69 The Cerebellum

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Definition

The principal signs of cerebellar dysfunction are the following:

Ataxia: unsteadiness or incoordination of limbs, posture, and gait. A disorder of the control of force and timing of movements leading to abnormalities of speed, range, rhythm, starting, and stopping.

Hypotonia: normal resting muscle tension is reduced, leading to decreased muscle tone and abnormal positions of parts of the body.

Tremor: an intention tremor of the hand on purposive movement is the most common, with coarse, rapid, side-to-side oscillations that increase as the movement goal is approached. Resting tremors of the limbs, head, and trunk can occur. At times, paroxysms of these tremors are severe enough to shake the entire bed and delude the unwary physician into suspecting seizure activity.

Gait: the station or manner of standing is abnormal; the legs are apart and there is swaying of the body. The patient staggers, reels, and lurches on walking.

Ocular motor abnormalities: saccadic dysmetria, impaired smooth tracking, fixation abnormalities, and various forms of nystagmus (see below).

Technique

Have the patient sit on the side of a bed or table. Note the position of the limbs; hypotonia can produce bizarre positions. The head can be deviated to one side. Look for resting tremors of the limbs and trunk. At times, these can be of such severity to cause the head to bob, or titubate.

Head

Observe the eyes in the primary position for nystagmus or problems with fixation. Observe for nystagmus as the patient follows a test object (such as the examiner's fingers) in the six diagnostic positions of gaze (see Chapter 60, Cranial Nerves III, IV, and VI). Ocular dysmetria is seen best when the patient on command moves the eyes from an eccentric position of gaze to the primary position of gaze. The examiner holds one hand directly in front of the patient, and the other about 45 degrees out from the primary position as the patient gazes straight ahead. The patient is asked to fixate on the eccentric hand and then, on command, to shift the gaze to the hand at the primary position. Observe the eyes for overshooting of the target as they fixate in the primary position.

Speech is frequently involved. Once the characteristic changes are heard in a full-blown case, they will never be forgotten. Begin by having the patient take a deep breath and maintain "ahhh" as long as possible. This procedure basically tests the expiratory muscles and vocal cords. Listen for variations in pitch and volume, and for tremor. Now ask the patient to say "la, la, la" as long as possible. This maneuver superimposes rapid alternating movements of the tongue upon function of the expiratory muscles and vocal cords. Ask the patient to say "me, me, me" as long as possible, thus testing rapid alternating movements of the lips. Finally ask the patient to read a simple paragraph, and listen to meter, volume, pitch, and enunciation.

Upper Extremities

Passively flex and extend each arm at the elbow and assess the tone. Then have the patient extend the arm in front with elbows slightly flexed and eyes closed. Observe for tremor. Assess postural fixation and tone by observing for drift, and by tapping sharply proximally, after explaining what you are about to do. With cerebellar dysfunction there is marked wavering of the arm with the tapping and difficulty in maintaining the posture of the trunk.

Now test rebound of the right arm by placing your right hand on the patient's right shoulder. This maneuver prevents the arm striking the patient's face if cerebellar dysfunction is indeed present. Grasp the patient's right wrist with your left hand. Ask the patient to flex the right arm sharply, and you suddenly let go. The patient with dysfunction will be unable to arrest the progress of the arm, and it will rebound markedly off your right arm.

Test rapid alternating movements of first one hand and then the other with the thigh-slapping test. Use the sitting position. Have the patient strike first the palm and then the dorsum of the hand upon the thigh just above the knee. Abnormalities are more likely to be brought out if you make sure that the hand is reversed between each strike, performance is as rapid as possible, and the length of the movement is about breast high. Observe for abnormalities of force and timing, and difficulty in alternating between the palm and the dorsum of the hand.

Test finger–nose–finger alternating movements by having the patient first touch the pad of your index finger with the pad of his or her index finger. Then the patient touches his or her nose with the pad of the index finger, thus requiring reversal of the hand. Have the patient do this as rapidly as possible. Observe for intention tremor, a coarse side-to-side tremor that increases as your finger or the patient's nose is approached. There are irregularities in control of the timing and force of the movement with cerebellar dysfunction. Abnormalities can be intensified if the position of the examiner's finger is moved each time.

An interesting little test that can be performed by some patients is to have them use their fingers to tap out a tune on the table. With cerebellar dysfunction, there can be disturbances in timing and force of striking that will be quite apparent. This is known as arrhythmokinesis.

Lower Extremities

First the patient stands at rest. Observe whether the feet are placed close together, as normally, or wide apart. Normally, individuals stand at rest with little or no shifting of feet or movement of the body. In cerebellar dysfunction the patient shifts about and the trunk wavers unsteadily. In many instances patients are unable to stand without support.

Ask the patient to walk in the usual fashion across the room and back. With bilateral cerebellar disease there is a reeling, rolling, lurching, staggering gait that resembles drunkenness. In unilateral cerebellar disease there is deviation to the side of the lesion, probably as a result of the hypotonia.

Test tandem walking. Ask the patient to walk a straight line heel-to-toe. This is probably the most sensitive test of function of the vermis of the cerebellum. It is the first function to be lost in alcoholic cerebellar cortical degeneration. This is a reliable and rapid screening test if alcoholic cerebellar dysfunction is a possibility.

Now have the patient lie faceup on the bed and observe performance of the following tests:

Heel–knee. The right heel taps the left knee gently, just as a hammer taps a nail. The arc of the swing should be about 60 cm. Observe for abnormalities of force and rhythm. Then test the left leg.

Heel–shin. The right heel starts on the top of the left knee and slides down the shin to the foot. The heel should stay exactly on top of the shin. When abnormal, there is a coarse side-to-side tremor as the heel goes down the shin. The knee is the optimal starting position. In some patients the abnormality is most marked at the knee and disappears as the heel goes on down the shin. Also insist that the heel be placed on top of the shin in the midline since this makes the dysfunction more prominent.

Basic Science

The field of cerebellar anatomy and physiology is one of the most complex and rapidly advancing areas in the neurosciences. In this section we give only the briefest background information, focusing primarily on some clinical correlations.

Anatomically the cerebellum can be divided transversely into three lobes, anterior, posterior, and flocculonodular, and longitudinally into a midline vermis and two lateral hemispheres (Larsell's nomenclature) (Figure 69.1). The simplified scheme of cerebellar organization given in Table 69.1 is modified from DeMyer (1974). There are exceptions to some of the generalizations.

The cerebellum can be divided into three longitudinal zones on the basis of afferent connections:

1. Vestibulocerebellum: afferents from vestibular nuclei
2. Spinocerebellum: afferents from the spinal cord

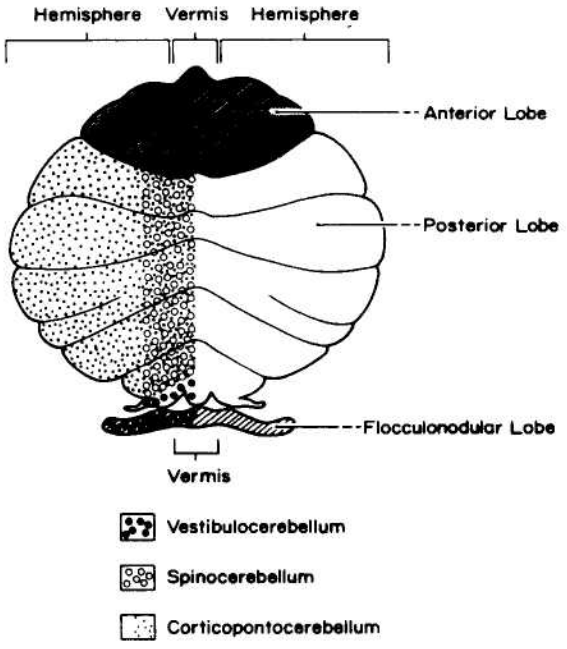

Figure 69.1
Anatomy and organization of the cerebellum. Modified from DeMyer W, Technique of the neurological examination: a programmed text, 2nd ed., McGraw-Hill, 1974; and Brodal A, Neurological anatomy, 2nd ed., Oxford University Press, 1969, using O Larsell's nomenclature, in Jansen J, ed., Comparative anatomy and histology of the cerebellum from myxinoids through birds, University of Minnesota Press, 1967.

3. Pontocerebellum or corticocerebellum: afferents from cerebral cortex via pontine nuclei

These divisions project onto the cerebellar nuclei and have efferent connections as outlined in the scheme. These three subdivisions do not exactly correspond to the anatomical divisions; there is considerable overlap.

DeMyer (1974) has put the function of the cerebellum lucidly by pointing out that the cerebellum probably evolved out of the vestibular nuclei. Using information provided by the vestibular system and other areas, the cerebellum equilibrates the contractions of axial musculature so that the eyes and head are properly positioned. In higher animals the cerebellum takes on the additional role of seeing to the smooth performance of voluntary movements by the limbs, working closely with the cerebrum. Thus the cerebellum, "sitting astride the vestibular nuclei," receives on the one hand information from the proprioceptive system, and on the other, information about commands the cerebral cortex is sending to muscles. The cerebellum sees to it that the movements are performed in a smooth, coordinated fashion, receiving constant feedback about what is actually happening.

Information as to what is happening in the muscles comes from muscle spindles, tendon organs, touch and pressure receptors, and from the labyrinth. These afferent impulses converge on the Purkinje cells of the spinocerebellar and vestibulocerebellar portions of the cerebellar cortex (see Figure 69.1), either directly or after synapse on granule cells. Efferent output from the Purkinje cells goes to the cerebellar nuclei (dentate, fastigial, and interpositus) and thence to the spinal cord (and therefore the lower motor neurons), vestibular nuclei, or cerebral cortex.

Table 69.1
A Simplified Scheme of Cerebellar Anatomy

Lobe	Phylogenetic subdivision	Afferent and efferent cerebellar peduncle	Midline nucleus
Anterior	Paleocerebellum (spinocerebellum)	Superior	Interpositus (emboliform and globose)
Posterior	Neocerebellum (cortico- or pontocerebellum)	Middle	Dentate
Flocculo-nodular	Archicerebellum (vestibulocerebellum)	Inferior	Fastigial

Interrelationships with the cerebral cortex are complex. Basically each area of the cerebral cortex that sends efferents to the cerebellum in turn gets efferents from that area of the cerebellum. The pathways from the cerebral cortex to the cerebellum can be divided into two groups (Brodal, 1969):

1. Routes via the inferior olive, pontine nuclei, and red nucleus, which show a precise topical organization.
2. Routes via the reticular nuclei principally. These nuclei are diffusely organized and thus can integrate impulses from many different sources before they reach the cerebellum.

Cerebellar efferents to the cortex go from the dentate nucleus mostly to the nucleus ventralis lateralis of the thalamus, thence to the cortex. Efferents from the cerebral cortex to the cerebellum go to the contralateral cerebellar hemisphere. Thus the right cerebellar hemisphere ultimately receives afferents from the right side of the body and the left cerebral cortex, and sends efferents to the same locations.

The cerebellum exerts its influence on motor activity via the cerebral cortex. It also directly influences the gamma fiber systems at the spinal cord level, thus influencing postural tone and reflexes. The nature of the cerebellar influence is still incompletely understood.

There are many problems with localization of cerebellar symptoms. The generalizations in Table 69.2 are crude but helpful.

Clinical Significance

There are a variety of clinical signs in cerebellar disease (Table 69.3). These signs have been studied in detail recently (Gilman, 1986; Gilman, Bloedel, and Lechtenberg, 1981).

Stance and gait abnormalities are the most common clinical signs. They reflect disease of the midline zone of the cerebellum. There is a broad-based stance with truncal instability during walking, causing falls to either side. The steps are irregular, and the feet may be lifted too high. Gait ataxia without limb impairment, occurring most commonly with alcohol damage and nutritional deficiency, indicates damage to the anterior superior vermis. Flocculonodular lesions can also produce stance and gait abnormalities. Tandem walking is often the earliest abnormality, and this maneuver is most severely affected.

Titubation consists of a rhythmic body or head tremor. There is a rotatory or rocking or bobbing movement. Clinically this has not turned out to have localizing value with respect to the part of the cerebellum involved.

The head can be rotated, or tilt to one side or the other. As with titubation, this does not have useful localizing value.

Oculomotor disturbances in cerebellar disease have been worked out in detail in the "pure" cerebellar degenerations (Leigh and Zee, 1983; Zee, 1984). The most frequent abnormalities include dysmetric saccades, fixation abnormalities, impaired smooth pursuit, postsaccadic drift, gaze-evoked nystagmus, rebound nystagmus, downbeat nystagmus, and positional nystagmus. Saccadic dysmetria most likely indicates dysfunction of the dorsal vermis and fastigial nuclei. The function of these structures is to control saccade amplitude. The vestibulocerebellum (flocculus) acts to provide stabilization of the retinal image, with dysfunction producing impaired smooth tracking, impaired fixation suppression of caloric nystagmus, and postsaccadic drift. (Also see Chapter 60, Cranial Nerves III, IV, and VI.)

Table 69.2
Localization of Cerebellar Symptoms

Disturbance	Localization
Limb ataxia (especially upper limbs) and hypotonia	Lateral lobes
Disturbed equilibrium—truncal ataxia: drunken gait, titubation of head and trunk	Flocculonodular lobe
Gait ataxia: inability to do tandem walking	Anterior lobe

Table 69.3
Clinical Signs of Cerebellar Disease

Abnormalities of stance and gait
Titubation
Rotated or tilted postures of the head
Oculomotor abnormalities
Decomposition of movement
Dysmetria
Dysdiadochokinesis and dysrhythmokinesis
Ataxia
Check and rebound abnormalities
Tremor
Dysarthria
Abnormalities of muscle tone

Decomposition of movement occurs with disease of the lateral zones of the cerebellum. This is reflected in difficulty with both simple and compound movements. Movement initiation and termination is affected.

Dysmetria is a trajectory disturbance; placement falls short of or extends beyond the initial goal, as in the finger to nose test. The heel–knee–shin test also demonstrates error in placement, as well as force. The lateral zone of the cerebellum is felt to be responsible for normal placement.

Repetitive movements, such as hand patting, are affected with dysfunction of the lateral zone of the cerebellum. The result is dysdiadochokinesis. Disorder of the rhythm of rapid alternating movements is known as dysrhythmokinesis. Disorders of the basal ganglia and corticospinal system can produce similar types of dysfunction.

Ataxia is the lack of smoothly coordinated movements. This incoordination is chiefly the combined result of dysmetria and decomposition of movement. Movements are imprecise, halting, awkward, and clumsy. Disease of the lateral cerebellar hemispheres causes limb ataxia.

Impaired check and excessive rebound are common signs in cerebellar disease. The patient, with eyes closed, is unable to return a limb that has been tapped and displaced to its original position. Overshoot occurs.

A static tremor, originating at the shoulder, can be brought out by having the patient hold outstretched arms parallel to the floor. A kinetic tremor (or intention tremor) is brought out by the finger to nose and heel to shin tests. It involves the proximal musculature. These tremors usually indicate disease of the lateral zone of the cerebellum on the ipsilateral side.

Dysarthria often occurs in severe cerebellar disease. In a sense, it is ataxia of speech. Articulation is uneven, words are slurred, and variations in pitch and loudness occur. Rhythm changes are prominent. Charcot applied the term "scanning speech" to a pattern heard in cerebellar disease; enunciation is difficult, words are produced slowly and in a "measured" fashion.

Muscle tone abnormalities in cerebellar disease were first described by Gordon Holmes in the 1920s. Hypotonia and pendular deep tendon reflexes are seen. These abnormalities are seen easily when there is unilateral cerebellar disease.

The cerebellum can be involved in a wide variety of systemic diseases, in addition to mass lesions and congenital afflictions. Alcoholism and remote cancer produce a cerebellar syndrome that at least initially begins within the anterior lobe and produces ataxia. Lead, mercury, dilantin, and other toxic or therapeutic agents can cause cerebellar degeneration. Various viral infections and hypoxia can produce prominent cerebellar involvement. Vascular disease can produce involvement of the cerebellum directly or by involvement of the cerebellar peduncles in the brainstem. Examples include the posterior inferior artery syndrome, the superior cerebellar artery syndrome, and the anterior inferior cerebellar artery syndrome. Cerebellar infarction and hemorrhage are other frequent manifestations of cerebrovascular disease. Common neoplasms include metastases, astrocytoma, medulloblastoma, angioblastoma, and acoustic neuroma.

Note that this chapter has not been concerned with ataxia due to involvement of the posterior columns of the spinal cord, the functions of which must be tested before considering ataxia to be of cerebellar origin.

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