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GUILLAIN-BARRÉ SYNDROME,
CIDP AND VARIANTS

Guidelines for Physical and Occupational Therapy



A PUBLICATION OF THE GBS/CIDP
FOUNDATION INTERNATIONAL

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This pamphlet is provided as a service of the GBS/CIDP Foundation International.

To our readers

DEAR THERAPIST/THERAPIST ASSISTANT,

Thank you for taking a few moments to learn more about what your patient with Guillain-Barré syndrome (GBS), chronic inflammatory demyelinating neuropathy (CIDP) or a variant has been experiencing and how you better can help them on their road to recovery. Perhaps they already are well along on their journey, or they just may be getting started, but their experience to this point undoubtedly has been a **frightening** and **stressful** one. Some of a patient's greatest fears regarding therapy are that their therapist will not know what to do with them and/or will not understand their limitations, or that therapy will be so painful and/or hard that they will not be able to move the next day.

We know that therapists and therapist assistants have a solid base of academic and clinical knowledge from which to work, but many have not encountered GBS or CIDP directly during their careers. Accordingly, the GBS/CIDP Foundation International (the "Foundation") has published this booklet and encourages patients to share it with their physical and occupational therapists. Regardless of your familiarity with GBS or CIDP, your taking the time to review this information will offer your patient peace of mind in knowing that you are interested in their care and sensitive to issues that are unique to their condition.

What are GBS, CIDP and related disorders?

GBS and CIDP are acquired immune-mediated inflammatory disorders of the peripheral nervous system. Their etiology is not completely understood, and neither disorder is contagious.

Most GBS cases appear to be precipitated by an infectious respiratory or gastrointestinal illness with diarrhea. In the U.S. and Europe, 60 to 80 percent of GBS cases occur within four weeks of a preceding infection. For an as yet unknown reason, the body's response to the infection goes awry and the immune system attacks the myelin and sometimes the axons of the peripheral nerves. As a consequence, neurological signals are slowed, altered or blocked altogether, resulting in paresthesias (e.g. numbness, tingling, "crawling skin"), loss of sensation and deep tendon reflexes, progressive muscle weakness, often general fatigue, sometimes pain and a number of other possible secondary complications. Motor and sensory involvement is symmetrical in proximal and distal muscles with symptoms progressing in an ascending fashion, the lower limbs usually being affected first. In severe cases, autonomic nervous system dysfunction also can occur, the presence of which is suggested by orthostatic dizziness, bowel and bladder function complications and/or cardiac symptoms.

Unlike multiple sclerosis and amyotrophic lateral sclerosis (a.k.a. Lou Gehrig's disease), GBS and CIDP generally do not cause damage to the central nervous system, although GBS patients may experience difficulty with swallowing (dysphagia), facial drooping and other deficits of the lower cranial nerves. Neither condition affects a patient's cognition.

GUILLAIN-BARRÉ SYNDROME

Guillain-Barré syndrome, also called acute inflammatory demyelinating polyneuropathy (AIDP), affects one to two new persons per 100,000 population each year. It can strike anyone without warning regardless of gender, age or ethnic background. About 50 percent of patients initially develop abnormal sensations such as tingling of the feet or fingers; twenty-five percent initially develop muscle weakness (e.g. difficulty climbing stairs, getting up from a chair and/or cramping) and 25 percent begin with a combination of abnormal sensations and weakness. Pain is also a common symptom, sometimes experienced as deep aching or cramping in the buttocks, thighs or between the shoulders.

Disability caused by GBS generally progresses over the course of a few days to four weeks, with weakness starting distally and ascending in a matter of hours to days. At the peak of the condition's progress, many patients experience flaccid paralysis of nearly all skeletal muscles, with talking, swallowing and breathing frequently being affected. Seventy percent of patients lose some strength in respiratory muscles that can lead to shortness of breath; in about one third of patients, intubation and a ventilator temporarily become required. Consequently, most newly diagnosed patients are placed in an intensive care unit for monitoring. Plasma exchange (PE) or a high dose of intravenous immune globulins (IVIg) often hasten recovery. Corticosteroids are not helpful for GBS, but frequently are used to treat CIDP.

Fortunately, GBS typically is self-limiting with improvement usually beginning spontaneously after weakness maximizes. The death rate is approximately three percent, and the recurrence rate less than five percent. Most patients eventually reach a full or nearly full recovery. Many patients will walk without aid after three months and experience only minor residual symptoms by the end of the first year following onset. Nevertheless, recovery can be extremely slow (stretching over the course of six months to two years or longer) and five to twenty percent of patients are left with significant residual symptoms that lead to long-term disability and prevent a successful return to their prior lifestyle or occupation.

ACUTE MOTOR AXONAL NEUROPATHY

This variant of GBS initially was recognized by westerners as epidemics of paralysis in children in rural northern China and first was named the Chinese paralytic syndrome. It often follows diarrhea from *Campylobacter jejuni*, does occur occasionally in the western world, but unlike classic GBS does not affect sensory nerves, and more often progresses sufficiently to require ventilator support.

Medical management and occupational and physical therapy methods (see below) are the same as for GBS.

MILLER FISHER SYNDROME

Miller Fisher syndrome (MFS), named after C. Miller Fisher, MD, who described the disorder, is an uncommon variant of GBS. It consists of the triad of areflexia, external ophthalmoplegia, that is, weak eye muscles that cause diplopia, and ataxia. Both the double vision and ataxic gait can contribute to impaired activities of daily living. Clinical features of MFS often accompany GBS. Principles of care for GBS also are applicable to MFS.

CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY

CIDP is a chronic counterpart to Guillain-Barré syndrome, and also is characterized by symmetrical weakness and sensory changes.

New cases of the condition are rare compared to GBS – 1.5 to 3.6 new patients per 1,000,000 population each year – but because CIDP can persist for years, there are estimated to be as many as eight cases per 100,000 population, or five to nine thousand people in the U.S. at any one time. In contrast to GBS, CIDP develops slowly, often over the course of two months or longer. It can manifest in a variety of patterns; however, the most common is a series of recurrent relapses and remissions of ascending weakness over the course of years. Occasionally, the disorder may run a slowly progressive deteriorating course without improvement.

Because of its typically slowly progressive nature, CIDP can present symptoms for months or longer before activities of daily living are impaired and the disorder is diagnosed. Once

it is determined that a patient has CIDP, first line treatment choices are corticosteroids, plasmapheresis (plasma exchange) or intravenous immunoglobulin (IVIG). In contrast to GBS, breathing, swallowing and speaking are rarely affected, though it is still important to establish a diagnosis and set a course of treatment as soon as possible. If left untreated, or if treatment is significantly delayed, CIDP can lead to severe nerve damage that may not be entirely recoverable. In fact, reinnervation and long-term disability rates are worse for patients with CIDP than for those with GBS, with the likelihood of progression and recurrence being much higher.

CAREGIVERS AND COMPASSION FATIGUE

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MULTIFOCAL MOTOR NEUROPATHY

Multifocal motor neuropathy (MMN) is a rare chronic inflammatory neuropathy characterized by episodes of right and/or left-sided, i.e., asymmetric, distal limb weakness, of the upper more often than the lower limbs. Thus weakness may occur at the wrist, fingers and/or ankles and reflects inflammation of their motor nerves with slowed conduction. The disorder can extend over two to as long as 20 years or more. Nerve conduction tests with selective nerve mapping aids the diagnosis. Because of its slow and variable presentation, MMN may not be recognized readily. Over time, ongoing weakness may lead to muscle atrophy. Fasciculations may occur. Treatment with IVIG is usually beneficial but not PE or corticosteroids. Various immunosuppressive drugs have been tried with variable benefit.

The principles of OT and PT care for GBS and CIDP as described below apply to MMN. The Plan of Care should be customized to the individual patient's disability. Orthotics may be indicated to compensate for distal weakness.

WHAT IS YOUR PATIENT FEELING?

In order to maximize the impact of your Plan of Care on your patient's prognosis, it is important to understand that their emotions frequently will override reason. When you first meet, the patient likely will have thoughts such as: "Will I ever walk again?" and "When will I get back to normal, or will I?" Even when it is time to be discharged from services, they likely will be asking themselves, "Will this ever happen to me again?" ***The therapist always should be encouraging and hopeful regarding the patient's recovery without making promises about the degree of recovery or the time frame.*** It can, however, be said with confidence that the vast majority of patients return to some level of independence. The following is a list of concerns that should be recognized as you work with your patient:

FEAR AND ANXIETY

Your patient went from being completely independent to experiencing at least some level of dependence on others for no apparent reason and in a short amount of time. If they are still in the acute stage of the condition, they and their caregivers are looking desperately for answers and may be thinking the worst.

Once stabilized, your patient probably will continue to experience some anxiety during quiet times alone or during treatment. Because GBS and CIDP are rare, often

not diagnosed immediately, and the reason for their having developed the condition uncertain, patients and caregivers often wonder whether their condition may worsen.

PAIN

Pain can be significant with GBS, occurs less often with CIDP and contributes significantly to patient anxiety. As sensory nerves begin to heal and grow back, the sensitive regenerating axons impulsively generate abnormal signals that can be exacerbated by weight bearing and exercise. Abnormal sensations, which usually occur distally in the feet and hands, can be difficult for the patient to describe but often interfere with daily activity.

Many GBS survivors report increased sensitivity to light touch. It is not uncommon to hear comments such as, “I can feel my fingerprints,” or “The wrinkle in the bed sheet is excruciating.” CIDP patients can experience “lightning bolts” of pain shooting from their lower and upper extremities. *Always* ask a patient for permission before touching them, and think of how their Plan of Care may affect their sensory system.

DEPRESSION AND GUILT

Patients almost certainly wonder if they ever will be able to contribute physically or financially to their family, friends or community again. Medical bills quickly accumulate at the same time that income and benefits diminish.

Many patients also demonstrate at least a period of low motivation toward participating in their Plan of Care (especially home programs) as they realize that their recovery is not always directly related to their personal efforts. This understanding is discouraging and can lead to apathy and/or depression.

Your greatest tool is the active involvement of your patient in their Plan of Care. Unless they can get past their feelings of fear, anxiety and guilt, it will be very difficult to move forward. The best way to help your patient conquer these challenges is to let them know that they are not alone.

The GBS/CIDP Foundation has many support groups – groups for CIDP patients, children, pregnant patients with GBS, etc. – with whom individuals may share experiences and learn from each other. (Patients may contact the Foundation for information.)

EVALUATION

Because the clinical features of GBS or CIDP can differ dramatically from one person to the next, a thorough physical therapy (PT) and occupational therapy (OT) evaluation is imperative to understanding the patient's particular needs. The evaluation will vary somewhat based on the setting in which a patient is being seen, as well as their current status in the pathology process. Patients with GBS frequently begin care in the intensive care unit of a hospital, then progress to a sub-acute setting in a rehabilitation department or outside nursing/rehab facility and eventually to home-based or outpatient therapy. Patients with CIDP, on the other hand, usually begin with outpatient or home-based therapy, and only visit a hospital or rehabilitation department if they experience severe symptoms associated with a relapse or if long-term dependent care is required.

It is not uncommon for OT to address primarily the upper body and activities of daily living (e.g. dressing, grooming and feeding) while PT focuses on the lower body and mobility; however, this is certainly not universal and may depend on a given site's policies and the specific state's practice acts. Regardless of who ultimately is responsible for each aspect of an initial evaluation, therapists should be sure to include the following components in their assessment:

PATIENT/CAREGIVER INTERVIEW

The most important part of any evaluation is the patient interview. Only the patient can tell you how they are feeling, what they have experienced and where they want to be after therapy.

Be conscious and respectful of their emotions. The interview is also a good time to assess the caregiver and environmental support that the patient has at home and what their needs may be after discharge.

SENSORY ASSESSMENT

Ask the patient if they are sensitive to touch, and if so, where and what types of touch are aggravating or painful, **before** placing a finger on them. There is no quicker way to lose a patient's confidence and set a negative precedence for the evaluation and future treatment than to begin by causing pain. Of the most commonly used tests to evaluate sensation, touch and pressure and touch localization are the most important to assess during the initial evaluation (use of a filament test kit or other acceptable means as tolerated). Sensation should be re-assessed frequently to track progress of reinnervation, monitor muscle soreness and avoid causing undue pain during therapy.

SKIN INSPECTION

After asking the patient for consent and explaining what you will be doing, check their skin for lesions or pressure spots. The skin over bony prominences (e.g. heels, sacrum and hips) is particularly susceptible to break down. If a patient is independently mobile, sufficiently cognizant, and has already been performing skin inspections, their report alone may be sufficient.

JOINT RANGE OF MOTION

Pay particular attention to the ankles, knees and hips, especially if the patient has spent a lot of time sitting or confined to a wheelchair. Foot and wrist drop are not uncommon and may require bracing or splinting to prevent contractures. If joint contracture is suspected, measure passive range of motion with a goniometer to guide stretching exercises.

MUSCLE TESTING

Manual Muscle Testing (MMT) grades of zero to five commonly are used to assess muscle strength (as tolerated). If a patient is unable to move a defined body part independently against gravity (grade 3/5), gravity reduced or eliminated positions may be used. If a patient exhibits at least a grade 3/5, the therapist can apply manual resistance to determine higher grades. Consider assessing grip and pinch strength via a dynamometer or pinch gauge, respectively, or through functional testing.

FUNCTIONAL TESTING

Depending on the findings from the previous measures, mat mobility, transfers, self-care tasks (e.g. grooming, feeding and dressing) and other functional tasks related to the patient's work and/or leisure activities may be assessed during the initial evaluation.

MOBILITY

If a patient is functionally mobile, a brief gait and/or wheelchair assessment/observation may be performed; however, take care to assure that the patient is not already exhausted from the previous activities. If this is the case, the mobility assessment may be postponed until the beginning of the next session.

OTHER

In addition to the preceding assessments, and in accordance with the client's current status, the following areas also may be evaluated:

- **Respiration Test** with a hand-held-monitor or spirometer. Vital capacity and inspiratory force or effort are measures of diaphragm strength frequently used to gauge breathing status of the GBS patient.

- **Deep-Vein-Thrombosis (DVT)** – A firm, tender, warm or swollen calf raises suspicion of DVT. Use the patient’s history or a venous duplex ultrasound study to guide the differential diagnosis of DVT.
- **Autonomic Dysfunction** – If the patient reports dizziness, consider obtaining a blood pressure and heart rate while supine and then standing to identify orthostatic hypotension.
- **Endurance Patients** should not be tested to exhaustion since recovery from fatigue can take some time and will delay the rehabilitation process. Note when the patient begins to show early signs of fatigue during the assessment and adjust or discontinue the activity accordingly.

TREATMENT

The principal goals of therapy are to:

- Help the patient to achieve optimal muscle use at a tolerable pain level as nerve supply returns; and
- Use supportive equipment and other functional adaptations to help patients with residual impairments to resume an activity level that is as close to their previous lifestyle as possible.

Therapy does not facilitate nerve repair; however, it does help the recovering patient to learn optimal use of muscles as the nerves heal and innervation improves. Every person with GBS, CIDP or variants responds differently to the physical manifestations of his or her condition as well as pharmaceutical and therapeutic/ rehabilitation interventions. Consequently, it is essential to keep in mind that the body only will do what it is capable physically of doing, regardless of the expectations of the patient or therapy staff. Use a ‘safety first’ approach by teaching your patient to perform only activities he or she can do safely.

ACUTE STAGE

During the acute phase of GBS or an exacerbation of CIDP, a patient may not tolerate, or be unable to participate in active movement. Although the condition still may be worsening at this point and care may be more medical than rehabilitative, physical and occupational therapy still play an important role. Initially, your Plan of Care likely will be

more consultative in nature, with patient participation being mostly passive during direct treatment. Include the following in this consultation process:

- Provide patient and caregiver with education and training for the prevention of contractures, DVT and bedsores, as well as proper positioning and the expected course of future rehabilitation. To this end, advise the patient and their caregivers of the following:
 - Avoid prolonged hip and knee flexion;
 - Change position at least every two hours in bed and perform regular pressure reliefs when sitting; and
 - Support weak upper extremities with armrests, a wheelchair tray and/or pillows to prevent stretching of shoulder muscles and joint tissues.
- Educate yourself about the patient's home life, work demands, recreational interests and support system in order to customize your Plan of Care to their needs.
- Anticipate the need for assistive devices and other adaptive equipment/technology and be prepared to train the patient and caregivers in their use.
- Use gentle passive range-of-motion to reduce the risk of contractures and DVT.
- Introduce breathing and coughing exercises to maintain good airway exchange.
- Communicate clearly with the patient before and during any physical interaction.

RECOVERY

As the patient begins to recover sensation and motor control, exercises may be progressed from passive to active-assisted range-of-motion. Initially, active movement should be performed at low repetitions and resistance with frequent rest breaks. A powder board (a smooth surface designed to enable gravity reduced active sliding of a limb), slings and even hydrotherapy, with exercise performed in a pool or large tub (as appropriate), can facilitate the active movement of muscle groups that are not yet able to move independently against gravity. It is essential to help the patient move on their own as soon as possible, without exercising to fatigue, in order to help reduce the progression of disuse atrophy and other complications. The “No pain, no gain” philosophy should not be applied. Exercising to exhaustion will delay recovery without benefitting the patient.

Rather, approach your Plan of Care in this manner:

- Expand activities gradually.
- Increase repetitions before resistance in order to avoid injury to muscles, tendons and joints. *Use of proprioceptive neuromuscular facilitation (PNF) techniques may be helpful.*

- Teach energy conservation (e.g. pacing and breaking tasks into steps).
- Train caregivers in proper body mechanics for transfers, positioning, etc. to decrease the risk of injuries to themselves and the patient.

Once sufficient active movement has returned, therapy sessions should begin to focus on the patient's ability to perform daily activities. Whenever possible, practiced activities should be "real world." Working on cones and peg boards with an OT does not approximate the realistic demands of a patient who needs to bring a spoon to his or her mouth without spilling, nor does performing 10 repetitions of knee extension with a PT represent the patient's ability to get up from the chair in which they are sitting. Several possible functional activities include bed mobility, transfers, gait and/or wheelchair mobility, sitting and standing balance while reaching, dressing, feeding, bathing/toileting, writing/typing, leisure activities and/or reintegration into the work environment. Remember that pain can be a prominent factor during the entire course of recovery. Most problems with sensation resolve over time; however, persistent pain may require treatment via various therapeutic modalities (e.g. TENS, moist heat pack, or sensory desensitization techniques). It is important to note that what may be comforting to one patient may cause discomfort or pain to another. To this end, you should:

- Customize exercises to strengthen weak muscles and watch for muscle substitution. For example, a patient may demonstrate hip-hiking and a circumduction gait pattern as a consequence of substitution for weak hip flexors.
- Plan multiple rest breaks during therapy if fatigue occurs. Exercising to exhaustion will require recovery time that can delay resumption of therapy. If complaints of increased fatigue last more than 12 to 24 hours, then the patient probably has worked too hard.
- Establish a home program that fits the patient's current activity level as soon as they and/or their caregivers have demonstrated a thorough understanding of the exercises. Assure that they are aware of the increased risk of falling as a result of their decreased strength.
- Provide a variety of additional activities that promote gross and fine motor skills and sensory stimulation and/or desensitization outside of therapy. Consider adding aerobic training – at a moderate Rate of Perceived Exertion on the Borg scale – as soon as the patient's doctor indicates that it is safe to do so.
- Provide your patient with adaptive/compensatory strategies and equipment as needed.
- Continue to perform follow-up assessments so that the Plan of Care may be altered to reflect new abilities.

IN CONCLUSION

Physical and occupational therapy are integral parts of the recovery and management of GBS, CIDP and variants. Their proper utilization can help a patient minimize pain, increase strength and endurance and prevent secondary complications and overuse damage to muscles and joints while improving balance and mobility and restoring functional activity at home, work and play. If you or your patient has additional questions regarding GBS, CIDP or variants, please contact the Foundation directly or visit our website at www.gbs-cidp.org. In many instances, one of our local contacts will be available to visit personally with your patient and provide support to them and their caregivers.

Again, thank you for your advocacy!

Matthew Hansen

Doctor of Physical Therapy; Santo Garcia, MOTR/L

ABOUT THE AUTHOR

Dr. Hansen is a practicing physical therapist, President of SOMA Healthcare Staffing & Consulting and a contributing writer for *I.G.Living* magazine as well as the GBS/ CIDP Foundation International's newsletter, *the Communicator*. Santo Garcia, Master's in Occupational Therapy, provides home-based OT. He teaches health care professionals as Adjunct Professor at Hodges University School of Allied Health as well as through his national educational programs. His personal experience with CIDP provides added expertise.

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A close-up photograph of a wheelchair wheel, showing the spokes and the hub. A semi-transparent green rectangular box is overlaid on the upper portion of the image, containing white text. The background is a light, neutral color.

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