Guillain-Barré Syndrome

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Author Disclosure
Dr Rosen has disclosed no financial relationships relevant to this article. This commentary does not contain a discussion of an unapproved/investigative use of a commercial product/device.

Educational Gap

The American Academy of Neurology published a practice parameter in 2003 outlining treatment recommendations for Guillain-Barré syndrome. The recommendations were based on strong data in adults and were extrapolated to children because no well-designed randomized studies have been done in children.

Objectives

After completing this article, readers should be able to:

1. **Understand the common causes of acute weakness and paralysis in children.**
2. **Understand the presenting features of Guillain-Barré syndrome in children.**
3. **Understand how to rule out other potentially dangerous and treatable causes of weakness and paralysis in children.**
4. **Understand the management of the child with Guillain-Barré syndrome.**

Definition

Guillain-Barré syndrome (GBS) is an acquired disorder of the peripheral nerves, described best as a polyradiculoneuropathy. With the widespread eradication of poliomyelitis, GBS is the most common cause of acute and subacute flaccid paralysis in infants and children. The disorder is characterized by progressive, symmetrical, usually ascending weakness, and diminished or absent reflexes. Diffuse pain also is a common presenting feature in many children with GBS, at times delaying accurate diagnosis. GBS was thought previously to be an inflammatory disorder that affected only the myelin sheath, resulting in diffuse demyelination. However, it is now recognized that the process can also attack the axon, leading to degeneration of the nerve itself.

Epidemiology

GBS occurs year round and in all age groups. Adults are affected more commonly than children. Some studies suggest that males are more likely to be affected than females. The overall incidence has been estimated to range from 0.4 to 2.4 cases per 100,000 per year, with 3,500 new cases per year occurring in the United States. The incidence in children is lower, with estimates between 0.4 and 1.3 cases per 100,000 per year. GBS can occur at any age but is rare in children under the age of 2 years.

Abbreviations

AIDP:	acute inflammatory demyelinating polyneuropathy
AMAN:	acute motor axonal neuropathy
AMSAN:	acute motor and sensory axonal neuropathy
CSF:	cerebrospinal fluid
GBS:	Guillain-Barré syndrome
IgG:	immunoglobulin G
IVIG:	intravenous immunoglobulin
MFS:	Miller-Fisher syndrome

Pathogenesis

GBS was long considered a single disorder, acute inflammatory demyelinating polyradiculoneuropathy (AIDP), in which the target is the myelin sheath. This variant of GBS remains the most common form seen in the United States and Europe. However, it has become clear that there are phenotypically similar disorders, acute motor axonal neuropathy (AMAN) and acute motor and sensory axonal neuropathy (AMSAN), in which the target is the axon itself, rather than the myelin sheath. AMAN is the predominant form seen in children in China and South America. In addition

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there are other variants considered to fall under the category of GBS, the most common of which is the Miller-Fisher syndrome (MFS), a disorder characterized by ophthalmoplegia, ataxia, and areflexia, usually with little to no weakness.

The pathogenesis of GBS is believed to be an abnormal immune response to an antecedent event, most commonly an infection. Approximately two-thirds of patients report a respiratory or gastrointestinal illness in the weeks preceding onset of symptoms. Many viruses and bacteria have been implicated, including cytomegalovirus, Epstein-Barr virus, *Haemophilus influenzae*, herpes simplex virus, and *Mycoplasma*. The strongest association is with the bacterium *Campylobacter jejuni*, which is believed to be the cause of 20% to 30% of GBS in the United States and Europe and a higher percentage of the AMAN cases seen in China and South America.

Vaccines also have been implicated as a triggering factor. Some evidence suggests that the "swine flu" vaccine used in the 1976 influenza season led to an increased incidence of GBS. There are no data linking other vaccines, including diphtheria-tetanus-acellular pertussis and measles-mumps-rubella, to GBS. In 2005, the Food and Drug Administration issued an advisory about the possible association between the MCV4 meningococcal vaccine and GBS and recommended that the vaccine not be given to adolescents or adults with a history of GBS. However, the 2009 *Red Book* published by the American Academy of Pediatrics says "the available data cannot determine with certainty whether MCV4 increases the risk for GBS and do not affect immunization recommendations". (1)

Rarely, GBS can be the presenting manifestation of lymphoma or systemic lupus erythematosus or can develop during the course of lymphoma or systemic lupus.

Pathophysiology

The pathophysiology of GBS is not completely understood. Both cellular and immune functions are involved. It is believed that, in a type of molecular mimicry, antibodies that form in response to antigens on the causative agent, usually a bacterium or virus, cross-react with gangliosides on the surface of the peripheral nerve. Antiganglioside antibodies, most commonly GM1, occur frequently in GBS. Some appear to target the myelin sheath, others, the nodes of Ranvier and the axon. The presence of antibodies to GM1 ganglioside is particularly common when GBS follows infection by *C jejuni*. The strongest connection with antiganglioside antibodies is found in the MFS, in which antibodies to G1QB ganglioside are found in >90% of patients who have this disorder. The nerves that innervate the extraocular muscles are enriched with G1QB ganglioside, which explains why ophthalmoplegia is the hallmark of this disorder.

Clinical Aspects

GBS is a triphasic illness characterized by an acute phase, a plateau phase of variable length, and a recovery phase that can take weeks to months. Initial symptoms usually present between 1 and 4 weeks after a respiratory or diarrheal illness.

The first phase is characterized by the onset and progression of symptoms. This phase can last from hours to weeks. The usual presenting concern is weakness that may manifest as a disturbance of gait such as refusal to walk, walking on a wide base, or difficulty with running or climbing stairs. The gait disturbance, although usually interpreted initially to be ataxia and therefore thought to represent central nervous system disease, usually is caused by weakness rather than cerebellar dysfunction. Vague paresthesias often are described. Significant, poorly localized pain is common, which can cause delay in diagnosis, because the child may be difficult to examine; early findings may be subtle, and pain is not commonly known to be associated with GBS.

As is typical in a peripheral neuropathy, weakness usually begins distally in the legs and ascends, at times rapidly, to involve the arms, face, and muscles of respiration. The weakness usually is accompanied by diminished or absent reflexes early in the course. Less commonly, weakness begins in the proximal muscles, a pattern that can lead to an incorrect diagnosis of myositis. Cranial nerve abnormalities are frequent, occurring in up to 50% of patients in some case studies. The facial nerve is the most commonly affected cranial nerve, and the weakness often is bilateral. Papilledema is a rarely reported finding in GBS and represents increased intracranial pressure of unclear etiology. Pupillary abnormalities are rare and should lead to consideration of another diagnosis, such as botulism.

Autonomic instability, including cardiac dysrhythmias, orthostatic hypotension, hypertension, paralytic ileus, and transient bladder dysfunction, is common and can occur anytime during the illness. Patients must be monitored closely for autonomic instability because it can be life threatening.

The acute illness usually peaks in severity 2 weeks after onset.

One recent study highlighted the course of 95 children with GBS in a prospective study. (2) Most study participants had AIDP, the demyelinating form of GBS. The disease progressed to a peak in a median of 7 days. At the peak, 40% were still able to ambulate, one-half of them

without assistance and one-half with assistance. Upper extremity function was abnormal in 76%, and 9% had no movement at all. Forty-three percent had cranial nerve weakness, and 13% required mechanical ventilation. An additional 18% were considered to have respiratory compromise. Fifty-one percent had a least one type of autonomic dysfunction, and 79% reported pain (Table 1).

Patients who had AMAN and AMSAN usually present in a similar fashion to the more common AIDP, with the addition of sensory symptoms in AMSAN. MFS presents with external ophthalmoplegia, ataxia, and areflexia without significant weakness. The first symptom in this disorder usually is diplopia, which progresses to clear paralysis of extraocular movements. The ataxia in MFS is not a pure cerebellar ataxia, although cerebellar circuits are almost certainly involved. Peripheral sensory nerves rather than motor nerves are affected predominately in this syndrome, leading to a "sensory ataxia." In this case, it is the loss of proprioception that leads to a widely based, unsteady gait.

Differential Diagnosis

The differential diagnosis of GBS is extensive (Table 2). The most important disorders to exclude are tick paralysis, botulism, transverse myelitis, and a compressive spinal cord lesion, because these treatable diseases have presentations that mimic GBS.

Tick paralysis is an acute peripheral neuropathy caused by a species of the *Dermacentor* tick in the United States and the tick *Ixodes holocyclus* in Australia. Patients can present with acute ataxia or ascending paralysis or both. Onset is usually more rapid than seen with GBS. Treatment involves removal of the offending tick, and recovery is usually complete. A careful examination is important, especially in children with long or thick hair, in whom finding a tick can be challenging.

Infant botulism is a disorder of neuromuscular transmission caused by colonization of the bowel by botulinum spores found in food or soil. It is predominately a disorder of infants <1 year of age, an age group in which GBS is rare. Much less frequently, botulism can be caused by botulinum toxin in food or wounds, usually in older individuals. Mild cases also have been seen in children receiving botulinum toxin for spasticity. Botulism differs from GBS in that it is usually a descending rather than ascending paralysis, beginning with the cranial nerves. Pupillary abnormalities and bulbar symptoms, such as chewing and swallowing difficulties, are common. If this disorder is suspected, treatment is available in the form of botulism immune globulin. Untreated patients can develop severe respiratory compromise; so treatment must be started before laboratory confirmation.

An acute disorder of the spinal cord can be confused with GBS, especially because reflexes may be absent in an acute spinal cord lesion. Transverse myelitis, secondary to

Table 1. Signs and Symptoms of Guillain-Barré Syndrome in a Prospective Study of 95 Patients

	At Presentation	At Diagnosis	At Peak of Illness
Symmetrical weakness		100%	
Hyporeflexia or absent reflexes		100%	
Gait abnormality	Unsteadiness of gait: 45% Inability to walk unaided: 24%	Inability to walk unaided: 60%	Inability to walk unaided: 80%
Pain	34%	>30%	79%
Dysesthesias	4%		
Cranial nerve dysfunction	Diplopia: 3% Facial weakness: 2%	27%	46%
Autonomic nervous system dysfunction		33%	51%
Upper extremity weakness			74%
Respiratory compromise		20%	13% intubated + 18% other respiratory compromise

Table constructed with data from: Korinthinberg R, Schessl J, Kirschner J. Clinical presentation and course of childhood Guillain-Barré syndrome: a prospective multicentre study. *Neuropediatrics.* 2007;38:10–17. Published with permission from Georg Thieme Verlag KG.

Table 2. Differential Diagnosis of Guillain-Barré Syndrome

Central nervous system
- Hysterical paralysis

Cerebellum
- Acute cerebellar ataxia

Spinal cord
- Transverse myelitis
- Spinal cord compression

Anterior horn cell
- Poliomyelitis

Peripheral nerves
- Tick paralysis (United States)
- Toxic neuropathy
- Diphtheria
- Porphyria

Neuromuscular junction
- Botulism
- Myasthenia gravis
- Tick paralysis (Australia)
- Neuromuscular blocking agents

Muscle
- Viral myositis
- Inflammatory myopathies
- Metabolic myopathies

Table 3. Factors That Cast Doubt on Diagnosis of Guillain-Barré Syndrome

- Marked persistent asymmetry of motor function
- Persistent bowel or bladder dysfunction
- Discrete sensory level
- Cerebrospinal fluid pleocytosis > 50 cells/mm^3
- Ptosis
- Pupillary abnormalities
- Prominent bulbar signs
- Descending weakness

Reprinted with permission from Asbury AK, Cornblath DR. Assessment of current diagnostic criteria for Guillain-Barré syndrome. *Ann Neurol.* 1990; 27(suppl):S22.

an infectious or autoimmune disorder, and spinal cord compression are the most common causes of acute spinal cord dysfunction. It is very important to distinguish these disorders from GBS because early corticosteroid treatment may improve outcome in transverse myelitis, and a compressive myelopathy often is a surgical emergency.

Several features distinguish a spinal cord lesion from GBS, although these characteristics may be difficult to appreciate in a young child. These findings include persistent localized back pain, asymmetric weakness, a discrete sensory level, and bladder dysfunction. Any suspicion of a spinal cord disorder should lead to emergency imaging of the spine (Table 3).

A central nervous system disorder such as acute cerebellar ataxia caused by cerebellitis is in the differential diagnosis, but strength and reflexes often are preserved. Myasthenia gravis, a disorder of neuromuscular transmission, is another cause of acute weakness, but the findings of ptosis and fluctuating weakness distinguish this disease from GBS.

Poliomyelitis, an infectious disorder of the anterior horn cell, can mimic GBS. However poliomyelitis is usually accompanied by fever and meningismus, which are unusual findings in GBS. Lyme polyradiculitis can also be confused with GBS, but is rarely seen as a feature of Lyme disease in the United States. Diphtheria and other toxic, infectious, and metabolic neuropathies are in the differential diagnosis, but are much less common.

Laboratory Tests

The diagnosis of GBS is made on clinical grounds. Laboratory tests can be confirmatory early in the course, but treatment never should be delayed awaiting results, especially because early in the course test results may be normal.

Cerebrospinal fluid (CSF) evaluation and electromyography are the two laboratory tests most useful in confirming the diagnosis of GBS. MRI of the spine with gadolinium can be useful as an adjunctive test if the diagnosis is in question or if electromyography is not available.

The typical CSF finding in GBS is elevated protein level without an elevated cell count (albuminocytologic dissociation). Protein concentration usually is normal in the first few days of the illness and begins to rise by the end of the first week, peaking in 4 to 6 weeks. In most cases, the CSF has <10 cells per mm^3, all lymphocytes. Greater than 50 leukocytes per mm^3 in the CSF should lead to consideration of other diagnoses, although this finding does not absolutely rule out GBS.

Early in the course of the disease, electromyography findings may be normal or minimal, such as absent F responses. Later in the course of AIDP, there are findings of prolonged distal latencies, conduction velocity slowing, and evidence of conduction block, which are all signs of demyelination. In the axonal forms (AMAN, AMSAN,

and MFS), there is reduced amplitude of compound action potentials without conduction slowing or prolongation of distal latencies, which is the characteristic finding in axonal injury.

The typical MRI finding in GBS is gadolinium enhancement of the spinal nerve roots.

Asbury originally wrote a summary of criteria for GBS in 1978, which was modified by Asbury and Cornblath in 1990 (Table 4). (3)

Management

Treatment of GBS, including the use of intravenous immunoglobulin (IVIG) as described below, should begin as soon as the diagnosis is determined clinically.

The most important aspect of treatment in the child with GBS is supportive care. All children with GBS require hospital admission until their course has stabilized. Those who have flaccid quadriparesis, a rapidly progressive course, vital capacity at or below 50% of expected (<20 mL/kg), bulbar palsy, or autonomic instability should be monitored in an intensive care unit setting (Table 5).

Close attention must be paid to pain management, nutrition, and psychosocial needs. Rehabilitation should begin as soon as diagnosis is made to prevent contractures and skin breakdown and to restore function.

Table 4. Diagnostic Features of Guillain-Barré Syndrome

Features required for diagnosis
- Progressive motor weakness of more than one limb
- Areflexia or marked hyporeflexia

Features strongly supportive of the diagnosis
- Progression over days to < 4 weeks
- Relative symmetry
- Pain, often significant, at onset
- Mild sensory symptoms or signs
- Cranial nerve involvement
- Autonomic dysfunction
- Absence of fever at onset of symptoms
- Onset of recovery 2–4 weeks after onset of plateau phase

Laboratory features
- Elevated cerebrospinal fluid protein level after one week of symptoms
- <10 leukocytes in cerebrospinal fluid
- Slowed conduction or conduction block on electromyography

Modified with permission from Asbury, AK, Cornblath, DR. Assessment of current diagnostic criteria for Guillain-Barré syndrome. *Ann Neurol.* 1990;7(suppl 1):S22.

Table 5. Indications for ICU Monitoring

- Flaccid quadriparesis
- Rapidly progressing weakness
- Vital capacity < or = 20 mL/kg
- Bulbar palsy
- Autonomic cardiovascular dysfunction

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The American Academy of Neurology published a practice parameter in 2003 outlining treatment recommendations for GBS. (4) The recommendations were based on strong data in adults and were extrapolated to children because no well-designed randomized studies have been done in children.

The practice parameter stated that there was good evidence in adults that both plasma exchange, started within 4 weeks of symptoms, and IVIG started within 2 weeks of symptoms hastened recovery in patients who were unable to walk independently. There was no additional benefit from the use of plasma exchange and IVIG together.

Because of the technical difficulty of the use of plasma exchange in children, IVIG has become the treatment of choice. The recommendation has been to treat those children who are nonambulatory with 2 g/kg total of IVIG in equally divided daily doses over 2 to 5 days. Common adverse effects of IVIG include fever, myalgia, headache, chills, nausea, and vomiting. Hypersensitivity and anaphylactoid reactions can occur. A full list of adverse effects is included in the 2009 *Red Book*. (5)

For children who remain ambulatory or for those children whose course has stabilized, only supportive treatment is recommended. Of course, every case must be considered individually, and many child neurologists would recommend treatment in any patient who has been diagnosed as having GBS, especially if there is bulbar weakness or respiratory compromise, even in the plateau phase.

Korinthenberg et al, in 2005, by using some of the patients included in the previously mentioned study, attempted to answer two questions about the benefits of IVIG use in children: (1) "Can early treatment with IVIG before the loss of unaided walking reduce the subsequent severity of the disease?" (2) "After the loss of unaided walking, does treatment with 1 g/kg per day body weight IVIG over 2 days result in faster recovery of

unaided walking than treatment with 0.4 mg/kg per day body weight over 5 days?" (6)

The authors could not answer the first question because the early treatment group consisted of only 21 children, 14 of whom were randomized to the treatment group. Of the seven in the non-treatment group, 4 became unable to walk and were randomized into the late treatment group, leaving only 3 children untreated.

In the late treatment group, the time to regain independent walking did not differ between the 2-day and 5-day regimens. However, patients in the 2-day regimen had a higher rate of early transient relapses that was statistically significant ($P = .049$); but the extent of the deterioration and the time to recovery were not described. Both regimens continue to be used.

A recent retrospective study looked at immunoglobulin G (IgG) levels in adults with GBS after treatment with IVIG and found a correlation with IgG levels and recovery time, suggesting that some patients may need higher or additional doses. (7) Although this finding needs to be replicated in a prospective study, the implications should be kept in mind if treating a patient who does not seem to be responding to treatment.

Corticosteroids have been tested extensively in adult GBS and are not indicated.

Course and Prognosis

The prognosis for childhood GBS generally is excellent. This outcome is quite different from adults, in whom morbidity and mortality are considerably higher, and severity of disease is correlated with outcome.

Although treatment with IVIG is believed to hasten recovery, and thus reduce complications associated with prolonged disability, IVIG therapy is not felt to influence ultimate outcome. Older studies in children report full recovery within 6 to 12 months, with the majority of those who do not recover fully having only mild disabilities.

In the study cited above, improvement was noted first at a median of 11 days, ability to walk unaided returned in a median of 43 days, and 75% became symptom free in a median of 106 days. (2) Sensory disturbances, early fatigability, and dyscoordination remained a problem for some.

Another study looked at 47 children 2 years after recovery from GBS. (8) On detailed examination, 46% had some residual neurologic findings, including 27% with decreased or absent deep tendon reflexes and 15% with sensory disturbances. Twenty-three percent had mild weakness in at least one muscle group. In this study, younger age and rapid progression correlated with long-term weakness, but this has not been a consistent finding in other studies of childhood GBS. Most importantly, however, no child tested had any functional disability.

Summary

- Based on strong research evidence, in countries where poliomyelitis has been eliminated, GBS is the most common cause of acquired paralysis in children. (9)
- Based on strong research evidence, GBS describes a spectrum of disorders caused by an autoimmune reaction against peripheral nerve components, including the myelin sheath and the axon. (10)(11)
- Based on strong research evidence, GBS usually is preceded by a bacterial or viral infection, less likely by vaccination in the 1 to 4 weeks before onset. The strongest relationship is with infection by *C jejuni*. (12)(13)
- Based on strong research evidence, GBS in children most often presents with symmetrical ascending paralysis, diminished or absent reflexes, and often severe pain. Pain may lead to a delay in diagnosis. (2)(3)(9)(14)
- Based on strong research evidence, the progressive phase peaks in 7 to 14 days and can lead to various levels of weakness, from abnormal gait to total paralysis, cranial nerve weakness, pain, respiratory compromise, and autonomic instability. (2)
- Based on some research evidence and consensus, children require hospitalization and often intensive care until their condition stabilizes because of significant risk of respiratory compromise and autonomic instability. (15)
- Based on strong research evidence in adults and some research evidence in children, IVIG and plasma exchange hasten recovery from GBS in patients with impaired ability to ambulate. (4)(6)
- Based primarily on consensus, IVIG is the treatment of choice in children with GBS. (6)(9)(15)
- Based on strong research evidence, the prognosis for full functional recovery in childhood GBS is excellent. (2)(8)(9)

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PIR Quiz

This quiz is available online at http://www.pedsinreview.aappublications.org. NOTE: Since January, learners can take *Pediatrics in Review* quizzes and claim credit online *only*. No paper answer form will be printed in the journal.

New Minimum Performance Level Requirements

Per the 2010 revision of the American Medical Association (AMA) Physician's Recognition Award (PRA) and credit system, a minimum performance level must be established on enduring material and journal-based CME activities that are certified for *AMA PRA Category 1 Credit™*. In order to successfully complete 2012 *Pediatrics in Review* articles for *AMA PRA Category 1 Credit™*, learners must demonstrate a minimum performance level of 60% or higher on this assessment, which measures achievement of the educational purpose and/or objectives of this activity.

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1. **A 7-year-old presents to your clinic after three days of worsening, unsteady gait. Among the following, the finding most typical of Guillain-Barré syndrome is:**
   - **A. Absent deep tendon reflexes.**
   - **B. Papilledema.**
   - **C. Proximal weakness.**
   - **D. Ptosis.**
   - **E. Sluggish pupils.**

2. **You are rounding on your 12-year-old patient, hospitalized in the ICU for Guillain-Barré syndrome. Among the following, the complication MOST likely to result from her Guillain-Barré syndrome is:**
   - **A. Autonomic instability.**
   - **B. Deep venous thrombosis.**
   - **C. Diarrhea.**
   - **D. Nephrolithiasis.**
   - **E. Sepsis.**

3. Which of the following characterizes infant botulism rather than Guillain-Barré syndrome?
   A. Age >1 year.
   B. Areflexia.
   C. Ascending paralysis.
   D. Pupillary abnormalities.
   E. Respiratory failure.

4. A 10-year-old boy is diagnosed as having Guillain-Barré syndrome after a week of wobbly gait that has now plateaued. He has remained ambulatory throughout his illness, with minimal if any assistance required. At no point has there been any evidence of bulbar weakness or respiratory compromise. Of the following therapies, an appropriate treatment having the lowest potential for adverse effects would be:
   A. Ceftriaxone.
   B. Intravenous immunoglobulin.
   C. Mechanical ventilation.
   D. Plasma exchange.
   E. Supportive care.

5. An 8-year-old girl presents to the emergency department for a history of falling and progressive gait ataxia for at least a week. Among the following, the finding most specific in the cerebrospinal fluid for a diagnosis of Guillain-Barré syndrome is:
   A. Glucose 15 mg/dL.
   B. Protein 72 mg/dL.
   C. 5 red blood cells/μL.
   D. 53 leukocytes/μL.
   E. 62 lymphocytes/μL.

Condolences

The staff of *Pediatrics in Review* would like to express our sorrow at the February 17, 2012, death of Dr. Donald Lewis, a highly respected colleague and valuable member of this editorial board. An article co-authored by Dr. Lewis will appear in the May issue.

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