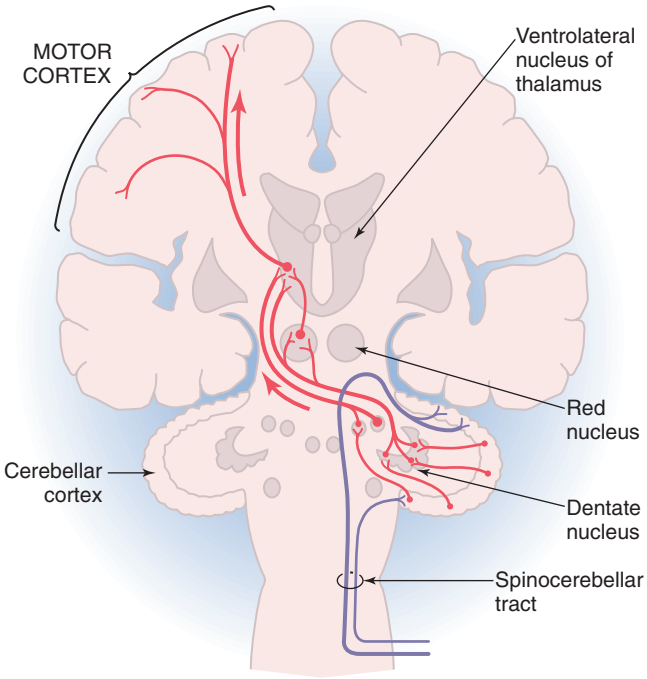

FIGURE 114-3 Corticocerebellar loop. The major cerebellar input is from the spinocerebellar tract. Outflow is to the motor cortex via the mesencephalon and thalamus.

monitoring and regulating motor actions. Eye movement abnormalities include square wave jerks on ocular fixation, jerky slow pursuit movements, hypo- and hyper-metric point-to-point saccades, and nystagmus. The characteristic speech pattern, a scanning dysarthria, displays an abnormal modulation of speech volume and velocity. Speech prosody is also affected. Voluntary movements are irregular and uncoordinated with irregularities in amplitude, velocity, and rhythm. With sustained posture a tremor may develop and increase in amplitude with prolonged sustention. On directed movements there is an irregular placement of the limb (i.e. dysmetria) with voluntary movements, such as finger to nose or heel to shin testing. An intention tremor (increasing amplitude of tremor as one approaches target) is characteristic of cerebellar dysfunction. Patients have difficulty regulating muscle contractions with an increase in rebound when the examiner pushes against a limb and the patient is asked to keep the limb still, or a reduced ability to check a movement when resistance to muscle contraction is suddenly removed.

SIGNS OF PERIPHERAL MOTOR SYSTEM DYSFUNCTION

Disorders affecting the peripheral motor system reflect disease of the motor unit: the anterior horn cell, peripheral nerve, neuromuscular junction, and muscles. All may be associated with weakness, muscle atrophy, hypotonia, hyporeflexia, fasciculations, and fibrillations. Anterior horn cell diseases cause a pure motor disorder with profound atrophy and prominent fasciculations. Paradoxically, the most common cause of anterior horn cell disease, amyotrophic lateral sclerosis, is associated with both upper and lower motor neuron syndrome: individuals have weakness, muscle wasting with spasticity, and increased muscle stretch reflexes. The motor neuron exits the spinal cord via a nerve root, in the limb multiple roots combine to form a plexus, and then individual peripheral nerves innervate specific muscles. Lesions of nerve roots, plexus, and peripheral nerves are often associated with both weakness and sensory symptoms in the regions innervated by the specific roots, plexus, or nerves. Muscle stretch reflexes are reduced or absent. The distribution of weakness, sensory, and reflex findings assists in accurate localization. Peripheral nerve disorders may affect single nerves, such as median nerve in carpal tunnel syndrome, or multiple nerves, such as polyneuropathy in diabetes. In the former setting combined motor and sensory findings are the rule, whereas in the latter, occasional pure motor neuropathies occur. Polyneuropathy is usually associated with a distal gradient of both motor and sensory abnormalities that can be slowly or rapidly ascending depending on the underlying etiology.

Neuromuscular junction disorders are characterized by fluctuating weakness and fatigability. Tone and reflexes are usually unaffected with the exception of Lambert-Eaton Syndrome, in which muscle stretch reflexes are reduced or absent. Muscle mass and sensation are preserved. On examination, an individual may demonstrate fatigable weakness with strength recovery after rest. Bulbar symptoms and signs may be prominent with a nasal speech pattern, various abnormalities of eye movements, ptosis, and neck weakness.

Disorders of the muscle classically result in proximal greater than distal limb weakness with some exceptions, such as inclusion body myositis and myotonic dystrophy, in which distal weakness predominates. Muscle diseases, especially toxic or inflammatory conditions, may be associated with myalgia and tenderness to muscle palpation. Muscle stretch reflexes are normal to slightly reduced. Sensation is preserved.

DIFFERENTIAL DIAGNOSIS OF PYRAMIDAL TRACT DISORDERS

Any disease affecting the nervous system can cause pyramidal tract dysfunction. Structural lesions due to stroke, tumor, infection, trauma, inflammatory and demyelinating diseases can cause pyramidal tract dysfunction with variable presentations depending on the underlying disease process and the variable involvement of non-motor systems. These disorders are discussed elsewhere.

Hereditary spastic paraplegia (HSP) is a rare and heterogeneous group of inherited disorders that causes progressive pyramidal tract dysfunction manifest clinically by spastic paraparesis.

DIFFERENTIAL DIAGNOSIS OF PERIPHERAL MOTOR SYSTEM DISORDERS

Similar to pyramidal tract dysfunction, lesions resulting from direct tumor involvement, infection, trauma, and inflammatory and demyelinating processes may all affect the peripheral motor system. In addition, specific inherited and acquired degenerative disorders may affect the peripheral motor system (Chapters 121, 122, and 123).

MOVEMENT DISORDERS

Movement disorders are a heterogeneous group of disorders associated with basal ganglia dysfunction. Movement disorders refer to the involuntary or abnormal movement, known as the