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-
    a. Rapid progressive neuro disorder with degeneration of upper and lower motor neurons that leads to progressive and eventually debilitating muscle weakness. Upper motor neurons degenerate and stop communicating that leads to muscles stiffening. Lower motor neurons don't produce or transport impulses which leads to flaccidity and atrophy of the muscles.
    b. S/S: Weakness spreading to multiple muscles, progressive muscle weakness and atrophy, dysphagia, dysarthria, pain, sleep disorders, spasticity, drooling, constipation, reflux, respiratory difficulties.
    c. No cure but symptomatic treatment with meds: Riluzole and Edaravone
2. Huntington's Disease
    a. Progressive, degenerative brain disorder that results in involuntary movements and mental deterioration. Men/ women affected equally ages 30-50. Course is chronic and progressive (no cure). Abnormal gene identified: with each pregnancy a affected parent has 50% of passing gene to child.
    b. Prominent clinical features are chorea and mental cognitive/psych decline. Chorea (jerky, brisk and purposeless movements, involuntary abnormal movements. Disease progresses writhing and twisting of the face, limbs and body can occur). Face (tic/grimacing). Speech (slurred, hesitant, explosive). Difficulty chewing. Incontinent.
    c. No cure, symptom control: Tetrebenzine and Neuroleptics
3. Multiple Sclerosis-
    a. Chronic, progressive degenerative disorder of the CNS with demyelination of nerve fibers. Unknown cause. Usually ages 20-50, women affected more than men. Genetic involvement. Autoimmune responds leads to chronic inflammation, myelin sheath damage by demyelination and gliosis which disrupts nerve transmission. Scar tissue causes plaques which interrupt nerve transmission. Permanent loss of function could occur if axons become continually damaged.
    b. Fatigue, limbs can feel weak or heavy, numbness and tingling, visual disturbances, auditory disturbances (tinnitus or loss of hearing), dysphagia, paresthesia, pain, decreased ST memory, concentration, remission, disease progression.
4. Myasthenia Gravis-
    a. Autoimmune disorder characterized by muscle fatigue and weakness from inadequate ach receptor stimulation due to Ach receptor antibodies that attack acetylcholine receptors. Antibodies are produced against ACH receptor sites. Results in fewer number of acetylcholine molecules cant attach to these receptor sites and stimulates normal muscle contraction. Cause is unknown.
    b. Primary feature fluctuating weakness of skeletal muscles, muscles affected (eyes, eyelids, chew, swallow, speak and breathe) Trunk, shoulder, neck weakness.

c. No cure but anticholinesterase drugs: pyridostigmine

5. Parkinson's Disease
   a. Chronic progressive neurodegenerative disorder characterized by slowness in the initiation and execution of movement, increased muscle tone, tremor at rest and gait disturbance. Unknown cause but 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.
   b. S/S: tremor, rigidity, akinesia and postural instability. Depression, anxiety, apathy, pain, fatigue, short term memory, sweating, flushing, orthostatic hypotension, urinary retention, constipation, sleep disorder.
   c. Treatment, control symptoms and maintain independence with antiparkinsonian drugs to restore balance of dopamine and acetylcholine.