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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-

Rapidly progressive, neurologic disorder with degeneration of upper and lower motor neurons that leads to progressive and eventually debilitation muscle weakness. ALS effects both upper and lower motor neurons. Upper motor neurons degenerate and stop communicating which leads to spasticity of the muscles (stiffness) Lower motor neurons cants produce or transport impulses from nerves which leads to flaccidity and atrophy of the muscles. Progression can include weakness spreading to multiple muscles. As this advance's dysphagia, pain, sleep disorders, spasticity, drooling, constipation, reflux, respiratory difficulties, can all occur. EMG to look at muscle activity can help diagnose. There is no cure but there is symptomatic treatment. Riluzole – which is a glutamate antagonist that may slow down the progression of ALS. Edaravone which relieves oxidative stress. Collaborative care with a nutritional consult, OT, speech, and PT. Emotional and psychological support is needed, because the patient may cognitively be ok, but the body is wasting away.

2. Huntington's Disease

Progressive, degenerative brain disorder that results in involuntary movements and mental deterioration. Men and women are affected equally usually between the ages of 30-50. Chronic and progressive but not curable. Abnormal gene identified, autosomal dominant disorder. A deficiency in the neurotransmitters GABA and Ach. Excess dopamine availability in HD, that leads to symptoms that are opposite of Parkinson's. The most prominent clinical features are, chorea and mental cognitive/psych decline. Chorea is jerky, brisk and purposeless movements as the disease progresses writhing and twisting of the face, limbs and body can occur. Slurred speech, disorganized gait, bladder/bowel control can be lost, personality and emotions are affected, and facial movements like tics and grimacing can occur. There is no cure, and medications can help with symptom control. May need a higher nutritional calorie intake per day. Long term care may eventually be needed.

3. Multiple Sclerosis-

Chronic, progressive, degenerative disorder of the CNS with demyelination of nerve fibers. Can happen to younger or older adults. Women are affected more than men. The cause is unknown and research for viral, immune, and genetic involvement. It is a autoimmune response that leads to chronic inflammation of the myelin sheath that is damaged by demyelination and gliosis (scar formation) which disrupts nerve transmission in the CNS. The brain and or spinal cord could be affected. Demyelination causes nerve transmissions to be slower. Scar tissue causes plaques which interrupt nerve transmission. Permanent loss of function could occur if axons become

continually damaged. Fatigue is common and disabling. Impaired limb movements and numbness and tingling may be present. Optic nerve disturbances, sensory disturbances, dysarthria and dysphagia. Classifications are characterized by flares, remission and disease progression. There is relapsing remitting, secondary progressive, primary progressive, and progressive relapsing. Lumbar puncture may be done to diagnose. Early drug therapy is most recommended. Goals of care are to maintain function, independence, well-being, and reduce exacerbations.

4. Myasthenia Gravis-

Autoimmune disease characterized by muscle fatigue and weakness from inadequate Ach receptor stimulation due to Ach antibodies that attack acetylcholine receptors. Nerve impulses arrives at a nerve ending, releases chemical called Ach. It travels across synaptic cleft and attaches at receptor sites, becoming activated by Ach and then causing muscle contraction once enough sites have been activated. Antibodies are produced against ACH receptor sites. Resulting in fewer number of Ach receptors sites so the Ach molecules can't attach to these receptor sites and stimulate normal muscle contraction. The cause is unknown. Primary feature is fluctuating weakness of skeletal muscles, can also have trunk, shoulder, and limb weakness. Anti-Ach R is a lab test. Anticholinesterase drugs are uses to help prevent destruction of Ach by inhibiting the enzyme acetylcholinesterase, enhancing availability of Ach and impulse transmissions across the muscle junction. Complications are myasthenic crisis- insufficient medication cause not enough ACH and cholinergic crisis- excessive medication causes too much Ach.

5. Parkinson's Disease –

Chronic progressive neurodegenerative disorder characterized by slowness in the initiation and execution of movement, increase muscle tone, tremor at rest, and gait disturbance. The cause is unknown 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. Some symptoms are tremors, rigidity, and postural instability. Onset is usually gradual, it's a prolonged course. TRAP is the cardinal sign. Clinical findings are presence of TRAP signs and positive response with anti-parkinsonian medications. Treatment, control and maintain symptoms. Anti-parkinsonian drugs to restore balance of dopamine and acetylcholine. Carbidopa/Levodopa which is converted to dopamine in the basal ganglia and carbidopa prevents enzymatic break down of Levodopa allowing for more levodopa to reach the brain which will then convert it to dopamine. Surgical management provides some relief but is not curative.

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