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Foundation International

Guillain-Barré Syndrome

**GBS: An Acute Care Guide
For Medical Professionals**

A publication of the
GBS/CIDP
Foundation International

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GBS/CIDP Foundation International

Serving the medical community and patients with Guillain-Barré syndrome
and related acute and chronic paralyzing disorders of the peripheral nerves.

Acknowledgements

Guillain-Barré syndrome (GBS) is a rare disorder. Some health professionals may not be familiar with treating it. A beautiful video by Tanya Ooraikul chronicled the superb care provided to her husband Kit during his recovery from GBS. His care at Gray Nuns Community Hospital in Edmonton, Alberta, Canada included 86 days in the intensive care unit. The video handsomely demonstrates the high quality of care that can be provided for this rare and complicated disorder in a community hospital. That video showed the value of compiling the pertinent nuances of acute care for GBS into a resource for nurses and therapists and resulted in this pamphlet. It is hoped that the information contained herein will assist acute care professionals in the care of their GBS patients. The video '*Kit's Journey*' can be seen at 'vimeo.com'.

The author of this pamphlet, Joel Steinberg, MD, PhD, was recovering from GBS in 1982 when he joined the GBS/CIDP Foundation International (originally the GBS Support Group) as a founding member and vice-president. I am indebted to several people who provided input to guide this pamphlet's preparation: my wife Susan Steinberg, RN, BS, formerly of Magee Rehabilitation Hospital, Philadelphia; Deborah Tolliver, RN, CRNP, MScN, of Anne Arundel Community College School of Nursing; Elizabeth Zink, RN, MS, CCRN, of the Johns Hopkins Neuroscience Critical Care Unit, all for general input on ICU and/or rehabilitation care; my colleagues at the Aria Health Hospital system, Drs. Gary Aaronson, pulmonologist, and Lawrence Bressler, nephrologist, for input on their respective specialties; members of the Foundation's medical advisory board, Drs. Arthur K. Asbury, Marinos Dalakas and Carol Lee Koski; Santo Garcia, OT (MOTR/L), member of the Foundation Board of Directors for input on occupational and physical therapy; Patricia Blomkwist-Markens Foundation secretary, for bringing the '*Kit's Journey*' video to my attention, and Elizabeth Emerson, JD, also a member of the Foundation Board, for general grammar editing.

Joel S. Steinberg, Montgomery County, PA 2012

An Acute Care Guide For Medical Professionals

Introduction

What is GBS? Guillain-Barré syndrome, more easily called GBS*, is a rare disorder of the peripheral nerves typified by rapidly ascending weakness, and often paralysis, evolving usually over days to one to four weeks. Paralysis is usually followed by a brief plateau period and then improvement, usually taking place over six to twelve months. In some, recovery can continue for up to two years and occasionally for as long as 5-7 years. GBS is perhaps best known to the public as the cause of paralysis from the 1976 swine flu immunization. Notable people in the public domain who developed GBS include Joseph Heller, author of 'Catch 22' and the actor Andy Griffith of TV's 'Matlock' fame.

There are several variants of GBS. By far the most common form, about 90% of cases in the western world, Europe and North America, is acute inflammatory demyelinating polyneuropathy (AIDP), the subtype discussed in this pamphlet.** In AIDP, weakness is often accompanied or even predated by a variety of sensation changes such as tingling of the fingers and feet, and/or pain in the back and proximal limbs. Paralysis can affect the muscles of respiration, with approximately one third of patients

*Older names for GBS include Landry's ascending paralysis and French polio.

**Of the other variants of GBS, acute motor axonal neuropathy (AMAN) is noteworthy. Originally called the Chinese paralytic syndrome, it was identified in rural Chinese children and young adults and frequently has its onset in the rainy season in summer and fall. Exposure to *Campylobacter jejuni*, a bacterium found in the chicken gut as well as their droppings, has been implicated as an etiology. AMAN is also frequent in Mexico and Central America. It is frequently preceded by diarrhea due to *C. jejuni* and often requires a period of respiratory support. In contrast to classic GBS, sensations are not affected, so paresthesias common to GBS, tingling, numbness, etc. are not experienced. See Section 8 on pain.

requiring mechanical ventilation (MV). In addition, irregular heart beat, cardiac arrhythmias, can occur and even be life threatening. Because of the risk of vital sign deterioration from cardiac or respiratory failure, most patients are initially placed in a monitored nursing unit and often, if MV is required, are transferred to the intensive care unit (ICU). Besides cardiopulmonary complications, other organ systems can be affected. This pamphlet highlights these issues and their care.

Pathogenesis. Approximately two thirds of GBS cases are preceded by and presumably triggered by an infection, such as the common cold or diarrhea. Current theory holds that the patient's immune system correctly identifies an infecting agent as foreign and mounts an immune attack. However, molecular structures on the surface of the nerve may closely resemble or mimic structures on the infecting agent. The similar appearance of surface structures on certain infecting agents and on nerves likely explains why the immune system may, in some persons, recognize nerve as foreign and, in error, attack it. The result of that transient nerve damage is GBS. Since the patient's own immune system does the injuring, GBS is considered an auto (against self) immune disorder. Typically, the nerve's myelin and sometimes the axon is attacked and injured after which healing begins.

Daily diligence. Mainstays of treatment for GBS include supportive care such as mechanical ventilation, rehabilitation and immune system modulation, via immune globulins or plasmapheresis. Paralysis and respiratory insufficiency place the patient at risk for nosocomial problems. A key to good outcome is diligent daily bedside attention to detail by the patient's care team. Consistent high level care will reduce the risk of such potential complications as deep venous thrombosis, stress ulcers, decubiti, atelectasis, etc.

Multidisciplinary care. Because of the potential for complications involving several organ systems, GBS is often treated by a multidisciplinary team of professionals: nurses, therapists and

physicians. The medical chart provides a means for them to communicate. Hence it is important that the chart contain clear concise information.

GBS/CIDP Foundation International. The Foundation, a non-profit 501(c)3 organization, is dedicated to the well being of patients with GBS and related disorders. This pamphlet is part of our services of support, education, research and advocacy.

Patient support. Most patients and families have not heard of GBS before their illness. The Foundation offer an *Overview for the Layperson*²¹ (Steinberg) designed to educate patients about this disorder. In addition, we assist interested persons to develop local support group chapters to educate and assist new patients. Contact us for our literature, a starter pack to help create a chapter, and other support/educational material, at:

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Note: Some medication recommendations in this pamphlet do not reflect FDA approval. To help apply the information contained herein, use clinical judgment and/or consult with appropriately trained and experienced specialists.

Initial Patient Evaluation

The newly admitted patient with suspected or diagnosed GBS warrants a disorder-specific initial assessment, a.k.a., intake evaluation, in addition to the basic history and physical exam.

Key issues that may serve to better treat the patient include the following:

History.

- 1) How many days have lapsed from symptom onset to hospital admission? Patients presenting within a few days are more likely to develop respiratory failure than those who present after a week.
- 2) Was there a precipitating infection? About two thirds of GBS cases follow within days to three weeks of an infection such as the common cold or diarrhea. Those with diarrhea are prone to have a more difficult clinical course. (See Appendix C)

PE. What is the patient's extent of weakness? Patients with substantial weakness, beyond impaired walking, such as loss of naso-labial fold and/or cranial nerve or bulbar palsy, with, e.g., choking (see end of Section 4) are more likely to develop respiratory failure and need mechanical ventilation or at least airway protection via intubation.

Upon admission, document the patient's functional status via, e.g., the Medical Research Council Scale

Grade 5: Normal strength.

Grade 4: Strength is reduced but muscle contraction can still move against resistance.

Grade 3: Limb can move against gravity but not against any added resistance.

Grade 2: Limb can only move if resistance to gravity is removed. For example, an arm can be slid on a bed but not raised.

Grade 1: Only a trace or flicker of movement, e.g., fasciculations, are seen or felt in the muscle.

Grade 0: No movement observed

Reflexes: In GBS, deep tendon reflexes become diminished or absent by four to five days after onset of neurologic symptom. If DTRs remain normal or hyperactive, consider other diagnoses.

Biochemical/laboratory evaluation. The differential diagnosis for GBS includes the several other causes of peripheral neuropathy. Accordingly, consider obtaining the following studies in addition to a standard compilation of tests (chest x-ray, electrocardiogram, urinalysis, pre-albumin to track nutrition, etc.): TSH, HbA1c, urine for heavy metals; urine for total porphyrins and their metabolites, delta-aminolevulinic acid, porphobilinogen; titers for Lyme disease, HIV; skin inspection for a tick bite, indicative of tick paralysis.

Cerebrospinal fluid (CSF): GBS patients usually develop cytoalbumin dissociation, that is, elevated CSF protein with normal cell count, within 10 days of onset of the disorder. If this is not found, entertain other diagnoses or the result of an early lumbar puncture. However, occasional patients with an ongoing viral infection, such as HIV or Lyme as the trigger of their GBS, may have cells in the CSF. Also of note, occasionally the finding of a normal protein level should not rule out GBS if other findings support that diagnosis.

Nerve conduction velocity-electromyography (NCV-EMG) testing will show slow or blocked nerve conduction. Other NCV markers of GBS include delayed F waves from nerve root demyelination, decreased amplitude of compound muscle action potential, and prolonged distal latencies. In the less common axonal form, acute motor axonal neuropathy (AMAN) conduction is normal but the nerve's action potential is small.

Respiration. Because breathing is often affected in GBS, it is prudent, along with admission vital signs, to obtain a set of baseline measurements of ventilation⁶ (Gorson). These include spirometry measurements of diaphragm function, e.g., maximal (negative) inspiratory force (NIF) and vital capacity, as well as measurements of ventilation, e.g., arterial blood gas or oxygen saturation by pulse oximetry. As respiratory muscle function declines, respiratory rate increases to compensate and maintain

oxygen levels while carbon dioxide rises. Non invasive carbon dioxide monitors are available in some units. In patients with chronic lung disease an arterial blood gas (ABG) may be more instructive.

Natural History of GBS: Implications for Patient Care

Natural history/outcomes. GBS can range from the mildest case, with only some gait ataxia and tingling of the toes, to the most severe, with respiratory failure and total paralysis. Most patients have never heard of this disorder and are thus understandably scared about their altered state. Fortunately, even those who require mechanical ventilation usually have a good outcome. The majority of patients, about 75%, will return to their pre-GBS health status within months to a year or two from symptom onset.

About 20% will have long term disability and require some assistive device (wheelchair, walker or cane) to get around. And about five percent of GBS patients die, usually from cardiopulmonary complications. Recurrence of GBS is rare, occurring in less than five percent of patients. During recovery, fatigue and neuropathic pain are not uncommon. And rarely, despite treatment, a patient may go on to develop a slower onset, recurrent cousin of GBS, called chronic inflammatory demyelinating polyneuropathy (CIDP).

Cautious optimism. Because most patients have a good outcome, caregivers, including medical professionals, may help by conveying optimism, tempered by the individual patient's particular medical status.

Communication. Hearing is only rarely impaired in GBS. Even if the patient is on a ventilator and locked in, that is, has total paralysis of limbs, extraocular muscles, etc., and so is unable to move voluntary muscles to communicate, they can usually still hear. Caregivers should converse at the bedside as though the patient can hear, and be mindful of what is said.

On approaching the patient, introduce yourself by name and job title or description and explain planned procedures so that the

patient is not unnecessarily scared. For patients on a ventilator and unable to speak, the Foundation provides a set of Communication Cards. This bound set of cards contains many topics of interest to GBS patients. The interviewer can point to them and the patient can blink, nod their head or communicate otherwise to provide 'yes' or 'no' answers about issues of concern. Table 1 below offers some suggestions to help allay patient and family concerns.

Table 1. Measures To Reduce Anxiety in the Acutely Ill GBS Patient and Family/Friends²¹ (Steinberg)

- Express optimism about a relatively good chance for recovery.
- Provide the patient on a respirator with a communication method to reduce frustration. *Communication Cards*, available from the GBS/CIDP Foundation, list in large print common patient problems. A nurse or family member can flip through these cards at the bedside, point to various items and get the patient's "yes" or "no" response by a head nod, eye movement to the right or left or eye blink.
- Explain procedures at the bedside to alleviate anxiety.
- Identify one family member to serve as contact with a hospital representative with good rapport (a physician or nurse), to provide accurate information on the patient's status and care plans. Having multiple members call risks confusion.
- Encourage frequent visits by family and friends to provide emotional support.
- A clock, electric calendar, radio and night light in the ICU room helps the patient track day and night hours, maintain awareness of the outside world and minimize confusion.
- Allow the patient to express emotions (anger, frustration, and fear) and help them deal with these issues. Consider a psychology consultation.
- Reduce the patient's sense of isolation during a prolonged hospital stay by inviting visitors to engage in bedside activities such as grooming, reading get well cards, etc.

Respiratory complications

Introduction. Respiratory failure occurs in about one third of GBS patients as part of this disorder's typical progressive ascending paralysis. The likelihood of respiratory failure is greater in patients with three features: 1) substantial weakness beyond difficulty walking; 2) medical care required within a few days of onset because symptoms develop rapidly; and 3) facial muscle or cranial nerve weakness (bulbar palsy) with such symptoms as choking or poor secretion handling²³ (Waalgard). Patients with a slower onset of illness may present after several days of symptoms and have a lower risk of respiratory failure – although it can develop subtly. Accordingly, early and regular monitoring of respiratory status is prudent until the patient's disorder plateaus and shows improvement. Even then a rare patient will relapse and again require ventilatory support.

To determine a need for mechanical ventilation consider both the clinical bedside evaluation of breathing as well as measurements of ventilation, i.e., oxygen saturation (by pulse oximetry) and spirometry measurements of diaphragm strength, i.e., negative inspiratory force and vital capacity. These criteria are detailed below.

Frequency of evaluating the patient can be guided by their pre-admission course. A more abrupt course, with only a few days from first symptoms until presentation plus facial weakness, warrants more frequent monitoring, perhaps every one to two hours rather than q four to six hours. Frequent assessment of strength and spirometry can help identify the failing patient sooner but also risks fatigue.

Clinical markers of failing respiration include:

1. the triad of:
 - a. rapid and substantial weakness, occurring over a few days, with, for example, inability to lift elbows or head off the bed,¹⁰ (Mehta) especially if accompanied by either
 - b. facial weakness (e.g. flattening of naso-labial fold,

inability to smile), and/or

- c. bulbar palsy (cranial nerve damage), indicated by, e.g., difficulty swallowing, with choking, coughing or drooling, absent gag reflex or slurred or weak speech;
2. shallow or rapid breathing with poor breath sounds in the bases of the lungs;
3. staccato speech, so only a few words are spoken with each breath;
4. autonomic instability (dysautonomia) as indicated by, e.g., fluctuating blood pressure and/or heart rate;
5. tachycardia and/or brow sweating, from stress induced adrenergic drive; and
6. paradoxical breathing, i.e., inward movement of abdominal muscles during inspiration, reflecting diaphragm fatigue; and/or episodic use of accessory muscles of respiration.

Lab markers of failing respiration include:

1. oxygen saturation less than 92%; $p\text{CO}_2 > 45$ mmHg, per ABG, in patients without chronic lung disease;
2. inspiratory effort (IF) (force generated by a maximal inhalation) less than 15 cm H_2O . Values > 25 are usually safe;
3. decreased vital capacity (VC) (measured as a full expiration after a deep inspiration), of $< 15\text{-}20$ ml/kg;
4. rapid fall, e.g., $> 30\%$, of IF or VC within 24 hours;
5. in the fatigued patient even a VC fall to 18 ml/kg may suffice to ventilate;
6. a drop of 500 ml in VC;
7. inability to count to over 10 with one breath provides a quick bedside marker of a VC down to 1 liter, signaling impending failure. A normal VC allows counting to twenty¹⁷ (Ropper); and
8. inconsistent or falling values of VC or IF at a single test session.

These findings may signal fatigue and/or the patient's weak lip grip of the spirometry mouthpiece.

Additional factors to guide intubation include the patient's age and chronic medical conditions, such as diabetes, obesity and/or chronic lung disease.

For patients on a ventilator additional care guidelines may apply:

1. **Stress ulcers:** to reduce the risk of this and other causes of upper gastrointestinal bleeding, use an H2 receptor blocker (e.g., famotidine [Pepcid®]) or proton pump inhibitor (e.g., pantoprazole [Protonix®]);
2. **urine retention:** prevent this with an indwelling bladder catheter (Foley);
3. **decubitus and foot drop prevention:** use modalities as listed in section 10, e.g., alternating pressure mode of specialty bed, frequent repositioning-e.g., every 2 hours, heel off-loading, ankle-foot orthosis, etc.
4. **oxygen desaturation:** if this occurs look for causes, such as atelectasis, mucous plug, endotracheal tube migration into a main stem bronchus, pulmonary embolism, pneumothorax, or new infiltrate;
5. **atelectasis:** treat with chest physical therapy, postural drainage, etc. and
6. **tracheostomy:** plan for this in the event that ventilation beyond two weeks is required.

Weaning

Discontinuing ventilator support is a two step process. First determine readiness to wean and then initiate weaning.

Monitor for returning strength, such as the presence of head, eye and shoulder movement, as a clue to weaning readiness.

Prior to weaning the patient should be hemodynamically stable and medications with sedating properties, e.g., opiates and anxiolytics,

should be discontinued. Daily interruption of these agents (sedation vacation), as clinically deemed safe, has been found to shorten duration of ventilation. Sedation vacation allows for assessment of neurologic status and measurement of vital capacity, key factors in determining the GBS patient's readiness for weaning.

Common methods for weaning include a T tube trial and pressure support ventilation.

In a **T tube trial**, intervals of spontaneous breathing off ventilatory support are provided through a T tube circuit. Limit the trial to two hours or less to determine if the patient is ready for extubation. If the patient fails they should be returned to full ventilatory support for 24 hours prior to reattempting weaning.

A potential disadvantage of the T piece trial is the lack of connection to a ventilator, thus requiring close supervision and demands on nursing staff. Return of neuromuscular function in GBS occurs slowly, in contrast to its abrupt onset. Thus with a T piece trial, care must be taken to assure that the time between T piece trials, that is, time on the ventilator, is sufficiently long so as to not exhaust the patient.

With **pressure support ventilation (PSV)** (i.e., pressure assisted ventilation) elevated airway pressure is utilized during each inspiration and discontinued during expiration. The additional inspiratory phase pressure, called the pressure support level, is used to lessen the work of breathing by improving ventilation. Pressure support compensates for the work of breathing caused by the resistance of the endotracheal tube and respiratory circuit and may also help improve alveolar expansion and thus gas exchange.

To begin the weaning process with PSV, set the ventilator to an initial optimal level of pressure that will enable the patient to generate a tidal volume of 10-12 ml/kg with a decreased respiratory rate. Accomplish this by increasing the PSV level, starting at a baseline value of 15 to 20 cm H₂O, in increments of 3 to 5 cm. Continue the incremental increase until the patient shows a decrease in respiratory rate while maintaining the tidal volume of 10 to 12 ml/kg. Gradually reduce pressure support,

based on the patient's clinical tolerance, by 3-8 cm water/H₂O while monitoring that the patient is maintaining their tidal volume. This method slowly transfers the work of breathing from the ventilator to the patient. Once the patient is stable on low pressure support, an extubation trial should be initiated.

Noninvasive ventilation. Patients with hypercapnic (elevated CO₂) respiratory failure can benefit from noninvasive positive pressure ventilation. Several trials have suggested that the use of inspiratory and expiratory or bilevel positive airway pressure (BiPAP) post extubation lowers mortality rate, lessens duration of ventilation and lowers the rate of nosocomial pneumonia.

Avoiding mechanical ventilation. In the patient with slowly evolving weakness, incentive spirometry and/or mini-nebulizer treatments may provide sufficient oxygenation to stave off mechanical ventilation. Do continue monitoring respiration in such a patient.

Bulbar palsy. Lower cranial nerves, IX through XII, come off the brain stem or bulb. Dysfunction of these nerves, called bulbar palsy, is typified by such findings as poor gag reflex, poor secretion handling, weak speech, choking or drooling. Patients with bulbar palsy are at risk for aspiration and high morbidity pneumonia. Provide these patients with airway protection via intubation even if oxygen saturation is acceptable.

BiPAP. Non-invasive mechanical ventilation via BiPAP is an unproven means to provide safe ventilation in the GBS patient and should therefore only be considered in selected patients^{16, 24} (Pearse, Wijdicks).

Dysautonomia and Cardiovascular Complications

Peripheral nerves of the autonomic nervous system – both sympathetic and parasympathetic branches – can be damaged in GBS. The result is dysfunction of the organ systems that it regulates, called dysautonomia. Potential complications of dysautonomia are described in this and the following sections.

Hypertension can be treated by any of the several classes of drugs commonly used for this problem. Short acting drugs, such as hydralazine, and labetalol are best used initially in case the pressure fluctuates or elevations are short lived¹² (Miller). If elevated pressure is accompanied by tachycardia, a beta or calcium channel blocker may provide practical treatment for both.

Hypotension is less common. It can be triggered by a seemingly innocuous event such as bringing the patient to a sitting position. The longer the patient is at bed rest, the more commonly this is seen. Potential treatments include leg elevation to return more blood to the central circulation, intravenous fluids to expand intravascular volume, compression hose, and, rarely, medications such as midodrine or IV vasopressors (e.g., norepinephrine, phenylephrine).

Sinus tachycardia may reflect dysautonomia of GBS. First rule out more common causes such as dehydration, fever, hypotension, infection, etc. If determined to be attributable to dysautonomia, consider treatment with chemical blockade, with, e.g., beta or calcium channel blockers.

Bradycardia is uncommon but can be severe. It can be triggered by seemingly benign procedures such as inserting an intravenous line or by otherwise stimulating excessive parasympathetic discharge, so called 'vagal spells.' For symptomatic sinus bradycardia, try 0.5 mg IV atropine. Ongoing symptomatic bradycardia (e.g., second degree, type II and third degree) warrants temporary pacing² (Flachenecker); asystole may occur in rare instances, requiring implantation of a pacemaker¹¹ (Mehul).

Deep venous thrombosis (DVT) is best prevented in critical care patients with subcutaneous administration of unfractionated or fractionated, i.e., low molecular weight, heparin, +/- intermittent pneumatic limb compression. Warfarin (Coumadin®) can be considered for the rare chronically bed bound patient.

Efficacy of mechanical methods (pneumatic or elastic compression) alone for prophylaxis in critically ill patients is unclear. Intermittent pneumatic compression alone, without heparin, is reserved for patients with high bleed risk⁵ (Geerts). If elastic compression is to

be used, consider thigh length compression gradient stockings, class II, 30-40 mmHg at the ankle. Anti-embolism stockings are of questionable value for prophylaxis. Early mobilization to prevent clots is usually impractical for GBS patients.

Bladder, Bowel Dysfunction

Urinary retention may occur as part of a dysautonomia picture, reflective of failing bladder reflexes, inability to sense bladder fullness and/or inability to relax the urethral sphincter²⁰ (Sakakibara). In older men, formerly compensated prostatic hypertrophy may become symptomatic. Treat the patient with an indwelling bladder catheter, that is, a Foley, or, for men with a resistant prostatic urethra, a Coude (bent) catheter. As strength improves plan a voiding trial.

Constipation may reflect parasympathetic dysautonomia with paralytic or adynamic ileus²⁵ (Zochodne). Poor gastric motility and emptying is less common. Other contributing factors may be inability to sense a full colon and inability to perform a 'push down' maneuver as well as issues common to many hospitalized patient, i.e., strange food and/or environment. Treat with any of the usual methods for dealing with constipation, such as dispensing prunes, milk of magnesia, dioctyl sodium sulfosuccinate (Colace[®]), psyllium (Metamucil[®]), lactulose (Chronulac[®]), polyethylene glycol (Miralax[®]), senna (Senekot[®], Ex-lax[®]), bisacodyl (Dulcolax[®], Correctal[®]), enemas, digital extraction, etc., depending on preferences of the patient and/or practitioner and severity of the problem.

Metabolism: Nutrition, Hydration, Electrolytes

Nutrition. In spite of bed rest and weakness, GBS patients, especially those on a ventilator, not infrequently have increased energy/caloric needs¹⁸ (Roubenoff). Factors contributing to this include:

1. pre-admission weight loss from, e.g., viral syndrome with decreased appetite and/or diarrhea;

2. poor intake due to bulbar palsy, with poor gag reflex and poor oral intake;
3. ileus with poor digestion and absorption; and
4. a combination of respiratory failure, endocrine, infection and inflammatory changes that create a hypermetabolic state.

Meet greater energy/calorie and/or protein needs by providing appropriate nutrition, via, as clinically indicated, a practical route, i.e., p.o., nasogastric, gastrostomy (PEG) or parenteral.

Nutrition options include, as clinically indicated, 1) elevated calorie feedings, e.g., 40-45 non-protein kcal/kg body wt./day, via carbohydrate and/or lipid sources; and/or 2) high protein feedings (2.0 to 2.5 g protein/kg body wt./day).

Consult with the dietician and/or surgical nutrition team to expedite appropriate nutrition per the hospital diet/formulary system. Utilize indirect calorimetry to assess caloric needs.

Positive protein balance limits muscle wasting, supports overall improved health and healing, supports visceral protein repletion to attain gastrointestinal tract integrity and promotes resistance to infection. Serum lab measurements as well as twenty-four hour urine collection for measurement of total urinary nitrogen and urine urea nitrogen can be used to calculate nitrogen balance⁸ (Mackenzie). Serum pre-albumin provides a ready marker of protein status. A low value with 'adequate' protein intake suggests a malabsorption or mal-utilization issue.

Hydration can be compromised by several factors including greater insensible water loss accompanying mechanical ventilation, infection and stress-related heightened sympathetic drive. Clues to poor hydration and intravascular volume depletion include poor urine output, hypotension, resting tachycardia, elevated urine specific gravity of > 1.020, dry mucous membranes and/or axillary skin, skin tenting, etc. Provide hydration as clinically warranted with p.o. or intravenous fluids and/or enteral water administered through a feeding tube. Tube feedings and even total parenteral nutrition do not always provide adequate free water to maintain

hydration. Supplemental free water, given intravenously or via a feeding tube may be warranted.

Electrolytes

Hyponatremia in GBS can be due to renal retention of water due to excessive secretion of anti-diuretic hormone, i.e., syndrome of inappropriate antidiuretic hormone secretion (SIADH) rather than by renal dysfunction. Up to one third of patients with GBS may develop hyponatremia from SIADH, in particular, those who require mechanical ventilation or are critically ill. In contrast to other and more common causes of hyponatremia, SIADH is characterized by inappropriately elevated urine osmolality from increased renal reabsorption of free water. This leads to the higher urine concentration, accompanied by decreased serum osmolality from retained water that dilutes the blood.

Treatment options for SIADH include fluid restriction or a combination of a loop diuretic (furosemide [Lasix®]) to generate a salt and water diuresis while also giving intravenous saline or hypertonic saline to replace sodium. If water restriction is not effective, consider, under guidance of a nephrologist, a course of medications. Demeclocycline (Declomycin®) increases renal water excretion, but may cause renal failure especially in patients with concurrent liver disease. The vasopressin receptor antagonists, Conivaptan (Vaprisol®) and tolvaptan (Samsca®) induce a water diuresis and may be useful in unusual select cases. When treating patients with hyponatremia, the rate of correction should not exceed 10meq/L during the first 24 hours and 18 meq/L over the first 48 hour period to avoid the possible complication of osmotic demyelination. A more rapid rate of correction, up to 4-6meq/L over the first two to four hours, may be necessary for severely symptomatic patients.

Pain

Early nociceptive pain. Even before weakness develops an occasional GBS patient may experience a variety of sensation problems. These often take the form of tingling, of the feet and/or fingers. Pain may be another initial complaint, felt in

the back or proximal limbs (thighs, hips, buttocks, shoulders and between them). This is nociceptive pain, that is, traditional pain due to tissue damage, perhaps from muscles¹² (Ono) trying to contract with inadequate innervation. Nociceptive pain can continue during the early hospital course and warrants treatment to improve patient comfort. Treatment options to be considered include non-steroidals (e.g., ibuprofen, or a short course of IV ketorolac [Toradol®]) to opioids such as morphine. Short acting agents can be tried initially in case the pain is short lived. Consider longer acting agents if pain persists. Exercise caution in the use of opiates since they can suppress respiration and cause grogginess. In patients protected with mechanical ventilation, opiates can be used more generously. For severe leg pain consider epidural anesthesia with morphine to avoid its systemic side effects. For prominent night time pain consider a trial of warm packs and massage.

Later neuropathic pain. Neuropathic pain from nerve demyelination tends to occur later in the course of GBS and can be a very real problem even without tissue injury¹⁴ (Parry). It can take several forms, such as frank pain, burning, tingling, electrical sensations, a sense of the body vibrating or formications, i.e., a sense of worms crawling under the skin. Typical treatment approaches include traditional analgesics such as acetaminophen but more often pain responds to a combination of newer anti-seizure and neurotransmitter inhibiting agents: gabapentin (Neurontin®) up to 4 g a day in divided doses (start low, 100 mg bid and increase dose slowly); pregabalin (Lyrica®); duloxetine (Cymbalta®); venlafaxine (Effexor®). These agents are often used in combination with a tricyclic antidepressant such as amitriptyline (Elavil®) or nortriptyline (Pamelor®) to provide greater efficacy.

ICU Delirium

Confusion or frank psychosis may be another issue for the ICU patient, especially with a protracted course of GBS. It can take several forms, such as hallucinations, agitation, nightmares, aggressive or other unusual behavior, paranoia or hearing voices.

Contributing factors may include an ICU unit without windows to differentiate day and night, disturbances of normal sleep duration and/or cycle (disturbed biorythm), pain, noisy vital sign monitors, critical illness pathophysiology, mind altering drugs (narcotics, benzodiazepines, other hypnotics and corticosteroids) and subclinical dementia that is exacerbated by the in-patient experience. Before treating the behavior change, rule out and/or address offending medical factors such as hypoglycemia, evolving sepsis, dehydration, etc. Treatment approaches include a calendar that marks the day, a radio station that frequently announces the time, frequent input from care givers and visitors as to the time of day (remember to say AM or PM with the time) and day of the week as well as removal of offending medications. Drugs such as haloperidol (Haldol®), quetiapine fumarate (Seroquel®) or olanzapine (Zyprexa®) may be considered to control agitation but should be used in moderation.

Skin

The paralyzed patient is at increased risk of developing a pressure ulcer or decubitus. The sensory loss of GBS adds to this risk: the patient may not be aware that his or her skin is under continued pressure and/or may not have the strength to reposition themselves. Utilize standard methods to prevent bed sores, including the following:

1. **Inspect**, *during each shift*, the skin overlying boney prominences (heels, lateral malleoli, sacrum, hips, pinna, elbows, etc.) for stage I decubiti, that is, non-blanching erythema upon brief compression of the skin.
2. **For stage I pressure wounds** implement off-loading (e.g., float heels via foam splints or pillows under the calves); for sacral or buttock skin damage, use frequent side to side repositioning and/or the low or alternating pressure setting of a specialty bed. Utilize a 'time and sign' sheet to document repositioning. Topical agents that contain hydrogel (e.g., Curasol®), petrolatum (e.g., Aquaphor®) or lanolin may help improve skin quality but do not replace aggressive off loading.

3. **For stage II decubiti**, i.e., partial skin thickness opening, add, along with off-loading, topical protective wound healing agents such as a hydrogel to moisturize the wound, petrolatum/lanolin product (e.g., A and D ointment), antibiotic such as silver sulfadiazine (e.g., Silvadine®), etc.
4. **For persistent or deeper wounds** consult a wound specialist to guide further care.

Musculo-Skeletal Issues, Occupational and Physical Therapy

Occupational and physical therapy (OT, PT) are part of the mainstay of care for GBS patients. Initiate physical medicine/rehabilitation modalities shortly after the patient's admission, even if in the ICU, as part of the 'attention to detail' approach. Early therapy with prn physiatry oversight helps prevent such untoward problems as skin breakdown, flexion contractures and other joint dysfunction.

Key elements of an OT/PT program include off-loading of boney prominences, joint and limb positioning, and foot drop prevention. Customize the program to the patient's degree of weakness. For example, GBS patients may experience orthostatic hypotension from dysautonomia and benefit from use of a tilt table to provide safer physical therapy. For additional details about occupational and physical therapy for the GBS patient, see the Foundation's brochure on this subject.

Fatigue. Fatigue that comes on with sustained active exercise is a feature unique to GBS. It is often heralded by paresthesias. During the early course of GBS, patients may not be ready for active exercises. If active exercises are begun, therapists should discontinue them when fatigue or paresthesias begin. Otherwise fatigued muscles will require time to recover, regain their baseline strength and thus delay, at least temporarily, the recovery process. Utilize methods to assure a good night's sleep to help provide a well rested patient who can more effectively participate in day time therapy. Accordingly, try to limit night time activities such as

vital sign measurement, repositioning, awakening the patient to take medications, etc. Hypnotics may also be helpful.

Note that fatigue can be a common problem after recovery and probably is related to extensive reinnervation. This happens even to ambulatory GBS patients after recovery and should be treated with periods of rest and non-fatiguing exercises.

Frog Position. An often comfortable position for the paralyzed patient is the 'frog' position, shown in Figure 1, with the patient supine, arms at the side, with elbows slightly flexed on pillows. Position wrists slightly cocked up, at about a 30° angle, in the so called 'functional' position (the position most frequently used during function). Fingers are gently flexed and thumb flexed in abduction. If the patient is sufficiently weak, a cock up wrist splint can provide this functional position.

Externally rotate the legs and draped them over pillows so the knees are gently flexed, with the heels floating in air, off the bed surface. Oversight by OT and PT specialists may help to institute correct functional positioning, with, e.g., the prn use of splinting.

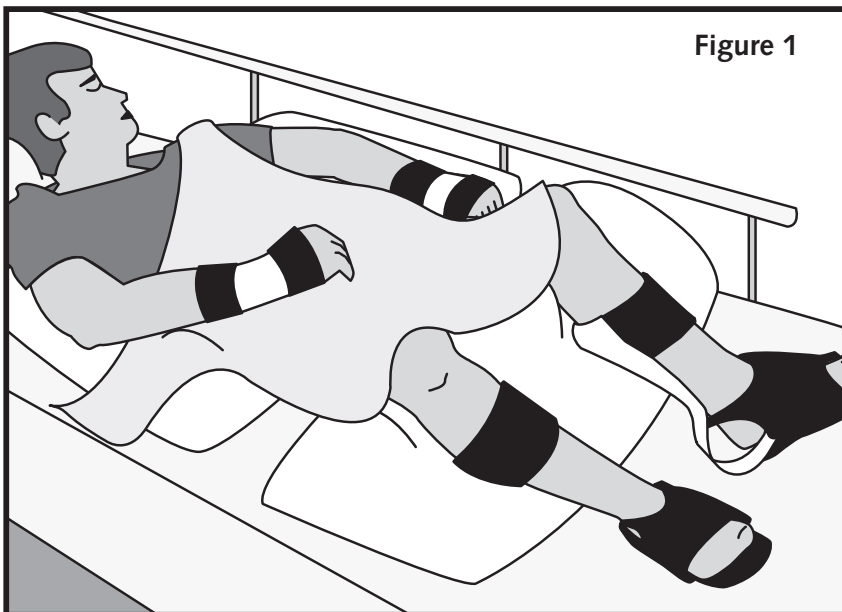


Figure 1

Foot drop. The weak patient at bed rest is subject to passive plantar flexion of the feet (a.k.a., foot drop) due to gravity. Prevention is warranted as, if this persists, the Achilles tendon can permanently shorten. The patient then cannot walk on their soles of the feet but rather will be forced to walk on their toes and surgical release is required to correct a shortened tendon. Accordingly, maintain feet at 90° to the leg. This can be provided via a light weight splint such as the Heelift® by DM Systems® or a comparable product which simultaneously off-loads or floats the heels. See Figure 1. In addition, range of motion/stretching exercises can be performed to assure normal range of the ankle joint, to about 10° dorsi-flexion. This will enable the normal gait with its push off phase.

Side to side repositioning. Implement a side to side regimen, using foam wedges behind the back or pillows for comfort, and, as importantly, to repeatedly off load the buttocks and back. Use a 'time and sign' sheet to document regular side to side off-loading. Some specialty beds have a decubitus prevention modality to provide alternating pressure and redistribution to help prevent and treat bed sores. During the night reduce frequency of turning to facilitate uninterrupted sleep and reduce limb pain if present¹⁷ (Ropper).

Compression nerve palsies. Some nerves pass over superficial bony prominences. If these sites are subjected to prolonged pressure function of the innervated muscle may be compromised. Accordingly, these areas should be off-loaded or cushioned. Principle sites at risk for nerve compression are the ulnar nerve at the medio-posterior aspect of the elbow, at the cubital tunnel (a.k.a., 'funny bone') and peroneal nerve at the antero-lateral aspect of the knee. Prevention is warranted as peroneal nerve damage can result in foot drop and loss of sensation of the dorsal foot.

Infection

The GBS patient is prone to infection, especially if paralyzed and on a ventilator. Common sites are the lungs, urinary tract, central

venous catheter, trachea and sinuses. Distinguish colonization from frank infection to avoid premature use of antibiotics and risk development of resistant organisms. Clues to infection include fever, white blood cell count elevation or shift, infiltrate on chest x-ray, productive cough and a cloudy urine.

Disorder specific treatments

High dose intravenous immune globulin (IVIG) and plasmapheresis, a.k.a., plasma exchange (PE) have been found to accelerate GBS patient improvement such as time to independent walking, and, if on ventilator, reduce time on mechanical ventilation. Because of its ease of administration and availability, IVIG has become a favored option. To achieve optimal benefit from their use either is preferably started within two weeks of onset of GBS symptoms. Giving PE followed by IVIG has not been found to provide greater benefit than using either treatment alone¹⁵ (PE/Sandoglobulin). Given that GBS is an inflammatory disorder, use of anti-inflammatory drugs, such as corticosteroids, would seem like an obvious treatment option. *However, systematic studies have found that 'steroids' not only do not improve the patient but may actually prolong their illness.*

IVIG is usually dosed at 0.4 g immunoglobulin/kg body weight daily for 5 days, to give a total course of 2 g/kg. Complications and side effects are uncommon and usually mild, such as headache, chills or muscle aches. These often can be managed with non-steroidal anti-inflammatory medications such as ibuprofen, and/or slowing the infusion rate. Other potential side effects include fever, hypertension, lightheadedness, flushing and renal insufficiency. More annoying side effects can often be handled with the administration, 30 minutes before the IVIG is started, of steroids (e.g., methylprednisolone, 60-100mg IV) and diphenhydramine, 25-50 mg IV. Another option is to switch to a different commercial product as they do have some compositional differences⁷ (Koski) with respect to osmolality, ph, and sugar and sodium content.

PE is administered via catheters into the vein, to draw blood out. The PE machine separates cells and discards the plasma; the cells are then mixed with albumin solution and returned to the patient. The exchange rate is usually 200-250 ml/kg body weight in exchanges divided over five to 14 days^{4,9} (GBS Study Group, McKhann). Each exchange takes about two to three hours. Side effects and risks include catheter related bleeding, infection and clots, hypotension and allergic reactions to replacement fluid, even anaphylaxis. Less common complications include arrhythmia from salt imbalance and citrate-induced hypocalcemia. PE reduces the platelet count and removes clotting factors. But these return to normal within 24 hours, except in the presence of liver disease. Catheter placement carries its usual risks of hematoma, pneumothorax, etc., depending upon site of insertion. In spite of such potential complications, the actual risks with PE have been found to be no greater than providing supportive care only, without IVIG or PE.

Relapse occurs in perhaps 10% of GBS patients after IVIG or PE. A second course of treatment may then be initiated although this has not been proven to be effective^{3,19} (Forcas, Rudnicki). Monitor the patient after the initial course of therapy to watch for an occasional relapse and even respiratory collapse.

Appendix*

A. A checklist to assist monitoring patients with suspected or diagnosed GBS

- (1) ___ Vital signs (including, as warranted, pulse oximetry, maximum inspiratory effort and vital capacity); see p. 8
- (2) ___ Strength of cough, speech, lack of drooling (unless intubated); see p. 9
- (3) ___ Disease-specific therapy (i.e., high dose intravenous immune globulins or plasma exchange); see pp. 22-23 and consult appropriate dept. and/or specialist to provide this
- (4) ___ Body positioning/re-positioning (especially off-loading at bony prominences); see pp. 20-21
- (5) ___ laboratory studies, including, but not limited to, serum electrolytes, albumin, pre-albumin); see pp. 5, 14-15
- (6) ___ Caloric intake and hydration; see pp. 14-15
- (7) ___ Sphincter status; see p. 14
- (8) ___ Pain control; p. 17
- (9) ___ Prophylaxis against peptic ulcer disease, DVT; see pp. 10, 13
- (10) ___ Involvement of relevant disciplines, as warranted (e.g., pulmonary physicians, OT/PT, respiratory therapy, speech therapy, psychiatry/psychology)
- (11) ___ Psychosocial issues; see, specifically p. 7

* This appendix should be used as an adjunct rather than a substitute for the complete text. For each numbered item please see the referenced page(s) for a more complete description and explanation of suggested care.

B. Diagnostic Criteria for Guillain-Barré Syndrome¹ (Asbury)

1. Rapid onset over a few days to 1-4 weeks of symmetrical weakness with loss of deep tendon reflexes in the extremities
2. Altered sensations, numbness, tingling or pain in affected limbs
3. Elevated spinal fluid protein, usually within 1-4 weeks of symptom onset, with normal cell count
4. Nerve conduction velocity-electromyography (NCV-EMG) evidence of nerve conduction slowing or blockage
5. Absence of other causes of a peripheral neuropathy
 - a. history of organic solvent inhalation, lead intake or intake of certain drugs, such as nitrofurantoin or dapsone
 - b. evidence of infectious causes of neuropathies, such as Lyme disease, HIV, diphtheria, and, in unvaccinated populations, poliomyelitis
 - c. findings of acute intermittent porphyria as evidenced by positive urine studies for porphyrin metabolites
6. Findings supportive of the diagnosis of Guillain-Barré syndrome
 - a. Illness occurs as a single event, i.e., without recurrence, with return of strength starting at about 2 to 8 weeks
 - b. Associated changes in blood pressure such as mild hypertension and rapid heart beat
 - c. A preceding infection, such as an upper respiratory infection or diarrhea, at 1 to 6 weeks before neurological symptom onset

Appendix

C. Prognosis

The Erasmus GBS Outcome Score (EGOS)²² (van Koning) helps predict the likelihood of recovery at 6 months*. It is based on three clinical characteristics, age, diarrhea preceding the GBS, and strength at 2 weeks into the disorder. It offers a guide to counsel patients about their prognosis.

Erasmus GBS Prognosis Score			
Prognostic Factor	Patient Characteristic	Score Points	The patient's points
1. Age (years)	< 40 y/a	0	—
	> 40 y/a	1	
2. Diarrhea within 4 weeks before GBS symptoms	No diarrhea prior to GBS	0	—
	Diarrhea preceded GBS	1	
3. Degree of disability at 2 weeks into illness	1. minor symptoms; able to run	1	—
	2. can walk unassisted but cannot run	2	
	3. needs cane or walker to walk	3	
	4. bed or chair bound	4	
	5. on ventilator, even briefly	5	
Meaning of Total Score		Total Score (1-7) =	—
1-3: very good chance of walking at 6 months 7: poorer recovery			

* Independent validation of this score system has not been published.

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