THE MANY FACES OF GUILLAIN-BARRÉ SYNDROME


**Take home points:**
1. Classic Guillain-Barré syndrome (GBS) presents as a rapidly progressive, ascending paralysis with distal areflexia and a mild sensory deficit.
2. We now know that GBS represents a spectrum of neurologic diseases with a common theme: rapidly progressive, symmetric neurologic injury that is due to immune dysregulation (molecular mimicry).
3. Think of GBS any time you have a patient with rapidly progressive, symmetric neurologic injury. Plasmapheresis is the only proven treatment, based on systematic reviews of the data. IVIG may be helpful but steroids are not helpful and are potentially harmful.

**Classic Guillain-Barré syndrome (GBS): accounts for approximately 80% of GBS cases**
- Also called acute inflammatory demyelinating polyneuropathy (AIDP) because most of the immune-mediated injury is directed at the myelin (in severe cases, axons can be affected as well).
- Rapidly progressive, ascending paralysis with distal areflexia and a mild sensory deficit.
- Proximal > distal weakness
- History of antecedent viral URI (most commonly due to CMV) or diarrheal illness (due to *Campylobacter*).
- On CSF exam, classic GBS presents with albuminocytologic dissociation (no WBCs but high protein).

**Acute motor axonal neuropathy (AMAN) variant: accounts for approximately 10-15% of GBS cases**
- Rapidly progressive, ascending paralysis with **normal reflexes** (sometimes associated with hyperreflexia) and a no sensory deficit.
- Respiratory failure is common.
- Good prognosis in most patients.
- On pathology there is widespread (and sometimes severe) destruction of axons.

**Acute motor sensory axonal neuropathy (AMSAN) variant: rare finding in GBS**
- Rapidly progressive, ascending paralysis with areflexia and moderate-to-severe sensory deficits.
- This is a fulminant form of GBS and is associated with widespread axonal injury.
- Prognosis is very poor; recovery is very slow and usually incomplete.

**Miller Fischer variant: accounts for approximately 5% of GBS**
- Classic triad of ataxia, areflexia, and ophthalmoplegia.
- Also can present with mild limb weakness, ptosis, facial palsy and bulbar palsy.

**Other variants of GBS:**
- Pure sensory
- Pure dysautonomic
- And many more continue to be described…

**Testing and treatment:**
- Make the diagnosis by CSF (see above) as well as EMG/NCV and biopsy if necessary
- Plasmapheresis is the only proven therapy (Cochrane systematic review)
- IVIG may be useful if plasmapheresis is not available
- **Steroids are not useful.**