

ESSENTIALS OF CLINICAL NEUROLOGY

CHAPTER 5: WEAKNESS AND GAIT DISTURBANCES

Normal voluntary muscle contraction can be described as an integrated sequential neuromuscular function. Impulses originate in the large upper motor neurons, primarily in the Betz cells, and travel through corticospinal tracts to lower motor neurons in anterior horns of spinal cord and motor nuclei of brain stem. The impulses from these lower motor neurons travel through the peripheral and cranial nerves to reach the nerve terminals, where acetylcholine that has been synthesized and stored in vesicles is released to reach the acetylcholine receptors on the postsynaptic muscle membrane. When acetylcholine reaches the receptor sites, there is a transient increase of permeability to sodium and potassium resulting in electrical depolarization of the muscle membrane. This initiates the sequence of events that leads to voluntary muscle contraction. Lesions at any site along these pathways can cause weakness. Clinical manifestations differ according to the lesion level. The clinician should be able to determine if weakness is due to a lesion in corticospinal tract, anterior horn cells, peripheral nerves, neuromuscular junction, or muscles. Observations that help the clinician are listed in Box 5-1.

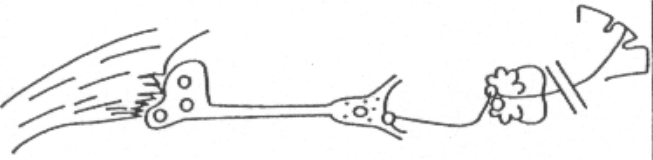
Box 5-1

1. *Location of weakness:* Is the weakness distal or proximal, symmetrical or asymmetrical? If asymmetrical, does it involve a group of muscles innervated by one nerve, several nerves, or a plexus? Does it involve ocular or bulbar muscles? Does it involve breathing muscles?
2. *Muscle atrophy:* Is wasting of muscles present? In what distribution: a nerve, root or plexus distribution? Is it distal, proximal, or generalized? Is it an early or late event in the disease? Are fasciculations present? If there is hypertrophy, is it localized to certain muscles such as the calf, or is it generalized?
3. *Stretch reflex:* Are the reflexes normal, hyperactive, or hypoactive? Are they symmetrical? Are pathologic reflexes present, such as Babinski sign?
4. *Sensory alterations:* Are there unilateral or bilateral sensory deficits, dermatomal or stocking-and-glove losses of sensation?
5. *Muscle tone:* Is it normal, increased (rigidity, spasticity), or decreased (hypotonia)?
6. *Myotonia:* When the muscles are contracted or percussed, can the muscle relax easily and fast, or do they relax slowly but without pain as in myotonia? Or do the muscles relax slowly and painfully as in muscle cramps? Are the cramps spontaneous, or are they induced by exercise or ischemia?

In Table 5-1, lesions at any level of the corticospinal tract (upper motor neuron) can produce paralysis or weakness (paresis) of large groups of muscles, increase in muscle tone (spasticity), hyperactive stretch reflexes, and pathological reflexes, including extensor-plantar response (Babinski sign).

TABLE 5-1 Neuromuscular Diseases

Level of Lesion	Site of Weakness	Muscle Atrophy	Stretch Reflexes	Sensation	Tone
Corticospinal tract	Large group of muscles; one extremity (monoplegia) or two (diplegia); if the arm and leg are paralyzed on the same side, this is hemiplegia; if both legs are paralyzed, this is paraplegia	Mild, disuse can develop	Hyperactive	Normal	Spastic
Anterior horn cell	Variable, proximal, and/or distal; small group of muscles	Severe, early; fasciculations muscles	Hypoactive or absent	Normal	Flaccid
Peripheral nerve	Distal extremities (feet, hands) are initially involved, and this can extend proximally	Mild to severe, early	Hypoactive or absent	Abnormal in sensory or mixed neuro-pathies	Flaccid
Neuromuscular junction	Fatigability; ocular, bulbar, or generalized	Minimal, late	Normal	Normal	Normal
Muscle	Proximal involvement	Late, skeletal abnormalities	Normal early, absent late	Normal	Normal or myotonia



All of these findings usually appear 1 to 3 weeks after acute onset of a destructive lesion, as in an infarct of brain or spinal cord vascular lesion, or evolve slowly, as in mass or degenerative lesions. Anterior horn cell lesions (lower motor neuron) tend to cause severe weakness and early atrophy of individual muscles or muscle groups, fasciculation, flaccidity, and loss of stretch reflexes but no pathologic reflexes. Lesions in peripheral motor nerves cause weakness in the muscles supplied by the nerve. In polyneuropathies, weakness usually begins distally in the extremities (hands and feet), can "ascend," and be associated with atrophy, flaccidity, and loss of stretch reflexes. Pathologic reflexes are not part of the syndrome. Sensory deficits can be present if sensory nerves are also affected (sensory neuropathy or mixed sensorimotor neuropathies). Diseases affecting the neuromuscular junction are manifested by fatigability (weakness after exercise which is relieved by rest) or weakness during exertion, variability of weakness that worsens during the day and improves with rest, minimal or no atrophy, no change in muscle tone or reflexes, and no sensory deficit (symptoms and signs of objective quantifiable fatigue). Primary diseases of muscle (myopathies) usually cause proximal muscle weakness with late atrophy, no changes of stretch reflexes except very late in the disease (areflexia caused by muscle atrophy or joint contractures), and no change in sensory functions. Myotonia or painless delay in relaxation of muscles after contraction occurs in some myopathies. Myotonia is frequently associated with weakness and atrophy of muscles in myotonic dystrophy and with hypertrophy in myotonia congenita.

When a patient complains of weakness, detailed inquiry into the symptoms should be made. By definition, weakness is a diminution of strength. Some patients complain of weakness when they are experiencing fatigability, numbness, or problems of balance or coordination. It is important to inquire about the onset of weakness, its progression or regression, its distribution, and any associated symptoms, such as cramps. A differentiation should be made between weakness as defined here and fatigability as defined by weariness during exercise. Objective ("pathologic") fatigability is seen in diseases that affect the neuromuscular junction. The fatigue should diminish with rest. Fatigue is also seen with other disorders that affect the motor system, for example, anterior horn cell disease, peripheral nerve disease, muscle disease, and upper motor nerve disease. The fatigue is most prominent after activity. If the patient reports fatigue after awakening or immediately after initiating activity, this suggests a psychogenic cause of the fatigue (neurasthenia). On describing the symptoms, the patient frequently localizes the site of the weakness. Patients with *hip weakness* complain of difficulty getting up from a sitting (chair) or squatting position; they also complain of having difficulty going up stairs and mention that they have to pull themselves up by pulling with their hands or nearby objects or pushing on their knees. *Weakness at the ankles* frequently affects dorsiflexion and produces footdrop. Patients complain of stubbing their toes, tripping, and twisting their ankles. *Weakness of the shoulder muscles* is described by patients as difficulty in combing their hair or difficulty with any activity that requires raising the arms above the head. Shoulder girdle weakness produces forward and lateral displacement of the scapulae with compensatory pronation of the arms (palms facing backward) when the arms are down. Patients with *weakness of the hand muscles* have difficulty turning door knobs, unscrewing caps of jars or bottles, or holding objects in their hands. Eating or writing becomes quite laborious. *Weak finger muscles* produce difficulty pinching, buttoning and unbuttoning, and using zippers. In *neck muscle weakness*, patients have difficulty lifting their heads when in a supine position, indicating involvement of the neck flexors. Rarely are the neck extensors primarily affected. Patients with *weakness of the facial muscles* have difficulty drinking through a straw, whistling, or blowing. Difficulty in chewing or closing the mouth

indicates weakness of the masseter and temporalis muscles. Difficulty in articulating or in removing food particles from between and around the teeth with the tongue can suggest weakness of tongue muscles. Diplopia can indicate weakness of extraocular muscles. Blurred vision can be caused by double vision, and this should be clarified during the interview.

Respiratory insufficiency caused by intercostal or diaphragmatic muscle weakness is a later and usually terminal event in neuromuscular diseases, is manifested by abdominal breathing, and should be differentiated from the different breathing patterns seen in central nervous system lesions. Respiratory insufficiency can be the first manifestation of some myopathies such as adult Pompe's disease (glycogenosis type II).

GAIT DISTURBANCES

Careful observation of the patient's gait as he or she walks into the examining room frequently can reveal what muscles are affected and where the lesion is even before any questions are asked.

Although impairment of gait can be caused by mechanical factors, such as diseases of bones, tendons, joints, or muscles; lesions at different levels of the nervous system are very important causes of gait abnormalities. Patients should be observed as they walk and then be asked to walk with their eyes closed, backward, around objects, on their toes, on their heels, and in tandem fashion (placing one heel directly in front of the other foot). Different patterns of gait develop when different muscles are affected, indicating different levels of lesions that can be considered distal to central involvement.

Steppage or Equine Gait

Steppage or equine gait results from weakness of the peroneal and anterior tibial muscles and is characterized by a slapping quality produced when the knee and hip have to be flexed and lifted to compensate for lack of foot dorsiflexion (foot drop). The slapping noise is produced when the anterior end of the foot has to be thrown upward to avoid tripping or twisting the ankle. This type of gait is seen in patients with ankle weakness that produces dropfeet, such as in the distal peripheral neuropathy of lower extremities, pressure or stretch injuries of peroneal nerves, and more rarely in distal dystrophic processes. Patients with this type of weakness are unable to walk on their heels when asked.

"Back-Kneeing"

Weakness of the thigh muscles can result in poor stabilization of the knee with resultant hyperextension of that joint (so-called back-kneeing) causing a peculiar gait. This gait is seen with some of the femoral neuropathies and with any other cause of quadriceps muscle weakness.

Waddling Gait

Waddling gait is a result of hip girdle muscle weakness. The lack of fixation of the hip by the weak gluteal muscles on taking a step makes the opposite side of the pelvis drop, producing compensatory lateral movements of the pelvis. Very frequently the hip and back extensors are concomitantly affected, which explains the marked lumbar lordosis with protrusion of the abdomen seen while the patient stands or walks. This type of gait is seen in all myopathies that affect the hip girdle musculature, such as in patients with most dystrophic myopathies (limb-girdle, Duchenne, Becker, and so forth), in the intermediate form of spinal muscular atrophy

(Kugelberg-Welander type), and inflammatory neuropathies (dermatomyositis polymyositis), and in hip dislocation.

Sensory Ataxic Gait

Sensory ataxic gait is a result of lesions in the posterior column of the spinal cord, posterior nerve roots, dorsal root ganglion, or peripheral nerves and is associated with loss of sensation in a variable distribution according to where the lesion is. The muscle strength is normal, and, if there is some weakness, it is out of proportion to the sensory deficit. In predominantly dorsal spinal cord disease there is loss of position and vibratory sense in the lower extremities and a positive Romberg sign (the patient falls or sways when standing with feet together and eyes closed, but not with eyes open). The inability to sense the positions of the joints in space and in relationship to themselves makes the patients produce an unsteady broad-based gait to correct the instability and causes the hard slapping character of the steps. The patient has to use his or her sight to maintain these spatial relationships and consequently has more difficulty walking in the dark or with the eyes closed. This type of gait is seen in patients with tabes dorsalis, subacute combined degeneration, Friedreich's ataxia, and some types of spinocerebellar ataxias (SCA1 and SCA2). In sensory neuropathies there is distal loss of all modalities of sensation and severe distal limb ataxia and truncal ataxia.

Cerebellar Ataxic Gait

Cerebellar ataxic gait is produced by cerebellar lesions, either focal, as in midline (vermal) lesions of chronic alcoholism, multiple sclerosis, and neoplasms, or diffuse (vermis plus both hemispheres), as in the various types of diffuse cerebellar atrophy, such as paraneoplastic immune disorders, degenerative diseases, and intoxications (mercury). In truncal ataxia, as seen in midline cerebellar neoplasms, and in chronic alcoholics with anterior (dorsal) vermal degeneration, the patient walks in a broad-based unsteady gait with short but somewhat regular steps and forward inclination of the trunk. In vermal cerebellar atrophy, usually due to alcohol, the upper extremities are not usually involved. In these patients, the upper limb movements are generally well coordinated, and the speech is not affected. Pancerebellar (involving both the midline vermis and lateral cerebellar hemispheres) disease and multiple sclerosis produce a very unsteady gait with irregular short or large steps, lateral or vertical truncal swaying, and severe incoordination of upper extremities frequently associated with slurred speech. Standing with feet close together results in marked instability, even with the eyes open. Patients are unable to walk tandem gait (heel-toe in a straight line).

Dyskinetic Gait

In dyskinesias, which are diseases that affect the extrapyramidal system, such as the choreas and dystonias, the gait exacerbates the abnormal involuntary movements and can result in jerky movements of one or several extremities or grotesque writhing movements of all four extremities, face, neck, and pelvis. The dystonic gait is slow and deliberate because of inversion of the feet or pelvic distortion (see Chapter 20).

Parkinsonian (Festinating) Gait

Parkinsonian gait is produced by nigrostriatal lesions as seen typically in Parkinson's disease and

is characterized by slowness in initiating movement and a shuffling character with a tendency to accelerate (festination). The steps are short and irregular (*marche à petits pas*). The patients have a characteristic forward displacement of their trunks and have difficulty stopping suddenly. Any change in the direction of the gait is difficult and laborious.

"Senile" Gait

Gait impairment and falls are frequent in the elderly population, worsen with age, and are more frequent in females than males. There is a significant correlation between hypodensity of the white matter on brain CT and MRI scans and both gait disturbance and balance disorders in the elderly. However, there is more likely a multifactorial cause, including vision and hearing deficits, decreased neuromuscular function caused by peripheral nerve and central nervous system deficits, and arthritis. The gait is characterized by a truncal and head forward flexion, diminished arm swing, and increased flexion of both knees and elbows. There is decreased walking speed and stride length.

Spastic Gait

Spastic gait is due to lesions of the corticospinal tracts and can be a unilateral hemiplegic spastic gait or a bilateral paraplegic spastic scissoring gait. Spastic hemiparesis is frequently the result of a lesion affecting one corticospinal tract. The leg is overextended at the hip and knee, and the foot is plantar flexed. The patient's inability to flex the hips and knees forces the leg to circumduct with each step (i.e., the leg is dragged in a semicircular movement external to the pelvis). The leg is circumducted and the upper extremity is held in an adducted and flexed posture. The upper extremity is immobile, held in a posture characterized by flexion of the shoulder, elbow, wrist, and fingers with adduction at the shoulder. In spastic paraparesis both legs are affected. The leg features described in the hemiparetic gait are similar. There can be shortening of the Achilles tendon and lumbar lordosis. The gait is slow, staggering, stiff, and shuffling with short steps, producing a scissoring quality to the gait.

Abnormal Gait in Frontal Lobe Disorders

Abnormal gait in frontal lobe disorders, such as normal pressure hydrocephalus or frontal lobe infarcts, is quite elaborate, very laborious, severely imbalanced, and frequently difficult to predict and describe. It has magnetic quality as if the feet were glued to the ground. The legs move easily and rapidly if the patient is lying in bed with the feet off the ground. There is no associated weakness of the legs.

Hysterical Gait

Hysterical gait, associated with no lesion of the neuromuscular system, has bizarre, nondescript qualities with an unpredictable changing character. At times normal movements can be detected along with abnormal ones. Patients may be able to run or walk backward or to the side without difficulty or may be unable either to stand or walk (astasia-abasia). There is no muscle weakness or abnormal tone and reflex examination is normal.

EXAMINATION

After observing the patient's ordinary routine gait, observing the performance of simple daily necessary activities will also help localize the area of weakness. Patients with hip-girdle weakness will have difficulty arising from a sitting position and will need to use their hands to push themselves up. These patients will place their hands on their knees or thighs to push themselves, push with their hands on the sides of a chair, or actually pull themselves with their hands by grabbing a static object. When back extensor muscles are also weak, the patient will get up from a chair and remain with the trunk flexed over the hips. He or she then has to get into an erect truncal position by using both hands to "climb up his or her thighs" as described by Gowers. Such patients will prefer to use a high stool at home so that arising from the sitting position is easier. Arising from the floor is a very elaborate and slow maneuver for patients with proximal hip-girdle weakness, and observing a patient during this procedure will provide information that can obviate complicated muscle strength testing, which can be particularly difficult in children. Such patients tend to break the movement into parts by first extending the knees in a sitting position, then rotating the trunk to place their hands on the floor, then flexing the knees and rising to an erect position by pushing themselves with their hands in an alternating displacement of the hands from leg to knee to thigh (Gowers' sign). This complicated maneuver can be modified according to the patient's degree of weakness. Observing a patient going up stairs or stepping into a stool will demonstrate mild hip weakness if the patient uses his or her hands on the knees or lower thighs to push himself or herself up. The patient may need to use handrails to pull himself or herself upstairs.

Manual Muscle Testing: Because of normal variations from person to person and differences in age, all muscle testing and grading are subjective. We prefer to describe the patient's ability to perform tasks, such as the ability to dress oneself, to eat, to brush one's teeth, to groom oneself, and so on. When using a grading system, we prefer the one recommended by the British Medical Research Council (see Box 5-2)

Box 5-2

- 1+ Flicker or trace of contraction
 - 2+ Active movement with gravity eliminated
 - 3+ Active movement against gravity
 - 4+ Active movement against gravity and resistance
 - 5+ Normal power
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After or during the observation of gait, the observation of a few simple tasks, and manual muscle testing, the examination of the neuromuscular system should include simple *inspection* of the muscle bulk. In neuromuscular diseases the muscles can be wasted or atrophic or they can be enlarged because of true hypertrophy or pseudohypertrophy (hypertrophy of some muscle fiber with infiltration of muscle by fat and fibrous tissue). During inspection, look for fasciculations, which are easily seen in thin persons but can be masked by subcutaneous fatty tissue in infants and obese patients. An accessible site to find fasciculation is the tongue. The tongue should be in a resting position within the mouth to avoid confusion with tremor. The irregular jumping of isolated muscle frequently associated with atrophy should be differentiated from the rhythmical generalized movements seen in tongue tremor. Facial inspection will detect ptosis, which is frequently compensated for by corrugating the forehead or tilting the head

backward. Atrophy of the temporalis and masseter muscles causes a cadaveric appearance of the face that frequently is associated with prominent puckered lips and tapiroid mouth caused by facial muscle weakness. Inability to close the eyelids and upward deviation of the eyeballs when trying to close the eyelids (Bell's phenomenon) indicate nuclear or infranuclear (peripheral) seventh nerve palsy if associated with flattening of the nasolabial fold and lack of facial expression. Sparing the eyelids and forehead in facial weakness indicates supranuclear (central) seventh nerve palsy, except in some myopathic processes (facioscapulohumeral dystrophy). Shoulder muscle atrophy makes the bony prominence more prominent and causes winging of the scapulae. The scapula deviates laterally, upward, and backward when the patient pushes his or her hand against the wall or when the arms are stretched in front of the chest. Shoulder weakness also will produce internal rotation of the arm in such a way that the dorsal side of the hand faces forward. Atrophy of the intrinsic hand muscles will cause a guttering appearance between the metatarsal bones. A "knife edge" configuration of the tibia indicates atrophy of the peroneal innervated anterior tibial muscles. An exaggerated variation of leg muscle atrophy with sparing of the thigh muscles results in the "stork leg" appearance or "inverted champagne bottle" sign.

Palpation of the muscles can aid in determining whether the muscles are of normal consistency or whether they are rubbery as in usually seen in the pseudohypertrophic calves of patients with Duchenne muscular dystrophy. Muscle nodules can be palpable in some cases of myositis. Subcutaneous calcifications can be palpable in the infantile form of dermatomyositis. Palpation of muscles can be painful in myositis, polyarteritis nodosa, poliomyelitis, and polyneuritis.

Percussion of muscles can help the examiner detect myotonia and myoedema. Percussion of normal muscles produces a sudden, brief contraction followed by a quick relaxation of the muscle. In myoedema the muscle contracts and produces a localized but electrically silent bulge. In myotonia the entire muscle contracts in a sustained involuntary fashion, then slowly relaxes and has electrical changes.

Skeletal abnormalities are frequently associated with hereditary, degenerative neuromuscular problems and include lumbar lordosis, kyphoscoliosis, pes cavus, and pectus excavatum.

SUMMARY

Gait can be impaired by nonneurologic disorders such as orthopedic, rheumatologic, vascular, or neurologic disorders (weakness of upper or lower motor neuron type, cerebellar, proprioceptive, vestibular, or basal ganglia disorders, normal pressure hydrocephalus, frontal lobe lesions). By examining the patient, the location of the neurological disturbance causing the gait impairment can be ascertained. This includes assessing base and station and the ability to arise from a chair; tandem walk; balance on either leg for 10 seconds with feet close together, stand with eyes open, and then, with eyes closed, walk on heels and toes; and walk forward and backward and around a chair. When the gait disorder is due to weakness, the pattern of weakness and accompanying motor, reflex, or sensory disturbances can determine the level of abnormality in the neuro-axis. This includes the corticospinal tract, anterior horn cell, spinal nerve root, peripheral nerve, neuromuscular junction, and muscle. By clinically determining the cause of the gait disorder or weakness the most cost-effective evaluation for each patient can be determined.

Suggested Readings

Weakness

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