



## Gillian-Barrè Syndrome

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### Introduction

Guillain-Barre Syndrome (GBS) is a serious disorder that occurs when the body's defense (immune) system mistakenly attacks part of the nervous system. This leads to nerve inflammation that causes muscle weakness. The term Guillain-Barré syndrome defines a recognisable clinical entity that is characterised by rapidly evolving symmetrical limb weakness, loss of tendon reflexes, absent or mild sensory signs, and variable autonomic dysfunctions. Since the virtual elimination of poliomyelitis, GBS has become the leading cause of acute flaccid paralysis in western countries.<sup>1</sup> The condition, however, occurs worldwide, affecting patients of all ages and both sexes. In the majority of cases, the neuropathy is triggered by a bacterial or viral illness. Weakness can develop acutely (within days) or subacutely (up to 4 weeks) and reaches a plateau, with subsequent spontaneous resolution of paralysis. Although the pathogenesis of GBS remains incompletely defined, there is increasing support for the concept that GBS results from an aberrant organspecific immune response.<sup>2</sup>

### Causes, incidence, and risk factors

GBS is an autoimmune disorder (the body's immune system attacks itself). Exactly what triggers GBS is unknown. The syndrome may occur at any age, but is most common in people of both sexes between ages 30 and 50. It often follows a minor infection, such as a lung infection or gastrointestinal infection. Most of the time, signs of the original infection have disappeared before the symptoms of Guillain-Barre begin. The swine flu vaccination in 1976 may have caused rare cases of GBS. However, the swine flu and the regular flu vaccines used today have not resulted in more cases of the illness. GBS damages parts of nerves. This nerve damage causes tingling, muscle weakness, and paralysis. GBS most often affects the nerve's covering myelin sheath. Such damage is called demyelination, and it causes nerve signals to move more slowly. Damage to other parts of the nerve can cause the nerve to stop working altogether. Some people may get GBS after a

bacterial infection. A similar syndrome may occur after surgery, or when someone is critically ill (neuropathy of critical illness).

### Symptoms

GBS can get worse very quickly. It may take only a few hours to reach the most severe symptoms, but weakness that increases over several days is also common. Muscle weakness or the loss of muscle function (paralysis) affects both sides of the body. In most cases, the muscle weakness starts in the legs and then spreads to the arms. This is called ascending paralysis. Patients may notice tingling, foot or hand pain, and clumsiness. If the inflammation affects the nerves to the diaphragm and chest and there is weakness in those muscles, the person may need breathing assistance.

Typical symptoms include:

- Loss of reflexes in the arms and legs
- Low blood pressure or poor blood pressure control
- Muscle weakness or loss of muscle function (paralysis)
  - In mild cases, there may be weakness instead of paralysis
  - May begin in the arms and legs at the same time
  - May get worse over 24 to 72 hours
  - May occur in the nerves of the head only
  - May start in the arms and move downward
  - May start in the feet and legs and move up to the arms and head

- Numbness
- Sensation changes
- Tenderness or muscle pain
- Uncoordinated movement

Other symptoms may include:

- Blurred vision
- Clumsiness and falling
- Difficulty moving face muscles
- Muscle contractions

- Palpitations

Emergency symptoms:

- Breathing temporarily stops
- Can't take a deep breath
- Difficulty breathing
- Difficulty swallowing
- Drooling
- Fainting
- Feeling light-headed when standing

### Signs and tests

A history of increasing muscle weakness and paralysis may be a sign of Guillain-Barre syndrome, especially if there was a recent illness.

A medical exam may show muscle weakness and problems with involuntary (autonomic) body functions, such as blood pressure and heart rate. The examination will also show that reflexes, such as the "ankle or knee jerk," are decreased or missing.

There may be signs of decreased breathing caused by paralysis of the breathing muscles.

The following tests may be ordered:

- Cerebrospinal fluid sample ("spinal tap")
- ECG
- Electromyography (EMG) tests the electrical activity in muscles
- Nerve conduction velocity test
- Pulmonary function tests

### Treatment

There is no cure for Guillain-Barre syndrome. However, many treatments are available to help reduce symptoms and treat complications. When symptoms are severe, the patient will need to go to the hospital for treatment, which may include artificial breathing support. In the early stages of the illness, treatments that remove or block the proteins that attack the nerve cells, called antibodies, may reduce the severity and duration of GBS. One method is called plasmapheresis, and it is used to remove the antibodies from the blood. The process involves taking blood from the body, usually from the arm, pumping it into a machine that removes the antibodies, and then sending it back into the body. A second method is to block the antibodies using high-dose immunoglobulin therapy (IVIG). In this case, the immunoglobulins are added to the blood in large quantities, blocking the antibodies that cause inflammation. Other treatments are directed at preventing complications.

- Blood thinners may be used to prevent blood clots.
- If the diaphragm is weak, breathing support or even a breathing tube and ventilator may be needed.
- Pain is treated with anti-inflammatory medicines and narcotics, if needed.
- Proper body positioning or a feeding tube may be used to prevent choking during feeding if the muscles used for swallowing are weak

### Expectations (prognosis)

Recovery can take months or years. Most people survive and recover completely but, around 30% of patients still have some weakness after 3 years. Mild weakness may persist for some people.

A patient's outcome is most likely to be very good when the symptoms go away within 3 weeks after they first started.

### CASE STUDY

#### Guillain-Barré syndrome, quadriparetic, chronic respiratory failure

19 year-old male admitted to the emergency room with difficulty walking and numbness to the feet, diagnosed to have Guillain-Barré Syndrome and received 3 doses of IVIG. The patient was moved to the ICU and placed on mechanical ventilation due to respiratory failure. Patient underwent five treatments of plasmapheresis and required a tracheostomy for long-term mechanical ventilation.

### Physical Evaluation

**General:** Patient was mentally alert and could follow simple commands. Due to severe facial weakness, patient communicated by blinking for yes/no.

**Respiratory:** The patient was in chronic respiratory failure requiring ventilator support. Ongoing weaning trials and pulmonary care were provided. Patient received routine tracheal suctioning, mouth care and chest physiotherapy. Due to underlying neuromuscular disease, Patient required a prolonged ventilator weaning process over 3 to 4 weeks. Patient made steady strides in weaning and was eventually weaned completely off the ventilator. Upon admission the patient had a degree of hypertension that was treated with Lopressor that was slowly tapered and eventually discontinued. The patient remained normotensive with no cardiovascular issues while in Hospital. The patient initially had significant dysphagia requiring nutritional support through a gastronomy tube. By working with the speech and language pathologist and with improvement in his respiratory status his dysphagia improved. He eventually started on an oral diet that advanced to a full regular diet. Once he was meeting his calories needs the G-tube was removed. He was slightly hypomagnesemic and was placed on magnesium supplementation.

**Strength:** Bilateral UEs grossly 2/5. Hip and knee musculature grossly 2/5, ankle strength 0/5.

**Bed mobility, transfers:** Dependent for rolling and supine-to-sit. Transfers not tested due to severe weakness.

**Sitting balance:** Poor static and dynamic. Dependent to sit upright at edge of bed.

### Hospital Treatment

Physical therapy was consulted to begin rehabilitation and the patient's education included an in-bed exercise program, breathing exercises, progressive

sitting, trunk control activities, and daily use of a dynamic tilt Sigma Q exercise platform to develop lower extremity partial weight-bearing (PWB) strength training. For the PWB workout, the patient performed three to four sets of 10 inclined squats with tolerable Sigma Q resistance. The first session started with approximately 15% body weight with the patient progressing to 55% body weight after one week. During this time, sitting balance improved from dependent to unsupported sitting at bedside. Sit-to-stand assessment showed that the patient was unable to support full body weight and required maximal assistance to stand. For this reason, PWB exercise on the platform was continued for lower extremity strengthening. The therapy treatments then alternated manual standing on one day with PWB exercise the following day. This regimen continued for one week, at which time the patient could stand with minimal assistance using a rolling walker for support. The exercise platform was then discontinued so the patient could focus on progressive standing and gait training with a walker for the next week.

#### **Hospital Discharge**

The patient was discharged to home, walking with a rolling walker 300', independent with transfers and bed mobility. The patient required supervision for ascending and descending stairs. The patient followed up with outpatient Physiotherapy to restore endurance and balance.

#### **Outpatient Treatment**

Before being discharged, the hospital care team coordinated a home-based therapy program using the sigma Q technology so the patient could continue recovery. The patient continued with aggressive outpatient physical and occupational therapy and followed up with a neurologist. The Sigma Q technology was used to exercises and improve motor skills, to strengthen muscles enough to function normally and possibly to regain normal strength for age. The follow up therapy contained 132 sigma Q treatments over an eight month period. 95 hourly and 37-30 minute treatments.

The range of motion was evaluated with a goniometer before therapy and regularly thereafter while grades of muscle strength was recorded throughout the therapy using a grading factor of 1-5.

### **Grades of Muscle Strength**

<b>Grade</b>	<b>Description</b>
5 or N	Full range of motion against gravity and full resistance for the patient's size, age, and sex
N-	Slight weakness
G+	Moderate weakness
4 or G	Movement against gravity and moderate resistance at least 10 times without fatigue
F+	Movement against gravity several times or mild resistance one time
3 or F	Full range against gravity
F-	Movement against gravity and complete range of motion one time
P+	Full range of motion with gravity eliminated but some resistance applied
2 or P	Full range of motion with gravity eliminated
P-	Incomplete range of motion with gravity eliminated
1 or T	Evidence of contraction (visible or palpable) but no joint movement
0	No palpable or visible contraction and no joint movement

N = normal; G = good; F = fair; P = poor; T = trace.

#### **Results**

Treatments	Scaling
0 - 10	0 - T
11 - 35	T - P+
36 - 75	P+ - G
76 - 132	G - 5G

#### **Conclusion**

Without doubt, there will be advances in treatment as the pathogenesis of Guillain-Barre Syndrome is further elucidated. These advances should address the needs of the patients who are left with severe motor control 1-2 years from onset. Muscle strength was increased with progressive resistive exercise using the Sigma Q technology. The pathogenesis of GBS show edematous changes proximal to the spinal nerve root at the junction of the anterior and posterior roots. The myelin sheaths soon become irregular at about the third day. Lymphocytes appear about the ninth day and phagocytosis on the eleventh day.<sup>5</sup>

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