

Guillain-Barré Syndrome

- ***Guillain-Barré syndrome (GBS)*** is a monophasic disease involving only myelinated nerves in the PNS.
- The most common form (>80%) , called *acute inflammatory demyelinating polyneuropathy (AIDP)*, appears to be due to an immune-mediated attack of peripheral myelin.
- *Acute motor axonal neuropathy (AMAN)* is more severe clinically than AIDP. The immune attack in AMAN appears directed against the PNS myelinated axon exposed at the node of Ranvier.
- The annual incidence of GBS about 1 to 2 cases per 100,000 persons. The incidence increases with age, and males slightly predominate.

Pathophysiology

- GBS occurs in the setting of an *antecedent illness* in about 60% of patients.
- Upper respiratory and gastroenterological infections are frequent, with the most common being *viruses* (cytomegalovirus and Epstein–Barr) and *bacteria* (*Campylobacter jejuni*).
- It is proposed that via *molecular mimicry* the patient develops an immune response against the infecting agent that *cross-reacts* with antigens on the patient's peripheral nerve myelin or axons.

Clinical Manifestations

- GBS manifests as rapidly evolving *areflexic motor paralysis* with or without sensory disturbance.
- The usual pattern is an *ascending paralysis*.
- Weakness typically evolves over hours to a few days and is frequently accompanied by tingling dysesthesias in the extremities.
- The legs are usually more affected than the arms, and *facial diparesis* is present in 50% of affected individuals.
- The lower cranial nerves are also frequently involved, causing bulbar weakness with difficulty handling secretions and maintaining an airway.
- Pain in the neck, shoulder, back, or diffusely over the spine is also common in the early stages of GBS, occurring in ~50% of patients.

- Most patients require hospitalization, and almost 30% require ventilatory assistance at some time during the illness.
- *Deep tendon reflexes* attenuate or disappear within the first few days of onset.
- Cutaneous sensory deficits (e.g., loss of pain and temperature sensation) are usually relatively mild, but functions subserved by *large sensory fibers*, such as deep tendon reflexes and proprioception, are more severely affected.
- Once clinical worsening stops and the patient reaches a *plateau* (almost always within 4 weeks of onset), further progression is unlikely.

- *Autonomic involvement is common* and may occur even in patients whose GBS is otherwise *mild*. The usual manifestations are loss of vasomotor control with **wide fluctuation in blood pressure, postural hypotension, and cardiac dysrhythmias**.
- Pain is another common feature of GBS; in addition to the acute pain described above, a deep aching pain may be present in weakened muscles that patients liken to having *overexercised* the previous day. Other pains in GBS include dysesthetic pain in the extremities as a manifestation of sensory nerve fiber involvement.

Variants

- **AMAN** is a variant of GBS. There is *motor axonal* degeneration and *little or no demyelination* or inflammation. Despite the axonal involvement, recovery is similar to the demyelinating form.
- The **Miller Fisher syndrome**: is characterized by *gait ataxia, areflexia, and ophthalmoparesis*. It is considered a variant of GBS because;
 - it is often preceded by respiratory infection,
 - it progresses for weeks and then improves,
 - and CSF protein content is increased.

There is no limb weakness, however, and nerve conduction studies are generally normal.

Major Laboratory Findings

- Major blood tests are normal.
- The CSF becomes abnormal during the first week.
 - CSF protein elevates to levels of 100 to 400 mg/dL *but*,
 - CSF has a normal glucose level and normal WBC count (*albuminocytological dissociation*).
- Neuroimaging of the spinal cord *should be* normal.
- Nerve-conduction study results become abnormal in AIDP by the end of the first week.
 - conduction blockage in the majority of motor axons.
 - The motor nerve conduction velocity reduces, reflecting the segmental demyelination.
 - Variable evidence of muscle denervation may be found on electromyography beginning after 2 to 3 weeks.

Principles of Management and Prognosis

- The key to successful management is excellent nursing care.
- Patients usually require hospitalization and placement in a *critical care* setting. About 1/4 of patients require *a ventilator*.
- *Cardiac monitoring* is recommended because *patients may be prone to severe arrhythmias* that may require electrical cardioversion and medication.

➤ *Plasmaphoresis* or *human immune globulin* is beneficial if given early in the course of AIDP.

Both equally *shorten the time to recovery* and likely *prevent progression* of disease to more severe stages.

➤ In contrast, *use of corticosteroids* is **not beneficial**.

➤ During recovery, *physical therapy* often improves function.

- In mild cases of AIDP, motor recovery can occur *over a few weeks*. For AIDP patients who cannot walk, ambulation often *takes 4 to 6 months*. In severe cases, recovery may continue for *up to 2 years*.
- In AIDP, about **85%** of patients fully recover and **15%** are left with minor sequelae such as loss of reflexes.
- Death following cardiac arrhythmias or infectious complications still occurs in 2% to 3% of patients with GBS.

Bell's Palsy

Bell's palsy or *idiopathic peripheral facial nerve palsy*

- is the most common cause of CN VII dysfunction.
- It has an equal sex distribution.
- Cases occur in all ages, but the incidence increases with age.
- It is rare for Bell's palsy to be bilateral or to recur.

Pathophysiology

- The pathogenesis of Bell's palsy remains poorly understood. MRI and pathologic studies show the facial canal as the site of pathology. The nerve becomes edematous with varying amounts of surrounding inflammation.
- Early theories suggested *ischemia* to the facial nerve led to nerve edema and nerve compression from the walls of the facial canal. Later, the ischemia concept was *dropped* and the nerve edema was considered idiopathic.
- Recently, *viral infection theories* have focused on *varicella-zoster* and *herpes simplex viruses* as potential viruses that reactivate in the facial nerve to cause nerve damage, edema, and inflammation.

Major Clinical Features

- The onset of Bell's palsy is fairly *abrupt*, maximal weakness being attained by *48 h* as a general rule.
- *Pain behind the ear* may precede the paralysis for a *day or two*.
- Complete interruption of the facial nerve paralyzes all muscles of facial expression,
 - The corner of the mouth droops,
 - the creases and skin folds are effaced,
 - the forehead is unfurrowed,
 - and the eyelids will not close.
 - Upon attempted closure of the lids, the eye on the paralyzed side rolls upward (*Bell's phenomenon*).

- The lower lid sags and falls away from the conjunctiva, permitting tears to spill over the cheek.
- Food collects between the teeth and lips, and saliva may dribble from the corner of the mouth.
- The patient complains of a heaviness or numbness in the face, but sensory loss is rarely demonstrable.
- Taste sensation may be lost unilaterally, and hyperacusis may be present.

Differential Diagnosis

- There are many other causes of acute facial palsy that must be considered in the differential diagnosis of Bell's palsy.
- *Lyme disease* can cause unilateral or bilateral facial palsies.
- *Ramsay Hunt syndrome*, caused by reactivation of herpes zoster in the geniculate ganglion, consists of a severe facial palsy associated with a vesicular eruption in the external auditory canal.
- in *sarcoidosis* and *Guillain-Barré syndrome* Facial palsy is often bilateral.
- *Acoustic neuromas* frequently involve the facial nerve by local compression.
- *Infarcts, demyelinating lesions of multiple sclerosis, and tumors* are the common pontine lesions that interrupt the facial nerve fibers; other signs of brainstem involvement are usually present.

- All these forms of nuclear or peripheral facial palsy must be distinguished from the *supranuclear* type.
- In the latter, **the frontalis and orbicularis oculi muscles** are *involved less* than those of the lower part of the face, since the upper facial muscles are innervated by corticobulbar pathways from both motor cortices, whereas the lower facial muscles are innervated only by the opposite hemisphere.
- In supranuclear lesions there may be *a dissociation* of emotional and voluntary facial movements and often some degree of paralysis of the arm and leg, or an aphasia (in dominant hemisphere lesions) is present

Major Laboratory Findings

- Remarkably few laboratory abnormalities exist in Bell's palsy. The patient has a normal hemogram, erythrocyte sedimentation rate, and serum electrolytes.
- The CSF is normal. If the CSF has a pleocytosis, the facial palsy etiology is likely due to an inflammatory or infectious process, such as varicella-zoster virus, Lyme disease or sarcoidosis.
- Cranial MRI with gadolinium may show enhancement of the facial nerve within the facial canal.
- The EMG, normal for the first 3 days, shows a steady decline in activity and after 10 days, denervation potentials begin to appear.

Principles of Management and Prognosis

Symptomatic measures include:

- (1) the use of paper tape to depress the upper eyelid during sleep and prevent corneal drying.
 - (2) massage of the weakened muscles.
- A course of *glucocorticoids, given as prednisone 60–80 mg daily during the first 5 days* and then *tapered over the next 5 days*, appears to shorten the recovery period and modestly improve the functional outcome.
 - *combination therapy* with *prednisone plus valacyclovir (1000 mg daily for 5–7 days)* may be *marginally better* than prednisone alone, especially in patients with severe clinical presentations.

- If there is incomplete paralysis of facial muscles, there is an excellent prognosis for full to satisfactory recovery that spontaneously occurs within 2 months.
- Should the facial paralysis be complete, full to satisfactory recovery spontaneously occurs in about 80% of patients over 1 to 3 months.