



NEWSLETTER OF THE GBS ASSOCIATION OF NSW INCORPORATED

RECOVERY

Dear Members,

It gives me great pleasure to welcome you to our brand new Recovery. Not only are we unveiling your new Look Logo and Newsletter in this edition but we are also announcing the details our new website too! Please be sure to pay us a visit. There is plenty of information, member stories, committee profiles and even access to past newsletters for members!

www.gbs-cidp-nsw.org.au



We have a new email address too so please make a note and drop us a line!

info@gbg-cidp-nsw.org.au

Speaking of the website, I could not possibly miss this opportunity to publicly acknowledge my dear friend Peter who has donated many hours of his own time to bring the website to life. Personally and on behalf of the Association thank you so much Pete for your patience and kindness in helping to realise the vision! We must also thank our former webmaster Max Valente who has managed our website for as long as I can remember and who has done a superb job, thank you too Max for your dedication to the cause!

We also hope you like our new Logo. Many thanks to all the people who participated in the voting process. It was a tough decision between two very good interpretations of our desire to communicate a supportive group who will be there to help people see a 'brand new day' when coming out from the worst of GBS or CIDP and other related disorders.

Unfortunately there could only be one chosen but I think it ticks most, if not all of the boxes we set ourselves.

The Association is definitely going forward in 2014. In addition to the new Logo, Newsletter and website, we are also undertaking to re-work our Association Constitution. It has been over 20 years since this has been done so perhaps it is overdue, but the time feels right to do it now. Look out for information in this regard as we share with our members in coming weeks.



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GBS | CIDP
GUILLAIN BARRE SYNDROME
CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY

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continued

But wait there's more we are heading back to our 'spiritual home' at the Royal Rehabilitation Centre at Ryde. We took our meetings to Ryde-Eastwood Leagues Club a few years back during the re-building of the Rehabilitation Centre and whilst the Club has served us well and been very helpful and supportive, we are excited to be returning 'home'. We hope the venue is convenient for people to attend meetings and it has the added benefit of being more accessible to in-patients and their families as well. We know this is probably the most important time for people to seek support and information so we hope by being closer during this period will be helpful to those in need. Our next meeting is **5th April**, see details below and on our new website.

Well I really hope you like what you see in the following pages and on the website. But remember, please provide me your feedback and/or your story for others to read, unfortunately I have no new personal cases to print this time, which is a shame. You may never know just how much your journey may touch and inspire, but rest assured it can have an impact - we know because we hear it from a lot of people!

Before I sign-off on this edition I would like to wish our Patron Ursula Carlile and Mary's husband Arthur our very best wishes for a speedy recovery. Both have not been quite at their best lately with varying health issues but we hear positive reports on their progress which is fantastic news.

You can see I have indulged myself with two pages this edition so will be off for now. However I hope to see some of you at the next meeting or share again in our next edition of Recovery. Take care.

Christine S-M



5th April

Royal Rehabilitation Centre Sydney

235 Morrison Road, Ryde

"Susan Schardt Conference Room" L1



09:30 to 11:00 - Committee business and administration

11:00 to 12:30 - Open forum for members and family / guest speaker

Visitors are welcome to both sessions or the Open Forum only if preferred.

Message from the Chair

Welcome to our first edition for 2014. You will have noticed the new look, new logo and hopefully by the time of this edition our new website will be up and running or very close. In this regard, I would like to publicly acknowledge the hard work of Christine who has driven the new website project and the website committee. Only those who have participated in such a project can realise the amount of decision making required and the infinite choices that must be made in order to build a top quality website. Further, Christine is also the Editor of Recovery and our Treasurer which makes her efforts building our website even more praiseworthy.

Touching further on the above, the support we provide to those impacted by GBS and CIDP is made possible by the Committee members who dedicate a portion of their own quality family time to ensure the Association runs day in and day out and is compliant with a myriad of legislative requirements. They do this without thought to payment because they know there are many current sufferers of GBS and CIDP, family and friends who need our support and whilst there remains no cure, many people will need our support in the future.

As Chair I know that to offer any payment to the Committee would be met with a resounding rejection and question, "What for?" They volunteer for many personal reasons but I believe there is an underlying 'calling' to do so and they know, all being sufferers themselves, the initial trauma and stress that comes with a diagnosis of GBS and CIDP, the challenges it presents and changes to an erstwhile healthy life. Their reward, for want of a better word, is seeing the expression on the face of a patient, family member or friend at a forum when they realise they are not alone or the change in the tone of the voice at the other end of the telephone conversation when they realise support is at hand. Our forums during the meetings are often an emotional roller coaster with many patients at first expressing their feelings of isolation and fear and even tears, from not only them but many at the meeting listening intently to a journey often not very dissimilar from their own, followed by a visible realisation I am in the company of fellow travellers on a journey to recovery. I walk away from these meetings 'fulfilled'.

At our next AGM all Committee roles will be vacated, persons nominated, votes counted and roles appointed. As current Chair, I often consider the long term viability of the Association, seeking new members, new ideas, new perspectives and those willing to consider a role helping to run the Association.

Another aspect of being on the Committee is discussing and deciding where the Association will direct its efforts in support of those with GBS and CIDP. During the years 2012-13 we were in receipt of a number of generous donations. If you have any ideas about where we might focus our efforts whether funding some specific research, equipment, aids or other area that might benefit sufferers etc. please drop us a line.

Looking forward to seeing you at our next meeting. Kindest regards,

Mark



2014 is a good year to become involved, why not join us for a meeting?

Our meeting forums provide an opportunity to meet new sufferers or maybe to hear a recovery update. If you have a GBS or CIDP story please consider coming along to a meeting or alternatively write your story, our Editor is always looking for interesting articles for our newsletter.



Article based on a
plenary lecture given at
the 2012 Peripheral
Nerve Society (PNS)
meeting

Update on Guillain-Barre Syndrome

Simon Rinaldi

Nuffield Department of Clinical Neurosciences, University of Oxford, Oxford, UK
Journal of the Peripheral Nervous System 18-99-112 (2013)

Abstract Understanding of Guillain-Barre' syndrome (GBS) has progressed substantially since the seminal 1916 report by Guillain et al. Although Guillain, Barre', and Strohl summarised the syndrome based on observations of two French infantrymen, 2012 saw the beginning of an ambitious collaborative study designed to collect detailed data from at least 1,000 patients worldwide (IGOS, www.gbsstudies.org/about-igos). Progress has been made in many areas even since GBS was last reviewed in this journal in 2009. GBS subsequently received prominent attention in light of concerns regarding H1N1 influenza vaccinations, and several large-scale surveillance studies resulted. Despite these developments, and promising pre-clinical studies, disease-modifying therapies for GBS have not substantially altered since intravenous immunoglobulin was introduced over 20 years ago. In other areas, management has improved. Antibiotic prophylaxis in ventilated patients reduces respiratory tract infection, thromboprophylaxis has reduced the risk of venous thromboembolism, and there is increasing awareness of the benefit of high-intensity rehabilitation. This article highlights some of the interesting and thought-provoking developments of the last 3 years.

Epidemiology

Most well-designed epidemiological studies of Guillain-Barre' syndrome (GBS) return an annual incidence of around 1–2 cases per 100,000. Nationwide hospital records from New Zealand were analysed from 1988 to 2010 to reveal an overall incidence of 2.32 cases per 100,000 per year. Interestingly, following a national programme aimed at reducing *Campylobacter jejuni* (C. jejuni) contamination of poultry, the incidence of GBS also fell (Baker et al., 2012). A shorter study from The Netherlands (1996–2008) returned a slightly lower annual incidence of 1.14/100,000, but here case records were manually reviewed by a neurologist (Vander Maas et al., 2011). The most impressive recent epidemiological data come from a comprehensive meta-analysis with strict inclusion criteria for quality, including the requirement for explicit use of an accepted case definition and review of the diagnosis by an expert (Sejvar et al., 2011).

The study encompassed 1,643 cases, 152.7 million person-years of follow-up, and confirms an increasing incidence with age and 1.8-fold excess of male cases. Regression equations were produced to allow an expected age-specific population incidence to be calculated and these equations are broadly in keeping with the oft quoted 1 in 1,000 lifetime risk of GBS.

H1N1/vaccination

Some of the recent attention to GBS epidemiology has been driven by concerns regarding the possibility that H1N1 influenza vaccine might trigger the disease, much in

the way that the publication of the original 1978 GBS diagnostic criteria was precipitated by the apparent aetiological association of some GBS cases with the 1976/7 "swine flu" vaccine (Asbury, 1978). Neither the 2009 "seasonal" trivalent H5N1 vaccine nor the "swine flu" monovalent H1N1 vaccine induced anti-ganglioside antibodies in mice or men (Yuki et al., 2012), unlike the 1976/7 swine flu vaccine and two other seasonal influenza vaccines (1991/2 and 2004/5, neither associated with an apparent increased risk of GBS) (Nachamkin et al., 2008).

This provided some limited initial reassurance regarding the safety of the contemporary vaccines. Subsequently, a number of different surveillance studies, using both population level and self-controlled methodologies, returned broadly similar results with around 1–2 excess cases of GBS per million vaccine doses administered, no different to previous risk estimates for seasonal influenza vaccine (Tokars et al., 2012; Wise et al., 2012; Yih et al., 2012). One study showed an excess of 5 cases per million doses with the monovalent "swine flu" vaccine vs. 1.1 cases per million with the trivalent seasonal vaccine.

However, it is informative to note that the 95% confidence intervals for these estimates overlap, and that the delayed production of the swine flu vaccine meant that this was administered concurrently with an influenza epidemic, whereas the seasonal immunisation programme started much earlier (Greene et al., 2012).

This small excess risk of GBS needs to be set against the potentially serious acute complications of influenza

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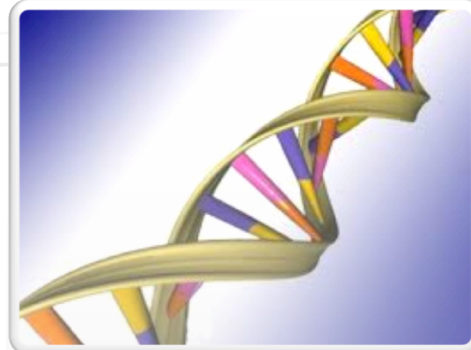
infection itself. Out of approximately 6.6 million H1N1 cases in California in 2009, 2,069 patients required admission to intensive care or died from their illness, and 419 suffered acute neurological complications (including encephalopathy/encephalitis, seizures, and meningitis) (Glaser et al., 2012).

Indeed, GBS itself can complicate influenza infection. An earlier study of the UK General Practice Database looked at the excess risk of GBS in the 2 months following consultation for a number of other reasons, comparing 553 cases with 5,445 controls matched for age, sex, season, and GP clinic. Although consultation for influenza like illness was associated with an approximately 18-fold increased chance of GBS diagnosis in the next 2 months (OR=18.6, 95% CI: 7.5–46.4), consultation for influenza vaccination was associated with no such risk, and in fact showed a non-significant trend towards being protective (OR = 0.16, 95% CI: 0.02 – 1.25) (Tam et al., 2007).

Clinical Features

A number of series have examined some of the often neglected clinical features of GBS, most notably sensory disturbance and pain. An Indian study of 60 patients (Karkare et al., 2011) found paraesthesia in 75%, but objective sensory loss in much smaller proportions – pin prick, proprioception, and vibration were impaired in 13.3%, 23.3%, and 18.3% of cases, respectively – echoing Guillain's original assertion of "paraesthesias with slight disturbance of objective sensation" (Guillain et al., 1916). Of course, GBS is most clearly characterised by "disorders of motor function and abolition of the tendon reflexes." Indeed, a significant proportion of patients have a pure motor form of the syndrome without any notable sensory disturbance (20% of patients in the Indian series had neither clinical nor electrophysiological evidence of sensory disturbance). The reverse pattern (pure sensory GBS) is much more rarely seen (Oh et al., 2001), and its very existence has sometimes been doubted (Windebank et al., 1990). Nevertheless, if GBS is used as an umbrella term for a monophasic, post-infectious, immune-mediated disorder of the peripheral nervous system, and the absolute diagnostic requirement for motor dysfunction is not upheld, then pure sensory GBS does seem to be a valid entity.

Pain is also a common feature, present in 50% of the Indian series and in 66% of 156 Dutch patients (Ruts et al., 2010). Interestingly, pain often preceded weakness. Back pain – affecting lumbar, intrascapular, and cervical regions – was particularly prevalent in the acute phase, although pain in extremities was most frequent overall. The pain was substantial in many, with 86% reporting moderate to severe pain despite the use of analgesics. Treatment with methylprednisolone did not alter this. The same group also reported that the intra-epithelial nerve fibre density as assessed by skin biopsy was significantly lower in GBS patients with pain than those without pain (Ruts et al., 2012), and observed damage to myelinated dermal fibres in the presence of a mononuclear cell infiltrate, further linking dysimmune pathology and neuropathic pain (Calvo et al., 2012).



Genetics

Two long appreciated facts about GBS strongly suggest that host factors influence the chances of developing the condition. First, at most 1 in 1,000 people infected with *C. jejuni* develop GBS (McCarthy and Giesecke, 2001; Tam et al., 2006), and a similar magnitude of risk has been estimated for cytomegalovirus infection (Orlikowski et al., 2011). Although some strains of *C. jejuni* clearly have an increased risk of inducing GBS compared with others, these strains rarely induce GBS in those they infect. There have been individual reports of GBS occurring in single family members in the context of familial outbreaks of *C. jejuni* gastroenteritis. Intriguingly, anti-ganglioside antibodies were found in other family members as well as in the patients, although the patterns and titres of antibody positivity differed (Ang et al., 2000; Hirano et al., 2003). In the latter report, levels of soluble ICAM-1 were higher in the asymptomatic elder brother when compared with the symptomatic younger brother. No other differences were noted in either study.

Second, the recurrence rate for GBS is usually quoted at around 5%, which is at least 50 times in excess of the risk in the background population. This was confirmed by a population-based Swedish study. In this series, 15 of 229 total GBS cases (6.6%) had at least one episode of recurrence. These patients tended to be younger at their first presentation and have a shorter episode duration, defined as time from disease onset to a clinically stable phase after remission (Mossberg et al., 2012).

(Genetics continued)

Despite these observations, a clear genetic risk factor for GBS has not yet been identified. Previous studies have found no association with HLA, T-cell receptor, CD14, or Toll-like receptor 4 (TLR4) polymorphisms (Ma et al., 1998; Geleijns et al., 2004; 2005). A 2006 study reported an association between CD1a/CD1e polymorphisms and GBS risk (Caporale et al., 2006), although concerns were raised regarding the statistical analysis used (Bang et al., 2007), and a subsequent, larger study showed no such association (Kuijff et al., 2008).

Most recently, an attempt has been made to systematically assess the available data on the genetic risk factors for GBS. Studies without a case – control design, with evidence of heterogeneity, and where genotype frequencies were not available were excluded from analysis. Furthermore, only polymorphisms reported by three or more separate studies were included. In this way, Wu et al. evaluated six genetic variants of three candidate genes (TNF- α , Fc γ R, and CD1), encompassing 1,590 GBS cases and 2,154 controls in total (Wu et al., 2012). They confirmed that there was no evidence for an association with the CD1 polymorphisms discussed previously. Although the meta-analysis appears to have inappropriately included 46 patients with chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) from a 2007 study (De Angelis et al., 2007), the conclusion would remain the same without these patients. Overall, only a 308AA polymorphism of the TNF- α gene was significantly associated with GBS. However, this association was not apparent within the sole included European study (Geleijns et al., 2007). Furthermore, the largest Asian study showed that the association only held for axonal subtypes of GBS, and not the demyelinating type that dominates in Europe, suggesting one potential reason for this discrepancy (Jiao et al., 2012).

Other as yet unidentified genetic polymorphisms may influence different aspects of the disease process aside from overall susceptibility. Indeed, earlier work has shown that polymorphisms in the mannose-binding lectin (MBL) gene alter serum levels and the activity of MBL, and that lower levels are associated with less severe GBS (Geleijns et al., 2006). This presumably reflects the involvement of MBL in the complement system and/or other immune processes. Nevertheless, this correlation is far from absolute, suggesting the involvement of other

genes. It is also likely that, as has been found for IVIg responsiveness and TAG-1 in CIDP (Iijima et al., 2009), certain polymorphisms will affect treatment responsiveness in GBS. Indeed, one presumes that the observed variations in delta-IgG measurements correlating with GBS prognosis (Kuitwaard et al., 2009), as discussed later, are a result of genetic polymorphisms governing IVIg pharmacokinetics.

Pathophysiology

Unravelling the finer details of the pathophysiology of GBS and integrating these into an overall disease mechanism have proved problematic. One major confounding factor is that there are clearly at least two pathologies that result in a clinical diagnosis of GBS – demyelinating pathology with acute inflammatory demyelinating polyradiculoneuropathy (AIDP) and axonal pathology with acute motor axonal neuropathy (AMAN). Furthermore, it is not always possible to accurately separate even these major GBS subtypes using standard clinical and electrophysiological assessments, as further discussed below. The applicability of animal models to the human disease is also open

to question. Unlike experimental autoimmune neuritis' (EAN) (Waksman and Adams, 1955), there has been no consistent detection of antibodies or T cells directed against myelin proteins in human GBS (Makowska et al., 2008). Similarly, murine models of ganglioside antibody-mediated GBS usually require passive transfer of antibody produced in ganglioside-deficient to ganglioside-overexpressing mice, along with the artificial provision of a source of complement (Goodfellow et al., 2005; Halstead et al., 2005a; 2008). Despite this, at least for particular disease subtypes, notably AMAN and Miller Fisher syndrome (MFS), a considerable body of evidence suggests that auto-reactive anti-ganglioside antibodies arise via a process of molecular mimicry following infection and drive complement-mediated damage to the peripheral nervous system (Goodyear et al., 1999; Perera et al., 2007; Yuki, 2007; Yuki and Kuwabara, 2007).

Continued next edition.....

Call - a - Friend

Hello friends, as you can see our Editor, Christine, has presented us all with a brand new layout of our newsletter Recovery. It will be a surprise for me to see but I know Christine has put a lot of time into this "new look" Recovery, therefore I am confident it will be great! Good reading!

Communications whether by email, mobile phone, text messages on mobiles, land line telephone or by ordinary mail, all methods of contact in today's life are acceptable and commonplace. To accommodate the dedicated mobile phone user the GBS Association has added a mobile contact number for your convenience.

Although 2014 is relatively new, the Association has received a wide range of calls, ranging from the mother of teenager to a senior all on their recovery path. At no age is it easy to manage one's life during the months or even years following GBS or CIDP rehabilitation.

Possibly childhood GBS onset is even more infrequent and uncommon than the instances suggested in adulthood. Nevertheless its effects can be just as overwhelming, for a child or teenager as well as for an adult. A young person often misses months of school work, finding themselves floundering when rehabilitating to "catch up" with their studies and school work and not being able "catch up" physically as well. Not to be able to walk quickly, or play sport, or even go out with class mates or friends even for a short time, leaves a young GBS patient isolated and for a time vulnerable emotionally as well as physically. No amount of reasoning that this time will pass, and recovery will hopefully happen, lessens the impact of GBS on the young. As a parent, realization of the physical and the emotional effects being endured by one's child is shattering. I am not sure there is any complete answer to this situation. Perhaps the only answer is time, time to heal, time to accept, time and patience. A tall order to suggest when one is only thirteen years old!



*"the phone number is
0487 843 723 and we
hope to hear you
'calling-a-friend' "*

I mentioned in a previous paragraph about one of our "seniors" who phoned, let's call him Errol. Yes Errol rings me now and again, just for a chat on this and that. He is currently a "senior" living alone and has had GBS just a few years ago. He is optimistic, positive, though struggling a little with GBS residuals, you all know the story, pins and needles, fatigue, weakness, all a familiar state of affairs, I'm sure to you all. Well Errol rang me on such a "high" some weeks ago re a medication he has been taking, (a doctor's prescription) which has given him a new lease on managing those usual GBS problems. Errol wants me to share his discovery with you all. I said I would do so, guardedly. To keep my promise completely it would mean I would have to mention the medication, this I cannot do publicly for ethical reasons. If any members would like to learn more about Errol's medication please contact the Assoc. I will give it to a personal call from a member only. Please remember I do not, or the Assoc. recommend this medication in any way or suggests it is suitable for GBS/CIDP or any other condition. All I might add is, for Errol it seems to have given him such relief. May I also add Errol is still in the rehabilitation period of his recovery, one might rightly also assume progress was as much to do with time as it might be to the medication. Wishing you all good health until next time.

Mary

Back Page Bits n Pieces

Thank You!**SMARTPRINT** for donating printing and labelling of our Newsletter**GBS Association of NSW**
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Please let us know if you would like to volunteer for your Association*We need your help to really make our Association supportive and effective. We are here for you – all on a volunteer basis. Can you be there for those who are going through what you did, or are still going through?*

Name:

Address:

Address:

Phone / Mobile:

email address: (if would you like your Newsletter via email)

☐ Hospital or home visits to new sufferers (remember how you felt)

Preferred area:

☐ Telephone contact (be a GBS or CIDP friend by phone)

Preferred contact number:

Committee Meetings

All are welcome to attend the GBS Association of NSW Committee meetings. Newly diagnosed and people recovering from GBS and CIDP will appreciate the contact, encouragement and support from fellow members.

2014 Meeting Dates			
5 th April	3 rd May AGM	5 th July	1 st November

Financial Year 2014

Members are reminded the Association's financial year is

1st January 2014 to 31st December 2014

GBS NSW would appreciate your continued support.

Disclaimer

Information presented in "Recovery", GBS Newsletter is intended for information sharing and general educational purposes and should not be considered as advising or diagnosing or treatment of the Guillain-Barre Syndrome or any other medical condition. Views expressed in articles and letters printed in Recovery are those of the authors and do not necessarily reflect the opinions or Policy of the GBS Association of NSW Inc.

Public Risk

The Guillain-Barre Association of NSW would like to inform all members, friends, guests and readers that the Association no longer has Public Risk insurance covering association meetings or association functions. We regret that due to increased costs we were unable to renew our Public Risk Insurance.

Contact the Editor

Do you have an interesting story to share with your fellow members? Perhaps you would like to share your experience with GBS/CIDP with us by writing your story for 'Recovery'. Maybe you just need some more information on an article appearing in the Newsletter? Whatever it may be you can contact Christine Simpson-Morgan:-

Mail: 8 / 36 Mobbs Lane EPPING NSW 2121**Email:** smorgan8@bigpond.net.au