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Polyneuropathies and Guillain-Barre Syndrome

Polyneuropathies

Polyneuropathies involve demyelination or axonal degradation of multiple peripheral nerves that lead to symmetric sensory, motor, or mixed sensory motor deficiencies. Typically the longest axons are involved first.

Guillain-Barre Syndrome (GBS)

Watch the video Guillain-Barre Syndrome (GBS)

Guillain-Barre syndrome (GBS) is a type of acute polyneuropathy that rapidly damages peripheral nerves and causes sudden onset of flaccid paralysis. It may hinder movement, sensation, or organ function depending on which nerves are involved. While rare, it is the most common cause of sudden-onset flaccid paralysis in the US. GBS is seen equally in men and women.

Patients with GBS typically experience absent reflexes, muscle weakness, and flaccid paralysis. A common symptom of GBS is **paresthesia**, which is tingling and numbness in the hands and feet due to damage to peripheral axons. Because electrical signals must travel the farthest to reach the arms and legs, symptoms of paresthesia and muscle weakness appear first in the hands and feet and then ascend upwards towards the torso (referred to as “toes to nose”). Paralysis of the diaphragm can be very dangerous and lead to patients requiring ventilatory support. Symptoms of GBS are symmetrical, which means they occur on both sides of the body. If present, most pain with GBS is experienced in the back, shoulders, and thighs. GBS can affect neurons of the autonomic nervous system as well. Therefore, symptoms may include postural or orthostatic hypotension, cardiac arrhythmias, facial flushing, and abnormalities of sweating and urinary retention. While there are differences in the rate of GBS progression between patients, maximum weakness usually occurs 2-3 weeks after the onset of symptoms.

Interestingly, GBS most commonly occurs after a gastrointestinal or respiratory infection. The most common gastrointestinal infection correlated with GBS is gastroenteritis/food poisoning caused by the bacteria *Campylobacter jejuni*. The most common respiratory infections are by the cytomegalovirus (CMV), Epstein-Barr virus (EBV), or *mycoplasma pneumoniae*. Occasionally, surgery or flu vaccination precedes contracting GBS as well.

There are two main subtypes of GBS:

- **Acute inflammatory demyelinating polyneuropathy (AIDP)** is the most common type of GBS. With AIPD, highly active T-cells and macrophages launch an immune attack directed at peripheral myelin sheaths. The axons may be damaged, but this occurs largely as a secondary consequence of the attack on the myelin.
- **Acute motor axonal neuropathy (AMAN)** is another common type of GBS. In AMAN, damage is mediated by the antibody IgG and complements destruction of the axon. Compared to AIDP, AMAN progresses more rapidly and may cause a prolonged paralysis and greater risk of respiratory failure. AMAN is associated with previous GI infections of *Campylobacter jejuni* and respiratory infections by CMV and EVB. The bacteria and these viruses contain antigens in their capsules similar to gangliosides in the axon of neurons. As the immune system tries to kill the pathogens, it may also erroneously injure the axons of neurons as antibodies against the pathogen cross-react with neurons. This is another example of molecular mimicry causing pathology.

GBS is diagnosed by observing symmetrical indicative symptoms, absent reflexes, and testing **cerebral spinal fluid (CSF)**. CSF results that suggest GBS are higher levels of protein in the CSF due to the nervous system inflammation and a low WBC count. Nerve conduction studies and electromyography can help validate a diagnosis by showing decreased conduction velocities of action potentials and decreased muscle recruitment.

Because GBS patients are immobile, anticoagulant therapy and manual movement of their limbs by caregivers is useful to prevent DVTs. Plasmapheresis is done to remove antibodies and other immune mediators that contribute to the destruction of neurons. IV immunoglobulin therapy from donors may also be useful in the treatment of GBS. The effects of immunoglobulin therapy are not well understood, but may involve the stimulation of negative feedback loops to diminish the autoimmune response of GBS. Antibiotics are used in the cases where an instigating infection is suspected to be the triggering event. GBS patients are often treated in the ICU because they must have mechanical ventilation. Unfortunately, 2-5% of patients die from complications of respiratory paralysis and cardiac arrest. After the acute phase, occupational, physical, and psychological therapy are important in recovery. Most patients with GBS have a full recovery, but 30% of patients still have weakness after three years and 3% have weakness and tingling after many years.