Clinical Practice of Physical Medicine & Rehabilitation

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What is Physical Medicine & Rehabilitation?: Disability and the Role of the Physiatrist

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Introduction

Dramatic health advancements over the past century have developed and shaped the medical specialty of rehabilitation. Today, more people are living longer and living with health conditions previously not compatible with life. These health conditions impact an individual’s ability to function in society. Researchers and clinicians have looked for ways to define, quantify and monitor this impact through models of disability. Rehabilitation medicine is a specialty that cares for individuals with disabilities. It incorporates the biopsychosocial model into caring for individuals with certain chronic medical conditions, focuses on a person’s function in an individualized plan and is often delivered by a team of health care providers. Your rotation with Rehabilitation Medicine will expose you to the different aspects of this medical specialty.

Definitions of Disability

Health care providers have used a variety of models to describe the relationship between health and disease. The medical model of health and disease served us well when diseases were more likely to result in death than disability. This model views health and function as a direct consequence of disease. Treatment plans are typically not individualized and everyone with a particular disease gets the same treatment. However, the biopsychosocial model is the preferred modern model. This model incorporates psychological and societal factors into understanding a person’s medical illness. It is clear every individual has unique psychological and social factors that effect the impact a certain disease will have on their ability to function. Consequently, modern clinicians acknowledge the complexity within which diseases affect an individual’s life.

Disability is a term frequently used to describe the negative impact a disease has on an individual’s ability to function. However, a concise definition for disability has not been universally agreed upon. As mentioned previously, the impact a disease has on an individual is complicated by each individual’s unique set of characteristics. One frequently used definition is that disability is caused by abnormal physiology and is a resultant limitation in performing tasks, activities or roles to the levels that are culturally expected.

A very intriguing aspect of disability is the variability of disability a certain condition causes. This variability depends on a wide range of interdependent issues and applies even when given the same degree of pathology. This is a phenomenon you will want to look for during your rotation on the rehabilitation service. Why do some people with a significant pathological condition, such as spinal cord injury, function at a higher level than another person with a seemingly much lower level of pathology, such as musculoskeletal back pain?
In an attempt to describe and predict disability, various organizations have developed disability models. The World Health Organization developed the “International Classification of Impairments, Disabilities and Handicaps” in 1980. The goal was to provide a way to categorize and organize disease consequence information. This model was linear and included disease, impairment, disability and handicap. The definitions used are as follows:

1) **Disease**: a condition of altered pathophysiology
2) **Impairment**: loss or abnormality of an organ or organ system in structure or function
3) **Disability**: limitation of performing tasks, activities and roles to the expected level considered normal for a human being
4) **Handicap**: lack of societal role fulfillment for an individual due to a disability

Listed in a linear model, disease leads to impairments, which cause a disability and may or may not ultimately cause a handicap. This model can help the practitioner transition from acute medical issues to functional issues.

The following case study illustrates the use of this model. A young 23-year-old man is admitted to the hospital after a motor vehicle accident and is unable to move either leg. He is diagnosed with a spinal cord injury after x-rays and MRI demonstrate a thoracic burst fracture with subsequent spinal cord compression and resultant T11 spinal cord injury. Applying the model, the disease, or altered pathophysiology, is abnormal function of anterior horn cells in the spinal cord as well as ascending and descending spinal cord tracts. A few of the impairments, or abnormally functioning organs, include bilateral leg weakness, bowel and bladder malfunction, and sensory loss in the lower extremities. Disabilities, or limitations is performing tasks, include difficulty ambulating, urinating and defecating. The extent to which these will be a handicap for this individual depend on the patient’s ability to compensate for the disabilities and/or the environment’s ability to be adapted. With having these terms described, one can’t define the central process of rehabilitation as the reversal or improvement of disability by restoration of functional losses.

In one scenario the 23-year-old man is a programmer at a software development company. He did not suffer a brain insult and does not have any cognitive impairment. He learns to propel a manual wheelchair, manages his bladder with intermittent catheterization and manages his bowel with a daily bowel program. He adapts his car to have hand controls and eventually returns to his old job in a wheelchair accessible building. In this scenario, we have not identified any handicaps. He is able to fulfill his societal roles as before through changing his behavior and adapting his environment.

The amount of handicap that the same pathophysiology may cause could be different given different personal variables and a different environment. In a second scenario, the young man is a lumberjack who did tree topping and lived on the 3rd floor of an apartment building that did not have an elevator. He completed 9th grade and is not interested in learning a new job. He feels the bowel and bladder issues are repulsive and is not interested in learning to do the bladder and bowel program himself. Living in rural Washington, there are no available visiting nursing services to help him with these issues even if he could get a more accessible apartment. He has no health insurance, savings or supportive family. Consequently, he is discharged to a skilled nursing facility with an application for permanent disability pending. He is unable to fulfill his societal role of even basic self-care due to difficulty changing his behavior and the inability of his social and work environments to change.

The initial WHO model was criticized for it’s linear nature and more importantly for it’s lack of description of how the environment and society play a role in enabling or disabling a person. The WHO revised their classification schema in 2001. Their new model organizes information about the individual into 3 components called dimensions. These are: 1) body functions and structure, 2) activities and participation, and 3) environmental factors. Within each dimension there are categories called chapters.
Each chapter is further divided into facets or blocks. The blocks are further divided into categories. Ultimately, there are greater than 3000 items of potential classification domain names. The individual’s functioning is then described by a series of numbers. These numbers would identify a level of function and could be compared for research purposes. The goal of the revision was not only to include the impact of the environment on an individual’s function, but also to develop a system for collecting research data. This new classification is not typically used in the clinical setting.

Other organizations have worked toward developing a model for disability. The National Center for Medical Rehabilitation and Research (NCMRR) was developed within the National Institute of Health and established through legislation. Its mandate was to provide and direct research to enable society to comply with the Americans with Disabilities Act, which had been signed in 1989. In 1993, NCMRR further refined a model of disability. They added environmental or social factors to their model and refined a non-linear concept. They used terminology similar to the WHO definitions. These included the following:

1) pathophysiology (instead of disease)
2) impairment (similar to the WHO definition)
3) functional limitations (instead of disability)
4) disability (instead of handicap)
5) societal limitations: added to address the impact society and the environment have on an individual’s function. “Restriction attributable to social policy or barriers (structural or attitudinal) which limits fulfillment of roles or denies access to services and opportunities that are associated with full participation is society”.

Refining an alternative to the linear approach was more complex. The NCMRR wanted to get away from the idea that disease caused disability as a unidirectional inevitability. In addition, they wanted to present the complexities of the rehabilitation process, show how the process would change depending on the part of the life cycle the individual was in and illustrate how parts of the model effect each other in a non-linear way.

Their model has the person and the rehab process in the center. See figure 1. Double arrows go between this center and 3 personal background factors. These are factors unique to the individual that impact their response to a given situation or stress. These are as follows:

1) Organic factors: factors inherent in the individual such as genetics or family history.
2) Psychosocial factors: include coping styles, belief systems, personality styles or cultural influences. Finally,
3) Environmental factors: include the physical environment, income, access to health care, transportation or educational resources.

In addition, there are double arrows between the person and the rehab process, and quality of life. Factors influencing the quality of life are termed survival factors, productivity and social and work relationships. They are as follows:

1) Survival issues: include maintaining health and preventing further or unnecessary medical complications.
2) Productivity issues: include meaningful participation in society such as employment, education and recreation.
3) Social and work relationships: includes ones ability to develop and maintain relationships.
Finally, underlining the entire model is the effect of the life cycle on disability. Figure 1 is a visual representation of this model. Clearly, this model achieves the goals of being non-linear and illustrating the complexities of disability and the rehabilitation process.

In 1997, the Institute of Medicine provided yet another revised model. This was presented in a text Enabling America. They made a few modifications on the NCMRR model. In addition, they presented an alternative and provocative visual representation of the concept of disability. See figure 2. This model has a person standing on a mat. The mat represents the environment. The left half of the mat is the physical environment and the right half is the social environment. Picture that the mat is three-dimensional and the amount of displacement of the mat from the person standing on it is proportional to the amount of disability they have. In other words, a person sunk deep into the mat has a lot of disability while a person standing on a flat mat has little disability. The strength or resilience of the mat is a function of social and environmental support. In this model, the degree of disability has more to do with society, the environment and the ability of those two to accommodate the individual than the individual’s chronic condition.

No longer is disability a given progression from an abnormal pathophysiological state. No longer is disability a part of the person. Disability is now an outcome born of interactions that consequently place a larger responsibility on society to adapt to the individual.

The Role of the Physician: The Physiatrist

Long before the advent of multiple models of disability, physicians have been caring for individuals with disabilities. However, changes in the patient population have refined and ultimately specialized the physician’s role. World War I, with its surviving injured soldiers, provided an arena for physicians to treat more chronic conditions. This was the major impetus for development of a medical specialty to care
for these individuals. Thus Physical Medicine and Rehabilitation started in the 1920’s and has developed over time to include Physical Medicine, Rehabilitation Medicine and Electromyography.

Physical medicine, probably nowadays better termed musculoskeletal medicine, includes a focus on modalities for treatment such as heat, cold, massage and exercise. The first formal full-time academic physician in physical medicine was John Stanley Coulter, MD in 1926. Later, Frank Krusen MD established the first Physical Medicine and Rehabilitation residency at the Mayo Clinic in 1936.

World War II resulted in even more need for sophisticated diagnosis and treatment of rehabilitation issues again due to the large number of veterans with debilitating war injuries. During the war, Howard A. Rusk, MD an internist for the military, was able to demonstrate that early, active rehabilitation of patients resulted in a dramatic increase in the rate and extent of recovery after severe injuries. After the war, he established the Institute of Rehabilitation Medicine at New York University Medical Center. He is credited with contributing significantly to the realm of rehabilitation medicine. He advocated early and aggressive physical therapy, involvement in sports activities and addressing the emotional and psychological aspects of rehabilitation as well.

Additionally, the Veteran’s Administration has contributed significantly to the advancement of Physical Medicine and Rehabilitation through its treatment of disabled soldiers. They continue to play an important role in training residents, conducting research and providing care for individuals.

The Advisory Board of Medical Specialties recognized the current specialty of Physical Medicine and Rehabilitation in 1947. Initially it was only Physical Medicine and 2 years later Rehabilitation was added to the name. Physiatrist is the term for Physical Medicine and Rehabilitation physicians. Physiatrists are also trained in residency to be competent with electrodiagnostic medicine.

Physical Medicine and Rehabilitation specializes in management of an individual’s medical and functional status as a result of health issues. This includes prevention, diagnosis, and treatment of disabling conditions as well as prevention of further decline or injury. The general focus is on an individual’s function. If an individual has an impairment that affects their function, the physiatrist and rehabilitation team help to eliminate the impairment or alter the individual’s environment to maximize their function. This means the physician, as well as other team members, address how to facilitate and aid an individual to do the important and necessary tasks of daily life. These tasks include a wide range of activities such as self-care, mobility, work, leisure activities, vocation, avocation and social role fulfillment. Thus, a simplistic, but appropriate, analogy to describe physiatrists is: cardiologists are heart doctors and orthopaedic surgeons are bone doctors as physiatrists are function doctors.

Conditions typically cared for by physiatrists can be grouped into 2 categories. First are neurologic related issues such as traumatic brain injury, stroke, spinal cord injury and multiple sclerosis. The second general group is musculoskeletal problems such as sports injuries, amputations, or spine pain. They regularly evaluate, diagnose, and treat medical issues encountered by individuals with these disorders. In addition, they include the patient’s ability to participate in work, hobbies and societal roles as basic components of every patient’s problem list.

The Physical Medicine and Rehabilitation Team

Physiatry is unique in its interdisciplinary approach. Physiatrists frequently work with a team, especially when treating inpatients. Team members include physical therapy, occupational therapy, speech therapy, nursing, social work, recreational therapy, vocational counseling, prosthetics/orthotics, and psychology. A complete description of each specialty’s role is beyond the scope of this introductory chapter. However, the following will highlight some of the unique aspects of each team member’s role. The therapists are
very familiar with the team approach and will welcome questions you may have on your rotation. It is
time well spent to watch individuals during therapies to see how the therapists elicit cooperation from
patients, incorporate the patient’s goals into therapy goals, and encourage and motivate patients.

Physical therapists work principally with gross motor functions such as range of motion and
strengthening to facilitate ambulation, mobility and balance. In addition, they evaluate and fit manual
wheel chairs, train the individual in the use of orthotics and prosthetic limbs, manage edema and evaluate
home environments. They are also trained in the use of modalities such as heat, cold, ultrasound and
massage.

Occupational therapists work principally with fine motor functions such as upper extremity range of
motion and strengthening to facilitate grooming, feeding, dressing, and toileting. They provide and train
individuals to use adaptive equipment to facilitate the functions mentioned. They evaluate and manage
power wheel chair issues, provide driving evaluations, home environment evaluations and educate on
energy-conserving methods of self-care and home management tasks. Some also work as certified hand
therapists.

Recreational therapists use leisure activities to facilitate the rehabilitation process by increasing attention
span, concentration, and providing an arena to work on social skills. They also use recreational activities
to assess a person’s ability to integrate into society after an injury. They take individuals and groups on
community outings to evaluate safety in the community, ability to function in public and ability to
function in a group. Finally, through these activities recreational therapists provide a structured, safe
environment for the individual to start processing their reactions to their limitations as well as the
reactions of others to them.

The rehabilitation psychologists are well trained in addressing psychological issues related to an injury.
At the University of Washington, their mandate includes not only the patient’s needs but the family’s
needs as well. They address issues related to adjustment to disability for the patient and their family,
depression, and sexuality issues. They also provide a variety of testing to evaluate personality issues and
cognitive functioning.

Nursing clearly plays an important role in all patient care. Specifically with rehabilitation, nurses can
apply for advanced certification in rehabilitation medicine. This specialty includes in-depth knowledge of
wound care and skin management, adjustment to disability, bowel and bladder issues, goal setting,
psychological support and medication management. Most importantly, nurses encourage patients to be
maximally independent with their care, to be involved with the goal setting process and ultimately to self-
direct their care.

Speech pathologists are also integral to a rehabilitation team. They are experts in speech function such as
pronunciation and word finding. They also evaluate swallowing function and safety. They provide
cognition and communication evaluations identifying deficits and providing compensatory strategies for
the patients, care givers, and team members.

Social workers are indispensable members of the rehabilitation team. They provide emotional support for
families, evaluate living situations and coordinate disposition. In addition, they identify community
services available for patients.

Vocational counselors provide a valuable service to the rehabilitation team. They evaluate an individual’s
potential to return to work, facilitate the return to work process, and evaluate options should the
individual not be able to return to work. Their interventions optimize the return to work process thereby
maximizing the patient’s successful return. They are knowledgeable in disability programs and can save
the individual hours of work.
The prosthodontist/orthotist works with the patient, physical therapist, occupational therapist and physician when prosthetics and orthotics are necessary. They evaluate, design, and make prosthetic limbs and orthoses. They also evaluate their fit and function after fabrication. Again, seeing your patient in therapy with the prosthodontist/orthotist and therapists will give you some idea of the problem solving skills necessary for optimal prosthetic use.

Conclusion
The impact a disease will have on a person’s function is a unique outcome of factors specific to the individual and their environment. Physiatrists treat individuals with a variety of functional disabilities and chronic diseases and require comprehensive medical knowledge about a variety of conditions. Physiatrists utilize the rehabilitation team members’ expertise in maximizing an individual’s function. The rehabilitation approach is unique in that it is team oriented, focuses on the functional aspects of a person’s life, and requires excellent interpersonal and problem solving skills. It is a rewarding medical specialty for people who value the individual’s progress from acute illness or impairment back to functional and productive community life.

REFERENCES


History and Physical Examination in Chronic Disease and Disability

Walter C. Stolov, MD, and Ross M. Hays, MD

The “classical” history and physical examination is unchanged when dealing with a patient with chronic disease. Details can be found in standard texts, and will not be repeated here. The workup presented here elaborates on features that are essential to reaching a diagnosis of disability in addition to that of disease. As a framework for conducting an adequate history and physical examination in a person with chronic disease or disability (see Table 2.1), the examiner must have a clear understanding of the terms disease, impairment, disability, and handicap which have all been introduced in the preceding chapter.

History

Historical data of prime importance for the diagnosis of disability are obtained from the “chief complaint,” “present illness,” and “social and vocational history.” The nature of the chief complaint may provide a hint to the existence of disability; the present illness data can determine the extent of lost function in basic self-care activities; the social and vocational history evaluates the environment, the family, and the social milieu and provides insight into the psychological background of the patient. The review of systems and past medical history contribute to the assessment of residual capacity.

CHIEF COMPLAINT

Chief complaints generally result from changes in health or well-being that create fear or anxiety, discomfort, or an inability to function. These unwelcome consequences lead the patient to seek medical attention. The chief complaints most likely to clarify areas of disability have to do with changes in the patient’s function. The class of diseases most likely to produce complaints of loss of function are those that involve the musculoskeletal, nervous, or cardiovascular system.

PRESENT ILLNESS

One of the hallmarks of disability is loss of independence. Historical information regarding dependence on others for activities of daily living (ADL) is best included under the category “Present Illness.” Such data are really part of a symptom complex of the disease and describe the disability to be addressed in rehabilitation. For example, a patient concerned about reduced ambulation may actually be providing a description of muscle weakness; or a patient who has developed a tremor may describe the abnormality historically as onset of difficulty holding a cup of coffee.
### Table 2.1

#### History and Physical Exam Outline

**Problem List**

Include functional, social, psychological, and vocational deficiencies as separate problems.

**Patient Profile**

1. **Social Function**
   - a. Current: architectural layout of the home; the persons in it, and their responsibilities
   - b. Past social history

2. **Vocational Functions**
   - a. Most recent job, either within or without the home
   - b. Job history
   - c. Avocational activities

3. **Psychological Function**
   - a. Lifestyle
   - b. Response to stress-past and current
   - c. Motivational factors

**Present Illness and Problems**

After developing historical specifics of the problems on your list, add a paragraph that describes current state and the evolution of impairment in the following categories:

1. Ambulation
2. Transfers
3. Dressing
4. Personal hygiene
5. Eating
6. Communication

Describe patient performance of these functions as: independent, requires standby assistance for safety or cues; requires partial physical assistance; or totally dependent.

**Physical Exam**

Include a thorough physical exam. Additionally, divide the neuromusculoskeletal examination into three parts.

1. **Musculoskeletal**
   - Use the screening exam described in this chapter.
   - Explore any abnormalities with a more detailed exam.

2. **Neurologic**
   - Use a basic screening exam with the additions mentioned in this chapter.
   - Include the expanded mental status exam.

3. **Functional Neuromuscular Exam**
   - Observe patient performance in ambulation, transfers, dressing, personal hygiene, and eating

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This one-page form is designed for use at the bedside. The points outlined are additions to consider in examination of a patient with chronic disease or physical disability.
The activities for which inquiries should be made can be divided into six categories:

1. Ambulation (Movement from place to place);
2. Transfer activities (changes of position in place);
3. Dressing;
4. Eating;
5. Personal hygiene including use of the toilet and bathing; and
6. Communication

The quantification of dependence in any of these activities of daily living is achieved by asking the patient: What type of assistance is needed? And who provides it? Assistance may take several forms:

- **Stand-by Assistance.** When the activity is performed by the patient, there may be concerns about safety or correct execution of the activity. The assistant then is “standing-by” to guard against the occurrence of accidents and to ensure completeness of performance by noting errors and omissions.

- **Partial Physical Assistance.** The patient is able to do a good part of the activity independently but an assistant is needed to provide partial help. For example, the assistant may buckle the patient’s belt after the patient has donned pants unaided; or the assistant may hold the wheelchair stationary while the patient transfers into bed.

- **Total Physical Assistance.** The patient contributes little or nothing toward performance of an activity. In these situations the patient is described as totally dependent. For the activity to occur, others must do it.

**Ambulation History**

In the broadest sense ambulation may be defined as travel from one place to another over a finite distance. It thus includes not only walking but also wheelchair travel, or even crawling. In order to assess the extent of disability with regard to ambulation, the patient’s capacity for moving in different environments must be obtained.

The environments of significance are the home, its immediate vicinity, and the community at large. A patient may, for example, be totally independent in walking around the house but not out in the community. The environments found in theaters, restaurants, subways, or downtown stores may require partial physical assistance in order to negotiate safely. The disability diagnosis for such a patient will therefore include the problem of “decreased ambulation in the community.”

If a patient uses a wheelchair, the extent of independence in its use needs to be known. For example, the examiner should determine whether the patient is able to maneuver it independently in the home or she can use it successfully out in the community unaided. Some questions in an exploration of ambulation skills are as follows:

- Are you able to walk without help from anyone?
- Do you use assistive equipment (canes, crutches, braces)?
- Do you use a wheelchair?
- Is there a limit to how far you can walk or use your wheelchair outside your home?
- Do you go out visiting friends or to restaurants or to theaters or stores?
- Do you fall very often?
- Do you drive?
- Can you climb stairs?
Transfer History

Transfers are movements that involve changes of position in place. They include such activities as turning over, moving from a bed to a wheelchair or a regular chair; going from a wheelchair to a toilet, bathtub, shower or car; and going from a wheelchair, regular chair, or toilet to a standing position. These activities are more basic than ambulation. For example, while a person may be independent in walking, he or she may need to depend on others for help to rise from a chair into the standing position. Independent ambulation therefore is not always available if assistance for the transfer into the upright posture is not always present.

Sample questions to begin an assessment of disability in transfers include the following

- Can you get in and out of bed unaided?
- Can you get on and off a toilet unaided?
- Can you get in and out of the bathtub without help?

Dressing History

A disabled patient’s ability to don and remove clothing must be carefully assessed. If the person is not independent in dressing, he or she is less likely to engage in community activities outside the home and less likely to receive guests. Dressing therefore has a significant impact on social adaptation after the onset of disability.

In obtaining a history of performance in dressing skills, it is not sufficient merely to ask: Do you dress yourself? An untrained, disabled patient may have, for sometime, abandoned the use of garments that are more difficult to don. Typically abandoned are shoes, socks, pants, clothes with buttons, and close-fitting undergarments. The patient therefore may answer “Yes” to such a question without realizing how few clothes he or she actually wears. A complete probe into dressing history is necessary to gain insight into performance and function. Sample questions to explore dressing abilities include the following.

- Do you dress in street clothes daily?
- Can you put on, with assistance, your shirt, pants, dress, undergarments etc.?
- Do you need help with shoes and socks?
- When you must go out, how much of your dressing do you do by yourself?
- How long does it take you to completely dress before school, work, etc.?

Eating History

The loss of independence in a person’s ability to feed himself or herself can be devastating to self-esteem. Unlike the activities previously discussed, it is one activity that must still persist even if totally physical assistance is required. The association of passive feeding with dependence is exceedingly strong in our society, and such individuals often isolate themselves socially. Eating skills include the use of fork, spoon, and knife, and the handling of cups and glasses. Sample questions for exploration of this area include the following.

- Can you feed yourself without assistance?
- Are you able to cut meat?
- How do you handle messier foods, such as soup and cereal?
- Do you have trouble holding glasses and cups?

Personal Hygiene History
Personal hygiene activities include the spectrum of skills concerned with cleaning and grooming: tooth brushing, hair combing, shaving, the use of the tub and shower, perineal care, and the successful handling of bowel and bladder elimination. Loss of independence in the performance of these skills is severely disabling. This is particularly true when a patient cannot handle bowel and bladder elimination in a socially acceptable manner. If he or she is concerned with the possibility of becoming soiled with feces, or urine, the emotional stress will be quite severe. Vocational rehabilitation efforts and improvements in social functioning will be unsuccessful until the person can develop a system for elimination that is consistent and successful.

It is more important to be continent socially than to restore elimination patterns that are anatomically and physiologically ideal to those of the nondisabled population. Socially acceptable elimination can be achieved by the majority of disabled patients. Patients with catheters can develop a successful system if they can handle the emptying of collecting bags and can satisfactory incorporate the necessary devices within their clothing. Same questions to explore the personal hygiene area include the following.

- Can you shave (use makeup) and comb your hair without assistance?
- Can you shower or bathe without help?
- Are you able to use the toilet unaided?
- Do you need help with any aspect of using the toilet?
- Are bladder and bowel accidents a problem for you? If so, how often?

**Communication**

The term *communication* includes a broad range of skills associated with listening, speaking, reading, and writing. Communication encompasses the breadth and depth of language, and therefore reflects the patient’s intellectual capabilities and educability. Listening and reading skills that form the receptive component of language function depend on the intact use of auditory and visual organs. Speaking and writing—the more expressive forms of language—depend on the integrity of motor functions associated with articulation and hand dexterity.

To obtain an accurate history of communication skills in patients with expressive communication deficits, the examiner will direct the inquiry to family members or others who have had a recent and long-standing relationship with the patient. To put the present level of disability into context, a clear idea of the patient’s premorbid language skills must be obtained. For communication deficits that are changing, the time course of deterioration should also be ascertained. The communication history will sometimes be self-explanatory. Recognition of expressive aphasia, dysarthric speech, and so on are logical components of the physical exam and are often elicited during the mental status examination or incidentally noted during the general history. Frequently, specific questions regarding expressive verbal language are unnecessary. Sample questions that may augment the examiner’s understanding of receptive language difficulty or deficits in written communication include the following.

- Do people often appear to mean something other than what you thought you heard them say?
- Do you have difficulty reading and understanding newspapers or magazines?
- Is writing possible for you? Do you have, or use, any special tools or methods to improve your ability to communicate either in speech or in writing

**General Principals in Determining Disability in Basic Functions**

Several principles must be kept in mind when exploring disability in the basic functions of ambulation, transfers, dressing, eating, and personal hygiene.
1. When the patient reports that he or she is not independent, determine the extent of assistance required for the particular skill in question.

2. If assistance is being supplied, determine who is assisting the patient.

3. Separately interview the persons (usually family members) who are supplying the assistance. Assistant(s) may report a greater degree of dependence than reported by the patient. The two may interpret differently what is occurring. A significant difference in their remarks may indicate that one or both are dissatisfied with the situation.

4. When it is expected or anticipated that the patient will be dependent, questions such as “can you” or “do you” should be rephrased. It may be more helpful to use questions beginning with “Who helps you…” Questions asked in this manner may yield more useful functional information because patients may initially wish to appear more independent than they actually are.

5. When the disability is one of acute onset, the inquiry should also include the premorbid level of independence. This is particularly important in the older patient or a person suffering an acute exacerbation of a long-term disability. Earlier disease or trauma may have left the patient with some residual dependence. An accurate understanding of the person’s prior skills will obviate the possible error of expecting rehabilitation to improve the patient’s functional status beyond that which was present in the premorbid state.

6. If there has been a loss of independence and the disease is of a progressive nature, determine the time course of this loss. Efforts to intervene in these situations are much more likely to be effective if the function was lost recently. Long-standing disability is likely to have already created some adaptive strategies.

    In many cases the answers to the preceding sample questions may be obvious from observation and physical examination. It is best, however, to assume less and inquire more, to prevent omitting significant data.

    When inquiry into the self-care functions of mobility (ambulation and transfers), dressing, eating, personal hygiene, and communication is complete, specific disability problems should be identified and documented separately in the patient’s list of problems, even though they may be interdependent. The pathologic condition underlying the functional disability may be irreversible in part or in whole, or the decreased function may not be irreversible in part or in whole, or the decreased function may not be eliminated by reversal of the disease process. Therefore disability problems may be more appropriately addressed individually with their own set of predisposing conditions, so that strategies may be designed to restore function when none are available to cure the disease and its impairments.

**Past Medical History**

Like the review of systems, the Past Medical History provides information regarding the patient’s residual capacity. Concurrent disease or previous trauma and surgery may have produced residual impairment which, although it no longer produces disability itself, may compound the disability of the present illness.

    A careful review of Past Medical History is therefore an essential component when evaluating the patient with disability. A simple recitation of past or concurrent disease or trauma may not be sufficient. The inquiry requires an understanding of the impact that past illness has had on the present disability, regardless how minor.

**Patient Profile**

The patient’s social and vocational history will provide the database necessary for understanding the interaction between the patient and his or her environment. A careful review of these issues will identify environmental
factors that are either secondary to or concurrent with the disease, and will also provide insight into the personality of the patient and his or her ability to adjust to the stress of chronic disability. A careful understanding of the complexities of adaptation to disability should be obtained as a foundation for further rehabilitation intervention. It is convenient to divide the personal history data into three categories: (1) social, (2) vocational, and (3) psychological.

**Social History**

The family unit may be compromised when a person becomes dependent on others for the performance of self-care skills, or when vocation is disrupted secondary to disease. Other family members’ plans may alter significantly due to the need to assist the disabled person in performing the activities of daily living, as well as by the loss of income incurred by chronic disability. A major disability on a family unit already beset with social difficulties is particularly threatening. Identification and inclusion of secondary and concurrent social issues into the patient’s history and problem list will allow the physician to begin to understand the environmental factors that influence the patient’s physiologic and medical problems.

The assessment of social impairment is obtained with inquiries into the stability of the family. Its history, the resources available to its members, and the responsibilities of the patient within his or her family should all be ascertained, as should a clear understanding of the physical environment of the patient’s home and community. In this context the word *family* must be broadly interpreted to mean “all interested parties within whom the patient interacts in an interdependent manner.”

The physical environment is important because dependence or independence in the performance of activities of self-care is directly related to the location in which the activity is performed. Sample questions that begin the search for problems in social functioning include the following.

- Do you live in the city? A rural community? The suburbs?
- Do you rent or own your residence?
- Are the bedroom, bathroom, kitchen all on the same floor?
- Are there entrance stairs or stairs within your residence?
- Do others live with you? If so, do any of them go to work? To school? Do they have any health problems of their own?
- Do your parents, brothers, and/or sisters live in your immediate vicinity? Do you maintain contact with them?
- Are you married? If so, for how long?
- What activities and functions did you perform for your family that you no longer are able to do (e.g., parental discipline, financial management, chores sexual activity, avocational activities)? How are these functions now being handled?
- Where were you born and where else have you lived? What did (or do) your parents and siblings do for a living? At what age did you leave your parent’s home?

The answers to these questions will provide the necessary information on social background and current resources, as well as some insight into current or potential problems. “Abnormal” responses to these questions should be pursued. For example, if the patient has experienced major disruptions within the family unit, further
questioning regarding the roles of individual family members may yield important clues regarding additional social stressors and past coping strategies.

**Vocational History**

A patient’s disease may also lead to unemployment. Whether there is, or will be, a significant disability in this area depends on the physical, intellectual, and interpersonal requirements of the patient’s job. Sample questions that will elucidate the interconnection between disease, lost function, activities of daily living, and employment are as follows.

- When did you last work?
- For whom did (do) you work?
- How long have you worked for them?
- Describe what you do (did) on the job. Be specific. Start with what you do (did) when you first arrive(d) at work and give a brief outline of your day.
- Was (is) your income sufficient to support your family or do you have other resources? Do you have significant debt?

Answers to these questions should provide information about the patient’s premorbid vocational pattern. If the description given by the patient of current or last job seems incompatible with present illness, even with rehabilitation management further inquiries may be necessary.

- What type of work do you plan to do in the future?
- What type of work have you performed in the past? How long ago did you do this type of work? Would you be interested in returning to it? What qualifications do you have (education, membership, licenses, special skills)?
- What is your educational background?

This additional information will indicate whether the patient has had work adjustment problems. The strengths in the person’s vocational background, which may be successfully exploited in the process of vocational adaptation, will become more clear in this portion of the history. If the patient has been unemployed, it is important to understand his or her current sources of financial support and their likely sufficiency for the future.

When working with persons who are not employed outside the home, or for those who live alone, it is important to inquire about household tasks and skills. Sample questions would include the following.

- What do you do with your leisure time (by yourself, with your family) after work and on weekends?
- What organizations or religious groups are important to you?
- When did you last participate in these activities?

**Psychological History**

Psychological function must be assessed in patients with chronic illness or physical disability for several reasons. Since the organic, pathologic changes may be incompletely reversible, the stress of the disease is often persistent. This stress may be of great magnitude. For example, a patient who loses a limb has to adjust not only to this loss but also the secondary stress of loss of employment if the physical requirements of the job are incompatible with activity limits of an artificial limb. The patient and his or her family may then have to relinquish established goals and patterns of activity. The patient may have to learn to be more active in protecting
his or her health than when premorbid. These new modes of behavior are likely not the patient’s prior preferred way of doing things. Thus the patient’s psychological background needs to be assessed and understood in order for new learning and adaptation to take place. A clear understanding of the reinforcers most likely to be successful in motivating a given patient will also be necessary to change behavior. For patients with brain damage from trauma or disease, an understanding of intellectual function will be required in order to successfully devise treatment strategies to reduce the disability.

Psychological problems should be included in the patient’s problem list when reaction to the stress of the disease interferes with adaptation to the disability, and when new learning is potentially impaired. While the mental status examination can provide some assessment of current function, the social and vocational data will yield a great deal of valuable information about the person’s personality. The same social and vocational data that identify the person’s lifestyle will also identify the types of activities that can be used as goals toward which the person will work to remove dependence during treatment. The likelihood of success in an activity, consistent with premorbid lifestyle, will serve as a motivational factor, even if the work the patient must perform to achieve his or her goals includes activities that are, in themselves, alien to his or her usual style. For example, an interpersonally oriented individual may be motivated to perform certain heavy exercises important for health if increased social contact with others will be possible when a required level of strength is achieved. Similarly, an intellectually oriented individual may be motivated to develop independence in transfer skills if successful transfers will afford opportunities to attend lectures or concerts.

Social and vocational data provide information that allows the clinician to build upon the patient’s strengths in planning for the removal of disability. Interpreting social and vocational data to yield psychological characteristics is a relatively simple matter when organized into four categories: (1) the patient’s previous lifestyle; (2) the patient’s past history or response to ordinary life stresses; (3) the patient’s current response to the stress of his or her disease; and (4) the activities likely to motivate the patient to adapt to his or her disability.

The Physical Examination

The information obtained from the physical examination of a person with a disability serves three functions:

- The examination searches for the signs that signify deviations from normal structure and function. Correlation of these signs with the patient’s history and laboratory data will yield the disease diagnosis.

- In examining a disabled patient, the physician searches for signs of secondary problems that are not necessarily a direct consequence of disease. Such secondary problems are physical complications that result from the loss of ability to initiate preventative health habits, or as a consequence of the medical treatment of the disease.

- The physical examination assesses the residual strength in the body systems that are unaffected by disease. These strengths provide the foundation upon which the patient and his or her physician will build a strategy to reverse or minimize the disability and reestablish loss functional skills.

The importance of secondary problems, whether unavoidable, treatment-induced, or due to omission of prevention measures, is that these add to the patient’s disability and may lengthen the treatment time necessary to remove the disability associated with the primary disease.

All patients with chronic disease and disability require a complete physical exam. The interaction between health-care professionals who provide rehabilitation or management of chronic disease and disability with professionals who provide acute care suggests that primary diagnosis issues have usually been addressed by the acute care providers. Frequently, new and important clues to previously unrecognized or underappreciated acute
medical concerns will become apparent in the process of carefully evaluating a patient from a fresh point of view with a complete physical exam.

In the thorough general physical exam, special attention should be paid to the cardiovascular and the respiratory systems to determine if there will be any cardiorespiratory limitations to retraining in self-care and activities of daily living. Because chronic disability frequently includes alterations in bowel and bladder function, special attention should be paid to the genital and rectal exam, especially in the evaluation of sacral mediated reflexes. Evaluation of loss of skin integrity or the incidence of skin breakdown is another essential aspect of the general physical exam, which may have significant implications for the disabled patient. In addition to the thorough general physical exam, the examination of the patient with disability must include an expanded examination of the nervous system, the musculoskeletal system, and the interaction between the two in functional activities; this is sometimes referred to as the functional neuromuscular examination.

**Nervous System**

The neurologic exam should be performed with the same care exercised by neurologists in searching for signs in a difficult diagnostic problem. All twelve cranial nerves should be carefully evaluated. The screening neurologic exam should be expanded with the addition of a careful sensory exam. The use of a standardized patient map illustrating spinal cord level dermatomes and the distribution of peripheral nerves is helpful in sorting out observed abnormalities and sensation. In addition to the sensation of superficial touch and pain, the examiner should test position sense, vibration sense, stereognosis and two-point discrimination, as well as hot and cold perception. Cerebellar and coordination functions should be tested in both gross and fine motor movements.

The general neurologic exam of deep tendon reflexes should be expanded to include the bulbocavernous reflex, in order to evaluate the S2 through S4 reflex arc. This is tested by involuntary contraction of the external anal sphincter in response to brief stimulation of the glans of the penis or clitoris, or to a tugging applied to an indwelling urinary catheter. The exam should include the evaluation of abnormal reflexes, including the Babinski reflex in the lower extremity, Hoffman’s reflex in the upper extremity, and careful evaluation of primitive reflexes in the brain-injured patient.

**Cardiovascular System**

Retraining to restore basic self-care skills that are lost as a result of musculoskeletal and neurologic disease usually requires specific therapeutic exercise regimens. An adequate cardiovascular reserve and optimized cardiovascular function are therefore essential.

Examination should therefore include the blood pressure (supine, sitting, standing,) liver size, peripheral pulses, carotid pulses, venous return systems, peripheral skin temperatures, peripheral skin hair, and peripheral edema. Cardiac size, cardiac rhythms, and cardiac sounds will need correct interpretations. All treatable abnormalities will need identification.

**Respiratory System**

Much like the cardiovascular system, the respiratory reserve must be assessed to evaluate exercise tolerance.

Examination should include the respiratory rate and rhythm, the chest shape, the fingers for clubbing, the facies for cyanosis, and the lungs for congestion and obstruction. Supplementary pulmonary function laboratory tests may also be required.
GENITALIA AND RECTUM

Particularly critical for patients with diseases that affect the functions of micturition and defecation are examination for cystocele and rectocele, prostate size, sphincter tone and anal wink reflexes, perineal sensation, the presence of orchitis and epididymitis, and the presence of the bulbocavernous reflex.

If present, the bulbocavernous reflex signifies that the sacral conus of the spinal cord at the level of S2 and S4 is intact. The afferent sensory stimulus is elicited by pressure on the clitoris or glans. For patients with catheters, a tug on the catheter will stimulate the response. The efferent response is contraction of the external sphincter. It can be detected by a finger in the anus; visualization of the anal opening may suffice.

MUSCULOSKELETAL SYSTEM

The functional unit of the musculoskeletal exam is the joint and its associated structures: the synovial membrane, capsule, ligaments and the muscles that cross the joint. Examination of this complex anywhere in the body cannot be completed unless the underlying anatomy is well understood. The screening examination is useful for localizing abnormalities when the disability problems are minor. However, examination of individual joints is necessary for conditions that result in major disability. Such examinations include inspection, palpation, passive range of motion, stability, active range of motion, and muscle strength.

Inspection

The two sides of the body should be observed for symmetry in contour and size, and any differences measured. Atrophy, masses, swelling, and skin color changes should be noted.

Palpation

The origin of a pain symptom may be localized by palpation of the various anatomic structures about the joint. Palpation of the bones may determine their discontinuity in fracture assessment. Palpation of masses and swelling for consistency can distinguish between bony masses, edema, and joint effusion. To determine the presence of a muscle spasm, muscle palpation with the patient at rest will identify sustained involuntary reflex contraction resulting from pain.

Passive Range of Motion

These tests are performed by the examiner while the patient is relaxed. When range of motion is limited, the examiner must determine if the limitation is due to joint surface incongruities, joint fluid excess or loose bodies, or to capsule, ligament, or muscle contracture.

Stability

To assess whether pathologic condition of the bone, capsule, or ligament is causing abnormal movement (subluxations or dislocations), the joint should be moved under stress in the direction its contour ligaments and capsule do not normally permit it to move, with the patient at rest. Tears in a ligament or laxity of the capsule will result in abnormal mobility. During movement, joint stability is also supported by active muscle contraction.

Active Range of Motion

These tests should be performed prior to strength tests in the event that pain is a problem. Muscle tension and joint compressions induced by an active movement are less than those in a strength test. If pain is minimal in the active range of motion, the examiner can more easily proceed with the strength test. When active range of
motion is less than the passive range of motion, the examiner must decide between true weakness, hysterical weakness, joint stability, pain, or malingering as possible causes.

**Muscle Strength**

Muscle strength can be tested if its prime action is known. The body part can be positioned to allow this prime action to occur. Grading systems are based on the ability of the muscle to move the body to which it is attached against the force of gravity.

**Grade 5:** Normal Strength. The muscle is able to move the joint it crosses through a full range of motion against gravity and against “full” resistance applied by the examiner.

**Grade 4:** Good Strength. The muscle can move the joint it crosses through a full range of motion against gravity with only “moderate” resistance applied by the examiner.

**Grade 3:** Fair Strength. The muscle can move the joint it crosses through a full range of motion against gravity only.

**Grade 2:** Poor Strength. The muscle can move the joint it crosses through a full range of motion only if the part is positioned so that the force of gravity does not act to resist the motion.

**Grade 1:** Trace Strength. Muscle contraction can be seen or palpated but strength is not sufficient to produce motion—even with gravity eliminated.

**Grade 0:** Zero Strength. Complete paralysis; no visible or palpable voluntary muscle contraction.

Grade 3 is the key muscle grade with regard to disability assessment. Determination of grade 3 is objective and independent of the examiner’s strength. Since any activity a patient may perform is done in a gravity field, muscles with grade 3 strength will allow the involved body part to be used. For grades less than 3, external support may be necessary to allow the involved part to be useful to the patient. Additionally, joints having muscles across them with less than grade 3 strength are prone to contractures.

Different examiners should concur about whether a muscle should be graded 0, 1, 2, or 3. For grades 4 and 5, ratings may vary among examiners depending on the expectations of different age groups and the amount of resistance that different examiners are able to apply. With experience the examiner’s accuracy will likely increase. In patients where motor strength deficits are asymmetrical, grades 4 and 5 are more readily appreciated because the unaffected body part can be used as a control.

This grading system is not as useful for the prediction of motor strength in conditions where weakness is associated with spasticity.

A useful abbreviated musculoskeletal exam is outlines in Table 2.2. It may be used as a framework for quickly evaluating the patient for gross impairments. When abnormalities are noted on the screening exam, a more detailed examination must be performed in order to clearly understand the nature of the impairment. The outline is a helpful guide to assist in the accurate assessment of the musculoskeletal system. It was developed at the University of Washington as a physical exam screening tool. Although the brief outline will not detect subtle pathophysiology, it will indicate areas of gross impairment that can then be further evaluated with more detailed examination. This method is easy to learn and especially appealing because the entire exam can be completed within a few minutes.
FUNCTIONAL NEUROMUSCULAR EXAMINATION

TABLE 2.2
Musculoskeletal System Exam Outline

A. Essential Symptoms to Evaluate in the History
   1. Pain
   2. Weakness
   3. Deformity (deviation from normal posture, including limitation of movement)
   4. Stiffness
   5. Injury
   6. Functional limitations

B. Screening musculoskeletal Exam to Localize Impairments
   1. Inspection of body of anatomical position
      a. From front, back, and side
      b. Symmetry, alignment
   2. Inspection of gait
      a. From front (or back) and side
      b. Look for:
         (1) Abnormal truncal movement,
         (2) Abnormal pelvic movement,
         (3) Stride length,
         (4) Base
         (5) toe, heel walking.
   3. Cervical spine movements
      a. Flexion is normal when chin can touch the sternum
      b. Extension is normal if the occiput comes within one finger width from the C7 spinous process
      c. Rotation left and right should allow the plane of the head to make an angle of 70° to shoulder plane
   4. Elbows extended and arms are placed in front of the body with the shoulder at 90° of forward flexion
      a. Inspect for symmetry
      b. Test hand intrinsic strength by having the patient abduct his or her fingers and attempt to squeeze them together
      c. Shoulder flexion strength is tested by resting upward motion of the arm
   5. Arms abducted overhead with external rotation
      a. Inspect rhythm of movement, symmetry of scapulae; smooth movement at the shoulder, sternoclavicular and acromioclavicular joints
      b. Can the upper arm touch the patient’s ears?
      c. Test lateral deltoid strength by resisting horizontal abduction with resistance applied to the elbows
   6. Test internal and external rotation of the shoulder by having the patient touch the inferior border of the scapula at the back to demonstrate full internal rotation and elbow flexion (external rotation is indicated by the ability to touch the back of the head)
   7. Any limitation of elbow, wrist or finger range of motion will be apparent from careful observation of the previous maneuvers; note any limitations.
   8. Upper extremity strength screen
With elbows at side flexed at 90°, the patient grasps examiner’s hands and holds himself or herself rigid while examiner tests finger flexors, wrist flexors, wrist extensors, forearm supinators and pronators, elbow flexors, elbow extensors, shoulder protractors and retractors by moving the direction that is resisted by these motions.

9. Examination of back
   a. With the patient’s back bare, have him or her standing facing away from the examiner check for level shoulders and pelvis; is the head in midline, spine straight? Observe for symmetry of space between trunk and upper limbs.
   b. Have the patient flex his or her trunk forward with knees straight; observe for a paraspinal prominence of thorax, or lumbar areas (i.e., fixed scoliosis).
   c. When the patient extends trunk check for flexibility.
   d. Have the patient rotate his or her trunk to right and left while the examiner stabilizes pelvis; the plane of shoulders should rotate to a 45° angle with the plane of the pelvis.
   e. Ask the patient to laterally flex his or her trunk to right and left; the fingertips should touch head of fibula.

10. Lower extremity hip and knee range of motion and extensor strength screen can be grossly evaluated by one maneuver: The examiner holds patient’s hands while he or she squats with heels remaining on floor and then returns to upright position. Limited extensor strength will result in loss of control of the smooth descent and extensor weakness will interfere with ascent. Any limitation in range of motion at the hips, knees, or ankles will be obvious during the maneuver. Any abnormality observed during the brief screening exam should be evaluated more thoroughly. A useful system for more detailed evaluation follows.

C. Specific Regional Exam of the Various Joint-Muscle Complexes Found To Be Impaired on History and Screening Exam

1. Inspection
2. Palpation of key anatomical structures in region (Consider percussion of muscles for myotonia)
3. Passive range of motion, using goniometer for accurate assessment
   a. Joint evaluation for contracture
   b. Assessment of muscle tone
   c. Search for rightness in two-joint muscles (e.g., hamstrings)
4. Stability test (stress joint in direction opposite to normal movement for ligament laxity or rupture)
5. Active range of motion, using goniometer for accurate measurements (Examine if added compression force of muscle contraction induces pain)
6. Muscle strength – use 0-5 grading system

   0=No visible or palpable voluntary contraction
   1= Visible or palpable contraction present but insufficient to produce range of motion (ROM)
   2= Sufficient strength to produce full ROM with gravity elimination
   3= Sufficient strength to produce full ROM against gravity
   4= Strength can be overcome with moderate resistance
   5= Normal, sustains against high resistance.

Once the musculoskeletal system is evaluated, the patient’s actual ability can be more accurately assessed with the functional exam.
The functional examination confirms the status of skills reported by the patient and the history regarding ambulation, transfers, eating, dressing, and personal hygiene. The functions to be tested include the following.

**Sitting Balance**

Sitting is a necessary prerequisite for transfer skills. It is tested by placing the patient in the sitting posture, with the feet on the floor and back unsupported; the patient’s hands are in his or her lap. If the patient can hold this position, then gently push him or her in various directions to observe the ability to use protective extension to recover from the tendency to fall (i.e., dynamic balance).

**Transfers**

Movements to be examined include turning from supine to prone and back, rising to a sitting position, rising from sitting to standing, and moving from a bed or a low examining table to a chair.

**Standing Balance**

This is a necessary prerequisite for safe ambulation. It should be assessed with support and, if adequate balance is present, the person should be gently pushed from side to side to assess the ability to use protective responses.

**Eating Skills**

Eating skills may be assessed by demonstrating hand-to-mouth abilities with various examining room objects or, for an inpatient, by means of actual observation at mealtime.

**Dressing Skills**

Range of motion and strength of upper extremities, including grip and fine motor skills as well as the flexibility of the lower hips, knees, and spine, are required for independence in dressing. Patients should be independent in dressing skills if they have the ability to touch the tops of their heads, the small of their back, and their feet; have the ability to flex at the hips and knees; and have the manual dexterity to handle snaps and buttons. Isolated parts of dressing, such as manipulating buttons or managing a garment, frequently can be simulated in the examination setting to represent the patient’s dressing skills.

**Personal Hygiene Skills**

Upper extremity strength and range of motion required for dressing are also required for personal hygiene skills. The motions necessary for face, perineal, and back care can usually be simulated by the patient in the examining room. Direct observation of a specific task when it is actually performed may be useful in clarifying some of the details of sequencing and organizing complex tasks.

**Ambulation**

Walking should be observed if the patient has standing balance. The person should wear minimal clothing so that major joints and the back may be observed as much as possible. Walking should be inspected with and without street shoes, and from the front and back as well as from the side. Abnormalities should be described in relation to the phase of gait in which they occur. If pain is present, it too should be related to phase of gait. The following format should be used in the systematic examination of gait.
• Observation and description should be systematically performed and recorded.
• Cadence should be observed for symmetry and consistency. Trunk position should be noted for flexibility or presence of abnormal posturing and especially for exaggerated movements in the anterior, posterior, or lateral planes.
• Arm swing—especially asymmetry—should be evaluated.
• The position of the pelvis should be observed for posture and placement. Abnormalities in anterior and posterior placement result in lumbar lordosis or kyphosis or obliquities associated with a lurching gait.
• The base should be observed to determine whether it is normal, narrow, or broad.
• Heel-strike and push-off should be carefully evaluated. Swing phase hip and knee flexion and circumduction should all be considered and noted when present.

If the person cannot walk, wheelchair mobility should be evaluated. The patient’s ability to travel in a straight line and to negotiate turns should also be observed. Careful attention should be paid to the patient’s seating and positioning in the wheelchair. If posture is not appropriate, recommendations should be made to improve positioning to allow the patient to have better use of the upper extremities for propulsion of the wheelchair. Seating and positioning may be critical for patients using a power wheelchair to allow them access to the electronic interface necessary to control its independent use.

MENTAL STATUS EXAMINATION

For a patient with chronic disease or disability, the screening neurologic evaluation should be expanded to include a comprehensive mental status exam. This examination and the psychological history provide the background for understanding the person’s basic personality structure and current emotional reaction to the disability.

Removal of the disability is an educational process that involves retraining and relearning. The mental status examination becomes particularly important in patients for whom disease or trauma has produced brain damage. The orientation of the mental status examination for the patient with physical disability differs from that usually emphasized in a psychiatric evaluation. The general categories to evaluate in the disabled patient include recent memory, perception, affect, and judgment.

Recent Memory

It is important to evaluate recent memory function in order to ascertain the patient’s ability to relearn skills and thus make progress in rehabilitation. The patient may, for example, need to learn a specific technique to execute a safe transfer, or to coordinate crutch-walking skills. Learning such new skills requires the patient to assimilate, retain, and reproduce new material, which may not have not been previously learned, or which may have been learned and lost.

Recent memory function with regard to language information may be assessed by asking a patient to remember, for example, an address provided. Retention is evaluated when the patient later is asked to reproduce the address, perhaps on the next day. A simple new motor task, taught during the evaluation, can be used to assess motor learning. Retention of this motor skill can then be assessed at a later date. A clear understanding of strengths and weaknesses in the recent memory area will guide the rehabilitation team in their approach to training the patient to use new adaptive skills.
**Perception**

Perception included the process by which the patient organizes sensory information about his or her environment. In this context the term *perception* is used to describe how the patient interprets his or her environment because disturbances of this interpretation may have an organic or a psychiatric etiology. The disabled patient will frequently have subtle disturbances in perception, relating to the ability to process visual information of form, space, and distance. These visual representations of the environment require correct interpretation in order for the patient to make a correct motor response. Errors in visual perception may result in serious accidents if they are not appreciated and incorporated into the adaptive strategies designed for the patient. For example, a patient with monocular blindness may suffer deficits in depth perception that may result in the inability to judge the correct distance for transfer from bed to wheelchair; this will result in accidents. A patient with right parietal lobe injury may not be able to accurately interpret the difference between the inside and outside of a garment and thus will require adaptive strategies for dressing.

Disturbances in perception of this type are more likely to occur with damage to the right cerebral hemisphere. These can be assessed by asking the patient to copy figures such as a square, a triangle, and a Maltese cross. He or she can also be asked to reproduce a clock face from memory. When disturbances in perception of form exist, these reproductions are distorted. The findings of disturbed perception suggests that the teaching of basic self-care will be more successful by verbal instruction than by demonstration.

**Affect**

A reactive depression is common following the acute onset of a major disability, or after a relatively sudden additional functional loss in a person with long-standing disease. It is a realistic response, and indicates that the patient has enough insight to recognize his or her losses. This same insight will be useful to the patient in helping him or her to remove the disability. A reactive depression requires remedial action if it is associated with eating or sleep disturbances or if it interferes with the patient’s ability to respond to treatment. The absence of reactive depression may be considered abnormal in some cases. If the patient is unable to confront his or her physical functional losses, the ability to overcome the disability created by the loss may be reduced.

Mood swings are another feature of affect to consider. Rapid transitions from laughter to tears and back can represent the lability associated with psychological disturbances or can be due to brain injury. Certain features may be helpful in delineating the emotional lability associated with organic brain disease from that associated with more traditionally psychiatric affective disorders. For example, vigorously changing the subject of the conversation or the simple immediate distraction of attention by snapping fingers will frequently alter the patient’s mood if the lability is of organic origin.

**Judgment**

In brain damage judgment factors related to difficulties that the patient may have in self-monitoring behavior. These may include the failure to detect errors, such as the omission of behaviors normally incorporated in eating or dressing. These problems must be distinguished from carelessness associated with depression. If such behavior is observed in the general assessment of the patient’s appearance and the various activities performed during the course of the evaluation, judgment problems may exist. Insight into judgment can also be obtained during observation of the patient as he or she performs the various tasks given the mental status exam. An organic origin is likely when such behaviors are associated with the onset of disease or trauma. When judgment errors are present, standby assistance and increased supervision may have to be provided as the person perfumes various functional activities.
The Problem List

A person with a disability is described here to illustrate how a problem list may be constructed.

A 19-year old woman fractured her cervical spine in a small plane accident, resulting in quadriplegia. Her male companion, with whom she had been living for the previous year and a half, was killed in the crash. Their relationship had been close and family-oriented; she served as a “stepmother” for her companion’s two small children from a prior marriage. Following the accident, responsibility for the children was legally assumed by the natural mother.

The patient was hospitalized for a short period on an acute neurosurgical service and then transferred to a comprehensive rehabilitation center for inpatient care. Following a complete comprehensive evaluation shortly after admission her problem list included:

1. C7 fracture dislocation
2. C7 complete quadriplegia
3. ambulation dependent
4. transfer skill dependent
5. eating, dressing, personal hygiene skills dependent
6. neurogenic bowel dysfunction
7. neurogenic bladder dysfunction
8. decreased respiratory function
9. potential for pressure sores
10. potential for deep venous thrombosis
11. history of impulsive behavior
12. reactive depression
13. home architecture incompatible with paralysis
14. financial obligations without immediate resources
15. diminished contact with her family of origin
16. unemployment without prior work history
17. absence of independent transportation

Seventeen problems create an impressive list. It may be argued that the list need not be this long because nearly all the problems are secondary to the first-C7 fracture dislocation-and that this one diagnosis should be sufficient. Such an approach might be valid if there were a therapeutic technique to reverse spinal cord damage and restore full nervous system function. Unfortunately, such a technique does not exist. There does exist, however, a set of techniques for each of the 17 individual problems. These can be used to minimize the severity of the problems and to fully resolve some of them.

Problems 1, 2, 6, 7, 8, 9, and 10 are examples of more “classic” medical problems. Problems 3, 4, 5, and 17, while also direct results of the trauma, relate to the patient’s physical disabilities. Problems 11 and 12 relate to the patient’s psychological condition and problem 13, 14, and 15, to the social sphere. Problem 16 succinctly identifies the vocational disability.
Conclusions

The diagnosis of disease alone is insufficient for the planning of a comprehensive treatment program. The symptoms and signs required to diagnose disease are not synonymous with those required to diagnose disability. An understanding of disability-this is, specific losses in physical, social, vocational, and psychological function-requires investigation beyond the history and physical exam ordinarily required in the treatment of acute disease. The techniques described will also identify those medical problems secondary to but not naturally consequences of a chronic impairment. To achieve a successful treatment program that removes disability, the clinician must also be prepared to assess and appreciate the patient’s residual strengths.

Following an appropriate evaluation, the clinician will be able to list all of the patient’s problems. This problem list will include disease, diagnosis, and secondary complications. It must also include the specific losses in physical, self-care, social, vocational, and psychological function. Once this problem list is established, the rehabilitation treatment process can begin. Successful strategies to remove disability can be planned for each of the problems on the list. Therapeutic techniques used will fall within one of six general areas:

1. Methods to prevent or correct secondary complications
2. Methods to enhance the capability systems unaffected by disease;
3. Two three Methods to improve the functional capacity of affected systems;
4. Methods to promote function through the use of adaptive equipment;
5. Methods to modify the social and vocational environment; and
   Methods derived from psychological theory to improve the patient’s performance
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Treatment Strategies in Chronic Disease

Walter C. Stolov, MD, and Ross M. Hays, MD, and George H. Kraft, MD

The history and physical examination of a patient with disability will identify the medical problems and the limitations of function that are in need of resolution, both of which constitute the problem list. A patient with chronic disease and disability will usually have a combination of both physiological and functional items on the list.

It is not unusual for such a patient to have an extensive problem list in which the physiologic and functional items are independent. The “classical” items on the problem list include pathologic and physiologic disorders that are frequently irreversible. The functional items, however, will be amenable to rehabilitation treatment as outlined in this book.

Many, and in some cases all, of the functional problems directly result from the classical medical problems. Notwithstanding, each needs a separate identification because they all have their own set of treatment techniques. Thus L2 paraplegia secondary to vertebral fracture causes an “ambulation dependency.” Removal of this dependency through treatment to achieve bipedal or wheelchair ambulation involves techniques that have nothing to do with treatments which deal with the fracture and the damage to the cauda equina, and do not change the paraplegia itself.

Strategies have to be carefully planned in order for the patient to achieve resolution or improvement of these specific functional deficits. Six classes of treatment strategies have been identified for patients with chronic disease. Their purpose is not to reverse the pathological derangement but to remove disability, and include the following:

1. prevention or correction of additional (secondary) disability;
2. enhancement of systems unaffected by pathology;
3. improvement of the functional capacity of affected systems;
4. the use of adaptive equipment to promote function;
5. modification of social and vocational environments; and
6. psychological techniques to improve patient performance.

Prevention or Correction of Additional Disability

Perhaps the most important initial treatment for the patient with chronic disease and disability is the prevention of secondary complications that would increase disability. Anticipatory guidance, based on knowledge of the primary pathology, guides the team and the patient in the development of strategies that will prevent secondary disability. Examples of such strategies would include:

- medication to avoid congestive heart failure in patients with atherosclerotic cardiovascular disease;
- medication to maintain control of diabetes, to reduce the chance of peripheral neuropathy and peripheral vascular disease;
- anticoagulation to reduce the risk of cerebral thrombosis in patients with transient cerebral vascular insufficiency;
• passive range of motion exercises for joints where motor strength is less than grade 3 to avoid the development of soft tissue contracture;
• appropriate management of the neurogenic bladder to prevent infection and hydronephrosis and resultant renal damage;
• progressive resistive exercise (PRE) to reduce muscle weakness associated with bed rest;
• a system of periodic relief of pressure at insensate body surfaces to avoid loss of skin integrity and decubitus ulcers; and
• time-contingent medication to prevent addition in the patient with chronic pain.

Enhancement of Systems Unaffected by Pathology

Impairments that result in disability are frequently not systemic in nature. Disability can often be reduced by either providing improvement beyond the previous level of function of body systems unaffected by the present disease, or by using those unaffected systems in novel ways to accommodate a loss of function elsewhere. Examples of the enhancement of systems unaffected by pathology include:
• exercise to strengthen muscles on the nonparalyzed side of a hemiplegic stroke patient or in the upper limbs of a paraplegic spinal-cord-injured patient;
• adaptive techniques using visual monitoring of hand function for patients with cutaneous sensory loss; and
• adaptive training using hearing and tactile sensation to improve mobility in a patient with low visual acuity.

Improvement of Functional Capacity of Systems Affected by the Disease

The disabled patient who suffers derangement of body function may be helped by directly addressing the deficits responsible for the disability. The strategies are most successful when the natural history of the impairment suggests that improvement will occur over time. In these cases the improvement initiated by the body’s own healing capacity can be augmented by well-planned, adaptive strategies. Examples of enhancement of functional capacity of affected systems include the following:
• graded exercise programs to improve systemic conditioning after myocardial infarction;
• exercise to improve the strength of muscles weakened by prolonged bed rest; and
• the use of visual cues in brain-damaged patients to assist impaired memory function.

Use of Adaptive Equipment to Promote Function

Perhaps the most rapidly growing area of medical rehabilitation is the use of adaptive equipment. The advent of computer-assisted technology and the miniaturization of electrical circuitry has allowed the development of a new generation of technical adaptations for disability. In many cases such technical aides may restore function in a patient who otherwise would remain severely disabled. It would be erroneous to assume that all adaptive equipment is complicated and electronic. Frequently, simple approaches will also have great benefit for the patient with disability. Specific examples of the use of adaptive equipment include the following:
• shoe modifications to improve standing balance;
• mechanical equipment to extend hand function for dressing-long shoe-horns, stocking pullers, and buttonhooks;
• canes, crutches, and braces to improve energy expenditure and safety in ambulation;
• wheelchair training for travel when walking is not possible;
prostheses for amputee patients to allow independent ambulation or to replace upper extremity function.

- adaptive hand controls for patients unable to use an automobile in the conventional fashion.
- computer-assisted augmentative communication systems for patients with the cognitive potential for language but with severe oral motor impairment.

**Modification of Social and Vocational Environments**

Disability is frequently as much related to the patient’s environment as it is to his or her impairment. Environmental barriers may combine with the individual’s disability to produce a handicap. For example, the patient is not handicapped, the patient has a disability, but lack of access to an elevator may be a handicap. Adaptations of the environment at home and in the workplace may restore function. In many cases modifications of a patient’s environment must be made before the patient can be discharged from the medical facility. Examples of environmental modifications include the following:

- procurement of a one-level home for patients unable to climb stairs;
- alterations to hallways and doorways to allow easy wheelchair access;
- addition of rails and grab bars to promote safety and mobility within the home;
- provision of assistance in the home for physical and homemaking needs;
- employment to reduce physical or mental demands on patients who have reduced motor strength or ambulation, or cognitive problems due to head injury;
- redesign of the work area for patients who will be expected to use wheelchair mobility; and
- training of family members to adapt to changes in the disabled individual in order to reinforce constructive behavior and discourage unnecessary dependence.

Support from health professionals and the family can exert a potentially powerful influence over the patient’s performance. Once a working relationship between the health-care team, the family, and the patient has been established, further work can progress to replace lost function. It is the responsibility of the health-care team to identify for the family which skills should be independently performed by the patient and which should be replaced, either by assistance from others or by technological adaptations. A clear differentiation of the tasks that can be expected from the patient will help prevent the frustration in the family caused by a mismatch of expectations after the patient returns to the community. As recovery continues, flexibility should be maintained to allow the patient greater independence as warranted by improved performance.

**Psychological Techniques to Improve Patient Performance**

Principles of behavior management can be extremely helpful in the recovery of function. Although certain behavior management strategies may be intuitive to the family, others may need to be pointed out by a more objective third party unemotionally tied to the patient. Examples of psychological techniques that can be used to improve patient performance include the following:

- the use of repetition in training self-care skills in patients with memory problems;
- adaptive strategies using demonstration for patients with receptive language deficits;
- the development of a consistent milieu to provide reorientation for the confused or agitated patient emerging from coma associated with traumatic brain injury; and
- the judicious use of operant conditioning techniques to improve performance and minimize unwanted or harmful behaviors.
Summary

In the presence of irreversible pathology, disability (i.e., functional problems) can be modified and reduced. The six treatment strategies emphasized in medical rehabilitation are not themselves directed at modifying the pathology. They are, however, effective means of reducing disability.

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Electromyography and Nerve Conduction Studies

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Introduction
Electromyography (EMG) and nerve conduction studies (NCS) are the tools that form the basis of the electrodiagnostic medicine consultation. With EMG and NCS, the practitioner can often diagnose, localize, and provide a prognosis for injury or diseases of the lower motor neuron and peripheral nervous system. EMG/NCS provides an assessment of the function of the nervous system which is often complementary to the anatomical picture provided by imaging studies.

This chapter will review the basics of NCS and EMG, and discuss the application of these studies to four typical clinical scenarios: traumatic nerve lesions, radiculopathies, peripheral nerve entrapments, and peripheral polyneuropathies.

Nerve Conduction Studies
Nerve conduction studies involve electrically stimulating a peripheral nerve and recording action potentials from nerve or muscle. Doing so allows one to measure the speed of conduction and the number of nerve and/or muscle fibers participating in the response. This then allows the examiner to make inferences about the degree of demyelination (producing slowing) or axon loss (resulting in small responses) that might be occurring in the nerve.

Sensory nerve conduction studies involve stimulating a peripheral nerve and recording from the same nerve either distally or proximally. When stimulating distally and recording proximally the study is referred to as orthodromic, i.e. proceeding in the same direction as occurring physiologically. Since axons conduct bidirectionally, one can just as easily record antidromic potentials, i.e. stimulating proximally and recording distally. An example of a sensory conduction study is seen in figure 1. Stimulation occurred proximally at the wrist and the response was recorded over the ring finger with electrodes placed circumferentially around the finger, 14cm distally. We know this is a sensory rather than muscle response since there are no muscles in the fingers.

Measurements of the response include the latency (time from stimulation to the peak of the response) and amplitude (size of the response in μV). The velocity in meters per second can also be calculated since both distance and time (latency) are known (velocity = distance / time).

Figure 1. Sensory NCS to the ring finger in a patient with CTS. Top trace is stimulation of median nerve. Bottom trace is stimulation of ulnar nerve. Note longer latency of median nerve, consistent with CTS
Prolongation of distal latency (i.e. slowing of conduction) can be due to both physiologic and pathologic factors. Mild slowing is known to occur with increasing age and with cold. Demyelination, from compression or disease, produces pathologic slowing. If demyelination is severe enough conduction can even be blocked completely.

Reduction in amplitude of the response typically results from loss of sensory axons. Loss of axons between the site of stimulation and recording will produce an immediate reduction in the response amplitude. However, loss of axons proximal to the site will, over time, also produce a loss of the potential amplitude. With more proximal lesions that separate the axon from the cell body (i.e. at or distal to the dorsal root ganglion) the axons will undergo Wallerian degeneration and by day 10 they will become inexcitable distally.

Motor nerve conduction studies are performed by stimulating a nerve proximally and recording from a distally supplied muscle. Since the neuromuscular junction operates only unidirectionally, it is only possible to perform orthodromic motor NCS (i.e. in the same direction as occurs physiologically). An example of a motor NCS is demonstrated in the top trace of Figure 2; the median nerve is stimulated at the wrist and recording electrodes are placed over the abductor pollicis brevis (APB) muscle 8cm distally. In this case latency and amplitude can be measured similarly to sensory NCS as above. However calculation of NCV is more complicated since the latency involves not only conduction along the nerve but also time to cross the NMJ (about 1msec) and possibly some time to conduct along muscle fibers. Thus taking the latency and dividing by distance is not an accurate method for measuring NCV. In this case the NCV is instead calculated by doing a second stimulation at a more proximal site (bottom of figure 2). Taking the distance between the two stimulation sites and dividing by the latency difference yields the NCV between the proximal and distal sites.

![Figure 2. Motor NCS to the abductor pollicis brevis muscle with stimulation of the median nerve at the wrist (top) and elbow (bottom). Nerve conduction velocity is calculated by dividing the distance between stimulation sites by the difference in latency.](image)

In the case of motor NCS, speed of conduction is affected by the same factors as for sensory NCS: primarily age, cold, and demyelination. However, since the response is recorded from muscle, the amplitude is affected not only by the number of nerve fibers, but also by the integrity of the NMJ and the number of muscle fibers. Moreover, since the motor neuron cell bodies are in the anterior horn of the spinal cord, it is from this point distally that any lesion will cause Wallerian degeneration with loss of motor amplitudes.
Figure 3. The motor unit and needle EMG. In the top figure, a normal EMG is demonstrated; the muscle is quiet at rest but a normal motor unit action potential (MUAP) is recorded with small voluntary contractions. In the middle figure, after denervation occurs, single muscle fibers will spontaneously discharge creating fibrillation potentials or positive sharp waves. When nearby axons send sprouts to reinnervate denervated muscle fibers

Needle EMG

The basic concept central to understanding the needle EMG exam is that of the motor unit. The motor unit is comprised of a single motor neuron and all the muscle fibers supplied by that neuron. The cell body of the motor neuron resides in the spinal cord (anterior horn cells). The number of muscle fibers supplied by the axon varies widely and depends upon the function of the muscle. Muscles that require small low force movements, such as extraocular muscles, have as few as six muscle fibers per axon. Muscles that require high force movements such as the quadriceps have hundreds of muscle fibers per axon.

Needle EMG involves placing a needle electrode into the muscle and recording potentials from muscle fibers both at rest and during contraction. Normally during rest the muscle should be electrically quiet. During contraction, the motor unit generates a motor unit action potential (MUAP) representing the synchronous discharge of all the muscle fibers supplied by the axon (Figure 3).

When the motor axon is interrupted by disease or injury and the muscle becomes denervated, the muscle fibers supplied by that axon will, after 2-3 weeks, start to fire spontaneously, i.e. not under voluntary control. These spontaneous single muscle fiber discharges, depending upon the integrity of the muscle fiber, will be recorded as fibrillation potentials or positive sharp waves occurring at rest. These potentials will persist either until the muscle is reinnervated or, in the case of complete denervation, becomes fibrosed.

When a muscle undergoes partial denervation a number of changes occur to the MUAP as well. After partial nerve injury, axons that are spared will start to sprout new branches to reinnervate the denervated muscle fibers. As their motor unit territory grows, the MUAPs change in appearance. Since these axons supply more muscle fibers the MUAPs grow in size. Moreover, the immature sprouts that supply these previously denervated muscle fibers will initially be poorly myelinated and hence conduct more slowly. As a result the MUAP will be larger, longer in duration, and more poorly synchronized (polyphasic) than normal. When these MUAP changes are seen, they indicate that reinnervation has occurred.
Traumatic Neuropathies

Nerve injuries after trauma are common. It is estimated that about 3 – 5 % of patients admitted to a Level I trauma center have some type of peripheral nerve injury. These are more common in more severe trauma, and patients on the rehabilitation service with traumatic brain injury, spinal cord injury, or multiple trauma are more likely to have peripheral nerve lesions.

Peripheral nerve lesions can be classified according the scheme proposed by Seddon. Neurapraxia is the most mild type of injury. It is usually a result of ischemia or demyelination with intact axons. Prognosis is favorable with recovery occurring within 3 months. Axonotmesis is defined by injury to the axons, but with at least partially intact supporting structures (endoneurium, perineurium, and/or epineurium). Recovery in this case is dependent upon axonal regrowth. Prognosis is dependent upon how many axons are injured, the distance to the muscle, and preservation of the endoneurial tubes for axons to regrow. Neurotmesis is the most severe nerve lesion with complete lesion of the axons and all the supporting structures, such as a complete nerve laceration. Prognosis is poor without surgical intervention.

EMG/NCS is useful to classify traumatic nerve lesions according to the above scheme. Neurapraxic lesions, without axon loss, will have intact responses when stimulation and recording occur distal to the lesion since Wallerian degeneration does not occur. There will, however, be conduction block when stimulating across the lesion. In contrast, axonotmesis and neurotmesis are lesions in which there is loss of axons and hence Wallerian degeneration occurs distal to the lesion. As a result, the amplitude of the response will be reduced or the response will become completely absent. Since the only difference between axonotmesis and neurotmesis is the integrity of the supporting structures, it is generally not possible to differentiate between these two possibilities using EMG/NCS. Both axonotmesis and neurotmesis will demonstrate fibrillations and positive sharp waves on needle EMG and, in the case of incomplete lesions, evidence of reinnervation.

Traumatic nerve lesions can often be localized using EMG and/or NCS. Conceptually, EMG can be used to localize nerve lesions by studying muscles along the course of a nerve. By knowing the branching pattern of the nerve one can presume that the nerve lesion is between the branch to the most distal normal muscle and the most proximal denervated muscle. NCS can localize neurapraxic lesions by demonstrating focal slowing or conduction block in a segment of the nerve.

Prognosis of traumatic nerve lesions can be estimated by measuring the amplitude of the motor nerve response when stimulation and recording are distal to the lesion. A large amplitude response indicates that many axons are intact and the lesion is neurapraxic; recovery can usually be expected within 3 months. A small amplitude or absent response indicates that severe axon loss has occurred and prognosis is guarded.

Radiculopathies

Radiculopathies, lesions of the nerve root, are commonly a result of a herniated intervertebral disk, neural foraminal stenosis, or central spinal canal stenosis. This lesion usually occurs proximal to the dorsal root ganglion (DRG), which sits near the intervertebral neural foramen.

Electrodiagnosis is helpful to diagnose radiculopathies and to evaluate the degree of axon loss. The most helpful electrodiagnostic test for radiculopathy is the needle EMG examination. Within 2 – 3 weeks after onset of acute radiculopathy the paraspinal muscles will start to show evidence of denervation (fibrillations and positive sharp waves) and soon thereafter muscles in the limb...
will show similar changes in a myotomal pattern. Diagnosis of radiculopathy requires finding evidence of denervation in two or more muscles supplied by the same root but different peripheral nerves. In more chronic radiculopathies there will be evidence of reinnervation with larger long duration polyphasic MUAPs.

Sensory nerve conduction studies are usually normal in radiculopathy since the lesion is typically proximal to the DRG. Motor nerve conduction studies are usually also normal, but when there is severe axon loss the motor responses can be decreased in amplitude due to Wallerian degeneration.

The H-wave is a specialized type of nerve conduction study performed in the lower limbs when assessing for S1 radiculopathy. It involves stimulating the tibial nerve at the knee and recording a late response from the soleus muscle. The response travels up the sensory fibers of the tibial and sciatic nerves, through the S1 root and after a central synapse, down the motor axons of the same nerves. It is similar to the clinical ankle jerk. In S1 radiculopathies the H-wave will show abnormalities before needle EMG changes occur.

In the assessment of radiculopathies, electrodiagnostic studies provide information complementary to that of imaging. Imaging studies of the spine (e.g. MRI, CT) provide an anatomical picture; while sensitivity is high, specificity is only moderate. It is estimated that 30 – 40% of asymptomatic healthy individuals will have disc bulging or protrusions on MRI. On the other hand EMG provides a functional assessment with a high specificity. Sensitivity, however, is probably somewhat lower than imaging modalities.

**Entrapment Neuropathies**

Entrapment neuropathies such as carpal tunnel syndrome (CTS), ulnar neuropathy, and fibular (peroneal) neuropathy are common sources of complaints of focal numbness and/or weakness. Electrodiagnostic studies are the initial evaluation of choice in most cases.

CTS, due to median nerve compression at the wrist, is the most common entrapment neuropathy. It usually affects women more than men, typically in the 5th decade of life. Risk factors include high force, highly repetitive work, obesity, pregnancy, diabetic polyneuropathy, and other medical conditions. In CTS the median nerve undergoes focal demyelination under the transverse carpal ligament; in more severe cases axon loss ensues.

Nerve conduction studies demonstrate slowing of median nerve conduction across the wrist. Sensory fibers are typically more affected than motor fibers due to their larger diameter. A common approach is to compare conduction in the sensory nerve with conduction in another nearby nerve (e.g. ulnar or radial nerve) as seen in figure 1. This approach compares two nerves at the same temperature, age, etc. and is preferable to comparing the results to normal limits.

Needle EMG will sometimes show changes in the median innervated thenar muscles consistent with denervation. However, this is a late change reflecting motor axon loss and is uncommonly seen.

**Peripheral Polyneuropathies**

Electrodiagnostic studies are very helpful in classifying and diagnosing peripheral polyneuropathies. Typically polyneuropathies affect the longest axons first and present as distal sensory complaints and distal weakness, involving the lower limbs more than the upper limbs. Presentation will vary, however, depending upon the specific etiology of the polyneuropathy.
Polyneuropathies are classified according to whether motor or sensory fibers are predominantly affected, whether the disease primarily affects axons or myelin, and, for demyelinating neuropathies, whether it is patchy or uniform demyelination. The six diagnostic categories usually used, with typical examples are:

1) Uniform Demyelinating: Inherited neuropathies such as Charcot-Marie Tooth.
2) Segmental (patchy) Demyelinating: Acquired neuropathies such as Acute Inflammatory Demyelinating Polyradiculoneuropathy (Guillain-Barré syndrome).
3) Sensory Axonal Neuropathy: e.g. paraneoplastic syndromes
4) Motor > Sensory Axonal Neuropathy: e.g. porphyria
5) Sensory and Motor Axonal Neuropathy: e.g. toxic or nutritional neuropathies
6) Combined Demyelinating and Axonal Motor and Sensory Neuropathy: e.g. Diabetes and Uremia

Nerve conduction studies and EMG are very useful to differentiate neuropathies into one of the above categories. Demyelination is inferred when there is marked slowing of conduction. Sensory and motor axon loss are detected by reduced amplitude of the sensory or motor responses respectively. Motor axon loss is also detected by evidence of denervation and/or reinnervation on needle EMG. Generally at least two limbs are studied in order to be sure the process is diffuse and not limited to a single limb distribution.

**Conclusions**

EMG/NCS is helpful for diagnosis, localization, classification and prognostication of a variety of peripheral nervous system and lower motor neuron lesions. These studies provide a functional assessment that is often complementary to the anatomical information afforded by imaging studies.

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Stroke Rehabilitation

David Yu, MD

Introduction

Stroke is a non-traumatic, non-convulsive, non-neoplastic brain injury caused by occlusion or rupture of cerebral blood vessels that results in sudden neurological deficit often characterized by loss of motor control, altered sensation, cognitive or language impairment, disequilibrium or coma. Stroke is a neurological syndrome caused by a heterogenous group of vascular etiologies requiring a diverse approach to medical and rehabilitative management. Most stroke survivors experience functional improvement but the pattern, rate and ultimate outcome differ across patients and their respective social environments. The manifestations of and functional recovery from stroke are highly variable with respect to impairment (physiology), activities (disability), participation (handicap) and quality of life. In this section, some basic issues relevant to stroke rehabilitation are discussed. Given the complexity of stroke management and the rate of knowledge accrual in stroke, this section should be considered a brief introductory overview of stroke rehabilitation.

Epidemiology

Stroke is the number one leading cause of adult disability and the third leading cause of death in the United States. There are over 700,000 new strokes per year and over 4 million stroke survivors living in the United States. The incidence and prevalence of stroke in the United States are expected to increase due to the rising average age of the population and improved management of cardiovascular disease.

Anatomy and Physiology

The causes of stroke can be grossly categorized as hemorrhagic or ischemic. Intracranial hemorrhage accounts for 15% of all strokes and can be further divided into intracerebral (10%) and subarachnoid (5%) hemorrhage. Subarachnoid hemorrhages typically result from aneurysmal rupture of a cerebral artery with blood loss into the space surrounding the brain. Rupture of weakened vessels within brain parenchyma as a result of hypertension, arteriovenous malformation, or tumor also causes intracerebral hemorrhage.

The remaining 85% of strokes are caused by ischemic brain injury resulting from large-vessel (40%) or small-vessel (20%) thrombosis, cerebral embolism (20%), and other less common causes (5%), such as cerebral vasculitis or cerebral hypoperfusion. Vessel occlusion from thrombosis in both large and small arteries occurs most commonly in the presence of atherosclerotic cerebrovascular disease. Vascular changes or lipohyalinosis found in small, deep, penetrating arteries as associated with chronic hypertension can lead to small-vessel thrombosis. Cerebral emboli are usually of cardiac origin and are frequently a result of chronic ischemic cardiovascular disease with secondary ventricular wall hypokinesis or atrial arrhythmia, both conditions that increase the risk for intracardiac thrombus formation.
The etiology of an acute stroke can often be inferred by classifying the temporal profile of the event using information gathered from the patient's initial history and physical examination. A transient ischemic attack (TIA) is an event in which neurological symptoms develop and often disappear over several minutes and, by definition, completely resolve within 24 hours. TIAs are most frequently associated with atherosclerotic carotid artery disease and they should provoke an urgent diagnostic evaluation so that appropriate preventive care can be instituted.

Embolic strokes generally have a quick onset and fully develop in a matter of minutes, whereas hemorrhagic strokes often evolve over 1 to 2 hours. Thrombotic strokes can have a rapid or a prolonged interval of onset, lasting many hours. Stroke in evolution denotes an unstable ischemic event characterized by the progressive development of more severe neurological impairment, and it is often associated with active occlusive thrombosis of a major cerebral artery. Once a stable neurological status is reached, clinicians refer to the event as a completed stroke. The therapeutic goal of many current acute stroke treatment protocols is to minimize neurological deterioration and to limit the neurological impairment and resulting functional disability once the stroke is completed.

Impairments resulting from stroke are variable depending on the location, severity and duration of injury to brain structures. Various mechanisms have been proposed to explain the clinically observed phenomenon of spontaneous recovery of neurological functioning. These mechanisms generally can be divided into two broad categories. The first category is the resolution of local harmful factors, which usually accounts for early spontaneous recovery. These processes include resolution of local edema, elimination of local toxins and recovery of partially damaged ischemic neurons. The second category is often referred to as neuroplasticity and can take place early or late after brain damage. The concept of brain plasticity refers to the ability of the nervous system to modify its structural and functional organization. The two most plausible forms of plasticity are collateral sprouting of new synaptic connections and unmasking of previously latent functional pathways. Other mechanisms of plasticity include assumption of function by undamaged redundant neural pathways, reversibility from diaschisis, denervation supersensitivity and proximal regeneration of degenerated axons. Experimental evidence indicates that plasticity can be altered by external conditions including pharmacological agents, electrical stimulation and forced use of the affected limb. Though the mechanisms have not been definitively elucidated, repetitive use of the affected limb appears to be an essential adjunct for most, if not all, interventions to promote neurological recovery after stroke. Therefore, stroke rehabilitation not only seeks to maximize functional recovery by compensating for impairment but also seeks to promote neurological recovery via mechanisms such as neuroplasticity.

Medical Management

Medical management of stroke has evolved rapidly over the past decade. The standard of medical care for stroke is constantly changing. Therefore, the reader is strongly encouraged to review the current literature. Medical management of stroke can be divided into acute medical interventions, strategies to save the ischemic penumbra, management of stroke complications, secondary prophylaxis and risk factor reduction.

At the time of this writing, intravenous (IV) tissue plasminogen activator (TPA) is the only medical intervention that is widely accepted to reduce neurological injury after an ischemic stroke. TPA delivered intra-arterially or via devices that theoretically introduce the TPA closer to the region of the thrombus remain controversial though they are used in some centers. Despite the consensus that IV-TPA is effective in reducing neurological injury, only a minority of stroke
patients receive it due to a number of stringent indications and contraindications, most importantly IV-TPA must be given within three hours of a documented non-hemorrhagic stroke.

Strategies to save the ischemic penumbra include permissive hypertension for weeks following stroke in patients at low risk for hemorrhage into the injured brain region. Lacunar strokes generally don’t benefit from permissive hypertension due to the lack of significant ischemic penumbra.

Having a stroke is a risk factor for further strokes. Secondary prophylaxis refers to use of antiplatelet therapies and anticoagulation to prevent additional strokes in patients who have had a previous stroke. The standard of care in this area is changing rapidly. In choosing the appropriate secondary prophylaxis, one must consider a number of factors including any stroke prevention therapy used prior to stroke, the type of stroke, the stroke etiology, the risk of hemorrhagic conversion and contraindications such as the risk of intracranial bleeding due to a fall.

Risk factors for stroke can be categorized as modifiable or non-modifiable. Age is the primary risk factor for stroke but cannot be modified. Modifiable risk factors include hypertension, diabetes, dyslipidemia, sleep apnea, smoking, obesity and sedentary lifestyle. Hypertension is the most important modifiable risk factor for stroke. Appropriate medical management is obviously critical for a number of important risk factors, particularly hypertension, diabetes and dyslipidemia. However, it may not be as obvious that behavioral management is not only critical for risk factors such as smoking, obesity and sedentary lifestyle but also to ensure compliance with medical management of all risk factors. Patient and family education about stroke and its risk factors is a core component of stroke rehabilitation and serves as the cornerstone of behavioral modification that ultimately seeks to minimize the risk of further strokes.

Rehabilitation

Stroke rehabilitation is an interdisciplinary process that is tailored for individual stroke survivors in order to address a wide spectrum of medical issues, neurological impairments, social environments and individual needs. The broad goals of stroke rehabilitation are to maximize function, independence and quality of life. These goals can only be met through a holistic approach to patient evaluation and management. In this section, general principles of stroke rehabilitation will be discussed.

Stroke Rehabilitation Settings

The optimal clinical setting for rehabilitation varies between individual stroke survivors and often within a single stroke survivor over time. Factors that are useful in determining the ideal setting of care for a patient at a particular point in time include the patient's cognitive ability, motivation level, prior and present level of functioning, medical stability, level of available social resources, medical and nursing needs, and likelihood of achieving significant functional gains during rehabilitation. The availability of appropriate services and programs in the specific community must also be considered. Unfortunately, the decisions as to which level or location of care a patient might be directed to are occasionally based on the preferences of third-party payers and on the availability of specific resources within the community, rather than solely on patient need.

Acute inpatient stroke rehabilitation refers to the traditional interdisciplinary hospital-based coordinated program of medical, nursing, and therapy services. Care in this setting is directed by a physician and carried out by a team. This level of care is most appropriate for patients who
need and can tolerate 3 or more hours of therapy a day, and who may need more intensive nursing care and physician supervision. Patients in acute rehabilitation must have a reasonable likelihood of achieving significant functional gains within this setting.

Subacute inpatient rehabilitation is appropriate for stroke survivors who need comprehensive and coordinated therapy services for functional training in an institutional setting, but in a less intensive program than is used at the acute level of rehabilitation. Patients in this level of care usually receive between 1 and 3 hours of therapy per day. Assessment of the intensity of therapy services that an individual patient needs is based primarily on his or her tolerance level and on the specific needs for functional training. Some patients are unable to tolerate the full course of intensive rehabilitation because of medical frailty, limited endurance, severe cognitive impairment or global language dysfunction. Some patients who receive subacute rehabilitation have strokes that are extremely severe, limiting their potential to participate in intensive rehabilitation. Subacute inpatient rehabilitation is usually conducted in a skilled nursing facility.

Many stroke patients need traditional outpatient therapy services. These services are also provided in an outpatient clinic setting, but they do not entail the coordination, comprehensiveness, and team conferences that characterize acute rehabilitation. Most patients requiring acute rehabilitation eventually transition to an outpatient rehabilitation program. Patients with less severe impairment often can be discharged from the acute medical service to an outpatient program. Occasionally an outpatient program will consist of therapies conducted in the patient’s home. The home is the most familiar environment for the patient and family, and therapy in the home allows the patient and family to learn specific functional tasks in the setting in which those skills will be used most often. However, a potential disadvantage of home therapy is the limitation in available resources such as specialized equipment and the availability of additional clinical staff.

**Common Post-Stroke Impairments and Conditions**

Of all diagnostic groups requiring rehabilitation, stroke may carry the most diverse spectrum of potential impairments. Some of the most common post-stroke impairments and their rehabilitation management will be briefly discussed. Though post-stroke impairments are discussed individually, the reader is reminded that rehabilitation requires an interdisciplinary, holistic approach to care of stroke survivors. In order to maximize a particular function, all impairments and conditions affecting that function must be considered. For example, in-home ambulation may be a goal that requires management of hemiparesis, spasticity, proprioceptive loss and hemineglect. Further, the social environment for a given patient must also be considered. For example, an ambulatory stroke survivor with moderate cognitive deficits may be safe to ambulate within a familiar home environment but may require supervision of a family member in the community setting due to inadequate safety awareness and judgment in higher stimulation settings outside the home.

*Cognitive impairment* after stroke is most common in patients with cortical lesions but may also be seen with subcortical lesions. Cognitive impairments may be diffuse but are more commonly multifocal. For example, patients with parietal lesions may have Gerstmann’s syndrome which includes acalculia, or difficulty with calculation, alexia, difficulty reading, right-left disorientation and finger agnosia, difficulty in identifying and understanding the use of the hand on one side of the body. Language is a cognitive function. Aphasias are an impairment of language function that may be receptive, expressive or most commonly mixed, having some receptive and expressive components. Research suggests that the majority of improvement in cognition and language after stroke occurs within the first few months but may continue for 2 years or more.
**Speaking and swallowing difficulties** are common after stroke, particularly in posterior circulation strokes when the brain stem is involved but can also be seen with injury to cortical and subcortical structures. Dysarthria is difficult speaking due to mechanical problems that may include muscle weakness (e.g. due to cranial nerve involvement) or incoordination of muscles of speech (e.g. due to apraxia.) Impaired swallowing is referred to as dysphagia. Aspiration pneumonia is a common complication of stroke that is associated with significant morbidity and mortality that can result from mechanical swallowing difficulty or cognitive impairment. All stroke survivors should have some form of swallowing evaluation after diagnosis, prior to the resumption of oral intake. In patients who are cognitively intact with normal cranial nerve function, a simple bedside evaluation of swallowing by the physician may be adequate. In patients with diminished sensorium, moderate to severe cognitive impairment or cranial nerve involvement, a more detailed swallowing evaluation conducted by a speech pathologist, occasionally under fluoroscopy, may be indicated.

**Hemiparesis** refers to weakness of half the body on the opposite side of the brain lesion. Though the extent of recovery from hemiparesis varies from patient to patient, the pattern and timeframe for recovery are predictable for most stroke patients. The majority of recovery of muscle strength occurs within the first 3 months after stroke and reaches a plateau by 6 months. However, severely impaired stroke survivors are expected to recover less fully and take longer to reach a plateau than moderately or minimally impaired patients. The pattern of recovery is predictable for most patients with middle cerebral artery territory strokes. Initially there is a period of flaccid paralysis followed by return of muscle tone and reflexes followed by increased tone and hyperactive reflexes. Movement begins with patterned movement of groups before patients are able to voluntarily isolate individual muscle movement. In the upper limb, a flexor pattern is typical with shoulder flexion, adduction and internal rotation, elbow flexion, wrist flexion and finger flexion. In the movement synergy stage of recovery, upper limb flexor strength may be adequate to grasp objects but reaching and hand opening are not possible precluding functional use of the upper limb. Most activities of daily living can be performed with one neurologically intact upper limb. Thus, stroke rehabilitation must employ both compensatory techniques to allow the earliest functional independence while also employing techniques to encourage recovery of the impaired limb via the neuroplasticity techniques described earlier. In contrast, an extensor pattern of movement is typically seen in the lower limb. In some cases, when lower limb muscle strength remains weak, the extensor pattern of movement may actually assist patients with standing and weight bearing. The rate, pattern and stage of recovery have important functional implications for stroke rehabilitation.

**Spasticity** is a defining component of upper motor neuron syndromes including stroke. For stroke patients, spasticity increases resistance to passive movement and may result in problems such as loss of range of motion and contractures. Spasticity may also interfere with function by limiting active range of motion due to co-contraction of antagonist muscle groups such as commonly seen in patients with upper limb flexor synergy patterns of movement. In contrast spasticity may aid in weight bearing by increasing extensor tone in the lower limbs. Tools to manage spasticity after stroke may act systemically (e.g. oral spasmylytic medications) or focally (i.e. botulinum toxin injections). Management of spasticity must include a clear picture of the functional outcomes.

**Neurogenic bladder** is a general term for bladder dysfunction resulting from neurological injury. Approximately 60% of stroke survivors will have bladder involvement. In 90% of these cases, normal voiding function returns within 6 months of stroke. When the bladder is involved, it most commonly results from damage to brain structures that inhibit the pontine micturition center. The resulting type of neurogenic bladder is referred to as an uninhibited bladder or spastic bladder that is characterized by detrusor spasticity and coordinated sphincter function resulting in low void
volumes, frequency and urgency. Timed voiding, a strategy where patients are instructed to void on a scheduled basis with increasing intervals between voiding over time, is the mainstay of retraining the bladder. Occasionally, sphincter dyssynergia results from posterior circulation strokes that involve the pontine micturition center. These patients may benefit from alpha blockade to reduce sphincter tone or may need instrumentation to permit bladder drainage when severe.

Summary

Stroke rehabilitation requires an interdisciplinary, holistic approach to patient care. Impairments, social environments and individual needs vary immensely from patient to patient and often within a single patient over time. Stroke rehabilitation serves several functions including optimizing medical management, ensuring appropriate secondary prophylaxis, reduction of risk factors, management of comorbidities, prevention of common complications and most of all, addressing a diverse constellation of impairments to maximize function, independence and quality of life.

REFERENCES


Traumatic Brain Injury

Kathleen R Bell, MD

Traumatic brain injury (TBI) is the most common injury to the central nervous system with over 1,000,000 persons evaluated in emergency departments each year. The majority of TBIs are mild to moderate (80%) in nature. However, about 230,000 people are admitted to the hospital and survive until discharge; another 50,000 die each year. Males are affected more frequently than females (2-3:1 ratio). Peaks of incidence occur in the age range of 15-25 years of age and over 75 years of age.

The causes of TBI vary according to age and severity of injury. In general, motor vehicle crashes are the most common cause of injury (48%), followed by falls (23%), violence (firearm-related 10% and nonfirearm-related 9%), and sports injuries (3%). However, in the age group over 75, falls on level ground are most common. Fatal TBI most commonly result from firearm-related injuries.

Types and Mechanisms of Traumatic Brain Injury

TBI can be caused by a direct blow to the head (as in a fall or assault) or by acceleration-deceleration forces applied to the head (as happens in some motor vehicle crashes). This means that injuries do not always require actual trauma to the head. The forces transmitted to the brain result in the primary injury. This may be a transient neurological dysfunction in a mild TBI or may be visible as a contusion or mass lesion from direct forces. Primary focal brain contusions occur most often at the frontal poles, orbital frontal lobes, temporal poles and lobes and cortex above the Sylvian fissure. Additionally, ruptured blood vessels may result in intracranial hematoma, extradural hematoma (associated with skull fractures), and acute subdural hematoma. Pressure causes nearby ischemia. Rotational forces may result in widespread white matter injury and petechial bleeding caused by axonal disruption in more severe injuries (also called diffuse axonal injury or DAI).

Perhaps as destructive, secondary injury to the brain occurs as a result of cell disruption and inflammatory reactions. Ionized calcium is the primary factor responsible for reactive axonal changes. Ca++ activates catabolic enzymes resulting in damage to structural proteins and activates phospholipases that break down cell membrane. This leads to the production of oxygen free radicals and superoxide radicals. Ca++ also initiates glutamate neurotoxicity by stimulating its release. Clinically, this appears as ischemia and spasm and edema.

Severity and Outcome

Generally speaking, factors that predict mortality after TBI (blood glucose, intracranial pressure, papillary reactivity) do not predict functional outcome. Clinicians use the Glasgow Coma Scale (GCS) to quickly define the severity of an injury. There are problems in its use, however, because of the early intubation clouding verbal response and ceiling effects for mild injuries. However, the motor score for the GCS is a relatively good early predictor of outcome. Not surprisingly, a lower GCS score correlates with worse outcomes. Other clinical predictors include the duration of coma and post-traumatic amnesia (PTA), the lack of continuous memory after the injury. The end of PTA is not easy to pinpoint; the time to follow commands is a more objective way to determine improvement.
Computerized tomography (CT) scans are useful in detecting focal trauma amenable to surgical intervention but does not contribute to predicting outcome except as a gross measure of injury burden. MRI scans are more sensitive to smaller lesions but still may not predict outcome, especially in mild TBI. Neuropsychological examination is the best way to determine the range of functional deficits and to plan rehabilitation.

**Consequences of Traumatic Brain Injury**

Mortality. As noted, about 50,000 people die each year from TBI. TBI is responsible for a quarter of the deaths occurring due to motor vehicle crashes. Perhaps another 10% of injuries result in prolonged or minimally responsive states. Half of those in these states will die within the first 12 months after injury.

Physical Consequences. The medical

<table>
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<tr>
<th>Cardiovascular System</th>
<th>Autonomic dyscontrol</th>
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<tr>
<td></td>
<td>Hypertension</td>
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<td>Respiratory System</td>
<td>Central and mixed sleep apnea</td>
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<td>Musculoskeletal system</td>
<td>Undiagnosed fractures</td>
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<td>Joint contractures</td>
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<td>Heterotopic ossification</td>
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<td>Nervous System</td>
<td>Sensory</td>
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<td></td>
<td>Traumatic vision syndrome</td>
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<td>Visual field loss</td>
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<td>Anosmia</td>
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<td>Motor control</td>
<td>Spasticity</td>
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<td>Rigidity</td>
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<td>Ataxia</td>
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<td>Tremors</td>
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<td>Apraxia</td>
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<td>Paresis</td>
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<td>Balance</td>
<td>Benign paroxysmal positional vertigo</td>
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<td>Late mass lesions</td>
<td>Chronic subdural hematoma</td>
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<td></td>
<td>Hydrocephalus</td>
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<td>Abscess, empyema</td>
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<td>Seizures</td>
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<td>Headache</td>
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<td>Endocrine System</td>
<td>SIADH, Cerebral salt wasting, Diabetes insipidus</td>
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<td></td>
<td>Hypothyroidism</td>
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<td></td>
<td>Hormonal disruption</td>
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<td>Gastrointestinal System</td>
<td>Dysphagia</td>
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<td>Genitourinary System</td>
<td>Urinary urgency</td>
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<tr>
<td>Immune System</td>
<td>Predilection to infection</td>
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<tr>
<td>Hematologic System</td>
<td>Thromboembolic disease</td>
</tr>
</tbody>
</table>

Neuropsychological and Neurobehavioral Function. Neuropsychological problems are best described as difficulties in concentrating, remembering new information, processing information quickly, flexibility in thinking, abstract reasoning, or solving new problems. In more severe injuries, this may also include very basic and specific alterations in key language.
capacities and in visual-perceptual and visual-mechanical skills. The significance of cognitive deficits is primarily linked to the severity of the brain injury. The degree of neuropsychological impairments is greater soon after injury with recovery occurring over days, weeks, months, and maybe years. Degree of improvement and degree of residuals depend on degree of original loss. Formal neuropsychological examinations, in which standardized tests of ability in many different areas (e.g., visual and auditory memory, problem-solving, visuospatial abilities, working and delayed memory, etc.) are tested, are the best way to objectively describe these deficits. However, these tests must be reviewed in the context of prior education and abilities and personality/affective influences.

Emotional behavioral difficulties are common sequelae of TBI and are major barriers to resuming prior social roles and responsibilities. These neurologically-based personality and behavioral changes can include problems with awareness of deficit, lack of initiative, impulsivity, disinhibition, rage reactions, loss of empathy or concern for others, flat affect, inappropriate comments and behaviors, uncontrolled laughing or crying, or emotional lability. These injury-related problems are all distinct from normal reactive emotions such as depression and anxiety, or from premorbid vulnerabilities.

The rate of depression in the first year after TBI is about 30% and is frequently accompanied by anxiety or irritability. Complicating the picture is the likely existence of acute stress syndrome (ASS) and post-traumatic stress disorder (PTSD) in many survivors of TBI. This is somewhat controversial as most patients with a significant TBI have no memory of the actual event which is thought to be necessary for the formal diagnosis of PTSD.

**Functional Status**

Traumatic brain injuries can leave those that survive them with various limitations or disabilities in everyday life. These limitations may involve basic areas such as personal care and ambulation to higher-level functions such as social relationships, work, and leisure. In addition, substantial problems may persist for long periods of time in areas of employment, recreation, and cognitive functioning and to a lesser extent in almost all areas assessed including in sleep and rest, emotional behavior and ambulation. In one study by Dr. Dikmen and her group, recovery to pre-injury levels ranged from 65% in personal care to 40% in cognitive competency, major activity and leisure and recreation in persons who were 3-5 years out from their injuries.

**Mild Traumatic Brain Injury**

Most cases of TBI are mild in nature. It is estimated that well over a million cases of mild TBI (MTBI) occur each year; firm statistics are not possible because many are not treated in emergency departments from which these estimates are gathered. At present, the recommended conceptual definition of MTBI by the Centers for Disease Control is an injury to the head as a result of blunt trauma or acceleration or deceleration forces that result in one or more of the following conditions:

Any period of observed or self-reported:
1) transient confusion, disorientation, or impaired consciousness,
2) Dysfunction of memory around the time of injury, and
3) Loss of consciousness lasting less than 30 minutes.
In addition, there may be observed signs of neurological or neuropsychological dysfunction, such as:
1) Seizures acutely following injury to the head,
2) For infants and young children, irritability, lethargy, or vomiting following head injury, and
3) Symptoms among older children and adults such as headache, dizziness, irritability, fatigue, or poor concentration, that when identified soon after injury can be used to support the diagnosis of MTBI.

While for most clinical purposes, MTBI are classified as such on the basis of the above criteria and a Glasgow Coma Scale score of 13-15, there have been a number of gradation scales that have been made for sports-related concussion that relate to return to play criteria.

Generally, recovery from a MTBI is rapid and the vast majority of people are back to baseline within three months. However, about 10% have persisting symptoms such as insomnia, memory difficulties, sensitivity to light and sound, fatigue, headaches, slow performance, poor concentration, anxiety, irritability, word-finding difficulties, distractibility, poor balance, and difficulty in thinking. Treatment consists of pain amelioration, remobilization, counseling, and assistance in returning to normal activities.

**Prevention of TBI**

Research has supported the use of protective head gear in reducing the mortality and morbidity associated with head injury. For instance, the use of bicycle helmets, for instance, reduced the risk for bicycle-related head injury in Seattle by 74-85% in a study by Rivera and associates. Although motorcycle helmets have also been proven to dramatically decrease mortality in accidents, many states have repealed helmet laws with a resultant increase in deaths and disability. There is also some evidence that the use of seatbelts and airbags combined with seatbelts results in a decrease in facial and head trauma. In sports with high incidences of concussion and head injury, improvements can be made somewhat safer with appropriate gear and behavior regulation.

**Glasgow Coma Scale**

<table>
<thead>
<tr>
<th>Eye Opening</th>
<th>Spontaneous 4</th>
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<tbody>
<tr>
<td></td>
<td>To Voice 3</td>
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<tr>
<td></td>
<td>To Pain 2</td>
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<tr>
<td></td>
<td>None 1</td>
</tr>
<tr>
<td>Verbal Response</td>
<td>Oriented 5</td>
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<tr>
<td></td>
<td>Confused 4</td>
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<tr>
<td></td>
<td>Inappropriate 3</td>
</tr>
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<td></td>
<td>Incomprehensible 2</td>
</tr>
<tr>
<td></td>
<td>None 1</td>
</tr>
<tr>
<td>Motor Response</td>
<td>Obeys Command 6</td>
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<tr>
<td></td>
<td>Localizes Pain 5</td>
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<td></td>
<td>Withdraws to Pain 4</td>
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<tr>
<td></td>
<td>Flexion 3</td>
</tr>
<tr>
<td></td>
<td>Extension to Pain 2</td>
</tr>
<tr>
<td></td>
<td>None 1</td>
</tr>
<tr>
<td>Total Score</td>
<td>1 – 15</td>
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Grading Systems for Concussion

<table>
<thead>
<tr>
<th>Grade</th>
<th>Cantu</th>
<th>Colorado Medical Society</th>
<th>AAN Practice Parameter</th>
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<tbody>
<tr>
<td>1</td>
<td>No loss of consciousness (LOC); posttraumatic amnesia (PTA) less than 30 minutes</td>
<td>Confusion without amnesia; no LOC</td>
<td>Transient confusion; no LOC; concussion symptoms or mental status abnormalities on examination resolve in ≤ 15 minutes</td>
</tr>
<tr>
<td>2</td>
<td>LOC ≤ 5 minutes in duration or PTA lasting ≥ 30 minutes but ≤ 24 hours</td>
<td>Confusion with amnesia; no LOC</td>
<td>Transient confusion; no LOC; concussion symptoms or mental status abnormalities on examination last ≥ 15 minutes</td>
</tr>
<tr>
<td>3</td>
<td>LOC for &gt; 5 minutes or PTA for &gt; 24 hours</td>
<td>LOC</td>
<td>Any LOC, either brief (seconds) or prolonged (minutes)</td>
</tr>
</tbody>
</table>

REFERENCES

History of Spinal Cord Injury

Before 1940, a complete spinal cord injury (SCI) was fatal. Patients died in months from sepsis, often due to a urinary tract infection, pneumonia or an infected pressure sore. For example, in 1881, James A. Garfield, the 20th U.S. President, died from sepsis within 3 months after a gunshot wound to the thoracic spinal cord. In World War I, 80% of those with spinal cord injury died within one year; those who survived had incomplete SCI. In the 1940s, thousands of veterans returned from World War II with spinal cord injury; through pioneering clinical care and rehabilitation by physicians such as Donald Monro, Ernest Bors, Estin Comarr and Ludwig Guttman, much was learned about preventing medical complications and how to return these patients to their communities. Since then, surgical, medical, and rehabilitative interventions have steadily improved and mobility aids, adaptive equipment and accessibility have increased, so that most SCI persons now approach normal life expectancies and are partly or completely independent (Table 1). Almost all with new onset SCI or non-traumatic myelopathy will benefit from multi-disciplinary rehabilitation, to optimize independence and to teach them how to prevent late complications.

Epidemiology/Classification

Traumatic SCI incidence in the U.S. is about 10,000 per year; prevalence is about 250,000. Roughly 80% are male and peak age at onset is 19. Vehicular accidents account for about 40%, falls about 22%, violence about 18%, and sports about 8%, with diving being the most common sport injury. About half have tetraplegia and half paraplegia; about 30% recover enough lower limb function to transfer independently and to do some walking.

Classification of traumatic SCI is by most caudal functional level and by degree of incompleteness. Key indicator muscles are used to classify the last functional level; the last normal sensory level is used, if there are no key indicator muscles for that level (Table 2). Completeness of SCI is classified by the American Spinal Injury Association (ASIA) Impairment Scale (Table 3) and by the clinical syndrome, reflecting the anatomy of the cord damage (Table 4). Neurologic exam of voluntary movement, sensation (pain- and temperature-sense for the crossed anterolateral spinothalamic tract; position- and vibration-sense for the posterior columns) and segmental reflexes allows physicians to describe the segmental and longitudinal extent of damage to upper motor neurons and lower motor neurons. Loss of segmental reflexes caudal to an acute SCI is called spinal shock; the bulbocavernosus reflex typically returns within days (unless there is lower motor neuron damage to the S2-S4 segments or cauda equina) and tendon reflexes return in weeks and then become hyperactive over several months, a major cause of spasticity.¹

Non-traumatic spinal cord disease or myelopathy often result from congenital spinal stenosis, degenerative spinal stenosis, spine instability and/or central disc herniation at cervical, thoracic or lumbar spinal levels. C1-C2 instability (i.e. atlanto-axial subluxation) is common in patients with rheumatoid arthritis or Down’s syndrome. Other non-traumatic causes of myelopathy include epidural abscess, spinal cord tumor, spinal cord vascular malformations, vitamin B12 deficiency, syringomyelia,
multiple sclerosis and HIV. Early diagnosis and treatment assure the greatest chance for recovery from
the myelopathy.

**Acute Management**

Spine instability with risk for spinal cord injury must be considered in all patients with major trauma
and in any patient with mild trauma and complaints of neck or back pain or complaints of sensory loss
or weakness. The spine must be immobilized during transport. In the ER, spine x-rays or CT scans are
used to rule out spine fracture or instability. If the patient has paraplegia or tetraplegia, then spinal
immobilization is maintained, Neurosurgery and/or Orthopedic Surgery are consulted to consider spine
surgery to decompress the spinal cord and to stabilize the spine. A urethral indwelling catheter is
placed to assure bladder drainage. Spine external immobilization is continued post-surgical
stabilization for 2-3 months with a cervical brace or a thoracolumbosacral orthosis (TLSO or body
jacket) until the bone fuses.

Aggressive preventive measures are instituted to address common acute complications. Low-
molecular weight heparin injections and sequential compression devices are begun to prevent
DVT and pulmonary emboli. Special mattresses and regular turning in bed are used to prevent
pressure sores. Pulmonary hygiene measures are started to prevent pneumonia. H2-blockers are
given to prevent stress gastritis. Daily bowel programs (i.e. rectal suppository & digital
stimulation) are used to prevent fecal impaction. When medically stable, patients are transferred
for initial rehabilitation.

**Initial Rehabilitation**

During initial rehabilitation, the SCI patient prepares to return to the community. A multi-
disciplinary rehabilitation team facilitates this process. Most will return home though some will
need caregiver assistance; 5-15% will go to assisted living or to a nursing home.

The SCI physician oversees the whole rehabilitation process, integrating the efforts of the multi-
disciplinary team, including physical therapists, occupational therapists, rehabilitation nurses, social
workers, psychologists, recreation therapists, vocational counselors and for children, a teacher. One
focus is to optimize functional recovery; this includes long-tract recovery in those with incomplete SCI
and zone-of-injury recovery in all with either complete or incomplete SCI. Interventions to optimize
recovery include:

1. assure spinal stability and adequate decompression of spinal cord and nerve roots (Fig 1),
2. assure adequate nutrition and stable metabolic status,
3. provide active exercise and functional training to optimize activity-dependent recovery of
   function,
4. give adaptive equipment and training to allow patients to regain function, and
5. facilitate home modifications.

Another focus is to prevent long-term complications; patients are provided with adaptive equipment
and educated in ways to prevent pressure sores, urinary tract infections, fecal impaction, contractures,
falls, pneumonia, autonomic dysreflexia, etc. A third focus is to facilitate the patient’s adjustment to
disability, which is more complex if there is associated psychiatric illness, traumatic brain injury or
recent alcohol / drug abuse; psychologists counsel patients, social workers discuss future finances and
inter-personal relationships, recreation therapists take patients into the community on outings, vocational counselors explore future work or school opportunities and the rehabilitation team educates family and caregivers regarding the patient’s future needs.

Neurologic recovery is common in those with incomplete SCI. This recovery is, in part, mediated by activity-dependent synapse growth in spared descending motor pathways; thus, active exercise is key to optimizing this recovery. Typically those with incomplete SCI who achieve some functional recovery below their SCI will begin to show some voluntary movement within a week of injury; this recovery will then progress over 3 to 6 months to functional recovery. Even those with complete SCI can regain one or two segmental levels of function at the zone of injury – e.g. a C5 tetraplegic may become a C6 tetraplegic patient over 6 months as they regain function in the C6 segmental level. Those with complete SCI at one month post-injury are not likely to regain functional use in their lower limbs. Much research is underway presently to attempt to achieve axon regeneration across a spinal cord injury and allow functional recovery, even in those with complete SCI; progress has been achieved in facilitating long-distance regeneration of spinal axons but much remains to be accomplished in assuring synaptic regrowth to appropriate neuronal targets in the distal cord.

Functional outcomes after SCI depend on the SCI level and SCI incompleteness, as well as on other factors such as age, other co-morbidities, and psychological status. Expected functional outcome for those with complete SCI are established (Table 5).

Sexual function and fertility are affected by SCI. The experience of orgasm is altered after SCI for both males and females. Sexual arousal is usually preserved but psychogenic erections are usually absent in males with complete SCI. Males often have reflex erections but these may be insufficient for vaginal penetration; external vacuum devices, penile injections of alprostadil and oral sildenafil can enhance erections. Male fertility is often impaired due to poor sperm quality and due to retrograde ejaculations; electroejaculation, artificial insemination and in-vitro fertilization can improve fertility. Females with SCI have normal fertility, though labor and delivery may be complicated by autonomic dysreflexia (AD) and there is more likelihood of caesarean section. SCI patients should be offered education and should be encouraged to ask questions about their sexual health during initial rehabilitation and later.

Health Maintenance / Long-Term SCI Complications

SCI patients need routine primary care but also have special preventive care needs due to their unique risks for long-term SCI complications.

- **Pressure sores** or decubitus ulcers are one such risk. With an appropriate wheelchair, wheelchair cushion, bed and mattress and with pressure releases every 15 minutes while sitting in the wheelchair and with regular turns while in bed every 2-4 hours and with daily skin checks and early treatment, most severe pressure sores can be prevented. If a severe pressure sore develops, then treatment with either prolonged bedrest such as in a fluidized sand bed (i.e. Clinitron) or a myocutaneous flap with 5-8 weeks post-flap immobilization / remobilization is usually needed.

- **Urinary tract infections** and **renal stones** are another risk, since most with SCI have a neurogenic bladder. Avoiding indwelling catheters can be achieved by training patients to perform intermittent bladder catheterization 4-6 times per day; for those with limited hand function, an indwelling catheter may be the most practical option. Obtaining kidney-ureter imaging yearly is advised to direct early treatment of hydronephrosis or renal stones, before renal function is compromised or before a renal stone displaces into the ureter and causes urosepsis, which may be clinically silent in tetraplegic and high paraplegic patients (i.e. they may not manifest ureteral colic).
• **Common gastrointestinal complications** are bowel incontinence, prolonged bowel programs and fecal impaction. Most SCI patients have an upper-motoneuron bowel, which will yield mass reflex evacuation of descending and sigmoid colon with rectal stimulation; thus, a daily or every other day bowel program consisting of a rectal suppository and digital stimulation x3 will often yield bowel results in 30-60 minutes and prevent bowel incontinence. If with aging this reflex evacuation wanes, then bowel programs can become excessively prolonged or patient’s may experience recurrent fecal impaction; in these persons, a colostomy is often warranted for efficient, effective bowel management. In SCI patients with a lower motor neuron bowel (i.e. conus medullaris or cauda equina SCI level), daily or twice daily bowel programs are usually needed and consist of digital stool removal from the rectum, since no reflex evacuation is possible; these patients may also benefit from a colostomy, if bowel care is problematic.

• **Common cardiovascular complications** include orthostatic hypotension and autonomic dysreflexia (also called autonomic hyperreflexia). Those with tetraplegia and high paraplegia will typically have low blood pressures – e.g 90/60. They may have orthostatic hypotension and may need to wear an abdominal binder and venous compression stockings and may need to take ephedrine or midodrine to prevent such orthostasis. These patients also have impaired temperature regulation and need extra protection from high and low environmental temperatures. They also can experience autonomic dysreflexia (AD) with acute hypertension, where bladder overdistension or other noxious input from below their SCI level (e.g. fecal impaction, acute abdomen, labor and delivery in a pregnant SCI female) elicits a reflex sympathetic vasoconstriction; such autonomic dysreflexia can cause acute hypertension with systolic blood pressures of 260 mm Hg or higher and put patients at risk of intracerebral hemorrhage. Treatments usually include: 1) elevate the patient’s head, 2) identify and reverse any treatable causes of AD (e.g. an obstructed urethral catheter), 3) apply nitropaste and/or give oral hydralazine. During rehabilitation, SCI patients are educated about how to manage orthostatic hypotension and AD at home.

• **Pulmonary complications** are common, particularly in those with tetraplegia; these include obstructive sleep apnea (OSA), late respiratory failure and recurrent atelectasis, mucus plugging and pneumonia. SCI patients have a higher prevalence of obstructive sleep apnea than able-bodied persons; this is partly due to medications for pain and spasticity and in partly due to altered respiratory mechanics. Sleep studies should be considered for those with OSA, treatment with continuous positive airway pressure (CPAP) or bilevel positive airway pressure (BiPAP) attempted. Late respiratory failure can develop, particularly in those with high level tetraplegia and a history of marginal respiratory function; pulmonary function tests should be monitored regularly in those at risk. Recurrent atelectasis, mucus plugging and pneumonia are common in those with high tetraplegia, particularly in those who continue to smoke. These individuals usually have ineffective cough due to low peak cough flows from reduced inspiratory volumes and absent forced expiration from absent voluntary abdominal muscle contractions. Using assisted coughs (manual abdominal compression) and a cough assist machine (i.e. mechanical insufflator-exsufflator) in conjunction with postural drainage and mucolytic agents can often achieve adequate pulmonary hygiene to prevent these complications.

• **Neurologic complications** include spasticity, neuropathic pain, inadequate spinal decompression, peripheral nerve entrapment and post-traumatic syringomyelia. Spasticity is managed with optimal wheelchair seating, bracing, oral medications (e.g. baclofen, tizanidine, diazepam, clonidine, gabapentin, dantrolene), nerve blocks (e.g. alcohol, botulinum toxin) or intrathecal baclofen pumps. An acute increase in spasticity and AD are often warnings of an acute medical problem (e.g. ureteral stone, acute abdomen, urinary tract infection) in tetraplegic patients with impaired sensation. Neuropathic pain is common in SCI patients and is severe in about 25%; there are no fully effective treatments but gabapentin and other anti-epileptics (carbamazepin, lamotrigine, etc.) and opioids can help reduce pain. Late neurologic decline can occur in about 20% of SCI patients; common causes are inadequate spinal
cord decompression or other spinal stenosis, peripheral nerve entrapment and post-traumatic syringomyelia. About 5% of SCI patients develop an enlarging fluid-filled cyst (i.e. syrinx) at the zone of spinal cord injury; such a syrinx can lead to ascending weakness and loss of function (Fig 2). Characteristic early findings are ascending loss of tendon reflexes and dissociated sensory loss (i.e. loss of pain & temperature sensation, but preserved touch & position-sense). Prompt diagnosis and treatment can minimize long-term functional decline. A spinal cord MRI is diagnostic. Surgical reconstruction of the subarachnoid space with dural adhesion dissection and a dural graft or surgical placement of syringo-peritoneal shunt can decompress the syrinx and prevent further functional decline in about half the cases.

Figure 1. Residual Cauda Equina Compression from L2 Burst Fracture

This CT scan of the L2 vertebral body shows markedly narrow spinal canal with likely persisting compression of the nerve roots of the cauda equina despite placement of posterior rods to stabilize the spine; this L2 burst fracture resulted from a sky-diving accident. The schematic shows the two posterior rods (thin arrows) and the normal canal size for accommodating the cauda equina (thick arrow), which is encroached upon by bone fragments from the L2 burst fracture. This patient with initial incomplete paraplegia, ASIA C, experienced declining lower limb strength after transfer to the rehabilitation hospital. He underwent subsequent anterior spinal decompression at L2 and then had marked recovery of lower limb strength and function.

Figure 2. MRI of Post-Traumatic Syrinx  This MRI shows a post-traumatic syrinx extending rostrally to the medulla from the T4 level of paraplegia; in this T1-weighted MRI, the fluid of the subarachnoid space and of the syrinx are dark and the neural tissue of the cerebellum, medulla and spinal cord are light. The schematic shows the medulla and cervical spinal cord as gray (arrows) and the syrinx as black. This syrinx caused loss of hand strength at 9 months post-SCI. Surgical decompression and a dural graft at T3 to T5 yielded full recovery of hand strength.
• Common **musculoskeletal complications** include **muscle / joint capsule contractures, heterotopic ossification, shoulder pain** and **osteoporosis**. Daily range-of-motion exercises can prevent muscle contractures and joint capsule tightness, although in tetraplegic patients a caregiver may need to provide this daily stretching. About 5% of SCI patients can develop severe abnormal bone formation in soft tissue around large joints, most often anterior to the hip joint. This abnormal bone formation is called heterotopic ossification (HO), and it typically begins within the first few months post-injury. For the majority of patients, there is no functional impact from HO; however, it can lead to impaired wheelchair seating and impaired transfers because of limited hip joint motion. Early diagnosis and treatment with NSAIDs, etidronate and/or local radiation therapy can prevent the functional decline due to HO. Many SCI patients will develop late pain in shoulders, overused by weight-bearing during transfers and manual wheelchair propulsion. An optimal manual wheelchair prescription, optimal training in manual wheelchair use and transfers, appropriate exercises, weight loss, and pain medications can help to manage such shoulder pain; for those with severe degenerative changes in the shoulders, a power wheelchair may be needed. Osteoporosis of lower limb long bones but not of the spine is characteristic of SCI; minor trauma can lead to femur or tibial fractures. There is, as yet, no known treatment to prevent or reverse this osteoporosis, as weight-bearing in the absence of muscular contraction (e.g. use of a standing frame) does not prevent this complication. Preliminary studies suggest that bisphosphonates may slow osteoporosis development in SCI, but long-term results from this treatment have not been reported.

**Summary**

SCI patients are often young at onset and live for decades with physical limitations and unique health risks of paraplegia or tetraplegia. Comprehensive initial rehabilitation and individualized long-term preventive care can optimize functional independence and quality of life.
Table 1: Years of Life Expectancy, if survive >24 hours

<table>
<thead>
<tr>
<th>Age @ Onset</th>
<th>Able-Bodied</th>
<th>ASIA D</th>
<th>Ventilator-Dependent</th>
<th>C5-C8 Tetraplegia</th>
<th>Paraplegia</th>
</tr>
</thead>
<tbody>
<tr>
<td>20</td>
<td>57</td>
<td>50</td>
<td>15</td>
<td>38</td>
<td>44</td>
</tr>
<tr>
<td>40</td>
<td>38</td>
<td>32</td>
<td>7</td>
<td>22</td>
<td>27</td>
</tr>
<tr>
<td>60</td>
<td>21</td>
<td>16</td>
<td>1</td>
<td>9</td>
<td>12</td>
</tr>
</tbody>
</table>

* from SCI Model Systems, U Alabama Birmingham

Table 2: Level of SCI – The Last Functional Level*

<table>
<thead>
<tr>
<th>SCI Level</th>
<th>Dermatomal Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>C1-C4</td>
<td>Use C1-C4 dermatomes</td>
</tr>
<tr>
<td>C5</td>
<td>Elbow Flexors</td>
</tr>
<tr>
<td>C6</td>
<td>Wrist Extensors</td>
</tr>
<tr>
<td>C7</td>
<td>Elbow Extensors</td>
</tr>
<tr>
<td>C8</td>
<td>Finger Flexors (flexor digitorum profundus, 3rd digit)</td>
</tr>
<tr>
<td>T1</td>
<td>5th Finger Abduction</td>
</tr>
<tr>
<td>T2-L1</td>
<td>Use T2-L1 dermatomes</td>
</tr>
<tr>
<td>L2</td>
<td>Hip Flexors</td>
</tr>
<tr>
<td>L3</td>
<td>Knee Extensors</td>
</tr>
<tr>
<td>L4</td>
<td>Ankle Dorsiflexors</td>
</tr>
<tr>
<td>L5</td>
<td>Great Toe Extensors</td>
</tr>
<tr>
<td>S1</td>
<td>Ankle Plantarflexors</td>
</tr>
<tr>
<td>S2-S4</td>
<td>Use S2-S4 dermatomes</td>
</tr>
</tbody>
</table>

* SCI level is most caudal segmental level with ≥3/5 strength (provided all rostral levels are 5/5). If no key muscles available, then SCI level is last dermatomal level with normal sensation.

Table 3: ASIA Impairment Scale*

| ASIA A | Motor & sensory complete SCI |
| ASIA B | Motor complete, sensory incomplete SCI |
| ASIA C | Motor & sensory incomplete SCI but motor non-functional® |
| ASIA D | Motor & sensory incomplete SCI with motor functional |
| ASIA E | Full motor & sensory recovery |

* ASIA is the American Spinal Injury Association.
  ® Incomplete sensory & motor require that anal sensation & anal sphincter muscle contraction are present respectively. Functional is defined as ≥50% of lower limb muscles have ≥3/5 strength.
### Table 4: Clinical Syndromes of Incomplete SCI

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Brown-Sequard</td>
<td>Asymmetric cord injury with more ipsilateral weakness and more contralateral loss of pain &amp; temperature sensation due to crossing anterolateral spinothalamic tract. Functional motor and bladder / bowel recovery are common.</td>
</tr>
<tr>
<td>Central Cord</td>
<td>Greater hand than lower limb weakness, due to cervical cord injury with greater gray matter &amp; central white matter damage than peripheral white matter damage of long-tracts. Functional motor recovery in lower limbs and bladder / bowel recovery are common.</td>
</tr>
<tr>
<td>Anterior Cord</td>
<td>Damage to anterior spinal cord, often infarct due to compromised anterior spinal artery, is the cause. Posterior columns are spared, so light touch, vibration and position-sense are spared. Pain &amp; temperature sensation are impaired, as is long-tract motor function. Functional motor &amp; bladder / bowel recovery are uncommon.</td>
</tr>
<tr>
<td>Conus Medullaris</td>
<td>Damage to caudal spinal cord neurons of S2 to S4 segments causes impaired bladder, bowel, and sexual functions. Recovery of voluntary bladder / bowel function is uncommon. May also have involvement of L5, S1 segments with impaired gait.</td>
</tr>
<tr>
<td>Cauda Equina</td>
<td>Damage to the cauda equina, coursing through the spinal canal at L2 to L5 spinal levels leads to paraparesis and bladder / bowel impairments. Common etiologies are central lumbar disc herniation or lumbar vertebral body burst fracture. Partial motor recovery is common.</td>
</tr>
</tbody>
</table>

### Table 5: Functional Outcomes Expected for Complete SCI

<table>
<thead>
<tr>
<th>SCI Level</th>
<th>Mobility</th>
<th>Self-Care</th>
</tr>
</thead>
<tbody>
<tr>
<td>C1-C4</td>
<td>Drive power WC with chin or sip tube control.</td>
<td>Use computer with voice-commands. Use ECU (environmental control unit).</td>
</tr>
<tr>
<td>C5-C6</td>
<td>Drive power WC with hand-control.</td>
<td>Feed self. Partially dress self. Use computer with hand-sticks.</td>
</tr>
<tr>
<td>C7-C8</td>
<td>Transfer self. Use manual WC. Drive an adapted van or car with hand-controls.</td>
<td>Bath and dress self. Perform bowel care. Use computer with hand-sticks.</td>
</tr>
<tr>
<td>T1-L3</td>
<td>Transfer self. Use manual WC. Drive a car with hand-controls.</td>
<td>MANAGE ALL SELF-CARE. USE A COMPUTER WITH TOUCH-TYPING.</td>
</tr>
<tr>
<td>L4-L5</td>
<td>Walk with bracing and crutches or walker. Use manual WC for long distances.</td>
<td>Manage all self-care.</td>
</tr>
<tr>
<td>S1-S4</td>
<td>Walk with limited bracing. WC not needed.</td>
<td>Manage all self-care.</td>
</tr>
</tbody>
</table>

WC = wheelchair, ECU = environmental control unit.
REFERENCES


Rehabilitation in Multiple Sclerosis

Kevin T. Caserta, MD and George H. Kraft, MD, MS

Background

Multiple Sclerosis (MS) is an autoimmune disease directed against the myelin of the central nervous system (brain, spinal cord, and optic nerve). However, we know from autopsy that axonal loss is also prevalent. There are a wide range of presenting symptoms with common complaints including visual disturbances, numbness, weakness, and fatigue. The disease is progressive and over time significant disability can occur. It is the number one cause of neurological disability in young adults between the ages of twenty and fifty. Prevalence has been described between 40-220/100,000. Women are affected over men 2:1.

The cause of MS, like most autoimmune diseases, is still unknown; however, there are genetic as well as environmental predispositions to the disease. Geographically, the further one is raised from the equator prior to the age of 15, the more likely one is to develop MS. Infections by certain viruses have also been postulated to contribute to the disease.

In MS CD4+ helper T cells cross the blood brain barrier, initiating macrophage and other inflammatory cell destruction of myelin. Central myelin, composed of the membranes of oligodendrocytes, allows rapid conduction of electrical signals critical for nerve function. Depending on the extent of damage, nerve conduction is delayed or even stopped completely. The oligodendrocytes can re-myelinate axons after the inflammatory cascade. However, over time sufficient damage prevents full neurological recovery and leads to the development of impairments. In later stages, and even early in the disease course, axonal death may occur. This axonal loss from the inflammation and/or independent degenerative processes eventually results in brain atrophy.

Diagnosis

Traditionally, MS has been primarily a clinical diagnosis, requiring two separate central nervous system attacks separated by space and time. With advancement in radiological techniques, MRI is becoming the gold standard for confirming MS. Evidence of demyelination is best visualized on the T2 and FLAIR MRI sequences. Classically, the lesions are periventricular and in advanced disease can radiate in a near fan like pattern from the lateral ventricles. Known as Dawson’s Fingers, this pattern of damage represents significant disease. In more advanced disease brain atrophy is present representing axonal loss. Gadolinium can be helpful to assess recent attacks, as it will aggregate in areas of recent damage to the blood brain barrier.

Evidence of central conduction slowing in visual evoked potentials and somatosensory potentials, and the presence of oligoclonal bands in cerebrospinal fluid help further to support the diagnosis.

Classification

There are three classic forms of MS, relapsing remitting, secondary progressive, and primary progressive. Recently a progressive relapsing variety has also been described. A relapsing remitting (RR) course is the most common initial presentation of the disease (85%). Often
taking several years to diagnose, people have a worsening of their neurological condition followed by an improvement as inflammation decreases, conduction block resolves, and remyelination potentially occurs. Unfortunately, this recovery is not always complete and permanent neurological sequelae may occur. There is typically no disease progression between relapses. The female to male ratio is 2.5:1. Most people affected are in the 20’s with most diagnoses occurring between 20 and 40 years of age. Although rare, there is also a juvenile presentation of MS, which most often takes a severe RR course.

About 50% of patients with the RR variety develop secondary progressive (SP) MS. Exacerbations can become less clear and the patient no longer improves significantly after an exacerbation. Over time axonal loss becomes more prevalent and a patient experiences a slow decline in function.

In the primary progressive (PP) form there is neurologic decline from the onset. There is some question if this is an independent disease process from the relapsing variety based on the differences in presentation and progression. Symptoms may plateau and there are no clear relapses. Axonal loss is more prominent and the spinal cord is more likely to be affected. Patients tend to be older at age of onset and it affects men and women relatively equally.

In the progressive relapsing (PR) variety there is progressive decline from the onset, but there are clear relapses. This presentation is uncommon and affects less then 5% of patients with MS. (Figure 1)

**Treatment**

For much of the history of multiple sclerosis we have had no effective treatment to slow disease progression. IV methylprednisolone (Solumedrol) given 1000 mg IV for 3-5 days has been shown to decrease the length as well as long term sequelae of an exacerbation. An oral prednisone taper is often used after the IV steroid. Pulsed IV methylprednisolone may also be effective in decreasing exacerbations although there is no benefit from oral prednisone. Low dose methotrexate, 7.5 mg a week, has also been suggested in PP and SP MS to slow upper extremity functional decline.

Fortunately, there are now five FDA treatments that have been clinically proven to decrease exacerbation frequency in RR MS and some cross over to SP MS. PP MS is more recalcitrant to current treatments although clinically medication management is often still attempted.

Interferon B 1a (Avonex and Rebif) and interferon B 1b (Betaseron) are felt to act by interfering with T cell activation, decreasing inflammation, and stabilizing the blood brain barrier. These medications are injected subcutaneously (Rebif and Betaseron) or intramuscularly (Avonex). There does appear to be a dose related response to the interferons; therefore, Betaseron and Rebif are recommended if the patient can tolerate one of them. Routine monitoring of liver functions and blood counts are recommended at least every six months while on these medications. The most common side effects are flu–like symptoms, which can be lessened with acetaminophen or non-steroidal anti-inflammatory medications.

Glatiramer Acetate (Copaxone), a small protein consisting of 4 amino acids, induces Th2 anti-inflammatory cells and inhibits T cells reactive to myelin basic protein. It may also have a separate “neuro-protective” component. It is given subcutaneously and side effects include chest pain although generally they decrease with time.
Mitoxantrone (Novantrone), which acts to decrease T-cell, B-cell, and macrophage activity, was initially used as a chemotherapeutic agent and now has FDA approval for the treatment of RR and SP MS. Compared to placebo, it has been shown to decrease gadolinium enhancing lesions, and decrease disease progression 60%. Its effects persist at least a year after therapy. Mitoxantrone, an IV medication, is dosed once every three months. It is potentially cardiotoxic; therefore, its use is limited to approximately 2 years. Also a screening echocardiogram should be performed prior to its use to assure a normal ejection fraction. Follow up echocardiograms are repeated every six months and then one year after completing treatment. Other side effects include myelosuppression and amenorrhea. With its high side effect profile, Mitoxantrone’s use is limited to disease unresponsive to other forms of treatment.

Rehabilitation in Multiple Sclerosis
With the multi-system manifestations of MS and its broad spectrum of severity, an understanding of the rehabilitative principles of MS assures a good basic knowledge in neuro-rehabilitation. Despite our treatments to slow progression of MS, rehabilitation is still the only treatment for MS related disability. The progressive nature of the disease makes treatment particularly challenging. A physician must adapt treatment strategies to a patient’s changing function and anticipate future status when prescribing medical equipment.

Impaired Mobility and Self Care
MS can cause weakness, sensory deficits, decreased cerebellar function and balance, and spasticity. All of these changes can result in disability. Fortunately, patients can benefit from training and strengthening through physical and occupational therapy to improve strength, endurance, and function. Exercise that avoids excessive fatigue is well tolerated and does not lead to disease progression. In severe disease, care giver training is essential to assure safe transfers and functional needs.

Strategies and equipment such as sock aides and large handled utensils that are used in other diseases are also beneficial in MS when indicated. Canes, walkers, and braces can decrease the risk for falls and extend the time before a wheelchair is needed. A patient’s strength, endurance, dexterity, cognitive function, and co-morbidities must be carefully considered when prescribing a wheelchair. One also must consider the patient’s long term prognosis based on the rate of disease progression, as many insurance companies will only pay for one wheelchair in a five year period. Lifts can make transfers safer for both the patient and care giver.

A good tool initially used in research to follow patient function overtime and monitor response to treatment is the Expanded Disability Scale (EDSS). Developed by Dr. John Kurtzke, the EDSS divides neurological disability in MS into eight functional systems (pyramidal, cerebellar, brain stem, sensory, bowel/bladder, visual, cerebral-total, and cerebral mentation). As one declines further in the EDSS, mobility and self care increase in significance. This scale ranges from 0, no impairment, to 10, death from complications of MS. Important functional levels include a score of 2.0, representing minimal disability in one functional system, 4.0 severely disabled, but still active and walking with out aides, and 6.0 only walking short distance with assistance. At level 7.0, one is essentially dependent on a wheelchair and at 9.0 one is confined to a bed, but can still communicate. (Figure 2)

Spasticity
Spasticity can be a significant problem, as one would expect in a disease of the central nervous. Increased tone normally starts in the lower limbs making gait, already potentially impaired by poor balance and weakness, more difficult. There can be benefits of spasticity including
maintaining muscle tone, improving vascular return, and facilitation of transfers so one must be careful to examine the patient and get a good functional history before treating.

Maintaining good range of motion with a home exercise program either for the patient or the caregiver is critical. Oral treatments include baclofen (Lioresal), tizanadine (Zanaflex), and dantrolene sodium (Dantrium). Neurolytics such as phenol or alcohol and myoneuromuscular blocking agents such as botulinum toxin can be injected for specific muscles with excessive tone. In severe lower limb spasticity, an intra-thecal Baclofen pump can be surgically implanted. Resting hand splints and ankle splints provide a prolonged stretch and are good at night in particular to decrease contracture formation.

**Pain**

Pain is a common complaint in MS. Neuropathic pain, described as burning in quality, often with parasthesias and a dysesthetic component, can be particularly troublesome to treat. Tricyclic antidepressants such as Amytriptyline and Nortryptyline and anticonvulsants like Carbamazepine and Gabapentin are often helpful. Some patients can also control symptoms with biofeedback strategies taught by a psychologist.

Pain may also be musculoskeletal in nature. Shoulder pain is common in patients with paraplegia who rely on their upper limbs for transfers. Spasticity and immobility resulting in contracture also generates pain in some patients. It is important to remember that patients with MS may get the same diseases as the general population and not to assume any new symptom of pain is from the MS. A thorough history, physical exam, and work up still must be performed. For instance, numbness in the right hand could represent an MS exacerbation, but also could be carpal tunnel syndrome or even a cervical radiculopathy.

**Cognitive Function**

Cognitive issues are one of the primary reasons that people with MS are medically retired. Short-term memory loss and difficulties with planning and multi-tasking are the most common symptoms. Verbal skills often remain fairly good even in advanced disease and often mask the true extent of disability. Neuro-psychologic testing is useful in quantifying deficits and a speech therapist can help a patient develop compensatory strategies to minimize the disability.

**Bowel and Bladder Care**

Urinary changes including detrusor hyperreflexia, detrusor sphincter dyssynergia, and detrusor hypocontractility are common in MS. Symptoms range from urinary frequency and urgency to full incontinence. Urinary tract infections and even serious damage to the kidneys and bladder can occur without good treatment. Once urinary symptoms do occur, it is important to routinely check post void residuals and consider catheterization if values are recurrently over 100 cc. Urinary frequency and urgency can be treated with anti-cholinergics although one must be careful to prevent urinary retention. Bladder patterns can change with disease progression and urodynamic studies are a good way of monitoring those changes and help with management. Routine surveillance with renal ultrasound and evaluation of creatine clearance should be assessed in cases of severe neurogenic bladder.

Bowel symptoms can occur, although not as frequently as urinary complaints. Generally the problem is constipation which can be treated with a number of stool softeners, suppositories, and laxatives. Metoclopramide (Reglan) can be used also to speed GI transit.
Treating Fatigue

Fatigue is the most commonly self identified symptom in MS. This fatigue is often very profound and even tasks as simple as walking to the restroom can be perceived as daunting. An occupational therapist can help with energy conservation techniques. Medical treatment for fatigue includes amantadine (Symetryl), modafinil (Provigil), and stimulants such as pemoline (Cylert) and methylphenidate (Ritalin).

Fatigue as well as other neurologic symptoms are worsened by heat. Poorly myelinated nerves do not conduct well in higher temperatures. Either the ambient temperature itself, or activity and illness that increase the core body temperature can increase symptoms. In fact, often a urinary tract infection or other illness can cause a pseudo-exacerbation that mimics true disease progression. Common sense such as avoiding high temperatures and doing physical activities during the cooler part of the day, as well as devices such as cooling vest are often beneficial.

Depression in MS

In a King County survey of 739 patients with multiple sclerosis 41.8% were suffering from depression. Depression tends to peak after diagnosis and in advanced disease. Depression can contribute to fatigue and memory loss and is often under treated. Treatments include counseling, support groups, and medications.¹

Conclusion

Besides the disease complications, one must consider how the patient is affected psychosocially by the MS. Focusing treatment on the patient’s functional and vocational goals maximizes patient care and satisfaction. MS can be one of the most challenging as well as most rewarding diseases to treat from a rehabilitation perspective due to the extent of disability as well as the diversity of symptoms.
<table>
<thead>
<tr>
<th>Score</th>
<th>Status</th>
<th>Score</th>
<th>Status</th>
<th>Score</th>
<th>Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Normal neurologic exam</td>
<td>4.0</td>
<td>Fully ambulatory without aid, self-sufficient, active 12 hours a day despite relatively severe disability. Able to walk without aid or rest for about 500 Meters</td>
<td>7.5</td>
<td>Unable to take more than a few steps; restricted to wheelchair; may need aid in transfer; wheels self but cannot carry on in standard wheelchair in a full day; may require motorized wheelchair</td>
</tr>
<tr>
<td>1.0</td>
<td>Patients fully ambulatory</td>
<td>4.5</td>
<td>Fully ambulatory without aid, active much of the day, able to work a full day, may otherwise have some limitation of full activity or require minimal assistance; characterized by relatively severe disability. Able to walk without aid or rest for about 300 meters</td>
<td>8.0</td>
<td>Essentially restricted to bed or chair or perambulated in wheelchair, but may be out of bed much of the day; retains many self-care functions; generally has effective use of arms</td>
</tr>
<tr>
<td>1.5</td>
<td>No disability</td>
<td>5.0</td>
<td>Ambulatory without aid or rest for about 200 meters; disability severe enough to impair full daily activities (e.g. working full day without special provisions)</td>
<td>8.5</td>
<td>Essentially restricted to bed much of the day; has some effective use of arm(s); retains some self-care functions</td>
</tr>
<tr>
<td>2.0</td>
<td>Minimal disability in one of seven Functional Systems (FS*)</td>
<td>5.5</td>
<td>Ambulatory without aid or rest for about 100 meters; disability severe enough to limit full daily activities</td>
<td>9.0</td>
<td>Helpless bed patient; can communicate and eat</td>
</tr>
<tr>
<td>2.5</td>
<td>Minimal disability in two FS*</td>
<td>6.0</td>
<td>Intermittent or unilateral constant assistance (cane, crutch, or brace) required to walk about 100 meters with or without resting</td>
<td>9.5</td>
<td>Totally helpless bed patient; unable to communicate effectively or eat/swallow</td>
</tr>
<tr>
<td>3.0</td>
<td>Moderate disability in one FS* or mild disability in three or four FS*, through fully ambulatory</td>
<td>6.5</td>
<td>Constant bilateral assistance (canes, crutches, or braces) required to walk about 20 meters without resting</td>
<td>10</td>
<td>Death due to MS</td>
</tr>
<tr>
<td>3.5</td>
<td>Moderate disability in one FS* and mild disability in one or two FS* or moderate disability in two FS* or moderate disability in five FS*</td>
<td>7.0</td>
<td>Unable to walk beyond about 5 meters even with aid, essentially restricted to wheelchair; wheels self in standard wheelchair and transfers alone; active in wheelchair about 12 hours a day</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
REFERENCES


Epidemiology and Classification of Major Adult Neuromuscular Disease

Amyotrophic Lateral Sclerosis (ALS) is a motor neuron disorder (MND). ALS rapidly destroys upper and lower motor neurons, causing spasticity and diffuse muscular atrophy and weakness. Most ALS cases are acquired and occur sporadically. Approximately 10% of all ALS cases are inherited in a family line. Of all familial cases of ALS, about 10% result from a specific gene defect on chromosome 21q12.1, leading to an abnormality in the production of the anti-oxidant enzyme Cu/Zn superoxide dismutase (SOD1). Current popular theories regarding the etiology of sporadic ALS include excessive glutamate activity in the brain and spinal cord due to defective cellular transport mechanisms and/or possible deficiencies in various neurotrophic growth factors.

ALS most commonly strikes people between 40 and 60 years of age with a mean age of onset of 58 yrs. The overall prevalence rate in the worldwide population is somewhere between 5 to 7 per 100,000, making it one of the most common neuromuscular diseases. Overall, the median 50% survival rate is 2.5 yrs, with death usually arising from pulmonary complications.

Charcot-Marie-Tooth disease (CMT), is a hereditary motor and sensory neuropathy (HMSN). CMT is among the most common hereditary neuromuscular diseases (NMDs), with prevalence ranging from 14 to 282 per million. There are many different genotypic and phenotypic forms of CMT. CMT type 1 (of which there are several subtypes) is primarily demyelinating and characterized by markedly reduced conduction velocities in peripheral motor and sensory nerves. CMT type 2 (of which there are also subtypes), exhibits predominant axonal loss, while conduction velocities remain relatively normal.

Spinal Muscular Atrophy

There are at least three forms of spinal muscular atrophy (SMA), all of which involve selective destruction of anterior horn cells. This is a lower motor neuron disorder. These disorders are inherited, autosomal recessive, traits. SMA I and II (also known as Werdnig-Hoffman disease) will be covered in the pediatric section. SMA III, also known as Kugelberg-Welander disease (KWD), is a chronic, later onset disorder, associated with significantly less morbidity. Signs and symptoms of SMA III, including weakness and atrophy, become apparent between ages 5 to 15 years. All types of SMA are caused by defect in the survival motor neuron gene located on chromosome 5. Prevalence rates for types II and III range from as high as 40 per million among children to around 12 per million in the general population.

Myotonic Muscular Dystrophy (MMD), an autosomal dominant multisystem hereditary muscular dystrophy with an incidence of 1 in 8,000, is characterized clinically by progressive, predominantly distal, muscle weakness and myotonia. Associated findings include frontal baldness, gonadal atrophy, cataracts, and cardiac dysrhythmias. In MMD, a DNA sequence within the gene, on chromosome 19q 13.3, is repeated many times leading to an enlarged,
unstable area of the chromosome. Called a triplet repeat mutation, the flawed gene grows by sudden leaps when transmitted from generation to generation causing the disease to occur at a younger age and in a more severe form (“genetic anticipation”). The repeated DNA sequences known as CTG (trinucleotide cytosine, thymine guanidine) are linked to the production of a protein (myotonin-protein kinase), which has important functions in smooth and skeletal muscle, eye, hair and brain.

Facioscapulohumeral Dystrophy (FSH) is a slowly progressive myopathy with autosomal dominant inheritance and prominent involvement of shoulder girdle and facial musculature. Prevalence is difficult to ascertain due to undiagnosed mild cases, but has been estimated at 10-20 per million. The abnormal gene is at the end of chromosome 4, although the protein product for this gene is not yet known. FSH can be quite heterogeneous in its clinical presentation and course.

Becker Muscular Dystrophy

Both Duchenne (DMD) and Becker (BMD) muscular dystrophy are inherited X-linked recessive diseases affecting primarily skeletal muscle and myocardium. Dystrophin, a large cytoskeletal protein in the subsarcolemmal lattice, which stabilizes the plasma membrane during muscle contractions, is absent in DMD and of abnormal molecular weight and/or reduced amounts in BMD. DMD is the most common neuromuscular disease of childhood with prevalence rates ranging from 19 to 95 per million with an estimated overall prevalence of 63 per million. This progressive myopathy will be covered in the pediatric section. BMD has a lower incidence than DMD with prevalence rates for BMD ranging from 12 to 27 per million with a recent estimated overall prevalence of 24 per million. BMD shows a similar pattern of muscle weakness to DMD but with later onset and much slower rate of progression. These men often live normal life spans, barring any cardiac complications.

Limb-Girdle Muscular Dystrophy (LGD) is a very heterogeneous group of myopathies that share some clinical features. There are many different subtypes and the expression may be in either sex, with primary involvement of the shoulder and/or pelvic girdle muscles with a variable rate of progression. LGD is no longer considered to be a distinct nosological entity and is felt to be a wide variety of muscle disorders predominantly caused by defects in the genes producing any number of transmembrane associated glycoproteins. These patients may clinically resemble a severe form of BMD, despite the normal expression of dystrophin.

Medical Issues

Restrictive Lung Disease: (RLD) is common in most NMDs due to weakness of the diaphragm, chest wall, and abdominal musculature. In the adult population RLD can become quite severe in SMA, MMD and ALS (see pediatric section for discussion of Duchenne muscular dystrophy). Severe scoliosis, if present, may also complicate RLD. Patients should be educated early in the disease process so informed decisions can be made later as the disease has progressed. Routine pulmonary function tests (PFTs), including forced vital capacity (FVC), maximal inspiratory and expiratory pressures (MIP/MEP), and peak cough flow (PCF) should be monitored closely. The MIP reflects diaphragmatic strength and the ability to ventilate. The MEP reflects chest wall and abdominal muscle strength and the ability to cough and clear secretions.

Ultimately, most patients with severe RLD develop hypoventilation, which leads to elevated CO₂ levels. Measuring only O₂ saturation levels with pulse oximetry may be inadequate. End tidal CO₂ levels or arterial blood gases (ABGs) should be measured periodically, depending on the clinical condition of the patient. A thorough review of systems will help define any problems. Patients that are hypoventilating will often become hypercapnic at night and complain
of a morning headache, restlessness, nightmares, and poor quality sleep. This may cause daytime somnolence. Insufficient respiration with hypoxia may occur later, especially if the lung is damaged by chronic aspiration.

Intermittent positive pressure ventilation by mouth (IPPV) avoids the need for tracheostomy and maintains reasonable quality of life. Bimodal positive airway pressure (BiPAP) is the best initial form. This can be done in the home and should be considered the preferred modality of assisted ventilation in NMDs. It generally takes some work with a respiratory therapist to get a good face or lip seal on the mask or nasal/oral orthotic interface. Patients may benefit initially from using assisted ventilation mainly at night. Other devices that are useful include an in-exsufflator, which is a device that helps produce an artificial cough. Coughing can also be manually facilitated with various physical maneuvers, including a controlled abdominal thrust against a partially closed glottis (“quad cough”). Suction machines are also helpful for pulmonary toilet. If better airway access becomes absolutely necessary and the informed patient wishes more aggressive care, a tracheostomy may be done.

In ALS and some rare forms of SMA, bulbar palsy and dysphagia may occur. Early signs of dysphagia include changes in voice patterns (“voice becoming hoarse”) and persistent coughing after swallowing liquids, which can indicate micro-aspiration. A speech therapist should be consulted early on for clinical swallowing evaluations and recommendations on dietary modification such as thickening liquids and preparing food that forms into boluses easily. A modified barium swallow (MBS) is helpful at accurately determining the presence of aspiration as well as defining which food textures the patient can safely swallow. A percutaneous endoscopic gastrostomy (PEG) tube may be needed for nutrition.

**Cardiac Complications**

Significant cardiac involvement occurs in BMD, MMD, and some cases of LGD. There is a very high (60-80%) occurrence of cardiac involvement in BMD subjects across all ages. Dystrophin has been localized to the membrane surface of cardiac Purkinje fibers and defective dystrophin expression likely contributes to the cardiac conduction disturbances seen in BMD.

The pattern of occasional life-threatening cardiac involvement in BMD may, on occasion, be more rapid than the progression of skeletal myopathy. The cardiac compromise may be disproportionately severe relative to the degree of restrictive lung disease in some BMD subjects. The evidence for significant myocardial involvement in BMD is sufficient to warrant screening of all these patients at regular intervals using ECG and echocardiography. Close follow-up will be needed for those with myocardial involvement. The slowly progressive nature of this dystrophic myopathy, which is compatible with many years of functional mobility and longevity, makes these patients suitable candidates for cardiac transplantation if end stage cardiac failure occurs. Successful cardiac transplantation has been increasingly reported in BMD subjects with cardiac failure who are otherwise still quite physically functional.

**Electrocardiographic abnormalities** have been extensively reported in myotonic muscular dystrophy. Approximately 33% of MMD subjects had first-degree AV block, 17% had left axis deviation, and 5% had left bundle branch block and rarely, complete heart block requiring pacemaker placement. Although few subjects show clinical signs and symptoms of cardiomyopathy, sudden deaths do occur in relatively young MMD subjects and bundle of His conduction delays have been reported. Any MMD subject with dyspnea, chest pain, or cardiac symptoms should receive thorough cardiac evaluation including bundle of His studies.

Cardiac involvement has only been recently recognized in LGD patients, and likely only occurs in a specific subset. As in BMD, the cardiac manifestations in LGD may predate the clinical evidence of skeletal myopathy.
**Nutritional Management**

In the more severe NMDs there is an increasing tendency towards obesity around or after the time of losing independent ambulation. Obesity is common in NMDs, particularly DMD where a 54% prevalence has been reported. However, in prior studies obesity was unrelated to strength decline, age of wheelchair reliance, functional grade status, timed motor performance, pulmonary function, likelihood of ECG abnormalities or age at death. Weight control has its primary rationale in ease of care, in particular ease of transfers and skin care.

In advanced stages of ALS and SMA malnutrition may be a significant problem. Nutritional compromise becomes particularly problematic if there is severe RLD with increased work of breathing, which may also influence caloric intake. Self-feeding often becomes impossible during this period. Nutritional counseling with energy intake and energy allowance estimations should be done for all NMD patients with severe (FVC < 50%predicted) RLD. Placement of a PEG tube may facilitate nutrition even if the patient can swallow because it allows for rapid and easy delivery of large amounts of calories and fluids. Patients should be reassured that they will still be able to eat food orally for enjoyment.

**Functional Issues and Approaches to Rehabilitation**

The goals of rehabilitation in patients with NMDs are to maximize functional capacities, prolong or maintain independent function and locomotion, inhibit or prevent physical deformity, and provide access to full integration into society with good quality of life. A team consisting of doctors, physical, occupational, and speech therapists, social workers, vocational counselors, and psychologists, among others, best carries out management. Treatment is goal oriented using various modalities.

**Weakness and the Role of Exercise**

Skeletal muscle weakness is the ultimate cause of the majority of clinical problems in NMDs. There have been several well-controlled studies looking at the effect of exercise as a means to gain strength in NMDs. In slowly progressive NMDs a 12-week moderate resistance (30% of maximum isometric force) exercise program resulted in strength gains ranging from 4% to 20% without any notable deleterious effects. However, in the same population, a 12 week high resistance (training at the maximum weight a subject could lift 12 times) exercise program showed no further added beneficial effect compared to the moderate resistance program and there was evidence of overwork weakness in some of the subjects. In a comparative study, CMT patients appeared to benefit significantly from a strengthening program whereas MMD patients showed neither beneficial nor detrimental effects. It is likely that the most effective exercise regimens for neuropathies and myopathies will be different although further studies are needed to confirm this. Due to the active, on-going muscle degeneration in the rapidly progressive disorders DMD and ALS, the risk for overwork weakness is great and exercise should be prescribed cautiously with a common sense approach. All patients with NMDs should be advised not to exercise to exhaustion due to the risk of muscle damage and dysfunction. Patients participating in an exercise program should be cautioned of the warning signs of overwork weakness, which include feeling weaker rather than stronger within 30 minutes post exercise or excessive muscle soreness 24-48 hours following exercise. Other warning signs include severe muscle cramping, heaviness in the extremities, and prolonged shortness of breath.

Exercise may induce muscle cramping in NMD, presumably due to sarcolemmal instability. Exercise may also cause symptomatic muscle spasms related to spasticity in ALS. Baclofen acts to facilitate motor neuron inhibition at the spinal levels and is a good initial agent to use. The benzodiazepines and other centrally acting muscle relaxants may induce somnolence
and respiratory suppression. Dantrolene, by impairing excitation-contraction coupling, is effective at reducing muscle tone but will also cause generalized muscle weakness and should not be used. Membrane stabilizing agents like procainamide and phenytoin may also be helpful, particularly in the myotonic disorders. Slow (30 second sustained), static muscle stretching is also helpful.

**Limb Contractures and Scoliosis.**

The occurrence of contractures appears to be directly related to prolonged static positioning of the limb, and these contractures often develop soon after wheelchair reliance. Several studies have shown that wheelchair reliance and lack of lower extremity weight bearing contribute to the rapid acceleration of contractures. Upper extremity contractures may occur in ambulatory patients with focal, proximal atrophy, particularly at the shoulder girdle, as is commonly seen in FSH. This may be worsened by subluxation. Slings may be helpful to support the joint but will not prevent contracture formation or subluxation. Gentle static stretching and splinting may slow the progression of contractures. Contractures may create further disability by impeding proper wheelchair seating, making transfers more difficult or impeding dressing and toileting.

Scoliosis is much more common in childhood NMDs however some adults may also develop this problem. Spinal bracing has not been shown to be effective in preventing progression of scoliosis in NMD. NMD patients with scoliosis should be followed closely with serial radiographs and have appropriate and timely spinal instrumentation and fusion which should be done before the primary curve becomes greater than 25 degrees and vital capacity (VC) has not gone below 50% of predicted.

**Bracing to Prolong or Maintain Ambulation**

The purpose of bracing at any level is to improve function and stability. Most CMT patients will require short leg braces or Ankle-Foot-Orthoses (AFOs). These should be custom made of a lightweight polymer such as polypropylene or carbon fiber resin and fit intimately to provide good stability and prevent pressure sores. The older, traditional double metal upright AFOs built in to the shoe may be too heavy and actually hinder ambulation. If there is severe distal muscle weakness and ankle instability the braces should be high profile, coming around the ankle in front of the malleoli. A pes cavus foot with a hammertoe deformity can be accommodated with a built up arch and metatarsal bar. Patients with CMT are at high risk for neuropathic arthritis (charcot joint) and skin ulcers. Their skin and weight-bearing joints should be closely examined at every clinic visit.

Patients with other NMDs may also benefit from bracing, depending on the distribution of weakness, gait problems and joint instability created by the weakness. In making the decision to brace, the clinician should also consider the added weight of the brace and whether or not the patient is willing to use a brace. If there are questions about whether a brace would help or not then the patient should be tried in an off-the shelf model first along with careful assessment by the clinician. Most patients will require a short course of physical therapy after the braces are made to help effectiveness.

**Equipment**

There are a number of useful equipment items that can substantially improve quality of life, including hand-held showers, bath tub benches, grab bars, raised toilet seat, hospital bed, commode chair, ADL aids (sock aid, grabbers, etc), and wheel chair ramps. Wheelchairs should be properly fitted (generally done by occupational therapist) and have adequate lumbar support and good cushioning to avoid pressure ulcers. Simply giving the patient a prescription for a
wheel chair often ends up with the patient getting a standard manual chair that does not fit properly. An electric wheel chair, although expensive, can be justified on that basis that it will help prolong independent mobility and thus markedly improve quality of life.

A good pressure-relieving mattress (air or dense foam) should be used on the bed at home, along with foam wedges to facilitate proper positioning. This will help prevent pressure ulcers and contractures. Wheeled walkers or quad (four point) canes may also help, depending on the pattern of weakness. Frequently severe weakness in the neck flexors and extensors will cause a "floppy head" associated with severe neck pain and tightness. This may be helped by a hard cervical (Philadelphia type) or a Freeman collar. Communicative aids including an alphabet, word board or speech synthesizer will help out in ALS with dysarthria. An occupational therapist and speech language pathologist will help define which, if any, of these devices will be useful to the patient.

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The rheumatologic disorders comprise a diverse group of diseases that cause dysfunction of the neuromuscular system. Arthritis is an important feature of most of these disorders and causes most of the impairment. This chapter will focus on the physical medicine management of arthritis in two disorders, osteoarthritis of a weight bearing joint, and rheumatoid arthritis involving the hand.

The Hand In Rheumatoid Arthritis
The goal of physical management is to develop long-term habits of hand use and self-management techniques designed to minimize deformity and maximize function. Control of the inflammation process is crucial. It should be noted that the emphasis in medication management has been to use disease modifying anti-rheumatic drugs (DMARD’s) earlier than in the past. (figure 1.) An excellent resource regarding medication management can be found in the Primer for Rheumatologic Conditions published by the Arthritis Foundation.

**Figure 1 Guidelines for the Management of Rheumatoid Arthritis**

**Evaluation**
The history should identify the degree and extent of disease activity and adequacy of control of joint inflammation. Functional requirements for the upper extremity need to be explored in both vocational and avocational activities.

Physical examination should identify range of motion (ROM) of the elbow, wrist, and hand. It is important to include an evaluation of the carpal joints, carpal-metacarpal (C-MP) joint of the
thumb, metacarpal phalangeal (MCP) joints and interphalangeal (IP) joints. Both ROM and joint stability are assessed. Intrinsic muscle shortening should be assessed. Each joint is palpated for synovial thickening, effusion, and tenderness. Strength of wrist flexors, extensors and hand intrinsics are examined with particular attention to the intrinsic radial deviators of the MCP joints. It is necessary to learn how to examine the hand from a skilled examiner and practice this.

**Education**

The patient needs to be engaged as a partner in their health care as with all of medicine. This is particularly true for physical management as it is the patient who will be responsible for implementing the management plan. The physician’s role is to plan an appropriate program and give the patient the necessary tools to carry it out; the patient must become an expert regarding his/her condition.

**Hand Protection**

In the presence of continued chronic inflammation, repeated strong grasping forces are associated with volar subluxation of the wrist and volar subluxation/ulnar deviation of the MCP’s (figure 2). Strong extrinsic finger flexor use by the flexor sublimus and profundus muscles provides the deforming force. Sudden forceful use of the hand is associated with tendon rupture or acute overstretch of the radial MCP joint capsule. For these reasons, patients with continued chronic synovitis should be asked to be gentle in the use of their hands and avoid regular or occasional forceful phasic use of strong grasp and twisting. Special care needs to be taken during periods of inflammatory flairs when the joint structures are more vulnerable. These principles are taught by demonstration in Occupational Therapy.

![Figure 2 Volar Subluxation/Ulnar Deviation of the Metacarpal-Phalangeal Joints](image)

**Strengthening**

Chronic inflammation is associated with loss of muscle mass beyond that attributable to aging sarcopenia (figure 3). Studies of exercise in rheumatoid arthritis (RA) support the role of exercise as the only known stimulus to reverse this process. In patients without acute synovitis, no increase in disease activity has been noted in these studies. These findings have supported the development of several community based self-help programs that utilize exercise. Unfortunately, there are as yet no studies of specific exercises to improve joint function in RA. For the hand, strengthening of the radial intrinsics should provide improved dynamic support of the MCP preventing stretching of the radial capsular structures during activity. Given the role of the extrinsic finger flexors in the development of volar subluxation/ulnar deviation, finger-
squeezing exercise should be avoided. Wrist extensor strengthening should help maintain wrist extension and improve joint stability.

Figure 3 Aging Sarcopenia

Stretching

Daily manual stretching is advised when loss of motion is observed on physical examination. Common motion loss problems in the hand include loss of PIP flexion associated with intrinsic muscle shortening and loss of wrist extension. Heating with a paraffin bath makes connective tissue more distensible and can facilitate stretching but requires time and greater commitment. Stretching orthoses can be fabricated for the stretching of greater degrees of contracture. (figure 4) Unfortunately, controlled trials of various stretching programs and devices have not been done.

Figure 4 Stretching Wrist-Hand Orthosis for the Proximal Interphalangeal Joints and Hand Interosseous Muscles
**Splinting**
There is a limited role for the use of splints. Resting splints can put the hand with uncontrolled active synovitis temporarily at rest, which diminishes inflammation. (figure 5.) Stretching orthoses can be used as noted above for the stretching of contractures. Functional splints can stabilize joints with instability such as severe ulnar deviation or hyperextension of the PIP (swan neck deformity) (figure 6, 6a, 6b, 7). Most of these orthoses are fabricated by occupational therapists or are commercially available.

![Figure 5 Resting Wrist-Hand Orthosis](image1.png)

![Figure 6(a) Functional Proximal Interphalangeal Extension Block Orthosis (Ring Splint)](image2.png)

![Figure 6(b) Swan Neck Deformity with Proximal Interphalangeal Hyperextension](image3.png)

![Figure 7 Functional Short Hand Orthosis to Prevent Ulnar Deviation](image4.png)

**Adaptive Aids**
A variety of devices may improve function in those with severe deformity. These range from modified utensils to allow easier grasp, reaches to allow grasp when ROM is limited, to power mobility devices.

**Management of Associated Carpal Tunnel Syndrome (CTS) or Tenosynovitis**
RA predisposes individuals to develop CTS. As with CTS unassociated with RA, management ranges from resting night splints, avoidance of prolonged flexed wrist postures, steroid injection into the carpal tunnel, and surgical release. Trigger fingers may develop early in the course of the disorder and can usually be managed with a local steroid injection followed by a finger splint for a few days. Tenosynovitis of the thumb extensor tendons also occurs and is managed with steroid injections and a few days rest in a thumb spica. Individual joints may flair and can be managed by local intra-articular steroids.

**DJD of Weight Bearing Joints**

**Evaluation**
The history should elucidate the course of pain, function, and previous treatment attempts. The review of systems should identify possible congenital, metabolic, endocrine or neuropathic problems. The examination should focus on the presence of inflammation, ROM, joint stability, alignment, muscle strength and associated mechanical factors such as leg length discrepancy and obesity. Weight bearing radiographs are helpful in confirming the extent of joint destruction and the presence of medial or lateral compartment collapse.
Education

DJD (Osteoarthritis) is by far the most common form of disabling arthritis affecting approximately 60% of the population over 35 years of age and is responsible for most total hip and knee replacements. The diagnosis of DJD in a weight-bearing joint does not necessarily imply relentless progression and loss of function leading to joint replacement. A recent study showed that over a period of 10 years in a population of subjects with DJD of the knee, 10% improved, 60% did not progress, and 30% worsened. The goal is to teach patients to optimally live with their arthritis, preserving cartilage and joint function. Simply prescribing analgesics is inadequate management. Therefore, engage the patient as a partner to do what is necessary to achieve this goal.

Pacing

Using pain as a guide, maintain weight-bearing time below the threshold for developing prolonged aching pain. Otherwise, continual pain is likely requiring a dramatic decrease in weight bearing activity. Analgesics can aggravate this problem if the patient forces weight-bearing activity to the point of pain when taking the analgesic. For this reason, it may be beneficial to use analgesics in a pulsed fashion. Continual use of NSAIDs does not modify the course of the disorder. Activity should be balanced during the day by rest from weight bearing. Vocational and avocational activities should be modified to make this possible.

Associated soft tissue pain syndromes may be responsible for much of the pain at the time the patient presents. Trochanteric or pes-anserine bursitis if present should be treated with NSAIDS, ultrasound or a steroid injection.

Unload the Joint

Optimization of body weight can increase function and slow progression. The use of a cane can significantly reduce compression forces at the hip and can provide some relief of weight bearing at the knee.

Minimize Impact Loading

Impact loading is poorly tolerated in experimental models of osteoarthritis. Sport activities involving impact should be adapted. Viscoelastic shoe inserts or soft soles can diminish peak impact forces.

Optimize Joint Motion and Stability

Stretch out or prevent flexion contractures. They cause increased joint loading and are associated with more rapid progression. This may require daily stretching and strengthening exercises to accomplish. Techniques include brief manual stretches, prolonged stretching with counterbalanced stretches, use of weights, dynamic splints, or serial casting depending on the severity of the contracture Rectus femoris and lateral retinacular shortening may be a factor in patellofemoral arthritis. Research is needed on the most efficacious methods of restoring full motion. Consider bracing for marked ligamentous laxity or varies/valgus deformity. Lateral shoe wedges may be helpful for valgus deformity of less than 5 degrees.
Strengthen Musculature

Randomized controlled trials show beneficial effects of strengthening on pain, self reported disability, walking performance and patients global assessment. Increased dynamic joint stability after strengthening may by an important factor in slowing progression.[9] Inadequate strength in the hips and quadriceps is associated with increased falls in the elderly leading to considerable secondary disability. The optimal techniques for improving strength have not been adequately studied. A reasonable approach is to start with isometrics at several joint angles, and subsequently add isotonic strengthening with little or no resistance in a pain free range of motion, gradually increasing the resistance if it can be done pain free.

Total Joint Arthroplasty

Total joint arthroplasty should be considered if the patient’s function and pain are unacceptable after adequate conservative management. Patients should ideally be educated regarding their post-operative rehabilitation program before their procedure. Following the procedure, initial weight-bearing status must be identified which varies with the kind of prosthesis. For the hip, instructions to limit postures (usually flexion, adduction and internal rotation) that could dislocate a total hip arthroplasty are essential. Initially, an abduction splint may be used. Adapted ADL activities may have to be taught to limit dislocating postures. Ambulatory aids may be needed at first to assure safe ambulation. Strengthening of the hip abductors and extensors are important to maximize safe transfers and ambulation. Stretching may be required to restore full hip ROM. Leg length discrepancies should be corrected with a shoe lift for length discrepancies not due to flexion contractures. Complications of heterotopic ossification, deep venous thrombosis, infection, or failure of the arthroplasty have to be identified and managed.

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More than 85% of all sports injuries are non-surgical, and all injuries (including surgical cases) require rehabilitation. The predominant factor of sports medicine resides in conservative non-surgical management and proper rehabilitation. This chapter will discuss the unique approach that a rehabilitation medicine physician can provide to a patient with a musculoskeletal injury. The broader term musculoskeletal medicine encompasses injuries that occur during sporting activities as well as those occurring in the workplace and everyday life. With a thorough knowledge of how the muscles and skeletal systems function, the underlying cause of the injury can often be determined and a comprehensive rehabilitation plan initiated. The ultimate goal is to restore function and prevent repetitive injury.

One of the main tenants of rehabilitation medicine is caring for people with disabilities. Those disabilities can arise due to a stroke, polio, or a sports related injury. Regardless of their cause, these disabilities can be addressed to provide an improved quality of life and restore function. Even if the cause is not curable, the disability it creates can be treated and often removed. Fortunately, many of the disabilities caused by musculoskeletal injuries are temporary, but remember that a disability is “any restriction or lack resulting from an impairment of the ability to perform an activity in the manner or within the range considered normal for a human being.” For a high level athlete, even a mild ankle sprain can cause significant disability.

The emphasis of medical school education is to recognize and treat disease. For a rehabilitation physician, recognizing the disease is just the first step. The impact the disease has on the person’s overall function becomes apparent through functionally directed questions. Once understanding how the injury or “impairment” affects the individual’s function, the disability becomes apparent. At this point, a plan can be formulated to treat not only the tissue injury, but also any disabilities that may be present.

The history is critical in the evaluation of a patient with a musculoskeletal complaint. This is perhaps the most important aspect, as it gives critical information regarding prior injury, mechanism of injury, and the temporal progression of pain and subsequent disability. A critical step is to classify the injury as acute, chronic, or acute exacerbation of a chronic condition. This initial determination will influence the treatment plan and provide some information as to the expected time for recovery.

A patient presenting with a chief complaint of “a sore ankle” gives insight into the location and possible differential of disease, but tells nothing of the disability. Further questions such as, “are you able to compete?” “are you able to drive and walk?” and “has this affected your work?” all provide vital insight into what disability is present. There is no way to predict the disability as the same disease process will manifest with different disabilities in every person; disability is person specific.

An acute injury is due to an inciting event in the past 3 months, and often the patient can recall a detailed history. These injuries most often occur due to a rapid disruption of connective tissue with a large inflammatory response. In contrast, chronic injuries may have an indolent onset with
variable symptoms that may escalate, or wax and wane over time. These injuries often occur due to repetitive micro-trauma to the tissue. An acute exacerbation of a chronic condition usually has a period that is relatively “pain free” or “healed” which reappears due to an acute reactivation. With any of these presentations, there can be eventual healing without pain, but does not mean the tissue has been restored to the normal strength or function. Following injury, there can often be contraction of the soft tissues, relative muscle imbalance and loss of range of motion. It is this loss of motion and tissue compliance that leads to eventual re-injury unless a progressive rehabilitation strategy is implemented. A key concept is that these imbalances may not manifest themselves under normal loading, but only during high-level dynamic activity or following muscle fatigue.

As with the rest of medicine, the more specific a diagnosis that can be determined, the more specific treatment can be provided. When evaluating bacterial infections, we determine the exact organism to determine the appropriate antibiotic. The same practice is true for musculoskeletal medicine. When evaluating a patient with shoulder pain, it is not sufficient to diagnose “bursitis” or “rotator cuff tear.” It is imperative to know which muscle(s) of the rotator cuff are involved. After determining if the injury is acute or chronic, the second step is determining the structures involved as specifically as possible.

The determination of the structures involved requires a thorough knowledge of anatomy and clinical provocative tests. It also requires understanding how each joint interacts with those above and below it. A joint does not function in isolation, but rather as just one in a series of joints that act together to move the limb or body though space. This concept of a kinetic chain helps us understand how one joint can affect the forces on another, and how the dysfunction of one joint can cause dysfunction of another. It is not always the first or most dysfunctional joint that presents as being painful.

The body is a dynamic structure with orchestrated movements that occur in a specific sequence to provide the most efficient movement. Each movement should occur without exceeding the inherent limitations of the tissues involved. Musculoskeletal injuries can affect the kinetic chain by restricting full motion (such as with tissue contracture), muscle imbalance between antagonistic muscle pairs across a joint, and change in joint stability or center of rotation. Any of these limitations may also change the sequence of activation along the kinetic chain.

Upon determining the structures involved (through a series of both passive and active range of motion and resistive testing), the first treatment step is to decrease pain and inflammation. This is done through a number of modalities and medications, and may include a period of immobilization.

Modalities are simple and without systemic side effects. Following an acute event or a chronic exacerbation ice is recommended for the first 72 hours. The cooling effect reduces the inflammatory cascade that contributes to tissue injury and pain. Following 72 hours of icing, heat therapy can be applied to provide improved blood supply to the injured tissue to assist with the healing process. Ice and heat should never be used if the case of suspected tissue infection or impaired sensation.

Medications are numerous and include pure analgesics (acetaminophen, opioids) and NSAIDs (aspirin, indomethacin, ibuprofen, naproxen, piroxicam). No single agent has been shown to be superior, and duration of therapy should be limited to the acute phase (2-3 weeks) to avoid gastrointestinal side effects. The role of muscle relaxants has remained controversial and they have not been shown to be more effective than NSAIDs. The use of corticosteroids (medrol dose
pack, prednisone) should be limited to treatment of acute injuries when NSAIDs are contraindicated or have been ineffective.

Regardless of the method of analgesia, the ultimate goal is to allow the patient to become active with their rehabilitation plan. Once the tissue inflammation has subsided, the tissue may begin to undergo gentle tension, usually via passive range of motion. This will load the soft tissues of a joint without causing high forces and possible re-injury. As tissues begin to heal, they undergo a process of shortening. As the patient tolerates, a progressive stretching program will help restore functional range of motion by lengthening the connective tissue. In this acute stage, the focus is to prevent further loss of strength with mild strengthening as tissues tolerate.

Once the patient has established near normal ROM and is tolerating the initial strengthening process, further emphasis is placed upon flexibility and specific progressive exercises including strengthening. This phase is often referred to as the recovery phase of rehabilitation and includes a change of focus from clinical symptoms to restoration of function. A fundamental that begins at this stage is the concept of proprioceptive neuromuscular control; a process to reeducate proper movement patterns and eliminate substitution patterns. This will allow better muscular activation at the appropriate timing to provide joint protection at an unconscious level. Since nearly all motion begins with support from the trunk, an evaluation of core (truncal) stability is necessary to allow proper initiation of movement within the kinetic chain.

Once the patient has achieved full pain-free range of motion, good flexibility, ~80% of normal strength and good muscle balance, they are progressed to the functional stage of rehabilitation which will address general fitness as well as progressive sport/work specific exercises. This will continue until strength is returned to normal and sport specific skills are demonstrated at full intensity. Following successful return to activity, the maintenance phase of rehabilitation provides ongoing conditioning and general fitness to prevent future injuries.

**Case example:**
A 38-year-old female, who is otherwise healthy, presents with a 6-week history of progressively worsening right > left knee pain without radiation or locking of the knee. The pain has come on slowly, without history of trauma or significant swelling. It is exacerbated by sitting for prolonged periods of time, as well as with descending stairs. Pain is relieved when she rests, and improved with over the counter NSAID therapy. Past medical history is notable for 2 children and GERD. Medications include ibuprofen 200-400 mg prn. ROS is notable for 10# weight gain in the last year. Functional history reveals she was a college level soccer player, and has returned to a regular exercise program including soccer and weightlifting after a 12-year period of relative inactivity. She is employed as a legal clerk, and feels significant pain throughout the day. Her mobility is only limited to the point she cannot run or perform her exercise regimen.

Physical examination is notable for internal rotation of the femur with “kissing patellas,” shortened ITB, relatively weak hip abductors and knee extensors, and lack of flexibility of the hamstrings and gastroc-soleus complex. The patella is relatively hyper mobile and reproduces her pain with compression and quadriceps activation. Her examination is negative for neurologic abnormalities, ligamentous laxity of the knee, and for infrapatellar fat pad impingement.

At this point, patellofemoral syndrome (PFS) appears to be the correct diagnosis. The pain is originating from the subchondral bone due to a repetitive stress exceeding the limitations of the tissue. To understand where the increased stress is coming from, we must understand how the patella works as part of the kinetic chain.
The patella acts as a fulcrum to provide increased extension forces at the knee joint. As the patella is drawn proximally by the contracting quadriceps, it is also undergoing compression in the knee joint. These forces are normally transmitted over multiple points in the trochlear groove. The patella normally tracts within the center of the trochlear groove due to a relative balance between the medially directing forces (vastus medialis), and the laterally directing forces (iliotibial band, vastus lateralis, lateral retinaculum). When there is an imbalance in these forces, the patella is usually pulled laterally within the trochlear groove, causing increased pressure to be transmitted by a reduced surface area (thus exceeding the limitations of the subchondral bone).

Proximal factors that can influence the tracking of the patella include increased lumbar lordosis, femoral anteversion, and weakness or contracture of the hip abductors, hamstrings, ITB, or hip rotators. Factors at the knee include vastus medialis vs. lateralis imbalance, quadriceps vs. hamstring imbalance, and patella alta or baja. Distal factors include Achilles contracture, foot pronation, cavus deformity, and internal rotation of the tibia.

| Diagnosis: | Acute patellofemoral syndrome (PFS) |
| Disabiliies: | • Exercise limitation  
• Vocational limitations due to pain |
| Structures involved: | Local: patellar subchondral bone due to abnormal biomechanics and patellar tracking |
| Disability treatment: | • Alternative exercise (ie. Swimming)  
• Workplace ergonomics (standing work station) |
| Acute phase: | Continue until full range of motion established and pain nearly resolved.  
• Ice  
• Naproxen 500 mg bid with meals to reduce GI upset  
• relative rest of knee extension  
• Gentle range of motion of knee/hip/ankle  
• Isometric contraction of vastus medialis |
| Recovery phase: | Continue until pain free range of motion and strength roughly 80%  
• Aggressive stretching of hamstrings, achilles, hip rotators, ITB  
• Progressive strengthening of quadriceps (to minimize compressive forces at the patella  
• Utilize closed chain exercises (leg press) from full extension to 45 degrees of flexion, and open chain exercises from 45 to 90 degrees of flexion  
• Progressive strengthening of hip abductors and external rotators  
• Propriocceptive training including balance  
• Trial of McConnell taping to stabilize patella |
| Maintenance phase: | Continued stretching and strengthening  
• Activity specific progression (addition of cutting drills and plyometrics to simulate soccer skills).  
• Return to full sport when able to perform simulations at 100% without pain |

As shown by the case example, musculoskeletal injuries require a comprehensive examination, specific treatment, and significant investment by the physician and patient to promote proper tissue healing and prevention of long term disability. Excellent communication between the physician, patient, and the treating therapist is crucial for success. The process of rehabilitation is an active process that requires participation by the patient. In such, proper motivation, positive feedback, and the setting of obtainable will instill a sense of accomplishment and increase the likelihood of success.
*Classify Acute/Chronic/Re-injury
*Determine disabilities
*Determine exact structure involved
*Initiate treatment plan

**Acute Phase**
1. Address disabilities
2. Decrease pain and inflammation
3. Restore pain free motion

**Recovery Phase**
1. Muscle reactivation and proprioceptive control
2. Kinetic chain restoration with core stability

**Maintenance phase:**
1. Sport/work specific activity progression
2. Continued stretch and strengthening program

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**REFERENCES**

Further Readings:

Lower Extremity Amputation Rehabilitation

Joseph M. Czerniecki, MD

Epidemiology

The number of amputees in the United States has been estimated to be greater than 2 million. In 1990 the National Center for Health Statistics estimated an annual incidence of 105,000 lower extremity amputations, excluding Veterans Administration military and public health hospitals. Of these, approximately 1/3 were above the knee, 1/3 were below the knee, and 1/3 involving the foot or the digits.

The etiology of lower extremity amputation is primarily related to what has been termed medical disease (vascular disease/diabetes), and much less commonly to tumor, trauma, and congenital etiologies. The relative prevalence of each of the underlying etiologies varies on a global level. It is dependant upon the age distribution in the population, dietary factors, presence of military conflict and its residua, such as land mines, as well as, the extent of industrialization. In most industrialized nations vascular disease and diabetes account for approximately 90% of lower extremity amputations, while tumor and trauma account for the balance.

Vascular disease and diabetes have special implications because both processes are generalized multisystem disorders. Atherosclerosis involves not only the peripheral vascular system, but also the cardiovascular and cerebral vascular systems. Therefore, this population may exhibit a high incidence of myocardial infarction, congestive heart failure, angina, TIA, or stroke. Additionally, diabetes commonly results in a sensori-motor polyneuropathy, and can also result in vision loss secondary to retinopathy. This has major ramifications when considering the rehabilitation process and outcome.

There is a high mortality rate among individuals with amputations secondary to vascular disease and diabetes. The survival rate 3 years after a patient's first amputation is approximately 50 percent, while at 5 years it is only 30%. In addition to an increased mortality, there is a high incidence of secondary amputation of the contralateral limb. In individuals with diabetes the probability of contralateral amputation is approximately 10% per year.

Pathophysiology

The increased risk of amputation in those with diabetes is related to peripheral vascular disease, neuropathic changes, and increased risk of infection. The most common causal cascade that leads to amputation in diabetes is the development of ulceration of the soft tissues of the foot. The sensory component of the neuropathy results in a “stocking” distribution sensory deficit. Due to this sensory deficit, the patient is often unaware of recurring or acute tissue injury, and is therefore unable to prevent or respond to soft tissue injury of the foot. The motor component of the neuropathy causes weakness and atrophy of the intrinsic muscles in the foot, leading to an unbalanced pull by the long extensors and flexors of the toes, which results in the typical claw toe deformity. This deformity leads to abnormal pressure distribution of the soft tissues of the foot during standing and ambulation, especially when the patient is wearing less than optimal...
footwear. Together the deformity in conjunction with the sensory deficit leads to mechanical injury of the soft tissues of the foot, ulceration and an increased risk of infection. Impaired vascular status subsequently results in an inability to heal the soft tissue ulceration.

The second most common factor that results in amputation is related to impaired perfusion. In diabetes there is an accelerated rate of atherosclerosis and the distribution of atherosclerotic changes also includes the medium sized vessels below the knee. Microvascular abnormalities also occur affecting capillaries and arterioles. The end result of vascular disease may be rest pain, tissue necrosis, or gangrene. All of which may result in amputation.

**Amputation Prevention in Diabetes**
With the use of appropriate preventive measures, the high risk of limb loss in the diabetic can be decreased. In some institutions, preventive programs led to a 50 percent reduction in major amputations. The key element in any prevention program is education. This can be done through specialized clinics and with preprinted educational materials.

If foot deformities develop, specialized footwear, footwear modifications, and orthotics may be necessary to reduce the risk of neuropathic ulceration. These recommendations should be made by a podiatrist or rehabilitation medicine specialist. In some instances, an orthopedic surgeon's opinion regarding a surgical approach to the correction of deformities may be necessary.

**COMMON AMPUTATION LEVELS**

**Partial Foot Amputations**
For conditions where pathologic involvement is limited to the digits, the amputation of single toes through the level of the metatarsophalangeal joint is a reasonable procedure. If the level of pathology extends proximally to the level of the metatarsophalangeal joints, the amputation may be extended to include a portion of the metatarsal. These amputations, if healed, provide little functional deficit and require only minor modifications to footwear.

The next most proximal level of amputation is the transmetatarsal amputation. This procedure is often performed on the diabetic patient when toe deformities increase the risk of recurrent ulceration, or when there is necrosis of a number of digits. This amputation is generally viewed favorably because it also preserves neuromuscular function about the foot and ankle. There may be a need for some simple modifications to footwear after this procedure, which may include a custom-molded insole or a spring steel plate in the sole of the shoe, with a rocker bottom adaptation.

The Lisfranc amputation is carried out through the tarsometatarsal junction, whereas the Chopart amputation is performed through the more proximal intertarsal region, commonly at the talonavicular and calcaneocuboid junctions. These amputations frequently result in an equinovarus deformity. These two amputation levels are not held in high regard at this time due to the risk of having a painful weightbearing residual limb, plus an increased risk of developing skin ulcerations on the plantar surface resulting from the abnormal pressure distribution on the plantar aspect of the foot remnant.

**The Symes Amputation**
The Symes amputation is performed through the ankle joint. The tips of the medial and lateral malleoli are resected to the level of the talotibial articulation and the plantar heel pad that normally exists under the calcaneus is brought up to cover the distal surface of the residual limb.
There are a number of benefits to this level of amputation. The patient has an end-bearing residual limb that can be used for short-distance ambulation in the house without a prosthetic device. The length of the residual limb provides greater proprioceptive feedback, and there is also a reduced metabolic energy demand at this amputation level compared to higher levels. The disadvantage of this amputation level is that it leaves a somewhat bulbous-shaped ankle that lacks the normal contours of the intact lower extremity. The Symes amputation represents a choice between the functional and the cosmetic needs of the patient.

**Transtibial Amputation**
The transtibial amputation is performed transversely across the tibia and fibula at any length between the ankle and knee joints. It should ideally be performed at three-fourths of the length from the knee joint to the musculotendinous junction of the gastrocnemius, or at least 19 cm below the knee. It is common, however, that in the dysvascular amputee the residual limb may need to be shorter because perfusion may be inadequate to heal a longer transtibial amputation. Compared with the through-knee or transfemoral amputation levels, preservation of the knee joint leads to a much higher level of function and mobility. There are many prosthetic options for this amputation level, including many suspension systems, socket designs, interface materials between the socket and skin, and prosthetic foot designs. The transtibial amputation level is a highly successful amputation level, and there are many choices of components to enhance function.

**The Through-Knee or Knee Disarticulation Amputation Level**
The advantages of this amputation level relative to the transfemoral amputation, are that there is a longer residual limb, and therefore a more powerful moment arm for controlling the prosthetic limb, and it is an end-bearing residual limb that provides greater proprioceptive feedback. Limitations of this amputation level include a bulbous appearance of the residual limb because of the widened femoral condyles, and the difficulty in achieving a cosmetic end result. This amputation level is particularly useful in the traumatic amputee where the below-knee segment cannot be salvaged.

**Transfemoral Amputation**
Amputation at the transfemoral level should ideally be performed approximately three inches proximal to the knee joint line. This will maximize the length of the residual limb preserving muscle power, the moment arm of the residual limb, and a greater surface area for distribution of tissue loads.

**Complications**
Pain is a significant problem in many lower-extremity amputees. After completion of surgical healing the most common causes of pain include phantom limb pain, pain related to neuromas, and mechanical irritation from the prosthetic socket. The evaluation of post-amputation pain requires a careful history and physical examination, as well as laboratory investigations.

**Phantom Limb Pain**
Phantom limb pain is defined as pain after amputation that is in the distribution of the amputated limb segment. It is important to differentiate phantom limb pain from phantom limb sensation. Phantom limb sensation is the presence of any non-painful sensory experience in the portion of the limb which has been amputated. These sensations may include sensations of length, volume
and movement of the limb, or it may only include the perception of exteroceptive tactile sensory experiences. Phantom limb pain may occur in up to 90 percent of amputees but is of significant functional impairment in only about 10 percent. The etiology of phantom limb pain is not well established. It is felt by most to be the result of deafferentation and the loss of sensory input. It typically decreases somewhat in the first 6 months to 1 year after amputation. If the pain persists beyond this time, there is little likelihood that it will spontaneously resolve.

The treatment of phantom limb pain is challenging and requires an evaluation of behavioral and psychologic aspects, in addition to, other possible sources of pain. Many pharmacological agents have been used to treat phantom limb pain including Tegretol, amitryptilline, nortryptilline, propranolol, mexilitine, and gabapentine. Unfortunately there are few well designed studies that have evaluated their efficacy in phantom limb pain. A recent study found that amitryptilline was no better than placebo and was frequently discontinued because of side effects.

Neuromas
Neuromas occur any time a peripheral nerve is cut. Axonal sprouts appear at the cut end of the nerve, and are associated with proliferation of myelin sheaths and Schwann cells, forming a tangled enlargement where the nerve was sectioned. They are symptomatic when their anatomic location leads to mechanical trauma from a prosthetic socket. Typically the pain is an electrical dysesthetic sensation that radiates in the distribution of the peripheral nerve. This diagnosis is supported when a palpable nodule is found in the residual limb in the anatomic location of a peripheral nerve, and when a Tinel like phenomena is elicited during palpation. Clinical management primarily revolves around altering the design of the prosthetic socket to provide relief over the affected area or by choosing an optimum interface material, to minimize mechanical trauma to the neuroma. If pain persists despite optimum prosthetic management, the residual limb may have to be revised, the neuroma excised, and the sectioned nerves relocated to an area that will be less likely traumatized.

Mechanical Pain
Probably the most common source of discomfort in the amputee's residual limb is mechanical irritation from an ill-fitting prosthetic socket. The cause of this discomfort can be due to abnormal alignment of the prosthesis or to abnormalities in the fit of the prosthetic socket. If this occurs, the patient's prosthetist should evaluate the problem and make necessary modifications to the prosthesis. It is common for an amputee's residual limb to change shape over many years and for new prosthetic sockets to be required every 18 months to 2 years.

Functional Outcome After Lower-Extremity Amputation
As a general rule the more distal the amputation the greater the level of function. In particular the difference in functional outcome between the transtibial amputation and the transfemoral amputation is profound. In addition to amputation level, functional outcome is profoundly affected by comorbid medical conditions such as diabetic retinopathy, nephropathy, peripheral neuropathy, or underlying cardio-pulmonary or cerebrovascular disease. Age as an isolated variable is not critical to the outcome.

One retrospective evaluation of functional outcome after amputation in a population of 134 amputees ranging in age from childhood to the ninth decade, found that amputation generally results in reduced mobility. In this population, only 1 percent of the unilateral below-knee amputees did not walk, while 19 percent of the above-knee amputees and 35 percent of the bilateral amputees did not walk. However, 45 percent of both the unilateral below-knee and
above-knee amputees felt they walked as much as a normal person.

Employment is significantly impacted by an amputation, especially if the patient's job demands a significant amount of standing, walking, or walking over complex surfaces. Approximately 60 percent of patients who were of employment age return to some type of employment, and of those, only 22 percent return to their pre-amputation occupation. The use of vocational rehabilitation services is important in order to understand the functional demands of the patient's employment, and whether the post-amputation function is compatible with a return to this employment. Simple adaptations to the job often can be made, or, if return is not possible, retraining may be required to achieve employment in a more sedentary occupation.

**Psychological Aspects**
The psychological reaction to the loss of a limb is complex, and has not been well studied. It depends on age, chronicity of disease prior to amputation, time since amputation, and the importance of physical appearance to the patient's psychological well-being. The young traumatic amputee is said to undergo acute psychological distress in the early postoperative period, involving feelings of anxiety, resentment, and defiance. This is followed by fairly rapid adjustment. In contrast the elderly dysvascular amputee often has had a prolonged period of illness with the limb being painful and dysfunctional. In the early post operative period they may actually be relieved to have had the amputation. Subsequently, the reality of potential loss of independence may lead to a sense of hopelessness and despair. These feelings may be amplified during the rehabilitative process, especially as discharge from the hospital approaches.

A number of studies have shown that with time, most amputees adapt psychologically and generally are comparable to the non-amputee population on the psychological domains of standardized tests such as the SF-36.

**Rehabilitation Management of the Lower Extremity Amputee**
The optimum management of the lower extremity amputee requires a team approach, involving the skills of prosthetists, physical therapists, occupational therapists, social workers, vocational counselors, and psychologists. An integrated and comprehensive approach to patient management is essential for achieving both immediate and long-term success.

**Pre-amputation Rehabilitation**
Many patients who have a limb at risk for amputation spend considerable time on medical or surgical wards to heal infected foot ulcers or deal with the management of trauma or revascularization procedures. During this time, there is a significant risk of deconditioning and the development of contractures. It is therefore imperative that a maintenance program of physical therapy be instituted to maintain joint range of motion, muscle strength, and cardiopulmonary endurance. Once the decision has been made to amputate, it is important that the patient be educated as to what to anticipate during the operative and postoperative phases. In many amputation centers, audiovisual presentations have been developed. Amputee support groups also can be used to help the patient deal with some of the psychological consequences and stress that surround the pending amputation.

**Early Post-amputation Rehabilitation**
The major goals during the early postoperative phase of rehabilitation are to continue the strengthening and endurance program, continue the psychological support of the amputee, ensure wound healing and stump maturation, and effective pain management.
Wound healing and stump maturation are integral to the successful long-term outcome of the amputee. There are many different techniques that have been used to enhance the likelihood and rate of healing and maturation.

**Soft Dressing with External Elastic Compression**
The traditional postsurgical management of the amputee has been with a soft dressing, with or without external compression through an elastic ace bandage or stump shrinker. The ace bandage is applied in a figure-of-eight wrap, with graded pressure applied distally to proximally. It is important in this type of dressing that no circumferential proximal compression occurs because the resultant tourniquet effect will increase swelling and may delay or prevent wound healing. The extensive use of staff resources makes this technique unfeasible in most settings, and teaching patients to use this procedure is difficult.

The stump shrinker is an elastic stocking that is pulled onto the residual limb, and provides external compression. Use of a shrinker avoids the potential problems of either a tourniquet effect, or lack of uniform pressure application, which may occur with the elastic bandage.

**Rigid Dressings**
A rigid dressing is used particularly at the transtibial amputation level. It is a plaster cast extending from the distal residual limb to the mid length of the thigh. It can be used without a pylon and prosthetic foot attached distally, and is termed a rigid dressing, or it can be used with a prosthetic foot and pylon attached at the distal end, in which case it is termed the Immediate Post-Op Prosthesis or IPOP. Using this technique, the amputee may begin some early weight bearing on the day after amputation. The rigid dressing is felt to enhance wound healing by preventing edema and mechanical trauma to the healing incision. It also prevents the development of knee-flexion contractures because the knee is rigidly immobilized in near full extension. It is also thought to reduce the incidence of phantom limb pain and cause a more rapid rate of stump maturation.

**Late Post-amputation Management**
This phase of rehabilitation typically begins 3-4 weeks after amputation. At this point, the typical transtibial residual limb has healed adequately to allow the initial fitting of a prosthetic limb. The patient begins partial weight bearing progressing quickly to full weight bearing, over the ensuing 2 week period. As the patient's balance and gait improve, he/she begins walking outside the parallel bars using a walker followed by two forearm crutches.

The residual limb will continue to shrink and change shape over the ensuing 3-6 months, often requiring a new prosthetic socket. The selection of prosthetic components for each amputation level is complex and takes into account many variables. At the below-knee amputation level, the process of choosing prosthetic components must include an evaluation of the patient's cardiopulmonary status, body weight, vocational and avocational interests, length of the residual limb, and whether the residual limb has special characteristics, such as split thickness skin grafts, scar tissue, or neuromata. The prosthetic prescription includes the following major categories: suspension, prosthetic socket design, interface between the prosthetic socket and the residual limb, shank characteristics, and prosthetic ankle-foot characteristics. The choice of components is often made in conjunction with the prosthetist who will manufacture the limb.

Another major focus of the rehabilitation process at this time is continuation of education in management and hygiene of the residual limb and care of the prosthesis. Patients are also provided with a home exercise program to maintain and increase the strength of their lower
extremity musculature. The most important muscle groups for using a prosthesis are the hip extensor and hip abductor groups, both at the below-knee and above-knee amputation levels. The preservation of range of motion after discharge from the hospital is essential, as hip and knee flexion contractures can occur from prolonged sitting. One of the simplest ways to avoid these contractures is to have the patient lie prone twice a day for 20 min.

Effective long-term care of the amputee requires follow-up by an individual experienced in prosthetic care: either a rehabilitation medicine physician or a prosthetist. Careful follow-up of these patients allows one to anticipate problems in prosthetic fit before it causes skin breakdown and the possible need for surgical revision. This is also an ideal opportunity to evaluate and reinforce appropriate care of the intact lower extremity, therefore preventing further amputation.
REFERENCES


An excellent textbook covering all aspects of amputation surgery and complications, and a review of current prosthetic principles.


An excellent up-to-date reference dealing with many aspects of the diabetic foot, including the pathological changes, the etiology of these problems, prevention of foot injury, and management of neuropathic ulceration.


An excellent review of pain issues related to amputation.


A review of factors contributing to functional outcome after lower extremity amputation.


A review of current issues in surgical and early post operative management of the amputee.
Low Back Pain and Radiculopathy

Karen P. Barr, MD

Chapter 13

A. Epidemiology

Low back pain is a very common condition. More than three fourths of all people will have low back pain at some time in their lives. It is the second most common symptom, after the common cold, for patients to visit a physician’s office. The natural history of back pain suggests that it tends to be a recurring problem with multiple acute episodes. After an episode of back pain, 25% of people will have recurrence within a year, and 75% of people will have another episode during their lifetime. However the prognosis for marked improvement, and in many cases complete recovery after each episode is good: 65% will recover within 6 weeks, 85% within 12 weeks. Since back pain is such a common condition, the cost to society for the few percentage who develop chronic low back pain is enormous. Low back pain is responsible for 10-15% of all work absences in developed countries, and is the most common cause for disability in those younger than 45. Two to eight percent of the work force is disabled by back pain annually.

B. Diagnosis/Classifications

Low back pain is a symptom that may come from many structures in the back. The cause of low back pain is often difficult to determine, and some studies estimate that for over 85% of patients, no specific pain generator can be found. This may change as sophisticated injection techniques designed to isolate the source of pain become more widely used. Some of the most common causes of low back pain are discussed below.

Muscle strain: this is a common diagnosis given for acute low back pain, but it is unclear how much back pain is actually caused by muscle strain because there is no specific imaging or other gold standard test in order to make the diagnosis. A typical history associated with a muscle strain is an acute onset associated with muscle overload (such as heavy lifting). Pain is localized to the muscles of the back, and resolves in a few weeks with conservative care.

Ligamentous strain: Again, a difficult diagnosis to make because of the lack of diagnostic tests available and nonspecific findings on physical exam. However, the spine has an extensive ligamentous stabilization system and these tissues are innervated with pain fibers, so it is logical that this is a source of back pain. History is similar to muscle strain.

Disk disease and radiculopathy: Disc disease is probably one of the most common reasons for low back pain. This type of pain is generally thought to be improved by extension activities, such as standing, and worse with flexion activity, such as sitting and bending forward, because flexion increased the mechanical load on the disk. Disk disease can be further subdivided into annular tears, which are tears in the outer surface of the disk, internal disk disruption, which is degeneration of the internal structures of the disk, and disk herniation, which is a focal extension of the disk beyond the vertebral endplate. Disk degeneration is part of normal aging. It can begin as early as the late teen years, and continues throughout life. However, many people with signs of disk degeneration on MRI do not have back pain. Studies done on normal subjects without back pain show that by age 60 almost everyone showed degenerative changes, over 80% had disk
bulges, and more than a third had herniated disks. Disk bulges and herniations may cause no symptoms, but they also may cause compression, inflammation and irritation of the nerve roots, which causes radiculopathy. **Radiculopathy** is characterized by pain radiating down the leg. Symptoms are determined by the level of the irritated nerve root. See Figure 13.1 for characteristic symptoms and physical exam changes with different root levels.

<table>
<thead>
<tr>
<th>Root</th>
<th>Dermatome</th>
<th>Muscle Weakness</th>
<th>Reflexes Affected</th>
<th>Paresthesias</th>
</tr>
</thead>
<tbody>
<tr>
<td>L1</td>
<td>Back, trochanter, groin</td>
<td>None</td>
<td>None</td>
<td>Groin, after holding posture, which causes pain</td>
</tr>
<tr>
<td>L2</td>
<td>Back, front of thigh to knee</td>
<td>Psoas, hip adductors</td>
<td>None</td>
<td>Occasionally front of thigh</td>
</tr>
<tr>
<td>L3</td>
<td>Back, upper buttock, front of thigh and knee, medial lower leg</td>
<td>Psoas, quadriceps—thigh wasting</td>
<td>Knee jerk sluggish, PTR positive, pain on full SLR</td>
<td>Inner knee, anterior lower leg</td>
</tr>
<tr>
<td>L4</td>
<td>Inner buttock, outer thigh, inside of leg, dorsum of foot, big toe</td>
<td>Tibialis anterior, extensor hallucis</td>
<td>SLR limited, neck-flexion pain, weak or absent knee jerk, ankle dorsiflexion limited</td>
<td>Medial aspect of calf and ankle</td>
</tr>
<tr>
<td>L5</td>
<td>Buttock, back and side of thigh, lateral aspect of leg, dorsum of foot, inner half of sole and first, second, and third toes</td>
<td>Extensor hallucis, peroneus, tibialis anterior, gastrocnemius, ankle dorsiflexor, hamstrings—calf wasting</td>
<td>SLR limited to one side, neck-flexion pain, ankle jerk decreased, crossed-leg raising—pain</td>
<td>Lateral aspect of leg, medial three toes</td>
</tr>
<tr>
<td>S1</td>
<td>Buttock, back of thigh, and lower leg</td>
<td>Calf and hamstrings, wasting of gluteals, peroneals, plantar flexors</td>
<td>SLR limited</td>
<td>Lateral two toes, lateral foot, lateral leg to knee, plantar aspect of foot</td>
</tr>
<tr>
<td>S2</td>
<td>Same as S1</td>
<td>Same as S1 except peroneals</td>
<td>Same as S1</td>
<td>Lateral leg, knee, heel</td>
</tr>
<tr>
<td>S3</td>
<td>Groin, inner thigh to knee</td>
<td>None</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>S4</td>
<td>Perineum, genitalia, lower sacrum</td>
<td>Bladder, rectum</td>
<td>None</td>
<td>Saddle area, genitalia, anus, impotence</td>
</tr>
</tbody>
</table>

Note: Manipulation and traction are contraindicated if S4 or massive posterior displacement causes bilateral sciatica and S3 pain. PTKB = Prone knee bendings; SLR = straight leg raise.

Figure 13.1

**Zygapophyseal joint arthropathy** (also commonly called facet joint disease): This is also a common source of back pain, and includes osteoarthritis of the joints, as well as other joint injury and joint capsule injury. Back pain secondary to the zygapophyseal joints generally improves with flexion of the spine, and worsens with extension of the spine because extension loads the joints. Because the joints are innervated by multiple levels of the spinal cord, they often cause referred pain into the buttocok and leg that may be difficult to distinguish from radicular pain on history, but does not show the neurological changes seen with radiculopathy.

**Sacroiliac joint pain:** Historically, this is a controversial source of buttock and leg pain that may be confused with radiculopathy.

**Lumbar spinal stenosis:** Spinal stenosis is caused by a combination of facet joint hypertrophy secondary to arthritis and age related changes, ligamentum flavum hypertrophy which occurs with aging, and disk bulges. The classic symptoms are back pain and leg pain that worsens with walking (i.e. spine extension) and improves with sitting (i.e. spine flexion). This type of leg pain is called neurogenic claudication, and is distinct from vascular claudication in that leg pain caused by vascular disease can be relieved by just standing and resting and does not require sitting and resting for symptom relief.
Vertebral compression fractures: This type of back pain is most commonly seen in women with osteoporosis compression fractures. It is most common in the thoracic spine. Pain associated with this is variable, from nearly asymptomatic to extreme pain.

Red flag diagnoses: These are a group of uncommon but serious medical conditions that may cause back pain. They include cancer, either primary bone tumors, or more commonly, metastatic disease, infection, abdominal aortic aneurysm, cauda equina syndrome, and spinal fractures. See Figure 13.2 for more information on these conditions.

<table>
<thead>
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<th>Table 17A.1 Red flags for potentially serious conditions</th>
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<td>Possible fracture</td>
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<td>From medical history</td>
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<td>Major trauma, such as vehicle accident or fall from</td>
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<td>height</td>
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<td>Minor trauma or even strenuous lifting (in older or</td>
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<td>potentially osteoporotic patient)</td>
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<td>From physical examination</td>
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Figure 13.2

Yellow flag diagnoses: These are a group of confounding psychosocial factors that appear to make back pain more disabling. Although they are not the cause of back pain, they may worsen the prognosis for recovery, whatever the underlying cause of back pain, and so need to be taken into consideration when diagnosing back pain. They include depression, anxiety, post traumatic stress disorder, a history of sexual or physical abuse, compensation issues pending, unhappy work environment, an oversolicitous spouse, tendency to catastrophize, disproportionate downtime or rest with pain episodes, and many more.

C. History and Physical Exam

The History:
The important historical features for low back pain are similar to other pain complaints. Ten key questions to ask include:

1) location: one side or both sides? How high up do symptoms go, how far down?
2) Are leg symptoms present? If so, where does the pain radiate? Is numbness or weakness present in the legs? Where? What percentage of symptoms are in the legs as opposed to the back?
3) Onset: did the pain start suddenly or gradually? What was the patient doing when the pain started?
4) Intensity: usually on a 1-10 scale. Intensity measurement should be at that moment, at its best over the last several weeks or months, and the worst it has been over the last several weeks or months.
5) Character: aching, sharp, burning etc, for both the back, and if it occurs, for the legs.
6) Alleviating factors: for example, stretching, lying down, heat, medication, etc.
7) Exacerbating factors: for example, lifting, sitting, bending, stress, etc.
8) Does the patient have any red flags such as bowel or bladder incontinence? Saddle anesthesia? Nonmechanical pain at night? Fever? Unexplained weight loss? Night sweats? Cancer?, etc.
9) Is this the first time the patient has had pain? Is this a recurrent episode with complete pain relief between episodes? Is this a chronic condition? If so, why did they seek help at this time?
10) Is this related to a work injury? MVA? Is a lawsuit pending? Is the patient being compensated for their back pain?

The past medical history should include questions regarding history of vascular disease (to help rule out AAA as cause of back pain and vascular claudication as cause of leg pain), Cancer (to help rule out metastatic disease to the spine) recent infection, etc.

Social history should include: smoking (people who smoke have more disk disease) exercise history, sport participation history, work history and work satisfaction, social support, recent stressors, etc.

**The Physical Exam:**

**Inspection**: The exam begins with inspecting the back. Patients should be in a gown with the back open. The examiner looks for asymmetry which may signify scoliosis or muscle spasm. Then posture is evaluated. Common abnormal postures associated with low back pain include increased lumbar lordosis, swayback posture, kyphotic lordotic posture, or excessive kyphosis, perhaps from multiple compression fractures (see Figure 13.3).
Palpation: Palpation may be quite difficult if the patient is obese. It often helps to localize the level of palpation first by finding the top of the iliac crest, which is normally at the L4 vertebral body. Structures to attempt to palpate include: spinous processes, interspinous ligaments and joint spaces, facet joints and muscles.

Range of Motion: Usually for the spine, only active (not passive) range of motion is tested. Patient is asked to bend forward, bend back, side bend, and twist. Range of motion of the spine is difficult to evaluate. It can be limited by pain, fear, or patient effort. Neighboring structures may limit motion rather than the spine; for example, with forward bending, tight hamstrings may prevent the motion as much as decreased spinal flexion. It is still useful to test however, to give the examiner an idea of functional restriction and as a way to measure rehabilitation progress.

Neurological exam of the lower extremities: Although it has a low yield in patients that do not have radicular complaints, most physicians include a neurological exam of the lower extremities in all patients with back pain, both to uncover unknown symptoms, and to document a baseline if leg symptoms develop in the future. This neurological exam should include manual muscle testing, dermatomal sensory testing, reflexes, and straight leg raise.

D. Rehabilitation

The rehabilitation of back pain and radiculopathy can range from simple reassurance and advice to complex, multidisciplinary programs. Because the majority of patients with acute low back pain recover without ever seeing a specialist, PM&R physicians tend to see patients with back pain who have more complex physical or psychosocial issues. Below is an outline of a “typical” evidence based rehabilitation approach to radiculopathy, just one cause of back pain. However even with similar diagnoses, each patient is unique with unique issues that must be addressed for a successful outcome. It is appropriate to rehabilitate a patient with radiculopathy UNLESS they
have cauda equina syndrome, a rare condition that requires immediate surgical intervention, the patient is having progressive neurological weakness, or there is a “red flag” diagnosis (tumor or infection) as the primary cause of radiculopathy. In these cases a surgical referral or appropriate medical management is more appropriate than rehabilitation. Otherwise, the rehabilitation is the first course of management.

Acute Rehabilitation (day 1 to 3 months)

1) Modified activity: for an acute onset of back pain and radicular pain, the first step is generally to tell patients to remain as active as they are able, and avoid movements that aggravate leg symptoms. Advice is given for comfortable sleep positions and ways to accomplish activities of daily living that do not overly aggravate the patients symptoms. For acute exacerbations of chronic low back pain and radiculopathy, deconditioning may be a significant source of the pain and more aggressive exercise are prescribed from the first encounter because they are beyond the early phase of modified activity.

2) Pain management: This is generally a graded approach beginning with modalities such as heat or ice to relieve pain and accompanying muscle spasms, NSAIDS, and acetaminophen. If this does not control symptoms, leg pain can be treated with medications to relieve neuropathic pain and assist with sleep such as tricyclic antidepressants or gabapentin. Often, in the early phases of pain management a short course of an oral steroid taper is used with the rationale that it decreases radicular inflammation, although the scientific evidence for this is equivocal. Many PM&R physicians feel that relatively early treatment with an epidural steroid injection or selective nerve root steroid injection is helpful in preventing disability and deconditioning, although this is somewhat controversial because injections are expensive and carry some risk, and many patients with radiculopathy will improve with time and not require injections. These injections can relieve pain to allow adequate participation in an active rehabilitation program. Many physicians will treat acute back pain with radiculopathy with a short course of opioids. The chronic use of opioids for nonmalignant back pain is also becoming more common, although remains controversial since it is not always clear that the use of these medications increases function and quality of life in chronic pain syndromes.

3) Physical therapy: Once pain is under adequate control to allow for some activity, physical therapy is standard treatment for radiculopathy and back pain. Generally passive modalities such as massage and ultrasound are discouraged except in the most acute cases, and active exercise is encouraged. It is unclear from the literature at this time what type of exercises is most beneficial. McKenzie exercises are commonly used to treat this problem. This is a series of movements designed to help the patient find positions that decrease their symptoms and then maintain these positions. Usually for radiculopathy, this is a series of spine extension exercises. Another common physical therapy approach is lumbar stabilization in which the patient strengthens the muscles that support healthy posture such as abdominal muscles and buttock muscles, as well as the smaller spine muscles (eg. multifidi training). Manual therapy and manipulation are also commonly used during this phase. Meta-analyses of the literature have found this to be generally helpful for relieving acute symptoms, but of questionable value in changing long term prognosis or in treating chronic conditions.
Subacute Phase of Rehabilitation (3-6 months)

This is generally the phase where the PM&R physician becomes most involved. The natural history of back pain suggests that most patients will improve dramatically by three months, and those who do not have a good chance of not recovering at one and two years after development of the pain. Steps during this phase include

11) Exploration of why the patient has not improved: this may include imaging if there are unusual features to the case to help determine an underlying cause, gaining further information about psychosocial factors that may be influencing the patients recover (the “yellow flags” of back pain), and questioning about the patients compliance with medication and physical therapy exercises at home.
12) More aggressive pain management: this could include medications for neuropathic pain such as gabapetin. It is also commonly includes antidepressants, both for their pain relieving features and to treat depression that is so common in patients with persistent pain problems. Opioids are to be avoided in this stage if at all possible so that the patient can develop other pain management strategies. Injections are generally offered during this stage if not done earlier. Manual therapy may be included for pain relief.
13) Extensive patient education: This should include reassurance that the patients back pain does not signify life threatening or disabling disease, encouragement to maintain or increase activity levels despite pain to avoid deconditioning, and information about the nature of back pain with an emphasis on what the patient can do to improve their condition to foster self efficacy. Individual concerns about ongoing pain should be addressed.
14) More aggressive physical therapy: aerobic conditioning should be stressed (walking, biking, water aerobics, etc. the patient can tolerate) and a home program for this should be established. Strengthening becomes more of an issue to combat deconditioning, especially core strengthening, lumbar stabilization, and balance training. The scientific literature supports that physical therapy and exercise training is beneficial in this stage of treatment, but again the exact types of exercises that are best remain unclear.

Chronic low back pain

This phase of rehabilitation is the most patient specific. It includes patients with pain anywhere from 6 months to many years. If not already done, imaging such as an MRI is generally indicated to further localize the source of the pain. Surgery consult may be an option if radicular leg pain is persistent and the patient has complied with conservative treatment without improvement. If a patient is not significantly disabled by their pain and surgery is not an option, the patient may benefit from a comprehensive pain program that includes a multidisciplinary approach. Behavioral modification, pain relieving techniques such as meditation and relaxation, psychological counseling, continued education about living with persistent pain, more structured exercise counseling and physical therapy, and medical trials with close monitoring are generally all included in a comprehensive pain program. Selected patients may be candidates for more invasive procedures such as spinal cord stimulators, morphine pumps, spinal fusions, etc. This is
generally done in conjunction with psychiatry to continue to promote the goals of function, increased activity, and in many cases, vocational rehabilitation and return to work.

E. Summary

Low back pain is extremely common. It is caused by a wide range of pathology, and the exact cause is often difficult to establish. A comprehensive rehabilitation evaluation is invaluable in the effective treatment of back pain. This includes a thorough history to rule out rare but serious causes of back pain, and to identify patients that may be slow to respond to treatment. A thorough physical examination includes both an exam of the musculoskeletal system and a neurological exam to identify radiculopathy. Treatment includes pain management with medication, modalities, and injections, and restoration of function with physical therapy and exercises. With this comprehensive approach, most disability from low back pain can be prevented.

REFERENCES


Chronic Pain

Heather R. Kroll, MD

**Definition**

Chronic Pain is often defined by the amount of time the patient has suffered from pain. Time frames vary from practitioner to practitioner, but three to six months is a typical range. More useful, however, is to define chronic pain as pain that persists beyond the expected time of healing.

Chronic pain patients can be divided into two broad categories: a) Those with chronic pain due to cancer, or malignant disease, and b) Those with chronic pain due to other causes. This second category, sometimes referred to as chronic “non-malignant” pain, has two further subdivisions: a) Patients with pain which is chronic because of recurrent or persistent acute pain from disease processes such as migraine, or arthritis, and b) Patients with pain which persists beyond the expected time of healing for whom there is no identifiable acute cause of their ongoing symptoms. This last group of patients often presents the greatest challenge to physicians.

Most of what follows in this chapter applies primarily to the patients in this last group; although, many of the principles are applicable to chronic pain of any etiology.

**Epidemiology**

Chronic pain is a very common and expensive problem. Prevalence varies from 2% to 40%, according to studies and methods used. A recent Finnish study estimated the prevalence of any chronic pain (once weekly to daily) at 35% with the prevalence of daily pain increasing in those over 40 years of age. Annual incidence of low back pain is estimated at 5% with a lifetime risk of 60% – 85%. Chronic pain of any cause impairs quality of life and is related to low self-rated health independent of age and other chronic diseases. It is a frequent cause of visits to primary care physicians.

The expense of chronic pain is due both to utilization of health care and to lost productivity. Low back pain, the most common cause of work days missed due to disability related to pain, is estimated to cost at least $16 billion per year, one-third allocated to medical costs and two-thirds for compensation. Pain not only causes workers to miss days of work, but also impairs their performance while they are at work. The estimated cost in the United States of lost productivity from reduced work performance due to pain is $61.2 billion per year.

**Etiology**

Chronic pain is a biopsychosocial phenomenon. Its etiology is usually multifactorial. Factors contributing to its development come from the nervous system, the individual and society. These factors interact to produce not only the symptoms that the patient complains of, but also the profound disability that may accompany these symptoms. The individual and societal factors are often more important in promoting and sustaining this disability than the nervous system factors.

The typical chronic pain patient starts out with an identifiable injury which generates nociceptive impulses and acute pain. In most cases, the injury heals, nociception diminishes and pain resolves. In some cases, however, despite healing, pain does not resolve and disability develops.
The nervous system can contribute to the development of chronic pain through changes that occur both peripherally and centrally. In the peripheral nervous system, pain messages are carried by the smaller, poorly myelinated or unmyelinated A-delta and C fibers. These fibers are stimulated by mechanical, chemical and thermal stimuli. Tissue injury causes the release of a variety of endogenous chemicals which have excitatory effects on nociceptors. Chemical changes in the microenvironment may sensitize the nociceptors to non-noxious stimuli. Patients with diseases that are characterized by chronic inflammation often experience allodynia, hyperalgesia and hyperpathia due in part to peripheral nerve excitation and sensitization. Injury to peripheral nerves also can be a cause of persistent pain. Injured nociceptive fibers may fire spontaneously leading to the ongoing experience of pain in the absence of new tissue injury.

Changes within the central nervous system also play a role in the development and perpetuation of chronic pain states. Sensory fibers, both nociceptive and non-nociceptive enter the spinal cord through the dorsal root and synapse in the dorsal horn. From the dorsal horn, pathways then ascend to the brain to synapse, or ultimately terminate in, the brainstem, midbrain, thalamus, limbic system and cortex. The neurons of the dorsal horn respond to multiple inputs. These inputs come, via interneurons, from nociceptors and other sensory fibers, as well as from descending fibers from the brain. Changes can occur in the spinal cord which lead to hypersensitization of this first relay station and consequent abnormal processing of both nociceptive and non-nociceptive sensory input. As a result, non-nociceptive input may be transmitted to higher centers as a pain signal and the intensity of nociceptive input may be effectively exaggerated. Higher centers, the limbic system for example, also have a powerful descending control over the spinal cord and can significantly influence the degree to which spinal cord signals are transmitted upward.

The complexity of the nervous system response to pain leads to individual factors which contribute to chronic pain. Injury and pain may lead the patient into a pattern of disuse, guarding and protection of the injured part. These altered patterns of movement change the biomechanical forces on joints and muscles and typically lead to loss of conditioning and flexibility. As a result, the ordinary activities of daily life may put stresses on these structures that they do not tolerate well, and which cause pain.

Patients believe that any pain is a signal that injury or harm is occurring to their body. They learn to avoid any activity that causes symptoms. As ordinary activities become painful the patients may become fearful of even simple daily living skills. They fall into a learned pattern of fear and avoidance. This contributes to increasing levels of inactivity, ongoing loss of physical conditioning and therefore less ability to tolerate the stresses of normal everyday life. These changes contribute to the patients’ developing conviction of disability.

Depression frequently develops as patients see themselves losing their ability to participate meaningfully in their own lives. The depression then feeds back into the system by increasing the patient’s sensitivity to pain signals and increasing the suffering that they experience on account of their pain. A downward spiral develops in which pain leads to inactivity leads to depression leads to more pain, more inactivity and worsening disability.

Patients not only learn to change activities in response to pain, but pain itself may become a learned behavior. It is here that the social milieu of the patient is most important. Patients exhibit a variety of behaviors (e.g. grimacing, complaining, limping) that let others know that they are in pain and are suffering. People in the environment respond to these behaviors and often inadvertently reinforce them. Pain behaviors can elicit positive benefits to the patient such as more attention from a spouse, or excuse from participating in work and other usual life roles. These positive benefits reinforce the pain behaviors. Concerned friends and family can also contribute to the development of disability in a patient by insisting that the patient rest and not participate in activities for fear that they will get injured, or worsen their pain. The health care system may also play a negative role. The endless search for a cause of the patient’s
symptoms together with a parade of ineffective treatments, reinforces the patient’s belief that, “There is something dreadfully wrong with me; it cannot be fixed and I will be disabled forever.”

The role of learning and social factors must not be underestimated or ignored. It is here that the key to effective evaluation and treatment lies.

Evaluation

Patients hope that their visit to the doctor will result in an evaluation which will reveal what is wrong with them and will identify treatment which will cure them of their ailment. Chronic pain, almost by definition, often precludes the identification of a specific pain generator which can somehow be fixed, thereby relieving the patient of their suffering. This does not mean that no evaluation should be undertaken; but attention always needs to be addressed to when it is time to stop the evaluation process and move on toward treatment.

As a biopsychosocial entity, chronic pain requires a biopsychosocial approach. Evaluation should include imaging and laboratory studies to look for identifiable and treatable diseases. Additionally, assessment must not overlook common associated conditions such as depression and sleep disturbance. Assessment of the degree of disability present in the patient’s home and work life is important for understanding the impact of pain on his/her life and also for understanding factors which may be contributing to the reinforcement of pain behaviors.

Treatment

Effective treatment of the chronic pain patient begins with establishing a therapeutic relationship with the patient. Most of these patients have seen a large number of providers in their search for a diagnosis and treatment of their symptoms. With no definitive test results, they feel that no one believes in their suffering. Previous treatments have often been ineffective or have made them feel worse. They may be taking large quantities of a wide variety of medications with little benefit and multiple side effects. They are commonly depressed, discouraged and sleep deprived.

Begin by listening carefully and believing in the patient’s symptoms. Review evaluations that have been done, and do things that have been missed. When no more is to be gained by further evaluation, then stop. Eventually, the patient must stop waiting for recovery and searching for a cure and begin actively participating in his/her own rehabilitation. Effective treatment begins by moving the patient toward the recognition and acceptance of this need to move on.

Educate the patient about chronic pain and its etiology so that he/she can understand how it is possible to have symptoms that are real and yet not amenable to simple explanation. Change the patient’s focus from the search for a cure, to the recovery of function and the return to control over his/her life.

The primary goal of the overall treatment plan is to improve function and eliminate disability. Improved pain control may occur as part of the process, but must not be the primary focus. The primary goal will be accomplished most effectively by a multidisciplinary approach that utilizes the expertise of a variety of professionals in a coordinated plan. Because learned behaviors can play such a large role in perpetuating chronic pain states, incorporating a behavioral approach into the treatment plan is important.

Most chronic pain patients have already experienced a wide range of passive treatments including massage, physical therapy modalities, injections and nerve blocks. They may have gotten temporary relief from these, but never a cure. These treatments may still have a role, but they should be used as adjuncts to help facilitate improved function. For example, in patients with lumbar disc herniation and radicular pain,
Epidural steroid injections can give temporary pain relief. This procedure is not curative. However, if it is combined with an active physical therapy program directed at recovering and improving strength, flexibility and body mechanics, it can be helpful in improving function.

Physical and occupational therapy play an important role in helping patients regain their strength, flexibility, coordination and endurance. As with any injury, the biomechanical consequences need to be analyzed and addressed with specific exercises to stretch, strengthen and regain coordination. However, the approach to the chronic pain patient is different than for the patient with an acute injury. With chronic pain, the therapist also needs to set out a gradual progression of activities focused on improving function in ordinary daily activities such as walking, sitting, standing, climbing stairs, lifting and carrying. The approach is behavioral, often described as “quota-based”. At initial assessment, patients are asked to do what they can, within their own perceived limits of pain and fatigue. A baseline of activity is then set 20%-30% below this level of performance. A gradual progression is outlined with each day’s activity for each particular exercise called the quota. The goal is for patients to perform this daily quota, and only this amount, regardless of the degree of discomfort and fatigue they are having on any particular day. This means that on a day when patients are feeling worse, they are still encouraged to do the quota for that day. When they are feeling better, they must avoid doing activities beyond the quota. The therapists give reinforcement for activities done appropriately and do their best to ignore and not reinforce pain behaviors. This behavioral approach helps to desensitize patients to the ordinary activities they have become fearful of and shows them that they can do more and improve without significantly aggravating their symptoms. It helps to bring patients back into control of their lives and to remove pain as the driving force behind decisions about activity.

The psychologist plays a key role in helping to manage these difficult patients. The focus is on cognitive-behavioral strategies for managing depression and improving coping skills. Frequently, there is a life-long history of poor coping which has contributed to the patient’s developing disability. Patients need to learn to recognize their negative and catastrophic thinking and maladaptive behaviors and to replace them with thoughts and behaviors that promote ability rather than disability. The psychologist can also work with the patient’s family members to help them understand the ways in which their actions have reinforced pain behaviors and disability within the patient. The goal is to move the patient back toward resumption of his/her normal life role. Treatment should also keep in mind the type of work the patient had been doing and help establish whether return to this type of work is realistic in terms of physical capacity or if other options should be explored.

Medications, perhaps, play the most controversial role in chronic pain management. Medications, like other passive treatments, should be used to facilitate improvement in function. There are a wide variety of medications that may play a role in chronic pain management; these include non-steroidal anti-inflammatory drugs, antidepressants, anti-neuropathic agents, muscle relaxers and opiates. All medications have side effects and their beneficial effects may fluctuate over time. Periodic reassessment of the risk-benefit ratio of medications used and the elimination of those whose benefits are not worth their adverse effects will help patients feel better.

Opiates, when used for chronic pain, are a class of medications about which there is much controversy, passion and opinion but very little data from controlled studies. These medicines are certainly among the most effective for acute pain. When used for chronic pain, over long periods of time, their utility is less clear. In general, pain-contingent use of short-acting opiates contributes to the patient’s ongoing focus on pain. It may lead to escalation of dosing and development of tolerance. Use of time-contingent, long-acting medications helps to facilitate removing the patient’s focus from his/her pain and onto activities to promote improved function. Opiates probably can play a useful role in some patients if it can be demonstrated that function actually improves with medication use. However, in many patients, long term opiate use leads to loss of medication efficacy and significant side effects, including depression and
cognitive dysfunction. Patients continue the medication only because they hope it is helping and they fear withdrawal symptoms. When gradually tapered, in the context of a multidisciplinary treatment approach, these patients typically do not experience increased pain levels and in fact feel much better as side effects go away.

Conclusion

Chronic pain is a complex, biopsychosocial condition which leads to tremendous amounts of suffering, disability, and cost to both individuals and society. Effective management requires recognizing this complexity and approaching treatment in a multidisciplinary manner. Maintaining a focus on function and the prevention or elimination of disability will lead to the best outcomes overall.

REFERENCES


The information for the section on epidemiology came from:

1. Jacobson’s chapter above.
Approximately 10-15 percent of children in the United States have some form of chronic disease. Ten percent of this group, or 1 percent of all children in the United States, have a significant disability that interferes with life on a daily basis. Chronic diseases and disabilities have replaced acute illnesses as the major cause of mortality and morbidity for children in this country. Physical disability differs in children and adults even when the underlying diagnosis is the same. The adult who acquires a disability suffers a loss of independence; the child who has or acquires a disability suffers loss in his or her potential to achieve independence.

**Common Childhood Disabilities**

**Cerebral Palsy**
Cerebral palsy (CP), defined as a disorder of motor control due to an injury or abnormality affecting the immature brain, is the most common condition associated with childhood disability. The incidence is 2 per 1,000 live births. Spastic, athetoid, ataxic, hypotonic, or mixed patterns of motor disability may be present. There may be severe involvement of all four extremities (quadriplegia); involvement primarily of the lower extremities with upper extremity deficits only present to a lesser degree (diplegia); involvement of an upper and lower extremity on the same side of the body (hemiplegia); or, rarely, involvement of a single extremity (monoplegia). The type of motor disability depends on the site of insult to the developing brain; spasticity results from involvement of deep white matter and cortex; athetosis is associated with damage to the basal ganglia. The severity is variable. The etiologies are diverse and can be divided into prenatal, perinatal, and postnatal factors. An example of prenatal etiology is congenital infection. Perinatal causes are associated with any period of anoxia or brain injury associated with labor and delivery. Postnatal etiologies include any insult to the brain in the early period of childhood, including neoplasia, trauma, or anoxic injuries to the central nervous system. The etiology of CP remains unclear in many cases.

While the prevalence of cerebral palsy remains stable, there have been changes in the incidence of the types of cerebral palsy that reflect changes in common etiological factors. There has been a reduction in pure athetoid cerebral palsy during the past generation of medical care, primarily due to a decrease in the incidence of hemolytic anemia, hyperbilirubinemia, and kernicterus. This is a result of practice of immunizing Rh-negative mothers and the clinical advances in the treatment of infants with hyperbilirubinemia. Hypoxia is now the most common factor associated with athetosis in CP. In contrast, there has been an increase in spastic diplegic cerebral palsy associated with the increased survival of premature infants who are predisposed to insults involving the periventricular vasculature of the immature brain.

The problem of motor control accounts for many of the functional deficits. Associated problems include poor oral motor control, feeding problems, gastroesophageal reflux, seizure disorders, sensory impairments, and secondary orthopedic deformities. Although children with CP can have impaired, average, or superior intellect, there is higher incidence of cognitive
dysfunction and communication deficits in this population. The clinical course of CP is non progressive, although clinical manifestations may change with normal growth and development.

Meningomyelocele

Meningomyelocele is the second most common congenital cause of pediatric disability. Its incidence is slightly less than 1 per 1,000 live births in the United States. The cause is unknown, but appears to be multifactorial and includes a polygenetic inheritance pattern and environmental factors. The defect results from failure of neural tube closure in the developing embryo at the end of the fourth week of gestation. Hydrocephalus associated with the Arnold-Chiari Malformation is present in over 90 percent of affected children. Medical management issues are related to the neurologic level of the neural tube lesion. Forty five percent of the lesions occur at the L-5/S-1 levels, whereas 92 percent occur at L-2 and below. Less than one-third will have completely flaccid paralysis below the level of the lesion; the remainder will have a combination of flaccid paraplegia, spasticity, and occasionally some spared voluntary activity. Virtually all children with lesions above L-2 will be wheelchair users. Approximately 50 percent of children with spared L-3 innervation will walk in childhood, and 20 percent will walk independently as adults. With L-4-5 sparing, nearly 100 percent of children will ambulate in childhood, but a lesser number will have independent ambulation in adulthood. The majority of patients with intact innervation to S-1 will be independent ambulators in adulthood.

All children with meningomyelocele have neurogenic bowel and bladder. Bowel management is of tremendous significance to the child’s self-esteem and socialization. Management of the neurogenic bladder is imperative in order to prevent secondary damage to the upper urinary tract and the life-threatening complications of infection and renal failure. The survival rate for children born with meningomyelocele in the 1950s was less than 1 percent; it is now greater than 90 percent. Diagnosis is now often made prenatally and adjustments made to labor and delivery in order to minimize additional neurologic damage to the infant caused by the trauma of delivery. Current medical management in the newborn period includes early surgical management of the open neural tube defect and ventriculoperitoneal shunting to reduce hydrocephalus. Continued aggressive management designed to prevent secondary complication associated with the urinary tract have dramatically decreased the incidence of morbidity and mortality associated with renal failure. The most common cause of death for children with meningomyelocele at this time is central ventilatory dysfunction. This complication is associated with the Arnold-Chiari malformation and abnormalities at the level of the brain stem.

Nearly 100 percent of these children will suffer skin breakdown associated with insensate skin. Maximum education and intervention should be provided to the child and his or her family to prevent the occurrence and the secondary complication of decubiti.

If the most efficient form of mobility for an affected child is independent ambulation, considerable effort should be directed to the provision of appropriate orthotics, orthopedic surgery, and therapy to maintain this skill. Wheelchair prescription is required if the child’s functional needs will be best met with augmentative mobility.

The first cohort of patients with spina bifida surviving with modern medical management is just now entering adulthood. Early studies directed toward evaluating the clinical needs of this population suggest that they have a high incidence of late neurologic complications, ongoing musculoskeletal complications, an unacceptably high rate of skin breakdown, and significant psychological, social, and vocational needs.

Neuromuscular Disorders

Neuromuscular disorders commonly encountered in children differ from those typically seen in adults. This is true for each level of the motor unit. The most common form of anterior horn cell disease in the pediatric age group is spinal muscular atrophy. Peripheral neuropathies are relatively rare in childhood and are more likely to be inherited. Some are caused by inborn errors
of metabolism affecting myelin, such as metachromatic leukodystrophy and Krabbe’s disease; others are associated with hereditary dysfunction of myelin (e.g., Charcot-Marie-Tooth disease or Déjerine Sottas interstitial neuritis). Peripheral neuropathy may also be acquired in childhood, as in the case of inflammatory polyneuropathy or Guillain-Barré syndrome, or following toxic exposure to chemotherapeutic agents and heavy metals. The more commonly seen disorders of the neuromuscular junction in children are myasthenia gravis and infantile botulism. Myopathic processes presenting in the pediatric age group include inherited muscular dystrophies, myotonic dystrophy, congenital myopathies, and inflammatory myopathies.

The most common muscle disease in children is Duchenne muscular dystrophy, an X-linked recessive genetic condition. This single-gene defect results in a deficiency of the muscle protein dystrophin and subsequent progressive deterioration of skeletal muscle. Affected boys characteristically have histories of delayed motor development; proximal weakness becomes apparent by the age of 3 to 4 years. Early manifestations of this weakness classically include the presence of a Gower’s sign, in which the hands are used to “walk-up” the thighs to assume a standing position. Diagnosis is aided by the finding of marked elevation in the serum creatine kinase, cDNA gene probe to identify the abnormality at the gene locus which is present in 65%, of cases those with deletions too small to detect are diagnosed by characteristic abnormalities in the muscle biopsy and specific histochemical assay to document the absence of dystrophin. Weakness is inexorably progressive. The period of ambulation may be prolonged with aggressive physical therapy. Over twelve clinical trials with prednisone have had encouraging results, with slowing in the rates of decline in strength and function. Loss of the ability to ambulate and complete dependence on a wheelchair usually occur before 12 years of age. The patient’s course may be complicated in the later stages of the disease by scoliosis, decreasing pulmonary function, and cardiac involvement. Approximately one-third of boys with Duchenne muscular dystrophy have intellectual impairment. Death usually occurs in the late teens or early in the third decade from respiratory compromise, a subset will have significant cardiomyopathy which will progress to congestive heart failure. Assisted ventilation in the end stage may prolong life for up to eight additional years.

Spinal muscular atrophy (SMA) describes a spectrum of genetic, autosomal recessive, disorders characterized by degeneration of the anterior horn cells. Classification systems vary; one useful system suggests the division into three major types. SMA-Type I is the acute infantile syndrome, Wernig-Hoffman disease; SMA-Type II is an early-onset childhood form; and SMA-Type III, or Kugelberg-Welander disease, has a later onset. Infants with SMA-Type I may present shortly after birth or within the first 6 months of life with respiratory distress and feeding difficulties. Weakness is generalized and symmetric; there is limited development of motor function, with affected infants typically never sitting independently. Death usually occurs in the late teens or early in the third decade from respiratory compromise by 2 years of age. At the other end of the spectrum of severity, SMA-Type III is usually clinically detected between the ages of 5 and 15 years. Proximal weakness occurs initially, with more distal involvement later in the course of the disease. Progression is highly variable, with some patients remaining ambulatory for decades and others losing the ability to walk by age 20. The majority of affected children have the intermediate form. The age at onset is variable and overlaps with the age at onset of SMA-Type I. Delayed motor development is characteristic; while some children will develop the ability to sit and stand independently, most will maintain these skills only briefly and will be dependent on a wheelchair for mobility by 2 or 3 years of age. Bracing and physical therapy may prolong ambulation briefly. Life span is extremely variable, but death usually occurs as a consequence of pulmonary complications. There is no intrinsic intellectual impairment associated with spinal muscular atrophy.

Limb Deficiencies
An absence of part or all of an extremity in childhood may be due to congenital deficiency or acquired amputation. Over two-thirds of children with limb deficiencies have congenital lesions.
The most common congenital limb deficiency (25 percent of the entire group) is the below the elbow transverse defect. Among children with acquired amputation, lower extremity amputation is more common. For unknown reasons, left-sided congenital lesions predominate, and boys are more affected than girls. The etiology is unknown in over 90 percent of children with congenital limb deficiency. These deficiencies may be associated with inherited syndromes of multiple malformations. Those involving the radius have been associated with congenital heart disease, blood dyscrasias, and vertebral anomalies. Trauma accounts for 70 percent of acquired amputation and malignancy is responsible for most of the remaining 30 percent. Prostheses are provided for children with limb deficiencies to accommodate function. Provision of the initial upper-extremity prosthesis has generally been recommended at the age at which the child is able to sit. There is a trend toward earlier prescription, and some clinicians provide prostheses for children as young as 3-4 months of age. The initial upper-extremity prostheses are passive devices that allow the infant to hold large objects, support weight bearing, and provide some sensory feedback. Cable activation of a prehensile terminal device is introduced at age 18-24 months when the ability to learn to operate the device has developed. For children with above elbow deficiencies, the elbow joint is activated at approximately 3 years of age. Consideration in selection of prostheses for children should include growth potential, durability, comfort, weight, and cosmesis. The devices should be as simple as possible in construction and in their operating requirements. One exception is the provision of myoelectric prostheses. In certain selected cases, upper-extremity prostheses with electrically powered terminal devices and/or elbow joints are used. Recommendations for myoelectric prostheses are based on the functional needs of the child and the user’s ability to adapt to the prostheses rather than on age alone.

Not all children with congenital or acquired amputations will benefit from prosthetic devices. There are high rates of rejections of prostheses, particularly at the extremes of residual limb length. Children with very distal amputations are usually more functional using their residual hand or foot than a prosthesis. Excessively high energy requirements may be required for the use of prostheses by children with short lower-extremity amputations. Prostheses provided for some proximal upper-extremity amputations may be so cumbersome that they offer only minimal functional advantage. It is important to note that children born with only one useable upper extremity will generally become completely independent in all aspects of self-care without a prosthesis. Children born with an absence of both upper extremities can be expected to be independent in most aspects of their self-care by using their feet.

Surprisingly, children with these deficiencies require very little formal therapy. The adaptive nature of the developing child encourages them to teach themselves one-handed or no-handed techniques. If a prosthesis improves the child’s function, it will usually be accepted. Upper-extremity prostheses are rejected at a high rate, both for functional and cosmetic reasons; in contrast, lower-extremity prostheses are so functionally useful in gait that they are nearly always accepted. Most children with limb deficiencies who become successful prosthesis users learn to use them without extensive periods of training in physical and occupational therapy.

**Spinal Cord Injury (SCI)**

Spinal cord injury (SCI) most often occurs in adolescents and young adults between the ages of 15 and 25 years. When SCI occurs in young children, the results are frequently more devastating. Several developmental anatomic and biomechanical differences in the immature spine predispose the upper cervical spine to the most severe injuries in younger children. These include increased ligamentous and joint capsule laxity, incomplete ossification of the vertebrae, immature shape of the vertebral bodies, and incomplete muscular development. All of these contribute to increased mobility in the developing upper cervical spine. The mass of the infant’s head is proportionally higher than in the adult, and the fulcrum of flexion-extension of the cervical spine is higher, with a gradual shift from the C2-3 level in the infant to a C5-6 level in the adult spine. These
developmental factors result in a higher proportion of quadriplegia in young children with SCI. With high cervical cored lesions, the child may be dependent on mechanical ventilation. Management of neurogenic bladder is critical to avoid renal damage. A child with quadriplegia may be expected to perform intermittent catheterization independently at approximately 5 years of age. Neurogenic bowel retraining and regulation is generally accomplished without great difficulty in younger patients. Injury during adolescence, when bone metabolism is very active, may be complicated by immobilization hypercalcemia. This problem should be suspected in teenagers with SCI who present with lethargy, anorexia, nausea, headache, polyuria, and polydipsia. Heterotopic ossification and deep vein thrombosis following SCI occur less frequently in children than in adults.

Pediatric Brain Injury

One million children sustain head trauma annually. Traumatic brain injury is the most prevalent cause of acquired disability in childhood and is presently the most common diagnosis leading to inpatient pediatric rehabilitation. Etiologies in children are, in order of frequency; falls, motor vehicle accidents, pedestrian accidents, bicycle injuries, and sports injuries. Child abuse is a common cause of serious brain injury among children less than 2 years of age. While motor vehicles do not account for the majority of all brain injuries in children, they are the major cause of severe brain injury. Severity of injury for children is greatest under 2 years and over 14 years of age. These extremes may be associated with the mechanisms of child abuse and motor vehicle accidents, respectively. The frequency and severity of pediatric brain injury in children is higher in boys than in girls.

Neurologic sequelae depend on the location and severity of the brain injury and the age of the child. Motor dysfunction reflects the site(s) of involvement in the motor pathways, and may result in spasticity, ataxia, and/or extraneous movements that impair the child’s mobility and self-care. Visual disturbances include deficits in acuity, visual fields, and perception. Hearing deficits are especially associated with fractures of the temporal bone. Conductive hearing losses are associated with longitudinal fractures, whereas sensorineural losses are associated more frequently with transverse fractures. Expressive language disorders following brain injury in children may include dysarthria and aphasia. Receptive deficits may be related to hearing loss or auditory perceptual problems.

Cognitive problems following traumatic brain injury typically include deficits in attention, concentration, memory, information processing and performance speed, cognitive flexibility, abstract problem-solving skills, and judgment. These deficits may persist even when intellectual function and academic performance test results are within normal ranges. These cognitive difficulties have obvious educational implications. Evidence of such deficits may not be detected initially following injury, but may become more apparent with time. For example, a child injured as a toddler will not demonstrate difficulty with tasks requiring abstract problem solving until demands are placed on him or her to participate in these activities in early elementary school. Changes in personality and behavior may also follow traumatic brain injury in childhood. Decreased frustration tolerance, poor anger control and increased aggressiveness and hyperactivity have been observed to follow even mild brain injury in children. While the sequelae of moderate and severe brain injury in childhood is most likely to warrant referral to pediatric rehabilitation, the effects of even minor injury may be more significant than is generally appreciated. Subtle neuropsychological and neurobehavioral change may impair a child’s ability to function in school sufficiently to warrant special education.

It is not clear that the outcome following brain injury is either better or worse for children than for adults. Physiologic differences exist between the child’s and the adult’s brain in parameters that include cerebral metabolic rate and mechanisms regulating blood flow. Skull compliance, cerebral water content, and extent of myelination vary with age and degree of brain development. Some of these factors increase, and others decrease, the likelihood of recovery of function in children.
Suggested Reading

2. Dystrophin and mutations: one gene, several proteins, multiple phenotypes, Lancet Neurology, Vol2 no 3
3. Brook MH. A clinician’s view of neuromuscular diseases (2d ed.). Baltimore, Williams & Wilkins, 1986
   This reference offers very readable, clinically oriented description of neuromuscular diseases affecting both adults and children. The chapter on symptoms and signs of neuromuscular disease provides an excellent discussion of clinical clues useful in evaluation of pediatric patients with these disorders, with an emphasis on functional evaluation.
   Traumatic brain injury in childhood is the topic of this issue, which includes relatively brief but comprehensive articles that provide a good introduction to the subject. Epidemiology and prevention, sequelae, and rehabilitative management of brain injuries are reviewed with an emphasis on aspects particularly relevant to pediatrics.
   Fundamentals of diagnosis and management in pediatric rehabilitation are presented in the first part of this text. The second part describes specific applications of rehabilitative interventions to the common disabling conditions of childhood.
   Etiology, classification, and management of congenital limb deficiencies and amputations acquired in childhood are described. The chapters on basic principles of prosthetics for the upper and lower extremities provide a good introduction to the subject of prosthetic components appropriate for use with children.
   The development of normal gait is described. Methods for studying gait in children are presented, and gait disorders commonly seen in pediatrics discussed.
   This is a current review of the etiology, epidemiology, and classification terminology of cerebral palsy.