Guideline for Neuromuscular Rehabilitation in Guillain-Barré Syndrome: What can we do?

Guia de Reabilitação na Síndrome de Guillain-Barré: O que podemos fazer?

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ABSTRACT

Introduction. Originally described by Landry in 1859 and Guillian, Barré and Strohl in 1916, Guillain-Barré Syndrome (GBS) is the most common cause of acute neuromuscular paralysis self-limited in developed countries. The annual incidence of GBS is 1.5 per 100,000. Although it had a favorable prognosis (“maladie bénigne et spontanément curable”) it’s mortality rate is about 5% and 10% of patients remaining severely disabled one year after neurological onset. Specialist teams, intensive care and rehabilitation are essential for patient management and should be provided in appropriate hospital units. Objective. To guide the health professionals about the utilization of the physical and respiratory techniques in patients with GBS, as well as it’s indication. Method. Update of articles about GBS and Rehabilitation, on the last years of the data bases Bireme, ScienceDirect, PubMed and SciELO. Conclusion. There is currently no consensus on the management of these patients in the acute, sub-acute and chronic settings. Many of them are being discharged without access to rehabilitation services. Our results suggest that the rehabilitation makes a measurable and significant difference and should be available to all patients with GBS.

Keywords. Guillain-Barré Syndrome, Rehabilitation, Physical Therapy.

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RESUMO

Introdução. Originalmente descrita por Landry em 1859 e Guillian, Barre e Strohl em 1916, a Síndrome de Guillain-Barré (SGB) é a causa mais comum de paralisa neuromuscular aguda autolimitada em países desenvolvidos. A incidência anual varia de 1,5 casos|100.000. Embora possua prognóstico favorável, sua taxa de mortalidade encontra-se entre 5% e 10% dos pacientes que permanecem gravemente incapacitados até 1 ano após os primeiros sintomas. Profissionais especializados, tratamento intensivo e reabilitativo são essenciais para o gerenciamento dessa clientela. Objetivo. Nortear profissionais da saúde sobre a utilização de técnicas fisioterapêuticas (motora e respiratória) em pacientes com SGB, assim como o momento correto de sua utilização. Método. Atualização sobre artigos envolvendo SGB e reabilitação, nos últimos anos nas bases de dados Bireme, ScienceDirect, PubMed e Scielo. Conclusão. Não existe na atualidade um consenso sobre o gerenciamento de pacientes com SGB nos estágios agudo, sub-agudo e crônico. Infelizmente muitos pacientes recebem alta hospitalar sem informação e acesso aos serviços de reabilitação. Nossos resultados sugerem que a reabilitação (motora e respiratória) promove diferenças significativas na restauração funcional desses pacientes.


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INTRODUCTION

The Guillain-Barré Syndrome (GBS) is the most common acute demyelization neuropathy self-limited in western countries and it is preceded in most - but not in all cases - by uncommon infectious disease complications due to bacterial or viral illness, such as Campylobacter jejuni or Cytomegalovirus\(^1\). The pathogenesis of GBS is recently associating characterized humoral and cellular immune dysfunctions\(^2\). One third of the patients require admission to the intensive care unit (ICU), associated with significant risk of morbidity, mortality, and incomplete recovery\(^3\).

It primarily affects the peripheral nervous system, although recent research has shown that in a few of the patients, the central nervous system is also involved. The acute phase often requires intensive care services. There is a growing recognition that the recovery is not as smooth and free of symptoms as previously thought. Following the recovery some people endure long-term residual symptoms, such as fatigue, weakness and pain\(^4\).

The care of a patient with GBS is challenging for the healthcare team. Due to the fact that the disease normally starts with a motor weakness and ends with the patient requiring months or even years of physical rehabilitation, the patient should be prepared for this extended period of treatment. It is only by collaboration of the entire healthcare team that such tasks as diagnosis, treatment, therapy and pharmaceutical interventions are performed in such a way to help the patient regain a previous autonomy level\(^5\).

A greater understanding of the nature and course of the disease and its ramifications can lead to more effective physiotherapeutic management and a faster rehabilitation process. Therapy should not over fatigue the motor unit, which has been associated with paradoxical weakening\(^6\).

Clinical aspects of GBS

GBS is one of the best examples of a post-infectious immune-mediated disease and offers insights to the mechanism of tissue damage in other more common autoimmune diseases. Controlled epidemiological studies have linked it to bacterial infection with Campylobacter jejuni and viruses including Cytomegalovirus and Epstein Barr virus. There are several variants presentation that will share some patterns such as monophasic time course, recovery, a probable similar immune pathogenesis and prognosis. The clinical spectrum is composed by classical SGB (acute demyelization pattern – AIDP), Miller-Fisher Syndrome, acute motor axonal neuropathy (AMAN), acute motor-sensory axonal neuropathy (AMSAN), pure sensory variant, acute pandysautonomies and pharyngeal-cervical-brachial variant. The syndrome includes several pathological subtypes, of which the most common is a multimodal demyelization disorder of the peripheral nerves in close association with macrophages. Evidence from histological examination of peripheral nerve biopsy and postmortem samples suggests that both cell mediated and humoral mechanisms are involved in the pathogenesis. Immunological studies suggest that at least one third of the patients have antibodies against nerve gangliosides which - in some cases -, also react with constituents of the liposaccharide/or oligosaccharide of Campylobacter jejuni\(^7\). The treatment with both intravenous immunoglobulin and plasma exchange reduces the time taken for the recovery to occur, although mortality remains around 8%, with about 20% of the patients remaining disabled\(^8\).

Disability and health-related quality of life in GBS

A prospective study reported the disability and health-related quality of life level in 42 patients (mean age 52 years) with GBS during the first two years after the acute onset. Evaluations were performed, primarily as home visits, at two weeks, two months, six months, one year and two years after the first symptoms. The disability level was measured using some functional instruments: the Katz Personal and Extended Activities of Daily Living Indexes, the Barthel Index, the Frenchay Activity Index and assessments of work capacity; health-related quality of life using the Sickness Impact Profile. After two years of the onset of the GBS, 12% of the patients were dependent of daily life activities and 26% of instrumental ADL. At two weeks, all of the patients that were working before the onset were unable to work owing GBS and at two years, 17% were unable to work. At two weeks, scores on Sickness Impact Profile were elevated in all dimensions; at two years, they remained elevated in the physical dimension and in the categories of home management, work and recreation, and pastimes. These results attempt to show that the impact of GBS on ADL, work, social activities and health-related quality is considerable after two years of the onset and presumably persists beyond this time point\(^9\).

Depending on the degree of the residual disability encountered, consequences can range from being relatively minor to being completely devastating. The former cannot be belittled as they might necessitate a change in profession, for example, although this is
seen in a different league when compared with a person who is left effectively quadriplegic, requiring a very high degree of personal care\textsuperscript{9}.

**Residual fatigue in GBS**

In spite of showing an apparently “good” neurological recovery after GBS in approximately three-quarters of all patients, it was noticed that the majority of the patients remained restricted in daily and social activities due to fatigue. Fatigue seriously affects quality of life, and most patients report a fatigue among their three most disabling symptoms\textsuperscript{9}.

A prospective study aiming to increased knowledge on the spectrum of the GBS and to increase insight the occurrence and path physiology of residual fatigue. All patients (139 GBS and 14 Miller-Fisher Syndrome) completed the Fatigue Severity Scale (FSS). Severe fatigue was found in 60\% of the patients and significantly more frequently in female (74\% females vs 45\% males: \textit{p}=0.003) and patients of 50 years of age or older (70\% vs 49\%: \textit{p}=0.03). No relation was found between the fatigue level, clinically antecedent infections and infection serology\textsuperscript{10}.

Unfortunately the fatigue cause is at present not known. It might be explained as a post-traumatic stress disorder after a potentially life-threatening disease. On the other hand, recent long-term follow-up studies showed that residual neuropathy is present in approximately half of all GBS patients, even when these patients showed apparently “good” neurological recovery, which might suggest possible involvement of “peripheral” disturbances in the path physiology of fatigue\textsuperscript{11,12}.

Other study determined the relative contribution of peripheral and central factors during a 2-min fatiguing sustained maximal voluntary contraction (MVC) in 10 neurologically well recovered GBS patients and 12 aged and sex-matched healthy controls. The protocol was based on the twitch interpolation technique. The results hypothesized that central fatigue in GBS patients is related to the decreased number of remaining motor units. The normal interplay between numbers of recruited motor units and firing frequency is disturbed. It could be that a limited number of motor axons in GBS induces the relatively fast increased in central activation failure during the first minute, as the dropout, where only a few motor units could induce a relatively large failure of central activation\textsuperscript{13}.

Management of an excessive fatigue can include use of energy conservation techniques, lifestyle changes, pacing, regular periods or naps during the day, and improvement of sleep. Treatment includes occupational therapy (OT), physical therapy (PT), and supervised desensitization therapy to enable patients to tolerate practicing their daily living tasks (eg. grooming, dressing)\textsuperscript{14}.

The OT can advise specific energy conservation strategies to manage fatigue and facilitate patient functional independence and provide adaptive equipment (eg. grabbers, sock donors, plate guards) for the patient to facilitate personal care. This continues over time to incorporate domestic (eg. making tea) and community tasks (eg. banking, crossing roads). Home modifications are also undertaken to make the environment safer and accessible (eg. nonskid mats, proper lighting, grab rails). In severe cases, electric wheelchairs may be provided for community mobility\textsuperscript{14,15}. Other longer term issues such as return to driving and work are also coordinated by the OT and PT professionals. Effective communication between other professionals and a rehabilitation team is imperative for improved functional outcomes and successful social reintegration\textsuperscript{15}.

**Possible predictive factors on GBS prognosis**

The outcome of GBS patients in relation to some clinical and laboratory prognostic features during the acute phase of the disease is summarized in (Table 1). Older age (>50 years), longer duration of plateau phase (>10days), longer latency between onset and maximum impairment (>10days), presence of respiratory impairment and, in particular, the need for assisted ventilation and absence of cranial nerve involvement tended to be associated with a worse functional prognosis as assessed by Hughes scale\textsuperscript{14,16}. However, low mean Compound Muscle Action Potential Amplitude (CMAP) is the most powerful predictor of poor recovery rate, which indicates a reduced number of functioning motor units\textsuperscript{17}.

**Table 1. Features associated with poorer outcome in GBS.**

<table>
<thead>
<tr>
<th>Features associated with poorer outcome in GBS\textsuperscript{14,16}</th>
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<tbody>
<tr>
<td><strong>Older age</strong></td>
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<tr>
<td>• Requirement for respiratory support</td>
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<tr>
<td>• Abnormal peripheral nerve function</td>
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<tr>
<td>• No plasmapheresis or Intravenous Immune Globulin performed</td>
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<tr>
<td>• Subgroup of GBS with primary axonal degeneration</td>
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<tr>
<td>• Patients with rapid onset</td>
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<tr>
<td>• Progression to quadriplegia</td>
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<tr>
<td>• Respiratory dependence</td>
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<tr>
<td>• Severe disease at presentation</td>
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<tr>
<td>• Campylobacter jejuni infection</td>
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<td>• Patients showing no improvement at 3 weeks of plateau of disease</td>
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</table>
Guideline of Physical Therapy Techniques in GBS

Table 2. Physical Therapy Techniques in GBS.

<table>
<thead>
<tr>
<th>Guideline of Physical Therapy Intervention \cite{17-21}</th>
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<tbody>
<tr>
<td><strong>Therapeutic exercises</strong></td>
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<tr>
<td>- Progressive resisted exercises should be performed with care to avoid overexertion\cite{17}.</td>
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<tr>
<td><strong>Functional training:</strong></td>
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<tr>
<td>- Safe transfer skills; balance and equilibrium in all positions and progressive ambulation; some patients may require progression on a tilt table to improve tolerance and decrease sensitivity to weight bearing.</td>
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<td>- Neurodevelopmental sequencing is effective with progressing the geriatric patient with GBS\cite{17,22}.</td>
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<td>- Use of partial body weight support systems can be beneficial during the rehabilitation of GBS patients as they can objectively progress the patient during gait\cite{17}.</td>
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<td>- Proprioceptive training using devices such as the podiatric (a motorized variable pitch wobble board with handrails) is helpful with functional ability, especially due to residual muscle shortening and loss of proprioception of the lower extremities\cite{20}.</td>
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<tr>
<td>- Manual Therapy: muscle shortening and contractures could be avoided with passive range of motion exercise and positioning is critical to achieve goal of avoiding shortening and contractures; positioning, turning programs, and mobilization are critical to offset the risks of deep vein thrombosis and decubiti.</td>
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<tr>
<td>- Prescription, application of devices and equipment: orthotics are beneficial to prevent contractures and minimize the effect of immobilization; bedding to prevent pressure sores should be considered; compression stockings can be used since there are some patients at risk of deep vein thrombosis due to bed rest; ankle foot orthosis (AFO) is sometimes used when beginning a gait program due to residual lower extremity weakness\cite{21}.</td>
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<td>- Airway clearance techniques: mechanical respiration may be required in some patients with proper techniques for maintaining hygiene and providing incentive spirometry to encourage recovery\cite{20}.</td>
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<td>- Integumentary repair and protection techniques: some patients with GBS depend on a catheter for a portion of their treatment duration\cite{20}.</td>
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<td>- Electrotherapeutic modalities: transcutaneous nerve stimulation (TENS) has been used effectively for pain control in patients with GBS\cite{20}.</td>
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<td>- Physical agents and mechanical modalities: hydrotherapy can be a valuable part of the treatment program with the GBS patient as it encourages mobility and muscle strengthening\cite{20}.</td>
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<td>- The Proprioceptive Neuromuscular Facilitation (PNF) concept provides tools for the therapist to help the patients gain efficient motor function and increased motor control\cite{21}.</td>
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Specific Complications and Rehabilitation Management

Table 3. Specific Complications and Rehabilitation Possibilities.

<table>
<thead>
<tr>
<th>Specific Complications On Gbs</th>
<th>Rehabilitation Techniques</th>
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<tr>
<td>Weakness and Fatigue</td>
<td>Therapeutic Exercises</td>
</tr>
<tr>
<td>Sensory Loss</td>
<td>Examination and Cleaning() Soaking Techniques</td>
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<tr>
<td>Autonomic Dysfunction and Pain</td>
<td>Compression Stockings and TENS</td>
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<tr>
<td>Neurogenic Bladder and Bowel</td>
<td>Biofeedback, Learn techniques, Orientation</td>
</tr>
<tr>
<td>Immobility</td>
<td>Position Changes, Splints and Exercises</td>
</tr>
<tr>
<td>Physical Deconditioning</td>
<td>Specific Aerobic Exercises</td>
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<tr>
<td>Respiratory Dysfunction</td>
<td>Therapeutic Techniques and Positioning.</td>
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Weakness And Fatigue Control

Weakness is often a predominant finding in GBS. The extent and severity of the weakness may range from total paralysis requiring mechanical ventilation to moderate/slight paresis of foot dorsiflexors or intrinsic hand muscles. Depending on the degree of weakness, exercises can be passive, active-assisted, active or resistive. Severely weakened or paralyzed muscles may require splinting and proper rest positioning to maintain the joint in a mobile and functional position until recovery occurs. During recovery, or when functional recovery is incomplete, functional orthoses may allow the patient with GBS to resume activities.\cite{22-24}. Aerobic training decreases fatigue and improves physical fitness and quality of life in GBS patients. In some cases, training may not be tolerated because of the initial complaints of severe fatigue or residual neurological deficits.\cite{23}.

A study determined the feasibility and effects on the fatigue severity of a structured 12-week bicycle exercise training program. In addition there should occur an evaluation of the effects on the physical fitness, muscle strength, functional outcome, anxiety and depression, handicap, and quality of life. The authors performed a 12-week study of bicycle exercise training in 20 patients with severe fatigue, 16 with relatively good recovery and 4 with stable chronic inflammatory demyelination polyneuropathy (CIDP). Training seemed to be well tolerated, and self-reported fatigue scores decreased 20%. Physical fitness, functional outcome and quality of life were improved.\cite{23}. 
Sensory loss

Decreased ability to perceive pain and touch, compromise the patients’ ability of protecting the involved extremity from injury. Injury may lead to difficult-to-heal skin ulcerations and infections. In GBS, the feet and hands are most commonly affected. When deep pain sensation is lost and kinesthetic and joint perception sense is impaired, neuroarthroplathic joints may develop from repetitive trauma. GBS patients with loss of kinesthetic and joint position sense, for example, cannot judge accurately the placement of their foot and the position of the foot and ankle, not only creating an ataxic gait but increasing the risk of falls.

The basics of treatment begin with education of the patient and a preventive program consisting of a careful daily skin examination and gentle cleaning and soaking techniques, followed by application of a lubricant to prevent skin fissuring. Removing pressure from red areas and use of appropriate custom shoes or shoe inserts also play a role. Special care should be taken in those wearing orthosis, with several inspections per day as well as periods of time when the orthosis is removed.

In patients with sensory ataxia, the sensory feedback is disturbed, which makes the development of new sensory engrams more difficult. Training consists of frequently repeating movements or certain tasks. For instance, the blind-folded patient receives different sensory stimuli. Afterwards, the GBS patient is allowed to look at the stimuli in order to correlate this with visual cues. When gait ataxia is present, balance exercises can be prescribed, gait aids may be necessary or patients may even become wheelchair-bound. GBS patients may benefit from shoes with a broadened heel and good stabilization around the ankle joint. Ataxia of the upper limbs may cause severe problems in performing daily and instrumental activities. Mechanical devices have been developed to aid the patients and a visual feedback control should be encouraged in the therapy.

Autonomic dysfunction and pain

An autonomic dysfunction can be seen in neuropathies affecting small fibers, diabetic neuropathy, and polyradiculoneuropathy (GBS), and is postulated to play a role in neurogenic pain and in regional pain syndromes. The symptoms include decreased sweating, orthostatic hypotension, gastro paresis, constipation, vomiting, diarrhea, impotence, and a flaccid urinary bladder.

Patients with orthostatic hypotension can be advised to arise more slowly and may also benefit from compression stockings. Gastric motility disturbances may be improved with diet and a proper bowel regimen and/or the addition of motility-enhancing drugs. Patients in the initial stages of acute GBS require cardiac monitoring.

Transcutaneous Electrical Nerve Stimulation (TENS) is beneficial in the treatment of many types of pain syndrome, but patients who respond best are those with pain of neurogenic and/or musculoskeletal origin and a relatively minor psychological component. There is no current research to support the use of one type of TENS (various strengths/durations, burst, modulated, etc.) over another, and patients should be encouraged to adjust the impulse as well as to try various lead placements to obtain optimal relief. In most situations, physical therapists work with patients using TENS in specific pattern. Adverse reactions to TENS are infrequent, the most common being skin hypersensitivity. TENS is contraindicated in patients with the need for pacemakers and in those who are pregnant.

Neurogenic bladder and bowel

Bladder and bowel dysfunction occur in a variety of neurological diseases, resulting in incontinence, urinary retention and infections. Serious complications include upper urinary tract infections, renal stones, renal damage and skin breakdown. Incontinence and the need to learn techniques such as self-catheterization may result in embarrassment and loss of self-esteem, and are often thought of as burdensome. It should be emphasized that bladder and bowel incontinence are major factors in determining whether or not a patient can remain in a home setting.

Pelvic floor muscle training is the most commonly recommended physical therapy treatment for women with stress leakage of urine. It is also used in the treatment of women with mixed incontinence, and less commonly for urge incontinence. Adjuncts, such as biofeedback or electrical stimulation, are also commonly used with pelvic floor muscle training. The role of pelvic floor muscle training for women with urge incontinence alone remains unclear. Many of the trials were small with poor reporting of allocation concealment and masking of outcome assessors. In addition there was a lack of consistency in the choice and reporting of outcome measures that made data difficult to combine.
Physical deconditioning

Rehabilitation nurses working with patients with chronic-relapsing GBS may have questions regarding the usefulness of exercise in the maintenance of health for GBS patients. However, those questions must remain largely unanswered because of a dearth of research regarding GBS and the effects of exercise on it.\(^{30,32}\)

The effects of low-intensity aerobic exercise was determined on one subject with GBS.\(^{30}\) Walking and cycling were chosen as the exercise modes, with a 10-week walking phase followed by a 15-week cycling phase. Changes in the amount of exercise tolerated over the course of the two exercise phases were compared. Also, measurement of pulmonary function (with a Spiro meter), grip strength (with a dynamometer), total body weight (with specific scales) and skin folds (with calipers) were completed before and after the walking and cycling phases. The activity provided appeared to have produced a number of functionally significant results for the patient. These outcomes support the idea that low-level aerobic exercise can be safe, and suggest that clinically important benefits may occur from such activities as walking and cycling. Rehabilitation nurses may find these study results encouraging for their own GBS patients, because they indicate that exercises may improve the quality of life for these individuals. Others studies also reported positive effects of low-intensity training exercises in neurological diseases.\(^{31,32}\)

Ambulation aids

Indications for ambulatory aids include weakness of the trunk or lower extremities, poor balance, and decreased ability to bear weight as a result of disease or injury (e.g., an infection or ulcer). Assistive devices make walking safer by providing a wider base of support, by redistributing weight to the upper extremities, and by allowing for larger shifts in the center of gravity before balance is lost.\(^{30}\) Unfortunately, physicians frequently write prescriptions for assistive devices without a prior physical therapy evaluation. A selection of a particular device is determined by the patients’ strength, stability, coordination, cardiovascular capacity and cognitive status. Maximum stability and support are provided by a walker, followed by auxiliary crutches, forearm crutches, two canes, a quad cane, and a single-tipped cane. The PNF patterns are important to improve stability and strength during ambulation.\(^{21}\)

Wheelchair

A wheelchair may be needed in cases in which the GBS patient has limited cardiovascular capacity or cannot meet the additional energy cost of ambulating with a cane and a brace. There are currently 8,000-10,000 production models of wheelchairs available on the market. The physician prescribing wheelchairs should be familiar with standard models and options, and should recognize when evaluation by a specialist for a custom chair is needed. For GBS patients who use wheelchairs, the seating system should maximize independence and minimize pressure over vulnerable areas while providing support for proper posture.\(^{33}\)

Respiratory function

Neuromuscular diseases that affect the respiratory system are a major cause of morbidity and mortality in both acute and long-term settings. In recent years, important advances have occurred in the understanding and management of the respiratory and critical care complications of neuromuscular diseases.\(^{34}\)

GBS is the most common neuromuscular disorder that requires admission in the Intensive Care Units (ICU). The indications for admission of patients with GBS in the ICU often include one or more factors: respiratory failure, severe bulbar weakness, autonomic instability and threatening pulmonary aspiration. Respiratory failure is a relentless problem among patients with the syndrome, and mechanical ventilation is required in 20 to 30% of the cases. The percentage of patients that requires mechanical ventilation varies across populations, but these variation maybe explained by the covered spectrum of the disease. Some papers used the entire spectrum of GBS, with some patients with mild patterns that usually do not require admission in the ICU and patients with extreme severe pattern. In GBS mortality is 12 to 20 percent and the median duration of mechanical ventilation is 18 to 29 days among intubated patients.\(^{35}\)

A study described the conditions for development of respiratory failure in patients with GBS. The author indicates four aspects: upper-airway compromise due to weakness of oropharyngeal and laryngeal muscles. These aspects could place the patient at risk for aspiration, because swallow and secretion clearance may be impaired. The second aspect is the weakness of the muscles from inspiration (diaphragm, intercostals and accessory muscles) resulting in restrictive disorder (low compliance), with microatelectasis, that lead to ventilation/mismatch and, consequently, hypoxemia.
A third aspect is the weakness of the muscles of expiration, which can reduce cough ability, increasing the risk of respiratory infections. Fourth, pulmonary complications (embolism and pneumonia) associated with the other conditions. To verify and determine the predictors for ventilation support and endotracheal intubation the author suggests an objective assessment of vital capacity (VC), maximum inspiratory pressure (MIP) and expiratory pressure (MEP) associated with the subjective assessment of the patients signs of respiratory muscle fatigue. The early identifications of patients at risk of respiratory failure are related to the “role” 20/30/40 (VC of less than 20 mL/Kg; MIP less than 30 cm H₂O; MEP less than 40 cm H₂O). Patients with GBS are poor candidates for Non-invasive Ventilation (NIV), because the upper airway protective mechanisms and inadequate cough are often impaired in the severe conditions of the disease. However, the patients without the conditions mentioned should benefit from NIV, if they need short-term ventilatory support. Observational studies in small numbers of patients suggest that Nasal intermittent positive pressure ventilation (NIPPV) may decrease the need for invasive mechanical ventilation, shortens ICU length of stay and decreases or reduces mortality. The role of tracheostomy in GBS is controversial. There is no optimal timing for performing the procedure. Another study proposed a post-intubation pulmonary function test to determine the need of tracheostomy. The score was calculated adding VC (ml/kg), MIP (cmH₂O) and MEP (cmH₂O). Immediately prior to intubation the caregivers calculated the score and in the twelfth day of mechanical ventilation they calculated the score again. If the result of the division of the two measured scores was less than one it is suggested that patient should need more than three weeks of mechanical ventilation and, in this case, tracheostomy should be indicated.

**Rehabilitation plan**

A rehabilitation plan is established based on accurate evaluation of function and considers de course of the GBS. After the evaluation, the data is summarized and a list of problems and a plan of rehabilitation are built. The rehabilitation plan considers in particular the disabilities and handicaps that result from the disease. Physical impairments (weakness, contractures, pain) are treated to improve function and to prevent progression. Furthermore, remaining capabilities are trained to ameliorate their function. When necessary, functional autonomy can be compensated by aids and environmental adaptations. Depending on the goals and objectives of the rehabilitation plan, patients have to call on the help of the different members of the rehabilitation team.

**Drugs in GBS**

A recent practice parameter recommended either intravenous immunoglobulin or plasma exchange, but not corticosteroids, as appropriate treatments for adults and probably children with severe GBS within 2 weeks from onset. Most patients in the trials of these treatments have had the demyelization form of the disease, and the benefits of treatment in uncommon subgroups, such as those with axonal disease, cannot be distinguished. Despite immunotherapy, 4% to 15% of the patients with GBS die from this syndrome and nearly 20% have a persistent disability. Death from GBS occurs mostly in mechanically ventilated patients. Supportive care remains the mainstay of treatment, but the evidence for the methods of supportive care is inadequate and consensus guidelines for treatment have not been published.

**DISCUSSION**

Patients and their relatives and friends will have been given a good idea of what to expect in the recovery phase of the illness. In most cases, these expectations are borne out and a steady improvement will be noticed. But sometimes improvement is just too slow and it becomes obvious that recovery will not happen within a few months. There are some rough pointers as to what may happen. Elderly patients with an explosive onset in the acute stage and those who need ventilation, especially for an extended period, tend to do less well. But these are only indicators. Some very old patients do very well. Some patients ventilated for long periods do recover completely or almost completely. Some patients who are not ventilated are left with significant disability. If the recovery remains slow doctors may order some electrical tests to ascertain axonal damage, if they have not already done so. If the phrase ‘axonal damage’ is mentioned, then it would certainly be worthy asking for an interview with the consultant neurologist to discuss the possible implications.

The prognosis varies enormously. GBS can prove fatal although this is usually because of breathing or heart complications or another coexisting condition. Some become so severely paralyzed that the ability to breathe never returns; others require ventilation for many months, even a year or more, before they can be weaned off the machines. These patients are often confined to wheelchairs and only have the rudimen-
tary use of their arms and hands. Such very severe cases are fortunately often rare.15,16

Although most patients with GBS need rehabilitation, there are no long-term rehabilitation outcome studies or comparisons of different methods. In neuromuscular diseases, “overuse” may impede recovery and cause paradoxical weakening. Attention needs to be paid on many details that cannot be summarized briefly. In GBS there is a danger of muscle shortening and joint contractures. Prolonged immobilization leads to a reduction of blood volume and increased episodes of postural hypotension. For some immobilized patients, a tilt table has been useful. Weight loss and significant sensory loss make patients susceptible to peripheral nerve compression and the development of decubitus ulcers, requiring proper bed positioning with frequent postural changes. In patients noted to have immobilization hypercalcemia, early mobilization was correlated with a therapeutic drop in the serum calcium levels. In the acute stage, patients lose weight and during recovery, they regain weight due to reduced activity levels. Treatment in the acute phase should include an individual PNF program of gentle strengthening involving isometric, isotonic, isokinetic and manual resistive and progressive resistive exercises. Rehabilitation should be focused on proper limb positioning, posture, orthotics and nutrition.6,9,15,21,45-47

CONCLUSION

Disabled patients should be treated by an interdisciplinary health team and should receive a specific assistance plan. General practitioners are primary caregivers for GBS patients and their families in the community. The prognosis from GBS is good, but recovery is prolonged. The rehabilitation team will assist the patient by minimizing disability, improving functional outcomes and his/her quality of life.

REFERENCES