Management of Guillain-Barré Syndrome

Kadarapura N. Gopalakrishna¹  Venkatapura J. Ramesh¹

¹Department of Neuroanaesthesia and Neurocritical Care, National Institute of Mental Health and Neurosciences (NIMHANS), Bengaluru, Karnataka, India

Address for correspondence Kadarapura N. Gopalakrishna, DNB, DM, Department of Neuroanaesthesia and Neurocritical Care, National Institute of Mental Health and Neurosciences (NIMHANS), Bengaluru 560 029, Karnataka, India (e-mail: gops868@gmail.com).

Guillain-Barré syndrome (GBS) is the most common, immune-mediated acute polyneuropathy characterized by variable degree of motor weakness, often presenting with quadriparesis. GBS with respiratory failure requiring endotracheal intubation with mechanical ventilation is common, affecting approximately 30% of patients. In addition, they present with sensory features, cranial nerve involvement, and autonomic disturbance. The majority of GBS patients have preceding respiratory tract infection or gastroenteritis. The diagnosis of GBS is based on clinical features supported by cerebrospinal fluid (CSF) examination and nerve conduction studies. Early course of the GBS requires meticulous monitoring and early initiation of immunotherapy. Plasma exchange (PE) and intravenous immunoglobulin (IVIg) are the proven therapies, and both have been shown to be equally effective. General supportive care is an important part of management of GBS. A multidisciplinary approach to prevent and manage potential complications in rapidly progressing GBS is important to reduce morbidity and mortality.

Abstract

Guillain-Barré syndrome (GBS) is the most common, immune-mediated acute polyneuropathy characterized by variable degree of motor weakness, often presenting with quadriparesis. GBS with respiratory failure requiring endotracheal intubation with mechanical ventilation is common, affecting approximately 30% of patients. In addition, they present with sensory features, cranial nerve involvement, and autonomic disturbance. The majority of GBS patients have preceding respiratory tract infection or gastroenteritis. The diagnosis of GBS is based on clinical features supported by cerebrospinal fluid (CSF) examination and nerve conduction studies. Early course of the GBS requires meticulous monitoring and early initiation of immunotherapy. Plasma exchange (PE) and intravenous immunoglobulin (IVIg) are the proven therapies, and both have been shown to be equally effective. General supportive care is an important part of management of GBS. A multidisciplinary approach to prevent and manage potential complications in rapidly progressing GBS is important to reduce morbidity and mortality.

Keywords

► Guillain-Barré syndrome
► immunotherapy
► plasma exchange
► immunoglobulin

Introduction

Guillain-Barré syndrome (GBS) is an acute or subacute onset, immune-mediated inflammatory disorder of the peripheral nervous system in which an autoimmune response is directed against peripheral nerves (myelin or axons) and spinal roots.¹ The annual incidence rate of GBS across different studies range from approximately 0.5 to 2 cases per 100,000 people.²⁻⁴ The incidence rates increase with age, especially in those 60 years or older. There is also a gender difference; males are affected more than females (1.5:1). GBS is the most common cause of acute paralytic neuropathy.²⁻⁵ It usually presents with an acute and rapid progression of symmetric weakness of limbs, hyporeflexia or areflexia, and with or without involvement of respiratory muscles or cranial nerve. The majority of patients have an infection before the onset of GBS that is within 4 weeks, most commonly in the form of respiratory tract infection or gastroenteritis.⁶ The diagnosis of GBS is based on clinical features criteria supported by cerebrospinal fluid (CSF) findings and nerve conduction studies. The common subtypes of GBS based on clinical features, pathological, and electrophysiological findings are shown in ▲Table 1.⁷ The complexity of interplay between infectious triggers and host factors determining the immune response shifting toward undesirable autoreactivity is unknown.⁸ The environmental and genetic factors contribution toward individuals vulnerable to develop the disease progression is unclear.⁹ GBS is most common paralytic neuropathy leading to respiratory failure, finally requiring mechanical ventilation. The poor prognostic factors include rapid progression, bulbar involvement, facial weakness (bilateral), and autonomic dysfunction.¹⁰ The early specific standard treatment for GBS is plasma exchange (PE) or intravenous immunoglobulin (IVIg), and in addition, it requires supportive care and meticulous monitoring.

We searched PubMed and MEDLINE using the search term “Guillain Barre syndrome,” and article published in English were included. This review article provides an overview of the etiopathophysiology, diagnostic criteria, and management of GBS.

Pathophysiology

The experimental evidence and clinical findings demonstrate that GBS is an immune-mediated disorder related to a complex synergistic interaction of humoral and cell-mediated immune...
response against the antigens of the peripheral nerve.11 Nearly 75% of patients report a preceding event, most commonly infection of the upper respiratory tract or gastrointestinal tract at least 1 to 4 weeks before the onset of GBS symptoms.4 The other triggers of GBS include vaccinations (including influenza virus vaccine, hepatitis vaccine, rabies vaccine, tetanus and diphtheria toxoids, and meningococcal vaccine),12 surgery, and trauma.13 The most common organism linked to GBS is Campylobacter jejuni–induced bacterial enteritis and is commonly linked to AMAN subtype.14 The other specific microbial agents,15 linked to GBS, include cytomegalovirus (CMV); hepatitis A, B, and E viruses16; Epstein-Barr virus, varicella zoster virus; Zika virus infection16; human immunodeficiency virus, Haemophilus influenzae17; and Mycoplasma pneumoniae.18 A preceding infection may result in an autoimmune response leading to production of antibodies against an infective agent that shares epitopes with the host’s peripheral nerve gangliosides. This phenomenon is called postinfectious molecular mimicry.19 Currently, identification of the preceding infective trigger in a case of GBS may help in prognostication, developing individualized targeted management20 and preventing GBS cases in the region in future.21

### Diagnosis and Evaluation

The classic form of GBS presents with rapidly progressive symmetrical weakness of the lower limbs. It ascends proximally over hours to few days and progresses to involve the upper limbs and facial, oropharyngeal, and respiratory muscles in severe cases.22 The diagnostic criteria for GBS were revised by Asbury and Cornblath in 1990 (Table 2).23 The severity of illness varies from mild form in which patients are able to walk unassisted, to severe form leading to near-total quadriplegia. Sometimes, motor weakness develops simultaneously in all the four limbs or develops first in the arms followed by the legs. Most patients develop areflexia or hyporeflexia, but tendon reflex may be normal during early course of the disease. Cranial nerve involvement is common resulting in facial, extraocular, and bulbar weakness.7 In case of Miller–Fisher variant, typical clinical features include a triad of ophthalmoplegia, ataxia, and areflexia.24 Nearly 30% of GBS patients develop respiratory failure requiring endotracheal intubation and mechanical ventilation.24,25 Clinical findings suggesting impending respiratory muscle failure include tachypnea, tachycardia, sweating, unsynchronized thoracoabdominal movements, and use of accessory muscles of respiration. Apart from motor weakness, patients may have burning or tingling sensations of the limbs,26 pain in the extremities,27 sensory loss mainly impairment of vibration sense, and autonomic dysfunction.28 The autonomic dysfunction may be present in the form of tachycardia or arrhythmias, hypertension, postural hypotension, sweating abnormalities, and bladder/bowel disturbances.28 In addition, excessive vagal activity triggered by endotracheal suctioning or other stimulus can result in episodes of bradycardia, cardiac conduction block, or asystole. Clinically significant autonomic dysfunction may progress for up to 4 weeks of the illness. Cardiac arrhythmias in severe form with unstable hemodynamics is more often seen in patients with severe quadripareisis and respiratory failure.28 The diagnosis of GBS in young children is difficult because they present with atypical clinical features, and it is more difficult to perform neurological examination in them.29 In children, pain is the most common presentation before the motor weakness and leads to delay in the diagnosis of GBS.27 In view of a wide range of differential diagnosis in children, it is challenging to rule out other causes such as brainstem lesion, spinal cord lesion, muscular abnormalities, or other neuromuscular junction abnormalities.20

The diagnosis of GBS is supported by CSF examination and nerve conduction studies. The characteristic finding of CSF examination is the increased protein levels without pleocytosis, known as cytoalbuminologic dissociation.31 Usually in the first week of disease course the CSF may be normal, but thereafter from second week onward, there is an elevation in CSF protein in almost 90% of cases.25 Transient oligoclonal bands and increased myelin basic protein may be detected in the CSF of few GBS patients.7

Electrodiagnostic features of nerve conduction are strongly supportive of GBS diagnosis. The electrodiagnostics features strongly supportive of GBS diagnosis include elevated CSF protein with cell count <10 cells/µL, Electrodiagnostic features of nerve conduction slowing or block.4

### Criteria for diagnosis of Guillain–Barré syndrome

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*Findings suggestive of an axonal neuropathy. Abbreviations: CSF, cerebrospinal fluid; GBS, Guillain–Barré syndrome.

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acquired demyelination from axonal degeneration subtypes of GBS. In case of acute demyelinating polyneuropathy, NCS features include prolonged latency of distal motor, decreased conduction velocity of nerve, increased temporal dispersion, latency prolongation of F-wave, and nerve conduction blocks. In addition, sural nerve sensory response is often spared. The characteristic electrodiagnostic feature of acute motor axonal GBS is reduced amplitude or absent distal compound muscle action potential (CMAP). A few patients with axonal GBS will have slowing or temporary conduction blocks that usually recover rapidly during the course of the illness. This phenomenon is known as early reversible conduction failure. The transient feature of the reversible conduction failure initially may suggest acute inflammatory demyelinating polyradiculoneuropathy (AIDP), but it should be distinguished from axonal neuropathy by serial NCS over weeks. Reversible conduction failure specific to axonal neuropathy suggest that it is due to anti-ganglioside antibody–directed mechanism, and in future, targeted therapies can be developed. Electrodiagnostic studies might also predict prognosis, since patients with NCS features of demyelination often require mechanical ventilation, and decreased CMAP are more suggestive of poor outcome. Patients with axonal neuropathy may recover either very slowly or incompletely, or sometimes recover rapidly because of restoration of temporary conduction blocks.

The anti-ganglioside antibodies levels can be used in the diagnosis of certain subtypes of GBS. Raised levels of anti-GQ1b ganglioside antibodies are found in more than 90% of Miller–Fisher syndrome (MFS) subtype of GBS. Preceding C. jejuni infection is linked to the AMAN variant and have elevated titers of anti-GM1, anti-GD1b, anti-GD1a, and anti-GalNAc-GD1a ganglioside antibodies of the IgG class. Raised serum antibodies to Mycoplasma, CMV, or C. jejuni can identify the preceding infection. Anti-galactocerebroside antibodies are found to be detected in patients with preceding Mycoplasma infection.

Neuroimaging, especially magnetic resonance imaging (MRI) of the spine with contrast, has been used for diagnostic utility of GBS and also useful to rule out differential diagnostic abnormalities. Typical GBS has enhancement and swelling/thickening of the spinal nerve roots/cauda equina on spinal MRI.

Other laboratory abnormalities in GBS include hyponatremia mostly in patients on mechanical ventilation, most likely due to inappropriate secretion of antidiuretic hormone. Transient mild elevation in liver enzymes in almost about one-third of GBS patients without detectable cause, and rarely renal dysfunction due to glomerulonephritis (microscopic hematuria and proteinuria) because of immune complex deposition.

Management

The mainstay of treatment in GBS includes general supportive care and specific treatment with immunotherapy.

General Supportive Care

General supportive care is an important part of treatment of GBS. A multidisciplinary approach to prevent and manage potential complications in rapidly progressing GBS is important to reduce morbidity and mortality. Especially during the progressive phase of GBS, it is important to continuously monitor for respiratory failure, bulbar dysfunction, physical ability to handle secretions to prevent lung aspiration, and hemodynamics. The management of respiratory failure, hemodynamics, and cardiovascular dysfunction is most important and provides better chances for the favorable outcome. In addition, other issues to be addressed are deep vein thrombosis (DVT) prophylaxis, management of pain, treatment for possible impaired bowel and bladder function, autonomic dysfunction, early physiotherapy initiation and plan for rehabilitation, and psychological support.

GBS patients with respiratory failure and bulbar dysfunction require mechanical ventilation. Predictors for the need of mechanical ventilation include rapid progression of disease course (onset of weakness to admission < 7 days), severity of motor weakness, bulbar dysfunction, presence of neck and facial weakness, or dysautonomia. Predictors for endotracheal intubation and ventilatory assistance in GBS are shown in Table 3. When the disease course strongly predicts for ventilatory support, tracheal intubation should be performed on an urgent basis before the crisis. Tracheostomy has to be performed when the ventilator support is longer than 2 weeks and there is no improvement in pulmonary function test from the baseline. If there is an improvement in strength and lung function test from baseline after 2 weeks, tracheostomy can be postponed for an additional week and meanwhile gradual weaning from the ventilator may be attempted. In centers experienced in percutaneous dilation tracheostomy, it is found to be a superior method as compared with the traditional surgical tracheostomy in GBS patients. Weaning from mechanical ventilatory support should be based on improvement in strength and serial bedside lung function test. Patients requiring prolonged mechanical ventilation are shown in Table 3.

Table 3 Predictors of intubation and ventilatory assistance in Guillain-Barré syndrome

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<td>• Clinical evidence of inability to cough, unable to clear secretions, and neck weakness</td>
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<tr>
<td>• Signs of impending respiratory failure such as tachypnea, use of accessory muscles, inadequate chest rises, and paradoxical breathing with inadequate effort</td>
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<tr>
<td>• Simple bedside assessment tool</td>
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<td>Single breath count (SBC) &lt; 20 Unable to complete the sentences</td>
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<tr>
<td>• Pulmonary function</td>
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<tr>
<td>Vital capacity (VC) &lt; 15–20 mL/kg</td>
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<tr>
<td>Maximum expiratory pressure (Pmax) &lt; 40 cm H2O</td>
</tr>
<tr>
<td>Maximum inspiratory pressure (Pmax) &lt; 30 cm H2O</td>
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<tr>
<td>&gt; 30% reduction in baseline VC, Pmax, and Pmax'</td>
</tr>
<tr>
<td>• Radiographic abnormalities include pulmonary infiltrates or atelectasis</td>
</tr>
<tr>
<td>• Arterial blood gas showing hypoxemia or hypercarbia or both</td>
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mechanical ventilation are at an increased risk of ventilatory associated pneumonia, sepsis, bleeding from gastrointestinal tract, and other complications.

In view of marked fluctuations in blood pressure or cardiac arrhythmias in GBS, continuous blood pressure and electrocardiogram (ECG) monitoring helps in early identification and timely treatment. Extreme care should be taken in using antihypertensive or vasoactive agents in patients with dysautonomia. A painful stimulus such as endotracheal suction can trigger episodes of bradycardia and hypotension. Extra precaution should be taken during tracheal suction and to monitor oxygen saturation, heart rate, and blood pressure. The other autonomic disturbances such as hyponatremia, decreased mucosal function of bronchus, and adynamic ileus may be anticipated and managed accordingly.

The management of pain in GBS is extremely important during the whole spectrum of disease. During acute phase of the GBS, considerable decline in pain severity was noted for gabapentin use and also with the use of carbamazepine. Large, high-quality randomized controlled trials (RCTs) are required to prove the efficacy of gabapentin and carbamazepine for pain intervention in the acute and recovery phase of GBS. The other drugs used for neuropathic pain include tricyclic antidepressants, lidocaine, SSRI (selective norepinephrine reuptake inhibitors), and topical capsaicin.

Adequate high-energy nutrition with high caloric protein diet should be instituted early through enteral feeding. Nutrition plays an important role to prevent muscle wasting and helps in weaning patients from ventilatory support. In contrast to bolus feeding, continuous enteral feeding is better tolerated.

Subcutaneous unfractionated heparin or low-molecular-weight heparin and calf compression devices should be used regularly in immobilized GBS patients to decrease the risk of DVT and pulmonary embolism. Prophylaxis for DVT should be continued until patients are able to walk with or without support.

In the intensive care unit (ICU), pulmonary infection and urinary tract infection are common in patients with GBS. It is important to take preventive measures and aggressively treat nosocomial infections. Chest physiotherapy with tracheal suction and periodic change of patient position helps in preventing atelectasis in patients with depressed cough. Proficient nursing care with regular care of the mouth, skin, eyes, bowel, and bladder is necessary.

In case of facial weakness, exposure keratitis is prevented by supplementing with artificial tears and using adhesive tape to close the eyes at night. Position-related nerve palsies are prevented by adequate padding of pressure points. Physiotherapy of limbs started early in the disease course helps prevent joint contractures and immobilization. Constant reassurance and psychological support of patients and family are important for recovery.

**Immunotherapy**

The evidence-based proven therapies for GBS are PE and IVlg, and both have been shown to be equally effective.

**Plasma Exchange**

Therapeutic PE has established benefit in acute GBS by decreasing the recovery time. PE initiated within 4 weeks of onset of weakness is proven beneficial in severe GBS, but substantial benefit was noted when started within 2 weeks. The recommended plasmapheresis includes a series of five PEs (40–50 mL/kg) on alternate days. The therapeutic PE procedure involves removal of blood from the patient to separate inflammatory mediators such as pathogenic autoantibodies, cytokines, and complement components. The cellular components and some plasma along with replacement fluid such as crystalloid, albumin, or fresh frozen plasma are transfused to the patient. There are several techniques for plasmapheresis, which include filtration technique (membrane plasma separator) or centrifugation. Filtration technique usually requires a central venous access, and in this method, cellular elements are separated, but large molecules are able to pass through the filter. In centrifugation technique, the whole blood is spun until the components are separated and plasma components are removed in addition to immunoglobulins and soluble large molecules. Immunoadsorption technique is an alternative to PE in which plasma components are exposed to an absorptive material of variable selectivity, and thus only certain specific inflammatory mediators are removed rather the plasma itself.

Based on moderate-quality evidence, PE improved the greater part of outcomes in comparison to supportive treatment alone. The time to regain strength to walk with support or without support and time to recover by 1 or 2 disability grades were decreased by PE. PE reduced the time on ventilator, and the percentage of patients on ventilator support was significantly shortened. PE did not change the risk of cardiovascular instability or severe infections but definitely reduced the possibility of cardiac arrhythmias. Long-term benefits of PE after 1 year include significantly increased number of patients with fully recovered muscle strength and reduced the percentage of patients with severe motor deficits.

Based on the severity of disease, a trial suggested appropriate number of PE, which included two sessions for mild GBS and four sessions for moderate to severe GBS. There are no randomized studies that have assessed the efficacy of PE in children aged < 12 years. The important considerations for using therapeutic PE are venous access and requirement of high flow rates. The clotting factors depletion during PE and heparin use predisposes the subjects to risk of bleeding. Sometimes, citrate is used as an anticoagulant, which may predispose the patient to hypocalcemia or metabolic acidosis. Cost analyses estimated in different studies showed a decrease in the cost in patients with GBS treated with PE. Small-volume PE is one option, especially in developing countries where cost is the limiting factor. In the case of small-volume PE, 15 mL/kg/day of volume of plasma is exchanged until the progression of disease is ceased or recovery started.

**Intravenous Immunoglobulin**

The recommended regimen of IVlg for GBS is the total dose of 2 g/kg, that is, 0.4 g/kg/day for 5 consecutive days. IVlg started...
within 2 weeks of onset of weakness accelerates recovery as good as PE.  The main content of IVlg is IgG, and other molecules present are IgM, IgA, various cytokines, and soluble anti-TCR. These antibodies supplied by IVlg have the potential to bind and neutralize pathogenic antibodies. In many centers, IVlg has replaced therapeutic PE as the choice of treatment because of greater degree of accessibility, availability, and convenience of infusion at any time. There is some evidence that rapid infusion over 2 days had a better response compared with a longer course, but data from most trials show no such benefits. Minor side effects of IVlg include myalgias, headache, arthralgias, flu-like symptoms, and fever. Minor infusion-related complications, though frequent, can often be prevented or controlled by slowing the rate of IVlg infusion or by symptomatic treatment. Sometimes serious complications such as anaphylaxis in IgA-deficient patients, congestive heart failure, transient renal dysfunction, and thrombotic complications are reported.

Corticosteroids
Trials have demonstrated oral or intravenous corticosteroids alone in GBS have no clear benefits in the recovery or long-term effects. Though GBS is an inflammatory neuropathy due to autoimmune response, the reason for ineffectiveness of steroids is unknown.

Combined Immunotherapy
The effectiveness of the combined therapy of PE followed by IVlg showed no significant benefit when compared with PE or IVlg alone. There are trials to show that combined therapy of IVlg and parenteral methylprednisolone had no clear benefit than IVlg alone.

Course and Prognosis
About 5 to 15% of GBS-treated patients deteriorate after initial improvement or stable phase of the disease. Treatment-related fluctuations occur due to transient treatment effects of the prolonged disease course. In such scenario, IVlg treatment can be repeated, but there are no RCTs for assessing the utility of repeat IVlg course. By standard definition, patients should reach maximum neurological deficit within 1 month of onset of disease, if disease progresses for beyond 1 month, but less than 2 months is categorized as subacute or more than 2 months classified has chronic inflammatory demyelinating polyneuropathy (CIDP). Acute-onset CIDP is to be considered if GBS disease severity worsens after 2 months or have more than twice the treatment-related fluctuations. It is important to differentiate between acute-onset CIDP from recurrent GBS or GBS with treatment-related fluctuations, because acute-onset CIDP may require maintenance therapy or a change of treatment from IVlg to corticosteroids.

After disease progression stops, plateau phase lasts for 15 days or 1 month or longer before recovery begins. Most patients with GBS recover functionally, but still 20% of patients have residual motor deficits even after 1 year. Approximately 70% of patients will have complete recovery in 1 year and 82% in 2 years. Up to 2 to 7% of patients may have recurrence following complete recovery and seen commonly in young patients, mild GBS course, and MFS subtype. AIDP subtype of GBS has more recurrence than axonal subtypes. In general, prognosis of GBS is influenced by patients age, severity of illness at its peak, and dependence on whether immunotherapy started early. In addition, infectious episodes and acute hypoxic ischemia may worsen the prognosis.

Conclusion
GBS is a common cause of acute paralytic polyneuropathy. Early course of the GBS requires meticulous monitoring and specific treatment with PE or IVlg. General supportive care, including management of respiratory failure, hemodynamics, and cardiovascular dysfunction, is most important and provides better chances for the favorable outcome.

Conflict of Interest
None declared.

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