GBS/CIDP *Foundation International*

Servicing patients of GBS, CIDP and Variants with support, education, research and advocacy

Guillain-Barré Syndrome • Chronic Inflammatory Demyelinating Polyneuropathy

Summer 2010

The Communicator

HIGHLIGHTS

Research Grants for Investigators

Letters of Intent for the next grant cycle are due to the office on November 1, 2010. Letters of intent can be emailed to the GBS/CIDP Foundation International at info@gbs-cidp.org.

CONTACT US

The Holly Building
104½ Forrest Avenue
Narberth, PA 19072
(866) 224-3301
(610) 667-0131
Fax: (610) 667-7036
www.gbs-cidp.org
email: info@gbs-cidp.org

MEDICAL ISSUE

This annual Special Medical Issue of the *GBS/CIDP Communicator* features articles and comments from experts in the field of GBS and CIDP. We thank all the contributors whose schedules are demanding but nevertheless considered the needs of our readership in bringing us the latest information on GBS and CIDP. We suggest that these newsletter issues be saved. Make them part of a reference library to serve as a ready resource for you or your physician. Additional copies are available upon request.

What an exciting time for our foundation as we mark our 30th anniversary and prepare for our 11th International Symposium being held November 5-7, 2010 at the Dolce Hotel-Valley Forge in King of Prussia, Pennsylvania. This year, in addition to the workshops being offered, the GBS/CIDP Foundation and Wayne State University are offering a special program for physicians. The course, *Inflammatory Neuropathies: The Impact of Clinical Practice on Outcomes* is being offered on Saturday, November 6th from 8:00 a.m. – 5:00 p.m. and physicians who participate will receive 6.50 AMA PRA category 1 Credits. This course is designed to increase awareness of health care professionals towards symptoms of acute and chronic demyelinating polineuropathy in order to encourage early diagnosis, treatment and improved clinical outcome. The CME brochure and registration information is on our website at www.gbs-cidp.org. **This program is recommended by the American Academy of Emergency Medicine.**

In May of this year the foundation awarded three grants out of 11 letters of intent that were received. If you are a CIDP patient who is registered with our organization you probably have received in the mail a package of information containing the "2nd Outcome Patient Survey." Please take the time to fill it out and return it in the envelope provided. Your participation is extremely important. One of the research projects funded by us is the Genome Wide Association Study of GBS being conducted by the University of Pennsylvania School of Medicine in Philadelphia. We have reached out to GBS patients in several northeastern states for your participation. Our new international *Centers of Excellence* program was launched in May and seven facilities thus far, including one in Germany, have received this important designation. The centers will form a network of healthcare facilities to provide expert diagnosis and management of inflammatory neuropathies for patients who are unable to consult with medical experts in the field in their own communities.

I hope you are making plans to join us in King of Prussia for our symposium. Attendees are coming from as far away as New Zealand! Brochures are in the mail. If you did not receive your copy by now, please visit our website at www.gbs-cidp.org for full details and to register.

I am looking forward to seeing you in November!

Continued good health,

Patricia A. Bryant

Patricia A. Bryant
Executive Director

We take this opportunity to thank ***CSL Behring*** *for their support in making this newsletter possible through an unrestricted educational grant.*

Printed on recycled paper.

Predicting and Improving Recovery in Guillain-Barré Syndrome

Dr. Bart C. Jacobs

Department of Neurology and Immunology, Erasmus MC, University Medical Center, P.O. Box 2040, 3000 CA Rotterdam, The Netherlands

Introduction

Research in the last two decades has made considerable progress in understanding the causes of the Guillain-Barré syndrome (GBS).[1] Important breakthroughs in this period were the discovery of the types of infections that precede GBS and how these infections trigger the disease process that results in damaging of nerves and subsequent clinical symptoms. This is now best understood for infections with the bacteria Campylobacter jejuni, that may be responsible for at least one-third of all cases of GBS. These bacteria contain structures that are very similar to those found on human nerves, a phenomenon called 'molecular mimicry'. Infection with these bacteria results in the production of antibodies as part of the normal immune defence. In some susceptible persons the antibodies raised against these bacteria cross-react with nerves, because of the molecular mimicry, and cause nerve dysfunction. Better understood are now the factors that control these processes and determine if a person will develop GBS or not. Despite this progress, the outcome of GBS in patients has remained largely unchanged during the last two decades. Still about one-third of patients require mechanical ventilation at an Intensive Care Unit (ICU), one-fifth has a considerable residual handicap and the majority suffers from severe fatigue and other long-term complications of the disease. One of the main challenges in current research is to apply the new insights in the basic mechanisms of GBS to develop more effective treatment.

Diversity of disease course in GBS

Finding a more effective treatment for GBS is complicated by the large clinical differences between patients. Some patients have a mild leg weakness that recovers spontaneously within days to weeks without leaving residual complaints. At the other end of the spectrum, patients have a devastating complete paralysis of facial, limb and respiratory muscles requiring artificial respiration at an ICU for months. Some of these patients can even die from the disease or suffer from a severe and permanent handicap. Despite these considerable differences, all patients, if treated, receive the same standard treatment with immunoglobulins or plasma exchange. It seems likely that the optimal treatment differs between GBS patients depending on the disease cause and severity. Personalizing treatment would require methods to predict the disease course in individual patients, preferably as early in the disease as possible when treatment is most effective. Such information to estimate the disease course may in the future be used to take precautions and to conduct selective treatment trials in patients with poor outcome. Last but not least, the patients and their relatives can better be informed about the expected disease course and eventual outcome.

Predicting the disease course in individual patients with GBS

Previous studies have already shown that the clinical course of GBS is influenced by demographic factors (such as age), types of preceding infection, antibody response and various clinical characteristics. This information was, however, difficult to use in individual patients in clinical practice. Recently we have developed so called 'prognostic models' that will enable clinicians to predict the clinical course in individual patients with GBS.[2,3] The Erasmus GBS Outcome Score (EGOS) is a simple yet accurate prognostic model to calculate outcome at 6 months after onset of disease (see figure).[2] The model is based on three clinical characteristics that can easily be defined at the bedside of a patient: (1) age of the patient at disease onset, (2) presence of an episode with diarrhea before disease onset, and (3) GBS disability score, which is an easy and frequently used score expressing the severity of clinical state. The scores of these three items are summed and the resulting EGOS score accurately predicts the chance of poor outcome, defined as not reaching independent walking at 6 months. Patients with the lowest EGOS scores have a probability of less than 5% of poor outcome compared to patients with the highest scores that have a probability of 85% (see figure). This prediction can be made with high accuracy as soon as 2 weeks after hospital admission without any further laboratory investigations. Such models will assist clinicians to inform patients and relatives and to conduct future trials selectively in patients with poor outcome.

A second prognostic model was developed to predict the probability that patients will need intubation and artificial respiration and referral to an ICU or to a hospital with such a facility.[3] This Erasmus GBS Respiratory Insufficiency Score can be used already at hospital admission and is also based on three easily obtainable clinical characteristics. These are (1) facial weakness or problems with swallowing, (2) rapidity of disease progression, and (3) severity of weakness in arms and legs. Patients with the highest summed scores have a probability of requiring intubation or ventilation of more than 80% and should be monitored carefully at or near an ICU. Future studies will have to define if patients with such a high risk should undergo elective intubation to prevent emergency intubations with the related complications. These two evidence-based prognostic models will help clinicians in making well-informed decisions that were previously based on clinical expertise only.

Optimizing treatment for GBS

From the new insights in the pathogenesis several possible new forms of therapy have evolved that need to be evaluated in clinical studies. Until now the most frequently used treatment for GBS are infusions of immunoglobulins (IVIg). At present all patients receive the same standard dose of IVIg of in total 2 gram per kilogram bodyweight. This is an effective treatment for a proportion of patients, but not for all. Some patients improve only temporarily after IVIg, and may require a second dose to induce further recovery. Others do not recover after IVIg or even further deteriorate. One possible explanation for this could be that these patients have a more severe form of disease. Alternatively, these patients with poor recovery could have a higher consumption of the IVIg and may benefit from a higher or second dose. IVIg consists predominantly of IgG and the blood levels after IVIg may give an indication of the consumption rate of the therapy. We were able to investigate the blood levels of IgG in a large group of patients with GBS treated

with the same standard dose of IVIg.4 To our surprise these patients showed a large variation in blood IgG levels after start of treatment compared to baseline levels. Some patients did not show any increase of IgG compared to base line at all, suggesting that all IVIg was rapidly removed from the blood and had probably been consumed already. Patients with the lowest levels of IgG also had the worst recovery. Probably, these patients have a higher consumption rate of IVIg and may require a second dose of IVIg.

It is important to note that the value of measuring IgG levels after IVIg to adjust treatment dose, has not been demonstrated and requires further investigation. In the meantime, however, we have started a new treatment trial for GBS in which we will focus on patients with the poorest predicted outcome according the prognostic models, and investigate the use of treatment with a second IVIg dose. In this study all patients will receive the usual first dose of IVIg. Half of the patients with a poor predicted outcome after this first course will receive a second dose of IVIg. The other half will receive a sham therapy to be able to measure the efficacy of this second dose of IVIg. By choosing this trial design we hope to be able to improve outcome in patients that need it most.

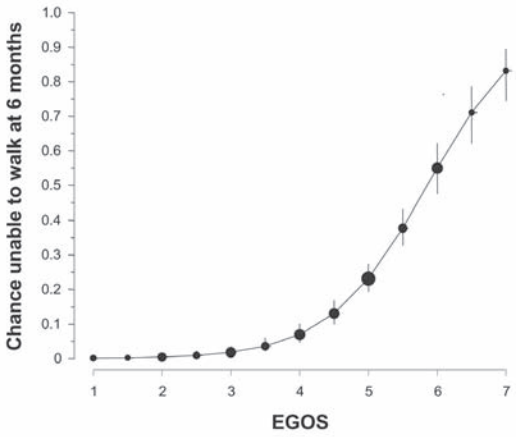
Future studies

More extensive studies are required to further define the mechanisms of disease progression and recovery and to predict in more detail the clinical course and outcome in GBS. In 2010 a group of experts on GBS from all over the world and united in the International Neuropathy Consortium, will start a unique collaborative study to bridge the gap between basic and clinical research. In this International GBS Outcome Study we aim to include more than a 1000 future patients to perform extensive studies on the genetics, immunology, clinical recovery and long-term outcome of GBS. A website for registration of patients in this study has already been developed with support of the GBS/CIDP Foundation International. In this study there will be an intensive collaboration between basic scientists, clinicians, patients and their relatives and representative organisations, including the GBS-CIDP Foundation International.

Better understanding the factors that influence the disease activity and recovery in individual patients will pave the way to develop personalized treatment for GBS.

Erasmus GBS outcome score (EGOS)

Predictors	Categories	Score
Age (years)	≤40	0
	41-60	0.5
	>60	1
Preceding diarrhoea	absent	0
	present	1
GBS disability score	0-1	1
	2	2
	3	3
	4	4
	5	5
EGOS		1 - 7



Legend to the figure.

The EGOS is a prognostic score, ranging form 1 to 7, which can be used to predict the chance of not walking independently at 6 months after onset of GBS (see reference 3). The EGOS can be calculated by the sum of the scores for the following three items: (1) age of the patient, (2) episode of diarrhea in the 4 weeks preceding the onset of GBS, (3) GBS disability score at two weeks after hospital admission (left panel). The GBS disability score is a measurement for disease severity grading 1 for minor symptoms, 2 for able to walk 10 meter without assistance but unable to run, 3 able to walk 10 meter across an open space with help, 4 bedridden or chairbound, 5 requiring assisted ventilation. For example, a patient of 67 years old (1 point), who had a history of diarrhea before GBS (1 point) and was bedridden at two weeks after hospital admission (4 points). This patient has an EGOS of 6 with a chance of being unable to walk independent at 6 month of 55%, although this patient may show further recovery afterwards.

Legislative Website Update

As our Foundation's website continues to evolve we want to be sure that everyone accessing the site has some advance notice of what's being added, deleted, modified and/or updated. Such is the case when you access the Legislative Action tab to the far right top of the home page. When you click on it you'll be able to access two entirely different, but very useful data resources. The Legislative Action link brings you into the realm of all things necessary when tracking both federal and state bills that have a connection to our Foundation in one way or another. It also provides Congressional contact info, voting records, media contact and so on. Folks, this site is a very powerful tool that allows you to communicate directly with all your elective officials by phone, email, or fax. Just follow the friendly prompts!

The new tab for Advocacy has been set up to be your gateway to subject matter that provides communication records of importance to the Foundation and our fellow patient advocacy groups. Included are congressional letters, Foundation letters, coalition letters, Foundation and coalition presentations and so on.

You'll also be able to access educational material concerning all key and relevant components of the newly enacted Patient Protection and Affordable Care Act of 2010. You'll be able to learn more about Medicare & Medicaid issues including how to select insurance coverage options based on the state where you live. We also added links to websites that will help you handle insurance problems.

This is not a complete list. Much more will be added over the next few months so please come back time and time again to keep current on the additions or changes. I would also encourage you to contact me with any suggestions on how we can improve this site. ed.gdula@gbs-cidp.org

1. van Doorn PA, Ruts L, Jacobs BC. Clinical features, pathogenesis, and treatment of Guillain-Barré syndrome. Lancet Neurol. 2008;7:939-50.
2. van Koningsveld R, Steyerberg EW, Hughes RAC, Swan AV, van Doorn PA, Jacobs BC. A clinical prognostic scoring system for Guillain-Barré syndrome. Lancet Neurol. 2007;6:589-94.
3. Walgaard C, Lingsma HF, Ruts L, Drenthen J, van Koningsveld R, Garssen MJP, Steyerberg EW, Jacobs BC. Prediction of respiratory insufficiency in Guillain-Barré syndrome. Ann. Neurol. 2010;67:781-7.
4. Kuitwaard K, Gelder J de, Tio-Gillen AP, Hop WCJ, Gelder T van, Toorenenbergen AW van, Doorn PA van, Jacobs BC. Pharmacokinetics of IV immunoglobulin and outcome in Guillain-Barré syndrome. Ann. Neurol. 2009;66: 597-603.

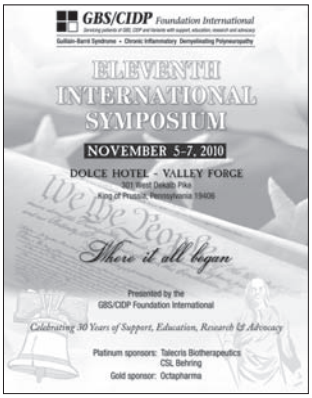
CIDP Treatment Update

Gareth J. Parry, M.D.
Department of Neurology, University of Minnesota

Chronic inflammatory demyelinating polyneuropathy (CIDP), as a distinct clinical entity, was separated from Guillain-Barre syndrome (GBS) only about 50 years ago. Prior to that time the two disorders had been largely lumped together and, indeed, they share many clinical, electrophysiological (EMG) and pathological features. The main difference is in their differing temporal evolution; GBS progresses for no more than four weeks whereas CIDP always progresses for more than eight weeks and usually for much longer. CIDP is uncommon in terms of new cases each year (incidence) but because of its chronic nature, large numbers of patients continue to suffer from this disease (prevalence); based on estimates of prevalence there are probably about 25,000 people with CIDP in the US. Another area where CIDP and GBS diverge is in the response to treatment although even here there are more similarities than differences. Importantly, corticosteroid medications (such as prednisone) have no role in the treatment of GBS but are effective in CIDP. Both conditions respond favorably to treatment with plasma exchange (PLEX) and intravenous immunoglobulin (IVIg). In CIDP treatment is most effective if started early, within two years of diagnosis. With prolonged duration of CIDP there is inevitable axonal degeneration which has limited ability to recover.

Like GBS, CIDP is an auto-immune (inflammatory) condition in which harmful antibodies attack the myelin sheath of peripheral nerves. Thus, treatment is aimed at removing antibodies or preventing their formation. There is no consensus on what constitutes the best treatment, probably because there is no one best treatment. It is important to note that some individuals respond to one treatment but not others and a trial of each treatment may be needed before declaring treatment failure and moving on to more aggressive and more dangerous therapies. What follows is a personal opinion shared by the neurologists at the University of Minnesota regarding treatment options in CIDP.

1. **Corticosteroid treatment:** Corticosteroids are chemicals that are produced in small quantities in the body by the adrenal gland. When used at high doses they have potent anti-inflammatory effects. Successful treatment of CIDP with steroids was first reported in the late 1950's and was established by careful clinical research trials in the 1970's. The most common approach was to use daily prednisone which was certainly effective; in the only study comparing steroids and IVIg the treatments were comparably effective. The problem with daily steroids is that long term treatment with high doses is usually necessary resulting in inevitable adverse effects such as weight gain, skin changes, fluid retention, hypertension, diabetes, osteoporosis and many others. More recently steroid treatment has been administered intermittently (known as pulsed treatment), usually once a week, at very high doses with no loss of effectiveness and a marked reduction in adverse effects. We administer the steroid methylprednisolone at a dose of 500 mg orally each week and have found that almost all patients improved and more than 50% are in complete remission, functioning normally and taking no medication. A recent Dutch study showed that daily steroids and pulsed steroids were equally effective in inducing remission. With weekly, rather than daily, dosing we have found the medication to be quite well tolerated with none of the side effects listed above except for osteoporosis in patients over the age of 50. Irritability and insomnia are common on the day of treatment but are usually mild and do not need treatment. About 30% of patients get mild indigestion or heartburn. Because this treatment is so effective, is so safe and well tolerated, is convenient to administer and is inexpensive we regard this as the primary treatment option in CIDP.
2. **Intravenous immunoglobulin (IVIg) treatment:** Immunoglobulin is a fraction of human plasma containing the antibodies that are important in fighting disease. When administered in large quantities immunoglobulin has been shown to be effective in fighting many autoimmune diseases, including CIDP. IVIg was first used to treat CIDP in the 1980's. Several small studies in the 1990's showed the drug to be effective and more recently a large international study confirmed the effectiveness. IVIg, steroids and PLEX are all equally effective in controlling CIDP. Although IVIg is effective in controlling the disease it probably does not induce remission and, therefore, has to be administered by way of regular intravenous infusions, usually every 2-3 weeks, often for many years. The infusions can be administered at an infusion center or through specialty home infusion services. IVIg is extremely well tolerated and safe with none of the side effects seen with steroids treatment although headache during the infusion may occur as can mild allergic reactions. Rare serious adverse effects including stroke, heart attack, pulmonary embolism (blood clots in the lungs) and kidney failure can occur, almost always in patients older than 60. The problem with IVIg is that it is expensive (more than $100,000 a year for average doses in CIDP) and inconvenient to administer. For these reasons we regard IVIg as second line treatment for patients who have failed or are unable to tolerate steroid treatment.
3. **Plasma exchange (PLEX):** PLEX is a technique for removing harmful antibodies from the circulation. It has been used to treat CIDP since the 1980's and is just as effective as both steroids and IVIg. Like IVIg it probably does not induce remission and requires long term treatment every 2-3 weeks. It requires complex and expensive equipment and has to be done at a transfusion or dialysis center, making it inconvenient. The cost is comparable to IVIg but because of the inconvenience we regard PLEX as third line treatment and rarely use it.
4. **Subcutaneous immunoglobulin (SQIg):** One strategy that is gaining momentum at present is to administer immunoglobulin subcutaneously (under the skin). The development of more concentrated forms of the drug has made this approach feasible. Preliminary studies suggest that SQ is as effective as IV administration and has the advantage of ease of administration; there is no need for venous access which can be difficult in many patients. SQ administration also seems to eliminate many of the risks of IVIg. More study is needed before this procedure can be widely recommended but it is promising.
5. **Unproven treatments:** Because a small proportion of patients do not respond to the first three treatments described above a number of other therapies have been tried but none has been studied in rigorous clinical trials. Immunosuppressive drugs such as **azathioprine** (Imuran) and **mycophenylate mofetil** (Cellcept) are often used in patients who respond partly to another therapy or for whom side effects or consideration of cost make the primary treatments unsatisfactory. More aggressive chemotherapy with drugs such as **cyclophosphamide (**Cytoxan) is also occasionally used and is effective but carries greater risk. **Rituximab** is a drug that specifically destroys the immune cells that produce the harmful antibodies and, theoretically, should be beneficial in CIDP but experience has been varied with as many negative as positive reports of the effects. Several other chemotherapy drugs are on the horizon and several hold out promise for CIDP but all will carry greater risk than the primary agents.



11th International Symposium

sponsored by the
GBS/CIDP Foundation International
is shaping up to be an exciting event!

Symposium brochures are in the mail.

If you have not already registered for this very special event please access our brochure and registration form online at www.gbs-cidp.org.

CIDP Patients – 2nd Outcome Study

The 2nd Outcome Study was mailed the week of August 16th to U.S. CIDP patients who are registered with the Foundation. This study is designed to provide our organization with data on the effectiveness of various treatment protocols and the quality of life being experienced by CIDP patients along various stages of treatment. Part III pertains to access to care and insurance issues. This information will be of tremendous importance in the Foundation's advocacy efforts on your behalf at federal and state levels and with insurance providers.

Please take the time to complete the study and return it by October 1, 2010 in the self addressed, prepaid envelope we have provided. All responses are anonymous.

Genome Wide Association Study of GBS

Investigators at the University of Pennsylvania School of Medicine in Philadelphia, Pennsylvania are conducting a study to examine genetic factors for susceptibility to developing Guillain-Barré Syndrome.

Eligibility for participating includes the following:

- Age 18 years or older
- Have had GBS at anytime in the past
- Be willing to provide limited medical records documenting a diagnosis of AIDP (GBS)
- Be willing to donate a blood sample for analysis

If you are interested in hearing more about this study and live in PA, NY, NJ, DE, DC, MD, CT and VA and potentially would like to participate, please contact:

Ms. Sarah Kirsch, UPenn-GBS Study Coordinator
University of Pennsylvania School of Medicine Email: gbsstudy@mail.med.upenn.edu

Research Grants Awarded!

On May 22, 2010 three research grants were awarded by the GBS/CIDP Foundation International to the following:

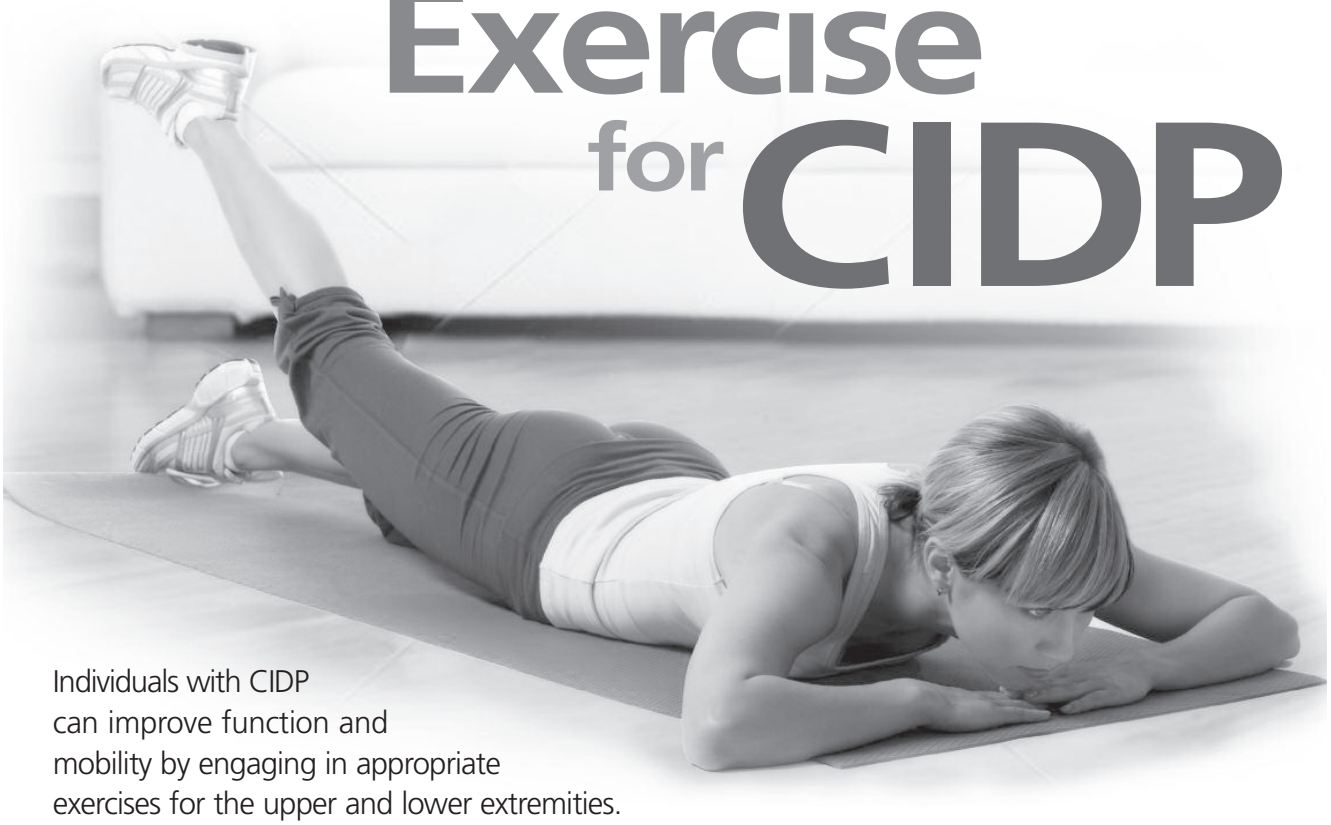
Genome Wide Association Study of Guillain-Barré Syndrome
Irving Nachamkin, M.D.; Arthur Asbury, M.D.

CCL2-CCR2 blockade as targeted immune-modulatory therapy for inflammatory demyelinating polyradiculoneuropathies
Eroboghene E. Ubogu, M.D.

The role of antibodies against proteins of the node of Ranvier in the pathogenesis of Guillain-Barré syndrome: finding novel immune targets
Jérôme J. Devaux, Ph.D.

Reprinted with permission from IG Living magazine, August-September 2010, www.igliving.com

Exercise for CIDP



Individuals with CIDP can improve function and mobility by engaging in appropriate exercises for the upper and lower extremities.

By Matthew David Hansen, DPT, MPT, BSPTS

Anyone who has experienced local anesthesia or has had an arm or leg "fall asleep" knows how frustrating loss of feeling and/or impaired control of a body part can be, even for a short while. Our brains have an inherent desire to know the status of our body parts at all times. When something is preventing — or interfering with — the signals that travel through our nervous system to and from our brain, it can have a dramatic effect on our behavior and our ability to function.

Think about the last time that you were given a shot of Novocain before having dental work performed. Can you remember what you probably began doing even before you left the office? You might have run your tongue along the side of your mouth that was numb; rubbed, pinched or tapped your face; or even gently bit your cheek to see how hard you could squeeze before feeling something. Why? The impulse comes from our brains as they try to figure out what's going on and to regain control of the chronic inflammatory demyelinating polyneuropathy (CIDP) is by no means a fair association. However, it may help those who have never suffered from a neuropathy to have some concept of what is experienced day after day by someone who has.

What Is CIDP?

CIDP, commonly considered the chronic equivalent of a similar condition named Guillain-Barré syndrome, is a disease of the peripheral nervous system that is caused by an abnormal immune response that mounts an attack on myelin (a fatty covering that protects nerve fibers and allows for a signal to be relayed quickly). As a result of the lost myelin, the affected nerves respond weakly or not at all to stimuli, resulting in progressive muscle weakness, fatigue, loss of deep tendon reflexes and atypical nerve sensations (tingling, burning, numbness and/or pain). Symptoms are usually symmetrical and frequently cause difficulties with walking and the coordination of other movements. The autonomic nervous system also may be involved, leading to complaints of dizziness when changing positions, heart symptoms and trouble with bowel and bladder function.

In most patients, the course of CIDP is slowly progressive; however, it is not uncommon for periods of recovery, lasting weeks to months, to occur between relapses. Although there is currently no known cure for CIDP, symptoms can be treated via corticosteroids to reduce inflammation, plasmapherisis to remove harmful antibodies from the blood, intravenous immune globulin (IVIG), immunosuppressant drugs (in some

severe cases) and exercise. Early medical treatment is important to confine nerve damage to the myelin sheath and to prevent harm to the axons (nerve fibers) themselves.

How Exercise Can Help

Appropriate exercise is a vital part of any CIDP intervention plan because of its potential to improve strength and endurance, thereby minimizing muscle shrinkage and improving function and mobility. Understanding some of the recommendations that have emerged from scientific research for those exercising with a peripheral neuropathy can help to establish a proper program.

First, patients should always visit with their medical doctor before beginning an exercise regimen. This is an important principle for any population; however, it is even more essential for those with a peripheral neuropathy, because the wrong exercise parameters can actually make a bad situation worse rather than better. The possibility of the autonomic nervous system being involved also means that the body may not respond to exercise in a typical manner.

Second, patients shouldn't overdo it! The damage caused by CIDP to myelin, and the possible damage to axons, results in the body's ability to recruit fewer muscle fibers to perform a task. Consequently, those muscle fibers that are engaged are at greater risk of being overworked. Some soreness after exercise may be expected, but it should dissipate within 12 to 48 hours. If pain persists, is exaggerated or is coupled by a loss in strength, the patient likely did too much.

Submaximal exercise is frequently recommended for peripheral neuropathies.[2] A doctor and/or a properly trained physical therapist can help patients find the exercise prescription (frequency, intensity, time and type of activity, known as the F.I.T.T. principle) that is currently right for them. Low-impact exercises like walking, swimming, riding a recumbent bike or performing "open-chain" arm and leg exercises (without bearing weight through the extremity) might also be good alternatives to high-impact activities like running or jumping.

Third, patients need to be aware of their physical limitations. Activities that put them at undue risk of falling or other physical injury should be avoided. And, they shouldn't hesitate to ask someone to help, or at least accompany, them during their workouts.

Fourth, muscle strengthening and aerobic conditioning are important. Science has demonstrated that strength exercise programs can improve muscle force in patients with peripheral neuropathies.[1,3,4] However, it also has been shown that aerobic conditioning is important in combating fatigue and other impairments, particularly in the later stages of recovery.[1,3,4]

And last, patients should wait until a muscle can work against gravity before stressing it against additional resistance. Fortunately, under normal circumstances, myelin and peripheral nerve fibers can regenerate, with muscle control gradually returning as it does. However, with CIDP, residual damage is not uncommon and recoveries can take some time. Therefore, it is important to progress exercises in a systematic way in order to avoid overstressing muscles and joints.

Appropriate exercise is a vital part of any CIDP intervention plan because of its potential to improve strength and endurance, thereby minimizing muscle shrinkage and improving function and mobility.

Choosing the Appropriate Level of Exercise

Those experiencing an immunological disease exacerbation probably find it difficult to imagine themselves exercising. The biggest hindrance may not be the weakness they are experiencing, but instead, the popular misconception that exercising means performing a workout à la Jane Fonda, Billy Blanks or even Richard Simmons. The reality is that there are multiple levels of exercise difficulty, each as achievable and as genuine a workout for those to whom they are prescribed as a typical exercise video would be for a fully able-bodied recreational athlete. For CIDP patients, the following exercise progression levels can be used for particular exercises, but which level is appropriate will depend upon what the patient is ready for.

Passive exercise: Gentle movement of the body (usually the limbs) is performed by a properly trained individual, without effort on the patient's part. Passive movement can be beneficial for maintaining or improving blood circulation and range of motion. Thinking about the movement and trying to assist may also help to re-establish nerve connections in cases where actual damage has occurred to the nerve and regeneration is under way.

Active-assisted exercise: Assistance is still required from another person, but the patient is able to participate in movement to some degree. Actual activation of the muscle(s) is occurring; however, it is still not strong enough to move the limb independently.

Active exercise (gravity eliminated): Independent movement is possible in a gravity "eliminated" position, but not against gravity. For example, a patient may be able to lift their knee toward their chest (hip flexion) while lying on their side in bed (gravity eliminated position), but not while standing (against gravity).

Active exercise (gravity reduced): Movement is possible against some gravity, but not against its full pull. To use the same example of hip flexion, a patient may be able to bring a knee toward their chest while lying on their back (gravity reduced), but not while standing (against gravity).

Active exercise (against gravity): Movement is possible in all planes (including standing for hip flexion), but without additional resistance.

Resistive exercise: The highest level of progression, but also the most variable level, limited only by the potential

continued on page 8 ➤

of the conditioned human body to produce force. Resistance may take the form of weights, resistive bands, household items, one's own body weight, etc.

Lower- and Upper-Extremity Exercises

There are several upper- and lower-extremity exercises for some of the most important gross motor (large muscle) actions performed by the body. The figures in this article provide one example of the progression levels for each exercise. These exercises also can be performed while patients are lying on their back, stomach, sides, sitting and standing.

Lower-extremity exercises. The lower-extremity exercises predominantly involve the hip, knee and ankle. Hip flexion (Figures 1-4) is the action performed when lifting the leg to walk forward or step up onto something. In addition to hip flexion, two other hip actions — extension and abduction — are fundamental to an individual's ability to walk and maintain balance.

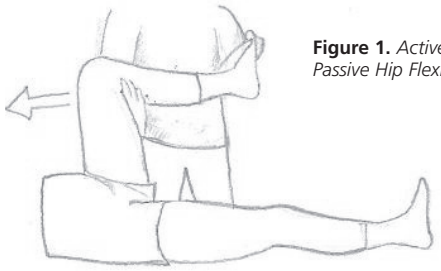


Figure 1. *Active-Assisted or Passive Hip Flexion*

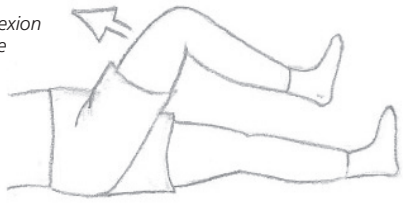


Figure 2. *Active Hip Flexion while lying on your side (Gravity Eliminated)*

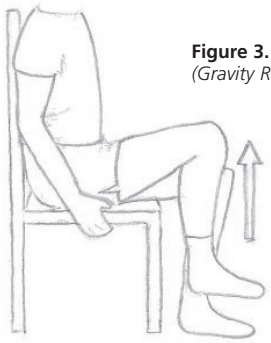


Figure 3. *Active Hip Flexion (Gravity Reduced)*

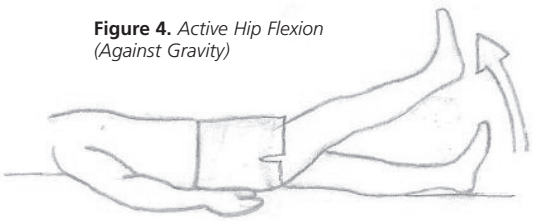


Figure 4. *Active Hip Flexion (Against Gravity)*

Hip extension (Figure 5) is the motion that helps to pull the leg back and propel the body forward after taking a step. Hip and knee extension also are needed to stand up from a chair, the floor or a squatting position, to jump or to climb stairs. For instance, an individual lifts their foot to the next step via hip flexion, but they progress, or pull, themselves up to the step via hip extension.

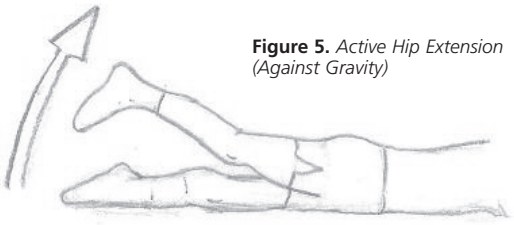


Figure 5. *Active Hip Extension (Against Gravity)*

Hip abduction (Figure 6) is most easily visualized as an open-chain exercise. However, the prime muscle of hip abduction, the gluteus medius, performs most of its work in a weight-bearing mode as a "closed chain" exercise when the leg may not even be moving. To experience this phenomenon, place an open hand over your hip; not the boney area at your waist line (that's part of your pelvis), but the area just below it. Now, if you are able to, lift the opposite leg (while stabilizing yourself against something with your free hand) so that you are standing on one foot. Did you feel anything happen under the hand that is placed over the hip? That's the gluteus medius contracting. If it didn't, your body would fall to the side of the leg that is being held up off of the floor.

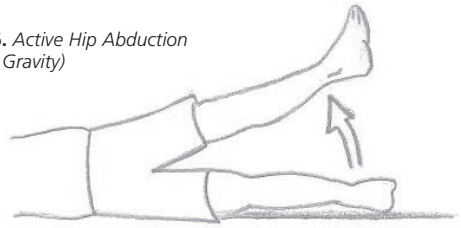


Figure 6. *Active Hip Abduction (Against Gravity)*

Besides its use in standing, jumping and climbing, knee extension (Figure 7) also is the motion of kicking — perhaps not something that individuals still do every day, but it was probably more important to people at some time during their lives!

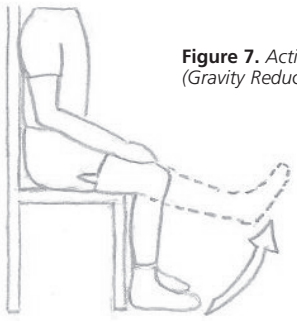
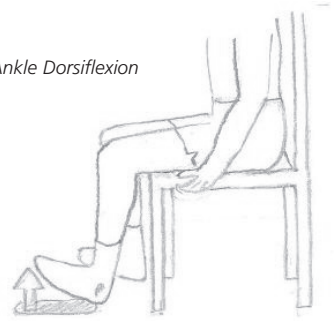


Figure 7. *Active Knee Extension (Gravity Reduced)*

The inability to dorsiflex the ankle/foot (Figure 8) is a frequent complication of peripheral neuropathies. The condition becomes especially troublesome when the toe drags or catches the floor when walking, causing a patient to

stumble or making it impossible to advance the limb forward without using an assistive device and/or dramatically changing the way that they walk.

Figure 8. *Active Ankle Dorsiflexion (Against Gravity)*



Upper-extremity exercises. The upper-extremity exercises predominantly involve the shoulder, elbow and wrist.

Shoulder flexion (Figure 9) is the chief shoulder action used by individuals to reach for something in front of their body or over their head (such as shaking someone's hand or getting something down from a shelf).

Figure 9. *Active Shoulder Flexion (Against Gravity)*

Elbow flexion (Figure 10) is used to lift and carry (such as carrying a box) and to bring objects that are grasped closer to the body. It also is used to simply lift something to one's mouth to eat or drink.

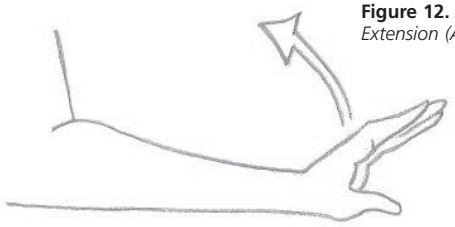
Figure 10. *Active Elbow Flexion (Resistive Exercise)*

Elbow extension (Figure 11) straightens the elbow and gives individuals additional length to reach an object when shoulder flexion alone isn't enough. When functioning as a closed-chain exercise, elbow extension also is fundamental in helping individuals to push themselves up to a sitting or standing position. This exercise is especially helpful when trying to get out of bed without using the arms.

Figure 11. *Active Elbow Extension (Gravity Reduced)*

There are also many fine motor (small hand muscle) exercises that can be performed to increase grip strength and improve precision handling of objects. For instance, the wrist extension (Figure 12) is a functional position used while eating, handwriting, keyboarding, driving, grasping objects and performing a number of other tasks.

Figure 12. *Active Wrist Extension (Against Gravity)*



Tailoring the Program to Each Individual

The muscles targeted by the exercises presented in this article are just a few of the 640 skeletal muscles found in the body, but they are some of the most important to everyday function. While it's true that CIDP and other immunological diseases certainly can be disabling, appropriate exercise to improve strength and endurance can lead to better daily function. Individuals with CIDP are capable of doing a lot, and there is a level of exercise that's right for everyone! ■

References

1. Bassile, CC. Guillain-Barre Syndrome and Exercise Guidelines. *Journal of Neurologic Physical Therapy,* 1996; 20(2): 31-36.
2. White, CM, Pritchard J, Turner-Stokes L. Exercise for people with peripheral neuropathy. *Database of Systematic Reviews,* 2004, Issue 4. Art.No.: CD003904.DOI:10.1002/14651858.CD003904.pub2.
3. Ruhland, JL, and Shields, RK. The effects of a home exercise program on impairment and health-related quality of life in persons with chronic peripheral neuropathies. *Physical Therapy*, October 1997.
4. Garssen, MP, Bussmann, JB, Schmitz, PL, Zandbergen, A, Welter, TG, Merkies, IS, Stam, HJ, and van Doorn, PA. Physical training and fatigue, fitness and quality of life in Guillain-Barre Syndrome and CIDP. *Neurology*, Dec. 28, 2004; 63(12): 2393-5.

MATTHEW DAVID HANSEN, *DPT, MPT, BSPTS, is a practicing physical therapist in Washington state and president of an allied healthcare staffing and consulting agency. He completed his formal education at the University of Utah, Salt Lake City, and has additional training in exercise and sports science, motor development and neurological and pediatric physical therapy.*

Illustrations by Veronica Hansen

GBS/CIDP Foundation
International Chapters EVENTS AND NEWS!

Annual Meeting of the Dutch GBS/CIDP Support Group
Saturday, October 2nd, 2010
in Veldhoven, The Netherlands
For further information and registration: www.vsn.nl
Phone: 035 - 5480480

Patricia Bryant, Executive Director of the Foundation, recently visited with members of the Australian Chapter in North Epping, Australia

Australia (NSW) Chapter members:
Top left - Ronald Nichols and Glenda Ford
Bottom - Mary and Arthur McAlister

Krista Kuitwaard Wins PJ Dyck Award

During the congress of the Peripheral Nerve Society in Sydney, Krista Kuitwaard, resident in Neurology at ErasmusMC in Rotterdam, was awarded the Peter James Dyck prize. This award is named to one of the most important researchers in peripheral nerve disorders and is awarded yearly to a young and promising researcher in the field of peripheral neuropathies. Krista Kuitwaard was awarded this prize for her research on identifying factors related to a favourable response to intravenous immunoglobulin in patients with chronic inflammatory demyelinating polyradiculoneuropathy (CIDP), the chronic variety of Guillain-Barré syndrome. This research was conducted in collaboration between ErasmusMC and researchers in Canada.

INTERNATIONAL NEWS

The GBS/CIDP Support Group of the United Kingdom and Ireland celebrated their 25th Anniversary in April, 2010. Unfortunately it coincided with the Icelandic Ash trauma. The venue had been chosen at the Radisson Hotel, Heathrow Airport, and London to enable easy access for international delegates to attend. Ha Ha Ha!

Though British patients, friends and relatives attended and one very welcome GBS/CIDP FI director, Phil Kinnicutt (he had flown in early so missed the chaos by a day and wondered if he would ever return home to Hawaii!). However, the reorganization did not stop a very successful conference (150 attending), with excellent speakers including; Dr. John Winer, a Children's Neurological Pediatrician, a fascinating insight into the Jubilee Sailing Trust (to enable able-bodied and disabled to crew a tall ship around the waters of the world) Dr. Mary Reilley, National Hospital for Neurology and Neurosciences, and by video conference link, Prof. Hugh Willison from Glasgow, Scotland. Prof. Willison was unable to fly to London because of the volcanic ash problem. The hotel had a completely different atmosphere, silent of planes taking off and landing and guests going nowhere! A fuller report on www.gbs.org.uk.

The UK Support Group has held many activities to celebrate and raise money in their 25th year. Howard Sanders, the Founder's husband, walked Hadrian's Wall and raised over £1,500. Glennys, the Founder and International Director of GBSFI, held a Ladies Luncheon for 75 friends with topics varying from hairdressing, flower arranging, adventures on The Tenacious Tall Ship around the Greek Islands and key speaker, Helen Young , accounting her career at the Meteorology Office presenting the BBC weather - with very funny anecdotes. Several people have run various marathons in London, San Francisco, Belfast and New York.

I have recently met the Pakistan liaison, Khalid Mehmood Zia, in London. Khalid is doing excellent work at the Plasmapheresis Center in Islamabad and has set up a mobile plasmapheresis service providing, free of cost, better care and rapid management for the needy and poor patients with Myasthenia Gravis, GBS, CIDP, and others related diseases. His tremendous work has saved many lives in Pakistan. However, since we met, Pakistan has experienced unprecedented floods and the necessary supply of plasmapheresis is not getting to the people who need it. How unfortunate.

The International Support Groups (23) now have or are developing websites for GBS/CIDP in their own languages which is great news for the patients and people in these countries.

Glennys Sanders, Co International Director

Glennys Sanders and Khalid Mehmood Zia

Help Support Our Organization Every Time You Search or Buy Online!

Add the GBS CIDP Foundation International toolbar to Internet Explorer or Firefox. Once you add the GBS CIDP Foundation International toolbar for Internet Explorer or Firefox, each time you shop at one of the more than 1,300 participating stores, a percentage of what you spend will be donated to us at no extra cost to you! You could even save money as the toolbar provides coupons and deals. The toolbar also has a search box and each time you search the Internet, about a penny is donated to us. No registration is required! Here is the link to download the Foundation's toolbar:

GoodSearch
YOU SEARCH WE GIVE
GoodShop

http://www.goodsearch.com/toolbar/gbs-cidp-foundation-international

Letters to the Editor

July 15, 2010

To Whom It May Concern:

My name is Emily Smith, and this past fall I was privileged to serve as Worthy Advisor of Silverdale Assembly #155 of the International Order of the Rainbow for Girls. Rainbow Girls, as it's called, is a Masonically-affiliated service organization for young ladies between the ages of eleven and twenty. Members are taught service and leadership. Each Worthy Advisor, when planning her six-month term, chooses a service project to support. Past Worthy Advisors in Silverdale Assembly have chosen causes such as Autism Speaks and the Kitsap Humane Society. I chose to support the GBS/CIDP Foundation International because I have personal experience with your wonderful organization.

In the winter of 2006, when I was sixteen years old, my mother Amy was hospitalized with Guillain-Barré Syndrome just before she was supposed to leave for a trip to Washington DC. She was taken to Harborview Medical Center in Seattle, then moved to Saint Joseph's Hospital in Tacoma for recovery. I had never heard of GBS before, and I was scared not knowing what was going to happen. Luckily, my mother recovered and returned to her job as an elementary school teacher the following year. Today she is doing well.

When I was planning my term as Worthy Advisor, I had many things to think about – the dress I would wear, what food I would serve at my installation, what trips my assembly would take, and countless others. However, it did not take me long to decide that if I had anything to say about it, my assembly would be supporting the GBS/CIDP Foundation. The work that you do, creating awareness of and providing support for people with these difficult conditions, is incredible and it touches my heart.

My main fundraiser was a Dinner Theater Dinner, held in October 2009. I chose this fundraiser because my theme for my term was Broadway, and it seemed like a unique way to raise money. Many people, including local Masons and Eastern Star members as well as parents and families of Rainbow Girls, came to support us. We served a pasta buffet and held a silent auction while the girls of the assembly performed skits to entertain the guests. I also set up a display board about GBS/CIDP and gave a speech explaining the seriousness of these conditions to guests who were unfamiliar with them. Thanks to the generosity of our local community, we raised over two hundred and sixty dollars!

I hope that this contribution can even come near expressing how grateful I am for the support that your organization provided to my family during a difficult time in our lives, and how wonderful and necessary the awareness that you raise is.

Sincerely,

Emily Margaret Smith, Past Worthy Advisor of Silverdale Assembly #155
Washington-Idaho International, Order of the Rainbow for Girls

Thank you to the Mechanical Contractors Association of NY for their generous contribution to the GBS/CIDP Foundation International in the amount of $15,000. The presentation was made at the North Hills Country Club in Manhasset, NY following the association's golfing event. The people in the picture are from left to right are: MCA Vice President, Anthony Bell, Promotion Fund Trustee, Reed Rickman of Crescent Contracting Corp., Steamfitting Industry Promotion Fund Chairman, Sal Barbera, along with Michael Bildner (Brooklyn liaison), Brie Landry (GBS survivor), Nancy Richter(Nassau County liaison), and MCA Executive Vice President, Al Gettler.

June 26, 2010

While reading your COMMUNICATOR newsletters I continue to be impressed with the courage and fortitude of GBS/CIDP patients. So many of them seem to endure a long recovery and very real uncertainty over the eventual outcome.

Our family has experienced GBS first hand, but our story is one that I hope can be a source of encouragement to your readers.

Antoinette, days before her 83rd birthday in April 2003, experienced the classic symptoms – loss of feeling and a tingling sensation in her feet and finger tips increasing weakness, sensory loss and inability to walk. She was seen at the neighborhood emergency medical center and diagnosed with a "severe case of dehydration." When symptoms persisted overnight, we took her to the ER for more extensive testing for "general weakness etiology unknown."

When the family refused to take her home, a neurologist was called in, correctly diagnosed GBS and arranged her immediate transport to University Hospital of Cleveland. She was intubated and spent 9 days in the neuroscience ICU before being moved to 28 days general care and inpatient rehab. Although GBS/CIDP patients are relatively rare, virtually every member of the staff – and especially the RN case manager assigned to her care became "experts" almost immediately.

This Easter (2010) our family helped Antoinette celebrate her 90th birthday. For the past 6 1/2 years since this episode she has been living independently in her own home, active in quilting, church activities, a weekly bowling league and visiting family out of town.

Antoinette's case is a great example of what can be accomplished by prompt diagnosis, knowledgeable medical staff, and informational support from your organization. This lovely lady has proven that age does not need to be an impediment to a successful recovery. God grant her story can be an inspiration to others.

I am enclosing a donation that hopefully you can use in some manner to help educate "entry level" medical personnel (ERs, MD offices etc) to recognize this serious but manageable condition.

Thank you for being there for us!

Dennis A. Waypa, Dayton, Ohio

GBS Survivors and CIDP Patients at the May 1, 2010 GBS/CIDP Foundation 5K Run and Walk — Charlotte, NC.

GBS/CIDP *Foundation International*
Serving patients of GBS, CIDP and Variants with support, education and research
Guillain-Barré Syndrome • Chronic Inflammatory Demyelinating Polyneuropathy

International Office
The Holly Building
104 ½ Forrest Avenue
Narberth, PA 19072

CHANGE SERVICE REQUESTED

DIRECTORY

Check the enclosed chapter directory and contact the chapter nearest you. In addition, our "subgroups" are listed below.

- **"CIDP" Group**
  For those having diagnosis of chronic inflammatory demyelinating poly-neuropathy. Please identify yourself to the National Office in order to be put in contact with others around the country.
- **Children with GBS**
  Lisa Butler • 670 Penllyn Blue Bell Pike, Blue Bell, PA 19422
  215-628-2771
  Son, Stuart had GBS at 5½ years old
- **Children with "CIDP"**
  For those children diagnosed with chronic inflammatory demyelinating polyneuropathy. A separate registry has been created. Please contact the National Office for details.
- **Pen Pals (for all ages)**
  Faith & Jeffrey Aronsky, GBS Pen Pal Coordinators
  P.O. Box 802954, Santa Clarita, CA 91380
- **Group for Having GBS Two Separate Times**
  Please call the National Office for contact with others.
- **Miller Fisher Variant Group**
  Please call the National Office for contact with others.
- **Wheelchair Limited Group**
  Please call the National Office for contact with others.
- **AMSAN Group**
  Please call the National Office for contact with others.
- **A Teenage Pen Pal Group**
  Arielle Challander • 4313 Shawn Drive, Traverse City, MI 49684
  231-946-7256
  E-mail: GBSTeenPenPal@hotmail.com
  Arielle had GBS in 2006 at age 13. She is willing to share experiences that others might not understand. To have a teenage GBS'er pen pal, write, call or e-mail to Arielle.
- **Pregnant Women with GBS**
  Robin Busch • 264 Oenoke Ridge, New Canaan, CT 06840
  203-972-2744
  Robin has offered to share her experience with GBS which came about during her pregnancy. We have many such cases and reassurance from someone who has gone through this is needed support.
- **Bereavement Group**
  A group for anyone who has lost a loved one due to GBS/complications. Please contact: Bereavement Group at the National Office.
- **The "Campy" Group**
  Those whose GBS onset was identified as a result of the campylobacter bacteria. Numbers to be used for research purposes.