Clinical Manifestations of Cerebellar Disease

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KEYWORDS

• Gait ataxia • Limb ataxia • Nystagmus • Dysdiadochokinesia • Dysmetria
• Kinetic tremor • Action tremor • Cerebellar mutism

KEY POINTS

- Clinical manifestations of cerebellar disease include ataxia and tremor as being the most prominent clinical signs, as well as nystagmus, dysarthria, and cognitive dysfunction.
- Recognition of the cerebellar pattern of disease can aid in the prompt and correct diagnosis, and lead to appropriate treatment and rehabilitation to minimize disability.

INTRODUCTION

Luigi Luciani initiated the study of cerebellar dysfunction in late 1800s, and later Gordon Holmes built and enhanced it in the early 1900s (**Tables 1** and **2**). The cerebellum is located in the posterior fossa of the brain. It controls the rate, direction, range, and force of voluntary movement through its vestibular connections (in the brainstem), and corrects and adjusts an individual's upright position in space. The cerebellum can be divided into 3 parts: midline, intermediate, and lateral. The midline cerebellum includes the cerebellar vermis and the fastigial, flocculus, and nodulus nuclei. The lateral cerebellum consists of 2 hemispheres, including the dentate nuclei. Damage to 1 hemisphere leads to symptoms that are most notable in the ipsilateral limbs.

Three cerebellar peduncles connect the cerebellum with the brainstem. The inferior cerebellar peduncle connects the cerebellum to the medulla oblongata. The important afferent fibers in the inferior cerebellar peduncle are the dorsal spinocerebellar, cuneocerebellar, olivocerebellar, vestibulocerebellar, reticulocerebellar, and the trigeminocerebellar tracts. The efferent tracts through the inferior cerebellar peduncle are the cerebellovestibular and cerebelloreticular tracts. The middle cerebellar peduncle is the largest of the 3 peduncles and connects the pons with the cerebellum, and the pontocerebellar tract is the main afferent tract. The superior cerebellar peduncle connects the cerebellum to the midbrain, and dentatothalamic and dentatorubral are the

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Table 1
Cerebellar connections

Peduncles	Connections	Afferent Tracts	Efferent Tracts
Superior	Midbrain	Ventral spinocerebellar Tectocerebellar Trigeminocerebellar Cerulocerebellar	Dentatothalamic Dentatorubral
Middle	Pons	Corticopontocerebellar	
Inferior	Medulla	Dorsal spinocerebellar Cuneocerebellar Olivocerebellar Vestibulocerebellar Reticulocerebellar Trigeminocerebellar	Cerebellovestibular Cerebelloreticular

main efferent fibers. The afferent tracts in the superior cerebellar peduncle include the ventral spinal cerebellar, tectocerebellar, trigeminocerebellar, and cerulocerebellar tracts. Damage or dysfunction of these pathways or the nuclei of the cerebellum results in various clinical manifestations (**Table 3**).

The most prominent sign of cerebellar dysfunction is incoordination. It is thought to be due to disruption in the timing of the normal patterning of agonist and antagonist muscle activity in the course of movement.[1] Lesion symptom mapping based on magnetic resonance imaging (MRI) is helpful in the study of the function of the cerebellar cortex and the cerebellar nuclei. Functionally, the cerebellum can be compartmentalized into 3 sagittal zones. In patients with cerebellar cortical degeneration, ataxia of stance and gait is correlated with atrophy of the medial (and intermediate) cerebellum, oculomotor disorders with atrophy of the medial cerebellum, dysarthria with the atrophy of the intermediate section, and limb ataxia with atrophy of the intermediate and lateral cerebellum.[2]

HYPOTONIA AND WEAKNESS

Although hypotonia can occur in acute cerebellar disease, it is not usually a major feature.[3] The patient may have pendular knee jerk responses. Hypotonia has been explained as a decreased response to stretch in the muscle spindle afferents. The inability of patients to check forearm movement in the rebound test is often said to result from hypotonia but may have other explanations. Although cerebellar lesions do not cause loss of motor strength in the traditional sense, many patients experience problems with sustaining a steady force with their hands. This may be mistaken for, or described by the patient as weakness.

Table 2
Functional zones of the cerebellum

Zone	Symptoms
Medial	Ataxia of gait and stance Oculomotor disturbances
Intermediate	Ataxia of gait and stance Dysarthria Limb ataxia
Lateral	Limb ataxia