Asperger's syndrome who have experienced victimization may ruminate about it for many years, and this leads to oversensitivity to slights, which can accelerate to aggression leading to social avoidance (Attwood, 2007). Antisocial and conduct problems, when they are part of the repertoire of a person with Asperger syndrome, cause great difficulty. A serious offense may lead to long-term hospital care or prison.

### ***Work-Related Problems***

Individuals with PDD have difficulty with work-related activities and employment (Fleisher, 2006). For example, they resist changes in daily routines and take prolonged periods of time to adjust after a long holiday, or refuse adaptation to a new or unfamiliar environment. Adolescents with Asperger's syndrome are likely to have increasingly conspicuous difficulties with planning and organizational skills, and completing assignments in time (Johnson, 2005). This leads to a decline in work outcomes, although not due to deterioration in intellectual abilities. Meanwhile, work-related difficulties become exasperated by stress and change.

Perspective employers may not hire people with Asperger syndrome thinking, sometimes rightly, they are not able to perform their jobs with satisfaction (Murray, 2006). These difficulties affect every person with disability, but have a particular demoralizing effect on the person with Asperger's syndrome, who cannot distinguish between self-presentations and intrinsic worth (Tantam, 2000).

### ***Health Problems***

As children progress toward adolescence, there is a high likelihood of developing various health problems that co-occur with PDDs (Odom, Horner, Snell, & Blacher, 2007). Failing at social tasks may continue throughout life and may be a constant source of distress. Individuals with PDD exhibit many behavioral difficulties including hyperactivity, attention problems, obsessive-compulsive phenomena, self-injury, tics, and affective symptoms. Problems with impulse control, disorganized behavior, aggression, and disruptions are prevalent (Leblanc et al., 2008).

Generalized anxiety and depression are more common in high-functioning PDD groups than adolescents of the same age, and typically continue to worsen throughout adolescence and into adulthood. Anxiety disorders are particularly usual expressions of distress, and long-standing anxiety may lead to depression and substance misuse; rituals may increase. Depression may cause increased social withdrawal and self-neglect, which leads to social breakdown (Kim, Szatmari, Bryson, Streiner & Wilson, 2000; Tantam, 2000).

Generally, causes of emotional disorder are due to (a) vulnerability of affective disorder (e.g., high "trait" anxiety), (b) family tension or breakdown, (c) higher than usual rate of adverse life events, (d) victimization, (e) awareness of difference, (f) over-sensitivity to slight ruminations about past injustice, and (g) broken relationships (Ghaziuddin, Weldmer-Mikhail, & Ghaziuddin, 1998).

### ***Sexuality***

Individuals with PDDs have less sexual knowledge and experience than their peers. Sexual knowledge is positively correlated with cognitive functioning. Consequently, those with higher cognitive impairments display more inappropriate sexual behaviors, engage in fewer privacy-seeking behaviors, and have less sex education than typically developing peers. Of particular concern is the likelihood that interests in the part of greater functioning individuals with PDDs might be misinterpreted as threatening or predatory behavior (Stokes, Newton, & Kaur, 2007).

As a result, only a small proportion of individuals with PDD develop intimate relationships in adolescence or adulthood (Larsen & Mouridsen, 1997). As mentioned, the centrality of social impairments in PDDs may cause difficulties in initiating and maintaining relationships, making them challenging or unappealing for potential partners. Sexual relationships are more difficult to achieve and they may find their self-esteem further eroded by rejections. Sometimes, when relationships do develop, it is on the basis of

exploitation of the person with Asperger syndrome, and the effect is undermining rather than rewarding (Attwood, 2007).

## Prevalence

The Centers for Disease Control and Prevention (CDC, 2007) estimates that 2–6 per 1,000 (from 1 in 166 to 1 in 500) children have a PDD. Increases in rates of PDD have been found in studies of both prevalence and incidence. Prevalence figures indicate how many individuals have the condition at a specific point in time. Incidence refers to the number of new cases occurring in a specified time period. If calculated accurately, incidence rates are more sensitive indicators of changes in etiological factors than prevalence (Attwood, 2007).

Although the prevalence of Asperger's syndrome has increased considerably, there has been significant variation (National Research Council, 2001). This is partly due to the lack of a clear distinction between individuals who have compensated well for Asperger syndrome, and those who are socially clumsy and take themselves to be, and are taken to be, normal. Also, many professionals continue to believe that Asperger syndrome is a term adopted by high-achieving families who do not want the stigma of a child diagnosed as having a learning disability (Tantam, 2000).

Possible reasons for increase in prevalence and incidence include: (a) changes in diagnostic criteria; (b) variations in methods used in studies; (c) increased awareness among professionals, parents, and the general public of the existence of autistic spectrum disorders; (d) the growth of specialist services; (e) probable causes and relation to age of onset; (f) the association of autism conditions with mental retardation, other developmental or physical disorders, besides psychiatric disorders of any type; and (g) possible true increase in numbers (Nebel-Schwalm & Matson, 2008; Wing & Potter, 2002). Nevertheless, most, if not all, of the reported rise in incidence and prevalence is because of changes in diagnostic criteria and greater awareness among professionals and parents.

## Etiology

According to the DSM-IV-TR (American Psychiatric Association, 2000), there is strong evidence from twin studies that genetic factors are of major importance in the etiology of over 90% of cases of autistic disorders (Rutter, 2000). Furthermore, compelling data have emerged to support the view of PDDs as a group of biologically-based brain disorders with a strong genetic basis (Muhle, Trentacoste & Rapin, 2004). However, genetic factors alone are very unlikely to be attributed to a real rise in yearly rates that appear to have occurred so rapidly and persistently. If there is an actual rise that is continuing, environmental factors must be involved that interact with PDD susceptibility genes.

Many suggestions have been made concerning other possible causes, including diet, environmental pollutants, antibiotics, allergies, mumps/measles/rubella (MMR) vaccines, and traces of neurotoxins, such as mercury in preservatives; however, none has been scientifically validated. With the MMR vaccine, current public concern that the vaccine is responsible for the observed increase in autistic conditions is based on the work of one particular group of researchers (Wakefield & Montgomery, 2000).

Most cases of Asperger syndrome are presumed to be congenital; it is therefore expected that the impairment will be life-long and that it, or its precursors, are manifest throughout life (Attwood, 2007). There are a number of neurological disorders with psychological manifestations which are associated with Asperger syndrome and may be causal. These include tuberose sclerosis, neurofibromatosis, and syndromes associated with chromosomal abnormalities (Tantam, 2000).

## Functional Limitations and Rehabilitation Potential

### *Functional Limitations*

Table 2 presents a summary of functional limitations in adolescents and adults with PDD.

**Table 2**  
**Summary of Functional Limitations of Adolescents and Adults with PDD**

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Cognitive	<ul style="list-style-type: none"><li>• Difficulty with planning and organizational skills</li><li>• May be eccentric intellectuals who speak their minds</li></ul>
Social/sexual	<ul style="list-style-type: none"><li>• Insensitivity</li><li>• Limited nonverbal skills</li><li>• Difficulty forming friendships</li><li>• Delayed use and understanding of speech</li><li>• Conversations limited to their topics of interest.</li><li>• Low participation in social and recreational activities</li><li>• Steer away from romantic relationships</li><li>• Changes in nature of friendship, body shape, etc., may precipitate crisis</li><li>• Social/sexual problems may cause depression and anger</li><li>• Suffer victimization of different types</li></ul>
Behavior	<ul style="list-style-type: none"><li>• Impulsive, display disorganized and disruptive behavior, aggression</li><li>• Hyperactivity and attention problems</li><li>• Engage in self-injury</li><li>• Low empathy</li><li>• Adherence to rituals</li></ul>
Health problems	<ul style="list-style-type: none"><li>• Mood disorders</li><li>• Depression, generalized anxiety disorders, obsessive compulsive disorder, conduct disorders</li><li>• Use of illegal substances to cope with daily challenges</li></ul>
Employment	<ul style="list-style-type: none"><li>• Inflexible</li><li>• Difficulty adapting to new environments</li><li>• Trouble completing assignments</li><li>• Interest in only a few types of activities</li><li>• May not discern the reason for rejection at job interviews</li><li>• Seldom plan for the future</li><li>• Hesitant to take up adult responsibilities</li></ul>

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### ***Rehabilitation Potential***

Individuals with ASD experience countless challenges in daily living. People with high-functioning ASD do know the social rules and can learn the skills but do not know how to apply these skills. They often lack the ability to recognize social cues, understand reciprocal conversation, and use humor appropriately. Social skills are a prominent part of employment success. While employers place the greatest importance on functional aspects of a job, socialization and integration in the workplace also are crucial. As a result, social impairment of individuals with ASD causes difficulties in employment and job retention. For those qualified individuals in the workplace, employment levels and occupational status are typically low; when employed, they are often underemployed. As a great majority of this population remains highly dependent, family members are often involved in job seeking while providing other supports (Hiller, Fish, Clopper, & Beversdorf, 2007).

Over the past two decades, a growing interest in rehabilitation of adults with ASD has been on the rise. While most adults with ASD live in institutions, there are non-institutional programs throughout the

nation. People with severe or profound autism have often been placed in sheltered workshops. Individuals with mild or moderate autism benefit most from supported employment. People with ASD working in an environment utilizing a supported employment model were found to achieve significantly higher than those in non-supported employment. Supported employment as an approach to hiring and retaining people with developmental disabilities has a mission to provide stable and predictable work environments where individuals with ASD can become contributing members of the productive workforce. A job coach or employment specialist is usually hired to provide individualized training for the “supported employee” (Villamisar & Huges, 2006). The chapter on *Mental Retardation* provides a discussion on supported employment.

Work is an important aspect of life. Having employment is a source of pride and accomplishment in American society. People with high-functioning ASD have many positive qualities to offer employers. Typically, they are punctual, detail oriented, appreciate routine and repetitive work, and are loyal to employers. However, overshadowing difficulties such as employment issues associated with social skills, problem solving, and on-the-job bullying may hide their positive qualities (Wehman, Targett, & Young, 2007). These difficulties bring the individual with ASD to the attention of rehabilitation professionals.

Job advancement is often a slow, difficult process. Job loss was more common than job advancement. Often, the reason for being terminated was poorly understood by the person with autism. Reasons that typical individuals understand, such as harassing coworkers of the opposite sex, could mean nothing. Finding new jobs become increasingly difficult with each job loss, particularly when periods of unemployment have to be explained to prospective employers. When helping these individuals select training and educational programs, rehabilitation counselors need to be particularly careful. It is a disservice when individuals with ASD enter programs that do not facilitate employment (Paxton & Estay, 2007).

Basic employment skills are often lacking among individuals with ASD. Handling of feedback and incorporating it to improve performance, along with adapting to new job routines are challenging. Individuals with ASD need to be individually trained. Besides on-the-job-training, structured teaching with written expectations, job responsibilities, and work procedures in great detail are crucial to overall success. At the workplace, it is important for employees with ASD to know whom to go to for help (Wehman et al., 2007).

Counselors must assess the potential ramifications of disclosure or non-disclosure with the client. Younger workers with ASD are more likely to disclose their disability. Recent support in the education system has taught young employees with ASD to maximize their strengths and how to ask for accommodation. They often have the support of career planning and more access to governmental supports than their predecessors. Earlier intervention has resulted in higher self-esteem among those individuals with ASD who have entered the work force in the recent years. ASD in high functioning people are usually invisible. Many people think of low functioning or nonverbal individuals when they hear of autism and may not believe that a high functioning person is on the spectrum. Although only 10% of people with ASD are considered savants (a rare condition in which persons with developmental disorders have one or more areas of expertise, ability, or brilliance that are in contrast with their overall limitations), many people on the autism spectrum are extremely intelligent and high functioning (Paxton & Estay, 2007). Nonetheless, decision-making on disclosure of disability should be individualized.

## Conclusion

There is no known cause or cure for autism; however, this condition is treatable. Individuals living with autism have unique strengths and weaknesses. Some may be of average to above-average intelligence, while others are below average. Academic and training goals and objectives need to be tailored to each person’s intellectual capability and level of functioning. With the correct services and supports, people with autism can live full, healthy, and meaningful lives.

## Case Study

Jamie Soto was a sophomore college student majoring in Informational Technology who has autism and is considered highly functioning. He is a client of yours at the university counseling and career center.



Recently, Jamie's mother called and conveyed concerns about Jamie because he became totally absorbed in computer games and often stayed up most of the night. He stopped accepting and returning telephone calls from his only friend and even became reluctant to go to school. Frequently, he withdrew into his room after school, refusing to come out even for meals. The night before Mrs. Soto called, Jamie expressed no interests in life or living.

Jamie's file indicated a sudden decline in academic performance along with frequent absences from class during the past six months. Knowing the urgency of Jamie's situation, you paid a visit to his home. At the Soto's residence, Jamie did not respond to his mother's request to come into the living room. You entered his bedroom where he was playing a video game. When he eventually looked up, his eyes were blood-shot and he seemed to have neglected personal hygiene. Jamie reported failing most of his classes. When asked why, he ignored the question and disclosed he was terminated from his part-time job at a fast-food restaurant a few months ago for a reason that he could not understand. He also commented that "people are evil" before putting himself in a rocking position. As he covered his head with his hands, you saw new and dated bruise marks on his forearms.

### **Questions**

1. What additional information do you need to develop an action plan; discuss how you will obtain this information.
2. Besides ASD, do you suspect any additional disorder? Explain.
3. Has "abuse" crossed your mind? If yes, what are the steps you need to take to properly handle the situation?
4. Would a career in computer technology be realistic for this client? Discuss.
5. What services will help Jamie get back on track? Describe these possible services and the potential benefits.

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## Chapter 28

# GENETIC COUNSELING

*Joseph E. Havranek, Ed.D., C.R.C.*

## Introduction

A subject of heightened interest in rehabilitation counseling is the area of genetic testing and genetic counseling. A genetic test involves “the analysis of human DNA, RNA chromosomes, proteins, and certain metabolites in order to detect heritable disease-related genotypes, mutations, phenotypes, or karyotypes for clinical purposes” (Holtzman & Watson, 1999, p.1). Genetic testing can be done for many reasons including the diagnosis of genetic disease in a fetus, a newborn, children, and adults (Burke, 2002). A comprehensive and continually updated listing of available tests can be found at the GeneTest – GeneClinics Website (<http://www.geneclinics.org>).

Genetic testing technologies are becoming both cost-effective and available for a wide range of hereditary diseases. In many countries, screening for hereditary disease is promoted as a cost-effective way to identify individuals who are at risk for such diseases. This allows health services to be targeted to individuals who are most likely to benefit from early intervention. Over the past 25 years, programs for genetic testing and counseling have become widespread as a result of the dramatic impacts of increased knowledge of hereditary diseases, identification of genetic processes, and DNA-based testing to identify potential carriers of genetic diseases.

This chapter reviews issues in genetic counseling, the purposes and procedures involved with genetic counseling, the potential for dissemination of genetic information, and individual counseling procedures. The chapter also reviews potential expanded roles for counselors, ethical issues, research, and a sample program in Australia. Three case studies for counselors and students to discuss conclude the chapter.

## Genetic Counseling

Genetic counseling is the application of what is known about human genetics in providing advice to (1) those concerned about the possibility of having a genetic disease themselves (genetic diagnosis); (2) those family members concerned about having the same genetic disease as an affected family member (familial risk); and (3) those concerned about passing a genetic mutation to their offspring (prenatal genetic testing). It is “a communication process which deals with the human problems associated with occurrence or risk of occurrence, of a genetic disorder in a family” (Fletcher & Evans, 1994, p. 27). Goals of genetic counseling include providing assistance with:

1. Client comprehension of medical facts including diagnosis, probable cause, and management of the disorder.
2. Understanding how heredity contributes to the disorder and the risks for family members.
3. Choosing and implementing the most appropriate course(s) of action considering the risks, resources, and long-term family goals.
4. Making the best possible adjustments to a disorder in an affected family member.
5. Dealing with the risks of recurrence of the disorder (Shiloh, 1996a, 1996b).

Genetic counseling addresses human problems associated with a genetic disorder, not the medical management of the disorder itself. Goals of genetic counseling include learning, understanding, choosing, and coping to help clients adjust to and handle unexpected circumstances. Physicians who specialize in

genetic disorders (medical geneticists) and counselors who have an academic background in genetics plus training in genetic counseling (genetic counselors) do genetic counseling.

Tremendous increases in knowledge about and potential intervention in genetic issues have resulted in both positive and negative consequences with respect to reproductive freedom. A positive result involves development of a family history that reveals potential risks, thereby allowing informed choice as to whether to conceive or not. A negative consequence of prenatal testing is that it may lead some individuals to abort an affected fetus. Availability of genetic testing has resulted in a series of choices that must be faced by individuals contemplating reproduction. These choices include:

1. Whether to procreate, with whom, and by what means.
2. Social context in which reproductive freedom takes place.
3. When to reproduce.
4. How many children to have.
5. Whether to proceed with a high-risk pregnancy.
6. Whether to proceed with a pregnancy that will result in a child with significant medical problems.

An even more challenging issue is the extent to which the broader society can legitimately claim a role in such decisions (Brock, 1994).

## **Definition of a Gene**

There are 23 pairs of chromosomes in each human cell that contain molecules of DNA. DNA contains the instructions that inform various cells throughout the body how to function and react under different circumstances. These instructions are arranged in segments of DNA referred to as genes. Each gene contains a code, and these codes in combination control everything in the body, from growth to circulation to breathing. By producing many different kinds of proteins, genes help the body conduct various functions. Each person has about 31,000 genes – these contain the coding for 100,000 to 200,000 proteins.

## **The Human Genome Initiative (HGI)**

Established in 1988, the Human Genome Initiative (HGI) was initiated in response to new diagnostic procedures and treatments for genetically-based disorders; the National Institutes of Health and the Department of Energy funded HGI (Beckwith, 1991; National Human Genome Research Project, 2007). The human genome is the complete set of human chromosomes. On April 14, 2003, the International Human Genome Sequencing Consortium announced the completion of the 13-year effort to map the human genome (Manasse, 2005). While this research may significantly impact the lives of people with disabilities, there are programmatic restraints on biomedical and clinical research that may result in some areas not being explored that might otherwise be of value (Houser & Lash, 1996; Jain, 1999; Phua, 2004).

Benefits of the HGI include coordination of genetic studies, increased funding, and targeted research to greatly accelerate development of the detailed genetic map (Gostin, 1991). The density of chromosomal markers identified will be so extensive that time required for a research group to locate a specific gene will be significantly reduced. Some people who are concerned about potential negative social consequences oppose the initiative, fearing that the information will be used to discriminate and deprive people of their civil liberties, especially with respect to insurance, employment, and other societal institutions (Beckwith, 1991). National legislative action on a genetic nondiscrimination bill remains a priority (Genetic Testing Prompts Legislation and Private Policy Development, 2005).

## **Role of the Primary Care Provider**

Many primary care physicians use the basic information developed in case histories to create a family genogram to identify potential risks. With the recent development of cost-effective and available genetic testing technologies this has become more common. Currently, genetic testing only occurs in primary care: (a) when there is an obvious underlying disease; (b) if the individual family history demonstrates the potential for rare conditions with grave implications for morbidity and mortality; and (c) as



part of a limited screening by medical practitioners who engage in prenatal and obstetrical care (McCrary & Allen, 1994).

This may alter the routine practice of primary care physicians who must implement formal policies and procedures for the secure handling of genetic testing records. McCrary and Allen (1994) have recommended that the following office policies and practices be considered: (a) educate office staff about the nature of genetic testing; (b) limit the number of staff members who have access to genetic test results (including access to computer records when information is contained therein); (c) segregate genetic test results from other medical records, if feasible; and (d) verification by the physician of a patient's written permission to release information regarding genetic test results.

Screening devices will soon be available to primary care practitioners and specialists in prenatal care that will assess the potential for some genetically based diseases. Unfortunately, there is currently a shortage of counselors trained in genetic issues. Those counselors who wish to accept referrals from physicians and other health care providers who are requesting information and advice must seek out further knowledge and specialized training in genetic counseling and risk assessment.

The most basic issue confronting genetic counseling is the development of standards for genetic counseling practice (Bennett, Pettersen, Niendorf, & Anderson, 2003; Veach, Bartels, & LeRoy, 2002). Goals to be achieved as identified by the National Society of Genetic Factors cover several areas. These are (a) clinical decision-making; (b) educating individuals and groups; (c) assessing and assuring quality of counseling; (d) increasing the consistency of client care; (e) guiding allocation of resources; and (f) reducing the chance of negligent or prejudicial counseling (Bennett, et al.). This leads to the need for evidence-based practice guidelines to determine what actually happens in genetic counseling and the impact on clients and their families (McAllister, 2001). These guidelines are being developed and refined as research becomes available (McConkie-Rosell et al., 2005).

## **Issues in Genetic Counseling**

The scope of genetically-based disorders can be appreciated when one considers that (a) the smallest complex genomes are about 100 million base pairs; (b) there are approximately 4,000 identified inherited genetic disorders which affect up to 3% of pregnancies; (c) due to advances in treatment and prevention of other diseases, about 40% of childhood mortality and 5% to 10% of all pediatric hospital admissions are due to genetic disorders; and (d) advances in diagnostic measures are dramatically expanding the potential for genetic screening and prenatal diagnosis (Godard et al., 2003; Hartl & Jones, 1999; Pembrey, 1996; Shiloh, 1996b).

### ***Gene Therapy***

The first attempt at gene therapy occurred in 1990 at the National Institutes of Health; the procedure for treating and possibly curing diseases using this therapy is still very experimental. Gene therapy involves delivering new genes to replace those that are missing or those that are improperly functioning at the cellular level. The hope is that these genes become incorporated in the cellular DNA and produce the necessary protein for appropriate functioning.

As researchers are able to identify more malfunctioning genes causing disease, additional efforts will center on genetic attempts at curing the dysfunction and hence the disease. Gene therapy will require long-term testing to determine that it is (a) effective and (b) safe to use. Clinical experiments that involve testing on people have difficulty receiving approval and are thoroughly monitored throughout the testing phase (Petechuk, 2004). Yet, the possibilities for treating chronic and debilitating diseases and the potential for cure are the hope for gene therapy.

### ***Genetic Testing***

Advances in genetic testing hold promise of becoming a medical breakthrough for treatment of a number of diseases including cystic fibrosis, sickle cell disease, Fragile-X syndrome, Duchenne's muscular dystrophy, hemophilia, Huntington's chorea, retinoplasty, neurofibromatosis, polycystic kidney disease, and various cancers. Other diseases not as severe as some of these, such as Type I diabetes and other

autoimmune conditions may eventually be preventable. Identification of the gene structures involved in these and other diseases will lead to effective therapies and preventive measures. Additionally, new technologies for genome research will find a myriad of applications in industry (Bailey, 2004; Pembrey, 1996; Scott, 2004). There are already tests for over 1,000 diseases and about two-thirds are available for order by practicing physicians. However, consumers need not utilize a physician to gain access to genetic testing. Direct-to-consumer marketing of genetic testing and other laboratory services is available on the internet, which has prompted concerns that companies will exploit consumers and sell tests without appropriate counseling (Wolfberg, 2006).

## **Genetic Testing Purposes and Procedures**

### ***Purposes of Genetic Testing***

The overall purpose of genetic testing is to facilitate informed decision-making about reproduction. According to Pembrey (1996), the objectives of prenatal testing are to: (1) allow the widest possible range of informed choice for women and their partners at risk of having children with genetic disorders; (2) provide reassurance and reduce the level of anxiety associated with reproduction; (3) allow couples at risk to embark upon having a family, knowing that if they wish they might consider avoiding the birth of a child seriously affected through termination of the pregnancy; and (4) prepare a couple who wishes to continue with the pregnancy of a fetus with a genetic disorder, ensuring early treatment of the child.

Most members of both the general public and at-risk groups express very favorable attitudes toward, and interest in, being tested (Croyle, 1995). Likewise, persons with disabilities who seek genetic counseling are generally favorable to that service (Chen & Schiffman, 2000).

### ***Procedures and Indications for Genetic Testing***

Amniocentesis is currently the primary means of prenatal testing. It is performed in a physician's office and involves a trans-abdominal puncture of the amniotic sac using a needle and syringe to remove amniotic fluid. The fluid is then studied in a laboratory to detect genetic and biochemical disorders, maternal-fetal blood incompatibility, and fetal maturity. Indications for undergoing amniocentesis include an older maternal age at time of conception, family history of a genetic disorder or inborn biochemical disorder, being in a high-risk population, a parent who is a known carrier of a genetic disorder, and a woman with abnormal levels of maternal serum alpha fetoprotein indicating an increased risk of chromosomal or neural tube disorder. Individual reactions to amniocentesis are directed at both the results of testing as well as any stress-provoking effects of the procedure itself (Croyle & Lerman, 1995; Evans & Biesecker, 2006).

In addition to amniocentesis, DNA analysis is available for mothers who are known carriers of genetic disorders. Another technique for those pursuing insemination is pre-implantation diagnosis (i.e., DNA testing of in vitro fertilized embryos) to ensure that embryos with known genetic disorders are not implanted (Cronister, 1996). This procedure is used when abortion is not an option; however, it is expensive and available at only a few genetic testing centers.

Recent advances in prenatal testing, which can determine who among the at-risk population has a genetic disorder, may resolve problems of uncertainty for potential parents. Variables associated with the use of prenatal diagnostic procedures include willingness to abort, family approval of abortion, need for reduced uncertainty, perceived severity of the disorder, prior experience with a child who has a genetic disorder, the woman's age, pre-counseling reproductive plans, desire to have children, and level of genetic risk. Test results can cause ethical and personal dilemmas (Coen, 1999; Veach et al, 2002). These dilemmas may be influenced by genetic counseling during which it can be explained that there is a wide variation in the expressed severity of genetic disorders. It should be recognized that severity is, at least in part, a product of one's personal perspective.

Prenatal diagnosis can only be understood in the context of the special emotional state of prospective parents, which is influenced by stress, ambivalence, changes in self and body images, emotional instability, and changes in thought processes (Evans & Biesecker, 2006). The counselor can discuss with the client that research has shown that while significant increases in short-term psychological distress

frequently occur in light of a diagnosis, there is little evidence that this distress remains high over the long term (i.e., more than six months after testing).

## **Genetic Discrimination**

### ***Discriminatory Practices***

As the ability to test for genetic disorders increases so does the potential for discrimination. Gostin (1991, p. 110) noted that “prejudice, alienation, and exclusion often accompany genetic disorders, even though the conditions are neither the result of willful behavior nor subject to the person’s control.” Testing for a genetic marker does not establish the onset date, nature, severity, course of a disease, or efficacy of treatment. Additionally, reliability and predictability of genetic tests are limited by the extent of knowledge about genetic disorders. All we know definitely is that “significant scientific uncertainty surrounds much genetic testing” (Gostin, p. 114).

Early in the process of identifying and evaluating genetic knowledge, activities were primarily aimed at improving the gene pool. This negative and discriminatory practice was evident during the late 19th and early 20th centuries. It was manifest in Germany where the term eugenics originated. This process also was evident in the United States; by 1937, 32 states had passed sterilization laws aimed at people with disabilities, especially those with mental retardation. Serious negative implications of this practice are obvious (Houser & Lash, 1996).

### ***Legislation***

Information about hereditary diseases is highly beneficial for people to the degree that it assists with prevention, diet, treatment, lifestyle choices, and reproductive changes. On the other hand, genetic discrimination (whether based on misconceptions about the disability or perceived extent of disability) is harmful to individuals, public programs, and society because it discounts the potential contributions of a significant segment of future society. The current legal system in this country protects individuals with disabilities at three levels: (a) the Americans with Disabilities Act (ADA) of 1990 and other federal disability rights legislation; (b) state mandates outlawing discrimination; and (c) local ordinances that reinforce federal and state initiatives. The ADA protects not only individuals with disabilities but also those perceived to have disabilities. This legislation covers those who may be subject to discrimination based upon their own genetic makeup or the genetic composition of potential offspring. The Civil Rights Act of 1964 (Title VII) bars discrimination based upon race or gender, an important protection because genetic testing often disproportionately impacts people who are most vulnerable.

The ADA protects job applicants from discrimination. Pre-employment inquiries can only be made if they relate to the capability to perform essential functions of jobs; inquires about an individual’s genetic history are illegal. A job applicant cannot be asked to undergo a medical examination unless a conditional offer of employment is made. If a medical examination reveals a disability, the employer cannot rescind the offer of employment unless the prospective employee cannot perform the essential functions of the job, with or without reasonable accommodation.

President Bill Clinton signed an Executive Order prohibiting employers from discriminating against job applicants who have genetic disorders. This was based on pending and potential cases of employer discrimination resulting from genetic test results secured by employers (Phua, 2004).

On May 21, 2008, President George W. Bush signed the Genetic Information Nondiscrimination Act (GINA) of 2008 (Public Law No. 110-233), culminating a 13 year legislative effort to achieve protection for consumers from discrimination by health insurers and employers on the basis of genetic information (Hudson, Holohan, & Collins, 2008). GINA makes it unlawful employment practice for an employer, employment agency, labor organization, or training program to discriminate against an individual or deprive such person of employment opportunities because of genetic information.

This legislation defines “genetic information” as including a person’s genetic tests, genetic tests of a person’s family members (up to and including fourth-relatives), any manifestation of a disease or disorder in a family member, and participation of a person or family member in research that includes genetic testing,

counseling, or education. Genetic services are defined as tests, counseling, or education. Examples of protected tests include: (1) tests for BRCA1/BRCA2 (breast cancer) or HNPCC (colon cancer) mutations; (2) classifications of genetic properties of an existing tumor to help determine therapy; (3) tests for Huntington's disease mutations; and (4) carrier screening for disorders such as cystic fibrosis, sickle cell anemia, spinal muscular atrophy, and fragile X syndrome. GINA expressly bans the use, disclosure, or collection of genetic information for purposes of insurance underwriting.

### ***Uses of Genetic Information***

There is currently very little use of genetic information in relation to employment (Godard, et al., 2003). However, markets for genetic testing worldwide are worth millions of dollars. The industry of genetic testing, a business now estimated at \$5 billion, is growing by 25% annually (Pollack, 2006). As more effective and efficient means of testing are developed at lower cost, the potential for abuse increases. It is conceivable that with the increasing costs of health, life, and disability-related insurance, employers and insurers could use screening devices to exclude those with potential for having expensive medical claims. Genomic information could become available to employers, insurers, educators, law enforcement, and others who might not have the skill to understand or correctly serve individuals with a propensity for disability (whether perceived or actual).

The role of the genetic counselor is to explain thoroughly what the genetic situation is but the decision must be a personal one between the husband and wife (Reed, 1955). Genetic counseling is a communication process which deals with the human problems associated with the occurrence or risk of an occurrence of a genetic disorder in the family. This process involves an attempt by one or more appropriately trained professionals to help the individual or family (1) comprehend the medical facts, including diagnosis, probable course of the disorder, and available management; (2) appreciate the way heredity contributes to the disorder, and the risk of recurrence in specified relatives; (3) understand the alternatives for dealing with the risk of occurrence; (4) choose the course of action which seems to them appropriate in view of their risk, their family goals, and their ethical and religious standards, to act in accordance with that decision; and (5) to make the best possible adjustment to the disorder in an affected family member and/or the risk of recurrence of that disorder.

Issues that should be determined before undergoing genetic testing include: (a) whether test results become available to other parties, particularly potential or current employers and insurance agencies; (b) whether genetic disorders make it impossible for applicants to obtain health, life, or disability coverage; and (c) whether insurance companies decrease back benefits for certain conditions including genetically based disorders (Resta, 2007). Gostin (1991) described the potential ills that might result from genetic screening. By excluding qualified individuals from education, employment, government service, or insurance, the marketplace is robbed of skills, energy, and imagination. If fear of discrimination deters people from genetic diagnosis and prognosis, renders them less willing to confide in physicians and genetic counselors, and makes them more concerned with loss of a job or insurance than with care and treatment, the benefits of genetic data collection will not be fully achieved.

## **Individual Counseling Procedures**

Early diagnosis, using amniocentesis or other testing, enables individuals to seek genetic counseling and thus make informed decisions about possible courses of action. When helping families arrive at informed decisions, the counselor can review the options of prenatal diagnosis, pre-implantation diagnosis, adoption, and egg and sperm donor programs. Counseling may assist couples with decisions and come to solutions that are available for them. However, couples are not the only parties affected. Siblings and other relatives feel the results of the decision as well.

Identification of a genetic disorder leads to various reactions. These include shock, denial, grief, guilt, anger, disappointment, diminished self-esteem, and difficulty making decisions. Family dynamics and marital relationships are disrupted. The mother will either feel external pressure to undergo an abortion or will bond to the pregnancy in hopes that it will prove to be without major complications (Cronister, 1996). For women who present with emotional problems or neurocognitive deficits, issues of shyness, social anxiety, paranoia, and mood lability need assessment. For those with poor self-perception, fear of



physicians, concern with being inappropriately labeled, and fear of stigmatization are additional areas of concern (Phua, 2004; Shiloh, 1996a).

Assessing and addressing the counseling needs of each client is paramount for counseling to be helpful. Appreciating a client's prior knowledge, experiences, personal attitudes, expectations, apprehensions, and philosophy about having children provides insight into how to approach counseling. "Explaining complex medical and genetic information in a manageable and meaningful way is a challenge that demands time, consideration, and practice" (Cronister, 1996, p. 273). Through emotional support and understanding of the counselor, issues such as anger, guilt, and disappointment can be resolved.

Ramifications of genetic counseling affect individuals and families in many areas of life (Chen & Schiffman, 2000; Darvey, Rostant, Harrop, Goldblatt, & O'Leary, 2005). As such, the counseling process should include sufficient time and commitment by the counselor for the client and family to establish adequate rapport and understanding needed to resolve their issues. For the family who chooses to proceed with the pregnancy, the counselor must be available to support the client and family as they adjust emotionally and begin to plan for the future. Conversely, for those who choose to terminate a pregnancy, ongoing counseling is necessary to allow resolution of issues related to loss.

An issue that has received considerable attention is the type of counseling to be provided. Non-prescriptive, or non-directive, counseling has generally been employed as a means of enhancing client self-esteem. During non-prescriptive genetic counseling, the counselor serves as a provider of information. In this position, the counselor assumes a neutral role. Prescriptive (or directive) counseling approaches, on the other hand, involve the counselor as a facilitator of the process. In this role, it is recognized that counselors cannot achieve total neutrality. Instead, the counselor helps the client through a process that includes value and resource utilization designed to enhance the ultimate outcome of decisions that are made. If they are to effectively counsel individuals about genetic issues, counselors must carefully examine their own ethics regarding many issues surrounding genetic counseling and come to terms with client choices that run counter to the counselor's own beliefs. Above all, counselors must never promote their own perceptions of "good and bad," "right and wrong."

## **Expanded Roles for Rehabilitation Counselors**

Several additional roles for rehabilitation counselors related to genetic counseling have been identified. These include education, support, and advocacy (Bennett et al., 2003; Evans & Biesecker, 2006). This author proposes an additional role – that of technical advisor.

### ***Education***

Education has been identified as a prerequisite for providing genetic services for both the public and professionals. Professionals need to be well versed and current in the areas of genetics and sensitivity regarding these highly emotional issues. In regard to education, rehabilitation counselors can assist in several activities. Counselors need to provide information concerning genetic processes and explain potential procedures and results in ways that are meaningful to clients. Specific tasks include providing information about diagnosis, prognosis, and medical procedures; referral for more formal genetic counseling; advice about prenatal care; and assistance with client preparation for parenting and related issues.

### ***Support***

In the supportive role, rehabilitation counselors perform a variety of services. These include assisting families to identify and address underlying psychosocial and cultural issues related to genetic concerns; referral to, or organization of, support groups for women and their significant others; assisting clients while making decisions and providing supportive follow-up; helping clients understand and cope with the reactions of other family members; establishing contact with persons who have had similar experiences for peer counseling; and making supportive contacts with family members and others who may be identified by clients.



### ***Advocacy***

In the role of advocate, rehabilitation counselors discuss issues concerning discrimination against clients. Discrimination involves employment, medical insurance, and social inequity. Potential counselor interventions include informing employers and insurers about legal protection afforded persons with disabilities and potential accommodations in the workplace, providing information to clients about their legal rights, and maintaining supportive contacts with family members and others identified by clients.

### ***Technical Advisor***

Rehabilitation counselors can help clients by providing technical assistance; this may involve construction of a life care plan for the potential child and family (Blackwell, Weed, & Powers, 1994). The life care plan can identify potential services and costs that may be required for the child and family, as well as resources to secure such services. This use of life care plans is a practical means by which to inform the client of responsibilities engendered by a child with a disability. It can serve as a basis for planning future life care services and avoiding lapses in the provision of services for the foreseeable future.

## **Ethical Issues**

A benefit of the Human Genome Initiative has been an increase in available information about genetic defects. The rapid increase in genetic knowledge has not been matched by similar increases in counseling abilities and skills necessary to deal with this information. This poses ethical, legal, and social questions about the implications of such knowledge.

Ethical issues in genetic counseling revolve around two stages. During the first stage, the process of counseling prior to prenatal diagnosis, ethical responses are provided to client inquiries about genetic testing procedures and their uses. This is a crucial component of genetic counseling because some rehabilitation service clients have developmental or traumatic disabilities that may lessen their ability to understand technical or multidimensional information (Manasse, 2005). The role of the counselor in such cases is to explain the issues to clients in a format that they comprehend and allows them to make informed decisions.

The second stage occurs after prenatal diagnosis. Potential ethical issues that arise during this stage include choices about abortion, disclosure dilemmas, confidentiality versus third-party interests, and twin, multifetal, and third-trimester dilemmas. Primary care physicians are of great assistance in providing referral to genetic experts and interpreting results of testing with consideration given to family and cultural values. Every effort must be made to safeguard this confidential information that can significantly affect a person's access to employment, insurance, and other resources.

## **Research Needs in Genetic Counseling**

Research in genetic counseling and its consequences should focus on: (a) provision of information to clients and understanding of the material; (b) roles of non-institution members on ethical review boards; (c) potential for establishing centers for genetic counseling where comprehensive services are offered, similar to the Clinical Genetics Centres in Great Britain; (d) establishing the efficacy of life care plans to aide with reproduction options; (e) determining ways to minimize the adverse psychological sequelae and maximize the benefits of testing; (f) discovering the perspectives of the various parties involved and their impact on the process; (g) developing new, creative, and efficacious counseling techniques to be used during genetic counseling; (h) determining the extent to which advances in genetic engineering impact information provided during the genetic counseling process; (i) assessing the extent to which society can legitimately play a positive role in genetics; and (j) determining the extent to which genetic information should, or should not, be shared with employers, insurance companies, and other parties (Houser & Lash, 1996).

## **Sample Program**

In Western Australia, genetic counseling is accomplished through a public health genetic service which offers diagnosis, counseling, investigation, and education. This service includes: information on the occurrence, probable course, and avenues for management of the condition; facilitation of psychological well-being; and facilitation of autonomous decision-making. The goal is for the client to determine the best

course of action in accordance with personal and family needs and desires, client expectations, psychological adjustment, and satisfaction with services provided. Ongoing genetic counseling is carefully assessed and evaluated. Changes in services are made depending on results of this evaluation (Darvey et al., 2005).

## Case Studies

Descriptions of three case scenarios adapted from Fletcher & Evans (1994) are provided to show the complexity and interaction of issues encountered during genetic counseling. Analysis of these cases will help the counselor more clearly comprehend the dynamics involved.

### *Case 1*

A 42 year-old woman is considering prenatal diagnosis for possible Down Syndrome, which causes moderate to severe congenital mental retardation. She and her husband already have one child with this condition. They tell the rehabilitation counselor that abortion is not an option and that she plans to carry the fetus to full term despite the results of testing. The family's reason for seeking the diagnosis was to provide more time for preparation for another child that may have Down Syndrome. They ask for the counselor's advice. How should the rehabilitation counselor proceed? Discuss various options.

### *Case 2*

A 25 year-old woman with no history of genetic disorders in her family is considering prenatal diagnosis. This client has a problem with alcohol but is in denial, believing she is only a social drinker. She is a client because she had an acquired below elbow upper extremity amputation as a result of an automobile accident. Although she did not have any abnormal sequelae, at the time of the amputation she received several blood transfusions and is concerned she may have contracted hepatitis or AIDS. The client appears very anxious about the normalcy of the fetus, even after being told that, in her case, the potential medical risks of the genetic procedure (including possible miscarriage) are greater than the likelihood of diagnosing an abnormality. She asks for the counselor's advice. As her rehabilitation counselor, describe what approach you might take.

### *Case 3*

A couple in their late 20s requests prenatal diagnosis to determine the sex of their fetus. The father is a client of yours due to a recent back injury. They already have four girls and feel desperate to have a son. They stated that if it is a girl, they plan to abort the fetus and keep trying to have a boy. If they cannot have testing, they will abort the fetus rather than face the possibility of having another girl. They have requested your advice. Discuss the ramifications of this case.

These case studies demonstrate the increasing complexity of issues surrounding genetic counseling. Case one concerns questions of planning. The second case potentially involves issues of AIDS, fetal alcohol syndrome, and mental illness on the part of the mother. The last case discusses the option of gender selection. These cases illustrate the need for counselors to have a firm understanding of their own beliefs. It is essential to provide meaningful responses that cause the client and significant others to carefully examine their personal beliefs, without actually giving advice about what choice to make.

## Conclusion

Currently, there is a shortage of trained genetic counselors. This is a crucial issue because genetic counseling involves far more than just preventing pregnancy by contraception or preventing birth through abortion. Instead, it encompasses a complex, multidimensional process of providing information incrementally to help arrive at a decision that is best for the individual client. In the future, genetic counseling will become an area of specialization for rehabilitation counselors. Rehabilitation counselor education programs have recently begun to incorporate information about genetics and genetic counseling into their graduate programs. Counselors interested in providing genetic counseling will need specialized training in the field of genetics.

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## Chapter 29

# ORTHOTICS, AMPUTATION, AND PROSTHETICS

*Lance R. Clawson, B. S., C.P.O.*

## Introduction

The fields of orthotics and prosthetics are unique disciplines in the health science field. Orthoses (orthotics), commonly referred to as braces, are devices designed to support, stabilize, and assist compromised segments of the body. Prostheses (prosthetics), most often referred to as artificial limbs, are devices designed to replace the function of missing segments of the body. Amputation involves the loss of all or a portion of a body part; a prosthesis is an artificial replacement of that missing part of the body.

Professionals providing braces (orthoses) and artificial limbs (prostheses) are referred to as orthotists and prosthetists. Upon receipt of a prescription from a physician, the orthotist or prosthetist is responsible for evaluating the patient's needs and then designing, fabricating, and fitting a device to meet the prescription criteria that will restore the weak or missing function or part to the greatest extent possible.

This chapter is divided into five sections. Section one briefly describes the history of prosthetics and orthotics. The second, orthotics, describes lower and upper extremity orthotics, spinal orthotics, other assistive devices, and physical and psychological limitations. Amputation, the third section, includes levels of amputation and potential complications, psychological considerations, family involvement, and employment issues. The fourth section explores prosthetics and covers treatment, types of lower and upper extremity prosthesis, and Medicare functional levels. Assistive technology and a case study conclude this chapter.

## History of Prosthetics and Orthotics

The study of orthotics began with the ancient art of splint and brace making. Bonesetters and brace makers eventually developed into what we now call orthopedic surgeons and orthotists (Kirkup, 2007). The study of prosthetics has been closely associated with amputation surgery performed as a lifesaving measure from the aftermath of war. Injured soldiers who returned home from battle with traumatic amputations utilized primitive wooden prostheses. Each major war has been a stimulus for the improvement of amputation surgical techniques and for the development of improved prostheses (Bunch, 1985).

It was not until the twentieth century when the most significant contributions to prosthetic/orthotic sciences were made; these contributions were stimulated by the aftermath of the first and second world wars and the polio epidemics of the late 1940s and early 1950s. Injured veterans who acquired musculoskeletal and neuromuscular impairments or traumatic amputation and polio survivors with diminished neuromuscular function increased the demand for orthotic and prosthetic services. To improve the quality and performance of assistive devices at the end of World War II, particularly for veterans with amputations, the United States Government sponsored a series of research and development projects under the auspices of the National Academy of Sciences that would forever change the manner in which orthotics and prosthetics would be performed (Fishman, 2001).

In the early 1960s, the National Academy of Sciences National Research Council began to promote multidisciplinary scientific research efforts into human locomotion, biomechanics, and the development of new materials and devices (Bunch, 1985). By the 1980s, the continuing introduction of new materials and methods spurred the profession of prosthetics and orthotics to rapidly evolve as a changing discipline. The

1990s saw significant advances in development of educational programs with the establishment of national education accreditation through a subsection of the American Medical Association (Fishman, 2001). Today, prosthetics and orthotics has developed into a burgeoning multifaceted profession. Continued technological advancements in engineering and medicine have led to advanced prosthetics technology including lighter, more comfortably fitting, and computer-assisted prostheses.

## **Orthotics**

Each year thousands of individuals in this country are affected by injuries and disease that result in failed or weakened parts of the body. A person who has fractured a finger may see a physician who then tapes the finger to a splint; by definition, the splint constitutes a simple orthosis. Physicians provide many simple orthoses. Other types of simple braces are found at local pharmacies and sporting goods stores. When a simple support is not sufficient and the physician believes a more complex device is needed, he or she prescribes an orthosis and directs the patient to an orthotist. Orthoses (orthotic devices) are divided into four categories: lower extremity, upper extremity, spinal, and other assistive devices. These are further divided into the categories of acute, chronic, and assistive orthoses. Orthoses are named after the joint areas of the body the particular orthosis stabilizes or affects (see Table 1). Formerly, braces were known by the name of the designer or institution that first made the device popular. A simple brace for the foot and ankle is referred to as an AFO (ankle-foot orthosis); one for the hand and wrist is called a WHO (wrist-hand orthosis). In the prescription for the orthosis, the physician indicates the type of motion to be limited, stabilized, or assisted, and the purpose of the device or goal to be achieved (e.g., greater strength, improved range of motion) (Bussell, 2000; Lusardi & Nielsen, 2000).

The physician may specify whether the orthosis is to be manufactured of metal or plastic, or a combination of both. In contrast, he or she may leave the prescription intentionally vague to allow the orthotist to determine what type of design will most likely succeed. In some cases, the patient requires both acute (short-term) and long-term orthoses. A patient who is post-cerebrovascular accident (stroke) and is at risk of losing range of motion in certain joints may be fitted with a soft, static orthosis. The purpose of a static orthosis is to keep the involved portion of the body in the desired alignment. Although this may result in the provision of two braces rather than one, prophylactic or acute fitting reduces the risk of joint contractures.

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**Table 1**  
**Categories of Orthoses**

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Ankle-Foot Orthosis	AFO
Knee-Ankle-Foot Orthosis	KAFO
Hip-Knee-Ankle-Foot Orthosis	HKAFO
Wrist-Hand Orthosis	WHO
Wrist-Driven Wrist-Hand Orthosis	WDWHO
Shoulder-Elbow Orthosis	SEO
Lumbosacral Orthosis	LSO
Thoraco-Lumbo-Sacral Orthosis	TLSO
Cervico-Thoraco-Lumbo-Sacral Orthosis	CTLSO

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### ***Lower Extremity Orthoses***

Following the acute episode of a cerebrovascular accident, the person may have the need to be fitted with a static or functional (dynamic) AFO (ankle-foot orthosis) or KAFO (knee-ankle-foot orthosis). The ankle-foot orthosis supports lower extremities that have instability about the ankle. It can be designed to simply compensate for paralytic dropfoot, preventing the foot from dragging during the swing phase of gait and to control the rate at which the forefoot contacts the ground. If more stability is needed, the AFO has a rigid ankle (at 90 degrees), thereby preventing the tibia and knee from collapsing forward in mid to late

stance during walking. The same device may be designed with movement, such as with an articulating joint. If the knee also has significant weakness or instability, the orthosis may span another joint (the knee) proximally, becoming a KAFO. The design may incorporate free motion, locking, offset knee joints, or operate as a rigid system without joints. This type of extension may continue to the hip and into the lower back depending on the person's paralysis or weakness (Lusardi & Nielsen, 2000).

### ***Upper Extremity Orthoses***

The arm, hand, and fingers are significantly more complicated to design and fit with orthotic devices than the lower extremities. Sensation, dexterity, and function of the fingers and hand are crucial to the activities of daily living and independence of every individual. Similar to the lower extremity, the hand and arm may be fit with a static or functional (dynamic) orthosis; a static orthosis is designed to maintain as functional a position as possible. With patients who have had stroke, there is typically less return of function in the upper extremity, and what motion does return trails the return in the lower extremity. When muscle function begins to return, both dynamic and functional devices are used and continually modified as the muscles slowly regain improved or normal function.

Specialized upper extremity orthotic devices, such as the wrist-driven wrist-hand orthosis (WDWHO), are used for specific levels of quadriplegia (paralysis involving the four extremities). The person with quadriplegia, who has been injured at the C-6 (sixth cervical) level, loses an ability to close the hands. This loss of pinch and grasp is accommodated by use of a wrist driven wrist-hand orthosis. With training from a skilled therapist, the individual with C-6 quadriplegia is able to perform many functions otherwise thought impossible (Bussell, 2000; Gallagher & Desmond, 2007).

### ***Spinal Orthoses***

Spinal orthotic systems are used to decrease pain, provide support, and prevent or reduce unwanted motion in the spine. Simple designs are commonly found in sporting goods and hardware stores where an elastic waist belt with suspenders is used for support during lifting and moving of heavy objects. While soft orthotic devices provide support, rigid devices are designed to prevent or control unwanted motion; they are used after injury or following surgery. For example, after a spinal fusion, spinal orthoses allow the spine to heal or fuse in an optimal position (Seymour, 2002). Spinal braces are designated by the segment of the vertebral spine that needs to be stabilized and supported, such as lumbosacral orthosis (LSO) or thoraco-lumbosacral orthosis (TLSO).

Cervical vertebrae are difficult to stabilize because there is little supportive structure. The human head is quite heavy and rests on top of the spine (the seven cervical vertebrae). The junction between the head and first vertebrae, C-1 (first cervical vertebrae), provides the flexion and extension (up and down tilt) of the head. A junction at C-1 and C-2 provides the rotational capability with C-3 through T-1 (first thoracic vertebrae) contributing to flexion, extension, rotation, and lateral bending of the neck.

Whiplash is a common injury to the cervical spine, often requiring an orthosis. A soft collar is used for comfort and to reduce motion of the head. Cervical orthoses (collars) can be more extensive and rigid to further stabilize the head. With severe injury, or when maximum cervical immobilization is needed, a halo fixation device is used (See Figure 1). Dr. Jacqueline Perry developed the halo fixation device at Rancho Los Amigos National Medical Center in Downey, California, for use on polio patients. In certain cases, the polio virus renders the muscles of the neck so weak that the head cannot be held erect. The halo holds the head in a functional position while protecting the spine until sufficient strength returns to the muscles of the neck and shoulders. Currently, the halo is used to stabilize the cervical region of the spinal cord while skeletal healing occurs (Lusardi & Nielsen, 2000).

### ***Other Assistive Devices***

These devices are as basic as canes, crutches, and walkers and aid in ambulation while an injury is healing or used on a permanent basis for support. Assistive devices include adaptive equipment such as self-help aids; included are devices fabricated to increase a person's daily activities such as dressing and grooming, eating, writing, and operating a computer keyboard. Examples of assistive devices include reachers, long-handled grippers, thickened writing implements, and adapted computer keyboards.

**Figure 1**  
**Halo Fixture**

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Modifications are made to automobiles such as relocating the brake and gas pedals and providing hand controls, more extensive rear-view mirrors, and easy-to-open door handles. Minivans are modified to accommodate the needs of an individual, making it possible for a person with quadriplegia to drive. Extensive modifications and van conversions for almost any kind of disability can be provided.

Orthotists and prosthetists, being well acquainted with manufacturing and fabricating custom devices, are called upon to design and provide special assistive systems (Falvo, 2009). Mouth sticks are used by individuals with high-level paralysis (quadriplegia). The mouth stick is a device with a portion made to fit into the mouth with a rod extending out to a point where a rubber tip is affixed. The person uses this device to push the buttons of a telephone, type on a keyboard, and perform other tasks. Wheelchair seating systems historically were custom made by orthotists until the wheelchair rental and sales companies took on that function. Special equipment is manufactured by certified orthotists that are only limited by the imagination and assessment skills of those involved (Clark, 1993).

There are hundreds of designs for wheelchairs, both power and manual. The orthotist can offer suggestions for modification so the individual can more readily use a wheelchair. The type of power chair is identified through evaluating the person's goals (sensory, cognitive, and anatomical areas of functioning) and level of transfer to get into and out of the chair.

### ***Physical Limitations***

A major purpose of orthotic devices is to improve function and thereby decrease physical limitations. Weak muscles are provided support both in the lower and upper extremities. An individual with leg weakness has increased support with orthotic devices, enabling that person to stand longer and walk greater distances. Adaptive devices for upper extremities support weak and inactive muscles, helping the person perform activities of daily living. Orthotic devices help the employee perform work tasks and can be custom designed for particular work functions, thereby improving vocational potential. Orthoses assist people who want to drive, consequently enhancing independence (Brodwin, Star, & Cardoso, 2007). This may allow an individual the transportation needed to secure and maintain employment. Orthotic devices for upper extremities increase gross and fine manual dexterity, coordination, and manipulation. For lower extremities, they enhance ambulation and overall physical mobility. The goal for assistive devices is to have them help individuals function independently at home, work, and in leisure activities.

## ***Psychological Limitations***

Orthotic devices help minimize functional limitations, thus assisting people to overcome physical limitations; as a consequence, emotional concerns may diminish. Use of orthotic devices helps people obtain and maintain employment; one sees a positive impact on a person's psychological state. Provision of orthotic devices assists with activities of daily living and independence, thus impacting self-esteem and confidence. More positive emotions can replace anxiety and depression, and help in adaptation to disability and loss (Dell Orto & Powers, 2007).

## **Amputation**

Amputation is the general term for loss of all or a portion of a body part. They can be congenital, the result of injury, or a surgical procedure to treat disease. The loss of a limb is devastating to both the person and the family. An amputation causes changes in body image and in functional capacity; much of this relates to a person's interpretation of the loss. Adaptation depends on the circumstances involving the amputation, usefulness of a prosthetic device, and the person's perception of the resulting disability (Carroll & Edelstein, 2006; Falvo, 2009). In cases of traumatic amputation, such as in a vehicular accident or an armed conflict, there is no time for the person to prepare; adjustment and adaptation becomes more difficult than for individuals who have had time to prepare for amputation, such as may occur with systemic illnesses (e.g., cancer, diabetes). A prosthesis can be perceived in a negative way by signifying loss, or in a more positive and productive manner as a means of restoring function, thereby diminishing limitations imposed by the resulting disability.

Statistically in the United States, the majority of amputations are a result of poor arterial circulation caused by complications of diabetes. Diabetes involves an inability of the body to produce sufficient insulin for glucose metabolism. One long term adverse health effect caused by diabetes is a vasculopathy (poor blood circulation to peripheral tissues), especially in the lower extremities, but also to the nerves, retina, heart, and kidneys. As a result, over time, the tissues become malnourished and die. Commonly, the person who has diabetes loses sensation in a lower extremity at the most distal segment first, such as in the toes. Because of a potential unrecognized injury, the person is also prone to infection (Pomeranz, Adler, Shenory, Macaluso, & Parikh, 2006).

Individuals in good control of their diabetes are less likely to have complications and, if they do, to a lesser degree. It is not likely that the person who has diabetes was healthy one day and requires an amputation the next. Diabetes is a chronic condition and a simple accident, such as a stubbed toe may be all that is needed to cause an injury from which the body of a person with diabetes has difficulty healing. Infection may set in and complicate already diminished circulation; the wound becomes more extensive and, in spite of the best medical care, lacks sufficient blood circulation. Infection may become so severe that surgical amputation becomes necessary (Carroll & Edelstein, 2006; Dunn, 1996). If circulation continues to be impaired at a higher level, further surgery may be required.

Historically, prior to the discovery of antibiotics, a person might survive a painful unanesthetized amputation only to die from ensuing infection. Today, with advances in modern surgery, the science of amputation is precise with scientific and clinical testing performed prior to an operation to determine the amputation level most likely to succeed and, at the same time, maintain the best physical functioning. Laboratory tests are performed to determine if preoperative antibiotics need to be used to reduce the risk of infection or the spread of an already existing infection.

## ***Levels of Amputation***

### ***Lower Extremity***

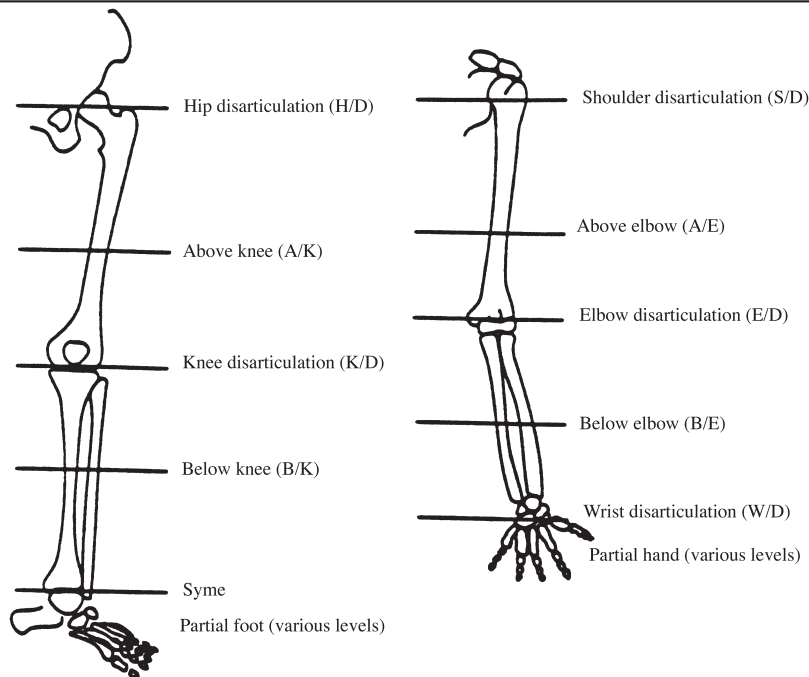
Amputations are classified according to their location relative to the nearest joint (see Figure 2). The lowest level is called a partial foot; as the name implies, it goes through a part of the foot. The next level up is referred to as a Syme and occurs through the ankle joint. Other levels include below the knee (BK), knee disarticulation (KD), above the knee (AK), and hip disarticulation (HD).



### Upper Extremity

Amputations involving the upper extremities are likewise categorized relative to the nearest joint (see Figure 2). The lowest level involves removal of part of the hand or fingers and is called a partial hand amputation. If the amputation occurs at the wrist level, it is referred to as a wrist disarticulation (WD). Other levels include: below the elbow (BE), elbow disarticulation (ED), above the elbow (AE), and shoulder disarticulation (SD).

**Figure 2**  
**Levels of Typical Amputations**



### Complications of Amputation

Complications may occur at the site of a recent amputation. These include edema, ulceration, contracture, infection, and pain (Seymour, 2002). Normally, a certain amount of edema (swelling of tissues) of the stump occurs following surgery; it has been found that immediate or early fitting of a prosthesis minimizes this type of edema. If abnormal or excessive edema occurs, immediate medical attention is necessary; otherwise, ulceration of the skin can occur. Skin ulcerations also are caused by infection, a poor fitting prosthesis, or excessive pressure where the prosthesis contacts the skin. Medical attention and a new prosthetic socket with an appropriate fit are essential in the remediation of this problem. Hot, humid environments can lead to breakdown of the skin; dirt and lack of cleanliness in the area can cause a serious problem and can lead to potential infection of the amputation area.

Muscular contractures can occur and are caused by lack of use of the extremity either before the limb was surgically removed or after removal when the prosthesis was not used. They occur because of limited range of motion or disuse of a joint; exercise or surgery usually resolves this problem. Lack of cleanliness can result in an ulcerative area, leading to infection. Treatment involves the use of antibiotics and possible surgical excision of a deeply infected area (Griffiths, 1995; Seymour, 2002).

The sensation of phantom-limb is a feeling following amputation that the amputated limb still is present. Phantom-limb sensation is a normal occurrence for most people who have had amputation; it is neither painful nor limiting. A sensation of pain in a removed part is abnormal and known as phantom-limb pain. Unlike phantom-limb sensation, phantom-limb pain can be disabling, and cannot always be relieved

even through surgery (Bower, 1995). Phantom-limb pain, if severe, may disable a person from most activities, including work. If a person experiences chronic pain, referral to a medical specialist is appropriate. There are surgical and non-surgical procedures that help alleviate chronic pain. Fortunately, there is a tendency for chronic phantom-limb pain to diminish over time (Carroll & Edelstein, 2006).

Physical implications of amputation are dependent both on the loss itself and on the person's adaptation to that loss. The higher the level, the more difficult and challenging the adjustment; functional loss is greater and fitting of a prosthesis is more formidable. A lower extremity prosthesis enables a person to stand and walk. As a result of increasing ability to ambulate, an individual is capable of more extensive work than just seated activity. Walking on uneven surfaces, rough terrain, and inclines/declines as well as pushing, pulling, and climbing are difficult with a lower extremity prosthesis. An individual with two lower extremity prostheses has a more difficult time walking and standing and may use a wheelchair. Many persons become proficient with two lower extremity prostheses and are able to ambulate with use of a cane or other assistive devices. Light or sedentary work not involving significant amounts of standing and walking is appropriate for a worker with bilateral lower extremity limb loss.

An upper extremity prosthesis increases a person's manual dexterity, bilateral dexterity, and allows for more complex tasks requiring both hands. Some people become quite proficient when using a prosthetic upper extremity. A hook terminal device allows for grasping, gripping, lifting, and carrying. Terminal devices can be specially designed and fabricated to meet certain needs within the workplace. Specialized devices may be fabricated and used solely for particular job functions.

### *Psychological Considerations*

The emotional impact of amputation of any body part is devastating; the person may experience anger, depression, resentment, fear, and withdrawal. People with amputation need to make permanent behavioral, social, and emotional adjustments to cope with the various problems presented with loss of a limb (Falvo, 2009). Yet, the human mind is flexible and most individuals learn, after a period of grieving over loss, to cope and adapt. After struggling with the loss for months, the person eventually comes to the realization that many activities, including work, still are possible. Early fitting of a prosthesis hastens adjustment; prompt intervention by a rehabilitation counselor makes the adjustment and adaptation less traumatic, and the focus is shifted from loss to the possibility of returning to work. A prosthesis increases capabilities and helps minimize limitations, thereby making emotional adjustment easier; it provides a feeling of "wholeness." Regardless of the reason for the amputation, above all, it is important to evaluate the individual's interpretation of the loss.

There are a plethora of emotional effects associated with amputation, and they are founded on factual occurrence and involve cultural perceptions. There is a naturally occurring grieving process involving loss of a part of the body, similar to the "stages of death and dying" identified by Dr. Elizabeth Kübler-Ross (1969). These stages include: (a) denial and isolation, (b) anger (rage, envy, and resentment), (c) bargaining, (d) depression, and finally, (e) acceptance or adaptation. Not all people go through these stages in the order listed, nor does everyone experience each stage. Some people never reach the final stage. Feelings of hope often persist through all these stages and are a positive indication of emotional recovery. A gradual transition takes place from the first four negative stages to the final positive stage – acceptance or adaptation. Time is needed to resolve feelings of loss and adaptation to a different body image.

An individual who is presented with a recommendation for amputation after months of possible loss of mobility, infection, pain, and failed medical treatment may agree that an amputation will improve the current quality of life. Quite differently, the person who is diagnosed with osteosarcoma (bone cancer) and requires immediate amputation has little time to begin the emotional adjustment process to the impending loss. A person who loses a limb in an accident likewise has no time to adjust. These individuals may need additional time to grieve and adjust to these traumatic losses (Carroll & Edelstein, 2006). Shontz (2007) noted several principles relating to disability and psychological adjustment. The counselor might keep these principles in mind when working with clients who have amputations.

1. Psychological reactions to the onset or imposition of physical disability are not uniformly disturbing or distressing and do not necessarily result in maladjustment.

2. Reactions (both favorable and unfavorable) to disabilities are not related in a simple way to the physical properties of the disabilities.
3. Environmental factors (e.g., architectural barriers, attitudes) are at least as important in determining emotional reactions to disabilities as are the internal states of people who have the disabilities.
4. Of all factors that affect the total life situation of an individual with a disability, the disability itself is only one aspect, and often its influence is relatively minor.

### ***Family Involvement***

Family counseling usually is made available so that each member of the family understands how best to fill the role of loved one and learn to help with the process of adaptation to a newly acquired disability and the process of rehabilitation. The family can help the person with the grieving process by assisting in adaptation to loss and changed body image. Capability of the family to provide emotional support is invaluable (Yetzer, Kaufmann, Sopp, & Talley, 1994). Yet, one must be aware that excessive attention can be dysfunctional to the person's full recovery. Psychological health improves as one adjusts and again begins to enjoy activities and social relationships.

### ***Employment Issues***

#### ***Lower Extremity***

Physical demands of work (walking, standing, lifting, and carrying) may need modification. For an employee with a lower extremity amputation, use of a wheeled cart for transporting material, a backpack for smaller items, powered carts or scooters for greater mobility, and minimizing the need for reaching up, reaching behind, bending down, and squatting is helpful both because these actions are difficult to perform with a prosthesis and involve energy expenditure (Brodwin, Parker, & De La Garza, 2003). Overall amounts of standing and walking need to be decreased; ambulation with a prosthesis requires substantially greater consumption of energy. Climbing, squatting, and kneeling are difficult or impossible to perform (Clark, 1993; Mueller, 1990).

#### ***Upper Extremity***

With an upper extremity amputation, even with use of a prosthesis, the person will lose a significant amount of manual dexterity. Because of the absence of feeling and sensation in the prosthetic device, function is further limited. A prosthetic hook only partially substitutes for the wide variety of movements and functions of a normal hand. At work, use of an upper extremity prosthesis increases manual dexterity, bilateral dexterity, eye-hand coordination, gripping, grasping, lifting, and carrying. Although a prosthesis does not replace a fully functional arm and hand, it allows a worker to perform many essential tasks. A workstation, with appropriate accommodation and modification, can be designed to make possible and efficient the work duties of a person with an upper extremity amputation.

#### ***Workplace Accommodation***

If a person has chronic pain (i.e., phantom-limb pain) but is able to work, workplace accommodation is beneficial. Employers who understand chronic pain are more willing to consider possible accommodations. Workplace accommodation for chronic pain is as variable as chronic pain itself. Each person with chronic pain needs individualized attention and case management (Brodwin et al., 2003).

Through careful evaluation of needs, limitations, strengths, and an awareness of functioning and the kinds of accommodations and assistive devices available, rehabilitation counselors can enhance the employability of people who have undergone amputation. Counselors should always remember the psychological trauma that has occurred and its impact on the total functioning of the individual within the home environment, at the workplace, and socially.

# Prosthetics

## *Prosthetic Treatment*

Immediate post-operative fitting of a lower limb prosthesis hastens the healing process. Emotional benefits of this procedure are well documented (Gallagher & Desmond, 2007; Lim, 1997). An immediate postoperative prosthesis (IPOP) is arranged by the surgeon or treating physician. It is a sterile, rigid dressing applied in the operating room immediately after surgery while the patient is still under anesthetic. A soft, sterile dressing is applied, followed by placing a rigid Plaster of Paris cast over the leg; a prosthetic pylon and foot are then added. An IPOP (Esquenazi, 1994; Wilson, 1998):

- provides a sterile environment for healing
- prevents flexion contractures
- protects the limb from external trauma
- allows partial weight-bearing (never greater than 30 pounds)
- maintains proprioception and a kinesthetic position of the limb
- accelerates healing by preventing edema and assisting in venous blood return
- gives a psychological perception and feeling of wholeness

### *Temporary Prostheses*

Generally speaking, after the surgical site has healed the patient is sent to the prosthetist to have a temporary prosthesis fabricated. The individual should have been seen by a licensed physical therapist with which the prosthetist and physician will consult to determine functional levels (see “Medicare Functional Levels” later in this chapter).

Upon completion of a temporary prosthesis, the patient is ready for gait training. A physical therapist begins the training process. Weight bearing and weight shifting capabilities precede attempts at ambulation. A person with a below knee amputation may acquire all the skills needed for ambulation within six weeks. Yet, a person may need six months for the limb and skin to toughen to the point that the tissues and bones tolerate the forces involved with ambulation.

Adjustments are routinely made by the prosthetist during the months in the temporary prosthesis. Time needed for the limb to mature is from four to eight months for below knee amputations and four to 18 months for those above the knee. The process of maturation includes reduction caused by muscle atrophy, decreased edema, and loss of adipose content. Each individual has a somewhat different adjustment period (Carroll & Edelstein, 2006).

### *Permanent or Definitive Prostheses*

Fitting with a permanent prosthesis is generally begun when the patient’s limb has matured and the majority of tissue volume reduction has occurred. It may be initiated when the prosthetist has exhausted all adjustments to the temporary socket and there is a need to replace the socket or upgrade the component parts. Sophisticated components may be selected for the permanent prosthesis, as appropriate, to increase function and longevity.

### *Factors Considered for Permanent Prostheses*

The prosthetist takes the following factors into account when fitting and designing a permanent prosthesis (Bussell, 2000; Carroll & Edelstein, 2006):

- types of terrain commonly traversed
- velocity of ambulation
- variance in ambulation velocity
- styles of shoes worn

- physical activities, including work
- body weight

### ***Purchase of Prosthesis***

Based on these factors, the prosthetist and patient select a group of components to be used in fabrication of the prosthesis. Most component manufacturers want their components to be used and allow them to be tried in the prosthetist's office, helping ensure the selection that best fits the person's needs (Lim, 1997). Reimbursement issues require consideration. Third-party payers are increasingly dictating levels of reimbursement and hence the sophistication of prosthetic systems being provided. The individual needs an awareness of what is available and necessary for desired functions. Except for Medicaid recipients, persons with limb loss have the legal right to purchase components or technology used in the prosthesis that may not be covered by insurance, by paying the difference in cost.

### ***Lower Extremity Prosthetics***

Components of a lower limb prosthesis include the limb interface, socket, mechanical joints, and foot assembly (Bussell, 2000). An individual with a new amputation will have several changes in limb volume (due to post-amputation swelling and changes in weight) and will require periodic adjustment of the socket. A tight, yet intimate fit is needed for maximum benefit and use. The person must learn appropriate gait through instruction and practice. Stair climbing and walking on uneven surfaces requires additional practice. Driving is accomplished with adaptive equipment (hand controls), if necessary. Most sporting activities can be done with a special prosthetic limb.

Activities are easier to perform with an amputation below-the-knee as only the ankle joint is missing. An individual with a BK amputation generally expends about 10% to 37% more energy for walking (Riley, 2005; Wilson, 1998). With an above-the-knee amputation, both the ankle and knee joints are artificial making standing and walking more challenging and energy consuming. Since a person with an AK amputation expends 60% more energy walking, some may choose to use wheelchairs for ambulation to save energy for work and other activities (Seymour, 2002; Wilson, 1998). The less excess standing and walking a job involves, the more energy the worker retains for other job functions.

Advanced prosthetics technology has led to development of lighter, more comfortably fitting, and technologically efficient lower extremity prostheses. Research is progressing to allow direct nervous system control of the prosthesis. The C-leg system is "the world's first completely microprocessor-controlled prosthetic knee/shin system with hydraulic swing and stance phase control." This product is so revolutionary that amputees who have been fitted with the device often state that its most obvious benefit is that patients do not have to think consciously about walking. This occurs because "the advanced microprocessor control does the thinking for them." (Holmes, 2005, p. 789).

### ***Upper Extremity Prosthetics***

Components of an upper limb prosthesis include the limb interface, socket, mechanical joints, terminal device, and suspension system. A harness system using the shoulder muscles on the opposite side allows the person to operate a body powered terminal device (hook). Normal functioning of a hand and arm cannot be replaced with a prosthetic device. Movements made by the hand are intricate; the hand has an ability to move in various directions and can perform complex motions and tasks, from gross manipulation to fine dexterity. The terminal device of a prosthetic upper extremity is a hook-type device that can grasp, pinch, and hold objects. It cannot feel, do fine dexterity, or manipulate; lifting and carrying are limited to lighter weights.

Functional or cosmetic hands can be used instead of hook devices. The functional hand has a three-finger pinch, grasp, and hold action, but lacks the power and stability of a hook. Cosmetic hands are used primarily for social purposes, as they resemble a natural hand in color, tones, and texture. A more sophisticated device is the external power myoelectric arm and hand. The myoelectric arm is computerized and contains electrodes that are placed on the skin over muscles. Electrical impulses from the muscles allow the person to open and close the hand simply by tensing the particular muscles. Although the myoelectric



arm has the appearance of an actual arm and hand, it is not usually prescribed due to the expense (Carroll & Edelstein, 2006).

### ***Medicare Functional Levels***

Medicare has established recognized definitions used to express functional levels of lower extremity amputation. The five levels are described below.

- Level 0 - Patient does not have the ability or potential to benefit from a prosthesis.
- Level I - Person has the ability or potential to use a prosthesis for transfers or ambulation on level surfaces at a fixed cadence. This level is typical of the limited and unlimited household ambulator.
- Level II - Individual has the ability or potential for ambulation with the ability to traverse low-level environmental barriers such as curbs, stairs, and uneven surfaces. Level II is appropriate for the limited community ambulator.
- Level III - Patient has the ability or potential for ambulation with variable cadence. This level is used for the community ambulator who has the capability to traverse most environmental barriers and may have vocational, therapeutic, and exercise activity that demands prosthetic utilization beyond simple ambulation.
- Level IV - Person has the ability or potential for prosthetic ambulation that exceeds basic ambulation skills, exhibiting high impact, stress, or energy levels. Level IV is appropriate for the prosthetic demands of a child, active adult, or athlete.

According to Medicare, the reason for these levels is to set a standard for which types of component parts are appropriate, based on the individual's projected needs.

## **Conclusion – Assistive Technology and the Workplace**

Assistive technology (AT) is defined in the Technology-Related Assistance for Individuals with Disabilities Act of 1988 (popularly referred to as “The Tech Act”) as “any item, piece of equipment, or product system, whether acquired commercially off the shelf, modified, or customized, that is used to increase, maintain, or improve functional capabilities of individuals with disabilities.” The goal of AT is to increase the functional independence for people who have disabilities. It is having a profound impact on improving the lives and employment opportunities of many individuals. AT is an effective and powerful device to help restore functioning, enhance quality of life, and remove physical and attitudinal barriers. Rehabilitation counselors are attempting to maximize this independence at the workplace (Brodwin et al., 2003). Through the use of orthotics and prosthetics, AT enables workers with upper or lower extremity limitations to reduce and perhaps minimize their functional limitations.

Through prosthetic and orthotic devices, rehabilitation counselors have seen their clients achieve greater productivity within education and employment, leading to enhanced self-sufficiency, independence, freedom to choose, and social re-integration. Many workers can continue working, return to work, or secure alternative employment following a disabling event; also, they are able to participate more fully and actively in all aspects of life with the provision of orthotic and prosthetic devices. Technology helps equalize the capacities of workers with disabilities when compared to workers who do not have limitations. Greater independence and satisfaction in daily life, including both work and leisure, become enhanced (Carroll & Edelstein, 2006). With daily use of orthotic and prosthetic devices, a person's emotional outlook is improved, as is the ability to interact more readily and comfortably at work and within the social arena (Brodwin et al., 2007).

## **Case Study**

Amy Nguyen, a Vietnamese-born 22-year old, has been residing in the United States for the previous 10 years; she has a Bachelor of Arts degree from a local university. At the present time, Amy is living with her parents in a large city. Several months ago, she was in an automobile versus bicycle accident. A serious right leg injury resulted in an AK amputation (Riley, 2005). Understandably, Amy has had great

difficulty accepting the loss of her leg, although recently she has asked to work with her treating physician and prosthetist for fitting of a prosthesis. Currently, she experiences “phantom-limb” and a moderate amount of “phantom-limb pain” (Bower, 1995).

Having recently graduated from college with a degree in liberal arts, Ms. Nguyen would like to become an elementary school teacher. She needs to attend one more year of college, including student teaching and passing a teaching certification examination. Her parents strongly suggested that she return to college and major in business accounting. At this time, Amy strongly believes she must follow her parents’ advise, even though she does not agree.

Amy’s only employment has been on a temporary or part-time basis. She has worked as a sales person during several Christmas seasons, a cashier and hostess in a restaurant, and a part-time cashier/ticket taker for a movie theater; these jobs took place during several summer and Christmas vacations and occasionally during the school year.

### **Questions**

1. Discuss Dr. Elizabeth Kübler-Ross’ stages of death and dying from the standpoint of adjustment to disability. Relate this discussion to the case study and the role of the rehabilitation counselor.
2. Describe the various levels of amputation, the functional limitations at each level, and psychosocial implications. Relate these limitations to school, work, and social activities.
3. What are Ms. Nguyen’s functional limitations as related to future employment? Will she be able to drive an automobile? Discuss the psychosocial factors relevant to this case.
4. How would you handle her parents’ strong guidance that Amy return to college and major in business accounting rather than becoming an elementary school teacher and Amy’s desire to fulfill her parents’ wishes? From a cultural perspective, how would the two opposing vocational goals affect this client and the provision of vocational rehabilitation services?
5. Amy is having great difficulty with adjustment to her disability. She is also experiencing “phantom pain.” As her counselor, offer recommendations in regard to these two conditions. Discuss “phantom-limb sensation” and phantom-limb pain.”

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## About the Author

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## Chapter 30

# PLASTIC AND RECONSTRUCTIVE SURGERY

*Neil E. Klein, M.D.*

## Introduction

“We bring back, refashion, and restore to wholeness the features that nature gave but chance destroyed, not that they may be an advantage to the living soul, not as a mean artifice but as an alleviation of illness, not as becomes charlatans but as becomes good physicians and followers of the great Hippocrates. For though the original beauty is indeed restored . . . the end for which the physician is working is that the features should fulfill their offices according to nature’s decree” (Gaspare Tagliacozzi, 1597).

This chapter examines different aspects of plastic and reconstructive surgery through in-depth coverage on wound healing, healing intentions, conditions required for wound healing, functional limitations, and rehabilitation potential. It is estimated that more than one million reconstructive procedures are performed in the United States by plastic surgeons every year. Reconstructive surgery helps patients of all ages and types - whether it is a child with a birth defect, a young adult injured in an accident, or an older adult with a problem caused by aging (Dolan, 2004).

The goals of reconstructive surgery differ from those of cosmetic surgery. Reconstructive surgery is performed on abnormal structures of the body, caused by birth defects (congenital deformities such as birthmarks, cleft-lip and palate deformities, hand deformities such as syndactyly [webbed fingers], or extra or absent fingers, and abnormal breast development), and developmental abnormalities acquired as a result of trauma or injury, infection, tumors, disease, and in some cases, aging. Burn wounds, lacerations, growths, and aging problems are considered acquired deformities. Reconstructive surgery is generally performed to improve function, but may also be done to approximate a normal appearance.

Primary goals of plastic and reconstructive surgery are to restore function and appearance (McCauley, 2005). Clearly, medical and surgical restoration leads to a decrease in functional limitations and thus improves rehabilitation potential. Plastic and reconstructive surgery helps minimize disfigurement and assist the person in adjustment to disability and interacting with the public.

In some cases, patients may find that a procedure commonly thought to be aesthetic in nature may be performed to achieve a reconstructive goal. For example, some older adults with redundant or drooping eyelid skin blocking their field of vision might have eyelid surgery. Another example is an adult whose face has an asymmetrical look because of paralysis might have a balancing facelift. Although appearance is enhanced, the main goal of the surgery is to restore function. Large, sagging breasts are one example of a deformity that develops as a result of genetics, hormonal changes, aging, and disease. Breast reduction (reduction mammoplasty) is the reconstructive procedure designed to give a woman smaller, more comfortable breasts in proportion with the rest of her body. In another case, a young child might have reconstructive otoplasty (outer ear surgery) to correct overly large or deformed ears.

Plastic surgery, whether cosmetic or reconstructive, encourages and promotes a strong, positive self-image. Even a small change on the outside can create an extraordinary change on the inside, allowing an individual’s self-confidence to flourish (Dell Orto & Powers, 2007).

In the specialty of plastic and reconstructive surgery, the word plastic refers not to the use of artificial materials for surgery, but to the Greek word “plastikos” meaning to mold or shape. The plastic



surgeon uses knowledge of the wound healing process to correct deformities and aid function to overcome disabilities. To evaluate the disability properly, it is essential to understand plastic and reconstructive surgery, as this surgical subspecialty is co-involved with wound healing. To know how a wound heals is to know, in part, how the patient will recover and the potential for return to employment.

Of major concern, along with the surgical aspect, is the rehabilitation of the patient to enhance function and minimize limitations. The purpose of surgery is not only to enhance healing, but to reestablish the patient's function and minimize limitations. For this reason, plastic and reconstructive surgeons work closely with psychologists, physical and occupational therapists, speech therapists, and rehabilitation nurses involving a multidisciplinary team approach. The goal of this team is to provide maximum function for the patient.

## **Wound Healing and Healing Intentions**

Wound healing is the way the body repairs itself following a physical insult. The process of wound healing may be considered a series of separate events. The healing environment may be manipulated to enhance or accelerate the healing process (Romo & Pearson, 2005). However, only the body can heal the wound; all a surgeon can do is remove impediments to healing. These impediments include infection, dead tissue, poor vascular (blood) supply to the injured tissues, or a general health problem of the patient that is preventing wound healing, such as a metabolic or immune disease or severe malnutrition. To help heal the wound, one must recognize the problem, correct it, and allow the body to heal itself (Thorne, 2007).

When faced with a simple traumatic wound, the physician (the terms physician and surgeon are used interchangeably throughout this chapter) treats it through cleansing and closure. In more complex or long-standing wounds, dead or severely damaged tissue must be removed by debridement (cleaning) processes. Infection is treated with antibiotics, either systemically (by mouth or injection) or topically (applying antibiotic directly to the tissues). Tumors must be surgically removed or treated. Once these procedures are complete, the plastic and reconstructive surgeon treats the resulting defect or wound.

### ***Types of Wounds***

Wounds are classified by how they will heal. A simple laceration or cut is sutured directly in a straight line and heals by **primary intention**. Intervention is not needed for the body to heal primary intention wounds. A wound that is missing tissue so that the skin edges of the wound cannot be brought together (such as the wound created by removal of a tumor, a significant burn, or a gouging type injury) heals by **secondary intention**. In other cases, when the wound defect created by the injury is too large to heal on its own, the plastic and reconstructive surgeon needs to transfer tissue from another part of the body to cover the wound.

The understanding of the mechanism of wound healing has increased dramatically during last few years. Today wound healing abnormalities are among the greatest causes of disability and deformity (Broughton, Janis, & Attinger, 2006).

### ***Scar and Epithelialization***

#### ***Scar***

The actual material for healing a wound is scar tissue – a specialized protein that pours into a wound to hold the wound together shortly following injury (Dolan, 2004). For example, in the process of primary intention healing of a simple cut in the skin (Figure 1), the physician first cleanses the wound. This cleansing process is crucial to healing as it minimizes chances of infection by mechanical removal of contaminants and infectious agents. Next, the physician brings together the edges of the wound, either by sutures, or by simple bandage if the wound is small. Surrounding tissue will start to attract fibroblasts – the cells that make collagen (Evans, 2000).

Collagen, a glue-like protein, is the chemical of scar. Figure 1 illustrates a sharply cut wound through the skin into the fat layer. In the second part of the illustration, the wound has been sutured and scar tissue is beginning to strengthen the bonding of the wound edges. The uppermost layer of skin heals by epithelialization (production of epithelial cells) – a regenerative process.

**Figure 1**  
**Healing By Primary Intention**



\*\*Figures 1-7 drawn by Ms. Crista Osterberg

During the early days following a wound, the collagen is soft and weak; young wounds may fall apart or pull apart if the physician removes the sutures too early. Gradually, the collagen toughens by creating more chemical bonding within itself and the scar tissue becomes stronger; this process is called scar maturation. Full maturation of a scar takes from six months to two years, depending on a person's physiology. During this period, the scar may be red, tender, pruritic (itchy), swollen, and unsightly. As the scar matures and toughens, metabolism within it decreases. There is less itching and swelling, the blood supply lessens, and the redness diminishes. Upon complete maturation, an ideal scar will be a fine line difficult to detect.

**Contraction** is a normal occurrence of scar maturation. All scars contract as they strengthen; this is a natural part of the healing process. However, scars do not always heal in a fine line. Some people have a physiological make-up that tends to produce unattractive or thickened scars known as hypertrophic scars or, in more severe forms, keloids. These may be aesthetically undesirable, painful, and pruritic. Also, a scar in a bad location, such as across a joint, may tighten and create a contracture (tightening or shrinkage) impairing motion in that joint.

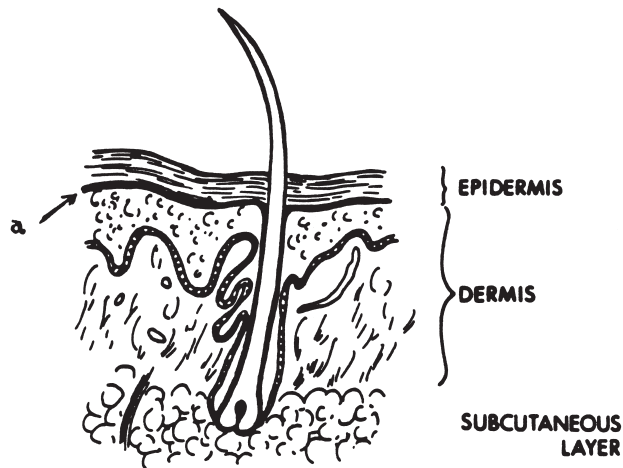
**Contracture**, however, is a pathological problem that occurs when normal scar contraction takes place in an unfavorable location. For example, the tissues around a joint may shorten and prevent full range of motion of that joint. Scar contracture is a common effect of burns involving joints. Scar contractures often need to be treated by therapy or surgical release.

### ***Epithelialization***

Epithelialization is an important aspect of wound healing. This process occurs whenever a wound breaks the skin (in both primary and secondary intention healing). It takes place concurrently with the early phases of scar formation during healing by primary intention, and is the process by which the top layer of skin heals itself. The process begins within hours of tissue injury. The uppermost layer of skin can heal itself by this regenerative process, as opposed to deeper layers that must be repaired by the body's scarring. While the deeper layers of the skin are laying down scar, germinal or formative cells are making new skin cells. These new cells migrate across the top of the healing wound until the approaching layers from both sides touch and close the wound. This is the process of epithelialization (Romo & Pearson, 2005; Thorne, 2006).

The germinal epithelial cells are located in the basal layer of skin and within all dermal elements of skin such as hair follicles, sebaceous tissue, and oil glands (Figure 2). In a partial thickness or 2<sup>nd</sup> degree burn or scrape where some deeper skin dermal elements are left behind, the skin regenerates or reepithelializes

Figure 2  
Normal Skin

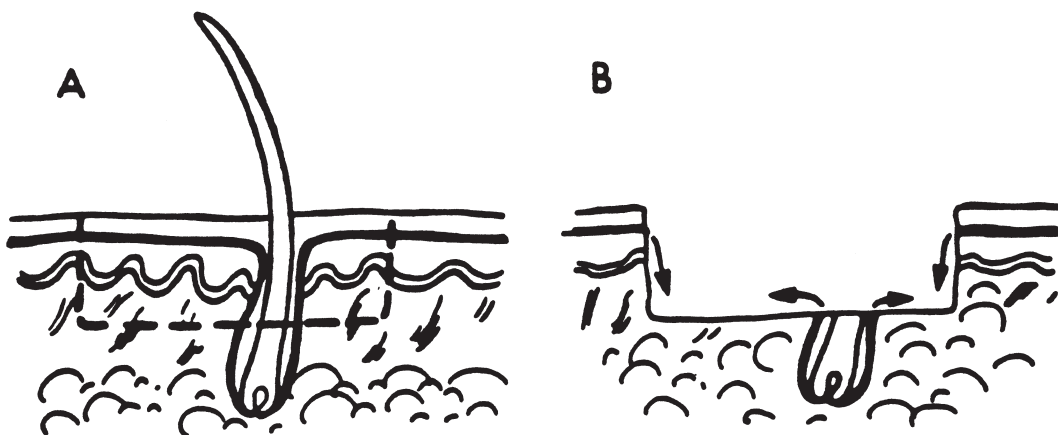


Note: Basal layer (a) lining skin and dermal elements.

without the need for surgical closure (Figure 3). In Figure 2, normal skin is depicted. The black line designated “a” is the location of the basal cell layer or regenerative layer. This layer, as previously noted, lines the skin and all the hair follicles and dermal elements.

A physician determines whether a wound or burn is full thickness (3<sup>rd</sup> degree) or partial thickness (2<sup>nd</sup> degree) by examining how much of the basal layer is left behind (see Table 1). Visualize a scraping wound of the skin that takes away the top layers of skin down to the deep dermis or fat. Such a wound is seen in Figure 3-A. The epithelial cells will grow from the basal layer and remaining hair follicles (Figure 3-B) grow to cover the open areas undergoing epithelialization. All skin in mammals has hair follicles or dermal elements, which can provide epithelial cells for partial thickness injuries to heal by epithelialization. The more superficial the wound, the more dermal elements remain to aid the healing process. A superficial wound (partial thickness) epithelializes much quicker than a deep cut. A very superficial wound, such as a

Figure 3  
Epithelialization



Note: Epithelial cells grow from basal layers of adjacent healthy skin and dermal elements.

**Table 1**  
**Classification of Burns**

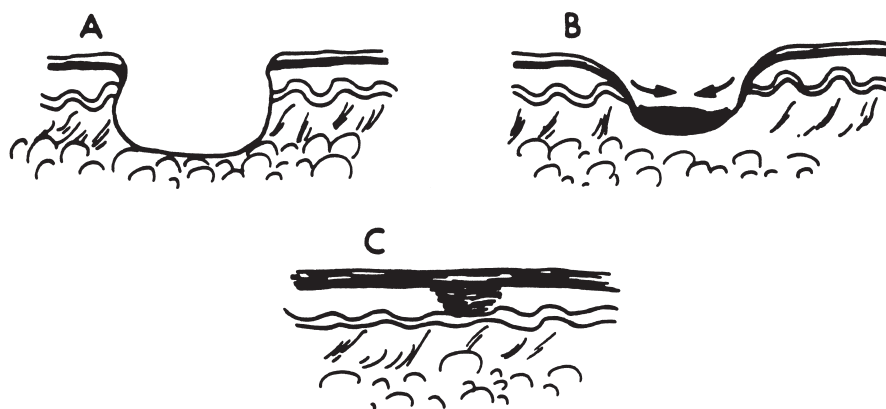
<b>Superficial</b>	Epidermis (uppermost layer) only.	1st degree burn
<b>Partial-Thickness</b>	Epidermis and dermis (inner portion of the skin).	2nd degree burn
<b>Full-Thickness</b>	Destruction of the entire skin, epidermis, and dermis.	3rd degree burn
<b>4th degree burn</b>	Extends deeply into subcutaneous fat, muscle, bone, or deeper tissue.	

minor scrape or “skinned knee,” for example, epithelializes so quickly that there is no time for scar formation. Scrapes or superficial partial thickness (2<sup>nd</sup> degree) burns heal with minimal or possibly no visible scar present.

Healing by secondary intention occurs in wounds with full thickness skin loss into the subcutaneous fat or deeper layers when no dermal elements or hair follicles are left behind (Figure 4). In this situation, the scar develops similarly to what occurs during primary intention, but as the sides of the wound are not together, the open wound edges cannot stick to themselves. The scar in this open wound bed will now contract as normal scar does; the contraction will continue until the wound edges touch. In general, a normal rate of closure is about 10%-15% wound reduction per week (Attinger et al., 2006); a slower rate of healing may lead the surgeon to reevaluate the strategy being used and perhaps consider a different modality of treatment. Interventions such as hyperbaric oxygen, negative pressure, vacuum-assisted closure, and platelet derived growth are experimental but aid the healing process. According to Janis and Attinger (2006), to date even the most advanced technologies are only supplemental to good basic wound care, such as debridement and redressing.

Epithelialization occurs while simultaneously the bed of scar is covered with a new top layer of skin. The epithelium, however, grows only from the edges since there are no dermal elements left in a full thickness wound (all structures of the skin have been destroyed or removed). Examples of this type of wound are the full thickness skin loss of 3<sup>rd</sup> degree burns and very deep gouging wounds. Due to the extensive destruction of tissue and the slow nature of scar contraction, healing by secondary intention takes longer than primary intention healing. When epithelialization takes place from the wound edges only, the healing

**Figure 4**  
**Healing By Secondary Intention**



Note: Contraction of the wound

process takes longer than if dermal elements are present within the wound, such as in a partial thickness injury.

If the wound is too large to heal by itself or if there is a physical impediment to healing such as bone in the wound, healing by secondary intention cannot occur and reconstructive surgery is required for proper healing. This type of wound is said to have a defect because of a deficiency or lack of tissue. The plastic and reconstructive surgeon identifies the actual tissue deficit and decides how to close the defect by replacing the appropriate tissue (Mathes, 2006; Mathes & Nahai, 1997).

### ***Surgical Closure of Wounds***

The first step in closing a wound is to clean the area and determine whether the patient is in generally good health. Infection, if present, must be treated and dead tissue must be debrided (cleaned out). Also, the patient must be metabolically capable of wound healing. When these factors are assured, the surgeon assesses the wound for repair. If the wound is simple and small, the surgeon closes it directly. If it is more complex, the surgeon determines the simplest, most effective means of wound closure. He/she evaluates what tissue is missing and to what extent, and then decides on the method of wound closure. In the clinical practice of reconstructive surgery, a problem one routinely encounters is skin and soft tissue defects, which require coverage. Coverage of such wounds requires primary/secondary closure, skin grafting, or flaps (Subramania et al., 2005).

#### ***Grafts***

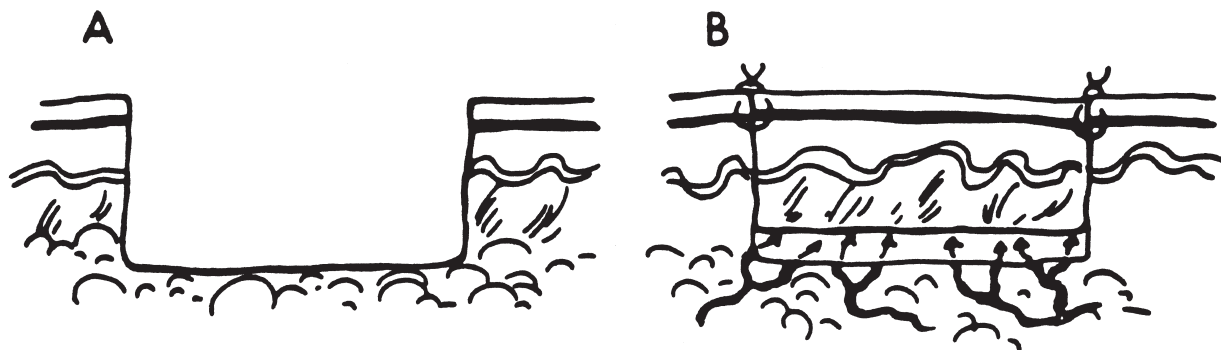
If a small amount of tissue is missing, grafting can be performed. A graft involves tissue that is transplanted or implanted in a part of the body to repair a defect. The surgeon takes a graft from an area of the body known as the donor site and transfers it to the deficient area known as the recipient site. Only tissue from one's own body may be used, except in particular cases, such as heart and kidney transplants. Many kinds of tissue may be grafted, the most common being skin (Platt, McKiernan, & McLean, 1996). Skin grafting is often performed to cover a wound such as that from a burn site. Other tissues commonly grafted are bone, tendon, and nerve.

A graft is limited in its size or thickness because its blood is supplied by the recipient site until new blood vessels grow into the graft to nourish it (Figure 5). Figure 5-A shows a wound defect requiring a graft. Note the full thickness wound with no residual dermal elements. In this case, the physician chooses a skin graft because the wound is too large to wait for healing by secondary intention. Figure 5-B shows the skin graft in place and the blood supply going into the donor skin from the recipient bed. Note that in the illustration, the skin graft is a thin shave of skin consisting only of dermis and above layers. A skin graft is

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**Figure 5**  
**Skin Grafting**

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Note: (A) Defect to be grafted; (B) Blood vessels from the defect supply the skin graft.

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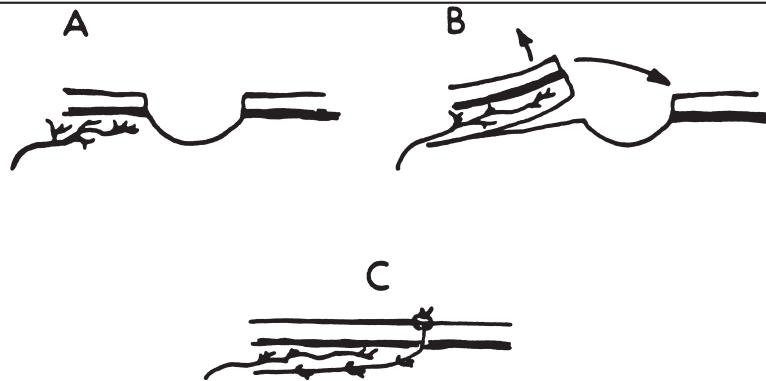
very thin, on the order of 10-15 thousandths of an inch; a bone graft is merely chips of bone wherein cells grow and form a solid structural support. Full thickness skin grafting or composite tissue grafting is done only in certain circumstances and requires special techniques to be successful.

Skin grafts normally require about one week to ensure growth of the blood supply from the recipient bed into the new skin. The donor site, from which the thin layers of skin were shared, may take as long as two to three weeks to reepithelialize (to regroup the skin layer).

### **Flaps**

If the physician needs larger quantities of donor tissue, or tissue with its own blood supply, a flap is used. Since a flap has its own blood supply, thickness is not a problem; yet, the surgery is more complex. A flap must be cut from tissue near the wound and rotated or transposed into the defect (Figure 6). Figure 6-A illustrates a defect with its nearby blood supply shown as the arborized line coming in from the left. The flap is the tissue adjacent to the defect, which is being freed from its bed with its own blood supply intact (Figure 6-B). Once this tissue is elevated and the surgeon is sure it is alive, healthy, and able to move easily into the defect, the flap is transferred, sutured over the defect (Figure 6-C), and then allowed to heal in its new location.

**Figure 6**  
**Flaps**



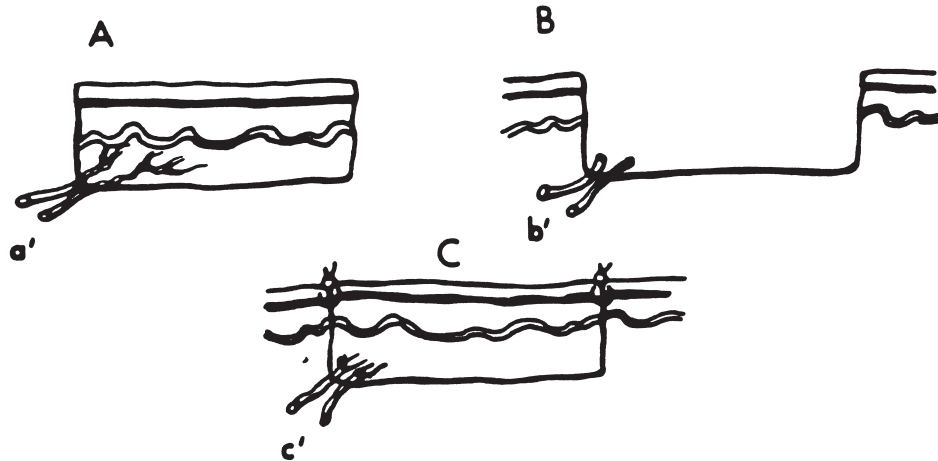
Note: (A) flap carries its own blood supply.

If for any reason this procedure cannot be performed (e.g., the local tissue is too scarred to use), the physician must resort to a very complex microsurgical technique called a free flap procedure (Figure 7). In free flap surgery, the surgeon proceeds only when the following criteria are met:

1. The donor site can spare the tissue.
2. The artery bringing blood into the tissue and the vein draining the tissue can be located.
3. The physician can identify an artery and vein of appropriate size near the recipient site.

After the criteria are met, the surgeon attaches blood vessels within the free flap to the recipient vessels using microsurgery (Figure 7-C). This donor tissue is used for the reconstruction, with the assurance that the donor tissue will survive at the new site. While a simple wound has sutures in place from two days to two weeks, complex grafts and flaps may require several days to weeks of hospitalization. Healing time for such procedures is dependent on location, size of defect, and type of tissue replaced. Medical complications can occur at any time, but become less likely as time passes. When the wound is strong enough to allow for use of the injured part, a rehabilitation counselor may begin intervention (Brodwin, Parker, & DeLaGarza, 2003).

Figure 7  
Free Flaps



- (A) Free flap taken from distant donor site with its blood supply (a' – the supplying artery and the draining vein)
- (B) Recipient site with recipient artery and vein (b')
- (C) Anastomosis (connecting of hollow tubes or vessels) of artery and vein with flap sutured in place (c')

## Conditions Required for Wound Healing

### *Burns*

A burn can be devastating in its effect. Emotional, physical, and socio-economic ramifications resulting from injury can catastrophically alter a patient's life. A burn is classified as full thickness, or 3<sup>rd</sup> degree, when all layers of skin have been destroyed. Partial thickness or 2<sup>nd</sup> degree burns, depending on depth, leave dermal elements thereby allowing re-epithelialization. If burns are superficial and cause only reddening of the skin with rarely any serious sequelae, they are categorized as 1<sup>st</sup> degree. Burns are caused by fire, electricity, or contact with hot substances that transmit heat. Chemical burns may result in similar wounds, but are associated with special problems particular to the damaging chemical (Dolan, 2004).

After assessment of depth, the physician evaluates the amount of the body damaged. This is often expressed as a percentage of total body surface area. While the total body surface area percentage burn is part of evaluating a disability, it must be remembered that only a small area of burn may result in a severe disability if a crucial structure is injured. For example, if the burn affects a hand, foot, eye, or face, the consequences may be severe. Even a small burn on the face that causes no functional damage may distort the features or leave a scar that causes serious emotional concerns that need treatment (McCauley, 2005).

Large surface area burns may involve many body systems and result in the patient's death. Survivors of such burns may go through weeks to months in the hospital, after enduring many surgical procedures to remove the dead tissue and cover the defects with skin. The problem of new skin coverage is compounded when a patient has a large burn, and there is less healthy skin available that can be used for grafting. A long time is required for healing before donor sites can be reharvested (used again). The surgeon can use only the person's own tissues for grafts and flaps. Skin taken from other human donors or animals can be used for temporary coverage but cannot be left in place as the body will eventually reject and destroy the foreign tissue.

Hospitalization treatment involves painful debridements, prolonged periods of immobility, concern about survival, and anxiety about the future. Depression and anxiety complicate recovery by diminishing motivation. After these problems are addressed, many patients need further surgery to reconstruct areas that were postponed while life-saving measures occurred. For example, scar contractures that limit function need

surgical release and skin grafting. Compression garments may be prescribed for months to years, as they diminish both contracture and scarring. These garments apply pressure to healing surfaces, providing some comfort to symptoms of immature scars (Mathes, 2006).

Physical deformities, facial scarring, loss of self-esteem, and other factors complicate the life of the survivor of a major burn. A burn survivor requires a compassionate long-term approach to recover, though some disability may be permanent.

Though smaller burns are easier to treat, the stigma of scarring can complicate a rather simple injury (Bentz, 1998). The surgeon must always consider the psychosocial effects of scars on the face or hands when evaluating patients with minor burns. If the burn occurs on a child, the surgeon may make different decisions than if the burn occurs to an adult.

### ***Hand Injuries***

Hand surgery is a sub-specialty of plastic and reconstructive surgery in which the surgeon combines knowledge of healing with the need to preserve function. For example, a hand that does not move and lacks strength restricts gainful employment and may be a source of emotional distress (Richter, 2005). Physicians use the principles of wound healing, along with medical and surgical techniques to allow as much range of motion as possible for moving anatomical structures. Tissues need to heal but they also must move and glide (as in tendons) or flex (as in joints). The physician uses knowledge of scar chemistry and properties of healing to achieve a maximum return of function. Many medical professionals consider this field of endeavor the essence of wound healing (Janis & Attinger, 2006).

### ***Congenital Anomalies***

When helping children with congenital anomalies, the plastic and reconstructive surgeon works on many areas of the body. These areas include cleft lip and palate, hand anomalies, congenital problems of the trunk, anomalies of the genitalia, and major anomalies of the head and neck. Anomalies of the head and neck involve combined cranio-facial operations that may revise the facial skeleton, and procedures that enter the brain cavity of the skull. The physician identifies the deficiency, assesses how to repair it, and decides what tissues to use; the defect is accurately measured and local tissues are selected to best match skin color and texture.

In congenital hand anomalies, function is of utmost importance. Children often learn to compensate for disability at an early age, and usually develop skills that allow them to function even prior to a surgeon's intervention. These adapted skills are taken into consideration when planning the most optimal surgical corrections (Attinger et al., 2006).

### ***General Reconstructive Surgery***

Head and neck tumors, skin cancer, and breast diseases are treated in the field of plastic and reconstructive surgery. The principles remain the same: remove the tumor, identify the defect, and use proven techniques to replace or reconstruct deficiencies (Evans, 2000; Janis & Attinger, 2006). Plastic and reconstructive surgeons also evaluate and treat deficiencies and defects of the abdominal wall and the extremities, including the legs. Osteomyelitis (infection of a bone) of the leg may occur following open fractures. This chronic disease is resistant to treatment and challenges the skills of the surgeon, as it is very difficult to eradicate. In addition, the lower part of the leg presents a complicated area of the body to cover with a flap. Lack of local tissue for use in flaps frequently requires free flap surgery. Scar from injury and infection invariably complicates surgery. Yet, modern surgical techniques have turned many near hopeless medical problems into highly manageable and treatable conditions.

### ***Cosmetic Surgery***

No writing on plastic and reconstructive surgery is complete without the mention of cosmetic surgery. Once thought of as reserved for the wealthy and vain, cosmetic surgery is now widely accepted. As individuals realize that disability greatly influences how we perceive ourselves, surgery to modify or eliminate defects in an attempt to raise self-esteem has become more acceptable and popular (Brodwin et al.,

2003). Plastic surgeons use the same skills and knowledge acquired in the healing of those with major deformities to treat aesthetic problems.

Cosmetic surgery may correct problems of the aging face or deformities of the nose, cheeks, or chin. Breasts can be made larger, smaller, or shapelier; the body can be contoured with liposuction, and abdominoplasty or thigh and buttock lifts are common. As with reconstructive surgery, cosmetic surgery is designed to restore the physical as well as emotional well-being of the person (Thorne, 2007).

## **Functional Limitations**

Functional limitations are classified as both physical and emotional. Physical limitations are more apparent to assess and treat, yet emotional or psychological limitations are of equal importance.

### ***Physical Limitations***

As stated at the beginning of this chapter, plastic and reconstructive surgeons work closely with other professionals using a multidisciplinary team approach to treatment. An individual needing plastic and reconstructive surgery often has multiple major physical disabilities. The goal for the team is to provide maximum restoration of function and the greatest degree of emotional adjustment possible.

People with lower extremity problems may have limitations in ambulation. Surgical intervention may restore some function, but many persons continue to have limitations in standing, walking, climbing, lifting, and carrying. Rehabilitation counselors must plan for the future with these limitations in mind. When functional limitations are not clearly addressed in medical reports, it is appropriate for the counselor to contact the treating physician for clarification.

A number of conditions result in functional limitations of the upper extremities. These conditions affect manual dexterity, fine and gross eye-hand coordination, lifting and carrying, and similar activities. One such condition is contracture, which involves tightening of tissue around a joint and limits full range of motion of the affected joint, resulting in functional limitations in physical activity. Through surgical intervention, the reconstructive surgeon attempts to minimize contracture and thereby maximize function.

When the rehabilitation counselor and client begin to establish educational and vocational goals, the counselor may benefit from consultation with the treating physician. For example, if limitations of movement of the hands caused by contracture are interfering with a specific vocational goal, the physician may be able to increase function of a certain needed movement through minor surgical intervention (Van Loey & Van Son, 2003). As part of the overall treatment, the physician also prescribes additional physical or occupational therapy.

### ***Emotional Complications***

Treatment of the psychological impact of severe disability and disfigurement is critical to the rehabilitation process. Emotional symptoms can persist long after a patient achieves surgical goals. The surgeon must be sensitive to the psychological and social factors involved in each individual case, and be aware that each person is affected in a different way.

The many months of medical isolation necessary for multiple surgeries and preventing infection may lead to problems with family, friends, and the community. This situation further aggravates the emotional consequences of the injury and isolation, though its impact is often overlooked. In addition, a person's reactions to injury may not be proportional to its severity. Relatively small injuries or disfigurements may result in serious psychosocial disability. Rehabilitation counselors must take this into account. People with clearly visible, severe disfigurement may have become socially isolated and their education or employment may have been disrupted. As a result of their injuries, restoration of self-image is as crucial to the rehabilitation process as the physical aspects of reconstruction (Irons & Irons, 1989).

Colmano and Tempereau (1989) described factors that help patients recover emotionally from severe burns and disfigurement. Burn survivors often share the same fears, such as losing their independence, re-entering society, and being viewed in public. The authors additionally recommended that patients seek treatment from a multidisciplinary team, since these professionals can address patients' physical, emotional, and spiritual needs in a holistic manner.

Due to the immediate and often life-threatening physical needs of the patient, professionals providing treatment often overlook his/her personal feelings and emotional concerns. A major function of the rehabilitation counselor is to recognize and treat the emotional concerns of the individual so as to provide timely and necessary support in the vocational and psychosocial readjustment process (Martz , Livneh, & Turpin, 2000). Much of the counselor's time during this early intervention phase may be spent helping the person rebuild self-esteem and confidence to eliminate destructive and negative self-perceptions.

## **Rehabilitation Potential**

Rehabilitation potential for individuals who have had plastic or reconstructive surgery varies, depending on the area of the body affected and the extent of functional limitations that remain following medical treatment. Both the counselor and client can work together to develop a rehabilitation plan emphasizing the person's abilities while accommodating limitations. A person who has had reconstructive surgery and is proceeding towards vocational rehabilitation may well have gone through a long period of intense physical and emotional upheaval (Phemister & Crewe, 2007; Van Loey & Van Son, 2003). Procedures are often painful, complicated, and difficult, and the net result is less than a return of normal function and appearance. A counselor needs patience and empathy when working with someone who has recently experienced loss of function, disfigurement, or both.

After enduring a long process of physical and psychological upheaval, individuals who have survived severe burns must begin to adapt to new or different lifestyles (Richter, 2005). As previously noted, people with severe burns may need to wear compression garments and/or splints for up to two years to minimize scarring. These garments, especially when worn on the face and arms, create a challenge not only to the client but to the rehabilitation counselor as well. Most of these individuals do not return to work during this time because of the reactions of other people to the unnatural appearance of these garments. Counselors must be actively involved with potential employers to explain the nature and purpose of the garments. The facemask, for example, may appear threatening and make jobs dealing with the public difficult or inappropriate. For people wearing splints or gloves, manual dexterity is affected and the individual must be careful to avoid substances that will damage the gloves.

Many people with severe facial disfigurements due to burns remain at home because of fears about how the public will react to them. Assistance can be discovered through interaction with local burn centers and hospitals (McCauley, 2005). Counselors need to take proactive positions and meet these individuals in the hospital or the home to discuss the benefits of rehabilitation services. Survivors of even extreme burns often have considerable rehabilitation potential and can benefit greatly from the provision of educational and vocational services.

The rehabilitation counselor can serve as a bridge between the person with an injury and a current or potential employer (Dell Orto & Powers, 2007). Employers need to understand such aspects as recovery time, possibilities of multiple surgeries, functional limitations, the wearing of pressure garments and/or splints, and reasonable accommodations at the workplace. Through becoming familiar with the specific feelings of individuals they help, counselors can carefully provide appropriate guidance and support toward realistic occupational goals. The desires and goals of the client must always be given primary and serious consideration when developing rehabilitation possibilities.

When evaluating people with disabilities and resulting functional limitations, understanding the role of plastic and reconstructive surgery is essential. The knowledge of how wounds heal allows rehabilitation counselors to be informed when working with treating physicians, be more accurate in predicting work status and disability, and have an enhanced understanding when providing help to clients. When evaluating the person who has had, or is currently undergoing plastic and reconstructive surgery, the counselor must take all factors into consideration.

## **Conclusion**

Although plastic and reconstruction surgeons help minimize actual damage and disfigurement that has been caused by congenital or catastrophic injuries, individuals may need substantial emotional counseling by professionals trained in this area. The person's family or significant others may require



emotional support, but often are left out of the process. Limitations due to disfigurement include society's negative perceptions and the accompanying discrimination that an individual who has an atypical appearance encounters. Employers and potential employers need to be educated in this area. Advocacy on the part of the counselor to minimize the effects of the "disabling environment" of society is essential to the rehabilitation process. A client's positive self-image, appearance enhancement techniques, improved relationships following catastrophic injury or illness, are able to help neutralize the negative reactions of others. Plastic and reconstructive surgeons begin this process as they attempt to improve people's functional capacities, diminished atypical appearance, and promote enhancement of self-image through modern surgical techniques.

## **Case Study**

Ms. Karine Manukyan is a 33-year old college educated elementary school teacher. She is an Armenian-American and relies on her close family ties. In an automobile accident, Karine sustained major burns to her face, neck, arms, hands, and a portion of her back. She suffered deep partial-thickness (2<sup>nd</sup> degree) and full thickness (3<sup>rd</sup> degree) burns over 25% of her body. Medical treatment at a burn center involved multiple surgeries and three months of hospitalization. Plastic and reconstructive surgery was successful to some extent, but disfigurement remained because of severe burns and scarring of the face, upper extremities, and back. Damage to Karine's fingers makes fine manipulation difficult, although she is still able to write well with her left, dominant hand. Ms. Manukyan has minimal strength in both hands; lifting and carrying are restricted to ten pounds with the right hand and five pounds with the left.

Emotionally, Karine is anxious and depressed. Although it is difficult for her to face the public, she forces herself to go out to the local shopping mall, visit friends and relatives, and dine at restaurants in the area, where people know her. Often, she comes home from these excursions exhausted and emotionally distraught. Although her husband and son are supportive, they are having problems coping with the extent of her disability and remaining disfigurement, and want Karine to have additional surgical procedures "until she looks as she did before the accident." Ms. Manukyan does not want additional surgery at this time, and is aware that surgery cannot make her appearance as it was before the accident.

Prior to working as a teacher, Karine was employed by the same school district as a tutor. Her unique ability to speak both the Armenian and Spanish languages fluently helped her obtain employment with this school district, which has a large population of Armenian and Hispanic students. Once Karine obtained her teaching credential, the school district promoted her from tutor to 5<sup>th</sup> grade teacher; her evaluations in both positions were excellent.

Ms. Manukyan held part-time and short-term jobs before her full time work as a tutor. These included cashiering in a car wash, printed circuit board assembly at a small electronics manufacturing company, office assistant at a medical clinic, and sales clerk at a department store.

## **Questions**

1. The client has recently completed her last surgical procedure. When should rehabilitation counseling be instituted?
2. Outline a vocational profile for Karine including age category, educational level, work history (including exertional and skill levels), occupationally significant characteristics, and transferable skills.
3. What kinds of employment positions are possible assuming Ms. Manukyan will have marked decreases in strength and range of motion of the hands and arms and residual facial scarring and disfigurement?
4. Identify steps to help Karine and her family deal with the emotional issues and concerns. Discuss the prospect of future plastic surgery and her family's idea that she will look as she did before the accident.
5. Discuss your feelings about working with clients who have catastrophic injuries and disfigurement.
6. Discuss multicultural dimensions of this case.

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## Chapter 31

# EVALUATING UPPER EXTREMITY FUNCTION AND IMPAIRMENT

*George W. Balfour, M.D.*

## Introduction

While the occurrence of upper extremity injuries has become increasingly common, treatment of these types of disabilities has been developing for centuries. As early as 1717, an Italian physician wrote about “the harvest of diseases reaped by certain workers” caused by “certain violent and irregular motions and unnatural postures of the body that impair the natural structure of the vital machine.” In the early 20th century, maladies that we call upper extremity disabilities were seen as distinct problems. These disabilities were named individually after the specific blue-collar occupation in which they developed. “Stitcher’s wrist,” “bricklayer’s shoulder,” and “cottontwister’s hand,” for example, were common documented disabilities. More recently, upper extremity disabilities have been documented within populations of “white collar” workers. Telegraphers were once noted to get “telegraphist’s cramp,” for instance, while workers with clerical occupations got “writer’s cramp.” Today, upper extremity disabilities are found in populations of workers in high-technology fields, such as software engineering, online brokering, and customer service.

According to the American Medical Association’s (AMA) *Guides to the Evaluation of Permanent Impairment*, 60% of the function of a “whole person” is assigned to a single upper extremity (American Medical Association, 2009). Of this 60%, 90% of the function of the upper extremity is defined to be in the hand. Hand function represents more than half (54%) of one’s total body functioning. From this medical definition of impairment, the rehabilitation counselor can more readily understand the impact of a hand injury on an individual’s ability to function in the workplace.

Impairment, however, differs from disability. Impairment is the loss of function in the physical sense, while disability is an inability to overcome or compensate for that impairment. Oskamp (1988) defined impairment and disability as follows: **Impairment** describes an abnormality or a loss of a physiological structure or function. **Disability** refers to the consequences of impairment – a restriction or lack of ability to perform some activity that is considered appropriate.

Injuries resulting in upper extremity disabilities occur in many different ways and in a variety of areas: the home, highway, sports field, battlefield, and workplace. In the workplace, injuries to the hand and upper extremity represent approximately one-third of all work-related injuries internationally. In 2006, approximately 285,000 people working in business and industry in the United States lost time from work due to a hand injury (Bureau of Labor Statistics, 2006). Upper extremity injuries, while commonly occurring, are additionally characterized by long periods of recovery. The better we understand these injuries, the more we can contribute to the rehabilitation of those injured by positively impacting the economic, personal, and social consequences of the injury, resulting disability, and functional limitations.

According to the National Health Interview Survey (NHIS), as reported by Smith et al. (2005) upper- and lower-body extremities accounted for more than 60% of work-related and non work-related injuries. The largest percentage of work-related injuries, however, occurred in the upper extremities. Over a third of these upper-extremity injuries (36%) occurred on the job.

Approximately four million people in the United States workforce have some form of limitation involving impairments of manual dexterity, range of motion, strength, and endurance of the hands and arms. Of this number, almost 300,000 employees are unable to fully use their upper extremities in a major way.

This figure includes people with paralysis or absence of part or all of one or both upper extremities. Many industrial injuries involve the hands and fingers, causing different degrees of impairment depending upon the extent of injury, loss of function, and whether the involved hand is dominant. Because the majority of occupations involve use of the hands, rehabilitation counselors must possess knowledge of hand function and impairment so that they may recommend rehabilitation plans and reasonable accommodations for workers with these disabilities (Brodwin, Parker, & DeLaGarza, 2003).

This chapter addresses the function and anatomy of the upper extremities by examining typical injuries and their evaluation, treatment, prognosis, and healing. It discusses assessment of impairment, functional limitations in commonly occurring hand injuries, and rehabilitation potential for individuals with upper extremity impairments. The goal of this chapter is to make available medical information on upper extremity injuries to aid the rehabilitation counselor in more effectively providing services for clients with these conditions and resulting functional limitations.

## Function and Anatomy

### *Function of the Hand*

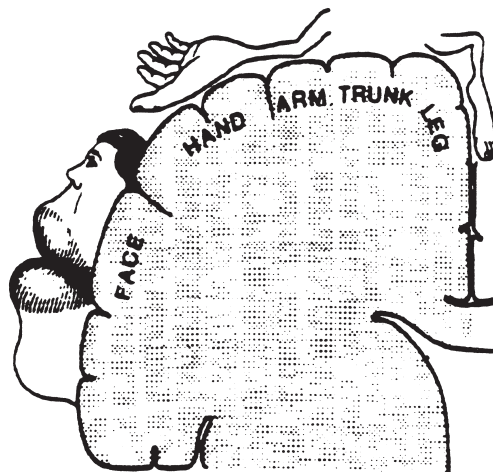
A variety of functions rely on the movement of the hands, including sensory, fine manipulation, gross gripping, cosmetic, and social. The hand is a sensory organ; a high concentration of nerve endings in the palms and fingers provide information about one's environment. Watch an infant at play. The infant looks at an object, picks it up and feels it, and discerns weight, hardness, surface texture, temperature, size, and shape. Lastly, the infant puts the object in his or her mouth further feeling and tasting it.

The somasensory surface of the brain receives the sense of touch. Almost as much brain area is devoted to hand sensation as to the rest of the body (excluding the face and mouth). Figure 1 is a schematic picture of the somasensory area of the brain that depicts the importance and proportions of brain tissue devoted to the sense of touch. Note that only the lips and face are given more area than the hand. In a sense, objects can be "seen" with the fingers. Other areas of the body, like the face or thigh are not able to read objects with the same accuracy or detail identification as the fingers. The amount of brain surface devoted to a body part is proportional to the sensitivity of that body part. The hand's high sensitivity to touch, for example, is a result of the high concentration of nerve endings in this area, particularly the palmar skin. A hand with significant sensory impairment can be considered "blind."

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**Figure 1**  
**Mannequin Illustration of Sensory Distribution of the Central Gyrus of the Cerebral Cortex**

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Source: Truex, R.C. & Carpenter, M.B. (1969), *Human Neuroanatomy* (6th ed., p. 559). Baltimore, MD: Williams & Wilkins. Reprinted with permission.

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Besides providing sensory input, the hands function as the terminal devices of the upper extremities for functions like grasping, lifting, and carrying. Hands also perform fine and gross manipulative tasks such as turning a key, holding a glass, doing fine crafts, writing, using tools, and operating office equipment. Additionally, hands are used in a social role: the shaking, holding, and gesturing of this part of the body signals characteristics such as feelings, emotions, greetings, and enthusiasm. The hands serve a cosmetic function; we paint fingernails, wear rings, and put on decorative gloves. Hands also function during activities requiring gross strength (i.e., in lifting, carrying, pushing, pulling, and while using large tools). When working with shovels, hammers, or saws, the function of the hand is to grip and hold the equipment; the principal force and motion occurs at the wrist, forearm, elbow, and shoulder. During the assessment of impairment and rehabilitation potential, each of these multi-faceted functions and any associated limitations need assessment (Strickland & Graham, 2005).

## ***Practical Anatomy and Physiology of the Hand and Upper Extremity***

### ***Terms and Hand Function***

**Terms.** The trunk (upper body) is described using two terms. Ventral (anterior) refers to the front or face side of the body; dorsal (posterior) is the term used when describing the back. Medial and lateral indicate closer to (medial) and farther away (lateral) from the midline of the body. When describing the hand, palmar (instead of ventral) and dorsal (instead of posterior) are the terms used.

**Hand function.** This begins in the cerebral cortex of the brain at the location of sensory and motor control where afferent nerve fibers conduct impulses to the brain and efferent fibers carry impulses from the brain to the rest of the body. Efferent fibers pass down the brain stem and exit through the nerve roots of the fourth to seventh cervical vertebrae (C4-C7) and from the first thoracic vertebrae (T1). On either side of the cervical spine, in the neck, these nerve roots combine to form the brachial plexus of nerves. From this plexus, three major nerves (the median, ulnar, and radial) reach the forearm and hand (Trumble & Allan, 2000; Weiss & Falkerstein, 2005).

### ***Neurological***

**Median nerve.** This nerve supplies sensation to the palm and palmar aspects of the thumb, index, long, and radial half of the ring finger. It is the most important sensory nerve to the hand and is the motor nerve for most of the muscles of the ventral forearm (forearm flexors), the muscles that bend and flex the wrist and flex or close the fingers and thumb. These median nerve controlled flexors provide power for gripping, lifting, and wrist flexion (Green, Hotchkiss, & Pederson, 2005). Below the wrist, the median nerve supplies motor and muscle innervation to the small, intrinsic muscles of the thumb (the thenar muscles) which aid in rowing the thumb out of the plane of the palm into opposition, and contribute to the power of pinch.

Median nerve injuries are classified as either “high” or “low” (Strickland & Graham, 2005). A “high” median nerve injury results in an inability to flex the wrist, fingers, and thumb. This type of injury makes gripping of objects impossible and causes numbness of the thumb, index, center fingers, and half of the ring finger. Contrastingly, a “lower” median nerve injury causes numbness and weakness of the intrinsic muscles of the thumb, preventing actions relying on these muscles, such as pinching, turning a key, or in touching the tip of the thumb to the tip of the little finger.

**Ulnar nerve.** This nerve arises from the medial cord of the brachial plexus. The ulnar nerve passes behind the medial epicondyle (the bony bump on the medial side of the elbow) an area called the cubital tunnel. It then passes down the ulna (little finger) side of the forearm to the hand. In the hand, it provides sensation to the little finger, the ulna half of the ring finger, and muscle innervation and control to the small intrinsic muscles of the hand. Intrinsic muscles provide balance to the fingers and fine manipulative control of the hand. Injury to the ulnar nerve principally impairs dexterity and fine manipulation, including functions like pinch and manipulation of small objects, such as screws, coins, and buttons.

**Radial nerve.** The radial nerve is the sensory nerve for the back (dorsum) of the hand and thumb. In the forearm, it supplies muscle innervation to the extensor muscle group. These muscles extend the wrist, fingers, and thumb. Radial palsy leads to wrist drop, a condition in which the hand drops toward the ground

because the person is unable to raise the hand or fingers against gravity. A wrist drop indicates an injury to the radial nerve. A lower radial nerve injury causes numbness (Trumble, 2002).

### ***Muscular***

**Forearm.** There are two sets of extrinsic muscles (those located outside the hand itself) in the forearm, the ventral (flexion group) and the dorsal (extensor group). These muscles are located within the forearm, but produce function in the hand. Ventral extrinsic forearm muscles, or flexors, reach the hand through tendons and act to flex or bend the wrist and fingers. The median nerve controls most of this function. An extensor group of extrinsic muscles function principally to extend the wrist, fingers, and thumb; they are innervated by the ulnar nerve (except those of the thumb) and balance finger motion, add strength to grip, and abduct and adduct the fingers (Green et al., 2005). To examine radial nerve function, the physician looks for wrist extension. To test median nerve function, thumb flexion is evaluated. In the case of ulnar nerve function, the physician assesses isolated abduction of the index finger.

### ***Vascular***

The blood supply for the upper extremity enters the arm as the large single axillary artery and continues as the brachial artery. At the elbow, it splits into the radial and ulnar arteries, which pass down their respective sides of the forearm and reconnect in the hand through both deep and superficial arches. From these arches, two digital vessels supply each digit, one to each side of a finger. A single vessel is adequate to supply a hand, but the hand may have some cold intolerance. Although two digital arteries supply each finger, a single digital vessel can usually provide an adequate blood supply.

**Vascular injuries.** Most arterial injuries are due to direct laceration, blunt trauma, or thrombosis. Lacerations compromise the blood supply to the affected part (Trumble, 2003). If there is no collateral circulation to a part, it will become ischemic (deficient in blood) and die. Collateral circulation means that two vessels supply the same part or area. If only one vessel is damaged, collateral circulation (the alternative blood pathway) may be adequate to keep the part alive and functional. Completely divided vessels usually retract, rapidly clot, and then bleeding ceases. Partially divided vessels cannot retract and, therefore, may allow serious blood loss to occur. If an intact vessel clots off because of blunt trauma, embolism, or arteriosclerosis, the result is loss of blood supply through that particular vessel (Green et al., 2005).

### ***Kinesiology***

**Upper extremity function.** Combined function of the shoulder, elbow, and wrist allows the hand to move anywhere within a large circular area. Shoulder motion involves the combined motion of the sternoclavicular joint, acromioclavicular joint, scapulothoracic motion, and the glenoid humeral joint. The shoulder girdle consists of the arm, scapula, and clavicle. The clavicle attaches to the sternum and to the scapula at the other end, functioning as a tie rod keeping the shoulder girdle to the side. Without a clavicle, the shoulder collapses inward toward the midline of the body, giving a narrow shoulderless appearance. The scapula lies on the thorax and glides back and forth across it. Muscles located on the back, anterior chest wall, and upper arm further contribute to motion of the shoulder (Strickland & Graham, 2005).

**The elbow.** The elbow is a complex set of three joints. The olecranon (proximal end of the ulna) articulates with the trochlear notch of the humerus (true simple hinge joint) for simple extension and flexion of the elbow. To create supination (palm up) or pronation (palm down), the radius rolls or radiates around the ulna. This last motion occurs both at the proximal ulnar-radial joint and the distal radial-ulnar joint within the wrist.

**The wrist.** The wrist involves a complex set of joints that function in supination and pronation, the turning of the hand palm up and palm down. It functions in dorsiflexion and palmarflexion movement of the hand upward and downward (as in the hand motion of saying good-bye), and in radial and ulnar deviation (motion of the hand from side to side). The wrist is involved with opposition of the small and little fingers, which is a limited motion that is important during grip and holding actions. The eight small bones of the wrist (carpal bones) are set between the forearm bones and the bones that form the palm of the hand (metacarpals). Joints of the fingers (metacarpals/phalanx, proximal interphalangeal, and distal interphalangeal joints) are hinge joints that function during flexion, extension, limited abduction, and adduction (Baratz, 2006; Geissler, 2005).

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## ***The Dermis***

**The skin.** Skin serves as an outer covering of the body, protecting it from the external environment. It is a constantly renewing wear surface that contains terminal sensory nerve endings, and is classified into specific types. Palmar skin is specialized in that it is thicker than the rest of the skin. It is more rigidly fixed to the deeper palmar fascia and, hence, less mobile. Palmer skin is hairless, but rich in sweat glands and sensory nerve endings. Scars in the palmar skin are initially very sensitive and thicken, but as they mature become durable and non-tender. The early healing phase typically takes three months to resolve, an important characteristic for the rehabilitation counselor to know when providing vocational services for workers with injuries involving the palms.

Defects in palmar skin caused by injury such as tears, lacerations, or burns can be covered or replaced with grafted skin or skin flaps. Skin graft is a thin layer of healthy skin removed from one site by a surgeon with a razor or dermatome and placed in an area needing skin coverage. These grafts retain the characteristics of their site of origin. Transferred skin, as compared to palmar skin, is thinner, more mobile, and less adherent to deep structures. Full thickness graft may involve skin with hair follicles. In persons with dark skin, the skin often becomes hyperpigmented (increased pigmentation). A physician may utilize a variety of flaps to provide thicker more durable skin. The type of skin on the back of the hand (the dorsal surface), for example, is characteristically thin, pliable, and stretchy, allowing the fingers to close and enabling the wrist to palmarflex. In contrast to palmer skin, this area has hair follicles (Jebson & Kasdan, 2002; Trumble, 2000).

**The nails.** The nails are specialized structures that act as protective covering for the tips of the fingers. An open wound near or under the nail is a source of pain and a viable route for infection. Nail injuries can heal abnormally with ridges or splits, also known as bifid nails (Jebson & Kasdan, 2002).

## **Evaluation and Treatment of Injury**

### ***Evaluation***

The surgeon's approach to injury follows a consistent procedure and order of priorities. The first step includes obtaining a detailed history and physical, as circumstances allow. The physical examination starts with inspection or a simple overview of the injury. During the physical, changes are noted and each injury is categorized in relation to the skin, color, and condition of the hand. Specific examinations reveal particular injuries. For example, palpation demonstrates losses of tendon continuity, fractures, dislocations, and torn ligaments. Loss of sensation is tested by touch or pinprick. Radiologic examination (x-rays) provides detail of bony injury and indicates the presence of metal and other foreign bodies.

### ***Treatment***

Once the surgeon understands the injury, he or she formulates a treatment plan. If the injury being treated involves an open wound, the first priority is to clean the exposed tissues and remove any bacterial contamination. If unstable fractures or joint injuries complicate the situation, stabilization, usually with metallic fixation devices, is required. Individuals with vascular injuries may need arterial and occasionally venous repair. If, however, the vascular injury is categorized as vascular insufficiency, the person must have emergency care. In the case of venous insufficiency, muscle tissue not receiving an adequate blood supply dies within six hours while other tissues of the hand become necrotic less quickly. The patient must undergo repair of tendons, nerves, and skin, though this procedure can be delayed. Skin coverage is done immediately and before any deeper structures are reconstructed to avoid infection and problems due to scarring. The simplest skin replacement is performed with a skin graft in which an extremely thin layer of skin is taken from a healthy place on the body and sewn over a defect or open wound (Trumble, 2000; Weiss & Falkerstein, 2005).

### ***Stabilization***

When a patient has a deformity that cannot be corrected by non-operative methods, he or she undergoes a fracture stabilization procedure. The usual reason for instability is that unopposed muscle or ligament forces create and maintain a deformity. Another indication for fracture fixation is when the fracture

involves a joint surface. Joints are like machine parts; if their smooth surfaces are allowed to heal unevenly, joint wear is accelerated which eventually leads to painful arthritis. The simplest method of internal (metal) fixation includes straight smooth pins which are drilled across the fracture to stabilize the area until it is healed. Many different types of fixation devices can be used.

### ***Ligaments***

Ligaments which hold joints together can be injured and torn. Many ligament injuries heal spontaneously, while others require casting or surgery. Incomplete tears heal independently with time, but complete tears need surgical repair. Torn ligaments that are not repaired become sources of chronic instability and impairment and, if neglected, lead to arthritis.

### ***Tendons***

Tendon injuries include acute laceration, rupture, avulsion from the bony sites of attachment, erosion (such as in rheumatoid arthritis), attrition, and tendonitis. Tendons are repaired end-to-end, replaced by tendon graft, or substituted by tendon transfer. Other conditions causing pain can be acute or result in chronic inflammation, such as those which occur in cases of tendonitis of an elbow or knee joint (Burke, 2006).

Tendon areas are divided into zones for purposes of prognosis and planning of tendon surgery. The palmar surface of the hand is divided into five tendon zones. Zone I extends from the pip joint (middle joint of a finger) to the end of the finger. Tendon surgery within this zone usually results in successful outcomes. In Zone II (mid-palm to the pip joints), the results of tendon repairs are far less predictable and surgical complications (such as scarring, adhesions, and stiffness) are more common. Complications involve scarring, adhesions, and stiffness. The proximal palm is referred to as Zone III. Within this area, tendon repairs are usually successful. The area of the carpal canal (from the wrist crease distally to the palm) is Zone IV and, because of tightness, tendon repairs are complicated by tendon adhesions. Zone V is the forearm, an area that has a positive surgical outcome.

Tendon graft operations are used to avoid problems of scarring by placing tendon suture lines outside of tight areas, lessening the risk of adhesion. These tendon transfer procedures involve rerouting of a muscle tendon complex to a new site of action to replace a lost motor function.

## ***Newer Developments***

Like all medical specialties, the discipline of hand surgery continues to evolve (Jebson & Kasdan, 2002). In the past few decades, microsurgery has developed from a research tool into a viable technique commonly performed throughout the country. Free tissue transfers have become common practice and are widely accepted. Arthroscopic surgery, initially developed for the knee, has become an accepted approach for the shoulder, elbow, and wrist. Arthroscopic procedures are being performed on small joints, such as metacarpal/carpal joints and metacarpal/phalangeal joints. Similarly, fracture treatment has continued to improve with newer, smaller, and stronger implants and progressively less invasive methods. Surgical treatment plans that only a few years ago would have been considered too risky to attempt have now become accepted surgical practice (Strickland & Graham, 2005).

## **Assessment of Impairment**

Assessment of impairment is accomplished using a scoring system which assigns specific values to various functions (Mink Van Der Molen, Ettema, & Hovius, 2003). Within the hand, evaluated functions include loss of parts (amputation), cosmetic defects, pain, tenderness, diminished sensation, limited range of motion, and loss of strength. An assessment must describe subjective pain, including intensity and frequency, along with the appearance of the hand, together with the presence of dirt or calluses, and condition of the nails. An evaluator describes sensation using a variety of testing methods; the report measures loss of parts, presence of scars, ranges of motion, and strength of grip, pinch, lifting, and resistance.

In the first part of the evaluation, the individual reports his or her functional history and present activities. It is best to ask about a variety of activities rather than simply having patients list their functional



limitations. The history needs to illustrate the patient's functional capabilities by describing duration and frequency of performed activities. Similarly, the report should explain activities that cannot be performed. Questions concerning lifting, carrying, pushing, pulling, gripping, pinching, manipulating, and sensing are all relevant to the physician's overall assessment of injury (Meyer, 2003).

### ***Medical Reporting***

Medical reports have a consistent format, beginning with subjective complaints and extending to the history of the medical problem or injury in which the individual's past occupational record and social functioning are addressed, and lastly followed by the physical examination. The physical exam contains inspection, palpation, range of motion, muscle testing, neurological examination, and results of special tests including x-rays, MRIs (magnetic resonance imaging), and CAT (computerized axial tomography) scans.

The physical exam typically begins with inspection, which includes a diagram of the palmar and dorsal surfaces of the hand and entire extremity. These diagrams illustrate amputations, nail deformities, scars, pigmentation alterations, calluses, and smooth areas. Amputations are further described according to same digit comparisons or identification of the most distal residual bone. Nail deformities are similarly measured by length, width, keloids, raised areas, and tender spots (neuromas). Areas of hyperpigmentation and hypopigmentation are illustrated on a drawing. The physician notes calluses suggesting heavily used areas, and clean soft areas suggesting lack of use.

Recently, there has been emphasis on patient self-reporting tests in the form of examinations such as patient completed hand diagrams. In such a diagram, the patient is asked to draw his or her symptoms, pain, and numbness on a drawing of a hand. Another form of patient reporting is the DASH (Disability Arm Shoulder Hand) score or other similar patient completed report (Solway, 2002).

### ***Pain and Sensory Deterioration***

#### ***Pain***

Pain is entirely subjective. Nosocomia is defined as a physically unpleasant experience; pain is the individual's perception of that experience. If you take a standardized needle and stick it an equal depth at an equal rate into a dozen individuals, each experiences an equal amount of nosocomia (unpleasant stimulation). Yet, each person describes a different degree and sensation of pain. Swanson, Goran-Hagert, and Swanson (1987, p. 896) defined pain as "a disagreeable sensation that has as its basis a highly variable complex made up of afferent nerve stimuli interacting with the emotional state of the individual and modified by past experience, motivation, and state of mind."

Even though pain is entirely subjective, an individual's complaints are supported by evidence. A person complaining of severe pain in the hand should have a clean, soft hand, evidence of purposeful disuse or avoidance; a dirty, callused, well-used hand raises doubts about the intensity of expressed subjective complaints. Contrastingly, some individuals have few or no complaints, but portions of their hands show evidence of disuse or avoidance. This is an indication that the person has pain but does not complain about it.

Pain can be classified according to its interference with the individual's performance of activities. "Minimal" pain is an annoyance (0-25% factor of impairment in the American Medical Association Guidelines). "Slight" pain interferes with activity (26-50% factor of impairment), while "Moderate" pain prevents activity (51%-75%). "Severe" pain prevents activity and causes distress (76-100%). Pain, besides being described by intensity, is described by duration. Severe pain, lasting a few minutes per week is of less significance vocationally than moderate pain occurring more often. "Occasional" pain is pain that lasts up to 25% of the time, while "intermittent" pain occurs 25-50% of the time. "Frequent" pain is pain that lasts 50-75% of the time, while "constant" pain is present more than 75% of the time (American Medical Association, 2009).

#### ***Sensory Deficits***

Subjective complaints of sensory deficits are measurable and, in reliable patients, quantifiable. Testing for sensory deficits includes distinguishing sharp from dull, static two-point discrimination, moving two-point discrimination, Von Frey hair testing, hot versus cold discrimination, proprioception, coin discrimination, and vibration. Proprioception is the sense that indicates body position. It tells us if our hands



are open or closed, if they are below our waists or over our heads, and allows us to perform activities such as typing without looking at the keyboard. Proprioception can be tested by placing the hand behind a screen to hide it from visual feedback (Geissler, 2005). Stereoagnosia is the sense that allows a person to read or see objects by feel; it is how one tells a dime from a quarter in a pocket without visual feedback, and how Braille is read. Patients with nerve lacerations and poor recovery, or following stroke or spinal cord injury often have severe sensory impairments, and may simply ignore the severely impaired hand; the hand then becomes nonfunctional solely on a sensory basis.

### ***Nerve Compression***

Individuals with nerve compression syndromes, such as herniated disks (causing pressure on nerve roots), thoracic outlet syndrome, cubital tunnel syndrome, and carpal tunnel syndrome, also experience decreased sensibility (capability to feel or perceive). Individuals experiencing these losses lose the sense of vibration more than the ability to perceive two-point discrimination. Anyone can experience the sensation of a compression syndrome by crossing the legs until one leg “falls asleep.” “Pins and needles” sensations, deep aching, a loss of the sense of position, and spontaneously dropping objects are associated with compression syndrome. In compression syndrome, the involved nerve is longitudinally intact but nerve impulse transmission is impaired by local pressure on a nerve at some point along its path into the hand (Green et al., 2005; Mackin, 2002).

### ***Range of Motion***

An examiner measures range of motion of each joint. The American Academy of Orthopaedic Surgeons has published standards for measuring joint motion. Each joint has a normal or typical range of motion. For any individual, the extremity on the opposite side is used as a normal standard. National standards define full extension as zero degrees of flexion. In some states, such as California, this is called 180° of extension, but the meaning is the same. Results of these measurements are recorded on a standard “range of motion chart” (Green et al., 2005).

### ***Grip Loss***

Atrophy can be caused by injury, disease, and disuse. Forearm circumference is measured as an estimation of atrophy; the physician compares the uninvolved extremity as a control to the involved side. Grip strength is measured with a calibrated dynamometer. Usually, the examiner obtains and records a minimum of three “good” efforts with each hand. If maximum efforts are made by the examinee, the dynamometer readings are consistent; if less than maximum efforts are made, the readings become variable. The ratio between the involved and uninvolved hand determines grip loss.

### ***Radiological Assessment***

An examination requires radiological evaluation to define the status of the skeletal system and the degree of joint injury, disorganization, and degeneration (Weiss & Falkerstein, 2005). Bone shortening and angulation is reported in degrees. Rotational errors are not seen on x-rays. Density changes, for example, from disuse (osteopenia) are seen on x-rays as thinner, less dense, and more radiolucent bone tissue. The examiner notes joint surface changes, narrowing, sclerosis, fractures, osteophytes, arthritic changes, joint displacements (subluxations), and any additional x-ray findings (Strickland & Graham, 2005).

## **Work and Physical Capacity Evaluation**

Work capacity evaluation measures the capability of a person to function over time. Under the direction of a physician, occupational therapists and work evaluation specialists observe the individual performing tasks over several days to provide an accurate assessment. Specific capacity testing enables the evaluator to quantify the capacity to perform work-related actions. If there is a particular job or specific job functions that are being considered, this can be evaluated.

Physical capacity evaluation documents a person’s hand function, including strength and range of motion (Mackin, 2002). The three components to physical capacity evaluation include administration of hand function tasks, standardized tests, and observation of the person performing the physical demands of

the job. By observing the person performing simulated job duties, the evaluator can report the functional limitations concerning job requirements. An accurate and detailed job description tailored to the specific worker's impairments helps a physician determine if the individual is capable of returning to the previous job.

## **Functional Limitations in Commonly Occurring Hand Injuries**

Hand surgery is a broad field addressing a variety of ailments. Hand injuries cause different functional limitations and thus have varying rehabilitation potential. Attitude and motivation play a significant role in the potential for rehabilitation and success in returning to work.

Fingertip injuries are the most common industrial accidents and the most frequent reason for amputation (Burke, 2006). Variables involved include level of amputation, amount of soft tissue coverage on the remaining part, presence or absence of tender neuromas or bone prominence, and nail deformities. Involvement of flexor tendons influences the person's strength, active motion, and range of motion.

An example is an injury and amputation through the middle of the nail bed where half the nail bed and one-third the distal part of the finger are gone. Medical treatment provides for adequate soft tissue padding; the nail may be rolled over and there is decreased sensibility. Since both flexor tendons are intact at this level, there is no loss of grip strength. The finger may fail to touch the midpalmar crease by one-half centimeter. With an injury similar to that noted above, the duration of time a person misses work is variable. Most heavy laborers require about 12 weeks recovery time before a return to work is possible. Even at 12 weeks, many people continue to experience tenderness that prolongs the time off work. Few amputations at this level cause severe functional impairment or require rehabilitation.

If the site of amputation is proximal to the distal joint, the fingernail is lost. In such an injury, the profundus tendon (the stronger of the two tendons to the finger) is also lost and with it some flexion of the residual digit, along with decreased grip strength. Impairment is different for each digit involved. As a guide, the entire thumb contributes to 40% of hand function, while the long and ring fingers contribute 20% each. The border digits (the index and little fingers) account for 10% each. The loss of several digits increases impairment more than the sum of these parts lost. Multiple digit injuries require longer recovery time, more hand therapy to regain motion and strength, and result in greater residual impairment (American Medical Association, 2009; Beasley, 2003). Injured workers with heavy exertion jobs or those with work involving manual dexterity are more likely to need a job change in order to continue working.

### ***Hand Fractures***

Fractures are another common hand injury in industry, with each kind of fracture having a different treatment and prognosis. Physicians can splint simple non-displaced, non-angulation, and non-articular fractures; usually, these fractures heal within three weeks. Most resulting stiffness in the fingers resolves within an additional three to four weeks. Even laborers can typically return to work within six to eight weeks. If the fracture is displaced, angulated, intra-articular, and therefore requires reduction or surgery, the time for healing to resolve stiffness, regain strength, and return to work is much longer, and some of the residual effects may be permanent. Residuals such as tendon adherence (scar between the tendon and bone) can prevent returning to customary work. Common sites for tendon adhesions following surgery are the extensor tendons over the middle and proximal phalanges. For an individual with tendon adherence, the only way to regain motion may be through surgical intervention. Full range of motion is rarely regained after surgery, and manipulative tasks are often impaired as a result.

### ***Wrist Fractures***

The most frequent wrist fractures involve the waist of the scaphoid, one of the eight small bones of the wrist, and are most often found in people with no other injuries (Baratz, 2006). Median time to union in non-displaced fractures is about 15 weeks, and the non-union (non-healing) rate is high (in some research reports as high as 40%). Following injury, stiffness and weakness are common; return to heavy labor takes an extensive period of time.

## ***Carpal Tunnel Syndrome***

The carpal tunnel is the final common pathway of multiple conditions, all of which increase pressure within the wrist (Katz & Simmons, 2002). Carpal tunnel syndrome is the most common nerve compression syndrome. Thyroid disease, masses, extra muscles (congenital), rheumatoid arthritis, amyloidosis, repetitive trauma, vibration disease syndrome, synovial hypertrophy, and prior wrist fractures are some of the causes of carpal tunnel, but many of these conditions are labeled idiopathic (unknown cause). Repetitive use of the hand, as in a cashier's use of an optical scanner throughout the work shift, or activities like typing, packing, and assembling, frequently are the basis of claims of an industrial etiology. Young workers may also develop carpal tunnel as the result of vibrator tools such as pneumatic tools, jackhammers, and power wrenches.

Factors which increase the occurrences of carpal tunnel syndrome in women include gender, age, generalized mild synovitis, and physique. Those at or near the age of 40, and women with thin or fragile physiques are more prone to carpal tunnel. Initial conservative treatment consists of decreased use of the wrist, splints, non-steroidal anti-inflammatory medication, and diuretics. If these methods fail, the diagnosis must be confirmed. The diagnosis of carpal tunnel syndrome is made based upon appropriate history, complaints, and physical findings. Test results indicating carpal tunnel include an abnormal nerve conduction time and an irregular vibratory sense test. If the diagnosis is confirmed, steroid injections into the wrist are given; if this fails and the symptoms are severe and prolonged, surgical decompression may be necessary. Following surgery, the individual is splinted in dorsiflexion for about a month; the scar can remain tender for up to six months. Return to work will take two to twelve weeks, depending upon the physical demands of the job in relation to the involved wrist and hand. Clerical or professional occupations take less recovery time than more physically demanding occupations. Carpal tunnel syndrome may be aggravated by vibration, such as when dealing with mechanics and pneumatic tool operators. These workers may require occupational change if they physically cannot tolerate return to tasks requiring use of vibratory equipment.

Carpal tunnel syndrome is one example of a cumulative trauma disorder. Practitioners believe these disorders are caused by the sum of multiple microtraumas. While this undoubtedly does occur, it probably happens less frequently than diagnosed. Other conditions that occur as the result of cumulative trauma include tendonitis, trigger finger, and tennis elbow. Arthritis is rarely caused by cumulative trauma but can be aggravated by activity. In many of these conditions, the precipitating event is a non-specific synovitis or thickening. This irritation or thickening contributes to carpal tunnel syndrome, tendonitis, and trigger finger (Trumble, 2002).

## **Rehabilitation Potential**

Potential for rehabilitation depends on multiple factors, including whether the injury involves the dominant or non-dominant hand. When the dominant hand is severely injured, training to improve use of the opposite hand is appropriate. Such a plan of physical rehabilitation includes:

- Strengthening exercises.
- Exercises to develop skills in activities of daily living.
- Writing exercises.
- Activities to improve manual dexterity.

Motivation and attitude are pivotal to the rehabilitation process (Brodwin et al., 2003). When serious injuries result in major impairment, the motivation and attitude of a person may mean the difference between successful medical and vocational rehabilitation and failure. Highly motivated individuals with severe impairment enjoy greater success rates than poorly motivated people, even if they have significantly less impairment.

Other factors contributing to rehabilitation potential include age, educational level, and work history. Younger, more educated persons with skilled or professional work backgrounds can more readily adjust to impairment and disability, while older workers are more tolerant of residual discomfort.

Injuries to the upper extremities cause limitations in eye-hand coordination, range of motion in one or both hands, manual dexterity, strength for lifting and carrying, as well as pushing and pulling activities (Meyer, 2003). Accommodating such an employee at the workplace may involve dividing heavier weights or large items into lighter or smaller units. Use of mechanical lifts and movable carts decrease lifting and carrying; the counselor can evaluate ways to diminish or eliminate lifting and carrying, exploring the possibility of replacing physical activities with job activities that are less exertionally demanding (Mueller, 1990). The most effective way to look at accommodation is to complete a detailed job analysis, paying particular attention to job functions requiring use of the injured hand(s). A step-by-step analysis allows the counselor and injured employee to work with the employer in suggesting reasonable accommodation which benefits both the employee and the employer (Brodwin et al., 2003).

As previously stated, many people who have serious hand injuries are unable to return to their previous employment because of residual impairments. As a result, some employers offer lighter duty work or entirely different jobs. Rehabilitation counselors additionally can provide suggestions concerning alterations to the work environment that will accommodate impairments. With the passage of the Americans with Disabilities Act of 1990, more employers are investigating the provision of reasonable accommodation for workers with disabilities and chronic illnesses.

Because the vast majority of jobs require use of the hands and fingers, counselors must be adept at providing accommodation for workers with problems in handling and fingering, manual dexterity, sensation, and hand strength. Adaptive devices, such as prosthetics and orthotics, improve function in many people. Prostheses are artificial devices used to replace a missing body part, and orthoses are specialized mechanical devices used to support or supplement weak or damaged joints and limbs (Brodwin et al., 2003). Individuals who use prosthetic and orthotic devices should cultivate an increased awareness of their environment to assure proper functioning of the devices.

A worker with a prosthesis needs to be careful in hot and humid places which can contribute to the deterioration of the prosthesis and can cause breakdown of the skin around the prosthesis. Dirt can be abrasive to skin and exacerbate skin problems, while dirty environments may also interfere with proper functioning of moveable prosthetic parts (Clawson, 2002).

In addition to possessing an enhanced awareness of their environment, persons with severe upper extremity injuries, including amputation, may face attitudinal, psychosocial, and emotional adjustments. Limitations in activities which can be performed and changes in body image may require a prolonged adaptation period. Many individuals also benefit from short-term counseling or in-depth psychological intervention (Meyer, 2003). Factors such as a person's interpretation of loss of function, circumstances involved with the injury, premorbid personality (personality before the loss), and extent of loss help the counselor analyze needed rehabilitation and psychological services.

There are a variety of orthotic and prosthetic hand devices that provide improved function. These devices are made to accommodate specific job duties. For example, an adaptive device allowed a filling station attendant, who had lost two fingers from his dominant hand, to pump gasoline. The device was worn only for this job duty and, in essence, was a custom tool that was put away when the worker performed other job functions. Orthotists and prosthetists custom design a variety of devices to help workers perform job duties.

Emotionally, everyone reacts differently to injury and impairment. Psychological counseling is an important aspect of rehabilitation for many individuals. Persons with disfigured hands may engage in various activities and do not feel self-conscious while in public. Others, with even minor injuries, attempt to hide their hand(s) from sight (Vash & Crewe, 2004). The counselor needs to evaluate the impairment considering individual needs.

## **Conclusion**

Ability to use the hand involves sensibility, dexterity, and mobility. Rehabilitation of hand injuries often requires time and effort; maximum return of function is the goal of the hand surgeon, hand therapist, work evaluator, and rehabilitation counselor. Counselors must work closely with other professionals to return the person to employment. At the workplace, use of upper extremity prostheses enhances manual



dexterity, bilateral dexterity, eye-hand coordination, grabbing, grasping, lifting, and carrying. Although a prosthesis does not replace a fully functioning arm and hand, it may allow an employee to perform necessary work tasks. A workstation, with accommodation and job modification, can help the worker with an upper extremity limitation to efficiently perform required work functions (Clawson, 2002).

## Case Study

Jeremy Moore is 34 years of age, married, father of two elementary-aged children, and right hand dominant. Currently, Jeremy is a Ph.D. candidate in biomedical engineering and has a Master's degree in both Mechanical Engineering and Industrial Design. Before being deployed to Iraq, Jeremy worked in the research and development of tools for biomedical robotic cardiac surgery for a major company. Much of his work involved use of computers.

In Iraq, insurgents ambushed Jeremy's patrol, detonating a device packed with shrapnel and explosives. Jeremy was wounded in the initial explosion, sustaining a massive crush-type injury to his right forearm and hand. The injury included an open proximal radius fracture, skin loss, a long finger metacarpal fracture, and a major muscle injury requiring debridement of non-viable tissue. Initial surgical treatment consisted of wound debridement, internal fixation of the fractures, an intrinsic muscle compartment fasciotomy (surgical incision and division), and split thickness skin grafts.

Within two months after his surgery, Mr. Moore's scars were healed and he could fully open and close his hand. Yet, range of motion in his wrist was diminished by 40% and his grip was classified as 50% decreased. Jeremy is interested in assistive technology (AT) as it relates to assisting him with his graduate studies and return to work options.

## Questions

1. Define "impairment" and "disability." Distinguish between these concepts in this case.
2. Discuss attorney involvement with this case and how it would impact your services.
3. As Jeremy Moore's rehabilitation counselor, identify several rehabilitation plans for assisting him in returning to gainful employment.
4. Can Mr. Moore return to his prior job on a physical basis? Is the job realistic for this individual?
5. Given Jeremy's education and interests, what AT can you identify that would assist him in his academic goals and return to gainful employment?

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## Chapter 32

# ALCOHOL-RELATED DISORDERS

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## Introduction

The process of fermentation occurs regularly in nature when sugars come into contact with airborne yeasts transforming them into a final product known as alcohol; however, alcohol use did not begin in earnest until the dawn of agriculture, when the types of sugars necessary for fermentation became available. The first alcoholic beverage, most likely date palm wine, originated in Mesopotamia. Through historical evidence, it is known that ancient Egyptians were alcohol drinkers; texts from this period referred to social problems associated with drunkenness. In Babylonian texts laws regulated drinking houses. The Romans worshipped Dionysus, or Bacchus, the god of wine. Greek literature abounds with warnings against intemperance – evidence of a people well acquainted with the health and social implications of excessive drinking. Even the Hebrew Bible cites references to alcohol consumption. Because of its ceremonial importance, overindulgence was frowned upon and alcohol consumption was under strict social control.

The prophet Mohammed banned alcohol partly to distinguish his followers from Christians and Jews. Alcohol remains prohibited by many Muslim traditions; many Buddhists and Hindu Brahmins also abstain for religious reasons. Alcohol's importance, however, has never been confined to the mystical. Throughout its history, alcohol has been used socially for many purposes such as calming feuds, providing courage in battle, sealing pacts, celebrating festivals, and seducing lovers (Holt, 2006). In medieval Europe more practical roles of alcohol included using it as a folk medicine and as a preservative. The abuse of alcohol, a substance long employed for its range of effects, is not new – ancient cultures were aware of its damaging potential.

Throughout the world religious groups and cultures condone, in various degrees, the use of alcohol. Logically, an increased likelihood of problematic drinking and alcoholism results from practices which more strongly advocate its use. From biblical times to the present, drunkenness has been condemned as degrading and perceived as a personal weakness. The general belief that those who frequently abused alcohol were lacking in moral character has continued to the present time (Schuckit, 2006).

Today, alcohol addiction is one of the most common addictions worldwide. Because of its legal availability and worldwide social acceptance, alcohol is relatively easy to abuse. The majority of those who consume alcohol do so at least once a week and for most people, regular drinking on a social level does not create a dependency on the substance (Carson-DeWitt, 2003). Alcohol addiction affects many people close to the alcoholic, including spouses, children, friends, employers, co-workers, and classmates.

According to the National Council on Alcoholism and Drug Dependence (NCADD, 2007), almost 18 million people in this country abuse alcohol. Yearly, more than 100,000 of these people die of alcohol-related causes. Alcohol is a factor in nearly half of all United States' deaths in car accidents. Alcohol-related problems range from mild to fatal. It is a major public health problem intricately connected with other social issues like crime, homelessness, teen pregnancy, and domestic violence (Donovan, 2005; Schuckit, 2006).

This chapter provides information about problem drinking and alcoholism. The conditions, causes, progression, and effects are addressed, along with current views about treatment approaches and recovery. Family issues and assessment are described; vocational rehabilitation, functional limitations, and

rehabilitation potential are discussed. The chapter also reviews factors that affect recovery and the vocational process, and concludes with a case study analysis.

## **Alcohol**

Alcohol is a sedative-hypnotic drug with anesthetic properties. One drink whether it be 12 ounces of beer, 4 ounces of wine, or 1¼ ounces of distilled alcohol all contain about the same amount of alcohol. Medically, if a woman or man consumes one or two alcoholic drinks per day, respectively, they are over 21 years of age and not addicted, pregnant, otherwise ill, or taking medications that interact with alcohol, then their consumption is not classified as problematic.

Alcohol affects the system of neurotransmitters in the brain and has an inflammatory effect on living tissue. Once ingested, alcohol passes through the esophagus into the stomach and small intestine, where it is rapidly absorbed in the bloodstream. On an empty stomach, absorption of alcohol occurs in 40–60 minutes; foods high in fat content slow down this absorption. Due to differences in physiology, absorption into the bloodstream occurs more rapidly in women than in men; therefore, its effects are likely to be felt by females more quickly. Blood alcohol content is the concentration of alcohol in a person's blood stream; it is measured by mass per volume. The size and weight of the person drinking is a factor; larger, heavier people require greater amounts of alcohol to reach the same blood level percentage than smaller, lighter individuals (Bryant, Windle, & West, 1997; Schacht, 2006). As of July 2007, all 50 states in the U. S. have laws defining driving with a blood alcohol content at or above a level of 0.08% as illegal (Highway Loss Data Institute, 2007).

After alcohol enters the blood, it circulates throughout the body. The liver detoxifies (removes from the body) approximately 95% of the alcohol and the remaining 5% is excreted through the skin and lungs. Areas that are higher in water content (the liver, brain, pancreas, heart, and muscles) pick up more alcohol than do dryer tissues (bone). As the blood alcohol level rises, functions such as judgment, impulse control, and alertness are diminished. In part, the “high” that drinkers experience is due simply to a decrease in inhibition and a perceived relief from cares and worries.

As additional alcohol is consumed, blood alcohol level increases and further impairs one's ability to drive and perform other complex tasks. Researchers generally agree that for most people, impairment occurs when three or more drinks are consumed within an hour (Miller & Swift, 1999; Perez, 1992). Stimulant drugs (including caffeine) do not hasten the process. Annually, countless motor vehicle deaths involve drivers who have been drinking. Over the past 25 years, considerable efforts have been made to reduce the number of people driving “under the influence.”

## **Problematic Patterns of Use**

Problematic or unacceptable consumption of alcohol has been described in several ways, including misuse, abuse, dependence, and alcoholism (alcohol addiction). This section elaborates on these concepts to clarify the difference and identify approaches to treatment that are effective for various conditions.

### ***Use and Misuse***

Use is the occasional drinking of alcoholic beverages that does not result in significant impairment or adverse consequences. The “misuse” of alcohol includes drinking by minors, pregnant women (or those anticipating pregnancy), individuals using hazardous or moving machinery, and people driving motor vehicles. Additional misuse of alcohol includes drinking while engaged in any hazardous activities, drinking in the presence of an illness that contradicts the use of alcohol, and drinking in situations where it interacts with medications (Perez, 1992).

### ***Alcohol Abuse***

The National Institute on Alcohol Abuse and Alcoholism (NIAAA) of the National Institutes of Health (2003) defined alcohol abuse as a maladaptive pattern of drinking that leads to clinically significant impairment or distress. An individual diagnosed with alcohol abuse drinks despite alcohol-related physical, social, psychological, or occupational problems. Alcohol abuse does not necessarily involve a consistent

pattern of heavy drinking, but is defined by the adverse consequences associated with the drinking pattern. These include failure to fulfill major obligations at work, school, or home (e.g., repeated absences or poor work performance related to substance use, substance-related absences, suspensions or expulsions from school, neglect of children or household).

### *Alcohol Dependence*

The NIAAA (2003) defined alcohol dependence as being characterized by multiple symptoms including tolerance, signs of withdrawal, diminished control over drinking, as well as cognitive, behavioral, and/or physiological symptoms that suggest that the person continues to drink despite experiencing significant alcohol-related problems.

### *Alcoholism*

Another term for alcohol addiction is alcoholism. The American Society of Addiction Medicine (ASAM) and the NCADD (2007) defined alcoholism as a primary, chronic disease that is often progressive and can be fatal. In historical use, alcoholism refers to any condition that results in the continued consumption of alcoholic beverages despite negative personal and social consequences. NIAAA (2003) stated that alcoholism is characterized by the following four symptoms: (1) craving, i.e., a strong need or urge to drink; (2) loss of control, i.e., not being able to stop drinking once drinking has begun; (3) physical dependence, i.e., the occurrence of withdrawal symptoms, such as nausea, sweating, shakiness, and anxiety after stopping drinking; and (4) tolerance, i.e., the need to drink greater amounts of alcohol to get “high.”

**Primary** alcoholism is a term used to highlight alcoholism as an illness separate from and in addition to any other illnesses that may be associated with it. Primary also means that alcoholism, as an addiction or dependence, is not a symptom of another physical or mental illness.

**Progressive and fatal** alcoholism indicates that the illness persists over time and that physical, emotional, and social changes that result are cumulative. Tissue damage from alcoholism causes premature death due to disease of the liver, brain, heart, digestive system, and other vital internal organs. There is an increased risk of death or disability due to accidents, homicide, suicide, and other traumatic occurrences. **Impaired control** indicates an inability to limit alcohol use or the amount consumed when drinking, the duration of the episode, quantity consumed, and behavioral consequences of drinking. **Denial** includes a range of psychological maneuvers that limit or eliminate awareness that alcohol use is the cause of the problems. Alcoholics cannot recognize symptoms or results that seem perfectly clear to others (Galanter & Kleber, 1994; McCrady & Epstein, 1999).

## **Etiology**

There is no single cause of alcoholism; causes of alcohol abuse, alcohol dependence, and alcoholism are multifaceted. Research has indicated that a genetic component may play a role in the abuse of alcohol (Begleiter & Kissin, 1995; Goodwin, 2000; Schuckit, 2006). For example, studies of identical twins show the son of an alcoholic father is four times more likely to become alcoholic than the child of a non-alcoholic father. Female children of alcoholics have an increased incidence of alcoholism and a much higher incidence of severe depression. Research indicates that there are changes in brain waves of children of alcoholics similar to those found in their alcoholic fathers, and genetic influences in the pattern of adolescent alcohol tolerance.

Aside from possible genetic factors, environmental and cultural influences play significant roles in substance abuse (Alters, 2007). In general, in groups or populations where heavy use of alcohol is more accepted, a greater number of incidences of alcoholism are found. There is no evidence of “an addictive personality,” and no valid tests exist to predetermine one’s predisposition to addiction. Certain personality characteristics develop during addiction (Powers & Dell Orto, 2004). Family history of addiction and prior addiction to other drugs, including tobacco, are strong predictors of alcoholism (Ruben, 2001).

Today, the concurrent use of alcohol and other drugs is an increasing problem (Schuckit, 2006). Many people who drink use other recreational or prescription drugs including sedatives, stimulants, diet pills, and marijuana. Combining alcohol with such intoxicants can result in synergistic and additive



interactions, with a greater incidence of disruptive effects. When multiple substances are used, withdrawal and recovery are complicated, prolonged, and more likely to require professional care in the early period of recovery. Additionally, for people who use alcohol with other drugs, the risk of early relapse is high.

Rehabilitation counselors and other helping professionals must possess a clear understanding of a client's alcohol and other drug use. Current alcohol abuse affects motivation and impairs physical and emotional recovery from other illnesses. Frequently, the state of the person's alcohol use may not have previously been assessed or diagnosed. When alcohol abuse is suspected, assessment by trained personnel can determine the extent of the problem and whether it is concurrent with another illness (dual diagnosis).

## **Effects**

The primary negative effects of alcoholism occur with use of the substance in excessive and damaging amounts. The secondary damage caused by the inability to control alcohol consumption is manifested in many ways. It is common for a person with alcohol-related problems to drink well after physical health problems surface. Such negative effects include cirrhosis of the liver, pancreatitis, polyneuropathy, alcoholic dementia, heart disease, cancer, nutritional deficiencies, sexual dysfunction, and even death.

### ***Social Effects***

The social problems arising from alcoholism are significant (Holt, 2006). Inebriation or its counterpart, a "hangover," during hours of employment can result in job termination. This can lead to immediate or eventual financial problems, including the loss of living quarters (Corley, Lawton, & Gray, 2005). Behavior caused by reduced judgment from alcohol consumption can have legal consequences, such as criminal charges or civil penalties. An alcoholic's behavior and mental impairment while drunk can profoundly impact family and friends, possibly leading to marital conflict and divorce, or contributing to domestic violence. This can damage the emotional development of the alcoholic's children, either immediately or even after they reach adulthood.

### ***Medical Issues and Illness***

All human tissues are negatively affected by alcohol use and prolonged alcoholism can lead to a variety of illnesses (Bertha, 2006). Alcohol alters neurotransmission and gradually alters neuronal structure. Ingestion of alcohol leads to slowed reaction time, impaired judgment, and loss of inhibition. Balance, learning, memory, and cognition are affected. Blackouts (amnesia for events which transpired while intoxicated) are a serious and advanced sign of the negative effects of alcohol on the nervous system.

Prolonged alcohol abuse may lead to disease states affecting nearly every system of the human body. Alcohol's affect on the gastrointestinal system includes bleeding throughout the gastrointestinal tract (especially the esophagus and stomach), acute inflammation of the pancreas or pancreatitis, acute and chronic liver disease, gallbladder and bile duct disease, and ascites (swelling of the abdomen). The genitourinary system is affected by bladder and prostate problems. The cardiovascular system is affected by high blood pressure, irregular heart beats, and alcoholic cardiomyopathy which may result in heart failure. The lungs can be affected by aspiration pneumonia (when stomach contents end up in the lungs) and sleep apnea.

Alcohol-related disorders affect the nervous system and the emotions. Frequent alcohol-related problems include anxiety, loss of concentration, short-term memory loss, blackouts, increased headaches, sleep disorders, fatigue, impaired judgment, loss of impulse control, and depression (Brozner, 2006). Sexuality and intimacy are affected; hormonal and emotional changes affect sexual performance, as well as the ability to maintain relationships. Sexual "acting out" or compulsive sexuality sometimes occurs as a result of alcohol abuse and dependence. Skeletal system effects include osteoporosis and damage from falls and accidents, including fractures and burns. Many illnesses are caused or complicated by alcoholism, and recovery from illness becomes more difficult with its continued use (Miller & Swift, 1999).

## Diagnosis

Frequently, hospital records of the alcoholic include a diagnosis of alcoholism. Such a diagnosis, however, may not have been made or recorded prior to an interview by the rehabilitation counselor. Many screening devices and questionnaires have been developed for this purpose. A skilled interviewer may use any of these instruments to do an assessment. Alcoholics are not accurate in reporting their own alcohol use or its effects. Questions addressing symptoms and experiences rather than quantity consumed are likely to reveal a more accurate assessment. Use of the following **CAGE** interview items may be helpful to enhance non-judgmental questioning of the individual (Beresford, Blow, Hill, Singer, & Lucey, 1990; Ewing, 1984):

Have you ever **C**ut down on your drinking?

Do you get **A**nnoyed when your drinking is criticized?

Do you feel **G**uilty about your drinking?

Do you use alcohol as an **E**ye-opener? (A drink first thing in the morning to get going).

A single positive answer raises suspicion concerning alcohol abuse or dependence. Although alcoholics are not likely to be accurate or candid about the actual amount used, any individual who notes having more than 14 drinks in a week or more than five drinks on any one occasion, is positive for abuse or dependence. Historical information can also be obtained from the family or significant others.

The rehabilitation counselor is unlikely to see a person in acute withdrawal. This condition is characterized by symptoms including hallucinations or delusions, rapid pulse, increased agitation, anxiety, excessive sweating, hand tremor, nausea, and vomiting. An individual with these symptoms may require immediate emergency medical attention.

## Treatment

Treatments for alcoholism are varied. Those who approach alcoholism as a medical condition or disease recommend different treatments than those who approach the condition as one of social choice. In general, most treatments focus on helping people discontinue their alcohol intake. These treatments include life training and/or social support strategies to help them avoid a return to alcohol use. Since alcoholism involves multiple factors which encourage a person to continue drinking, each must be addressed individually to successfully prevent a relapse once drinking is stopped (Perez, 1992; Schuckit, 2006).

### *Detoxification*

Alcohol detoxification is only the first step in recovery. Although symptoms of withdrawal are treatable, addiction, which has both physical and mental characteristics, must be treated over an extended period of weeks or months. While some alcoholics can stop drinking for a period of time (days, weeks, or even months), alcoholic drinking generally recurs. Rarely, people who show signs of alcoholism are able to maintain prolonged abstinence without assistance (Stimmel, 2002). For most people alcoholism is a life-long problem and extensive treatment is required. Studies (O'Connell, 1998; Perkinson, 2004) have shown that a minimum of nine months is necessary for a stable recovery. Because a return to asymptomatic drinking is unlikely, the commitment to long-term abstinence is needed to prevent further alcohol-related problems. Sobriety is achieved by many through regular participation in a recovery program such as Alcoholics Anonymous (*Alcoholics Anonymous*, 2001; *The Twelve Steps and Twelve Traditions*, 1981).

The American Society of Addiction Medicine published an excellent document called the Patient Placement Criteria-II-R (PPC-II-R) (Gartner, Mee-Lee, & Shulman, 2001), which has been adopted by many states and several federal government agencies. The PPC-II-R outlines whether a person could be safely detoxified at home or needs a non-medical social model or a medically supervised program. Following detoxification, a client may require an ongoing structured living environment such as a recovery home. Utilization of these resources generally enhances recovery.

**Withdrawal** ranges from mild to severe (life threatening) and sedative hypnotics, such as the benzodiazepines and barbiturates, decrease the symptoms of withdrawal. When these drugs are not used, a process of "kindling" may take place in the brain, making it more sensitive to future withdrawal syndromes.

“Hangovers” are the mildest form of alcohol withdrawal syndrome. They usually consist of nausea, headache, thirst, and dysphoria (exaggerated feelings of depression and unrest). Additionally, tremulousness and irritability may be present. Moderate withdrawal symptoms include severe agitation, tremulousness, irritability, insomnia, difficulty concentrating, anxiety, and increased dysphoria. Symptoms arise as early as two to four hours after the last drink and reach a peak within 12 to 72 hours. Occasionally, tonic clonic (grand mal) seizures (usually one, but can be multiple) occur during this time.

The most severe form of the alcohol withdrawal syndrome is delirium tremens (DTs). This consists of delirium, increased pulse rate, elevated blood pressure and body temperature, and severe agitation. Delirium tremens has a mortality rate of about 10%; early and intense medical intervention is required. Cardiac arrhythmia can occur during withdrawal from alcohol. Treatment for all stages of alcohol withdrawal syndrome includes large doses of thiamine (Vitamin B1), sedative hypnotics, fluids, multivitamin supplements, and rest. Most people do not go through detoxification in hospitals but do so at home or in “social model” recovery homes. Symptoms of mild withdrawal usually last for two to three days; more severe withdrawal syndromes last much longer (Goodwin, 2000).

DTs usually begin three days after the last drink and can be severe for five to ten days. Seizures may occur for two weeks after the last drink, although they usually are experienced within the first three days. Multiple medications have been used during withdrawal to minimize prolonged abstinence syndrome and seizures; the use of thiamine and sedative hypnotics is the most effective (*Hatherleigh Guide*, 1996; Stimmel, 2002). The goal is to become free from all chemicals that have the potential for addiction.

### ***Medications***

Although not necessary for treatment of alcoholism, a variety of medications may be prescribed as part of treatment. Some may ease the transition to sobriety, while others cause negative physical symptoms when alcohol is used. In most cases, the desired effect is to help an alcoholic abstain from drinking (Brozner, 2006; Gray, 2004).

**Antabuse** (disulfiram) prevents the elimination of acetaldehyde, a chemical the body produces when breaking down ethanol. Acetaldehyde itself is the cause of many hangover symptoms from alcohol use. The overall effect is severe discomfort when alcohol is ingested, causing an extremely fast-acting and long-lasting hangover. These results are intended to discourage an alcoholic from drinking while in recovery. Heavy drinking while on antabuse can cause severe illness and result in death.

**Naltrexone** is a competitive antagonist for opioid receptors, effectively blocking a person’s ability to use endorphins and opiates. It also appears to act on glutamate neurotransmission. Naltrexone is used in two very different forms of treatment. The first treatment uses naltrexone to decrease cravings for alcohol, thus encouraging abstinence. The other treatment, called pharmacological extinction, combines naltrexone with normal drinking habits to reverse the endorphin conditioning that causes alcohol addiction. Naltrexone comes in two forms. Oral naltrexone is a pill and must be taken daily to be effective. Vivitrol is a time-release formulation that is injected once a month.

**Acamprosate** (also known as Campral) is thought to stabilize the chemical balance of the brain that would otherwise be disrupted by alcoholism. The Food and Drug Administration (FDA) approved this drug in 2004, stating “While its mechanism of action is not fully understood, Campral is thought to act on the brain pathways related to alcohol abuse...” (FDA Approves New Drug for Treatment of Alcoholism, 2006). While effective alone, it is often paired with other medications to improve success.

**Sodium oxybate** is the sodium salt of gamma-hydroxybutyric acid (GHB). It is used for both acute alcohol withdrawal and medium to long-term detoxification. This drug enhances GABA neurotransmission and reduces glutamate levels. *Baclofen* has been shown in animal studies and small human studies to enhance detoxification.

## **Recovery Process**

Detoxification treats withdrawal symptoms but does not resolve the illness of alcoholism or alcohol dependence (Carson-DeWitt, 2003). Rapid recovery usually occurs during the first week of abstinence manifesting in physical and cognitive improvements. Recovery is affected by factors such as severity and

duration of alcohol dependence, age, concurrent emotional and physical diseases, and intensity of medications used to help withdraw the person from alcohol. During the first three weeks of abstinence, cognitive abilities and emotional stability rapidly improve, and continue to do so at a slower rate for the first two years of abstinence. Cognitive improvement persists as long as five years and, at a lesser rate, for as long as ten years while brain tissue heals from injuries caused by alcohol.

## Alcoholics Anonymous

Alcoholics Anonymous (AA) is a fellowship of men and women who share their experiences of strength and hope with each other to resolve their common problem and help others recover from alcoholism (Gray, 2004). The only requirement for membership is a desire to stop drinking. There are no dues or fees for AA membership; it is self-supporting through voluntary contributions. Alcoholics Anonymous is not allied with any sect, religion, political group, organization, or institution. An AA member's primary purpose is to stay sober and help other alcoholics achieve sobriety (Cheever, 2004).

The 12 steps of AA were put into written form in 1938 and elaborated in 1981 (Cheever, 2004; *The Twelve Steps and Twelve Traditions*, 1981).

1. We admitted we were powerless over alcohol, that our lives had become unmanageable.
2. Came to believe that a Power greater than ourselves could restore us to sanity.
3. Made a decision to turn our will and our lives over to the care of God as we understood Him.
4. Made a searching and fearless moral inventory of ourselves.
5. Admitted to God, to ourselves, and to another human being the exact nature of our wrongs.
6. Were entirely ready to have God remove all these defects of character.
7. Humbly asked Him to remove our shortcomings.
8. Made a list of all persons we had harmed, and became willing to make amends to all of them.
9. Made direct amends to such people wherever possible, except when to do so would injure them or others.
10. Continued to take personal inventory and when we were wrong promptly admitted it.
11. Sought through prayer and meditation to improve our conscious contact with God as we understood Him, praying only for knowledge of His will for us and the power to carry that out.
12. Having had a spiritual awakening as a result of these steps, we tried to carry this message to alcoholics, and to practice these principles in all our affairs.

Of great importance in AA is service to other alcoholics and practicing a program of complete honesty, tolerance, and service in daily living. Recovery in AA is viewed as a life-long process, requiring consistent effort and regular involvement in the organization. AA does not profess to be for everyone or the only answer to the alcoholic illness. Its effectiveness as a recovery program is widely accepted by the helping professions, the judiciary, and AA is attested to by hundreds of thousands of sober alcoholics world wide (*Alcoholics Anonymous*, 2001; *The Twelve Steps and Twelve Traditions*, 1981).

## A Chronic Disease

Alcohol dependence is a chronic disease that is subject to relapse. A relapse should not be viewed as a treatment failure, but rather as a symptom of the illness. Though never desirable, relapses can be therapeutic in that, if properly handled, re-enforce the need for sobriety. With prolonged or repeated relapses, however, recovery becomes increasingly difficult. A person who has met the criteria for the illness of alcoholism rarely can return to moderate drinking on any consistent basis. Individuals recovering from alcohol dependence must abstain from alcohol totally to ensure the best recovery outcome.

### *Dual Diagnosis or Co-Occurring Disorders*

Individuals who experience a dual diagnosis are often faced with a wide range of psychosocial issues and may experience multiple interacting illnesses. The term "co-occurring disorders" is becoming a common term to refer to dual diagnosis, or co-occurring substance abuse disorders and psychiatric or emotional illnesses.



Dual Recovery Anonymous (an independent, self-help fellowship) defines “dual diagnosis” as meaning that an individual has two separate but very interrelated diagnoses:

1. A psychiatric diagnosis.
2. A substance abuse diagnosis, which may include both drugs and alcohol.

A dual diagnosis occurs when an individual is affected by both alcoholism and an emotional or psychiatric illness. Both illnesses may affect an individual physically, psychologically, socially, and spiritually. Each illness has symptoms that interfere with a person’s ability to function effectively, and interfere with relating to self. Not only is the person affected by two separate illnesses, but they interact with one another. The illnesses may exacerbate one another. Symptoms can overlap and even mask each other making diagnosis and treatment more difficult. An individual may sincerely try to recover from one illness and not acknowledge the other. As one neglects his or her mental illness, that illness may recur. This recurrence may, in turn, lead a person to feel the need to “self medicate” through alcohol use.

Although a person’s initial use of alcohol may have been a self-medicating drug for a physical or emotional disorder, once developed, alcohol dependence and alcoholism are classified as primary diseases. The presence of another illness or disorder may be masked by alcohol use, and the ability to diagnose or treat the other disease may not emerge for weeks or months after withdrawal from alcohol (O’Connell, 1998). Thus, alcoholism must be treated first.

With emergence of another disorder, additional treatment is required. If the additional disorder is psychiatric, it can be treated simultaneously with the alcoholism. A person diagnosed as having two disorders requires treatment for both. However, some emotional symptoms present in the first days or weeks of sobriety that appear to be symptoms of a separate emotional disorder may diminish or resolve with continued treatment for the alcoholism. As a result, counselors must remain alert for emerging symptoms of psychiatric disorders to diagnose and treat them effectively.

### ***Alcoholism as a Family Disease***

Living with a person who is alcohol dependent has powerful emotional repercussions. Families and close associates of individuals with this disease should seek support systems that provide information and counseling. ALANON, a group mutual-help program for adult relatives and friends of alcoholics, enhances such support. Similar 12-step oriented programs exist for children of alcoholics. If an alcoholic is not willing or able to become sober and maintain sobriety, support groups help the family to free themselves of the emotional bonds of guilt, hopelessness, and despair which are often felt by individuals connected to those with alcoholism (Brown & Lewis, 1999; Ruben, 2001).

Human resource departments frequently provide emotional and referral information about alcoholism. The local chapter of the National Council on Alcoholism and Drug Dependence, for example, provides referral to Alcoholics Anonymous, ALANON, and other recovery support programs, as well as information about addiction. Some universities maintain referral lists of local recovery agencies. Counselors need an awareness of the role of AA and other treatment programs; a visit to local programs for information, including referral criteria, is recommended to provide further support for clients in recovery.

## **Assessment for Rehabilitation Services**

Generally, individuals in treatment for alcoholism are not referred for vocational rehabilitation during the initial phases of treatment (detoxification period or during the first month of treatment) (Alters, 2007). Persons evaluated for vocational rehabilitation services should undergo an assessment procedure that is similar to that which is provided for other chronic illnesses and disabilities. Attention is given to physical and psychological limitations, work history, training and education, assessment of specific skills acquired, and areas of interest for employment. Many alcoholics have sporadic work histories, with numerous job and career changes. Some have a history of self-employment in the skilled trades or professions, which may have allowed for greater flexibility to accommodate periods of reduced functioning due to heavy drinking (*Hatherleigh Guide*, 1996). Certain jobs where alcohol and/or drugs are present, such as bartending and pharmacy technician, are contraindicated for people in recovery.



A proper assessment includes physical status, medical problems, psychosocial functioning, functional limitations, the nature of current treatment, and whether or not the individual is still drinking. If currently sober, questions need to be asked regarding efforts to remain sober. Many people with alcoholism have had experiences in treatment with varying degrees of success. Because alcoholism is a chronic disease, many recovering alcoholics do not achieve or maintain long-term sobriety on their first attempts (Barrett, 1996). Vocational rehabilitation may be deferred for persons who are repeatedly relapsing or unable to maintain sobriety.

During an assessment for rehabilitation services, special attention is given to attitudes toward work and career. Overall, persons in the early stages of recovery may not have the ability to make accurate self-assessments of their capabilities and limitations. Thus, the rehabilitation counselor and others involved with treatment and recovery play a key role in developing accurate evaluation regarding motivation to return to gainful employment.

## **Vocational Rehabilitation**

In the early stages of recovery many alcoholics need work levels that are less demanding, stressful, and potentially less rewarding than the employment performed when at maximum functioning. Focusing on the acquisition of appropriate work habits and a more stable work history are positive initial vocational rehabilitation goals for the recovering alcoholic; the specific job chosen is less crucial. Likewise, job finding and retention skills are an integral part of the vocational rehabilitation program. Changing attitudes toward work and maintaining employment is a considerable challenge for alcoholics during the first year or two of recovery. Because career progress can be initially slow and challenging, several years are often required before a person reaches his or her “maximum career success.” Alcoholics often display impatience with this slow progress in their careers. Work stability is a primary goal; the counselor needs to stress that work habits and attitudes have more to do with success than any specific experience in an occupation. Overall, maintenance of sobriety is considered the first priority (Brown & Lewis, 1999).

### ***Need for Retraining***

Because some individuals lack marketable vocational skills, they have difficulty obtaining work appropriate to their age and educational background. Once stable sobriety is achieved, job retraining is frequently needed. Many recovering alcoholics indicate considerable interest in returning to school to complete education they abandoned at an earlier time. As with anyone in society, the acquisition of marketable skills is essential.

Living and working without alcohol can be an anxiety filled and apprehensive experience. Once familiar activities may seem new and difficult. As a result, many alcoholics have lost confidence in their ability to adequately perform past work at any skill level. If one experienced failure at work, due in part to alcohol and drinking-related problems, there will be increased anxiety and stress related to return to work (Alters, 2007; Schuckit, 2006). From the beginning of the assessment through long term and stable employment at the highest level achievable, the counselor’s office is a safe place for the person to talk about fears and discuss realistic, attainable goals. Counselors provide assistance by placing prior experiences in the proper context. Honest feedback, along with encouragement from the counselor, is beneficial in correcting misperceptions. The person needs to view his or her occupation as only one part of recovery – which requires time and patience. Retraining can thus be a way to “ease into” a return to work.

### ***Vocational Rehabilitation and the Recovering Alcoholic***

Vocational rehabilitation counselors occupy a central role during recovery. To be of assistance, they must possess familiarity with the difficulties facing the recovering alcoholic. Counselors may become involved in assisting recovering alcoholics at any point in the recovery process following initial detoxification, depending on the setting where recovery takes place. Hospital-based programs, for example, may either have a rehabilitation counselor on staff or refer clients for state-provided vocational rehabilitation services. Similarly, non-medical or social model recovery programs, such as 12-step oriented recovery homes, have staff trained to provide vocational exploration and counseling (Schuckit, 2006).

When the client's identified disabilities do not include alcoholism, but whose current drinking affects abilities to maintain employment, the counselor may have to take the lead in addressing the alcohol use, although it is not an "identified disability." As a result, it is necessary for the counselor to acknowledge the reluctance a person has in facing the recovery process. In some cases, a formal "intervention" is required, which results in a structured confrontation by several persons who are closely involved with the individual (*Hatherleigh Guide*, 1996).

### ***Untreated Alcoholism in Disability Clients***

Persons with active alcohol problems are more likely to have industrial accidents and chronic illnesses that lead to disability. The identified disability may not include the alcohol abuse or dependence – which is often undiagnosed or hidden. Additionally, the pain and injury associated with some illnesses and injuries can lead to excessive use of pain medications which may then lead to an increased use of alcohol. If undiagnosed alcoholism is present and not appropriately treated, it presents a virtually insurmountable obstacle to successful rehabilitation. When a counselor suspects alcohol abuse or dependence, directly confronting the client may not be helpful (Alters, 2007). The person will likely deny or minimize the extent of use of alcohol or pain medication, unable to see any connection between the demands of rehabilitation and the use of drugs. Assessment by a professional qualified in addiction counseling is helpful in this situation; the rehabilitation counselor should seek out an appropriate referral early in the process. Community resources, such as the local chapter of the National Council of Alcoholism and Drug Dependence, are available for referral information (Leonard & Blane, 1999).

## **Functional Limitations**

Physicians knowledgeable about addictions can help determine what residual functional limitations, emotional and physical, are present in individuals who are recovering from substance abuse (Bryant et al., 1997). The rehabilitation counselor needs to assess a person's (a) ability to work effectively both with others and with supervisors; (b) limitations due to mood or emotional disorders ("dual diagnosis") (O'Connell, 1998); (c) ability to work independently without close supervision; and (d) capacity to tolerate stress.

Some late-stage alcoholics become overly dependent on others and are unable, at least for a time, to work independently. These individuals can benefit from more structured work settings to make the successful transition to gainful employment. The recovering person also faces a decreased tolerance for stress during the first months of return to work, and this may continue for up to two years of sobriety. Employment involving low stress during the early months of a person's sobriety is likely to result in a more successful transition. Recovering alcoholics, however, may feel irritated at the necessity of performing work "beneath" their self-perceived capabilities. Positive feedback, like reassurances that this process is necessary, is beneficial for the person's recovery.

Though complete recovery from alcoholism leaves few specific functional limitations, the recovery to maximum functioning is slow and gradual, occurring over several years. A return to productive employment improves one's overall recovery by enhancing self-esteem and beginning the process of resolving financial difficulties (Brozner, 2006). It is for this reason that some recovery programs emphasize an early return to some type of work, generally beginning after one month of participation and sobriety. This is standard practice in alcohol recovery home settings.

## **Rehabilitation Potential**

Assessment of the potential for rehabilitation is a challenging task for counselors. Clearly, many factors affect rehabilitation potential including age, educational background, prior work history, functional limitations, and motivation to work. Though not always immediate, most recovering alcoholics are eventually able to return to productive employment. Factors likely to affect the potential for effective participation in vocational rehabilitation include (a) length of sobriety and active participation in a recovery program; (b) history of relapses and their frequency; (c) commitment to maintaining sobriety; and (d) residual functional limitations (physical, cognitive, and emotional) resulting from the alcoholism or other chronic conditions (Goodwin, 2000; Carson-DeWitt, 2003). Once sober, the recovering alcoholic needs to

demonstrate the same traits as any other person in rehabilitation, including willingness to accept responsibility for behavior, personal well-being, capacity to meet vocational expectations, and personal determination to succeed.

Most alcoholics demonstrate moderate rehabilitation potential and eventually return to gainful employment because of financial necessity. Functional limitations diminish over time with continuous sobriety and active participation in an on-going recovery program. Those with repeated arrests for more serious legal charges present special challenges for rehabilitation; yet, there are surprising, unexpected recoveries. Individuals on probation or parole need coordinated efforts with the legal system.

### ***Emotions and Attitudes Affecting Vocational Rehabilitation***

Paradoxically, the alcoholic who has been seeking a “high” from drinking often experiences intense, negative emotions as consequences of prolonged drinking. These feelings include depression, guilt, exaggerated fearfulness, anxiety, intense loneliness, and feelings of emotional isolation (Perkinson, 2004). Frequently, some of these intense negative emotions are present in the early months of sobriety, and may recur periodically for many years, with reduced frequency and intensity. During the first months of recovery, coping with these intense emotions can be extremely difficult. Participation in recovery programs that emphasize interpersonal interactions is most effective in regulating the emotional extremes.

Alcoholics use alcohol to block out or sedate negative perceptions and feelings, especially anger, resentment, and fear (Brozner, 2006). Because these emotional responses are not “processed,” the person may experience alienation from self and others. Newly sober alcoholics often display child-like responses to precarious situations and lack adult maturity and emotional balance. Impatience, low frustration tolerance, and explosive or excessive emotional reactions are common (*Hatherleigh Guide*, 1996).

Rehabilitation counselors need to understand and be familiar with the difficulties faced by recovering alcoholics. They may test the patience of most counselors by being manipulative and untruthful. Alcoholics in recovery are often highly sensitive to feeling “judged.” Clearly, appropriate counseling precludes being judgmental. A recovering person often responds to negative, judgmental attitudes in a strong way and becomes angry, wishing to withdraw from the situation (Frances, Miller, & Mack, 2005). This response requires the counselor’s intervention, suggesting alternative ways to deal with frustration, anger, and anxiety.

Improvement in coping with emotions and negative attitudes is noticeable within the first few months of recovery, though more permanent changes require time (*Alcoholics Anonymous*, 2001). Although progress is noticeable early in recovery, it is unrealistic to expect a person with many years of alcoholism to be fully recovered in a few weeks or even several months. The counselor must regularly acknowledge the long-term nature of recovery. Additionally, when depression or severe anxiety persist, or other signs of emotional illness evolve, referral to a mental health professional experienced in substance abuse counseling is appropriate.

## **Conclusion**

Alcohol consumption has negative consequences for not only the health and well-being of the client, but also the health and well-being of those around him or her. Alcohol addiction is common and costly while imposing a negative impact on all aspects of the individual’s life. Alcoholism is a disease in a sense that once an alcoholic drinking pattern is present, if left untreated, the severity and consequences of drinking are generally progressive and sometimes fatal. Although the effects vary widely, detoxification from alcohol is the first step to recovery. The desired goal is to abstain from drinking and maintain sobriety. Rehabilitation counselors need to understand and familiarize themselves with the process, stages, and difficulties facing both recovering alcoholics and clients whose current drinking affects the ability to maintain employment or return to work. Appropriate interventions and holistic supports are the keys to rehabilitation for individuals with alcohol-related disorders.

## Case Study

Sally, a 32 year-old woman, is currently undergoing detoxification at a local alcohol treatment center. Two previous attempts to become sober and maintain long-term sobriety have failed. She is married, although separated, and has two children, three and six years of age. The present attempt at sobriety was prompted by the departure of her husband as a result of her continued drinking behaviors. The children are living with her mother in a nearby city.

There is a history of alcoholism in Sally's family. Her father was a diagnosed alcoholic and died at age 47, the result of physical complications resulting from alcoholism. The mother's present husband was previously married to an alcoholic. Sally states that her parents are not sympathetic or supportive of her sobriety. Her parents indicate that they are willing to provide care for the children, but that Sally is not welcome in their home when she is drinking.

Drinking did not become a problem for Sally until she became an adult, although she began drinking at about the age of 12, drinking secretly from her father's stock of alcohol and enjoying the feelings she experienced. Drinking continued in high school. Once she was expelled from a school dance due to drinking. Serious trouble from alcohol consumption began during her freshman year in college; Sally experienced problems in classes due to absences. She had to move from her apartment on three occasions when her roommates refused to put up with her drinking. College life thus became impossible and Sally dropped out after the second term.

At the age of 20, Sally secured employment with a ticket agency, where she met and married a band musician. Six months later, she was divorced, again the result of her drinking and socially unacceptable behavior. She enrolled in an alcohol treatment program at the time of the divorce but left the program after five days. Continuing to drink, she was eventually fired from her job and returned home to live with her parents under the condition that she seeks help for her drinking problem. She attended several meetings at a women's center and enrolled in an outpatient treatment program at a local hospital. She continued participating in the outpatient program for three months, but stopped when she married her current husband, a produce manager at a local supermarket.

Sally controlled her drinking for the next eight years, during which time she gave birth to two children. During these years, she worked part-time at a local printing shop doing general office work. When she was 30, drinking again became a serious problem for Sally. While at home, she would drink secretly, but her practices soon became obvious to all around her. On several occasions, she would leave the children unattended to stay out all night. She lost her job, and the week after her 31<sup>st</sup> birthday, her husband left after she had destroyed much of the furniture at home in a drunken rage. Sally's parents took her and the children home and again insisted that she enter some type of treatment program.

Alcoholics Anonymous (AA) became Sally's support system; she attended meetings regularly but continued to drink. Her mother and stepfather eventually told Sally that she could no longer reside with them but that they would continue to care for the children until she regained the capacity to care for them. Sally was angry, frustrated, and hurt, but enrolled at a county detoxification center where she is currently seeking help from a counselor.

### Questions

1. Identify any factors which might have contributed to Sally becoming an alcoholic.
2. What are the positive factors in this case for rehabilitation?
3. As Sally's counselor, identify the first three measures you would recommend.
4. In your intervention, how would you involve Sally's significant others?
5. What types of intervention would you suggest for this individual?

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## Chapter 33

# ILLICIT AND PRESCRIBED DRUG USE

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## Introduction

Substance abuse is a serious problem that affects all strata of society. Escalating costs prompt governmental and nongovernmental agencies and organizations to enact laws and establish policies that attempt to reduce and minimize the adverse effects of substance abuse.

Many of these efforts center on how the problem of substance abuse is viewed or defined. For example, the Americans with Disabilities Act (ADA) of 1990 qualifies a person with a history of illegal drug use as having a disability when the individual has successfully completed or is participating in a supervised rehabilitation program (U. S. Equal Employment Opportunity Commission, 1997). In addition, the American Psychiatric Association (2000) included substance abuse and dependence as a mental disorder in the *Diagnostic and Statistical Manual of Mental Disorders IV – Text Revision (DSM-IV-TR)*.

At an individual level, consequences from substance abuse can lead to physical, emotional, educational, and vocational dysfunction and disability. To effectively assist individuals who have substance abuse problems to maximize their functioning, independence, and well being, it is necessary to understand the basic concepts of substance abuse. This chapter provides a general overview of substance abuse which includes definitions, prevalence, pharmacology, etiology, drug categories, treatment, and vocational implications.

## The Disease/Problem

The consumption of mood-altering substances is not a new phenomenon but one that has existed since prehistoric times. The opium poppy was cultivated by the Sumerians in lower Mesopotamia as early as 3000 B.C. (Stimmel, 2002). Around the same period, coca leaves were being ingested by people in South America before the rise of the Inca Empire (Office of National Drug Control Policy [ONDCP], 2006b). Cannabis seeds have been traced in Chinese food as far back as 6000 B.C., while its use as a medicine occurred around 2700 B.C. (ONDCP, 2006a)

Although many substances may have origins in religious or medicinal uses, some are cultivated or synthetically manufactured for their mood-altering qualities. This affect on mood often promotes substance abuse by a large number of people. This chapter focuses on these substances, their effects, and the consequences of their abuse.

## *Patterns of Use*

Terminology for patterns of substance use and their definitions are not consistent in the field of substance abuse treatment (Goode, 2008). Depending on the approach to treatment, terms or labels are applied to the pattern of drug use and the individual. For example, Alcoholics Anonymous (AA) utilizes a disease model and labels the pattern of use as an “addiction” and the individual as an “addict” whereas other treatment models identify the pattern of use as “abuse” or “dependence” and the person as having an “abuse” or “dependence” disorder. There are ongoing disagreements on the utility of placing a label such as “addict” on individuals due to the negative reactions that such labels elicit based on existing social values. Despite these disagreements, there is an effort to establish uniform terms when referring to patterns of substance use.

The American Medical Association has presented a ten-stage continuum of drug use which includes: nonuse (abstinence), appropriate use, misuse, experimental use, abuse, habituation, dependency, addiction, withdrawal, and craving (Stimmel, 2002). In addition, the American Psychiatric Association (2000) utilizes the categories of abuse, dependence, intoxication, and withdrawal in the diagnosis of mental disorders associated with substance use.

**Nonuse and abstinence** is defined as not using a drug, with abstinence implying that there had been use but the individual is no longer using. **Appropriate use** is using a drug in an accepted or approved manner recommended by the manufacturer. An example is taking prescription drugs as instructed by a physician. **Misuse** is taking a drug in a manner that varies from its prescription, for example, taking more sleeping pills than prescribed in an effort to increase its effects. **Experimental use** is the unintentional misuse or abuse of a substance. This is the most common initial usage by most people, particularly adolescents. The assumption in experimental use is that a substance is used a minimal number of times. **Abuse** is the intentional and inappropriate use of a drug or drugs that results in harm in one, or more than one, areas of human functioning. These areas can include but are not limited to physical, emotional, legal, economic, and social harm (Stimmel, 2002).

**Habituation** results when continued use of a drug is needed to avoid the feelings of anxiety that result when not taking the drug. A person who drinks coffee every morning is an example. **Dependency** is the continued need to take a substance for its euphoric effects with the development of physical and/or psychological changes when drug use is discontinued and the development of tolerance to the drug. **Physical dependence** occurs when the body adapts to a drug and cannot function normally without it. When the drug is discontinued, the person will have withdrawal symptoms. **Psychological dependency** is the emotional state of craving a drug for its euphoric effects or to avoid the feelings of discomfort caused by not taking the drug. **Tolerance** is the physical adaptation to the effects of a drug where it takes larger doses to get the same effects. **Addiction** is the compulsive use of a drug with loss of control (cannot stop using) and continued use in spite of negative consequences (Stimmel, 2002). As with drug dependency, drug addiction usually includes drug tolerance and withdrawal symptoms. The most widely used diagnostic categories are dependency and abuse as defined by the *DSM-IV-TR*.

### ***Prevalence and Incidence***

The Substance Abuse and Mental Health Services Administration (SAMHSA) (2005) reported in the National Survey on Drug Use and Health (NSDUH) that in 2004, 19.1 million Americans (7.9 percent of population) age 12 and older identified themselves as current users of illicit drugs. Marijuana was the most widely used illicit drug with 14.6 million current users followed by cocaine users at 2.0 million. NSDUH reported for 2004 that 6.0 million persons were current taking psychotherapeutic drugs for non-medical purposes. The most commonly used psychotherapeutic drugs were pain relievers at 4.4 million, tranquilizers at 1.6 million, stimulants at 1.2 million, and sedatives at 0.3 million.

In relation to employment, 19.2 percent of unemployed adults age 18 and older reported being current illicit drug users, compared to 8.0 percent of those employed full time and 10.3 percent employed part-time. Of 16.4 million illicit drug users age 18 and older, 12.3 million were employed. In the area of treatment, 3.8 million people age 12 and older received treatment in the past year for drug or alcohol use problems. Of these, 2.3 million received treatment at a specialty facility for substance abuse. It is estimated that 21.1 million people needed but did not receive treatment (SAMHSA, 2005).

### ***Protective and Risk Factors***

Prevention research on drug abuse has concentrated on adolescents between the ages of 12 and 18, as these are the years of highest risk for initiating drug use (Goode, 2008). The area of drug abuse prevention with adolescents has identified protective factors as well as risk factors. This research has found the following critical factors.

Protective factors:

- strong and positive family bonds;
- parental monitoring of children's activities and peers;

- clear rules of conduct that are consistently enforced within the family;
- involvement of parents in the lives of their children;
- success in school performance; strong bonds with institutions, such as school and religious organizations; and
- adoption of conventional norms about drug use.

Risk factors:

- chaotic home environment, particularly those in which parents abuse substances or suffer from mental illnesses;
- ineffective parenting, especially with children who have difficult temperaments or conduct disorders;
- lack of parent – child attachments and nurturing;
- inappropriately shy or aggressive behaviors in the classroom;
- failure in school performance;
- poor social coping skills;
- affiliations with peers displaying deviant behaviors; and
- perceptions of approval of drug-using behaviors in family, work, school, peer, and community environments (National Institute on Drug Abuse [NIDA], 2005, pp. 1-2).

This research suggests that an interaction between protective factors and risk help to predict the probability of a person’s likelihood of engaging in social, nonconforming behaviors. Strong and positive bonds with family and others appear to be a mitigating factor in preventing substance abuse.

## **Pharmacology**

The most commonly abused substance or “drug of choice,” as they are often referred to, changes over time. Their use and abuse is dependent on various factors including availability, cost, legal status, social status, and expected effects (Goode, 2008). Consequences from abuse of drugs may result in barriers to successful vocational rehabilitation. This section presents substances that are currently abused by major categories such as stimulants, hallucinogens, depressants, cannabis, and nicotine. Alcohol is not included in this discussion as it is presented in detail in another chapter. The pharmacology of the selected substances includes their source, appearance, actions, and uses. In addition, pharmacokinetics (how the body responds to drugs) and psychopharmacology (interaction between psychoactive drugs and the mind) are included in this section.

### ***Controlled Substances Schedules***

The Controlled Substances Act (administered by the Drug Enforcement Administration [DEA] within the U. S. Department of Justice) established a system for classification of all substances that are regulated by federal law. Substances are placed into one of five schedules based on harmfulness, medical use, and potential for abuse or addiction (See Table 1) (Drug Enforcement Administration, 2006).

## **Drug Categories**

### ***Stimulants***

Stimulants are a group of drugs that increase activity in the central nervous system (CNS). When taken in moderation stimulants enhance mood, increase alertness, and relieve fatigue (Abadinsky, 2004). Two extremely powerful stimulants discussed are cocaine and amphetamines. Both these drugs have devastating physiological and psychological effects when they are abused.

**Table 1  
Controlled Substances Schedules I through V**

<b>Schedule</b>	<b>Criteria</b>	<b>Substances/Drugs</b>
I	a) high potential for abuse b) no current accepted medical use in United States c) lack of accepted safety	heroin, LSD, marijuana, MDMA, mescaline, peyote, Psilocybin, GHB
II	a) high potential for abuse b) current accepted medical use with severe restrictions c) abuse of substance may lead to severe physical or psychological dependence	morphine, cocaine, methamphetamine, LAAM, methadone, oxycodone, PCP, Oxycontin
III	a) potential for abuse less than the substances in Schedules I and II b) current accepted medical use in treatment in United States c) abuse of the substance may lead to moderate or low physical dependence or high psychological dependence	codeine, anabolic steroids, Rohypnol, Buprenorphine
IV	a) low potential for abuse relative to substances in Schedule III b) current accepted medical use in treatment in United States c) abuse of the substance may lead to limited physical dependence or psychological dependence relative to substances in Schedule III	Benzodiazepines Valium, Xanax, Ativan
V	a) low potential for abuse relative to substances in Schedule IV b) current medical use in treatment in United States c) abuse of the substance may lead to limited physical dependence or psychological dependence relative to substances in Schedule IV	codeine mixtures

Cocaine (street names: coke, snow, blow) has had a long history in our civilization. It has been used for several purposes including medicinal, spiritual, and recreational. Currently, it is a Schedule II drug. Cocaine is derived from the leaves of the coca plant producing cocaine hydrochloride (powder) and its freebase form, crack-cocaine.

Cocaine affects the neurotransmitter dopamine by increasing activity in dopamine receptors and blocking its reuptake as well as blocking the reuptake of norepinephrine and serotonin (Kalat, 2001). The resulting experience for a cocaine user is an intense feeling of euphoria and increased energy. At larger doses, physiological effects include elevated blood pressure, slower digestive process, seizures, and increased body temperature. Psychological effects include mood swings, depression, paranoia, and hallucinations. The intensity and duration of the effects of this drug depend on the form that is utilized and the route of administration. When cocaine is injected or inhaled the effects are instantaneous but the pleasurable feelings described last for a short period of time, usually 30 to 40 minutes. If snorted, the effects occur within a few minutes and last for 60 to 90 minutes (Goldberg, 2003).

Unlike cocaine, amphetamines are synthetic drugs formed in a laboratory. They have been medically used to treat asthma, anxiety, depression, obesity, hyperactivity, and narcolepsy (Abadinsky, 2004; Goldberg, 2003). As with cocaine, amphetamines are classified as Schedule II drugs. Methamphetamine (meth, crystal meth, ice), a derivative of amphetamine, is increasing in use as a drug of choice for abuse. This drug has become popular because it is easily manufactured and the effects or high last up to 12 hours compared to 15 to 30 minutes for cocaine. Methamphetamine produces a greater “high” than other amphetamines. As with other drugs, the repeated use of amphetamines for medicinal or recreational purposes can lead to abuse and dependence.

Once amphetamines enter the body, they affect the reuptake of the neurotransmitters dopamine and norepinephrine. Similar to cocaine, this effect causes an individual to feel euphoria and increased energy. Repeated use of amphetamines cause physiological symptoms such as tremors, high blood pressure, and



convulsions (Goldberg, 2003). Psychological effects of repeated amphetamine use include anxiety, obsessions, paranoia, agitation, and hallucinations.

### ***Cannabis***

Cannabis (marijuana, pot, weed, grass, hashish, hash, rope) is a sedative-hypnotic drug and is the most abused illegal drug in the United States. The rates of use range from 14.6 million current users to 96.7 million lifetime users who are 12 years of age or older (SAMHSA, 2005). Cannabis comes from the hemp plant with the dried leaves, stems, seeds, and flowers producing marijuana and the resin from the top of the plants producing hashish (Stimmel, 2002). The active psychoactive agent in cannabis is delta-9-tetrahydrocannabinol, commonly known as THC (Goode, 2008). It is THC that produces the effects on the body, affecting the neurotransmitters norepinephrine and dopamine.

Marijuana and hashish are smoked using joints (cigarettes, bong, and pipes) or ingested by mixing with food. When marijuana or hashish is smoked, THC is absorbed into the blood stream through the lungs to the brain. The absorption process takes seconds with physiological and psychological effects occurring within a few minutes and feelings of euphoria lasting from one to three hours. In contrast, when cannabis is ingested, the effects are not experienced for a few hours due to slower absorption through the digestive system. The slower absorption rate allows the effects to last up to 12 hours (Goldberg, 2003).

Cannabis causes several physiological and psychological effects. Physiological effects include dilatation of peripheral blood vessels resulting in blood shot eyes, increased heart rate, decrease in testosterone levels for men, disrupted menstrual cycles for women, and increased respiratory problems. Psychological effects include a sense of euphoria, increased appetite, and a feeling of being “in tune” with oneself. Other effects may include anxiety, delusions, disorientation, and hallucinations.

### ***Hallucinogens***

Hallucinogens are drugs that cause changes in a person’s perception of reality. Approximately 929,000 persons 12 years of age and older are current users of hallucinogens (Goode, 2008). Hallucinogenic substances are found in plants as well as synthetically produced. Hallucinogens stimulate serotonin receptors that affect mood and emotions (Kalat, 2001). The most commonly abused hallucinogens are lysergic acid diethylamide (LSD), phencyclidine (PCP), psilocybin (mushrooms), and mescaline (peyote).

LSD (acid, dots, sugar cubes) is considered the most powerful conscious-altering substance and a Schedule I drug (Goode, 2008). Users experience both physiological and psychological effects. The effects of the drug are felt within 30 to 90 minutes after ingestion and may last 8 to 12 hours. Physiological effects include increased blood pressure and heart rates, dizziness, nausea, sweats, loss of appetite, dry mouth, and tremors. The major effects are psychological with changes in mood, thoughts, and perceptions. Mood may rapidly shift from fear to euphoria and back. Perceptual effects are experienced where colors, smells, sounds, and other sensory stimuli are dramatically intensified. Users of LSD report having both pleasant and unpleasant experiences referred to as “acid trips.”

Tolerance to LSD does not last long and the user returns to normal functioning after a few days of not using the drug. Although no withdrawal symptoms are reported, there are two long term effects associated with LSD use: hallucinogen persisting perception disorder (HPPD) and persistent psychosis. HPPD is more commonly known as “flashbacks” and can occur after a single use of the drug. Drug induced persistent psychosis can include mood swings, visual disturbances, and hallucinations that may last for years (NIDA, 2001a).

PCP (angel dust, zombie, rocket fuel, peace) is a white crystalline powder that is usually sprinkled on marijuana, tobacco, and oregano and smoked. It is also taken orally in tablet form or snorted. PCP causes both physical and psychological effects that are felt within minutes of administration and can last for several hours. Physical effects include rapid breathing, increased blood pressure, increased body temperature, nausea, dizziness, and blurred vision. Psychological effects include detachment from reality, hallucinations, and feelings of invulnerability and increased strength. PCP is addictive with reported psychological dependency and withdrawal symptoms with prolonged use.

Two types of hallucinogenic substances derived from plants are psilocybin and mescaline (peyote). There is no evidence of dependency for psilocybin but tolerance can occur over a short period of time. Mescaline is the active hallucinogenic substance found in peyote. Although there is no report of dependency with mescaline, tolerance develops rapidly with repeated use (ONDCP, 2006c).

### ***Depressants***

Drugs that depress the CNS are categorized as depressants and are sub-divided into sedative hypnotics and narcotic analgesics. SAMSHA (2005) reported that, in 2004, approximately 6.7 million persons 12 year of age and older were current users of some type of depressant, not including alcohol. Sedative hypnotics include alcohol, barbiturates, benzodiazepines, and gamma-hydroxybutyrate (GHB). Narcotic analgesics presented in this section include opium, morphine, heroin, codeine, and Oxycotin.

#### ***Sedative Hypnotics***

These depressants have been used medicinally to relieve anxiety and stress and to promote sleep. While they have significant benefits when used as prescribed, sedative hypnotics are frequently abused with a high potential for development of tolerance and dependence (Goode, 2008).

Benzodiazepines have less potential for abuse and are the leading group of medications used to treat anxiety. Although there is a lower potential for abuse, users may become dependent or addicted to these medications over time. Examples of benzodiazepines are Valium, Xanax, Ativan, and Centrax. These drugs are categorized as schedule IV medication.

Two drugs included in this category that have emerged as “club drugs” or “date rape drugs” since the 1990s are gamma-hydroxybutyrate (GHB, Liquid E, Liquid X) and Flunitrazepam (Rohypnol, Roofies, R-2). The two drugs have been associated with drug facilitated sexual assaults due to their anesthetic producing qualities. At large enough doses, victims become incapacitated and are unable to consciously reject sexual advances. GHB is categorized as a Schedule I drug while Rohypnol is categorized as a Schedule III drug. The DEA is evaluating the possibility of reclassifying Rohypnol as a Schedule I controlled substance (ONDCP, 2003b).

#### ***Narcotic Analgesics***

These substances relieve pain and produce euphoria by reducing anxiety and inducing relaxation. The term analgesic means to relieve pain. Narcotics are referred to as “opiates” or “opioids.” Opiates are substances that are naturally occurring derivatives from opium while opioids are produced synthetically. Narcotics act on the CNS by stimulating the endorphin receptor sites that regulate pain and emotions, thus relieving pain and causing euphoria (Kalat, 2001).

Opium is a naturally occurring substance that is extracted from certain poppy plants. Its derivatives include morphine and codeine while synthetic opioids include heroin, methadone, and oxycodone (Goode, 2008). With the exception of heroin, a Schedule I drug, most narcotic analgesics are Schedule II drugs with accepted medical use. In terms of abuse, this group of drugs has a high potential for tolerance and dependence due to their strong euphoric effects. They also produce withdrawal symptoms with discontinued use.

Morphine and codeine are derivatives of opium that have been medically used to reduce pain and suppress coughs since the 1800s. Morphine is the more potent of the two derivatives and has a higher potential for abuse (Drug Text Web Lab, 1995).

Heroin is synthesized from morphine and is considered the fastest acting narcotic. It is usually injected but can be snorted and smoked. More heroin users are smoking or snorting it due to increased health risks associated with sharing needles, such as HIV transmission. Heroin crosses the blood brain barrier quickly, creating the feeling of euphoria or “rush” within seconds of being injected. This quick acting effect makes heroin the narcotic with the highest potential for tolerance and dependence. Withdrawal symptoms from heroin include muscle and bone pain, nausea, vomiting, restlessness, sweating, and tremors. These severe symptoms can be stopped or reversed by re-administration of heroin or other narcotics. This phenomenon contributes to the high incidence of relapse for heroin users seeking treatment or attempting to

quit on their own. A treatment of choice for many chronic heroin and narcotic users is methadone (Drug Text Web Lab, 1995; ONDCP, 2003a).

### *Inhalants*

Inhalants are commonly found in household and commercial products. These substances are abused by inhaling them through the mouth or nose (“huffing” or “sniffing”) to produce intoxicating effects. Examples of inhalants are lighter fluids, adhesives, paint, correction fluid, and nitrous oxide; children and adolescents are the most common users. The danger of inhalants is that they are readily available to children and inexpensive to purchase. Prolonged use can result in damage to the kidneys and liver, or even death from asphyxiation (McNeece & DiNitto, 2005).

### *Nicotine*

Nicotine, one of 4,000 chemicals found in smoke of tobacco products, is the primary compound that affects the CNS. Tobacco is smoked, chewed, or snuffed. Nicotine stimulates nicotinic receptors within the CNS which in turn increases the release of dopamine. Increased dopamine levels appear to be involved in mediating feelings of pleasure and the desire to consume drugs. This suggests that nicotine has a high potential for abuse and addiction; hence, the number of individuals that attempt to reduce or stop cigarette use and fail. It is reported that approximately 35 million smokers make attempts to quit each year but less than 7% achieve abstinence for more than one year (NIDA, 2001b).

An estimated 70.3 million Americans aged 12 and older reported current use of tobacco products (SAMHSA, 2005). The most common method of tobacco use is cigarette smoking. NIDA (2001b) reported the following:

“Cigarette smoking is associated with coronary heart disease, stroke, ulcers, and an increased incidence of respiratory infections. It is the major cause of lung cancer and is also associated with cancers of the larynx, esophagus, bladder, kidney, pancreas, stomach, and uterine cervix. It is the major cause of chronic bronchitis and emphysema” (p.1).

## **Etiology**

Research has identified two general conceptual approaches in the etiology of drug abuse. “One major direction focuses on psychological/behavioral and social/environmental variables and their interaction leading to the abuse of psychoactive substances. The second approach focuses on the neurobiological mechanism affecting, or affected by, the chemical actions of drugs” (Gordon & Glantz, 1996, p. 1).

Interactions between personality and environmental factors are important determinants in the development of problem drug use. The behavioral/environmental approach attempts to identify characteristics in people that may lead to drug use and abuse. This approach has helped to identify risk factors as well as protective factors to drug abuse (discussed previously in this chapter).

Risk factors have generally focused on understanding variables that may contribute to the onset of drug taking behaviors in adolescents. These variables are subdivided into individual (personality traits), family, school, and peer factors. Goldberg (2003) identified the following personality characteristics: low self-esteem; poor interpersonal skills; need for immediate gratification; defiant feelings toward authority; little tolerance for anxiety, frustration, and depression; impulsivity; risk-taking; and a low regard for personal health.

**Family factors** identified as critical by the NIDA (2005) include: chaotic home environment, particularly in which parents abuse substances or have mental illnesses; ineffective parenting, especially with children who have difficult temperaments or conduct disorders; and lack of parent–child attachments and nurturing. **School factors** include inappropriately shy or aggressive behaviors in the classroom and failure in school performance. Additionally, there is a low commitment to education as a factor related to drug use (McNeece & DiNitto, 2005). **Peer factors** are some of the strongest predictors of adolescent drug use. Affiliation with peers that engage in deviant behaviors is a critical risk factor identified by NIDA. Positive bonds to family and school decrease the potential for involvement with drug using peers.

While the personality/environmental approach has resulted in identifying risk factors, the neurobiological approach has helped identify drug receptor sites and the mechanism of action for various drugs, thus laying the foundation for the development of pharmacological treatments for drug abuse. The most well known pharmacological treatments are methadone for heroin dependency, and disulfiram (brand name Antabuse) for alcohol dependency.

## **Treatment**

Prevention and intervention strategies are primary, secondary, and tertiary interventions that target all drug usage patterns (Goode, 2008). Primary prevention targets nonusers and appropriate users through educational programs aimed at preventing development of drug abuse. Secondary prevention targets abuse, dependence, and addiction, with inpatient and outpatient treatment programs aimed at reducing the consequences of drug abuse. The goal of most programs is abstinence and restoring a person to pre-abuse levels of functioning. Tertiary prevention targets abstinence, withdrawal, and cravings via support groups aimed at relapse prevention.

### ***Traditional Treatment Modalities***

It is estimated that approximately 3.8 million people age 12 and older received some form of substance abuse treatment in 2004. It was also estimated that in 2004, 21.2 million persons needed treatment but did not receive it (SAMHSA, 2005). Several types of treatment are available. The focus in this section is on the following treatment modalities: residential, inpatient, intensive and non-intensive outpatient, self-help groups, and methadone maintenance.

Residential treatment consists of therapeutic communities (TCs). TCs are drug free facilities in which individuals commit to treatment for one to two years. TC treatment emphasizes that a person's substance abuse disorder cannot be cured but the behaviors that lead to abusing drugs can be addressed. Utilizing group therapy, TCs attempt to teach and promote changes in behaviors that eradicate substance use and abuse. The intent of intense treatment is to provide an environment in which residents can learn appropriate behaviors and develop skills to assist them in being able to adapt to their "outside" social environment in a drug free manner. Other services such as vocational and educational training are implemented to assist residents in improving self-esteem.

Inpatient treatment programs focus on removing individuals from the environments in which they have been using drugs. These programs are located in hospitals and tend to be costly. A person's treatment occurs over a period of 20 to 60 days. This type of treatment has a strict regimen of rules and expectations (Goldberg, 2003).

Outpatient treatment is the most utilized form of treatment. Two types of outpatient treatment are available that differ in intensity of treatment. Intensive outpatient treatment usually provides group therapy for three to five hours a day, five days a week. Groups focus on relapse prevention and drug education. An individual also receives case management services. Referrals are made to assist in addressing a multitude of barriers that are encountered by this population. Barriers to treatment consist of housing, unstable environment, conflict with family, low income, legal involvement, and childcare (Abadinsky, 2004). Intensive outpatient treatment usually lasts for 6-12 months. The advantage of utilizing outpatient treatment versus inpatient treatment is that it allows a person to maintain employment. Less intensive outpatient treatment provides similar services, usually for one to two hours a day, three to five days a week.

Self-help groups grew out of the idea that individuals with substance abuse disorders can assist themselves in recovery rather than utilizing professional services. Alcoholics Anonymous (AA) was the original self-help group for self-identified alcoholics. From AA, other self-help groups were formed, such as Narcotics Anonymous (NA) and Cocaine Anonymous (CA). These self-help groups utilize a 12-step recovery approach to treating alcohol and drug abuse. Core to the 12-steps is "the individual's powerlessness over alcohol, the need to recognize one's shortcomings and make amends, and reliance on a higher power" (McNeece & DiNitto, 2005. p. 182).

Methadone is a synthetic opioid that has been used in the treatment of narcotic addiction for over 40 years. It is a Schedule II drug with a high potential for tolerance and dependence. Methadone has similar



analgesic and sedative effects as other narcotics, but they last longer (12-24 hours) and are less intense. Approximately 20% of heroin addicts in this country receive methadone maintenance treatment (MMT) (ONDCP, 2003a). Benefits of utilizing MMT are that it reduces withdrawal symptoms from narcotics and inhibits euphoria experienced from narcotic use. A major concern with this practice is that individuals tend to become dependent on methadone. The substitution of one substance dependency for another is the main criticism of MMT. Yet, the effectiveness of methadone in reducing crime, death, disease, and drug use is well documented (Drug Policy Alliance, 2006)

### ***Effectiveness of Substance Abuse Treatment***

Does treatment work? This question is often raised in the field of substance abuse treatment by service providers and funding sources. Prendergast, Podus, Chang and Urada (2002) conducted a meta-analysis on the effectiveness of drug abuse treatment to answer this question. These authors concluded that substance abuse treatment is effective in reducing drug use and crime. Programs that were rated as being well implemented provided better results.

## **Vocational Factors**

For rehabilitation counselors, the probability of working with individuals that have substance abuse problems is high (Rubin & Roessler, 2008). It has been reported that there are high co-morbidity rates of substance abuse with other disabilities (Christensen, Boisse, Sanchez, & Friedmann, 2004). For individuals with disabilities, substance abuse can become a coping behavior to relieve negative interpersonal feelings related to disability. Vocational factors include the importance of employment, rehabilitation counselors' attitudes toward substance abuse and consequences of these attitudes, assessment of substance abuse during vocational intake, available vocational service delivery systems, incorporation of relapse into vocational planning, and programs to treat and prevent substance abuse at the workplace.

Employment facilitates success and is a goal in rehabilitation for a person in recovery, providing both stability and structure. Being employed places an individual in recovery within an environment with the responsibility for being on time, working a set amount of hours, and being rewarded monetarily. This assists in establishing a routine, which may have been previously nonexistent. By being financially stable, an individual is actively involved in alleviating stressors associated with a former loss of income.

Employment gives meaning to the individual as people often identify themselves through their occupation. Return to work provides a positive association and improves quality of life for individuals in recovery. This process is facilitated by rehabilitation counselors holding positive perceptions of all clients they serve, including those recovering from substance abuse. A perception of many counselors that is very detrimental is the view that persons in recovery will not successfully achieve planned vocational goals. Counselors need to address their own biases towards substance abuse. The following section examines vocational factors crucial for rehabilitation counselors to understand when providing services for individuals with substance abuse issues.

### ***Assessment***

The intake interview is a crucial time for rehabilitation counselors to obtain information to be used in developing a vocational plan. The importance of assessing for substance abuse is that a significant number of individuals with disabilities abuse substances. Rates of substance abuse among people with disabilities are higher when compared to the general population. Counselors need to utilize questions that focus on determining the extent of substance abuse. Important areas to examine include: stage of recovery, involvement with treatment, substance abuse history, frequency of use, methods to obtain drugs, and triggers that cause drug use (Becker, Drake, & Naughton, 2005).

Many clients are not forthcoming about their drug abuse during the intake interview. People with drug abuse issues have various areas of their lives that have been affected by their drug use. These areas consist of family, friends, work, finances, physical and mental health, and education. Some people may have been involved with the criminal justice system or are currently in this system due to drug use.



### ***Vocational Service Delivery Models***

Vocational service delivery models for individuals with substance abuse disorders are categorized as traditional and supported employment. Unfortunately, a majority of substance abuse facilities do not incorporate vocational services or the services offered are limited. There are four types of vocational service delivery models that include employment as an expected outcome: (a) work as positive outcome, (b) work infusion, (c) contingent sanctions, and (d) work as reinforcement (Magura, 2003).

The first model, “Work as Positive Outcome” is defined as an individual obtaining employment after completing substance abuse treatment. The justification for utilizing this model is that after completion of substance abuse treatment, the individual is able to obtain employment due to abstinence from drugs. Unfortunately, studies have shown that successful completion of substance abuse treatment does not insure obtaining employment.

The next model, “Work Infusion,” integrates vocational services within treatment. Providers that utilize a work infusion model view employment as a means of fostering abstinence. With this model, employment allows the individual to remain occupied and it provides structure. Employment is seen as a means of shaping positive self-identity (Magura, 2003).

The final two models implement vocational services into treatment as a means of influencing behavior in treatment. A “Contingent Sanctions” model focuses on a system where there are punitive consequences for not obtaining and maintaining employment. “Work as Reinforcement” utilizes employment as a positive outcome for remaining abstinent (Magura, 2003).

Vocational service systems are categorized as follows: traditional vocational services, supported employment, and customized employment services. These vocational service delivery systems incorporate variations of the described models.

Traditional vocational services provide vocational counseling to individuals in substance abuse treatment. Job training and placement is often outsourced to other agencies and vocational services tend to be minimal. An important characteristic of this delivery system is that users are required to have been abstinent from drugs for a period of time, usually six months (Blankertz et al., 2004). The next two models, supported employment and customized employment services, are more intensive and focus on expediting the return to employment.

Supported employment has been established as an effective means of returning individuals with severe disabilities to employment (Becker et al., 2005). The main focus of supported employment is to place the client in a job and train onsite. A job coach provides employment support.

Customized Employment Services (CES) is a vocational service delivery system established for clients receiving methadone maintenance treatment (Blankertz et al., 2003). The focus of CES is to provide comprehensive vocational services to expedite a successful and meaningful return to work. CES utilizes methods from both traditional vocational services and supported employment. The intent of CES is to address multiple barriers that deter individuals receiving Methadone Maintenance Treatment from returning to work. To address the multiple barriers, CES incorporates intensive case management services. A major barrier is self-efficacy. Self-efficacy is described as an individual’s thoughts and feelings that assist in determining his or her behavior. Placing clients into competitive employment facilitates improving self-efficacy by developing positive associations with work. CES encourages clients to consider and obtain various types of employment (i.e., part-time, self-employment, volunteer work) as a means of achieving long term vocational goals (Blankertz et al., 2004).

## **Federal Legislation and Individuals with Substance Abuse Disorders**

Substance abuse disorders are similar to other disabilities in the way federal legislation influences the delivery of vocational services. In addition, federal legislation affects individuals with substance abuse disorders in the legal, financial, and social realms. This section will examine the Rehabilitation Act of 1973 and its amendments, the Americans with Disabilities Act (ADA), and welfare reform acts that have helped shape policy towards people with substance abuse disorders.

The Rehabilitation Act of 1973 established disabilities as a protected status; however, substance abuse disorders were not considered a disability until the Rehabilitation Act Amendments of 1992. A substance abuse disorder is not protected when current use of substances affects job responsibilities or there is serious potential harm to self or others. Protection was expanded with the ADA to encompass both the public and private work sectors.

The ADA was designed to remove barriers to employment for people with disabilities (Rubin & Roessler, 2008). For individuals with substance abuse disorders, there are certain conditions that are necessary for protection under the ADA. Specifically, people are not covered if they are currently abusing substances and have not successfully completed or are currently participating in a supervised rehabilitation program (Shaw, MacGillis, & Dvorchik, 1995). A qualified person protected under the ADA is eligible to receive reasonable accommodation. Accommodation will not be implemented unless the individual has disclosed his or her disability to an employer during the hiring process. However, disclosing a substance abuse disorder may lead to being stigmatized and discriminated against, and can easily become a barrier to employment.

Due to this paradox, a rehabilitation counselor needs to discuss the issue of disclosure with an employer. Disclosure can be complicated since a substance abuse disorder is considered a chronic condition with potential for relapse. A counselor can explore these issues by working with a client to understand the positive and negative aspects of disclosure to an employer. Rehabilitation counselors can help employers view their role in reinforcing the process of recovery.

If an employer chooses to hire a person with a substance abuse disorder, reasonable accommodation can be implemented to ensure that the essential functions of the job can be performed. Accommodations may consist of the following: a flexible work schedule for the employee to attend ongoing treatment, job restructuring to reduce stress, and use of an employee assistance program to obtain brief counseling and referrals for treatment (Shaw et al., 1995). The intent of providing these accommodations is to prevent relapse and support abstinence.

## **Relapse and Vocational Planning**

There is a high probability for drug relapse. Relapse is the abstinence from drugs followed by a subsequent return to drug use. A rehabilitation counselor may experience difficulty in framing relapse into vocational rehabilitation services. The counselor needs to view relapse as a temporary part of recovery.

Other issues involved with relapse are related to protection in the workplace. As stipulated in federal legislation, people with substance abuse disorders are only protected if they are abstinent from drugs; relapse can result in job termination. An employer that has knowledge of relapse may become suspicious and unfairly drug test employees with this disability.

Because the intent of vocational services in substance abuse treatment is to facilitate reentry into a work environment as well as abstinence from drugs, it is crucial for the rehabilitation counselor to determine an appropriate work environment (Becker et al., 2005). For example, a counselor would not want to support a vocational plan for employment in a pharmacy for an individual that has been addicted to pain medication.

### ***Employment Assistance Programs and Disability Management***

A major factor affecting all employers is the issue of substance abuse. The Drug and Alcohol Services Information System (DASIS) (2004) reported that one-third of individuals admitted to substance abuse treatment programs were employed. Additionally, the National Survey on Drug Abuse Use and Health (NSDUH) reported that in 2004, 8.0% of adults employed full time and 10.3% of those employed part-time were illegal drug users. In addition, NSDUH also indicated that of the 16.4 million illicit drug users age 18 and older, 12.3 million were employed (SAMHSA, 2005). These statistics are noteworthy because of the large number of employed substance users and potential negative consequences for both employees and employers. Employers incur costs such as increasing health benefits, accidents, workers' compensation claims, absenteeism, and loss of worker productivity (Akabas, Gates, & Galvin, 1992; Reynolds & Lehman, 2003).

Corporations often utilize preventive measures (e.g., drug testing) to assist in identifying employees who are using substances. Once a problem has been identified, the employer can implement either punitive measures (i.e., termination) or rehabilitation. One resource that can be utilized by corporations is an Employee Assistance Program (EAP). EAPs assist in evaluating substance abuse, providing brief counseling, and referring to substance abuse treatment programs.

EAPs have been successful but there is a low percentage of employees that utilize this service (Reynolds & Lehman, 2003). Two reasons that explain employee reluctance in utilizing this service are lack of knowledge about the service and the stigma attached to receiving these services. For EAPs to increase outreach to employees who abuse substances, employers need to promote awareness of the program and its services. The stigma can also be addressed by providing a work environment that stresses the health of its employees (Calkins, Lui, & Wood, 2000). Although EAPs have been shown to assist identified employees, it remains a secondary measure to address substance abuse in the workplace. Corporations that utilize a disability management or Workplace Managed Care approach address the preventive measures necessary to curb workplace substance abuse.

Disability management has been a major force in changing the way disabilities are addressed in the workplace. Akabas et al. (1992) defined disability management as:

“a workplace prevention and remediation strategy that seeks to prevent disability from occurring or, lacking that, to intervene early following onset of disability, using coordinated, cost conscious, quality rehabilitation service that reflects an organizational commitment to continued employment of those experiencing functional work limitations” (p. 2).

Disability management has proven effective in lowering costs that result from disabilities (Calkins et al., 2000). It has been modified in some corporations to include all disability-related benefit programs including workers' compensation, public and private health insurance, short and long-term disability, and EAPs. Disability management emphasizes early intervention and preventing lengthy leaves of absence from work. Early intervention improves the possibility of a successful return to work. In the case of substance abuse, a disability management specialist works with an employee to provide reasonable accommodation to assist in a return to work. One possible accommodation involves provision of a flexible work schedule, allowing time for attending a substance abuse treatment program.

These innovative practices address substance abuse in the workplace with the intent of diminishing or eliminating drug use. An employer utilizing a preventive or secondary program helps alleviate the negative effects of substance abuse on both employees and employers. An employer may use an integrated approach, including both preventive and secondary services to reduce the number of employees that abuse substances and limit the costs and problems associated with substance abuse in the workplace.

## **Conclusion**

Prevalence of drug abuse is a significant problem within our society. Over \$80 million per year is spent on drug treatment, prevention, and drug-related law enforcement. There are multiple risk factors, including individual personality traits and environmental factors while relapse is a highly probable occurrence. Levels of negative impact on a substance abuser may vary and range from physical, mental, and emotional disabilities to attitudes and psychosocial issues. Gainful employment helps in retention of people who are in treatment. Working along with available support systems, rehabilitation counselors play a major role in improving quality of life for recovering clients.

## **Case Study**

Kompany Nguyen is a 28-year old Cambodian immigrant. Before coming to America, she was a middle school student and worked at her parents' farm. Her daily chores included feeding the farm animals, keeping the farm clean, and taking care of her three younger siblings while her parents worked in the fields. She speaks fluent Cambodian, Vietnamese, and minimal English. When Kompany first came to this country, she worked as a baby-sitter for her neighbors and kitchen assistant in a small Cambodian restaurant. Additionally, she was employed as a computer programmer and an assistant manager for a child care center.

Ms. Nguyen was arrested and jailed during a police raid for prostitution in the inner city 17 months ago. During the initial deposition, Kompang disclosed that she came to America as a mail order bride at age 18. Three years into the marriage, her husband found out she was unable to bear children; he became physically and sexually abusive and eventually forced Kompang into prostitution. Feeling guilty and shameful, she cut her wrists several times. To prevent her from further suicidal attempts and as a method to control her, the husband injected her with heroin for several years. Kompang was recently released and referred to your agency by her parole officer. When asked of her plans for the future, Ms. Nguyen expressed an interest in culinary arts and moving away from the Cambodian community.

### Questions

1. Assign Kompang a vocational profile and suggest necessary additional information about her work history that will be helpful in rehabilitation planning.
2. As the rehabilitation counselor, what services might be appropriate for Ms. Nguyen?
3. Identify possible job options for Ms. Nguyen given her education, work history, and transferable skills.
4. Identify important factors for developing a vocational objective with Kompang, and how you would assist her in proceeding with a vocational rehabilitation plan?
5. What kind of support system would be most beneficial for Ms. Nguyen and why?

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