Chapter 19
Disorders of Motor Function

Components of the Neuromuscular System

- Neuromuscular unit containing motoneurons
- Myoneural junction
- Muscle fibers
- Spinal cord
- Descending pathways from the brain stem circuits
Classifications of Muscles

- **Extensors**
  - Muscles that increase the angle of a joint

- **Flexors**
  - Muscles that decrease the angle of a joint

Mechanisms Controlling Coordinated Movement

- **Agonists**
  - Promote movement

- **Antagonists**
  - Oppose movement

- **Synergists**
  - Assist the agonist muscles by stabilizing a joint or contributing additional force to the movement
Requirements of Motor Systems

- Upper motoneurons project from the motor cortex to the brain stem or spinal cord.
  - Directly or indirectly innervate the lower motoneurons or contracting muscles
  - Motor unit is a motor neuron and all the muscle fibers it innervates.
- Sensory feedback from the involved muscles
  - Continuously relayed to the cerebellum basal ganglia and sensory cortex
- Functioning neuromuscular junction that links nervous system activity with muscle contraction

The Motor Unit

- The motor unit consists of the motor neuron and the muscle fibers it innervates.
**Neuromuscular Junction**

- Serves as a synapse between a motor neuron and a skeletal muscle fiber

- Consists of the axon terminals of a motor neuron and a specialized region of the muscle membrane called the end plate

- The transmission of impulses is mediated by the release of the neurotransmitter acetylcholine from the axon terminals.

- Acetylcholine binds to receptors in the end plate region of the muscle fiber surface to cause muscle contraction.

**Motor Systems**

- **Pyramidal Motor System**
  - Originates in the motor cortex
  - Provides control of delicate muscle movement

- **Extrapyramidal System**
  - Originates in the basal ganglia
  - Provides background for the more crude, supportive movement patterns
Question

• Which motor system is responsible for crude muscle movements?
  
  – A. Pyramidal motor system
  
  – B. Extrapyramidal motor system

Answer

• B. Extrapyramidal motor system

• Rationale: Extrapyramidal motor system originates in the basal ganglia and provides background for the more crude, supportive movement patterns.
Movement Planning

- The primary motor cortex is responsible for execution of a movement.
- The premotor cortex for generating a plan of movement
- The supplemental motor cortex for rehearsing motor sequences of movement

Disorders of Motor Function

- Upper Motoneuron Lesions
  - Can involve the motor cortex, the internal capsule, or other brain structures through which the corticospinal or corticobulbar tracts descend, or the spinal cord
- Lower Motoneuron Lesions
  - Disrupt communication between the muscle and all neural input from spinal cord reflexes, including the stretch reflex, which maintains muscle tone
Disorders of Skeletal Muscle Groups

• Muscular Atrophy
  - If a normally innervated muscle is not used for long periods, the muscle cells shrink in diameter, lose much of their contractile protein, and weaken.

• Muscular Dystrophy
  - Genetic disorders that produce progressive deterioration of skeletal muscles because of mixed muscle cell hypertrophy, atrophy, and necrosis

Alterations of Neuromuscular Function

• Drugs and agents can alter neuromuscular function by changing the release, inactivation, or receptor binding of acetylcholine.
  - Curare acts on the postjunctional membrane of the motor end plate to prevent the depolarizing effect of the neurotransmitter.
    • Used during many types of surgical procedures
  - Clostridium botulinum blocks ACH and results in paralysis.
Myasthenia Gravis

• Definition
  – Disorder of transmission at the neuromuscular junction that affects communication between the motoneuron and the innervated muscle cell.

• Cause
  – Autoimmune disease caused by antibody-mediated loss of acetylcholine receptors in the neuromuscular junction

Components of the Peripheral Nervous System

• Motor and sensory branches of the cranial and spinal nerves

• The peripheral parts of the autonomic nervous system

• Peripheral ganglia
Peripheral Nerve Regeneration

- Damage to a peripheral nerve axon
  - Due to injury or neuropathy
    - Results in degenerative changes, followed by breakdown of the myelin sheath and Schwann cells

- Regeneration factors
  - Proximity to soma
  - Crushing versus cutting

Peripheral Neuropathy

- Definition
  - Any primary disorder of the peripheral nerves

- Results
  - Muscle weakness, with or without atrophy and sensory changes

- Involvement
  - Can involve a single nerve (mononeuropathy) or multiple nerves (polyneuropathy)
Mononeuropathies

- Caused by localized conditions such as trauma, compression, or infections that affect a single spinal nerve, plexus, or peripheral nerve trunk
  - Fractured bones may lacerate or compress nerves.
  - Excessively tight tourniquets may injure nerves directly or produce ischemic injury.
  - Infections such as herpes zoster may affect a single segmental afferent nerve distribution.

Polyneuropathy

- Involves demyelination or axonal degeneration of multiple peripheral nerves that leads to symmetric sensory, motor, or mixed sensorimotor deficits

- Typically, the longest axons are involved first, with symptoms beginning in the distal part of the extremities
Causes of Polyneuropathies

- Immune mechanisms (Guillain-Barré syndrome)
- Toxic agents (arsenic polyneuropathy, lead polyneuropathy, alcoholic polyneuropathy)
- Metabolic diseases (diabetes mellitus, uremia)

Question

- Lead toxicity would result in which of the following conditions?
  - A. Mononeuropathies
  - B. Polyneuropathies
  - C. Upper motor lesion
  - D. Myasthenia gravis
**Answer**

- B. Polyneuropathies

- Rationale: Polyneuropathies would result due to the systemic exposure to lead.

**Nerve Root Injuries**

- Ruptured intervertebral disk
  - Sensory deficits
    - Spinal nerve root compression
    - Paresthesias and numbness
      - Particularly of the leg and foot
      - Knee and ankle reflexes also may be diminished or absent
  - Motor weakness
The Cerebellum

- Coordination of motor movement
- Cerebellum-associated movement disorders
  - Causes
    - Congenital defect, vascular accident, or growing tumor
  - Types
    - Vestibulocerebellar ataxia
    - Decomposition of movement
    - Cerebellar tremor

Basal Ganglia

- A group of deep, interrelated subcortical nuclei that play an essential role in control of movement
- They receive indirect input from the cerebellum and from all sensory systems, including vision, and direct input from the motor cortex.
  - They function in the organization of inherited and highly learned and rather automatic movement programs.
  - They also are involved in cognitive and perception functions.
Structural Components of the Basal Ganglia

- Caudate nucleus
- Putamen
- Globus pallidus in the forebrain

Four Functional Pathways Involving the Basal Ganglia

1. A dopamine pathway from the substantia nigra to the striatum
2. A γ-aminobutyric acid (GABA) pathway from the striatum to the globus pallidus and substantia nigra
3. Acetylcholine-secreting neurons, which are important in networks within the neostriatum
4. Multiple general pathways from the brain stem that secrete norepinephrine, serotonin, enkephalin, and several other neurotransmitters in the basal ganglia and the cerebral cortex
Characteristics of Disorders of the Basal Ganglia

- Involuntary movements
- Alterations in muscle tone
- Disturbances in body posture

Types of Involuntary Movement Disorders

- Tremor
- Tics
- Chorea
- Athetosis
- Ballismus
- Dystonia
- Dyskinesias
**Parkinson Disease**

- **Definition**
  - A degenerative disorder of basal ganglia function that results in variable combinations of tremor, rigidity, and bradykinesia

- **Characteristics**
  - Progressive destruction of the nigrostriatal pathway, with subsequent reduction in striatal concentrations of dopamine

- **Clinical syndrome**
  - Parkinsonism

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**Amyotrophic Lateral Sclerosis (ALS)**

- **Definition**
  - A devastating neurologic disorder that selectively affects motor function

  - The disease typically follows a progressive course, with a mean survival period of 2 to 5 years from the onset of symptoms.
Locations of Motoneurons Affected by ALS

- The anterior horn cells of the spinal cord
- The motor nuclei of the brain stem, particularly the hypoglossal nuclei
- The UMNS of the cerebral cortex
- Death of LMNs leads to denervation, with subsequent shrinkage of musculature and muscle fiber atrophy.

Multiple Sclerosis (MS)

- A demyelinating disease of the CNS
- Most common nontraumatic cause of neurologic disability among young and middle-aged adults
- Characterized by exacerbations and remissions over many years in several different sites in the CNS
  - Initially, there is normal or near-normal neurologic function between exacerbations.
  - As the disease progresses, there is less improvement between exacerbations and increasing neurologic dysfunction.
Question

- Demyelination is the causative factor in which disease?
  - A. Parkinson disease
  - B. ALS
  - C. Multiple sclerosis

Answer

- C. Multiple sclerosis

- Rationale: Multiple sclerosis is caused by an autoimmune attack on the oligodendrocytes of the CNS.
Spinal Cord Injury (SCI)

- **Definition**
  - Damage to the neural elements of the spinal cord

- **Causes**
  - Motor vehicle crashes, falls, violence and sporting activities

- **Involvement**
  - Most SCIs involve damage to the vertebral column and/or supporting ligaments as well as the spinal cord.
  - Commonly involve both sensory and motor function

Types of Injuries to the Vertebral Column

- Fractures
- Dislocations
- Subluxations
Types of Incomplete Spinal Cord Injuries

- Central cord syndrome
- Anterior cord syndrome
- Brown-Séquard syndrome
- Conus medullaris syndrome

Areas Affected by SCI

- Spinal reflexes
- Ventilation and communication
- Autonomic nervous system
- Temperature regulation
- Edema and deep vein thrombosis
- Sensorimotor function
Areas Affected by SCI (cont.)

- Skin integrity
- Pain reception
- Bladder and bowel function
- Sexual function