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# GUILLAIN-BARRE SYNDROME: A COMPREHENSIVE REVIEW

**Nursing**

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## ABSTRACT

**GUILLIAN–BARRE SYNDROME**

Guillain-Barré syndrome is that the commonest and most severe acute paralytic neuropathy, with about 100000 people developing the disorder per annum worldwide. Under the umbrella term of Guillain-Barré syndrome are several recognizable variants with distinct clinical and pathological features. The severe, generalized manifestation of Guillain-Barré syndrome with respiratory failure affects 20–30% of cases. Treatment with intravenous immunoglobulin or plasma exchange is that the optimal management approach, alongside supportive care. Understanding of the infectious triggers and immunological and pathological mechanisms has advanced substantially within the past 10 years, and is guiding clinical trials investigating new treatments. Unfortunately, morbidity and mortality rates are still high despite the present understanding of the pathophysiology and available treatment options. Investigators of giant, worldwide, collaborative studies of the spectrum of Guillain-Barre syndrome are accruing data for clinical and biological databases to inform the event of outcome predictors and disease biomarkers. Such studies are transforming the clinical and scientific landscape of acute autoimmune neuropathies.

## KEYWORDS

## INTRODUCTION

One of the earliest descriptions of what we know today as Guillain-Barré syndrome is found in Landry's report on 10 patients with "ascending paralysis" in 1859. In 1916 Guillain, Barré, and Strohl described two French soldiers with motor weakness, are flexia, and "albuminocytological dissociation" in the cerebrospinal fluid. Subsequently several cases with similar manifestations were reported, and this clinical entity was named after Guillain and Barré. Later, different types of the syndrome with characteristic clinical features were identified. This distinction is possible today on the basis of clinical features, aetiology, and electrophysiological characteristics.

### Definition

Guillain-Barré syndrome (GBS) also known as landry's palsy and has rapid onsent of muscle weakness and non-polio acute flaccid paralytic illness . GBS is an acute, monophasic, symmetrically progressive, peripheral ascending demyelinating polyneuropathy characterized by rapidly evolving symmetrical limb weakness, are flexia, absent or mild sensory signs, and variable autonomic disturbances.

### Causes

The exact cause of Guillain-Barre syndrome isn't known. The disorder usually appears days or weeks after a respiratory or digestive tract infection. Rarely, recent surgery or vaccination can trigger Guillain-Barre syndrome. Recently, there have been cases reported following infection with the Zika virus.

In Guillain-Barre syndrome, your immune system — which usually attacks only invading organisms — begins attacking the nerves. In AIDP, the most common form of Guillain-Barre syndrome in the U.S., the nerves' protective covering (myelin sheath) is damaged. The damage prevents nerves from transmitting signals to your brain, causing weakness, numbness or paralysis.

### Risk Factors

Guillain-Barre syndrome can affect all age groups. But your risk increases as you age. It's also more common in males than females.

Guillain-Barre syndrome may be triggered by:

- Most commonly, infection with campylobacter, a type of bacteria often found in undercooked poultry
- Influenza virus
- Cytomegalovirus
- Epstein-Barr virus
- Zika virus
- Hepatitis A, B, C and E
- HIV, the virus that causes AIDS
- Mycoplasma pneumonia
- Surgery
- Trauma
- Hodgkin's lymphoma
- Rarely, influenza vaccinations or childhood vaccinations

## PATHOGENESIS

Multiple antecedent and potentially triggering events have been reported. The association with infections is established in not only C jejuni but also cytomegalovirus, Epstein-Barr virus, influenza A, Mycoplasma pneumoniae, Haemophilus influenzae, hepatitis (A, B, and E). and Zika virus. The risk of GBS from influenza vaccine varies from 3 cases per million to as low as zero. Surgery may predispose patients to GBS (more likely in patients with prior malignant or autoimmune disorders) but is exceedingly rare in our experience. Guillain-Barré syndrome is often a postinfectious, immune-mediated nerve injury. Three phenotypes are likely purely demyelinating, purely axonal, and demyelinating with axonal involvement. Immunopathogenesis differs in each of these conceptual models, and outcome (possibly a response to treatment) is also different. The current working hypothesis of GBS immunopathogenesis is depicted in Figure 1.

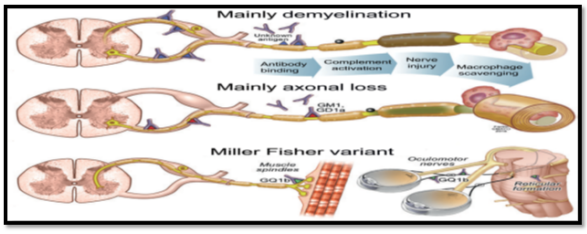


**FIGURE 1.** Current understanding of Guillain-Barré syndrome pathogenesis and clinical variants. In demyelinating Guillain-Barré syndrome, unequivocal antigens have yet to be identified but are inferred by complement activation, myelin destruction, and cleanup by macrophages. In axonal and Miller Fisher variants, specific gangliosides (GM1, GD1a, GQ1b) are targeted by immunoglobulins and share antigenic epitopes with various bacterial and viral antigens. These antigenic targets are at nodal structures, at roots,and located at the end organs. In Miller Fisher syndrome, the GQ1b antigen also exists within the brain stem. In this variant, the macrophages clean up the axon debris and come in from the nodes.

Although both elements of the immune response (T cells and B cells) play a role, current understanding holds that GBS is antibody mediated. Not all antigangliose antibodies are neurotoxic, but antibodies binding to GM1 or GD1a gangliosides (at nodes of Ranvier) activate myelin-destroying complement. The predominance of motor axonal involvement has led to the designation acute motor axonal neuropathy. Campylobacter jejuni infection is the main known

instigator of this mechanism, and molecular mimicry between C jejuni lipooligosaccharide and GM1 and GD1a has been found. For patients presenting with the ataxic sensory variant, the most commonly identified ganglioside antigen target is GQ1b. Another motor-sensory axonal form of GBS (acute motor-sensory axonal neuropathy) can also be grouped under a more general term, axonal GBS. The target molecule in purely demyelinating disease is yet unknown. Despite this differentiation into subtypes, treatment is similar. Prognosis is worse with a more protracted course in axonal forms and more long-term disability.

**Clinical features of Guillain-Barre syndrome**

- Motor dysfunction

Symmetrical limb weakness: proximal, distal or global
Neck muscle weakness
Respiratory muscle weakness
Cranial nerve palsies: III–VII, IX–XII
Areflexia
Wasting of limb muscles

- Sensory dysfunction

Pain
Numbness, paraesthesiae
Loss of joint position sense, vibration, touch and pain distally
Ataxia

- Autonomic dysfunction

Sinus tachycardia and bradycardia
Other cardiac arrhythmias (both tachy and brady)
Hypertension and postural hypotension
Wide fluctuations of pulse and blood pressure
Tonic pupils
Hypersalivation
Anhydrosis or excessive sweating
Urinary sphincter disturbances
Constipation
Gastric dysmotility
Abnormal vasomotor tone causing venous pooling and facial flushing

- Other

Papilloedema

**INVESTIGATIONS**

**Serum biochemistry**

Urea and electrolytes are usually normal but may have evidence of the syndrome of inappropriate ADH secretion (SIADH) or renal dysfunction. ALT and gamma GT may be raised in 33% of patients. Creatine kinase may be raised.

**Inflammatory Markers**

Erythrocyte sedimentation rate is usually raised and C - reactive protein is sometimes elevated.

**Anti-ganglioside antibodies**

Anti-GM1 is positive in 25% of patients and is associated with a worse outcome. Anti-GD1a is associated with AMAN subtype of GBS. Anti-GQ1b is associated with Miller-Fisher syndrome.

**Infection screen**

Serology tests for *Campylobacter jejuni*, Cytomegalovirus, Epstein-Barr virus, Herpes simplex virus, Mycoplasma pneumonia, HIV antibodies should be considered. Stool cultures looking for evidence of gastrointestinal infections particularly *Campylobacter jejuni*.

**Radiological**

A CT brain is indicated to exclude other causes of symptoms and evidence of raised intracranial pressure prior to performing a lumbar puncture. An MRI of the spine may show selective anterior spinal nerve root enhancement with gadolinium and will exclude cervical nerve impingement.

**Lumbar puncture**

Increased protein levels and cell levels in CSF are indicative of GBS

**Nerve Conduction Studies**

Findings depend on subtype of GBS. The majority show demyelinating pattern while some patients may show evidence of axonal loss with little or no demyelination.

**Respiratory function tests**

These may show reduced vital capacity, maximal inspiratory and expiratory pressures. Arterial blood gases may indicate progressive respiratory failure.

**Differential diagnosis of Guillain–Barre syndrome**

The differential diagnosis of Guillain–Barré syndrome is broad and highly dependent on the clinical features of the individual patient. Here, we present an overview of the most important differential diagnoses categorized by location in the nervous system.

**CNS**

- Inflammation or infection of the brainstem (for example, sarcoidosis, Sjögren syndrome, neuromyelitis optica or myelin oligodendrocyte glycoprotein antibody-associated disorder)[a]
- Inflammation or infection of the spinal cord (for example, sarcoidosis, Sjögren syndrome or acute transverse myelitis)
- Malignancy (for example, leptomeningeal metastases or neurolymphomatosis)
- Compression of brainstem or spinal cord
- Brainstem stroke
- Vitamin deficiency (for example, Wernicke encephalopathy[a], caused by deficiency of vitamin B1, or subacute combined degeneration of the spinal cord, caused by deficiency of vitamin B12)

**Anterior Horn Cells**

- Acute flaccid myelitis (for example, as a result of polio, enterovirus D68 or A71, West Nile virus, Japanese encephalitis virus or rabies virus)

**Nerve Roots**

- Infection (for example, Lyme disease, cytomegalovirus, HIV, Epstein–Barr virus or varicella zoster virus)
- Compression
- Leptomeningeal malignancy

**Peripheral Nerves**

- Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)
- Metabolic or electrolyte disorders (for example, hypoglycaemia, hypothyroidism, porphyria or copper deficiency)
- Vitamin deficiency (for example, deficiency of vitamins B1 (also known as beriberi), B12 or E)
- Toxins (for example, drugs, alcohol, vitamin B6, lead, thallium, arsenic, organophosphate, ethylene glycol, diethylene glycol, methanol or N-hexane)
- Critical illness polyneuropathy
- Neuralgic amyotrophy
- Vasculitis
- Infection (for example, diphtheria or HIV)

**Neuromuscular Junction**

- Myasthenia gravis
- Lambert–Eaton myasthenic syndrome
- Neurotoxins (for example, botulism, tetanus, tick paralysis or snakebite envenomation)
- Organophosphate intoxication

**Muscles**

- Metabolic or electrolyte disorders (for example, hypokalaemia, thyrotoxic hypokalaemic periodic paralysis, hypomagnesaemia or hypophosphataemia)
- Inflammatory myositis
- Acute rhabdomyolysis
- Drug-induced toxic myopathy (for example, induced by colchicine, chloroquine, emetine or statins)
- Mitochondrial disease

**Other**

- Conversion or functional disorder

**MANAGEMENT**

There is currently no cure for GBS, but medications are available to ease symptoms.

Guillain-Barré is an autoimmune inflammatory process that's self-limiting, means it will resolve on its own. However, anyone with this

condition should be admitted to a hospital for close observation. The symptoms can quickly worsen and can be fatal if untreated.

- In severe cases, people with Guillain-Barré can develop full-body paralysis. Guillain-Barré can be life-threatening if paralysis affects the diaphragm or chest muscles, preventing proper breathing.
- The goal of treatment is to lessen the severity of the immune attack and support your body functions, such as lung function, while your nervous system recovers.
- Treatments may include:

**1) Plasmapheresis (plasma exchange)**

- The immune system produces proteins called antibodies that normally attack harmful foreign substances, such as bacteria and viruses. Guillain-Barré occurs when your immune system mistakenly makes antibodies that attack the healthy nerves of your nervous system.
- **Plasmapheresis** is intended to remove the antibodies attacking the nerves from your blood.
- During this procedure, blood is removed from your body by a machine. This machine removes the antibodies from your blood and then returns the blood to your body.

**2) Intravenous Immunoglobulin**

- High doses of immunoglobulin can also help block the antibodies causing Guillain-Barre. Immunoglobulin contains normal, healthy antibodies from donors.
- Plasmapheresis and intravenous immunoglobulin are equally effective. It's up to you and your doctor to decide which treatment is best.

**3) Other treatments**

- Gave medication to relieve pain and prevent blood clots while you're immobile.
- Physical and occupational therapy. During the acute phase of the illness, caregivers will manually move your arms and legs to keep them flexible.
- Once you begin to recover, therapists will work with you on muscle strengthening and a range of activities of daily living (ADLs). This can include personal care activities, like getting dressed.
- Breathing support or a breathing tube and ventilator, if the diaphragm is weak.
- Proper body positioning or a feeding tube to prevent choking during feeding, if the muscles used for swallowing are weak.

**Prognosis**

After the acute phase of illness, Guillain-Barre syndrome (GBS) patients tend to do well. More than 80% achieve independent ambulation after 6 months. Mortality during the acute phase of the illness is less than 5%. However, there is a subset of patients, less than 20%, who continue to have significant disability despite receiving the standard of care for GBS. Studies are underway to try to identify these patients early. Early identification of poor prognostic factors could lead to trials of further treatment specific to this subgroup. In a cohort of Dutch patients, a prognostic tool, Erasmus GBS Outcome score, utilizes the patient's physical examination, age, and presence of diarrhea to predict the patient's ability to walk in the near future. Patients with a significant likelihood of residual disability would be most amenable to further therapeutic trials.

Studies to assess whether plasma exchange followed by IVIG would have an additional benefit were not significant. Additionally, a number of studies regarding the addition of corticosteroids to IVIG were also not significant; however, there may have been a benefit in patients with worse prognostic factors (such as age and GBS disability score). According to a small case series, two courses of IVIG has been suggested as a possible intervention. There may be some limitations of IVIG use based on adverse effects, however. Currently, there is an ongoing randomized controlled trial of 2 courses of IVIG with patients with refractory GBS. Additionally, there is much interest in the key role of complement activation in the pathogenesis of GBS; therefore, a randomized controlled trial of eculizumab in patients with GBS is being studied.

Other clinical features have been shown to predict the need for ventilation during the illness. These include fulminant course (onset to admission < 7 days), bulbar weakness, and neck flexion weakness. These predictive factors would suggest triage to an intensive care unit rather than a stepdown unit.

Following recovery, patients may continue to contend with residual fatigue, pain, and paresthesias for up to several years. (Evidence level III)

**REFERENCES**


1. Seneviratne U. Guillain-Barré syndrome. Postgrad Med J. 2000 Dec 1;76(902):774–82.
2. Sarkar UK, Menon L, Sarbapalli D, Pal R, Zaman FA, Kar S, et al. Spectrum of Guillain—Barré syndrome in tertiary care hospital at Kolkata. J Nat Sci Biol Med [Internet]. 2011 [cited 2020 Jul 16];2(2):211–5. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3276017/
3. Guillain-Barre syndrome - Symptoms and causes [Internet]. Mayo Clinic. [cited 2020 Jul 15]. Available from: https://www.mayoclinic.org/diseases-conditions/guillain-barre-syndrome/symptoms causes/syc-20362793
4. Wijdicks EFM, Klein CJ. Guillain-Barré Syndrome. Mayo Clin Proc. 2017 Mar 1;92(3):467–79.
5. Tandel H, Vanza J, Pandya N, Jani P. Guillain-barré syndrome (GBS): A Review. Curr Med Res Opin. 2016 Jan 31;3:366–71.
6. Leonhard SE, Mandarakas MR, Gondim FAA, Bateman K, Ferreira MLB, Cornblath DR, et al. Diagnosis and management of Guillain–Barré syndrome in ten steps. Nat Rev Neurol. 2019;15(11):671–83.
7. Guillain-Barré syndrome: Symptoms, causes, diagnosis, and treatment [Internet]. 2017 [cited 2020 Jun 30]. Available from: https://www.medicalnewstoday.com/articles/167892
8. Nguyen TP, Taylor RS. Guillain Barre Syndrome. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2020 [cited 2020 Jun 30]. Available from: http://www.ncbi.nlm.nih.gov/books/NBK532254/