Gait, Cerebellar Function, and Movement Disorders (Dr. Merchut)

Gait

1. Essentials for normal walking

Obviously, **strength** in the lower limbs and trunk is required to walk, which is dependent on functioning upper and lower motor neurons, neuromuscular junctions and muscles. The cerebellar and extrapyramidal systems help provide the **coordination** and **postural control** of gait. Intact **sensation**, especially proprioception, is needed to walk safely in the dark. The **memory or concept of walking** is required to execute or perform this learned motor activity "on command." Patients with gait apraxia have adequate strength, coordination, postural control, and sensation to walk, but are immobile when asked to walk.

Standing up is the first action prior to actual walking. Patients with significant weakness, dizziness, or pain may be unable to stand up on their own. In other cases, the **Romberg sign** is present, where a patient can stand steadily with the feet together, but sways and breaks stance or topples if the eyes are closed. The Romberg sign suggests a problem with impaired proprioception, either from involvement of the posterior or dorsal column pathways or their afferent sensory nerves. It should be noted however that elderly patients may readily sway or lose balance when standing with closed eyes, often due to dizziness or other factors, and the Romberg sign may not truly be present. Other patients may stand up, but spread the feet apart widely to keep balance, swaying or falling over if the feet are placed together. This finding suggests a cerebellar problem since intact vision cannot compensate for this type of imbalance.

2. Types of gait abnormalities

In the **broad-based ataxic gait** (Fig. 1A), the feet are spread apart for greater stability when standing or walking. Unsteadiness worsens when the patient tries to walk tandem "on a straight line" or "heel to toe." This may occur with lesions of the posterior columns or proprioceptive sensory nerves, where it is worse with eyes closed, or may be due to cerebellar dysfunction. The **hemiplegic gait** (Fig. 1B) is often seen with stroke patients. The affected lower limb is stiffly extended and swung or circumducted when walking while the ipsilateral upper limb is flexed at the elbow and wrist with decreased armswing. The **tabetic gait** occurs with tabes dorsalis from neurosyphilis, and has a "foot slapping" characteristic. The patient compensates for impaired sensation in the feet by forcibly planting the feet down to "feel" the floor. **Steppage gait** (Fig. 1C) occurs in a patient with foot drop or weak dorsiflexion of the foot. To prevent tripping over the toes when walking, the hip is flexed or pulled up even higher to elevate the drooping foot, which is then lowered to the floor toe first. Unilateral foot drop may occur from a lesion of the peroneal nerve or L5 root. Bilateral foot drop may be seen with severe polyneuropathy, motor neuron disease or bilateral L5 root lesions.

The **duck waddle or waddling gait** occurs from weakness of the hip girdle muscles, usually seen in muscle disease (myopathy). Hip and pelvic muscles support the weight of the patient "on one leg" when the other leg is elevated while walking. The

patient may topple over if these muscles are weak. To compensate for this, the patient leans or bends the trunk to the left as the right foot is raised, and vice versa when lifting up the left foot, alternately tilting the pelvis and hips side to side like a walking duck. In the **scissors gait** (Fig. 1D), the advancing leg or foot tends to cross over the opposite lower limb, similar to the closing blades of a scissors. This usually occurs from upper motor neuron (corticospinal tract) lesions affecting the lower limbs, as in spastic paraparesis. The increased spastic tone and tightness in the adductor muscles of the thighs tends to force the lower limbs together when walking. The **parkinsonian gait** (Fig. 1E) is slow and shuffling, with decreased armswing and a "stooped forward" or "bent over" posture. Patients with Parkinson's disease may exhibit festination of gait, having to lean forward in order to walk, followed by uncontrollable running to "catch up" with the center of gravity.

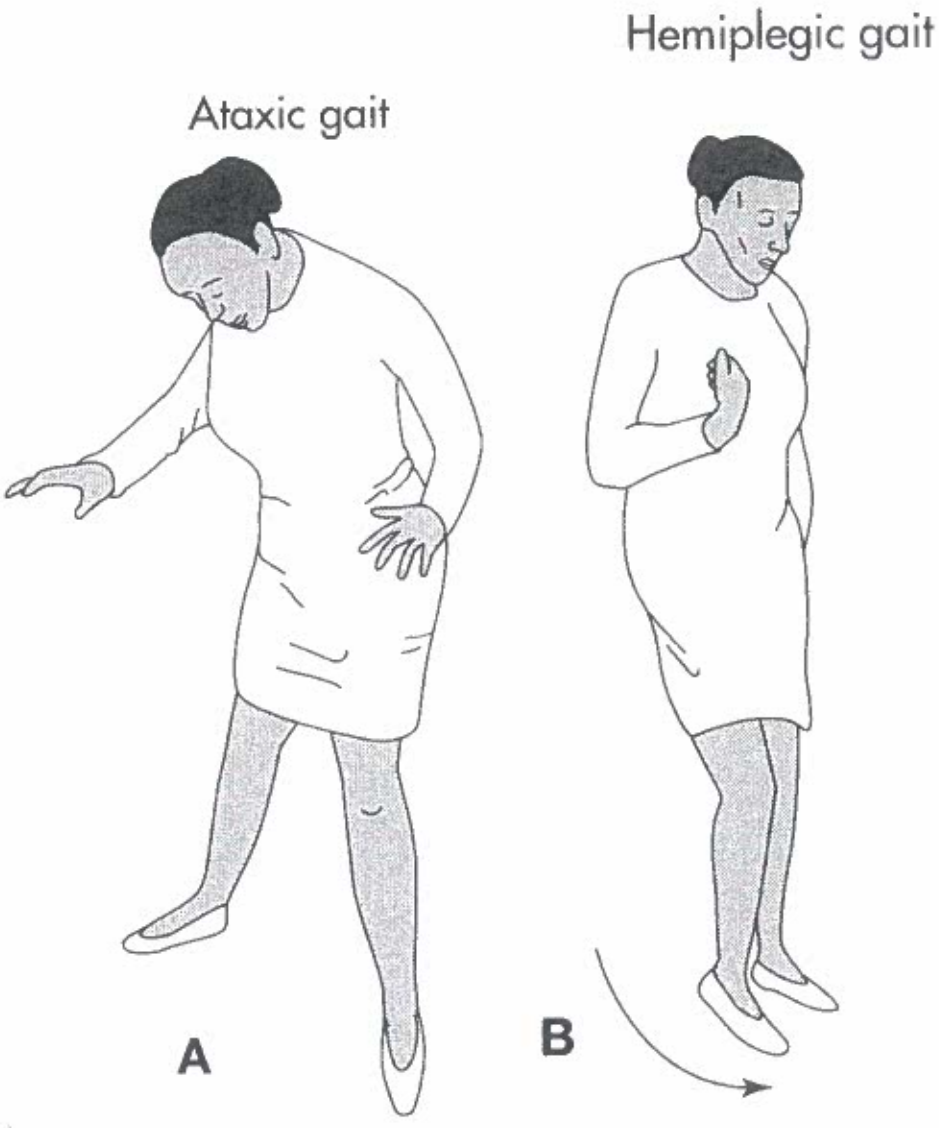

Fig. 1 Gait abnormalities

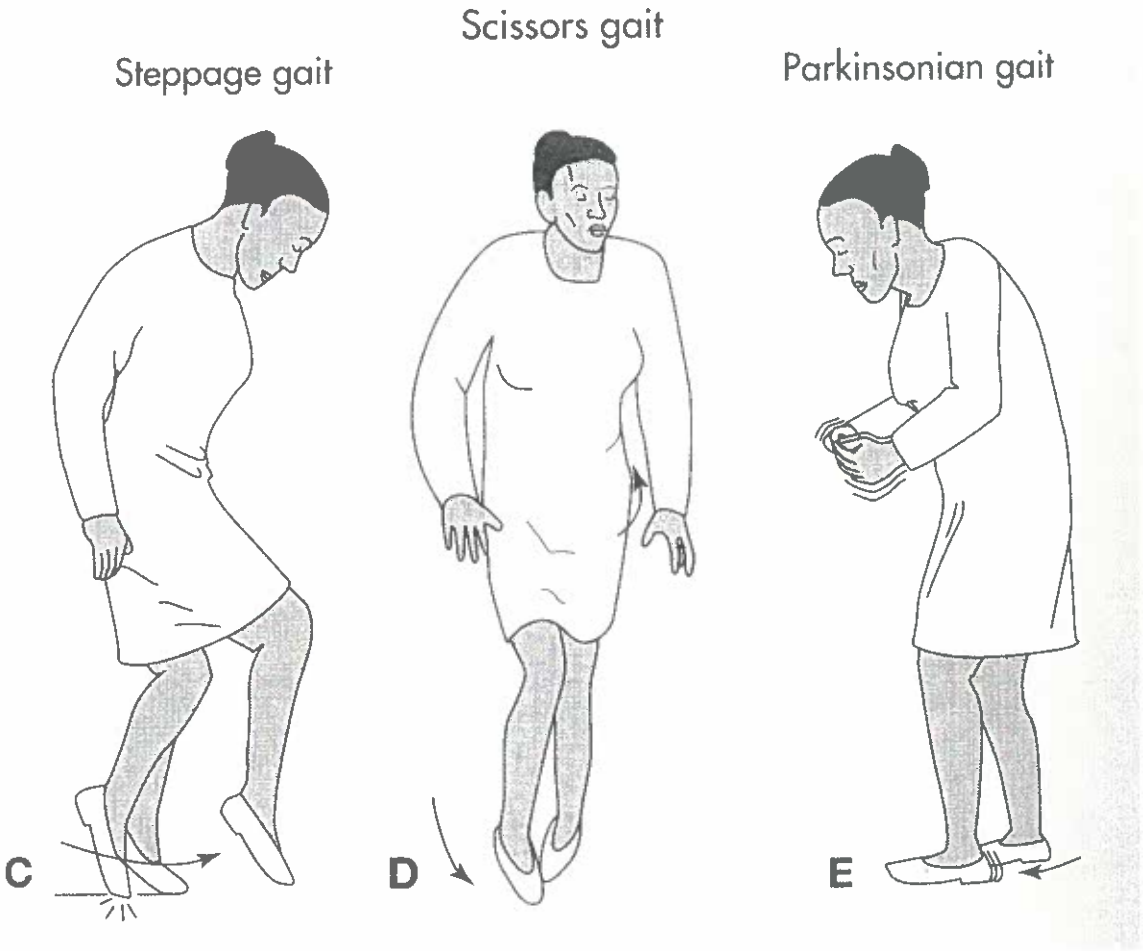

Fig. 1 (continued) Gait abnormalities

Cerebellar Function

1. Clinical cerebellar deficits

The cerebellum helps maintain the smoothness and precision of movements for the limbs, trunk, eyes, and voice. Deficits may be obvious by merely observing the patient or demonstrated by various bedside neurological tests. The **finger-nose-finger** test consists of the patient using the index finger to alternately touch his or her own nose and then the extended finger of the examiner (Fig. 2A). In the **heel-shin-knee** test (Fig. 2B), the supine patient places the heel on the opposite knee and slides it up and down the shin. Jerky, "broken down," imprecise, or off-target movements are typical of cerebellar deficits in the absence of significant weakness. While performing these tests, a patient may show a **kinetic or action tremor** consisting of rhythmic oscillations of the hand or foot while it is moved. **Dysmetria** is the term used to describe the overshooting or undershooting of the target by the hand or foot. **Rapid alternating movements** are also tested, such as a patient quickly slapping his or her own knee (or contralateral palm) with

alternating pronation and supination of the hand (Fig.2C). Cerebellar dysfunction may create uncoordinated, nonrhythmic, sloppy hand movements termed as **dysdiadochokinesia**. A **rebound phenomenon** or abnormal "check reflex" may be found in an upper limb with cerebellar deficits creating an imbalance between agonist and antagonist muscles (Fig. 2D). The patient is asked to contract the biceps muscle against the examiner's efforts. If the examiner suddenly "lets go," normally the patient's triceps (antagonistic muscle) should reflexively contract to "check" or stop the unopposed elbow flexion by the biceps. In the presence of cerebellar disease, the persisting elbow flexion may cause the patient to strike his or her chest or face unless protected by the examiner.

Cerebellar disorders may also cause a characteristic type of slurred speech or **cerebellar dysarthria**, most often associated with involvement of the left cerebellar hemisphere. Here, the speech is less distinct, "thick," erratic, jerky, or explosive. Syllables are often "broken down" or "hyphenated" with nonrhythmic or unequal emphasis or force. Eye movements may appear jerky or erratic with cerebellar disease, sometimes exhibiting multidirectional **nystagmus** (which may also occur with lesions of the vestibular system and brain stem).

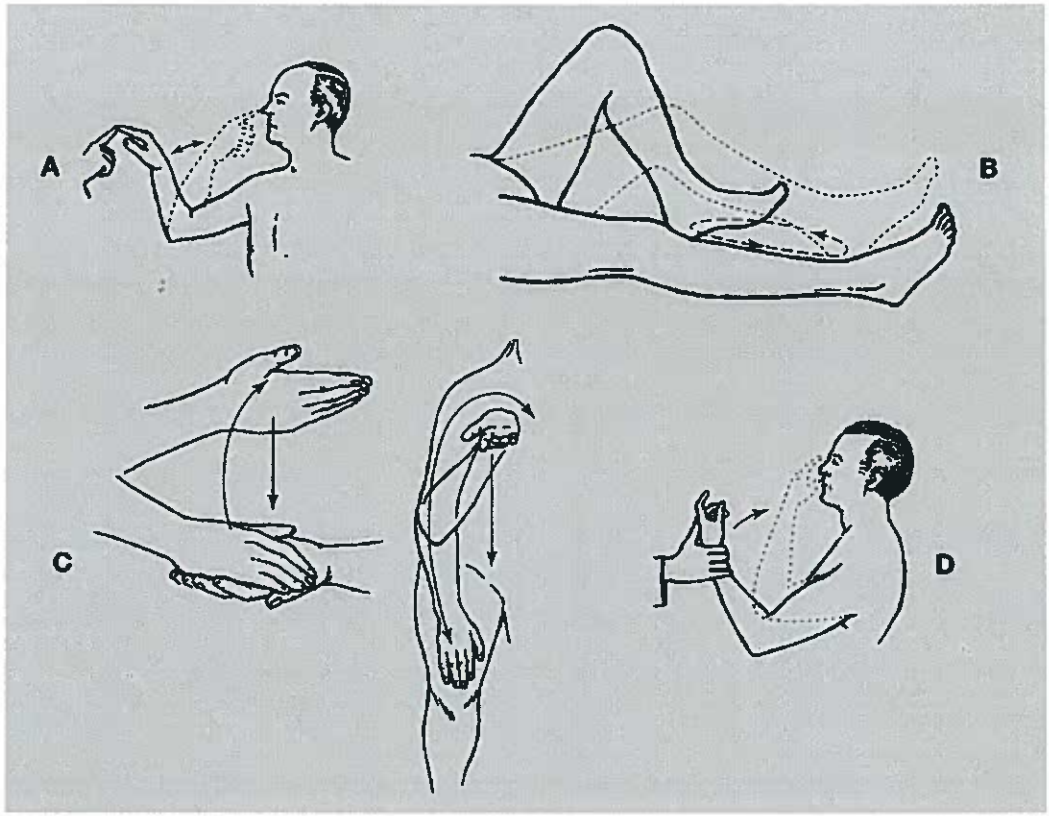

Fig. 2

Limb movement tests. A, Finger-nose-finger; B, heel-shin-knee; C, rapid alternation; D, rebound. (From Lindsay KW, Bone I, Callander R: *Neurology and neurosurgery illustrated*, ed 2, New York, 1991, Churchill Livingstone; Swartz MH: *Textbook of physical diagnosis*, New York, 1989, WB Saunders.)

2. Cerebellar syndromes

A lesion of a cerebellar hemisphere predominantly affects the ipsilateral limbs, causing kinetic tremor, limb dysmetria, dysdiadochokinesia, and rebound phenomenon. Common examples of unilateral cerebellar hemispheral lesions include ischemic infarction, hemorrhage, tumor, and multiple sclerosis. Bilateral involvement of the cerebellar hemispheres may be seen with various degenerative or toxic diseases. **A midline lesion of the cerebellar vermis predominantly affects the trunk**, causing truncal unsteadiness while standing or walking, with impaired balance and gait ataxia. Common causes include hemorrhage, tumor, multiple sclerosis, and degenerative or toxic disorders. **Alcoholic cerebellar degeneration** is an example of the latter, where chronic alcoholism leads to atrophy of the anterior-superior vermis, where the trunk and lower limbs are represented (Fig. 3). Deficits here include gait ataxia, truncal unsteadiness, and lower limb dysmetria.

Other degenerative cerebellar diseases are the **spinocerebellar degenerations or ataxias**, a group of several hereditary disorders with unknown cause and no curative treatment. Specific nuclei and tracts of the spinal cord and cerebellum are affected in progressive fashion, such that older patients become wheelchair-dependent. The most common type is **Friedreich's ataxia**, an autosomal-recessive disorder which begins in school-age children and gradually worsens. Although the cerebellum is affected, most signs and symptoms here are related to lesions in the dorsal or posterior spinal cord. Spinocerebellar tract lesions, with patchy loss of cerebellar Purkinje cells, lead to limb dysmetria, gait ataxia, and dysarthria. Corticospinal tract lesions produce weakness and Babinski signs. Lesions of the dorsal root ganglia and dorsal or posterior columns initially affect the lower limbs, causing loss of vibration, position sense, and absent reflexes. Non-neurological features include scoliosis, pes cavus (high-arched feet), cardiac hypertrophy, and potentially fatal cardiac arrhythmias. The clinical diagnosis is confirmed by a blood test revealing multiple trinucleotide repeats from a defect in chromosome 9.

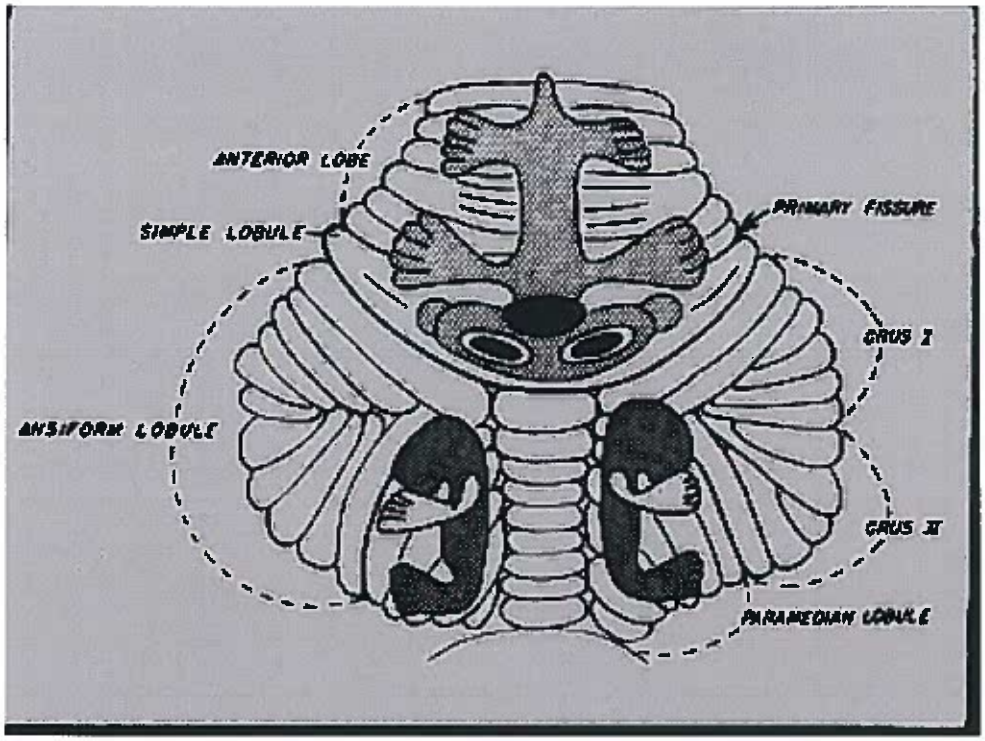

Fig. 3 Cerebellar afferents mapped out in an animal model, with ipsilateral limbs represented in each hemisphere and the trunk represented in the vermis.

Movement Disorders (Hyperkinesias)

1. Types of hyperkinesia

Hyperkinesias are spontaneous, involuntary movements with characteristic clinical features. Some of these abnormal movements are associated with specific anatomical lesions or disorders while the underlying lesion or etiology of other movement types is unknown. **Tremor** is a spontaneous, rhythmic, oscillatory movement of hands, limbs, head, or voice, and is fairly common. A **resting tremor** of the limbs or head, primarily noticeable during restful sitting or reclining, is typical of **Parkinson's disease**. A **postural tremor** is more obvious when the limbs are maintained in various positions, such as holding an object or extending an arm or leg. Postural (and kinetic) tremor without other neurological signs or symptoms is often due to **familial essential tremor,** the lesion and etiology of which remains unknown**.** The voice and head may also be affected in essential tremor. **Kinetic tremor** primarily occurs in a limb moving towards a target or performing a task. It may accompany other signs and symptoms of **cerebellar disease**.

Athetosis is the term for slow, writhing, fairly continuous movements of the distal limbs. It often coexists and blends in with chorea, which consists of purposeless, random, nonrhythmic movements of the limbs, face, neck, and trunk. **Choreoathetosis** is the combination of these brief, irregular movements that flow together in dancelike fashion (the Greek *choreía* means "dance"). Choreoathetosis patients (Fig. 4) appear

somewhat restless, fidgety and "antsy." This abnormal movement is produced by lesions in the **caudate nucleus or its connecting pathways**. A well-known example is Huntington's disease, which is further discussed in "Disorders of the Basal Ganglia."

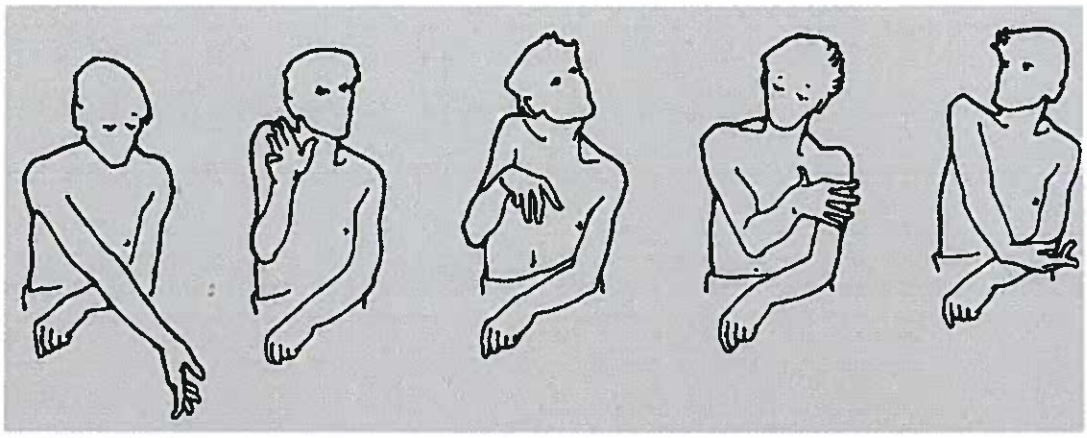

Fig. 4

Choreoathetosis. Sequential abnormal movements of the right arm and neck. (From Mumenthaler M *Neurologic differential diagnosis*, New York, 1992, Thieme Medical.)

Hemiballismus consists of rapid, violent, flinging movements of the proximal limbs on one side. (The Greek *bállein* means "throw" and is the root for "ballistics," the study of projectile movements, which aptly describes the "throwing" characteristics of this hyperkinesia.) A lesion in the contralateral subthalamic nucleus, usually an ischemic infarction, causes hemiballismus, which may improve or disappear over time. **Dystonia** is the continual, sustained, often painful contraction of muscles leading to spasms, turning, and twisting of the limbs, neck, head, or trunk into unnatural positions or fairly fixed postures. It can be **focal**, confined to muscles in the neck or shoulder (cervical dystonia or torticollis) or it may be a **generalized** dystonia which is often hereditary and progressively disabling. Its anatomical substrate or lesion is uncertain.

Tics (not to be confused with "ticks," which are blood-sucking insects) are brief, stereotyped, often repetitive, focal muscle contractions that appear semipurposeful, such as an eyeblink, facial twitch, or sniff. After the patient tries suppressing these movements for a period of time, there may be an irresistible urge or "need" for the tics to occur. Such motor tics occur with vocal tics (grunts, growls, or vocalizations) in **Tourette's syndrome**, a hereditary condition more commonly seen in boys and often accompanied by attention deficit and behavioral disorders. It has been suggested that decreased motor inhibition in the basal ganglia causes tics, but the specific anatomical lesion or pathology underlying Tourette's syndrome is still unclear.

A diffuse encephalopathy occurs when a toxic, metabolic, infectious, or inflammatory disorder affects the brain as a whole. Systemic illnesses such as renal failure or liver dysfunction thus indirectly impair several functions of the brain, which are further discussed in "Behavior, Cortical Function, and Alzheimer's Disease." Movement disorders which accompany encephalopathy include myoclonus (myoclonic jerks) and asterixis. **Myoclonus** refers to the rapid, shocklike, lightning movements or jerks of the

limbs and trunk, which is usually bilateral but asynchronous and irregular. It may also be observed in patients with Creutzfeldt-Jakob disease, which is discussed later. **Asterixis** is the semirhythmic loss of postural control of hands and feet, so that the extended hands or feet appear to have a "flapping tremor." This movement resembles bouncing a ball or tapping the foot to music. When occurring unilaterally it is due to structural brain disease such as an ischemic infarction.

2. Pharmacotherapy for hyperkinesias

Some of these medications were found helpful in reducing or lessening these abnormal movements by coincidence, while other drugs were the result of directed research. The **resting tremor** of Parkinson's disease is treated with **anticholinergic drugs** if it is more of an isolated or predominant symptom, while **levodopa and dopamine agonist drugs** are used if other troubling parkinsonian symptoms accompany the resting tremor. **Essential tremor** may improve with **beta-adrenergic blocker drugs** or **barbiturates**. **Choreoathetosis, hemiballismus, and tics** may improve with **dopamine antagonist** drugs. **Dystonia** is treated with **anticholinergic drugs, benzodiazepines and botulinum toxin injections** into the affected muscles. For medically refractory, severe cases, advanced neurosurgical techniques now allow the implantation of "deep brain stimulators" to inhibit areas in the thalamus and subthalamic nucleus to help patients with essential tremor and Parkinson's disease, respectively.