Guillain-Barré-Strohl Syndrome

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Objectives: To understand this rare, acute polyneuropathy, and in particular the clinical indices of deterioration that indicate requirement for ventilatory support.

What is it? Guillain-Barré syndrome is the historical name for a heterogeneous series of acute immune mediated demyelinating polyneuropathies, which most frequently present as an acute illness following some infectious episode. Progressive, ascending symmetrical muscle weakness beginning in the legs, absence or depressed deep tendon reflexes, and a paucity of sensory findings, are the hallmarks of the condition. Bladder dysfunction is unusual at presentation. It is the most common cause of acute flaccid paralysis in children in the post-poliomyelitis era.

Causes: An immune response to a preceding infection, due to cellular mimicry. Campylobacter jejuni is the most commonly associated pathogen in the U.S., and might be associated with a worse prognosis. Influenza, CMV, HIV, Zika and EBV are also common culprit organisms. About 60% of cases of GBS will have a history of recent URI or GI infection. Very rarely it can follow immunization as an idiosyncratic reaction.

Presentation: Symptoms typically begin with symmetrical muscle weakness in the legs and progress proximally, but arms, face and bulbar symptoms also occur. Differing presentations give rise to different syndromes. See below.

Severity varies from mild difficulty with ambulation to complete paralysis. Patients present within a few days to 2 weeks after onset. 90% of cases are at their maximum within four weeks.

Paresthesias occur frequently in extremities, but sensory abnormalities on clinical exam are absent or mild. Back pain secondary to nerve root inflammation can be a presenting feature.

Facial nerve palsies are variable, and can be bilateral. Oropharyngeal and oculomotor weakness occur in 15-50% of cases. Dysautonomias are common, bradycardia, tachycardia, diarrhea/constipation, SIADH, hyponatremia, and a reversible cardiomyopathy. Perhaps most importantly, progressive respiratory muscle weakness requiring ventilatory support is present in 10-30%.

What presenting features make a diagnosis of GBS UNLIKELY?
Back pain with bowel or bladder dysfunction at onset
Fever (If fever present at onset, think transverse myelitis; especially in HIV or post infectious patients)
Any significant loss of sensation
Marked asymmetry of muscle weakness

Diagnosis
Primarily clinical. Imaging rarely helpful, although MRI, if you can get one, may show enhancement of spinal nerve roots, thecal sac and potentially brain changes.
CSF protein is elevated, especially after one week.
CSF WBC is usually normal, although a mild pleocytosis, can occur.
This combination of elevated protein and normal CSF WBC is known as albuminocytologic dissociation.

Electrodiagnostic nerve conduction studies and EMG are useful both for diagnosis and prognosis, and for differentiating the main variants of GBS. These are not available in the emergency department.

The above criteria are diagnostic of classical GBS, the most common form in the U.S., earning the moniker Acute Inflammatory Demyelinating Polyradiculoneuropathy, AIDP.

But… up to 20 variations of GBS have been described (what?!). The most likely to be met on board exams is Miller Fisher Syndrome, (MFS) which typically displays muscle weakness and areflexia in the upper extremities, along with ophthalmoplegia and ataxia. This is an insanely rare condition and therefore limited reports on optimal management.

3 Hallmarks of Guillain-Barré
1. Progressive symmetric muscle weakness
2. Absent or decreased DTRs
3. Paucity of sensory findings
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MANAGEMENT:
Let's set the record straight- no therapy will reverse the disease process. Potentially severe autonomic/hemodynamic dysfunction mandates ICU admission with close monitoring until the disease severity is established and/or nadir has been passed. Patients may require vasopressor and inotropic support if there is severe dysautonomia.

Plasma Exchange or IVIG will speed recovery. There is no added value in using both. These will be performed in the ICU, not the ED.

Up to 30% will develop respiratory failure requiring ventilator support.

The Numbers:
Forced Vital Capacity < 20 mL/Kg
Negative Inspiratory Force, (NIF), < 30 cmH2O
Maximum Expiratory Pressure < 40 cmH2O.

These indicate impending respiratory failure and are indications for intubation.

Caution: succinylcholine is contraindicated during intubation. The reason is muscle denervation results in up-regulation of acetylcholine receptors at neuromuscular junctions. The membrane is therefore super sensitive to succinylcholine, leading to hyperkalemia and potentially life-threatening arrhythmias.

CLINICAL features that are concerning for respiratory failure:
Bulbar dysfunction, swallowing difficulties are ominous clinical signs of impending respiratory failure.
Inability lift head
Rapid onset of disease (i.e., < 7 days from onset to admission/nadir)
Inability to cough or stand
Vital capacity < 60% of predicted
Severe muscle weakness
Elevated LFT’s

PAIN CONTROL
Gabapentin and carbamazepine are primary agents as pain is neuropathic
Use opioids with care in the setting of autonomic dysfunction
 Epidural morphine may be useful.

Prognosis is spotty. Only about 60% of patients regain full neurological function in one year. Yikes.

References