Guillain-Barré Syndrome

AN AUTOIMMUNE DISORDER OFTEN CONSIDERED A POSTINFECTIOUS POLYNEUROPATHY INVOLVING MAINLY MOTOR BUT ALSO SENSORY AND SOMETIMES AUTONOMIC NERVES.

THIS SYNDROME AFFECTS PEOPLE OF ALL AGES AND IS NOT HEREDITARY.

CLINICAL MANIFESTATIONS

- ✔THE PARALYSIS USUALLY FOLLOWS A NON SPECIFC GASTROINTESTINAL(*CAMPYLOBACTER JEJUNI, HELICOBACTER PYLORI*)OR RESPIRATORY INFECTION (*MYCOPLASMA PNEUMONIAE*) BY APPROXIMATELY 10 DAYS.
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- ✔ GUILLAIN-BARRÉ SYNDROME IS REPORTED FOLLOWING ADMINISTRATION OF VACCINES AGAINST RABIES, INFLUENZA, AND POLIOMYELITIS (ORAL) AND FOLLOWING ADMINISTRATION OF CONJUGATED MENINGOCOCCAL VACCINE, PARTICULARLY SEROGROUP C.
- ❑ADDITIONAL INFECTIOUS PRECURSORS OF GUILLAIN-BARRÉ SYNDROME INCLUDE MONONUCLEOSIS, LYME DISEASE, CYTOMEGALOVIRUS, AND *HAEMOPHILUS INFLUENZAE* (FOR THE MILLER-FISHER SYNDROME

INITIAL SYMPTOMS INCLUDE NUMBNESS AND PARESTHESIA, FOLLOWED BY WEAKNESS. THERE MAY BE ASSOCIATED NECK, BACK, BUTTOCK, AND LEG PAIN. WEAKNESS USUALLY BEGINS IN THE LOWER EXTREMITIES AND PROGRESSIVELY INVOLVES THE TRUNK, THE UPPER LIMBS, AND, FINALLY, THE BULBAR MUSCLES, A PATTERN KNOWN AS **LANDRY ASCENDING PARALYSIS.**

PROXIMAL AND DISTAL MUSCLES ARE INVOLVED RELATIVELY SYMMETRICALLY, BUT ASYMMETRY IS FOUND IN 9% OF PATIENTS.

THE ONSET IS GRADUAL AND PROGRESSES OVER DAYS OR WEEKS;

PARTICULARLY IN CASES WITH AN ABRUPT ONSET, TENDERNESS ON PALPATION AND PAIN IN MUSCLES ARE COMMON IN THE INITIAL STAGES.

AFFECTED CHILDREN ARE IRRITABLE. WEAKNESS CAN PROGRESS TO INABILITY OR REFUSAL TO WALK AND LATER TO FLACCID TETRAPLEGIA.

MAXIMAL SEVERITY OF WEAKNESS IS USUALLY REACHED BY 4 WK AFTER ONSET

- **Bulbar involvement** occurs in about half of cases. Respiratory insuffciency can result.

Dysphagia and facial weakness are ofen impending signs of respiratory failure. The facial nerves may be involved.

Some young patients exhibit symptoms of viral meningitis or meningoencephalitis. Extraocular muscle involvement is rare

- Tendon reflexes in Guillain-Barré syndrome are lost, usually early in the course, but are sometimes preserved until later; areflexia is common but hyporeflexia may be seen; 10% may have normal reflexes.
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  The **autonomic nervous system** is also involved in some cases. Lability of blood pressure and cardiac rate, postural hypotension, episodes of profound bradycardia, or tachycardia and occasional asystole occur. Cardiovascular monitoring is important. A few patients require insertion of a temporary venous cardiac pacemaker.

- **Congenital Guillain-Barré syndrome** is rarely, manifesting as generalized hypotonia, weakness, and areflexia in an affected neonate, fulflling all electrophysiologic and CSF criteria and in the absence of maternal neuromuscular disease. Treatment might not be required, and there is gradual improvement over the 1st few mo and no evidence of residual disease by 1 yr of age.

- **LABORATORY FINDINGS AND DIAGNOSIS**

CSF studies are essential for diagnosis. The CSF protein is elevated to more than twice the upper limit of normal, the glucose level is normal, and there is no pleocytosis. Fewer than 10 white blood cells/mm3 may be found.

- The results of bacterial cultures are negative, and viral cultures rarely isolate specifc viruses. The dissociation between high CSF protein and a lack of cellular response in a patient with an acute or subacute polyneuropathy is diagnostic of GBS.

- MRI of the spinal cord may be indicated to rule out disorders , MRI fndings include thickening of the cauda equina and intrathecal nerve roots with gadolinium enhancement. These finds are fairly sensitive and are present in >90%

- Motor nerve conduction velocities are greatly reduced,
- sensory nerve conduction time is often slow.
- Electromyography shows evidence of acute denervation of muscle.
- *Serum creatine kinase level may be mildly elevated or normal*.
- Antiganglioside antibodies,

- Muscle biopsy is not usually required for diagnosis; specimens appear normal in early stages and show evidence of denervation atrophy in chronic stages. Sural nerve biopsy tissue shows segmental demyelination, focal inflammation

- Serologic testing for *Campylobacter* and *Helicobacter* infections helps establish the cause if results are positive but does not alter the course of treatment. Results of stool cultures are rarely positive because the infection is self-limited and only occurs for about 3 days, and the neuropathy follows the acute gastroenteritis

- **TREATMENT**
  Patients in early stages of this acute disease should be admitted to the hospital for observation because the ascending paralysis can rapidly involve respiratory muscles during the next 24 hr.

Respiratory effort **must** be monitored to prevent respiratory failure and respiratory arrest..

- Rapidly progressive ascending paralysis is treated with intravenous immunoglobulin (IVIG), administered for 2, 3, or 5 days.

A commonly recommended protocol is IVIG 0.4 g/kg/day for 5 consecutive days, but some studies suggest that larger doses are more effective (1 g/kg/day for 2 consecutive days) and related to improved outcome.

- Plasmapheresis and/or immunosuppressive drugs are alternatives if IVIG is ineffective.
- Steroids are not effective.
- Supportive care, such as respiratory support, prevention of decubiti in children with flaccid tetraplegia, nutritional support, pain management, prevention of deep vein thrombosis, and treatment of secondary bacterial infections, is important

- relapses usually respond to another course of plasmapheresis. Steroid and immunosuppressive drugs are another alternative, but their effectiveness is less predictable. High-dose pulsed methylprednisolone given intravenously is successful in some cases.
- The prognosis in chronic forms of the Guillain-Barré syndrome is more guarded than in the acute form, and many patients are left with major residual handicaps.
- Even if *C. jejuni* infection is documented, treatment of the infection is not necessary because it is self-limited,
  For the treatment of chronic neuropathic pain following GuillainBarré syndrome, gabapentin is more effective than carbamazepine

- **PROGNOSIS**
  The clinical course is usually benign, spontaneous recovery begins within 2-3 wk.
- Most patients regain full muscular strength, although some are left with residual weakness.
- The tendon reflexes are usually the last function to recover. Improvement usually follows a gradient opposite the direction of involvement: bulbar function recovering first,
  and lower extremity weakness resolving last.
- Bulbar and respiratory muscle involvement can lead to death if the syndrome is not recognized and treated. Although prognosis is generally good and the majority of children recover completely,

3 clinical features are predictive of poor outcome with sequelae:

- cranial nerve involvement,
- intubation
- maximum disability at the time of presentation.

- Easy fatigue is one of the most common chronic symptoms,
- Most patients with the axonal form of Guillain-Barré syndrome had a slow recovery over the 1st 6 mo and could eventually walk, although some required years to recover.
- EMG and NCV studies do not necessarily predict the long-term outcome