

## Ticket to Enter: Degenerative Disorders Fact Sheet

Briefly define it, state the cause if known, name a few classic S/S you may see in a patient:

### 1. Amyotrophic Lateral Sclerosis

Definition: rapidly progressive, neurologic disorder with degeneration of upper and lower motor neurons that leads to progressive and eventually debilitating muscle weakness

Cause: unknown (pathophysiology: upper motor neurons – degenerate and stop communicating which leads to spasticity of the muscles; lower motor neurons – cannot produce or transport impulses from nerves which leads to flaccidity and atrophy of the muscles)

S/Sx: early – progressive muscle weakness and atrophy, advanced – dysphagia, dysarthria, pain, sleep disorders, spasticity, drooling, constipation, reflux, respiratory difficulties (many will die due to respiratory complications)

### 2. Huntington's Disease

Definition: progressive, degenerative brain disorder that results in involuntary movements and mental deterioration

Cause: genetic disorder - autosomal dominant disorder (pathophysiology: HD abnormal gene causes a deficiency in the neurotransmitters GABA and ACH and excess dopamine availability in HD)

S/Sx: chorea; mental cognitive/psych decline; facial movements: tics/grimacing; speech: slurred, hesitant, explosive; chewing/swallowing: difficulty; gait: disorganized; bladder/bowel: control can be lost; cognition, personality, and emotions are all affected

### 3. Multiple Sclerosis

Definition: chronic, progressive, degenerative, disorder of the CNS with demyelination of nerve fibers

Cause: unknown; research for viral, immune, genetic involvement (pathophysiology: autoimmune response leads to chronic inflammation, myelin sheath damage by demyelination, and gliosis (scar formation) which disrupts nerve transmission in the CNS; brain and/or spinal cord can be affected; demyelination causes nerve transmissions to be slowed, scar tissue causes plaques which interrupt nerve transmission; permanent loss of function could occur if axons become continually damaged)

S/Sx: fatigue (severe, disabling), impaired movement (early – limbs can feel weak or heavy, numbness and tingling may be present, as disease progresses – stiffness or gait problems, poor balance), visual disturbances (distortion of red/green colors, blindness in one eye), auditory disturbances (tinnitus or loss of hearing), dysarthria, dysphagia, bowel/bladder function loss

possible, sensory disturbance (paresthesia, pain), cognitive and emotional problems (decreased ST memory, concentration, emotional stability)

4. Myasthenia Gravis

Definition: autoimmune disease characterized by muscle fatigue and weakness from inadequate ACH receptor stimulation due to ACH receptor antibodies that attack acetylcholine receptors

Cause: unknown; (pathophysiology: antibodies are produced against ACH receptor sites; this results in a fewer number of acetylcholine receptor sites so the acetylcholine molecules cannot attach to these receptor sites and stimulate normal muscle contraction)

S/Sx: fluctuating weakness of skeletal muscles (muscles that move the eyes, eyelids, chew, swallow, speak, breathe); can also have trunk, shoulder, limb, neck weakness

5. Parkinson's Disease

Definition: chronic, progressive neurodegenerative disorder characterized by slowness in the initiation and execution of movement, increased muscle tone, tremor at rest, and gait disturbances

Cause: unknown; thought to be due to lack of dopamine in the brain due to degeneration of dopamine producing neurons causing an imbalance of dopamine and acetylcholine (pathophysiology: decreased dopamine levels due to destruction of dopamine producing cells in the substantia nigra located in the mid brain)

S/Sx: tremors, rigidity, akinesia, postural instability