Guillain-Barre Syndrome

Waleeja Nasir

1st Year MBBS, Islamabad Medical and Dental College, Islamabad, Pakistan.

Key points:

- Overview & clinical presentation
- Pathophysiology
- Diagnostic methods
- Treatment options
- Prognosis
- Conclusion

Introduction

Guillain-Barré syndrome was first discovered more than a century ago.[1] It is a rare autoimmune neurological disease that affects the peripheral nerves and causes muscle weakness. It is further characterized by a high rate of morbidity and mortality. Guillain Barré syndrome (GBS) is the most frequent, rapidly progressive, severe, and potentially lethal form of polyneuropathy.[2]

Subtypes

Guillain-Barré syndrome can be divided into subtypes, mainly:

- AIDP (Acute inflammatory demyelinating polyradiculoneuropathy)
- MFS (Miller Fisher syndrome)
- AMAN (Acute motor axonal neuropathy)
- AMSAN (Acute motor-sensory axonal neuropathy)

The most common subtypes are acute inflammatory polyradiculoneuropathy (AIDP) and acute motor axonal neuropathy (AMAN). Approximately 90% of people with GBS in North America and Europe have AIDP.[3] The most common symptom of AIDP is muscle weakness that originates in the lower extremities of the body and gradually spreads upward.

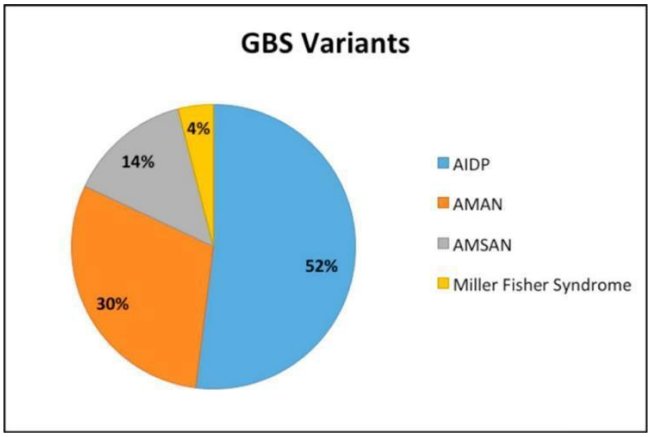

Figure 1: *GBS Variants: AIDP - acute inflammatory demyelinating polyneuropathy, AMAN - acute motor axonal neuropathy, AMSAN - acute motor-sensory polyneuropathy* https://nsj.org.sa/content/25/5/369

Clinical presentation

As GBS affects the peripheral nerves of the body (polyneuropathy) and since our nerves control our bodily functions and movements, people with GBS generally experience:

- Breathing difficulties: Weakness or paralysis may extend to the muscles controlling breathing which may be fatal.
- Blood pressure fluctuations and cardiac arrhythmias.
- Pain may be experienced.
- Bowel and bladder function problems may arise.

- Blood clots: Immobility associated with GBS increases the risk of developing blood clots.
- Pressure sores: Immobility raises the chances of developing bedsores.
- Relapse: A small percentage of individuals may experience muscle weakness even years after symptom resolution.
- Rarely, complications such as respiratory distress syndrome and heart attacks may result in death.

Patients with the AIDP subtype of GBS typically have weakness that starts in the legs and spreads to the arms, as well as decreased or absent reflexes. In more than 50% of these patients, nerves that originate in the brain stem (cranial nerves) are affected, which may cause facial weakness, difficulty swallowing, and eye muscle weakness or paralysis. Approximately 25% to 30% of patients develop severe weakness or paralysis of the muscles used to breathe. GBS commonly causes symptoms of low back pain and limb numbness and tingling, and fluctuations in blood pressure or an irregular heart rhythm can also occur. [3]

Causes

The exact cause of the disease is unknown. Approximately two-thirds of patients have diarrheal or respiratory illness within 4 to 6 weeks prior to the onset of GBS symptoms. Other, less common events or conditions that may trigger GBS include recent surgery, pregnancy, and immunosuppression.[3] This condition's low prevalence but high morbidity and mortality, variable atypical patient presentations, and challenging diagnosis make it a high-risk and low-prevalence disease.[4]

Risk factors

GBS affects people worldwide, and the lifetime risk of GBS is estimated at 1 in 1000. Although individuals of any age can develop GBS, the incidence increases with age, and males are slightly more likely to develop GBS than females.[3]

Pathophysiology

GBS is an autoimmune disease- when your body's nervous system attacks itself. Patients with infections linked to GBS frequently produce antibodies against the gangliosides of the human peripheral nerves. This results in the entry of viruses or bacteria into the body. Due to molecular mimicry, these antibodies target various lipo- oligosaccharides in microbes and structurally similar gangliosides in the brain. Target on gangliosides by the antibodies leads to demyelination of neurons, leading to loss of neurons that leads to GBS.[5]

When the myelin sheath, the protective covering of neurons, sustains damage, the damage interferes with the transmission of signals to the brain, resulting in symptoms such as weakness, numbness, or paralysis.

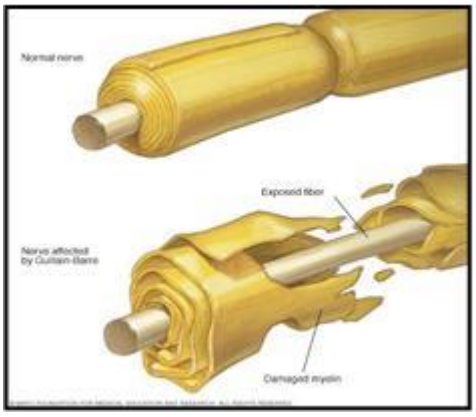

Figure 2: *Normal nerve vs. nerve affected by GB* https://www.mayoclinic.org/diseases- conditions/guillain-barre- syndrome/symptomscauses/syc-20362793

Diagnostic methods

The diagnosis is challenging and often subtle, as only 25–30% of patients are diagnosed on their initial healthcare visit. Clinicians should consider GBS in patients with progressive ascending weakness involving the lower extremities associated with hyporeflexia, although the cranial nerves, respiratory system, and autonomic system may be involved.[4]

The diagnosis of GBS relies upon a combination of clinical features, often with support of electrodiagnostic and laboratory features. Most diagnostic criteria for GBS require a combination of history, neurological examination, cerebrospinal fluid (CSF) and electrodiagnostic results.[6]

Requirements for the diagnosis of sensory-motor or motor GBS are:

- Progressive weakness of arms and legs.
- Absent or decreased deep tendon reflexes in affected limbs.
- Progressive worsening for no more than 4 weeks.[6]

GBS is a spectrum, and most cases have limb weakness and fulfil the requirements for the diagnosis. Some GBS variants and MFS do not fulfil all requirements for GBS since there is not always progressive weakness of arms and legs. In some patients, the deep tendon reflexes initially can be normal or hyperactive.[6]

Treatment options

While there is currently no cure for Guillain-Barré syndrome, treatments are available to accelerate recovery and alleviate the severity of the illness.

Current recommended treatments for GBS are intravenous immune globulin (IVIG), an infusion of antibodies, or plasma exchange, which involves removal and replacement of the liquid component of blood. About 40% to 50% of patients with GBS do not improve within 4 weeks after IVIG or plasma exchange and need prolonged supportive care. Physical, occupational, and speech therapy are important to help patients regain strength and function.[3]

Prognosis

Most patients with GBS gradually improve and can have a complete recovery over 6 to 12 months. However, some patients have residual symptoms, including fatigue, pain, numbness, tingling, and muscle weakness. Some factors associated with a higher risk of death due to GBS include older age, more severe disease, and need for mechanical ventilation.[3]

Conclusion

To summarize, Guillain-Barré syndrome presents a complex challenge, characterized by its autoimmune nature and diverse clinical symptoms. Diagnosing the disease relies on a combination of clinical assessment and diagnostic tests, while treatments like IVIG and plasmapheresis offer hope for symptom management. Despite the absence of a cure, ongoing research and therapeutic advancements contribute to improved outcomes. In the future, it's crucial for doctors, researchers, and patients to work together to better understand GBS and improve patient care.

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