Guillain

Learn what to expect so this paralyzing disease doesn't paralyze you.

By Susan Simmons, PhD, RN, ARNP-BC

CE
2.0
ANCC
CONTACT HOURS

ALTHOUGH IT'S USUALLY reversible and survivable, Guillain-Barré syndrome (GBS) is a terrifying ordeal for patients and their families—and not much easier for healthcare providers. GBS is the leading cause of acute flaccid paralysis; its most common presentation is characterized by ascending, symmetric paralysis that usually affects the cranial nerves and the autonomic nervous system.[1] This results in musculoskeletal paresis and paralysis and, potentially, further complications or even death. Fortunately, GBS is rare, affecting 1 to 3 people/100,000 in the United States each year.[1,2]

The syndrome has no known cure, but with supportive care, especially improved respiratory management, and intensive rehabilitation, it resolves completely within 6 to 12 months in 85% of cases.[1,3] Full recovery can take up to 2 years.[1-5]

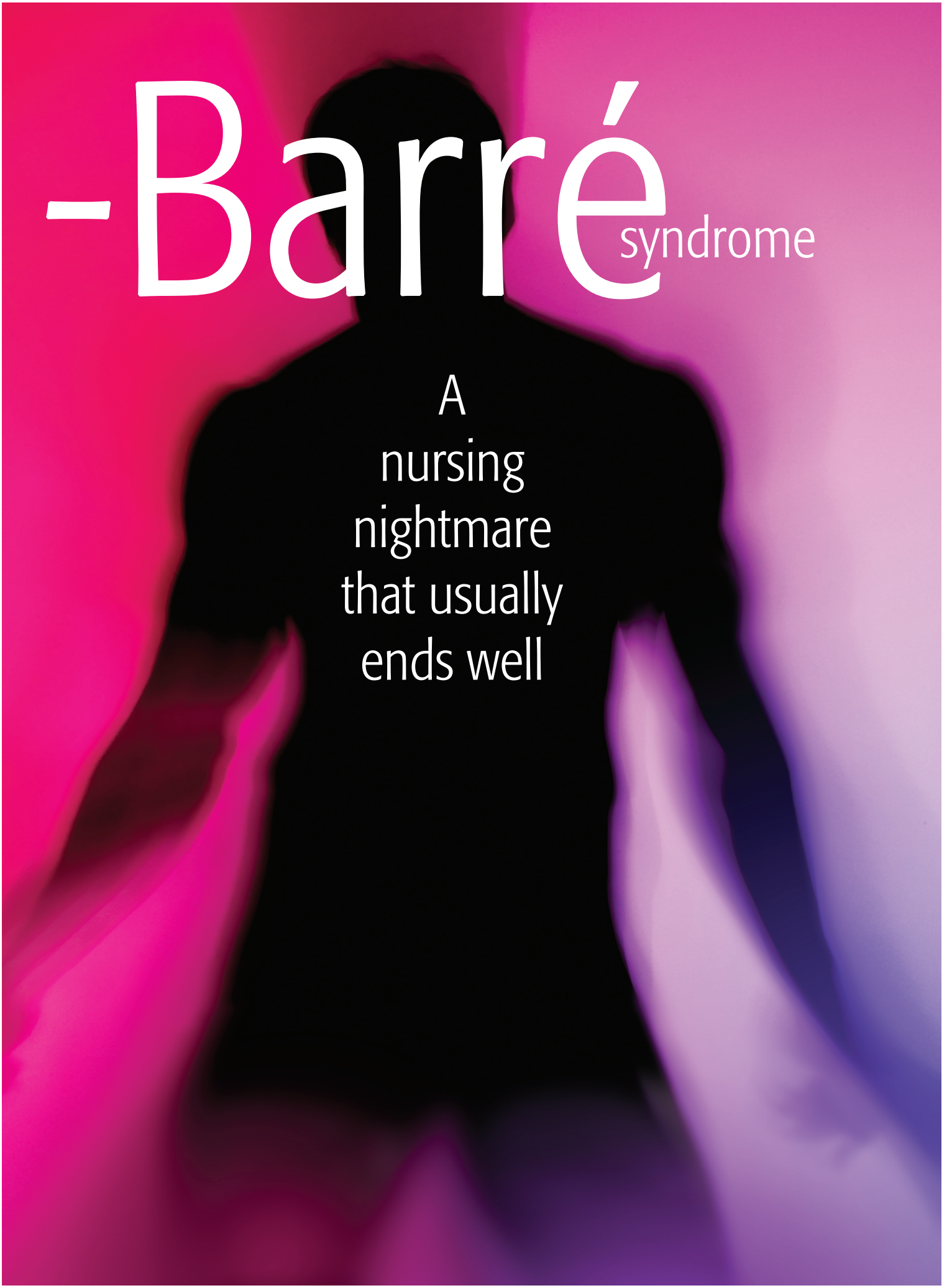
In patients who survive but don't fully recover, neurologic problems are most common.[1,3] In 2% to 10% of cases, complications of GBS prove fatal. Mortality rises with patient age.

What is GBS?

An acute inflammatory polyneuropathy, GBS has several subtypes. The most common is acute inflammatory demyelinating polyneuropathy. In the acute phase of this form of GBS, humoral and cell-mediated immune responses result in antibody production that destroys the myelin sheath surrounding cranial and spinal nerves, sometimes accompanied by varying degrees of axonal injury.[6] Highly myelinated peripheral nerves, such as motor nerve fibers, are affected more severely than less-myelinated nerves, such as those responsible for temperature and pain sensation.[2]

This first stage of GBS progression, known as the *acute stage*, begins with the onset of signs and symptoms resulting from peripheral nerve demyelination, edema, and inflammation, and ends when no additional signs and symptoms or deterioration are noted. This stage can last up to 4 weeks.[2,4,6]

When demyelination eventually subsides, the patient enters the *plateau stage* lasting from a few days to a few weeks, in which signs and symptoms remain constant.[2,4]


-Barré syndrome
A
nursing
nightmare
that usually
ends well

Finally, remyelination and axonal regeneration begins, signaling the *recovery stage,* characterized by gradual improvement in signs and symptoms.

Healing occurs in reverse: The nerve cells affected last are the first to recover.[2]

What causes GBS?

We don't know for certain. However, in two-thirds of cases, patients experience the onset of GBS signs and symptoms 1 to 3 weeks after an acute viral infection that was respiratory or gastrointestinal (GI) in origin, leading most researchers to believe that GBS may result from an autoimmune response.[2,4]

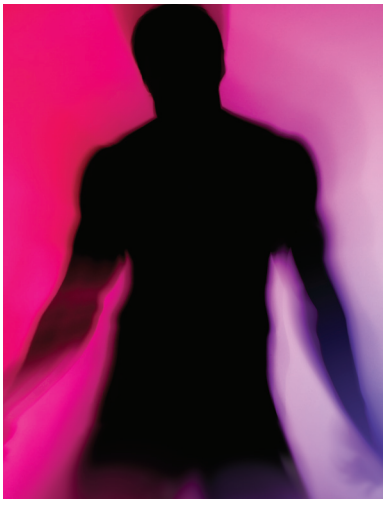
GBS can occur at any age and affects both sexes about equally. Incidence increases generally with age, peaking most sharply from ages 50 to 74.[3,5]

Besides a recent acute respiratory or GI infection, risk factors for GBS include HIV infection and, in rare instances, recent surgery, trauma, bone-marrow transplantation, or an influenza or meningococcal conjugate (Menactra) vaccination.[3]

Patients are less likely to recover well and more likely to die if they're older than 60 years, have a rapidly progressing variant of GBS, experience axonal loss, or require prolonged mechanical ventilation.[2]

Signs and symptoms: A domino effect

Signs and symptoms may take days or weeks to develop and days, months, or even years to fully disappear. The severity of your patient's condition depends on the extent of nervous system involvement—that is, the extent of demyelination. Cranial nerve involvement is present in 85% of cases, with the facial nerve (cranial nerve VII) most often affected. The glossopharyngeal (cranial nerve IX), vagus (cranial nerve X), spinal accessory (cranial nerve XI), and hypoglossal (cranial nerve XII) nerves also may be affected.



Treatment of GBS is mainly supportive, aimed at reducing severity, complications, suffering, and recovery time.

Signs and symptoms of facial nerve dysfunction include inability to smile, frown, whistle, or use a straw when drinking. Paralysis of cranial nerves IX and X can cause dysphagia and laryngeal paralysis. Autonomic dysfunction may occur if the vagus nerve is affected.

Motor weakness and paralysis usually begin in the legs, progressing to the trunk and arms. If sensory symptoms are present, patients may complain of paresthesias and pain. They also may experience mild to severe pain that's usually worse at night and most commonly involves the shoulder girdle, back, buttocks, and upper legs. Research indicates pain may be due to spontaneous firing in affected, demyelinated nerves.[1,4]

After the first few days of weakness and paresthesias, physical assessment will reveal diminished (hyporeflexia) or absent deep tendon reflexes (areflexia) in the involved extremities. Areflexia is considered a key assessment finding in GBS.[1-3] Respiratory failure is thought to be caused by involvement of the muscles of respiration, including the diaphragm and the intercostals. Up to 40% of patients have respiratory involvement.[5] GBS doesn't affect the patient's cognitive function, level of consciousness, or pupillary signs.[6]

Autonomic dysfunction is more common than once thought. Signs and symptoms include cardiac dysrhythmias, paroxysmal hypotension, orthostatic hypotension, paralytic ileus, urinary retention, and syndrome of inappropriate secretion of antidiuretic hormone.[6]

Respiratory compromise increases the likelihood that your patient will need endotracheal (ET) intubation and mechanical ventilation to ward off respiratory arrest. An intubated, ventilated patient is at risk for trauma from the ET tube and ventilator pressures, and ventilator-associated pneumonia (VAP). The patient is also at risk for malnutrition and stress ulcers.[2-4]

Recognizing GBS

Diagnostic criteria for GBS include:

- progressive weakness of two or more limbs caused by neuropathy
- areflexia
- exclusion of other causes of signs and symptoms
- history of recent viral infection
- elevated protein levels in cerebrospinal fluid (CSF) with a normal cell count on lumbar puncture
- abnormal electromyography (EMG) results (nerve conduction velocity will slow soon after the patient develops paralysis).[6]

Any patient suspected of GBS should be hospitalized and monitored for worsening signs and symptoms, especially respiratory insufficiency and dysphagia.

Medical treatment

Treatment of GBS is mainly supportive, aimed at reducing severity, complications, suffering, and recovery time. The main medical approaches today are I.V. immunoglobulin G (IgG) and plasma exchange, also known as plasmapheresis; CSF filtration is a relative newcomer.[2-4] Corticosteroids

haven't proven to be effective in GBS and aren't recommended.[3,7]

IgG therapy can shorten the recovery phase by 50%, is easy to use, readily available, and unlikely to infect your patient with diseases such as hepatitis B or C—but it's expensive.[1,3] This therapy is recommended for patients who need help to walk within 2 to 4 weeks after neuropathic symptoms start.[8] Common adverse reactions to IgG include self-limiting, flulike symptoms.[2-4] Anaphylaxis occurs rarely, mainly in IgA-deficient patients.[5,9] Other rare adverse reactions include aseptic meningitis and acute renal failure.[9] Fluid overload can occur, especially in patients with heart failure or renal insufficiency.[3]

Plasma exchange is recommended for patients who aren't walking within 4 weeks after symptom onset, and for patients who are walking within 2 weeks of symptom onset.[8] This therapy can cut recovery time in half but has significant downsides: Plasma exchange may lower immune system function and increase the risk of sepsis.[3]

The technique involves filtering problematic immune factors out of the plasma, which is replaced, preferably, with a colloid substitute (such as albumin). Plasma exchange reduces length of stay and the need for mechanical ventilation most effectively if begun within the first week of symptoms.[2,3,9] Common adverse reactions include hypotension, bradycardia, fever, chills, and rash.[4] It also requires more time, staff, and specialized equipment than IgG therapy and is more uncomfortable for the patient.[3,4] The two therapies (plasma exchange and IgG) shouldn't be used sequentially.[8]

Cerebral spinal fluid filtration, a newer technique, involves filtering out immunoglobulins along with bacteria, endotoxins, and inflammatory mediators. Adverse reactions are minimal and include headache.[2] However, the American Academy of Neurology hasn't found sufficient evidence to recommend this treatment.[8]

Even patients who can't communicate easily (or at all) can still hear, see, feel, and think.

Common issues and interventions

Considering the high likelihood of autonomic dysfunction and cranial nerve involvement in GBS, your patient will probably face these issues:

Feeling trapped, frightened, and isolated. Make communication a priority. Remind yourself and others that a patient who can't communicate easily (or at all) can still hear, see, and think, and still has sensation. Tell the patient frequently what's going on. Put a clock and calendar within view. Because of ET intubation or paralysis of muscles used for speech, the patient will need an alternate means of communication. Patients who can make a small puff of air, move their lips, blink their eyes, click their tongue, or exert even minimal pressure can use a communication board.

Don't leave the patient alone without some way to get help, such as a call device that can be activated by mouth. Arranging open visitation for family and significant others can help the patient feel less isolated.

Because of sleep pattern disturbances related to pain and altered autonomic function, the patient's sleep-wakefulness cycle may be disturbed, leading to sleep deprivation.[6] Schedule regular rest periods, which will also help to prevent ICU delirium.

If you suspect that fluctuations in your patient's heart rate and BP coincide with anxiety, consult with his healthcare provider; an anxiolytic might help. The patient might also benefit from an antidepressant through all phases of the illness.

Risk of cardiac instability. Monitor BP via an arterial line or automatic noninvasive BP monitor. Monitor hemodynamic status, via a pulmonary artery catheter if indicated.

Vagal stimulation due to supine positioning (which puts pressure on the vagus nerve), suctioning, rectal stimulation, or coughing can cause cardiac dysrhythmias. Monitor your patient for alterations in cardiac rate and rhythm and intervene as indicated. Also monitor for alterations in electrolyte balance, which can contribute to dysrhythmias.

Risk of respiratory dysfunction. Closely monitor the patient's respiratory rate, quality of respirations, Spo_2, and arterial blood gas results (especially if the patient isn't intubated). Assess respiratory depth, effort, and breath sounds frequently. Administer supplemental oxygen as prescribed, and monitor for signs and symptoms of respiratory insufficiency (such as tachypnea or dyspnea). For red flags that can signal the approach of respiratory arrest, see *Knowing when to intubate.*

ET intubation is indicated for GBS patients with decreased vital capacity, weak cough, or ineffective airway clearance. Monitor the patient's vital capacity frequently to avoid emergency intubation and increased risk of aspiration. Compare the patient's current

reading with the optimal level prescribed by the healthcare provider (usually 12 to 15 mL/kg).[6]

If your patient is intubated, suction only as needed, for the shortest amount of time needed to clear secretions. Nasogastric suctioning may be needed for gastric decompression, but can stimulate the vagus nerve and increase the risk of aspiration. Prevent aspiration by monitoring gastric residuals according to facility policies and procedures. Administer enteral nutrition (using small-lumen tubing) via the jejunum rather than the stomach. Prevent gastric distension by avoiding opioids and anticholinergics whenever possible; these medications slow gastric motility.

To reduce your patient's risk of VAP, use the ventilator bundle of interventions recommended by the Institute for Healthcare Improvement, including keeping the head of the bed elevated 30 to 45 degrees, unless contraindicated.[10]

Check the patient's maximum inspiratory pressure and forced vital capacity (FVC) every 2 to 4 hours or as ordered, as well as gag, cough, and swallow reflexes, to see when extubation may be attempted.[4] The patient may require a tracheostomy if respiratory parameters haven't improved in 2 weeks.[9,11]

Weaning can begin when FVC exceeds 30% of predicted capacity and negative inspiratory force is 20 cm H_2O or greater.[4] After extubation, continue vigorous pulmonary toileting, including incentive spirometry, to avoid reintubation and pneumonia.

Facial nerve dysfunction. Keep your patient's eyes moist with eye lubricants, such as artificial tears. Using an eye mask to gently close the patient's eyelids during sleep may be fine, but don't tape the eyes shut. Covering the eyes at all may increase the patient's anxiety.

Immobility. Implement prophylaxis for venous thromboembolism as ordered. Inspect your patient's skin daily, with special attention to areas at high risk for pressure ulcers (sacrum, back, buttocks, heels, and elbows). Minimize skin exposure to moisture from incontinence, perspiration, or wound drainage. If the patient's skin is too dry, apply moisturizers. Optimize nutrition and hydration (obtain a nutrition consult as needed), monitor weight daily, and obtain a swallowing evaluation. Use pressure-relieving surfaces and turn and reposition the patient at least every 2 hours.[10] Prevent joint contracture with passive range-of-motion exercises, physical therapy, and orthotics.

Knowing when to intubate[3,9]

Patients require intubation if they meet these three criteria:
- FVC < 20 mL/kg
- maximum inspiratory pressure < 30 cm H_2O
- maximum expiratory pressure < 40 cm H_2O.

Patients probably require intubation if they meet at least four of these six criteria:
- symptoms began less than 7 days ago
- can't lift elbows off bed
- can't lift head off bed
- can't stand up
- can't cough effectively
- liver enzyme levels are elevated.

Pain. Avoid nonepidural opioids whenever possible, as they can worsen autonomic dysfunction of the renal and GI systems.[3,9] Better options include epidural opioids or a mix of opioids and nonopioids such as carbamazepine and gabapentin (for acute or chronic pain) and long-term adjuncts such as tricyclic antidepressants, tramadol, or pregabalin (for chronic pain).[3,9] Other ways to alleviate pain include combating anxiety, repositioning, massage, music therapy, relaxation techniques, biofeedback, transcutaneous electrical nerve stimulation, ice, heat, orthotics, and physical therapy.[2,4,11]

Malnutrition. Provide nutrition that's high in calories, protein, and essential fatty acids, with reparative vitamins and minerals such as vitamin C, B vitamins, and zinc.[2,4] If the patient can't eat because of the risk of aspiration, implement enteral feeding as ordered.

Urinary retention and paralytic ileus. Monitor urine output, assess for bladder distension, and auscultate bowel sounds; assess for nausea, vomiting, abdominal pain, and constipation. Drugs to increase gastric motility are contraindicated in GBS because of autonomic dysfunction, but erythromycin or neostigmine may help with ileus.[9,11] Your patient may need intermittent or indwelling urinary catheterization.

Anhydrosis (decreased ability to sweat). Keep the skin moist with lotion and monitor temperature, intervening as needed.

Planning and education

A multidisciplinary conference early on, with family present, will make the care team's plan more cohesive and help the family understand the complexity of GBS as well as potential length of hospitalization, rehabilitation, and disability. Intensive care may be

needed for a month or more, until the patient has clearly reached the plateau phase.

When stable, your patient may be moved to a step-down or monitored unit, and eventually to a rehabilitation facility to continue physical therapy. The care team will need to help the patient and family address long-term financial, physical, and emotional concerns.

Educate your patient and the family in the ICU, but repeat the information in a calmer environment. Up to 10% of patients relapse and need to undergo further IgG or plasmapheresis.[11] From 10% to 40% of patients suffer permanent neurologic effects (including neuropathic pain syndromes, sensory ataxia, intrinsic hand muscle wasting, and bilateral footdrop). In some cases, permanent disability is severe.[1,3,5]

"Challenging" may not cover it

Because GBS has no cure, varies considerably in its course, and can be fatal, caring for patients with GBS will require all your nursing skills and powers of observation. Keeping communication channels open is crucial as you work to extricate them from this imprisoning disease. ■

REFERENCES

1. Miller A. Guillain-Barré syndrome. http://www.emedicine.com/emerg/topic222.htm.
2. Atkinson SB, Carr RB, Maybee P, Haynes D. The challenges of managing and treating Guillain-Barré syndrome during the acute phase. *Dimens Crit Care Nurs.* 2006;25(6):256-263.
3. Newswanger DL, Warren CR. Guillain-Barré syndrome. *Am Fam Physician.* 2004;610:2405-2410.
4. Sulton LL. Meeting the challenge of Guillain-Barré syndrome. *Nurs Manage.* 2002;33(7):25-30.
5. Davids HR. Guillain-Barré syndrome. http://www.emedicine.com/pmr/topic49.htm.
6. Hickey JV. *The Clinical Practice of Neurological & Neurosurgical Nursing.* 6th ed. Lippincott Williams & Wilkins; 2008.
7. Hughes RA, Swan AV, Van Koningsveld R, van Doorn PA. Corticosteroids for Guillain-Barré syndrome. *Cochrane Library.* 2006; 4.
8. Hughes RA, Wijdicks EF, Barohn R, et al. Practice parameter: immunotherapy for Guillain-Barré syndrome: report of the Quality Standards Subcommittee of the American Academy of Neurology. *Neurology.* 2003;61(6):736-740.
9. Vriesendorp FJ. Treatment and prognosis of Guillain-Barré syndrome in adults. *UpToDate.* 2006.
10. Institute for Healthcare Improvement. 5 Million Lives Campaign. http://www.ihi.org/IHI/Programs/Campaign/Campaign.htm?TabId=2.
11. Hughes RA, Wijdicks EM, Benson E, et al. Supportive care for patients with Guillain-Barré syndrome. *Arch Neurol.* 2005;62:1194-1198.

RESOURCES

Kollef MH. Prevention of hospital-associated pneumonia and ventilator-associated pneumonia. *Crit Care Med.* 2004;32(6):1396-1405.

Vriesendorp FJ. Pathogenesis of Guillain-Barré syndrome in adults. *UpToDate.* 2006.

Susan Simmons is a nurse practitioner at the Walk-In Healthcare of Olathe, Kan., and Quivera Family Care in Overland Park, Kan.

The author has disclosed that she has no financial relationships related to this article.