



Research article

## GUILLAIN-BARRÉ SYNDROME

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### ABSTRACT

Guillain-Barré syndrome (GBS) is a rapid-onset muscle weakness caused by the immune system damaging the peripheral nervous system. During the acute phase, the disorder can be life-threatening with about 15% developing weakness of the breathing muscles requiring mechanical ventilation. Some are affected by changes in the function of the autonomic nervous system, which can lead to dangerous abnormalities in heart rate and blood pressure.

#### Key words:

Auto Immune disorder, peripheral nervous system, ascending paralysis, immunoglobulins, plasmapheresis.

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## INTRODUCTION

### Definition

Guillain-Barré syndrome is a disorder characterized by progressive symmetrical paralysis and loss of reflexes, usually beginning in the legs. The paralysis characteristically involves more than one limb (most commonly the legs), is progressive, and is usually proceeds from the end of an extremity toward the torso. Areflexia (loss of reflexes) or hyporeflexia (diminution of reflexes) may occur in the legs and arms.

### Etiology

The cause is unknown. The underlying mechanism involves an autoimmune disorder in which the body's immune system mistakenly attacks the peripheral nerves and damages their myelin insulation. Sometimes this immune dysfunction is triggered by an infection or, less commonly, surgery or vaccination.

### Signs and Symptoms

The first symptoms of Guillain-Barré syndrome are numbness, tingling, and pain, alone or in combination. This is followed by weakness of the legs and arms that affects both sides equally and worsens over time. The weakness can take half a day to over two weeks to reach maximum severity, and then becomes steady. In one in five people, the weakness continues to progress for as long as four weeks. The muscles of the neck may also be affected, and about half experience involvement of the cranial nerves which supply the head and

face; this may lead to weakness of the muscles of the face, swallowing difficulties and sometimes weakness of the eye muscles. In 8%, the weakness affects only the legs (paraplegia or paraparesis). Involvement of the muscles that control the bladder and anus is unusual. In total, about a third of people with Guillain-Barré syndrome continue to be able to walk. Once the weakness has stopped progressing, it persists at a stable level ("plateau phase") before improvement occurs. The plateau phase can take between two days and six months, but the most common duration is a week. Pain-related symptoms affect more than half, and include back pain, painful tingling, muscle pain and pain in the head and neck relating to irritation of the lining of the brain.

Many people with Guillain-Barré syndrome have experienced the signs and symptoms of an infection in the 3–6 weeks prior to the onset of the neurological symptoms. This may consist of upper respiratory tract infection (rhinitis, sore throat) or diarrhea.

In children, particularly those younger than six years old, the diagnosis can be difficult and the condition is often initially mistaken (sometimes for up to two weeks) for other causes of pains and difficulty walking, such as viral infections, or bone and joint problems.

On neurological examination, characteristic features are the reduced power and reduced or absent tendon reflexes (hypo- or areflexia, respectively). However, a small proportion has normal reflexes in affected limbs before developing areflexia, and some may have exaggerated reflexes. The level of consciousness is normally unaffected in Guillain-Barré

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syndrome, but the Bickerstaff brainstem encephalitis subtype may feature drowsiness, sleepiness, or coma.

### ***Respiratory failure***

A quarter of all people with Guillain–Barré syndrome develop weakness of the breathing muscles leading to respiratory failure, the inability to breathe adequately to maintain healthy levels of oxygen and/or carbon dioxide in the blood. This life-threatening scenario is complicated by other medical problems such as pneumonia, severe infections, blood clots in the lungs and bleeding in the digestive tract in 60% of those who require artificial ventilation.

### ***Autonomic dysfunction***

The autonomic or involuntary nervous system, which is involved in the control of body functions such as heart rate and blood pressure, is affected in two thirds of people with Guillain–Barré syndrome, but the impact is variable. Twenty percent may experience severe blood-pressure fluctuations and irregularities in the heart beat, sometimes to the point that the heart beat stops and requiring pacemaker-based treatment. Other associated problems are abnormalities in perspiration and changes in the reactivity of the pupils. Autonomic nervous system involvement can affect even those who do not have severe muscle weakness.

### ***Diagnosis***

The diagnosis of Guillain–Barré syndrome depends on findings such as rapid development of muscle paralysis, absent reflexes, absence of fever, and a likely cause. Cerebrospinal fluid analysis (through a lumbar spinal puncture) and nerve conduction studies are supportive investigations commonly performed in the diagnosis of GBS. Testing for antiganglioside antibodies is often performed, but their contribution to diagnosis is usually limited.<sup>[4]</sup> Blood tests are generally performed to exclude the possibility of another cause for weakness, such as a low level of potassium in the blood. An abnormally low level of sodium in the blood is often encountered in Guillain–Barré syndrome. This has been attributed to the inappropriate secretion of antidiuretic hormone, leading to relative retention of water.

In many cases, magnetic resonance imaging of the spinal cord is performed to distinguish between Guillain–Barré syndrome and other conditions causing limb weakness, such as spinal cord compression. If an MRI scan shows enhancement of the nerve roots, this may be indicative of GBS. In children, this feature is present in 95% of scans, but it is not specific to Guillain–Barré syndrome, so other confirmation is also needed.

### ***Spinal fluid***

Cerebrospinal fluid envelops the brain and the spine, and lumbar puncture or spinal tap is the removal of a small amount of fluid using a needle inserted between the lumbar vertebrae. Characteristic findings in Guillain–Barré syndrome are an elevated protein level, usually greater than 0.55 g/L, and fewer than 10 white blood cells per cubic millimeter of fluid ("albuminocytological dissociation"). This combination distinguishes Guillain–Barré syndrome from other conditions (such as lymphoma and poliomyelitis) in which both the protein and the cell count are elevated. Elevated CSF protein

levels are found in approximately 50% of patients in the first 3 days after onset of weakness, which increases to 80% after the first week.

Repeating the lumbar puncture during the disease course is not recommended. The protein levels may rise after treatment has been administered.

### ***Neurophysiology***

Directly assessing nerve conduction of electrical impulses can exclude other causes of acute muscle weakness, as well as distinguish the different types of Guillain–Barré syndrome. Needle electromyography (EMG) and nerve conduction studies may be performed. In the first two weeks, these investigations may not show any abnormality. Neurophysiology studies are not required for the diagnosis.

### ***Treatment***

#### ***Immunotherapy***

Plasmapheresis and intravenous immunoglobulins (IVIG) are the two main immunotherapy treatments for GBS. Plasmapheresis attempts to reduce the body's attack on the nervous system by filtering antibodies out of the bloodstream. Similarly, administration of IVIG neutralizes harmful antibodies and inflammation. These two treatments are equally effective, but a combination of the two is not significantly better than either alone. Plasmapheresis speeds recovery when used within four weeks of the onset of symptoms. IVIG works as well as plasmapheresis when started within two weeks of the onset of symptoms, and has fewer complications. IVIG is usually used first because of its ease of administration and safety. Its use is not without risk; occasionally it causes liver inflammation, or in rare cases, kidney failure. Glucocorticoids alone have not been found to be effective in speeding recovery and could potentially delay recovery.

#### ***Respiratory failure***

Respiratory failure may require intubation of the trachea and breathing support through mechanical ventilation, generally on an intensive care unit. The need for ventilatory support can be anticipated by measurement of two spirometry-based breathing tests: the forced vital capacity (FVC) and the negative inspiratory force (NIF). An FVC of less than 15 ml per kilogram body weight or an NIF of less than 60 cmH<sub>2</sub>O are considered markers of severe respiratory failure.

#### ***Pain***

While pain is common in people with Guillain–Barré syndrome, studies comparing different types of pain medication are insufficient to make a recommendation as to which should be used.

#### ***Rehabilitation***

Following the acute phase, around 40% of people require intensive rehabilitation with the help of a multidisciplinary team to focus on improving activities of daily living (ADLs). Studies into the subject have been limited, but it is likely that intensive rehabilitation improves long-term symptoms. Teams may include physical therapists, occupational therapists, speech language pathologists, social workers, psychologists, other allied health professionals and nurses. The team usually

works under the supervision of a neurologist or rehabilitation physician directing treatment goals.

Physiotherapy interventions include strength, endurance and gait training with graduated increases in mobility, maintenance of posture and alignment as well as joint function. Occupational therapy aims to improve everyday function with domestic and community tasks as well as driving and work. Home modifications, gait aids, orthotics and splints may be provided. Speech-language pathology input may be required in those with speech and swallowing problems, as well as to support communication in those who require ongoing breathing support (often through a tracheostomy). Nutritional support may be provided by the team and by dietitians. Psychologists may provide counseling and support. Psychological interventions may also be required for anxiety, fear and depression.

### Prognosis

Guillain–Barré syndrome can lead to death as a result of a number of complications: severe infections, blood clots, and cardiac arrest likely due to autonomic neuropathy. Despite optimum care this occurs in about 5% of cases.

There is a variation in the rate and extent of recovery. The prognosis of Guillain–Barré syndrome is determined mainly by age (those over 40 may have a poorer outcome), and by the severity of symptoms after two weeks. Furthermore, those who experienced diarrhea before the onset of disease have a worse prognosis. On the nerve conduction study, the presence of conduction block predicts poorer outcome at 6 months. In those who have received intravenous immunoglobulins, a smaller increase in IgG in the blood two weeks after administration is associated with poorer mobility outcomes at six months than those whose IgG level increased substantially. If the disease continues to progress beyond four weeks, or there are multiple fluctuations in the severity (more than two in eight weeks), the diagnosis may be chronic inflammatory demyelinating polyneuropathy, which is treated differently.

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