Notes from 2008 GBS/CIDP Colorado Statewide/Regional Meeting
St. Anthony Central Hospital, Birch Room
May 3, 2008

About 27 people consisting of GBS/CIDP patients and their families from Nebraska, Fort Collins, and the greater Denver area came together to discuss, question, and provide perspective on all issues concerning GBS and CIDP. Coffee, hot and cold tea, sandwiches and a very special cake were served (thank you Maureen and Joy). Registration opened at 12:30PM and Jeff began an excellent presentation shortly after 1PM and concluded around 2:30PM.

Ann Brandt began the meeting with some news of members who couldn't attend and some needs and desires of our organization. There is a need to distribute information on GBS/CIDP to hospitals. There is also a need for someone to assist in writing cards to recent GBS/CIDP patients around the country. Contact Ann Brandt if interested.

Jeffrey Raff, Ph.D., is the author of 4 books and has been in private practice in Colorado since 1976, is a certified Jungian Analyst, and is a member of our group. The following are a combination of notes from Jeff's presentation (with some additions and references from the webmaster):

I should say that I can only speak about residuals and their management from the perspective of one who has had GBS (May 2005). People with CIDP will have to see if what I say has relevance for their situation. Hopefully, when we have our discussion, CIDP people will speak about that.

Residuals were, until very recently, the surprise package that came along with GBS. No doctor, the whole time I was in the hospital and in rehab, ever mentioned them or said anything but that I could expect a complete recovery. Only my physical therapist in the hospital hinted that I would need a cane for a very long time. When I got out of rehab there was no provision for me to continue physical therapy and I was told by the physical therapists that there was nothing they could do for me. My neurologist finally acknowledged the existence of residuals but also informed me that there was nothing he could do for me and there was no need for me to see him anymore. I felt adrift and very lost because my residuals were, and are, serious and I didn't know what was happening to me. Fortunately, I discovered this support group and the support groups online where people shared their experience about residuals and I learned how serious they could be. Because I was a writer and researcher I decided to study residuals for myself and see what I could learn. In that process I also found a doctor of rehabilitation medicine who worked with me on how to live my life within the boundaries created by residuals.

There is now a theory about why residuals come about and seems, though not fully accepted, to be the best model there is. I won't go into the gory details but we all know that GB S is a disease that inflicts damage to the myelin sheath and, in cases of severe paralysis, to the axonal level as well. In the majority of cases this nerve damage heals and a person is able to walk and resume all normal activities. However, it seems that nerves may actually not fully heal. While they heal enough to allow walking, some of the nerves may not heal, so that the nerves that have healed have to work harder than in normal cases. Whereas before GBS you had 10 healthy nerves working away, after GBS you may have 4 healthy nerves doing the work of ten. The result is that they

get tired. If you have 8 out of 10 nerves that recover you may get less tired, but if have only 1 nerve doing the work of 10 you will get more tired. Because nerves heal slowly, and axonal nerves heal extremely slowly, this condition may last years. Age and amount of paralysis, speed of onset, and time of beginning of treatment are factors that can affect the cumulative amount of nerve damage.

So residuals come from the damage done to nerves and because doctors in the past defined full recovery as the ability to walk they were right according to their definition that most people were fully recovered. Only in the last few years has research begun that shows almost all people with GBS experience residuals to one degree or another. In one study only half the people interviewed indicated that they experienced residuals to the point that they interfered with normal life. In some cases 80% of the people interviewed indicated that they suffered from residual fatigue. So you are not alone. The other unfortunate fact revealed by research is that residuals may come at any time after the disease. People who indicated that they had no residuals immediately after the disease may come to experience them years later. It seems possible that they healed nerves in these cases get fatigues over years of having to work too hard. There is also some debate about what is called the post-polio syndrome in which nerves previously healed become damaged again as aging continues. Alternatively, because nerves can heal over many years there is hope that residuals will go away in time-especially for the young. As with some many aspects of GBS there is no way to predict results for a particular individual. However, it has been said that certain factors mitigate having more problems with residuals. These are age: the younger the patient, the greater the chances of more rapid or complete nerve healing while the closer to 60, the less rapid and less complete healing of nerves. The more rapid the onset of the disease the more likely fatigue and other residuals are, the degree and severity of paralysis with the more serious paralysis indicating more residual problems. And finally whether one needed a ventilator or not, with those needing a vent having more residual problems. However, it is also important to say that these are not binding laws but guidelines. It has been clearly proven, for example, that even mild cases of GBS with little or no paralysis can leave fatigue as a severe problem.

So what are the most common residuals people must deal with. These are fatigue, fatigability, pain, numbness and tingling, sexual dysfunction, depression and memory or cognitive problems.

Coping with GBS/CIDP Residuals:

General Guidelines:
1) Get the right doctor. The right doctor is one who listens and gives treatment and medication. A good doctor works with the patient.
2) Have a good support group – family, friends, GBS support groups
3) Be aware of your own symptoms and be willing to tell others about them – especially your caregiver. Be aware of what causes pain and what helps. You need to communicate about what's going on.
4) Be aware that there is no medical cure for residuals. Be patient because residuals will only gradually go away and in some case they will never go

away. Don't expect too much too soon. Recovery takes time. Be patient with yourself.

5) Denial is tempting but ultimately destructive. So try to avoid denial. It gets in the way of telling people what is going on with you and what you really need. Hope is important but wishful thinking is also not helpful.

6) Find your own meaning of what has happened/is happening to you. Maybe something positive can come out of it. Become stronger mentally.

7) Find your passion. Find your own bliss within your limits. Find something that you are passionate about, that you connect with, and that will take you away from your GBS residuals.

Common Residuals:

Fatigues and fatigability:
1) Know your limits and live within them as much as you can. Don't cross your own line. You can have both physical and mental fatigue.

2) Moderate exercise seems to help. Don't do too much. Do as much as is comfortable, rest, and do it again when you are ready. It seems best not to exercise every day but every other day or every third day. Knowing your own limits is critical.

3) Fatigability is the degree to which you get fatigued during/after exercise. For example, walk for 10 minutes and see how long before you can walk again. The level is all due to the degree of nerve damage.

Pain:
1) Pain relates to how much damage occurred during onset. So don't do what causes you pain.

2) Get the right medication. There are many types work with your doctor to find what is right for you. Don't worry about addiction or dependence – find a way to control the pain and do what it takes. Then worry about possible addiction.

3) Avoid overdoing things that cause fatigue and fatigability and contribute to pain.

4) There are several types of pain post GBS: Muscle cramps, sharp stabbing pain, deep aching pain, and burning pain. Make sure to explain to your doctor what pain you have.

Depression:
1) PTSD from GBS is common. GBS is a traumatic event for the body and as goes the severity of the case, so goes the level of trauma and the importance of therapy, medication, and a support structure including family and groups. Depression can be an aid to fatigue but fatigue can also add to depression as well. In fact, sometimes depression comes from the fatigue and is not true depression.

2) Must deal with depression. Coping with depression is achievable using the right tools (antidepressants).

Sexual Dysfunction:
1) Can be short termed or long lived. It may get better with time.
2) Usually in the form of a lack of sensation for women and erectile dysfunction for men. There are currently many medications for erectile dysfunction. Get help if you need it.

Memory and Cognitive Issues:
1) Have concentration issues.
2) Many doctors deny it, but recent studies have shown hallucinations and dream disturbances in as much as one-third of GBS cases. Research is currently being conducted in this area.

Heart Disorders:
May have dysautonomia - heart palpitations, shortness of breath, and light headedness. Close relative of postural orthostatic tachycardia syndrome (POTS).

The presentation ended with questions and more discussion. Refreshments followed.

The next meeting of the Colorado GBS/CIDP Support group will be on the first weekend in October and the annual social potluck meeting will be held the first weekend in December.

Several references were found on the WWW related to GBS residuals and GBS cognitive issues. They are included here.

TITLE:	Residual Problems Following Recovery from Guillain-Barre′ Syndrome: Do Depression and Anxiety Play a Part?
TOPIC AREA:	Psychology
KEY WORDS:	Guillain-Barre′ Syndrome; depression; anxiety
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Guillain-Barre′ Syndrome (GBS) is a relatively rare disorder affecting 1 to 2 people per 100,000 per year. Steinberg (2000) describes GBS as being "typified by the rapid onset of weakness, and even paralysis, often accompanied by abnormal sensations. These changes reflect damage of the peripheral nerves of the body, those outside the brain and spinal cord." (p.3). The cause of GBS is unknown although a number of events may trigger it; for instance, the common cold, bacterial infections, surgery, and insect stings (Steinberg, 2000). The illness can be fatal, although the vast majority of sufferers do recover (Lennon, Koblar, Hughes, Goeller, & Riser, 1993). For many, the recovery is not complete; a significant number are left with varying residual problems. For example, Meythaler, De Vivo, and Braswell (1997) estimate that at any one time between 25,000 and 50,000 individuals in the USA have some form of long term disability secondary to GBS. These include muscle weakness (especially limb muscles), abnormal sensations, shortness of breath, pain, and extreme fatigue.

Most GBS research has focussed on its medical and clinical aspects, such as its causes and treatment options. Research into the psychosocial impact of the disorder is scarce. Nonetheless, it is commonly reported by GBS sufferers that they experience severe anxiety and depression at the peak of their illness, and for several weeks afterwards during the initial phase of recovery (Renaud, 2000). Our concern was with those sufferers who never fully recover, those who are left with residual problems. Does living with the knowledge that one may permanently suffer pain, fatigue, or abnormal sensations affect one's psychological state? The purpose of the present study[1] was to examine the role played by depression and anxiety in those sufferers experiencing long-term residual problems. Participants provided information about their residual problems including their severity, and then completed a questionnaire that was used to assess their levels of anxiety, depression, and overall quality of life.

METHOD

Participants
Potential participants were contacted through the New Zealand GBS Support Group. This group has a membership in excess of 250, but only 49 members volunteered to take part, and of these, 44 completed and returned the questionnaire. No attempt was made to find out what proportion of members suffered from one or more residual effects. Thus, it is not known whether our 44 respondents were representative of this group as a whole. Participants' ages ranged from 32 – 82 years (**M** = 62.0, **SD** = 12.4).

Measures
GBS Questionnaire. This questionnaire, specifically designed for the present study, provided biographical details, information on the nature and timing of the illness, the type of treatment, the length of time in hospital, the severity of the diagnosis, and past and present medications. In addition, information was requested for residual effects and their severity, and there was a range of general questions asking about the GBS experience and the recovery process.

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[1] The data reported here are taken from the first author's Masters thesis (Bourke, 2001).

The McMaster Health Index Questionnaire (MHIQ). This questionnaire was used because it focussed on emotional as well as physical aspects of the illness. Further, it is a relatively simple questionnaire to complete with reasonable reliability and validity scores (Bowling, 1997; Chambers, 1993).

Beck Depression Inventory–2 (BDI-2). The BDI-2 is a frequently used test for assessing depression in adolescents 13 years and over and adults. It has been extensively normed and validated and takes only about 10 minutes to complete (Beck, Steer, & Brown, 1996; Dozios, Dobson, & Ahnberg, 1998).

Six-item State Trait Anxiety Inventory (STAI-6). As for the BDI-2, the STAI-6 is well normed and validated. It has been in use for more than 30 years (Spielberger, 1989). The STAI-6 takes only 5 – 10 minutes to complete and gives results that are highly correlated with the full-scale (40 item) test (Marteau & Baker, 1992).

Procedure
Questionnaires were delivered and returned by mail via the NZ GBS Support Group secretariat. Each questionnaire was coded to preserve participant anonymity.

All aspects of this study were scrutinised and approved by the Massey University Human Ethics Committee.

RESULTS AND DISCUSSION

In the present sample, the mean age of onset of GBS was 54 years. Sixty four percent of the sample was between the ages of 60 and 79 years when the survey was completed. Mean age since diagnosis was 8 years with almost two thirds of the sample receiving a diagnosis of severe GBS. All bar 2 respondents were hospitalised with just under half staying in hospital for 3 or more months.

Table 1: Number of participants who report common residual problems following GBS.

Residual	Mild	Moderate	Severe	<u>n</u>	%
Fatigue	38.6	50.0	4.5	41	93.2
Pain	34.1	22.7	9.1	29	65.9
Reduced Mobility	22.7	29.5	13.6	34	77.3
Muscle Weakness	34.1	34.1	15.9	37	84.1
Limb Weakness	29.5	34.1	15.9	35	79.5
Numbness	34.1	29.5	2.3	29	65.9
Nerve Tingling	36.4	25.0	9.1	31	70.5

Table 1 shows the number of participants reporting the 7 most common residual problems. Forty-one of the 44 respondents (93%) reported fatigue as a residual problem with 50% reporting the level as moderate. This finding, that fatigue is a very

common residual problem, is consistent with previous research (e.g., Bernsen, Jacobs, de Jager, & van der Meche, 1997). Reduced mobility (77%), muscle weakness (84%), and limb weakness (80%) were also commonly reported residual problems. It is notable that two thirds of the sample reported some degree of pain. All 7 of the residual problems shown in Table 1 were experienced by 16 (39%) of the participants. Sixteen further residual effects were reported, each by between 1 and 3 participants (e.g., reduced coordination, burning sensations). All of the residuals reported are consistent with peripheral nerve damage.

Winer, Greenwood, Hughes, Perkin, and Healy (1985) estimate that approximately 16% of GBS patients are left with residual deficits such as those reported above. The high rate of reporting in the present study may be due to the fact that most people volunteered for the study only if they had residual problems.

It is possible that the length of time since diagnosis interacted with the number of residual problems reported. However, as Table 2 shows, this does not appear to be the case. Splitting the sample into those diagnosed within the last 6 years and those diagnosed more than 6 years ago revealed little or no difference in reporting rates. A similar analysis showed that the severity of each residual problem was also unrelated to the time since diagnosis. Previous research has also found that residual effects, such as pain, do not decrease with time (Tempest-Roe, 2000).

Table 2:   Number of participants who reported 7 most common residual problems as a function of time since diagnosis.

	Group 1 (%) (≤ 6 years)	Group 2 (%) (> 6 years)	n
Residual Problem			
Fatigue	51.2	48.8	41
Pain	51.8	48.2	29
Mobility	50.0	50.0	34
Muscle Weakness	48.6	51.4	37
Limb Weakness	48.6	51.4	35
Numbness	48.3	51.7	29
Tingling	48.4	51.6	31

Quality of Life: MIHQ. The MIHQ consists of 3 subscales – physical, emotional, and social functioning. Participants completed these subscales twice, once for the time when GBS was at its worst (Time 1) and again for the time at which the survey was completed (Time 2). Overall quality of life was within the normal range at both Times 1 and 2 for all 3 subscales, with the exception that the physical functioning subscale was abnormally low when GBS was at its worst (Time 1). The fact that physical functioning dramatically improved between Times 1 and 2 is probably explained by the fact, that during the acute phase of the syndrome, most sufferers reported being 'completely paralysed'. As the paralysis subsided during the first few weeks of recovery, improvement in physical functioning would have been great. There were

also statistically significant (p<.001) improvements in social and emotional functioning between Times 1 and 2 even though functioning appeared to be within the normal range at both times.

Depression: BDI-2. A total of 37 of 43 participant scores (86%) fell within the 'minimal depression' range on the BDI-2. Five participants were mildly depressed and one moderately so. Therefore, our results do not support the hypothesis that GBS sufferers with residual problems are generally depressed. There was no relationship between level of depression and the degree of severity of each of the 7 most common residual problems. However, the *number* of residual problems did correlate with depression scores (r = .28, p = .06); the more residual deficits suffered the greater the depression. It must be noted, though, that this relationship is only weak and needs to be treated with caution. Nonetheless, more research is required to examine this possible link between depression and number of residual problems.

Anxiety: STAI-6. Possible scores on the STAI-6 range from 6 to 24. The overall mean in the present sample was just 9.7 (**SD** = 3.6), well within the normal range. Therefore, as for depression, we found no support for the hypothesis that long-term residual problems following GBS will be accompanied by high levels of anxiety. Furthermore, none of the participants were being treated for anxiety and none were on anxiolytics. Additionally, we found no relationship between anxiety and type of treatment, anxiety and severity of diagnosis, or anxiety and severity or number of residual symptoms. In sum, our participants seemed to be a remarkably anxiety-free group! One possible explanation is that current anxiety levels may have seemed relatively very low when compared to the levels experienced at the peak of the illness. Until further research is conducted our explanation must be seen as only tentative.

CONCLUSION

In general, our results demonstrate that depression and anxiety are uncommon sequelae to GBS, even when the sufferer may be experiencing a wide range of residual problems. This conclusion must be seen as tentative at the present time for at least three reasons. First, the sample was small and self-selected. We have no way of knowing how representative it is of New Zealand GBS sufferers, let alone those in other countries. Second, the study was retrospective in nature. Participants were asked to recall events and psychological states from the past, often many years ago. Smith, Leffingwell, and Ptacek (1999) found that people tend to overestimate their daily coping ability when required retrospectively to recall events. Because of the very small numbers of GBS sufferers in New Zealand at any one time, running a prospective study in New Zealand alone with a large enough sample may not be possible. Third, Renaud (2000) reported that psychological distress (depression, anxiety) may be more frequently reported earlier in the recovery phase than at the time our participants responded. Future studies will need to look into how psychological distress varies across the recovery period as well as at the time when participants have realised that the residual effects they suffer from are probably permanent effects.

REFERENCES

Beck, A.T., Steer, R.A., & Brown, G.K. (1996). *BDI-II manual.* The Psychological Corporation. San Antonio: Harcourt Brace.

Bernsen, R.A., Jacobs, H.M., de Jager, A.E., & van der Meche, F.G. (1997). Residual health status after Guillain-Barre′ Syndrome. *Journal of Neurology, Neurosurgery, and Psychiatry, 62,* 637-640.

Bourke, C. (2001). *Relationships between depression, anxiety, and residual problems following recovery from Guillain-Barre′Syndrome: A New Zealand survey.* Unpublished Masters thesis, School of Psychology, Massey University, Palmerston North, New Zealand.

Bowling, A. (1997). *Measuring health: A review of quality of life measuring scales (*2nd ed.). Buckingham: Open University Press.

Chambers, L.W. (1993). The McMaster Health Index Questionnaire: An update. In S.R. Walker, & R.M. Rosser (Eds.), *Quality of life assessment: Key issues in the 1990s.* (pp. 131-149). London: Kluwer Academic Publishers.

Dozois, D.J., Dobson, K.S., & Ahnberg, J.L. (1998). A psychometric evaluation of the Beck Depression Inventory-II. *Psychological Assessment, 10,* 83-89.

Lennon, S.M., Koblar, S., Hughes, R.A., Goeller, J., & Riser, A.C. (1993). Reasons for persistent disability in Guillain-Barre′ Syndrome. *Clinical Rehabilitation, 7,* 1-8.

Marteau, T.M., & Bekker, H. (1992). The development of a six-item short-form of the state scale of the Spielberger State-Trait Anxiety Inventory (STAI). *British Journal of Clinical Psychology, 31,* 301-306.

Meythaler, J.M., De Vivi, M.J., & Braswell, W.C. (1997). Rehabilitation outcomes of patients who have developed Guillain-Barre′ Syndrome. *American* Journal *of Physical Medicine and Rehabilitation, 76,* 411-419.

Renaud, A. (2000). *People with Guillain-Barre′Syndrome: Biopsychosocial impact. (A first New Zealand survey.)* Unpublished Masters thesis, School of Psychology, Massey University, Palmerston North, New Zealand.

Smith, R.E., Leffingwell, T.R., & Ptacek, J.T. (1999). Can people remember how they coped? Factors associated with discordance between same-day and retrospective reports. *Journal of Personality and Social Psychology, 76,* 1050-1061.

Spielberger, C.D. (1989). *State-Trait Anxiety Inventory: A comprehensive bibliography* (2nd ed.). CA: Consultant Psychologists Press.

Steinberg, J. (2000). *Guillain-Barre′Syndrome (Acute Inflammatory Polyneuropathy): An overview for the layperson* (9th ed.). Wynnewood, PA: Guillain-Barre′ Syndrome Foundation International.

Tempest‑Roe, J. (2000). *Information for patients recovering from Guillain‑Barre' Syndrome.* GBS support group of the UK.

Winer, J.B., Greenwood, R.J., Hughes, R.A., Perkin, G.D., & Healy, M.J. (1985). Prognosis in Guillain‑Barre' Syndrome. *The Lancet, 1,* 1202‑1203.
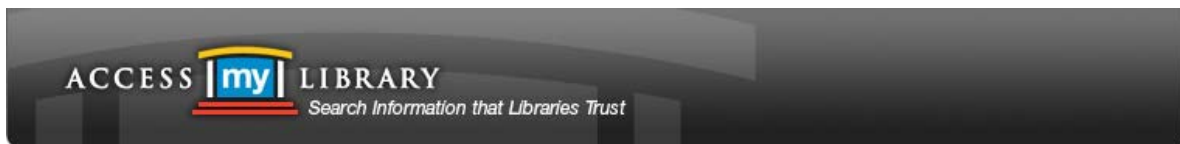
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Understanding residuals in Guillain-Barre Syndrome: peripheral nerve damage, rather than strained muscles, can cause fatigue and breathlessness for those suffering the long-term effects of GBS.(practice)

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GUILLAIN-BARRE SYNDROME (GBS) is a fascinating neurological disease, if such there can be, for it is rare, occurs with sudden onset, and creates acute effects, particularly on the peripheral nervous system.

Essentially, the peripheral nerves are attacked by the body's defence system, an auto-immune attack, and as a result, the myelin sheath and axons of nerves are impaired. When sufficient damage occurs, paralysis may result. Neurological examinations, intensive hospital care and plasmapheresis treatments lead most people with a severe case back to recovery and rehabilitation in a few weeks or months. While the onset and collapse may be dramatic, the ensuing years are dull in comparison. However, the nature of the disease can hide both the long-term physical effects and the psychological reactions.

The residual effects of GBS have been noted by American physician Joel Steinberg who stated: "Well over 50 percent, and probably up to 90 percent, of patients eventually reach complete or nearly complete recovery and resume their prior lifestyles. About five to 15 percent of patients will have significant long-term disability. Perhaps 35 percent will experience long-term mild abnormalities, such as foot drop or numbness." (1) Steinberg also commented that patients might develop fatigue, particularly with sustained activity, and demonstrate poor endurance, even with normal muscle strength. This could lead to serious problems for those working long hours and/or with physically demanding jobs.

American neurologist Gareth Parry, who has spent some years working at Auckland Hospital, added to our understanding with a thoughtful analysis of what appears to happen physiologically. (2,3) Based on recent research, Parry stated teat a study of 83 patients found 80 percent experienced severe fatigue that interfered with their life. Also and significantly, these patients experienced fatigue that did not seem to decrease over time. Meanwhile, he noted that these people had relatively normal strength. "The basis ... is probably axonal degeneration."...

Residual Effects
Following
Guillain-Barre'

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Gareth J. Parry
Consultant Neurologist Auckland Hospital, Professor of Neurology University of Minnesota

Guillain-Barre syndrome is a disorder whose excellent prognosis is invariably emphasized. Widely accepted figures suggest that 75%-85% of patients make a complete recovery, however, many of my patients have complained to me of minor but annoying persistent symptoms continuing for years after the initial paralytic event. Although I have made no systematic study of the proportion of patients with these residual complaints it is certainly more than the 15%-25% that the figures in the literature would suggest. The great majority of studies of GBS outcome are based on telephone interviews or retrospective chart reviews and seemingly minor complaints may have been missed or disregarded. Thus, patients are often asked if they have returned to their previous work or other previous activities but they may not have been asked whether they have more difficulty performing their former activities. A note of caution was sounded in one small study from Dr. J. McLeod and his colleagues in Australia who objectively evaluated a small group of 18 recovered GBS patients and found that half of them had residual neurological abnormalities. Even then the residual abnormalities were considered to be significant in only four patients. A recent important paper from Dr. I.S.J. Merkies and colleagues in Holland (Neurology 1999; 53:1648-1654) has established that residual effects from both GBS and CIDP are much more common than has been generally reported and that seemingly minor neurologic abnormalities may still result in annoying disabilities. The study used a validated index of fatigue severity to assess residual disability. It included 83 patients who had suffered from GBS an average of five years previously. About 80% of these patients experienced fatigue that was considered severe enough to interfere with their life despite the fact that the majority had normal strength or only minor weakness. They noted also that the fatigue did not seem to improve over time; the fatigue index score was the same in patients in whom many years had elapsed as it was in patients whose acute illness had occurred only 6-12 months previously. This paper provides sound scientific support for the validity of the observations of my patients who regularly complain of fatigue even when they have returned to all or most of their former activities and who are working full time at their former jobs. Although their strength may be normal when they are examined in the doctor's office they are clearly unable to sustain the same level of physical activity that they had performed prior to their GBS. A second under-appreciated symptom that may persist for many years is pain. Certainly, severe disabling pain is very rare. However, a number of my patients complain of persistent discomfort in their feet. The discomfort may take the form of annoying paresthesias (tingling) or there may be a vague aching discomfort. The symptoms have the same characteristics as typical neuropathic pain in that they tend to be worse in the evening or at night and are particularly annoying following days during which the patients have been up on their feet a lot. The discomfort is not particularly responsive to analgesics but usually does respond to drugs such as gabapentin or amitriptyline, drugs typically used in the treatment of neuropathic pain. However, these medications have to be taken on a daily basis to be effective and one problem with deciding whether to treat this residual symptom is that the discomfort is usually rather mild. Thus, patients may be daily irritated by their symptom but be reluctant to take a drug every day for a symptom that significantly bothers them only once or twice a month. I have seen no mention in the medical literature of this phenomenon. It is possible that I see a highly selected group of patients in my practice who had initially been more severely affected and that the prevalence of this annoying residual symptom is much higher in my patients than in the usual population of recovered GBS patients. I would be most interested to learn whether the group of patients reported by Merkies and colleagues also suffered from minor persistent discomfort.

The basis for both of these seemingly minor residual symptoms (fatigue and pain) is probably axonal degeneration. During the acute illness the predominant underlying pathology in most patients is segmental demyelination, a completely reversible phenomenon. However, some degree of axonal degeneration is almost invariable. As recovery occurs function is restored by a number of mechanisms. Axonal regeneration of motor axons probably plays very little role in restoration of function except in the more severe cases. Rather, surviving axons send out small branches called collateral sprouts that restore the nerve supply to those muscle fibers whose nerves have been damaged. This process of collateral sprouting is very effective at restoring strength to a muscle but the efficiency of the muscle suffers - the muscle must work harder to achieve its goals. Thus, fatigue may result even when there appears to be full restoration of strength. On the sensory side, even a small number of damaged sensory axons may be sufficient to generate spontaneous discharges that are registered as pain or discomfort.

It is entirely appropriate that the good outcome of GBS should be emphasized during the acute illness. During this time,

the patient is losing control of many motor functions, sometimes including life preserving functions, and constant reassurance from the attending physicians plays a vital role in the recovery process. However, it is equally important to be aware that residual problems are experienced by "recovered" GBS patients. Acknowledgement that such residual problems exist will go a long way towards helping patients deal with the frustration of their incomplete recovery.

More research is needed to discover an effective treatment for the residual fatigue. In addition, since these persistent symptoms are probably related to the degree of axonal damage that occurs at the time of the initial attack, we also need to continue to strive for earlier and more effective treatment of the acute stage of the disease so that these residual problems are minimized.

Article from the Spring 2000 GBS Newsletter

Disability After "Recovery" From GBS

**What's In a Name? Important Differences
Between GBS, CIDP and Related Disorders**

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Disability After "Recovery" From GBS

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Guillain-Barre syndrome (GBS) is the most common cause of acute neuromuscular paralysis in developed countries. Most patients recover and return to productive, independent lives. In a recent representative survey of 140 GBS patients, 70% made a complete neurological recovery within a year, 22% could walk but were unable to run, 8% were unable to walk unaided, and 2% remained bedridden or ventilator-dependent after a year. Thus, despite the good prognosis for recovery, GBS can cause long-term disability. Persisting disability is largely the result of weakness from the motor nerve injury that occurred during the acute illness. An estimated 25,000 to 50,000 persons in the United States alone are experiencing residual effects from the disease. Most research on GBS has focused on understanding the cause and finding better treatments. Much less attention has been paid to the long-term disability caused by GBS. In addition to the previously mentioned residual weakness, there may be pain, fatigue, psychosocial dysfunction, possible relapses of the illness, and late progression of weakness.

Pain

Moderate to severe pain is a well-recognized symptom during the course of acute GBS. For some patients neuropathic pain, consisting of abnormal painful sensations, may persist after recovery from the disease. In a recent prospective study of 55 GBS patients followed for up to 24 weeks, pain occurred during the course of the illness in almost 90% of cases. Whereas deep aching back and leg pain were the most common early on, abnormal painful sensations and myalgic-rheumatic type pain were observed during the recovery period. Musculoskeletal pain was common in association with physiotherapy. Painful abnormal sensations in the extremities tended to persist after 8 weeks, and were still present in some patients after 24 weeks. In two cases the pain was severe. Overall, pain can be effectively relieved with an escalating regimen of analgesic medications, starting with nonsteroidal anti-inflammatory drugs or acetaminophen, and if necessary including oral or parenteral opioids. Even severe pain can be controlled, sometimes with the addition of patient-controlled analgesia. In a large series of GBS patients treated for pain, these medications were generally effective, and no adverse effects on breathing function or narcotic addiction occurred.

Chronic fatigue

Fatigue following GBS is underrecognized by neurologists and rehabilitation physicians, because attention is directed toward the more objective weakness and sensory disturbances. In a recent study of 83 patients recovering from GBS, severe fatigue was reported as one of the three most disabling symptoms by over 80%. The incidence of fatigue did not correlate with age, or motor and sensory residual deficits, but fatigue was more common in women. Fatigue was unrelated to the time since the acute phase of the GBS, a median of 5.2 years in this group. Another study of 123 GBS patients, evaluated 3 to 6 years after the acute illness, concluded that psychosocial functioning, especially in areas such as sleep and rest, alertness, emotional behavior and social interaction, was seriously affected. This was true even when "complete" physical recovery was reached, or only minimal residual deficits were present. Deconditioning and less engagement in physical activities were discussed as possible explanations for persistent fatigue. A supervised training program and low-intensity aerobic exercise may reduce daily fatigue, with improvements in activities of daily living and functional capacity. Specific treatments for other factors associated with fatigue, such as sleep disturbances, pain, and daytime inactivity, are available.

Psychosocial dysfunction

Reports of long-term psychological sequela after GBS are rare, although this issue may be a major factor in psychosocial dysfunction of patients recovering from the disease. Many psychological factors could contribute to chronic fatigue and social dysfunction, including fear of disability, inability to cope with physical limitations, and depression following a major illness. The role of depression in psychosocial dysfunction after GBS is not fully understood. The sickness impact profile of GBS survivors was found to differ from the profile of other patients with depression. Nevertheless, further study of the long-term psychological impact of the disease is necessary, and depression should be considered on an individual basis when appropriate. Both supportive psychotherapy and/or pharmacologic treatment can be effective.

Post-traumatic stress disorder (PTSD) has been reported in a patient following severe GBS with paralysis and a prolonged intensive care stay. The GBS-induced PTSD shared the features of PTSD seen following other traumatic events. Even such profound psychological problems following GBS can be treated with supportive psychotherapy and appropriate medications. They may at least in part be prevented by adequate pain management and the use of a communication system, such as clear lucid letter-board in the event of near complete paralysis. Better understanding, prevention and treatment of these issues may have a positive impact on the quality of life for GBS survivors. Moreover, it is important for patients and their families to know that their psychosocial problems are also experienced by other patients after GBS.

Recurrence of GBS

Although GBS is thought to be a one-time disease, relapses and chronic recurrent forms can occur. Patients are often concerned about the risk of having additional episodes of GBS. In a study of 220 GBS patients, 15 were found to have a relapsing course, with one to 4 recurrent episodes. The interval between episodes ranged from 3 months to 25 years. Antecedent events such as a viral infection preceded most relapses, and patients presented each lime with the typical clinical and laboratory findings of acute GBS. All patients had long asymptomatic periods between the episodes. In a more recent study of 476 patients following GBS, 2.5% experienced a recurrence of the acute illness, with a mean period of 16 months between the episodes (range 2-47 months). One patient had three episodes. The authors found no relationship between the risk of having a recurrent episode and the severity of the first episode. Furthermore, the severity of the subsequent episode did not correlate with the intensity of the first episode. Reaching a correct diagnosis may be challenging in these cases. Even GBS experts may find it difficult to separate a "relapsing variant of GBS" from chronic inflammatory demyelinating polyneuropathy (CIDP), especially early in the course. Recurrent episodes of true GBS, although rare, may occur following similar preceding illnesses, and should be treated in the same way as the initial episode. They respond well to the same established treatment modalities.

Delayed progression

Weakness from GBS reaches its maximum during the first two or three weeks of the disease. This is the active or acute phase of the illness. After a plateau period of days or weeks, recovery begins, lasting between weeks and two years. During this time strength improves steadily. Strength and sensory function plateau after about two years. However, many decades after GBS, recovered muscles once weakened by the disease may again grow weak. This is a slow process that occurs over years, and may at first escape the patient's notice. It is likely that this delayed weakness is the effect of the normal gradual age-related nerve cell loss on muscles that have a reduced reserve nerve supply from earlier GBS. The same phenomenon has been observed after poliomyelitis ("post-polio syndrome") and other forms of acute nerve injury. The incidence of slowly progressive late weakness in GBS is unknown, but it is rare. When it does occur, the patient's physician must recognize that the new weakness of seemingly recovered muscles does not necessarily indicate a second attack of GBS.

For many patients recovering from GBS, residual motor or sensory deficits may be only one aspect of the long consequences of the disease. Other issues described here may have a considerable impact on their quality of life. Effective treatments are available for most of these problems.

References available on request.
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Article from the Spring 2000 GBS Newsletter

- The vexed question of residuals in Guillain Barre Syndrome -

Lawrence Kaplan and Robert J. Gregory

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Numbers in parentheses: Please refer to the reference list at the bottom.

Guillain Barre Syndrome (GBS) is a fascinating neurological disease, if such there can be, for it is rare, occurs with sudden onset, and creates acute effects, particularly on the peripheral nervous system.
Essentially, the peripheral nerves are attacked by the body's defence system, an auto-immune attack, and as a result, the myelin sheath and axons of nerves are impaired. When sufficient damage occurs, paralysis may result. Neurological examinations, intensive hospital care and plasmapheresis treatments lead most people with a severe case back to recovery and rehabilitation in a few weeks or months. Dramatic as the onset and collapse may be, the ensuing years are dull in comparison. However, the nature of the disease can hide both the long-term physical effects and the psychological reactions.

The residual effects of GBS have been noted for some time.(1) American researcher John Steinberg stated that, "Well over 50 percent, and probably up to 90 percent, of patients eventually reach complete or nearly complete recovery and resume their prior lifestyles. About five to 15 percent of patients will have significant long-term disability. Perhaps 35 percent will experience long-term mild abnormalities, such as foot drop or numbness."(1) Steinberg also commented that patients might develop fatigue, particularly with sustained activity, and demonstrate poor endurance, even with normal muscle strength. This could lead to serious problems for those who worked long hours and/or had physically demanding jobs.

New Zealand researcher Gareth Parry added to our understanding with a thoughtful analysis of what appears to happen physiologically.(2,3) Based on recent research, Parry stated that a study of 83 patients found 80 percent experienced severe fatigue that interfered with their life. Also and significantly, these patients experienced fatigue that did not seem to decrease over time. Meanwhile, he noted that these people had relatively normal strength. "The basis . . . is probably axonal degeneration."(2) Further, he commented that, "surviving axons send out small branches called collateral sprouts that restore the nerve supply to those muscle fibres whose nerves have been damaged."(3)
Thus while strength to a muscle stays roughly the same, the nerves that are restored are less strong, and so the efficiency of the muscle is reduced, resulting in fatigue. In a recent presentation, Parry noted that "Residual effects from both GBS and CIDP are much more common than has been generally reported."(2)

Recently, New Zealand psychologist Cecilia Bourke noted that 93 percent of her sample of 44 persons who had GBS reported varying residuals.(4) She found that 38 percent were mildly, 50 percent moderately and four percent severely fatigued. Pain was reported by 66 percent, nerve tingling by 70 percent and reduced mobility by 77 percent of the participants. In addition, 84 percent claimed muscular weakness, while numbness was felt by 66 percent. Interestingly, a remarkable 39 percent of the 44 persons

interviewed claimed to experience all seven of these symptomatic problems. This finding is contrary to the frequent assurances that, after initial acute phases of GBS, recovery is total. Given that Bourke found anxiety and depression were within a normal range as measured by psychological testing, the large number complaining of physical residual effects was surprising.

What exactly is taking place?

What may take place is that the myelin sheath in nerves and the axons themselves are damaged from GBS. Some of those wounds recover, heal and the person then gets on with their life. Some of the damage, however, does not heal, in particular the damage in axons. What may occur then is that relatively weak collateral nerves take over the transmission duties for nervous system messages. These alternative circuits through the nervous system have to do extra duty to replace the functions of the axons of nerves that no longer work well. Those collateral nervous circuits are simply not as strong or as resilient, and are simply not so capable as the originals.

Therefore, when a person with GBS-damaged axons and nerve tissue exercises, these collateral nerves are rapidly overloaded, and slow or even stop functioning fairly quickly. The person comes to a screeching halt -- a neurologically induced crash. Others may look at the person and say, "You are tired and exhausted and fatigued," thinking that it is muscles and overloaded muscles that will recover easily with rest. However, it is not the muscles that are faulty; it is nerves that are limiting functioning abilities. Thus there are significant and real differences in the cause and consequences of fatigue. Even those with GBS may believe they have tired muscles, for muscular tiredness is a common experience. That does not seem to be the case, however. The nerves just can't handle the extra exertion, and when stressed, they do not recover as quickly as muscles do. Tests for muscular strength show up just fine, for the muscles do work and are possibly or even probably stronger than in other people. But the nerves are rarely, or not tested, or suspected.

Some of those nerves affected are essential to lung function and breathing and that may account for developing shortness of breath. Even though individuals may experience this effect, they may not be able to explain it to their family, doctor or friends. These people have no experience other than muscular weakness, and therefore, they cannot understand that there are differences in cause and effects. That may be one reason why those who have had GBS are rather unique!

Case studies

Case A: David, aged about 50, was carrying an extremely heavy and stressful workload. One day, he noted tingling in his feet, then peripheral neuropathy. His physician claimed, "She'll be right," in typical Kiwi optimistic fashion, and David carried on. With some walking and breathing difficulties, acute leg pains, and generalised nervous aches and pains, David continued to work, albeit at a reduced and less effective pace. He noted that he had virtually no knee reflexes, that he slept 14 hours a night and was still exhausted, and endured a variety of "system problems," such as diverticulitis, thyroid failure, and depression. A visit to a different physician led to laboratory and neurological tests, and a diagnosis of GBS, nearly a year later. David began thyroxine treatment, but there was nothing that could be done at that late date for the GBS, nor for continuing symptoms.

For the next 10 years, David continued to have extreme leg pains, generalised weakness, fatigue and tingling sensations that sometimes kept him awake at night. Gradually these aches and pains left, but then reoccurred periodically. The symptoms abated for the most part after 10 years. But, at about 15 years from onset, David noted shortness of breath, a marked lack of endurance even though he retained an ability to handle

tasks that required muscles, fatigue, and the return of some tingling and other painful sensations. Currently he has concerns about the trajectory of the disease and what may lie in his future.

Case B: Philip contracted GBS in 1981. He was ill for two or three weeks with flu-like symptoms, then diarrhoea, and numbness of legs and fingers. Slight paralysis of his legs followed. This progressed without his doctor being concerned, until he became about 30 percent paralysed. It was at this time, the end of the third week, that his medical practitioner hospitalised him. He was promptly diagnosed with GBS. Due to the severity of the disease, and as it rapidly progressed, he spent the next two weeks in cardiac intensive care. Three weeks' nursing care was followed by two weeks' rehabilitation. Plasmapheresis was not an available treatment at that time. At week six, Philip was released from hospital, against the doctor's orders, but he did continue outpatient rehabilitation for another eight weeks.

From 1982 through 1983, Philip gradually increased his work activities to 10 or even 14 hours per day, plus included bicycling and mild sports as part of his continuing self-rehabilitation. He hoped to build greater endurance. From 1984 through 1994, Philip succeeded and led a fairly normal life, but still felt various residual effects, including constrained breathing, frequent and fast onset of fatigue, sudden feelings of complete exhaustion, and tingling sensations between his shoulders. He also noted back and leg soreness, aching knees, and persistent discomfort when laying one knee against the other. Philip also found he had clumsy feet and fingers. He noticed squeakiness in his voice.
From 1995 through 2002, Philip led a very active lifestyle, with reduced impact from the GBS residuals. He worked eight to 12 hours per day, led a sports club, and engaged in physically demanding sports between eight and 20 hours per week. Symptoms such as constrained breathing, fast onset of fatigue and exhaustion, and back and leg soreness were rare. Though he still had tingling sensations between his shoulders, he had no aches in his knees, and only occasional discomfort from knees adjoining or becoming clumsy. He stated that although the residuals had abated significantly, when they occurred, they were intense. Symptoms lingered, and quickly reappeared if and when his activity level decreased.

In 1998, Philip was diagnosed with a heart murmur. In 2000, he had sinusitis and prolonged bouts of upper respiratory infections, requiring surgery. In 2003, the heart murmur led to a repair of the mitral valve, and during his period of rehabilitation, GBS residuals returned in the form of increases in exhaustion, sudden onset of fatigue and constrained breathing. Now aged 48, he hoped for a quick and easy recovery, but that did not happen. Six months later, he felt he had regained only half of his activity levels. He suffered soreness at the incision site, bouts of sudden fatigue, and frequent onset of constrained breathing. An extensive series of diagnostic tests revealed nothing. His medical professionals, knowing little about GBS, let alone about long-term residuals, had no opinion regarding this conclusion and deferred to a diagnosis of "de-conditioning". This ongoing physical incapacity, mixed with the medical professionals' inability to accept the relevance of GBS, brought on depression as well. Ten months following surgery, a significant return of GBS symptoms was evident, including "crashing" and extreme fatigue after even mild exercise. These symptoms collectively were sufficient to be rated as debilitating.

What can be done?
GBS symptoms and residual effects do present a challenge, and are very important to the individuals concerned. Nurses can help in a number of ways. Perhaps the first and most important point is for nurses to be aware that those who have had GBS are few and far between, that most recover and that persistent optimism is valuable.

In addition, however, it is important to really listen to GBS patients,

whether in an acute stage, or throughout the rest of their lives. Their bodies and nervous systems have been affected, and they may well have quite unique problems and issues to face. As with other invisible disabilities, families, friends, neighbours, work mates, and even health care personnel, may ignore complaints. The continuing pains, aches and fatigue that those who have had the disease report are real, and should not be lightly dismissed or ignored. Individualised treatment plans and actions, careful, patient instruction about anatomical and physiological terminology as related to their case to help them communicate and make sense of the unusual sensations and deficits they may encounter, and empathy for these people will all prove helpful.

Further research can be of value to patients, families, and other caregivers, for not enough is known about recovery and rehabilitation from GBS. Certainly the long-term effects are not well understood, and need to be studied.

References

1) Steinberg, J. S. (1998) Guillain-Barre Syndrome: An overview for the layperson. Wynnewood, Pennsylvania: Guillain-Barre Syndrome Foundation International.
2) Parry, Gareth J. (2003a) Residual effects following Guillain-Barre. The Communicator, GBS Newsletter, Spring, 5-6.
3) Parry, Gareth J. (2003b) GBS and CIDP -- what's new? Proceedings: Inaugural Conference of the Guillain-Barre Syndrome Support Group New Zealand Trust, 24-27 April, ed Bob Stothart.
4) Bourke, C. (2003) Psycho-social aspects of GBS. Proceedings: Inaugural Conference of the Guillain-Barre Syndrome Support Group New Zealand Trust, 24-27 April, ed Bob Stothart.

Excerpt from Autonomic Neuropathy

Synonyms, Key Words, and Related Terms: syndrome of acute pandysautonomia, postural orthostatic tachycardia syndrome, POTS, Guillain-Barré syndrome, GBS, acute inflammatory demyelinating polyneuropathy, AIDP, Lambert-Eaton myasthenic syndrome, LEMS, Holmes-Adie syndrome, idiopathic distal small-fiber neuropathy, human immunodeficiency virus, HIV, Chagas disease, Chagas' disease, botulism, chronic idiopathic anhidrosis, familial amyloid polyneuropathy, FAP, diabetes mellitus, uremic neuropathy, hepatic disease, vitamin B-12 deficiency, paraneoplastic autonomic neuropathy, Sjögren syndrome, acute intermittent porphyria, variegate porphyria, hereditary sensory autonomic neuropathy, HSAN, Fabry disease, autonomic dysreflexia, AD, acquired immunodeficiency syndrome, AIDS, autonomic nervous system, ANS, autonomic reflex screen, ARS, composite autonomic scoring scale, CASS, collapsin response–mediator family, CRMP-5, cerebrospinal fluid, CSF, vasopressin, DDAVP, electromyography, EMG, inhibitor of klight polypeptide geneenhancerin Bcells, IKBKAP, mitochondrial neurogastrointestinal encephalomyopathy, MNGIE, M-phase phosphoprotein-1, MPPI, multiple system atrophy, MSA, nerve conduction studies, NCS, progressive autonomic failure, PAF, primary biliary cirrhosis, PBC, Purkinje cell cytoplasmic antibody-2, PCA-2, Parkinson disease, PD, positron emission tomography, PET, peripheral nervous system, PNS, quantitative sudomotor axon reflex test, QSART, quantitative sensory testing, QST, single-photon emission computed tomography, SPECT, serine palmitoyltransferase, SPT, sympathetic skin responses, SSR, selective serotonin reuptake inhibitor, SSRI, thermoregulatory sweat test, TST

Please click here to view the full topic text: [Autonomic Neuropathy]

Background

Autonomic neuropathies are a collection of syndromes and diseases affecting the autonomic neurons, either parasympathetic or sympathetic, or both. Autonomic neuropathies can be hereditary or acquired in nature. Most often, they occur in conjunction with a somatic neuropathy, but they can also occur in isolation.

The autonomic nervous system modulates numerous body functions, and therefore, dysfunction of this system can manifest with numerous clinical phenotypes and various laboratory and electrophysiologic abnormalities. Often, a patient may present with symptoms related to a single segment of the autonomic system. The physician must be wary of other affected parts of the autonomic system.

In some forms, the degree and type of autonomic system involvement varies extensively. In some patients, the degree of autonomic dysfunction may be subclinical or clinically irrelevant, while in others symptoms may be disabling. Several clinically important features of autonomic neuropathies can be treated with either conservative or pharmacologic therapies; therefore, the physician must be alert to these features.

Pathophysiology

The pathophysiology of the autonomic neuropathy depends on the etiology of each particular type. These may range from genetic disorders with specific gene defects to metabolic disorders with accumulation of toxins and to autoimmune disorders with identifiable autoantibodies. Although it is accepted that a loss of somatic C fibers is associated with autonomic deficits, selective involvement is now known to occur for specific autonomic neuropathies. For example, diabetic neuropathies are associated with somatic and autonomic C-fiber impairment, while neuropathic postural tachycardia syndrome is associated with selective distal autonomic deficit (Singer, 2004).

Pandysautonomia

The syndrome of acute pandysautonomia includes both parasympathetic and sympathetic dysfunction (Low, 1983). An immunologic basis for acute pandysautonomia remains most likely, often with onset after a viral illness. Approximately 50% of patients may test positive for an autonomic ganglionic acetylcholine receptor antibody.

Postural orthostatic tachycardia syndrome

Postural orthostatic tachycardia syndrome (POTS) is a syndrome most common in young females with orthostatic intolerance characterized by palpitations with excessive orthostatic sinus tachycardia, sensation of lightheadedness, and near-syncope. POTS may be associated with an infectious prodrome and thus may represent the chronic sequelae of a form fruste of postviral pandysautonomia (Low, 1999). Antibodies against ganglionic receptors are found in 9% of patients with POTS (Novak, 1998).

Guillain-Barré syndrome

Guillain-Barré syndrome (GBS), or acute inflammatory demyelinating polyneuropathy (AIDP), is an acute autoimmune somatic neuropathy commonly associated with prominent autonomic dysfunction that can lead to both morbidity and mortality (Zochodne, 1994, Panagyres, 1989). As many as 50% of patients report a viral illness 1-4 weeks prior to onset.

Autoantibodies can be found against gangliosides, such as with anti-GM1 antibodies. Pathologic studies of the autonomic nervous system in GBS may demonstrate edema and inflammation of autonomic ganglia and destruction of peripheral ganglion cells. Chromatolysis, mononuclear cell infiltration, and nodules of Nageotte can be found within sympathetic ganglia (Zochodne, 1994).

Lambert-Eaton myasthenic syndrome

Lambert-Eaton myasthenic syndrome (LEMS) is an acquired neuromuscular transmission disorder with antibodies present against presynaptic voltage-gated P/Q-type Ca^{2+} channels. LEMS is frequently associated with clinical and electrophysiologic evidence of dysautonomia, which can be severe in 20% of patients with LEMS (O'Suilleabhain, 1998). In 50% of cases, LEMS is associated with a neoplasm, most commonly small cell carcinoma of the lung.

Holmes-Adie syndrome and Ross syndrome

Holmes-Adie syndrome is probably autoimmune in nature and manifests as tonic pupil or pupils associated with tendon areflexia. In rare cases, it is associated with an autonomic neuropathy with prominent orthostatic hypotension. Ross syndrome is a related condition where segmental anhidrosis occurs in conjunction with Adie pupil.

Idiopathic distal small-fiber neuropathy

Idiopathic distal small-fiber neuropathy is a chronic peripheral somatic neuropathy affecting sympathetic postganglionic sudomotor fibers. Clinical features may include allodynia, sympathetic vasomotor changes, pallor and rubor, cyanosis, and even mottling (Stewart, 1992).

HIV infection

HIV infection may lead to autonomic neuropathy, particularly in late-terminal stages of disease. Often, this occurs in conjunction with a somatic neuropathy related to HIV infection or complications of AIDS.

Chagas disease

Chagas disease due to infection with *Trypanosoma cruzi* is occasionally associated with autonomic neuropathy during the chronic stage of infection. Parasympathetic dysfunction tends to be greater than sympathetic dysfunction.

Autoimmune destruction of the peripheral nervous system (PNS) and autonomic nervous system may occur, especially of autonomic nerves supplying the cardiovascular and gastrointestinal systems. Other infections such as leprosy, diphtheria, and Lyme disease can be associated with autonomic neuropathy rarely.

Botulism

Botulism produces neuromuscular paralysis due to prevented release of acetylcholine from the presynaptic terminus, as well as an acute cholinergic neuropathy.

Chronic idiopathic anhidrosis

Chronic idiopathic anhidrosis is an acquired generalized loss of sweating without other autonomic features.

Amyloid neuropathy

Three main forms of amyloid neuropathy exist: (1) hereditary amyloid neuropathy; (2) neuropathy associated with hematologic disease, such as multiple myeloma; and (3) acquired neuropathy. Of all autonomic neuropathies, amyloidosis probably causes the most severe forms, with universal autonomic dysfunction common. A somatic neuropathy is often coexistent.

Familial amyloid polyneuropathy

Familial amyloid polyneuropathy (FAP) is a rare and severe hereditary form of amyloidosis caused by a genetic mutation of the transthyretin gene. Mutant transthyretin, produced in the liver accumulates as amyloid deposits in the PNS and autonomic nervous system.

Rarely, a mutation in the gelsolin gene, which produces a protein important in cytoskeletal actin function, may also lead to amyloid deposition in autonomic nerves. Acquired amyloidosis with accumulation of immunoglobulins kappa or lambda light chains may be associated with the presence of multiple myeloma. Another acquired amyloidosis occurs with dialysis, with beta2-microglobulin deposits in the nervous system. In syndromes of amyloidosis, the development of generalized autonomic failure significantly worsens the overall prognosis. At present, liver transplantation, currently the most effective treatment for FAP, may not slow the development of autonomic neuropathy (Delahaye, 2006).

Diabetes mellitus

Diabetes mellitus is associated with many forms of neuropathy, and autonomic involvement is present in many (Zochodne, 1999). Parasympathetic abnormalities are thought to precede sympathetic abnormalities, but this has not been verified.

Laboratory evidence of autonomic dysfunction is frequent, perhaps as frequent as nerve conduction abnormalities in diabetic neuropathy (Zochodne, 1999).

Acute diabetic autonomic neuropathy appears as acute pandysautonomia and may be associated with ganglionic antibodies in some patients. Diabetic radiculoplexopathy is associated with prominent autonomic dysfunction, which may have an immunologic cause with destruction of both large and small nerve fibers (Zochodne, 1999).

Diabetes affects autonomic neurons differently; sympathetic neurons from the celiac/superior mesenteric ganglia develop pathological changes, while sympathetic

superior cervical ganglion neurons do not. This selectivity may be related to increased sensitivity to oxidative stress (Semra, 2006).

Uremic neuropathy

Uremic neuropathy is a primarily somatic neuropathy commonly associated with coexistent autonomic neuropathy, either symptomatic or subclinical. The cause of uremic neuropathy remains unknown, although either accumulated toxins or lack of a neurotrophic factor may be responsible because renal transplantation reverses autonomic dysfunction while dialysis does not.

Celiac disease

Autonomic neuropathy may occur in approximately 50% of adults with celiac disease, leading to clinical features of presyncope and postural nausea (Gibbons, 2005). Autonomic denervation may be related to antineuronal antibodies; the condition does not appear to respond to a gluten-free diet (Tursi, 2006).

Hepatic disease–related neuropathy

Hepatic disease–related neuropathies, as with primary biliary cirrhosis (PBC), can be associated with autonomic neuropathy in 48% of patients. The cause of autonomic neuropathy in hepatic disease remains unclear, but it may be associated with toxic metabolite accumulation or related immune-mediated mechanisms. It may be reversible following liver transplantation. Maheshwari et al (2004) hypothesized that patients with autonomic neuropathies are more likely to develop hepatic encephalopathy due to a decreased intestinal transit time. Although this group's study did not show an independent effect of autonomic neuropathy on hepatic encephalopathy, their findings did demonstrate that patients with autonomic neuropathies were more likely to develop new-onset hepatic encephalopathy.

Vitamin deficiency and nutrition-related neuropathy

Deficiency of vitamin B-12 and alcohol- or nutrition-related neuropathy may also be associated with parasympathetic dominant autonomic dysfunction.

Toxic and drug-induced autonomic neuropathy

Toxic and drug-induced autonomic neuropathies may occur with chemotherapeutic medications such as vincristine, cisplatin, carboplatin, vinorelbine, Taxol, and suramin. Other therapeutic agents associated with a toxic autonomic neuropathy include acrylamide, pyridoxine, thallium, amiodarone, perhexiline, and gemcitabine.

Paraneoplastic autonomic neuropathy

Paraneoplastic autonomic neuropathy may occur as a component of paraneoplastic neuronopathy with anti-Hu antibodies in 23% of patients. Autonomic dysfunction appears to result from autoimmune destruction of autonomic postganglionic and myenteric neurons.

Additional antibodies against ganglionic receptors are found in 41% of patients with idiopathic or paraneoplastic autonomic neuropathy. A reversible and dose-dependent association between the level of ganglionic binding antibodies and severity of autonomic dysfunction occurs.

A variant of paraneoplastic autonomic neuropathy is an enteric neuronopathy that exists with antibodies directed against the myenteric plexus. Other paraneoplastic autonomic syndromes may have autoantibodies against neuronal cytoplasmic proteins of the collapsin response–mediator family (CRMP-5) and against Purkinje cell cytoplasm (PCA-2).

Sjögren syndrome

Sjögren syndrome may lead to peripheral and autonomic neuropathy without characteristic systemic symptoms. A small-fiber neuropathy associated with Sjögren syndrome can be associated with widespread anhidrosis. Also, a sensory neuronopathy due to Sjögren syndrome can be associated with autonomic dysfunction. The cause of neuropathy in these patients is likely to be autoimmune, but this remains unclear.

Rheumatoid arthritis, systemic lupus erythematosus, and connective tissue disorders

Rheumatoid arthritis, systemic lupus erythematosus, and other connective tissue disorders may have abnormalities of sympathetic postganglionic function. Some of these patients may have autoantibodies to ganglionic acetylcholine receptors. Autoimmune thyroiditis, as with chronic thyroiditis and Hashimoto thyroiditis, can be associated with some features of Sjögren syndrome such as xerostomia. Patients with systemic sclerosis and mixed connective tissue disorder may have abnormalities of autonomic functioning of esophageal motor activity.

Acute intermittent porphyria and variegate porphyria

Acute intermittent porphyria and variegate porphyria can both have forms of peripheral neuropathy. Attacks can be triggered by exposure to particular drugs. During episodes with acute polyneuropathy that may mimic GBS. Autonomic dysfunction, particularly cardiac and vascular in nature, can be prominent.

Hereditary sensory autonomic neuropathy

Currently, 5 types of hereditary sensory autonomic neuropathy (HSAN) have been defined. HSAN I has an autosomal dominant inheritance, and the disease is characterized by distal limb involvement with marked sensory loss and susceptibility to painless injuries.

HSAN I has been associated with point mutations in serine palmitoyltransferase (SPT) at chromosome arm 9q22.1-q22.3 (Bejaoui, 2001). SPT is the rate-limiting enzyme in synthesis of sphingolipids, including ceramide and sphingomyelin. Ceramide is necessary for regulation of programmed cell death in a number of tissues, including the differentiation of neuronal cells.

HSAN II is inherited as an autosomal recessive condition and is more severe with a congenital onset. HSAN II has a pansensory loss with early ulcers, and nerves demonstrate a marked loss of myelinated and unmyelinated fibers. HSAN III (Riley-Day syndrome) is autosomal recessive in Ashkenazi Jews, with early childhood onset of autonomic crises. The genetic defect in HSAN III is in the inhibitor of kappa light polypeptide gene enhancer in B cells, kinase complex-associated protein (IKBKAP) at chromosome arm 9q31. HSAN III nerve pathology shows absence of unmyelinated fibers with essentially normal myelinated fibers.

Patients with HSAN IV present with widespread anhidrosis and insensitivity to pain. The genetic defect in HSAN IV is in the tyrosine kinase receptor A or nerve growth factor receptor at chromosome arm 1q21-q22. This defect is autosomal recessive. Recently, two novel missense mutations in the tyrosine kinase domain were found in a 10-year-old patient with HSAN IV (Ohto, 2004). This finding may provide a better understanding of the neuropathophysiology of HSAN IV.

Patients with HSAN V present with pain insensitivity and preservation of other sensory modalities. Some patients with HSAN V have similar genetic abnormalities as those with HSAN IV. The genetic mutation has been isolated to the nerve growth factor beta gene (Einarsdottir, 2004).

Types of HSAN

HSAN	Mode of Inheritance	Onset	Symptoms	Signs
Type I	Autosomal dominant, point mutations in SPT, 9q22.1-9q22.3	Second decade of life	Distal lower-limb involvement, ulceration of the feet, particularly the soles	Low sensory action potential amplitude
Type II, Morvan disease	Autosomal recessive	Congenital onset	Pansensory loss of upper and lower limbs, also trunk and forehead; early ulcers	Loss of myelinated and unmyelinated fibers
Type III, Riley-Day syndrome or familial dysautonomia)	Autosomal recessive, 9q31	Childhood onset, predominantly Ashkenazi Jews	Pallor in infancy, irregularities in temperature and blood pressure; Difficulties in eating and swallowing	Absence of unmyelinated fibers
Type IV	Autosomal recessive, 1q21-1q22	Congenital onset	Widespread anhidrosis, lost sense of pain, mental retardation	Loss of myelinated and small unmyelinated fibers
Type V	Autosomal recessive	Congenital onset	Pain insensitivity in extremities	Not applicable

Fabry disease is an X-linked recessive disorder with mutations in the gene for alpha-galactosidase. Somatic and autonomic neuropathy is due to accumulation of glycolipids. Attacks may be triggered by changes in temperature or exercise. Nerve pathology demonstrates loss of both small myelinated and unmyelinated fibers.

Mortality/Morbidity

Most cases have a gradually progressive course. In several forms of autonomic neuropathy, development of autonomic dysfunction worsens overall prognosis. This is particularly true in amyloidosis, diabetic neuropathy, and GBS. Patients with severe dysautonomia can have sudden death secondary to cardiac dysrhythmia, as has been documented in GBS and diabetic neuropathy.

- Single-photon emission CT (SPECT) and positron emission tomography (PET) have demonstrated that cardiac sympathetic dysfunction is commonly present in both type I and type II diabetes mellitus.
- When associated with vascular complications, dysautonomia related to diabetic neuropathy is also associated with increased mortality.
- In other disorders, other forms of systemic dysfunction may lead to mortality, such as with kidney failure in Fabry disease.

Race

One form of autonomic neuropathy, HSAN III (Riley-Day syndrome), is inherited in an autosomal recessive form in Ashkenazi Jews.

Sex

In general, no predilection for autonomic neuropathies exists with regard to sex. POTS is more common among young females. Fabry disease is inherited as an X-linked recessive disorder; therefore, it manifests in males.

Age

In general, no predilection for autonomic neuropathies exists with regard to age. Patients with most of the forms of HSAN (except HSAN I) present at birth or in childhood.

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Case report

Hydrocephalus associated with Guillain-Barre syndrome

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Abstract

A 68-year-old man developed progressive four-limb weakness and areflexia 17 days after an influenza vaccination. He was diagnosed with Guillain-Barre syndrome (GBS), and remained ventilator dependent and bed-bound for 3 months, despite plasmapheresis and immunoglobulin infusion. However, cognitive impairment, excessive daytime sleepiness, and motor disability were still present, even when he was no longer ventilator dependent. Brain computerized tomography scan and isotope cisternography was consistent with normal pressure hydrocephalus. His motor control and cognitive function recovered almost completely after insertion of a ventriculoperitoneal shunt. Although hydrocephalus is not frequently associated with GBS, our case report indicates that brain imaging is necessary in GBS patients whose cognitive functions deteriorates after disease onset.

Article Outline

1. Introduction
2. Case report
3. Discussion
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Vivid dreams, hallucinations, psychosis and REM sleep in Guillain-Barré syndrome

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▸ **Abstract**

We conducted a prospective controlled study of the clinical and biological determinants of the mental status abnormalities in 139 patients with Guillain-Barré syndrome (**GBS**) and 55 patients without **GBS** placed in the intensive care unit (ICU controls). There were mental status changes in 31% of **GBS** patients and in 16% of controls (odds ratio = 2.3; $P = 0.04$). In **GBS** patients, they included vivid dreams (19%), illusions (30%, including an illusory body tilt), hallucinations (60%, mainly visual) and delusions (70%, mostly paranoid). They appeared a median 9 days after disease onset (range 1-40 days, during the progression or the plateau of the disease), and lasted a median 8 days. Seven (16%) patients experienced the symptoms before their admission to the ICU. Hallucinations were frequently hypnagogic, occurring as soon as the patients closed their eyes. Autonomic dysfunction, assisted ventilation and high CSF protein levels were significant risk factors for abnormal mental status in **GBS** patients. CSF hypocretin-1 (a hypothalamic neuropeptide deficient in narcolepsy) levels, measured in 20 patients, were lower in **GBS** patients with hallucinations (555 ± 132 pg/ml) than in those without (664 ± 71 pg/ml, $P = 0.03$). Since the mental status abnormalities had dream-like aspects, we examined their association with rapid eye movement sleep (REM sleep) using continuous sleep monitoring in 13 **GBS** patients with ($n = 7$) and without ($n = 6$) hallucinations and 6 tetraplegic ICU controls without hallucinations. Although sleep was short and fragmented in all groups, REM sleep latency was shorter in **GBS** patients with hallucinations (56 ± 115 min) than in **GBS** patients without hallucinations (153 ± 130 min) and in controls (207 ± 179 min, $P < 0.05$). In addition, sleep structure was highly abnormal in hallucinators, with sleep onset in REM sleep periods (83%), abnormal eye movements during non-REM sleep (57%), high percentages of REM sleep without atonia ($92 \pm 22\%$), REM sleep behaviour disorders and autonomic dysfunction (100%), reminiscent of a status dissociatus. The sleep abnormalities, that were almost absent in non-hallucinated **GBS** patients, were not exclusively related to ICU conditions, since they also appeared out of ICU, and were reversible, disappearing when the mental status abnormalities vanished while the patients were still in ICU. In conclusion, the mental status abnormalities experienced by **GBS** patients are different from the ICU delirium, are strongly associated with autonomic dysfunction, severe forms of the disease and possibly with a transitory hypocretin-1 transmission decrease. Sleep studies suggest that mental status abnormalities are wakeful dreams caused by a sleep and dream-associated disorder (status dissociatus).

Keywords:
Guillain-Barré syndrome; hallucinations; hypocretin; ICU syndrome; REM sleep; REM sleep behaviour disorders; status dissociatus.

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[*] These authors contributed equally to this work
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Cutaneous innervation in Guillain-Barre syndrome: pathology and clinical correlations.

Articles

Pan, Chun-Liang 1; Tseng, To-Jung 2; Lin, Yea-Huey 1; Chiang, Ming-Chang 1; Lin, Whei-Min 2; Hsieh, Sung-Tsang 1,2

Abstract:

Guillain-Barre syndrome (GBS) is traditionally considered to be a large-fibre neuropathy. However, the presence of hypo-aesthesia, dysaesthesia and dysautonomia in GBS patients raises the possibility that small-diameter sensory and autonomic nerves may also be affected. To investigate small-fibre neuropathy in GBS, we performed a skin biopsy from the distal leg of 20 patients with the demyelinating form of GBS. Skin sections were immunohistochemically stained with antiserum against protein gene product 9.5 (PGP 9.5), a ubiquitin C-terminal hydrolase. Cutaneous innervation was evaluated by measuring epidermal nerve density (END), and END was further correlated with various clinical and electrophysiological parameters. In GBS patients, END values were much lower than in age- and gender-matched control subjects (5.03 +/- 1.18 versus 10.16 +/- 0.87 fibres/mm, P < 0.001). Eleven patients (55%) had reduced epidermal innervation with pathological evidence of active nerve degeneration in the dermis: fragmentation of subepidermal nerve plexuses and a beaded appearance of dermal nerves. GBS patients had significantly elevated thermal thresholds with higher warm threshold temperatures (44.54 +/- 1.04 versus 39.00 +/- 0.35[degrees]C, P < 0.001) and lower cold threshold temperatures (25.57 +/- 1.11 versus 29.05 +/- 0.21[degrees]C, P=0.032). Reduced END values were associated with an elevated warm threshold (P=0.027), ventilatory distress (P=0.037) and dysautonomia (P=0.001). END values were negatively correlated with disability grade on a scale of 1-6 (slope -0.134 +/- 0.038, P=0.0018). Patients with reduced END values tended to have a slower recovery than those with normal END values (P=0.013, median time 12 versus 2 weeks). Patho logically, sudomotor innervation of the skin was reduced in five of 17 (29.4%) GBS patients in whom sweat glands could be recognized. These findings suggest that small-fibre sensory and autonomic neuropathies exist in a significant proportion of GBS patients, and that END values are correlated with functional disabilities. In summary, GBS should be considered a global neuropathy instead of a pure large-fibre neuropathy.

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CORRESPONDENCE

S100B in Guillain–Barré syndrome

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Editor—Guillain–Barré syndrome (**GBS**), a subacute inflammatory demyelinating polyneuropathy, is the most common cause of acute neuromuscular paralysis. Up to one-third of patients with **GBS** require mechanical ventilation (MV) in the ICU,[1] although outcome is generally good,[2] acute mortality remains relatively high and ~20% of hospitalized patients may have a long-term disability.[3]

Serum protein S100B is a recognized marker of traumatic **brain** injury. We observed an increase in S100B levels in two **GBS** patients, pointing to a potential acute influence of peripheral neuropathy on levels not related to CNS injury.

Serum S100B was measured from venous blood samples in two **GBS** patients requiring MV, on admission to ICU and after 10 days. S100B values >0.2 µg litre^{-1} are considered abnormal. The patients were examined by a neurologist and the diagnosis was based on the National Institute of Neurological and Communicative Disorders and Stroke diagnostic criteria.[4]

The first patient was treated with a total of 10 plasma exchanges and steroids and required 13 days of MV. The patient was discharged from hospital at 40 days with severely limited lower limb activity and confined to bed. On admission, the S100 serum level was raised at 0.495 µg litre^{-1}, and at 10 days the level had reduced to 0.093 µg litre^{-1}.

The second patient was treated with plasma exchange and steroids. The patient recovered spontaneous ventilation after 62 days and was discharged, still confined to a chair, 90 days after SGB onset. On ICU admission, the S100B serum level was very high (2.61 µg litre^{-1}), and was still abnormal 10 days later (0.891 µg litre^{-1}).

The serum S100B increases observed in these two **GBS** patients exceed those attributed to **brain** tissue damage patients with mild traumatic head injury.[5] The physiological role of this neuroprotein is not yet clear, but in **GBS** it may act as a regeneration stimulus or as marker of neuronal damage. A comparison of CSF and serum levels of S100B would be useful for the

understanding of the role played by this protein in **GBS**. S100B in **GBS** might be an expression of neurotrophic and neuroprotective activity but no conclusion should be made without extensive and focused clinical and experimental studies.

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References

1 Sharshar T, Chevret S, Bourdain F, Raphaël J. Early predictors of mechanical ventilation in Guillain-Barré syndrome. *Crit Care Med* 2003; **31**: 278–83[CrossRef][ISI][Medline]

2
Henderson RD, Lawn ND, Fletcher DD, McClelland RL, Wijdicks EF. The morbidity of Guillain-Barré syndrome admitted to the intensive care unit. *Neurology* 2003; **60**: 17–20[Abstract/Free Full Text]

3
Dornonville de la Cour C, Andersen H, Stalberg E, Fuglsang-Frederiksen A, Jakobsen J. Electrophysiological signs of permanent axonal loss in a follow-up study of patients with Guillain-Barré syndrome. *Muscle Nerve* 2005; **31**:70–7[Medline]

4
National Institute of Neurological and Communicative Disorders and Stroke ad hoc Committee (NINCDS). Criteria for diagnosis of Guillain-Barré syndrome. *Ann Neurol* 1978; **3**: 565–6[CrossRef][Medline]

5 Ingebrigtsen T, Waterloo K, Jacobsen EA, Langbakk B, Romner B. Traumatic **brain** damage in minor head injury: relation of serum S-100 protein measurements to magnetic resonance imaging and neurobehavioral outcome. *Neurosurgery* 1999; **45**: 468–75[ISI][Medline]

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