Guillain-Barré Syndrome (GBS) Joel Steinberg

Definition: rapidly ascending symmetric weakness, often accompanied by paresthesias, reflecting underlying inflammation-mediated damage of peripheral nerve myelin.

Additional Description:

- Weakness usually occurs first in lower limbs leading to difficulty walking and ascends to involve the trunk, arms, face, cranial nerves and breathing muscles, sometimes with respiratory failure.
- Diagnostic features: absent or decreased deep tendon reflexes; elevated spinal fluid protein.
- Additional names for GBS include: Landry's ascending paralysis; Landry-Guillain-Barré-Strohl syndrome; post-infectious polyneuropathy; acute inflammatory demyelinating polyradiculoneuropathy (AIDP).

Causes:

- Acute upper respiratory track or gastrointestinal (GI) infection with diarrhea are frequent causes of the most common form of GBS in North America and Europe, AIDP.
- Other prodromal events or triggers include surgery, pregnancy, 1976 swine influenza immunization; the chicken gut bacterium, *Campylobacter jejuni*, triggers the GBS variant, acute motor axonal neuropathy, in Chinese children that play in mud contaminated with droppings.
- A popular explanation for the cause of GBS is the molecular mimicry-innocent bystander theory. Per this theory, immunogenic molecules, epitopes, of a microbe appears similar to or mimic molecules in the peripheral nerve. Entry of a microbe into a person triggers their immune system to attack it. And because nerve components look similar, the immune system also attacks the nerve. The nerve has become the innocent bystander. In most cases, myelin is the principle target of the immune attack.

Scope of the Problem: Incidence is 1 to 2 new cases per 100,000 population per year, making GBS the most common cause of acute flaccid paralysis. Age range: 6 months and upwards, with increased frequency above age 50. Gender predilection: male 1-1/2 fold over female. No ethnic predilections.

Differential Diagnosis: anxiety, hysteria; Miller Fisher variant of GBS, with the triad of diplopia, ataxia and areflexia; spinal stenosis, myasthenia gravis, amyotrophic lateral sclerosis, polio, hypokalemia, dehydration, sepsis, hypothyroidism, hyperventilation, panic attack.

Diagnosis

History:

- Typical scenario (70% of patients) – rapid onset, over days, following a recent mild upper respiratory or GI infection with diarrhea, of paresthesias and weakness.
- Paresthesias, e.g., numbness, tingling, pain, of the feet, hands, back, thighs, may predate weakness by a day or so.
- Weakness is usually ascending and symmetric, starting in the legs, then to the hands and breathing muscles, presenting as clumsy walking, difficulty arising from a chair, climbing stairs or dropping objects.
- Clinical course can vary widely, from a very mild case, with fleeting paresthesias and a waddling gait, to total paralysis with need for mechanical ventilation.
- Diagnosis early in disorder's course can be difficult when only paresthesias and mild weakness are present, when the differential can include anxiety or neurosis.

- Can become a diagnosis of exclusion, when common causes of weakness (hypokalemia, dehydration) are absent.

PE: - Difficulty climbing onto examining table, standing on toes; duck-like waddling gait.
- Absent or diminished deep tendon reflexes in weak limbs.
- Subtle sensory changes such as impaired two-point or sharp versus dull touch discrimination.

Lab:- Spinal fluid protein usually elevated by day 10 of symptoms without elevated cell count (cytoalbuminologic disassociation).
- Nerve conduction velocity-electromyography test shows slowed nerve conduction, F wave loss, temporal dispersion (non-uniform conductions speeds), other markers of demyelination.

Imaging: CNS imaging excludes brain or spinal cord involvement.

Treatment:
Two tracks of care are used, disease specific and general supportive care.

1. Both high dose intravenous immune globulins and plasma exchange (plasmapheresis) shorten time to walking and/or time to weaning off ventilator. Either but not both are indicated if the patient is sufficiently symptomatic, and started within two weeks of onset of symptoms. Corticosteroids may actually prolong paralysis and are thus contraindicated.
2. Supportive care:

- Track respirations (e.g., inspiratory effort) closely and initiate mechanical ventilation prn.
- Treat autonomic dysfunction prn, e.g., beta blockers for tachycardia and/or hypertension; prn temporary pacer for symptomatic bradycardia.
- For bulbar palsy, of cranial nerves IX to XII, with, e.g., choking on secretions, use prn intubation to protect lungs.
- DVT, PUD prophylaxis
- Bedside PT/OT, with, e.g., bedside range of motion to maintain joint mobility, prevent contractures, repositioning to prevent decubiti, float heels in bed, oral hygiene.

Recovery:
- Maximal weakness, nadir, is reached within 4 weeks of onset of symptoms.
- Following a plateau phase, momentary to weeks, improvement follows, with strength returning in a descending pattern, over weeks to up to 2 years and occasionally longer.
- Wean off ventilator as strength returns.
- Transfer to rehab setting when medically stable.
- Relapse is rare.
- During rehab, if strong muscles substitute for weak muscles, customize strengthening exercises.
- For fatigue, pace activities with rest periods to prevent exhaustion and collapse.
- Most patients reach full to nearly full recovery.
- 3-5% succumb, usually to cardiorespiratory complications. 15% may experience chronic major weakness, requiring wheelchair and/or crutch use (think Franklin D. Roosevelt).
- Chronic paresthesias, e.g., pain, can be treated with NSAID's, moist heat, gabapentin, etc.
- Professional and patient literature and emotional support available via GBS/CIDP Foundation.

Clinical Pearl:
- In the newly weak patient with paresthesias, check ankle and knee reflexes.
- Because of the risk of cardiopulmonary collapse, treat GBS as a medical emergency.