The 2013 Annual General Meeting and Conference

Caroline Morrice

This year we met in the De Vere Village Hotel on the Wirral on 20 April. Many people arrived on the Friday where we gathered for an informal meal and chat.

The day started with a review of the charity in 2012 before the main business of the AGM.

Caroline Morrice – The support of patients and their families remains at the heart of our work.

In 2012 we had 500 requests for support, 75% with GBS, 4% with Miller Fisher syndrome, 20% with CIDP and 1% unidentified diagnosis. Of these 198 asked to speak to a local contact, 29% of the total, and 73 local contacts were employed. This is an increase on last year, but we are still finding that many people rely on the internet and using social media to chat with others in a similar situation. Although the use of the internet and social media does not suit everyone, as a charity we need to ensure that our information is available to all and in a format to suit those affected by the illnesses.

The helpline received 800 calls in 2012, a drop of 17% over previous year. Although we have seen a drop in calls coming through, the fact is that the facility is there, and is extremely important. If we only spoke to one person and helped them it makes it worthwhile. We used 20 volunteers to man this valuable resource.

The welfare fund continues to receive requests for support, and we have been able to offer help to many families again this year. Sixteen applications were received of which 13 were approved, 1 rejected and 2 withdrawn.

The Branches around the UK continue to be active, but as ever there is scope to increase the number of Branches across the UK.

We have seen an increase in the number of volunteers coming forward with 20 being taken on during the year. This ensures that we have a good mix of people available to help out when needed. Local contacts are provided with a pack of information about all the variants as they may have to visit someone with a slightly different diagnosis to the one they are familiar with. On the down side we have more volunteers than people to visit; maybe these volunteers might like to consider how else they can support the Charity like the Helpline, starting a branch or creating a fundraising team. We have plenty of ideas so please ask us what else you could do.

Membership has dropped overall by 52 during 2012 and friends have increased by 308.

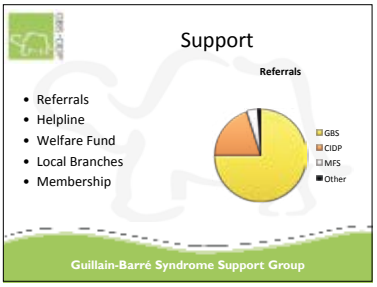
The internet is now our most important first point of contact for those seeking help and information and the new website is almost ready and will be up and running for July. It will have a fresh look and greater capability; one of the new elements will be to allow the newsletter to be received electronically.

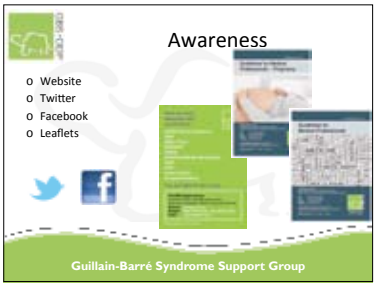
The GBS Support Group site on facebook continues to be a major forum for discussion and now has over 2000 members. This forum is monitored by a number of people and they do their best to answer questions and weed out the problems.

Twitter is also a popular tool for spreading the word and we are looking at ways of increasing our followers and utilising this to support the Charity.

The information booklets are still issued to all new referrals and this information is up to date as we have a rolling programme of review by our Medical Advisory Board. New information leaflets are considered and will be added to the list as appropriate. Again many people download the information from the internet and the new website will make this a little easier to achieve.

2012 saw a drop from last year, however last year there was a legacy of £110,000 which we have not repeated this year! Given the current economic situation fundraising activity is very good, and it is worth noting that most money raised for the group is by people that are not registered with us. Many





remain anonymous, choosing to use an on-line fundraising site such as Justgiving or Virgin money giving.

An amazing range of events take place throughout the year to raise funds, from cake sales to the more energetic cycling, running and climbing events. However you raise money, we sincerely thank all of those people who raised funds in 2012 and who continue to raise funds for the Group. In fact just before we left the office yesterday we received £1000 from a livery company.

Finally, and yes I know I mention it every year for which I make no apology, we have all heard of Gift Aid and would like to remind people what a difference gift aiding your donation makes to the charity's income. As long as you are a UK tax payer you can tick the gift aid box. This allows us to get "free" money from HMRC worth 25pence for each £1 donated. In 2012 we received an additional £5972.22 as a result of gift aid. 2013 changes to the system will now allow us to claim gift aid on the proceeds of collection boxes, so please ensure that you complete the form and return to Lesley everytime you empty the box. This has the potential to boost our funds and we should never miss an opportunity to receive money from the tax man!

So what does the future hold? We are always in the market for applications for new trustees so if anyone is interested please have a chat with me and I will get an application pack to you. It is a voluntary role and one that anyone with an interest in the governance of the charity would find rewarding.

2016 marks the 100th anniversary of the pioneering research carried out by Guillain and Barré. Some of our supporters have started fundraising against the Centenary appeal but we still have a way to go to reach the £1m we would like to use for essential research. If you have any ideas or a desire to scale Everest or the cake stall at a garden party, Lesley would love to hear from you.

We have four guaranteed places in the cycling event RideLondon-Surrey100 2014. This event attracts top cyclists on the back of London2012 which started people thinking 'sport' so if you or anyone you know wish to take part we would love to hear from you. You don't have to be Bradley Wiggins or Victoria Pendleton to take part – obviously beating them would get you noticed!

We wish to continue developing the ways in which we can support patients and members alike to ensure we can reach all affected by these illnesses. The introduction of a mobile website to run alongside our new website is happening and embracing Facebook and Twitter must be a part of the future.

More recently we have received a lot of media coverage through the national and local press, with one story making it into all the national papers and also on to BBC tv and radio in Yorkshire and Lincolnshire. Also on the back of that we were on the BBC South East tv news talking about another case. I am aware that some of you saw these broadcasts and I hope that we will be able to build on this around the UK.

We are continuing to target the medical professionals with information, but we do rely on members and friends to let us know of any gaps so that we can add them to the mailing list.

The newsletter *In The Know* has over the last couple of years changed from a simple newsletter into a full blown glossy magazine and so it has been decided that *In The Know* and *Reaching Out* will be amalgamated and the Summer Issue will be a bumper conference issue. Please keep sending your articles for the magazine to Lesley; you will find the deadlines for submissions in each publication.

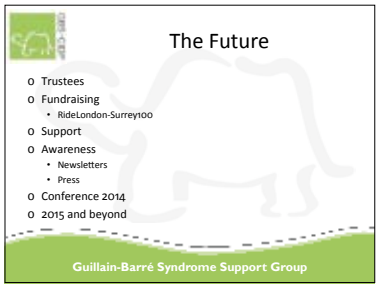
The 2014 conference will be held on 17 May in The Village Hotel – Swansea. Any suggestions you make on the feedback form that can help with planning next year would be welcomed.

Obviously 2016 is due to be a big year and the conference will be held in Scotland on the weekend of 25 June; planning has started and we will give updates as we get nearer the event.

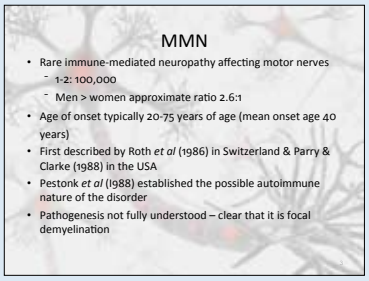
We have to consider the future conferences and 2015 may be the time to hold an AGM with a lunch and with a speaker ; there will be more about this in the October magazine.

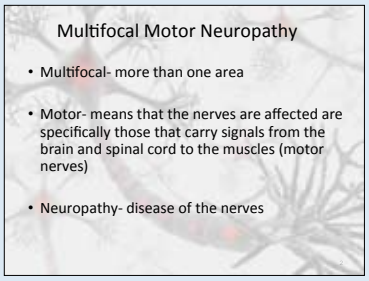
One of the proposals put to the members resulted in the simplification of the membership categories being adopted. This gives members and supporters the option of receiving all their correspondence by email. We can report that members and supporters renewing their subscription have responded well to the new system, with 25% opting to receive newsletters by email.

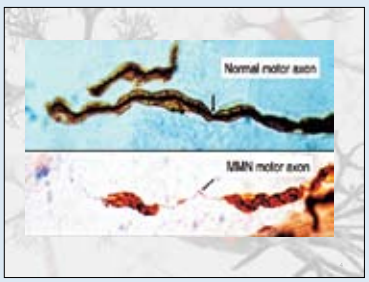
The proposal to convert to a Charitable Incorporated Organisation (CIO) was adopted and is scheduled to come into being on 1 January 2014. Watch out for details on an additional General Meeting taking place later this year to formalise the changes. The new CIO would come with a new name which embraces all those people we aim to support and help – Guillain-Barré and Associated Inflammatory Neuropathies – GAIN. A competition to design a logo was held and your opportunity to vote for your favourite entry is at the back of the magazine.



Amanda Woodhall







Amanda Woodhall on Multifocal Motor Neuropathy (MMN) – this is a rare disorder affecting only motor nerves. About 1 in 200,000 have it; typical age of onset is 40. It was first recognised 1986. It presents with progressive limb weakness, normally in the hands and fingers. For diagnosis, we look for antibodies in the blood, protein in the spinal fluid, and nerve conduction block.

Unlike CIDP there is only one treatment: IVIg. Plasma is taken from about a thousand donors and mixed to produce IVIg. The treatment works for from two to eight weeks; most patients need treatment at varying intervals. We're not exactly sure how it works. It's very expensive so its use is governed by the Department of Health to ensure it's only being used for the right patients.

In 2008 we began switching MMN patients to subcutaneous immunoglobulin (Subcut). They can then manage treatment at home. It's less intrusive, but we need expert patients – patients who are able and willing to undertake their own infusions. On the whole our patients on subcut are doing very well, and we are about to start CIDP patients on subcut. Michael Tooze, one of my patients, will talk about his experience.

Michael Tooze: I'm now 67, a retired company director. I was diagnosed sixteen years ago with MMN, despite which I'm still going strong – I play golf, do Scottish dancing, gardening, and I volunteer with the NHS, so I stay fairly busy.

Twenty-five years ago I had a new company car and found I couldn't push the button on the hand brake; I assumed the problem was just that it was a French car. Then I got twitching in my shoulder, which I still have. Then began problems with my left foot – it was then that I went for medical advice. The only further deterioration I've had is lifting my arm in certain ways, but I have learned to adapt. That's what this disorder is all about – adapting. My left foot has improved because other muscles have taken over, so I'm no longer limping. I was on BUPA, who had never heard of MMN.

IVIg was an all day event; I had to wait round for a doctor to cannulate me because at the time nurses weren't allowed to do it. When it was determined that I had a chronic condition, I moved my care from BUPA to the NHS.

Amanda put me on more concentrated IVIg, then in time invited me to join home treatment – which has been a very positive step. Every two months they telephone me to determine what I need and then deliver it to my home. Amanda gave initial training. [He shows the pumps and indicates where he infuses the Ig.] I can put the pump in a bag that I carry round my waist, then can walk around: it's much more convenient than going to hospital for infusion. Rather than nurses having to record batch numbers, etc., Baxters gave me a smart


Michael Tooze

phone to provide information to Amanda.

There are a number of benefits with subcut: I can control when and where I have treatment; I don't get peaks and troughs as I was when having treatment every five weeks because I can have treatment every week. Early on I had a few side effects – swelling, etc. – as my body got used to it, but I persevered and it's working very well now. It's stabilised my condition. My hope is that this helps with NHS expenses. I'm very concerned about health care in the community – I write software for this, working to get patients out of hospitals and into home.

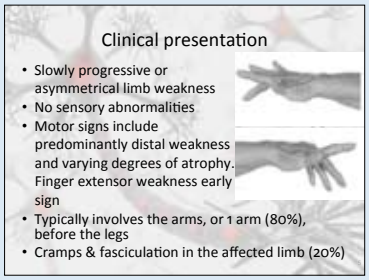
Question: Can you drink alcohol in moderation?
Michael: I think I'd be abusing the system to be overdrinking, but I can drink a bit. I've had to adapt in some ways. I've found that MMN has improved my golf in some ways – the way I swing my arms sometimes gives me a better hit.

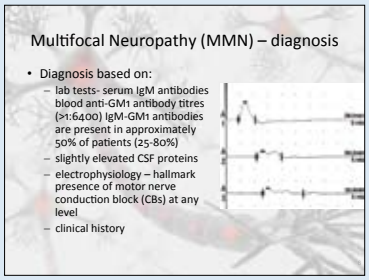
Question: What about foreign travel?
Michael: I can take everything with me.
Amanda: one patient is an underwater photographer who travels all over; he takes everything with him. He carries a letter explaining why he has the drugs and advises airlines ahead of time, and so far has had no problems.

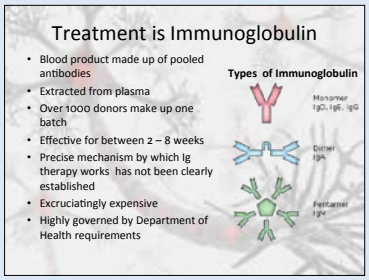
Question: Does the Ig need to be refrigerated?
Michael: I have a fridge, but the product will last a long time without refrigeration.
Amanda: I would advise patients to take the drugs in a cool bag to avoid high temperatures. But it won't be problematic.

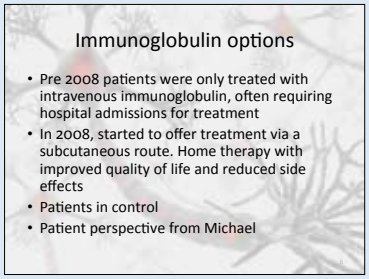
Question: How do you know it's working?
Michael: I could tell I was going down on treatment every five weeks. With MMN if you lose a part of your body it's liable not to come back, so it's important to hold stable. When I was in Nigeria in the heat I found I didn't go down as badly, but in colder climes I need level treatment.
Amanda: I do on occasion challenge patients to try to reduce their dose to see if they can do without the full amount.
Michael: I can sense when I'm going down. I'm concerned about the cost but also want to maintain a good quality of life.

Question: Does MMN progress to other muscles?
Michael: it seems to: right now it seems to be affecting my right hand. But I'm also getting older. Of course, if I start forgetting dance steps I'll start to worry!
Jamie: I am a CIDP patient and on subcut and would encourage everyone to try. It does not work for everyone.









John Winer

Jane Pritchard

Hugh Willison

Ask the Experts

Members of our Medical Advisory Board Chairman – Dr John Winer, Queen Elizabeth Hospital; Secretary – Dr Jane Pritchard, Charing Cross Hospital; Professor Hugh Willison, University of Glasgow; Dr John Nixon, Lancaster; Dr Shakti Agrawal , Paediatrician Queen Elizabeth Hospital; and Dr Claire White, Kings College London were joined by Dr Bart Jacobs, Netherlands to answer questions from the floor.

Question: We hear that there are only a few people annually who contract GBS, but now it seems more. Is it becoming more common?

John Winer: Probably not. Sometimes the numbers depend on the database being accessed. And it may be that perhaps there are more some years, and fewer in others.
Jane Pritchard: There are clusters and outbreaks on occasion.
Hugh Willison: More and more people know someone who has had GBS: it's not unusual in seventy years of life to meet people who had it. And there is more interest in rarer diseases these days, which brings more cases to light. We are better, too, in the West diagnosing it; but in Africa, etc., other diseases overwhelm the numbers of GBS patients – they have to deal with malaria, TB, other more common diseases that we don't see often in the West. But in some countries there are more incidents of GBS, e.g. Asia with axonal forms while more demyelinating forms in Britain. It's a big project to gather information.
Bart Jacobs: I have worked in Bangladesh, and half of intensive care units are filled with GBS patients, but it is possible that perhaps all severe cases of any illness are gathered in that hospital. It would also be that illnesses that bring on GBS are more common in those countries.

Question: More vaccinations, more GBS?

Shakti Agrawal: In paediatrics we don't know if it's vaccinations or that children are more likely to have infections, etc., which bring on GBS. In our area are more children who travel outside the UK. But regarding the relation of vaccinations to GBS, we haven't seen any that are definitive.
Bart Jacobs: The swine flu thirty years ago in the USA indicated that there was something in the vaccination that triggered GBS. But to make a definitive statement we also need to know the background of the flu and the drug, and it needs to be remembered that the more new vaccines, the more likely we'll see side effects. Often it is more advantageous to have the vaccination even at a risk.

Question: Is the change from Vigam™ to Privigen™ good or bad?

John Nixon: Each product differs in how it's made. There's no clear evidence that one product is better than the other; some are licenced when others aren't because of lack of clinical studies. I have found that some patients note improvement or worsening; individuals may find differences, and sometimes doctors can even note the differences. We simply use the new product for a time to see how it works and move the patient back if necessary. Some centres only use one brand, others may use several brands.
Jane Pritchard: Some PCTs rotate brands because of supply.
John Winer: We don't really know what in IVIg works, so can't determine which one works best. And even different batches of the same brand differ – some have less of the active ingredient.
Hugh Willison: We don't know how it works, and have problems determining whether patients are better or worse – for CIDP it's hard to put a hard number on patients, and hard to know if improvement is from treatment or from the disease itself changing. Ig costs about £30 per gram, so about £1,500 per treatment, so we as prescribers are constantly under pressure to control prescription.

Question: For a person with IVIg, what about using steroids and steroid-sparing agents. What are the side effects?

John Winer: There's been a big discussion on this, but no consensus of thought whether to use a single type of treatment, or to add additional drugs. If you need a high dose of steroids, it might be better to also have an immunosuppressant to cut down the steroid. Which immunosuppressant to give: there is no clear guideline yet. We want to set up databases of CIDP patients to try to determine this. The starting steroid level is about 60mg, then reduce to get a maintenance dose which is as low as possible. If a patient requires a have high dose, perhaps it would be better to have a sparing agent.


John Nixon


Shakti Agrawal


Claire White


Bart Jacobs

John Nixon: There are different choices for different people. I have more on IVIg than on steroids, but that is much based on patient desire, often based on side effects, etc. For immunosuppressants, we try to work out whether it works or not, e.g. I use methotrextate but if in a few months it doesn't work, I take the patients off it.
Jane Pritchard: Most people know the side effects of steroids and we can do a lot to counteract them, but others we have to monitor more closely, and some side effects put people off. IVIg isn't without side effects for some people.
Bart Jacobs: We start with IVIg usually, but with a couple who didn't work we found steroids have worked – it makes us wonder if maybe CIDP isn't a single disease but several related ones which responds to different treatments.

Question: How do you deal with a doctor who doesn't believe there's anything wrong with you?

Jane Pritchard: Keep going back, or see a different doctor.
Hugh Willison: One problem in clinical medicine is that doctors are going to be wrong on occasion. GPs can't refer people for every symptom. The art of medicine is to determine when something will go away on its own. With GBS some have nonspecific symptoms. Both patient and doctor need to be persistent.
Jane Pritchard: Ask GP for second opinion, or if possible ask the neurologist for a second opinion.
Shakti Agrawal: In neuropaediatrics, in a young person or child who presents with inability to walk, etc., it can be difficult. I have a two-week old with GBS. Diagnosis is not necessarily black and white. We get referrals from other paediatricians who aren't sure what's wrong. Are there functional changes? We need to do investigations, and if everything appears okay then may have to consider psychological evaluation.
John Nixon: Things can be missed but there's a better understanding now than when I was in medical school. It's still not very straight forward, but understanding of these diseases is improving.
Bart Jacobs: Sometimes a doctor doesn't recognise longer-term symptoms; sometimes a GP doesn't recognise muscle cramps that return some months later. There's a book of some 250 pages dealing with GBS – it's a bit too much for a GP who may never see a case. Shorter material written for doctors is very helpful.
Hugh Willison: Myasthenia gravis takes several years to diagnose – the main symptom is feeling tired. How many here had problems being diagnosed?
Delegates: One patient says she took three months; others were diagnosed by junior doctors when the disease was missed by neurologists; some say that they have continuing problems although the doctors say there's nothing wrong with them – some days you feel fine and others awful.
Hugh Willison: GBS leaves irreparable damage, so residual symptoms are well known. When the doctor says there's nothing wrong, they often mean there's nothing they can do about it. We also need to be aware that there is recurrent GBS or that it can change to CIDP, rather than assuming that the patient is just suffering residuals.
John Winer: Sometimes information given is old, while better information is available.
Delegate: Patients are very frustrated sometimes: GPs can be lethal when they don't know that they don't know; everything needs to put in writing rather than just talk so they'll have to respond.

Question: In Miller Fisher, what causes hallucinations? I was in hospital for nine months.

Hugh Willison: This sounds like MFS with GBS. Intensive care GBS patients often have hallucinations.
John Winer: Drugs, the constant lights in ICU, etc., can cause hallucinations in any patient, but it's perhaps worse for neurological problems.
Delegates: I had hallucinations even without the drugs; you can't communicate that you're having hallucinations to doctors; I thought members of staff were trying to kill me while I was in ICU; you lose track of reality.
Jane Pritchard: If doctors don't know you're having hallucinations they can't do anything to help you. Often those on vents are sedated. Infections can also cause hallucinations – many things can cause them.
Bart Jacobs: Doctors should ask their ICU patients if they're having hallucinations. We need to pay more attention perhaps.
Delegates: It seems a lot of people have visions of death and hell.

Question: Regarding MFS, why do I have a lack of hunger and thirst after nil by mouth and on a peg?

Jane Pritchard: That's not something I've ever come across. The drive to want to eat and drink is from the central nervous system. Most patients with pegs still want to eat.
Caroline Morrice: I came across a site that deals with people with alternative forms of feeding. www.pinnt.com Patients on Intravenous & Nasogastric Nutrition Therapy

Question: Can we donate our bodies to science in an effort to help find a cause and cure?

Jane Pritchard: Tissue banks are available, but I don't know of anything for GBS/CIDP. Routine post mortems don't normally take nerve tissue.
Hugh Willison: We prefer to study patients while alive. We've done a lot of study of end of life GBS patients, etc., but not at the early stages: these are where we need studies. But we can't do some tests on live people. Additionally, nerve biopsies aren't done easily. We really need other methods to examine nerves.
Bart Jacobs: Biopsy means that the nerves don't work again. But in skin nerves the nerves can regenerate. We have done some skin biopsies in the acute stage of GBS to try to understand what's going on at the beginning – we can sometimes determine if the patient will have autonomic problems by skin biopsies.

Question: Is there a connection between GBS/CIDP and diabetes?

John Winer: Peripheral neuropathy is common in diabetes, and some with diabetes also have CIDP – it appears to be more common than by chance. Perhaps the genes for diabetes also cause CIDP.
Jane Pritchard: Sometimes with patients with diabetes it's hard to differentiate between that and CIDP.
John Nixon: If I had a CIDP patient with diabetes, I would be more careful about treatment because of steroid use.
Hugh Willison: Statistics can be confusing, and trying to differentiate between what's causal and what's coincidence is hard.
John Winer: CIDP is more common amongst diabetics.

Question: What are the ten most important facts about GBS and CIDP from a medical standpoint?

Bart Jacobs: No two patients are the same: cause, preceding infection, prognosis, etc.
Jane Pritchard: It's necessary to measure vital capacity – even without machine indicators, we need to determine whether or not the patient needs ventilation.
John Winer: Start treatment as early as possible to prevent nerve damage.
John Nixon: GBS can be mistaken for other diseases, so the patient needs to be seen by neurologist as soon as possible.
Hugh Willison: We really do need to make a huge amount of progress is determining the cause – until then we can't find a cure.
Shakti Agrawal: Pain is always a presenting symptom in children and needs to be managed aggressively. Once the child is walking we have to determine on-going problems, particularly pain and residuals.
Hugh Willison: What do patients say is most important?
Delegates: Listen to patients; don't forget physio; let the person know there's life after GBS; nurses need to understand what patients can and can't do – often they don't think you're trying hard enough; need to communicate with family; listen to parents in cases of children with GBS or CIDP.
Claire White: Patients need to have physio, and the rest of the team need to work with the patient, particularly in pain control.
Shakti Agrawal: We like to think we always listen to parents, but sometimes there's a lack of communication, particularly when the acute and critical stage is over. On the general ward, there are not as many doctors and nurses.
Delegate: My son got info from GBSSG that enabled him to ask the right questions.

Question: Is 2016 still the goal?

Hugh Willison: That year is a big anniversary which is why we set the goal for that year. If a hundred years after the disease was first described it could still not be cured, we need to work harder. Progress so far has been remarkable, but it's tiny little bits of information that add up. I don't think we're too far away from an answer, but the real challenge is to convert that to new treatments. I'm optimistic that we'll improve treatment, and that toward the mid part of this century we'll learn more of regenerative methods – there's much work being done on the brain, but not on peripheral nerves. Although the peripheral nerves will regrow, they don't always grow normally.

Research Updates

Claire White

Claire White: HINT – starting in June 2012 a study of the effect of exercise on longstanding residual neuropathies was undertaken. The benefits of general exercise are well known: reduction in health problems normal for older people, improvement in mood and wellbeing. This is known to be true in specific health problems. But will exercise work with inflammatory neuropathies, particularly aerobic and strengthening exercise? Thus we have undertaken a trial of prescribed amounts and duration of exercise. Thus far we've found clinical improvement and also changes in disability levels.

But we need a large-scale randomized clinical trial to ensure that such exercise will be helpful generally and cost effective. We need volunteers who have had disease for some time and are stable and who are not getting much on-going treatment – we hope to look at those with residuals, who have difficulty with some everyday activities. We're looking for those who are generally recovered but not badly affected, etc. They need to be able to walk a short distance without help. We will contact their doctor to ensure their diagnosis and that there are no other health problems. The trial will mean that some patients get advice only, others will get a full clinical assessment and individualised specific exercises. We will be able to provide some funding for gym activity, may be able to hire a static bike if necessary.

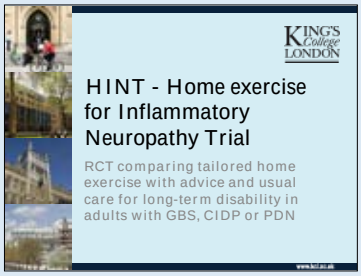
Jane Petty is doing part of an assessment, a questionnaire, looking for the impact of the diseases on everyday life, particularly such things as fatigue, mood, quality of life, how much physical activity the patient can do and any improvements you've seen, and how you feel about your disease. Do you use walking aids? How does it impact on employment – how does it impact on society? At twelve weeks we'll re-evaluate for immediate changes, and do assessments periodically thereafter.

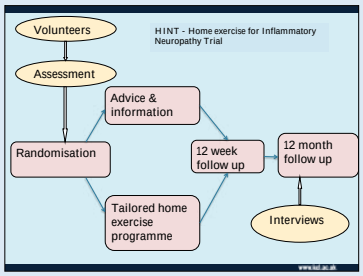
We would be interested in anyone who would like to join. We need seventy people at least – we have fifty-six so far, with thirty-nine screened for eligibility. The patient will need to be within two hours of London.

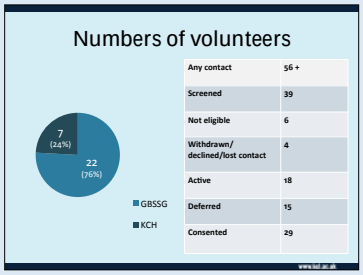
We won't have data for evaluation for another eighteen months at least – we hope in two years to have results regarding the usefulness of exercise. We have already discovered that we need more information on people's ideas of their own health, what's it's like living with the condition and exercise, so we want to do another small study. What is your experience of living with the disorders? These need to be people who aren't part of the main trial.

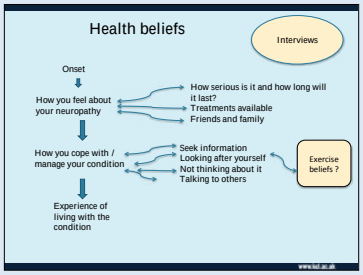
Question: Is the objective to be fit enough to return to work, etc.?
Claire White: Not necessarily, but we aim to see if exercise will help with the return to more normal life.

Question: With the government looking at benefits, if I could resume work they might take my benefits away.
Claire White: We're aware of that so are doing discussions of the cost effects as well before taking on patients.









What to do if you are interested

- Talk to one of us at lunch or teabreak – 15 volunteers for interview about health beliefs
- Contact us at: claire.white@kcl.ac.uk or on 020-7848-6331
- Look out for adverts on www.gbs.org.uk

Hugh Willison

Govind Chavada

Bart Jacobs

IGOS: the way forward for GBS research and discovery (a summary of their conference presentations).

Govind Chavada, Bianca van den Berg, Hugh Willison and Bart Jacobs.

IGOS was introduced to the GBS Support group at the Liverpool AGM in April 2013 by Dr Govind Chavada, our IGOS UK clinical research fellow jointly funded by the GBS Support Group and the University of Glasgow. Dr. Bart Jacobs, international leader of IGOS from the Rotterdam GBS Research Group, Netherlands, also attended the meeting in Liverpool, along with Professor Hugh Willison and Dr. John Winer who are also both involved in coordinating IGOS UK. Dr. Bianca van den Berg, the clinical research fellow in Netherlands who coordinates international activities and co-authored this article was holding the fort in Rotterdam. After the AGM, Bart, Govind and Hugh returned to Glasgow for further in-depth discussions about IGOS plans for the future. A four hour car journey with non-stop debate about GBS ensued.

Although we have achieved a huge amount of progress in our understanding of GBS over recent decades, in terms of cause of disease and available treatments, there are several major issues which still remain unsolved. One of the major aims of IGOS, the International GBS Outcome Study, is to comprehensively address these issues on a worldwide scale, bringing together patients and researchers in a manner that has never been achieved before. Many local and national surveys and studies have been conducted on GBS and gathered extremely useful data in the process. It is on the shoulders of these studies that IGOS has been constructed. In doing so, we can truly state that IGOS is a worldwide first, and hopefully sets out the model for future studies that will inevitably follow over the coming decades.

What research questions is IGOS addressing, and hoping to achieve answers to?

Firstly, GBS still remains a clinical diagnosis in that there is no single test which can confirm the diagnosis. This means that clinicians have to rely on patient history and clinical examination findings that do not always point in the right direction, as many support group members will know from their own experience of the early phase of the disease. Lumbar puncture and nerve conduction studies can be normal in the initial stage of GBS, which doesn't help the diagnostic situation. These factors may lead to delay in the diagnosis and treatment and therefore there is an urgent need for development of the early diagnostic marker of the disease, ideally in the form of a simple blood test.

Secondly, GBS has a variable clinical course in different individuals. Whilst many patients completely recover, others are left with severe residual disability. In the initial phase of the disease, around one third of cases require intensive care therapy. In the longer term, about 20% of GBS patients do not recover well, remaining unable to walk 1 year after onset. In up to 5 % of patients, GBS is fatal. Currently there is little information available to predict the disease course and overall outcome in individual cases and therefore we need good prognostic markers which can reliably identify patients with poor outcome so they can benefit from clinical trials of more intensive treatment.

To address the above important issues, a group of peripheral nerve specialists within the group called Inflammatory Neuropathy Consortium (INC) have launched a worldwide observational study called International GBS Outcome Study (IGOS). IGOS is a web-based registration system that provides the framework to recruit at least 1000 GBS cases around the world over a 3 year period. IGOS has been funded by a range of organizations including GBSSG UK, GBS/CIDP Foundation International, and in Glasgow by the University of Glasgow and by grants to Hugh Willison from The Wellcome Trust.

The aim of IGOS is to identify the factors that are responsible for the varying clinical and pathological characteristics of the disease in different patients. This is achieved by collecting and analysing the clinical and laboratory data during the course of the disease over a one year time scale. Analysing these data will allow researchers to predict the disease course and its outcome in groups of patients and in individual cases, and to develop biomarkers of disease. This is important because it will help doctors to identify patients destined for a poor outcome earlier in the course of disease than is currently possible. Such patients might benefit from additional therapies.

Another major aspect of IGOS is to develop biomarkers for GBS. Principally, these are laboratory tests looking at immune, genetic and nerve factors that predict the course, subtype, and severity of disease. For example, researchers have recently identified antibodies in the blood of GBS patients that are believed to be directly responsible for damaging nerves. Currently their precise roles and predictive value in GBS are unknown. To investigate these and other biomarkers further, blood samples are required from GBS patients. The unique genetic makeup of individuals may contain vital information on how nerves withstand injury and how well they are able to regenerate. By correlating these laboratory data with clinical information we aim to identify the underlying significance of these biomarkers in relation to particular disease characteristics.

How far have we got with establishing IGOS?

IGOS has a web based data entry system. After almost 2 years in the planning stage the website was officially launched in May 2012 (www.gbsstudies.org). At present, 177 centers from 18 different countries have expressed their interest in participating in the project, which requires local ethical and administrative approvals. Out of 177 centers, 77 centers have achieved this and are actively recruiting patients. 117 patients have been recruited in to the study so far and majority of them have been in last 6 months. This was due to increase in the number of participating centers. In the UK, we have 20 centers which are actively recruiting patients. As of June 2013, the UK has recruited highest number (38) of the patients into the study, followed by Denmark (18), Netherlands (17), USA (17), Italy (12), Germany (6), Japan (4), Spain (3) and Belgium (1). In the UK, we hope to recruit several hundred patients into the database.

IGOS has made remarkable progress over last 12 months. As more centers are coming on board we are expecting a substantial increase in the worldwide recruitment figures over next two years. IGOS will make an important contribution to the existing information available to predict the disease outcome and development of new biomarkers. Eventually such discoveries will lead to better treatment planning for patients with GBS.

This has been a great opportunity to introduce IGOS to the UK GBS community, and also for the study coordinators to meet and further develop plans. The IGOS UK team are extremely grateful for the financial support that the GBSSG has provided. The results of IGOS will be published over the coming years. The IGOS team has been delighted with the interest in the projects from patients, the vast majority of whom have consented to be involved. It is important to note that patients are only able to be recruited into if they are within 14 days of the onset of GBS, as the database is designed to include data from all stages of the illness, early and late. Fully or partially recovered patients are not eligible for entry, but may be the subject of future research projects.

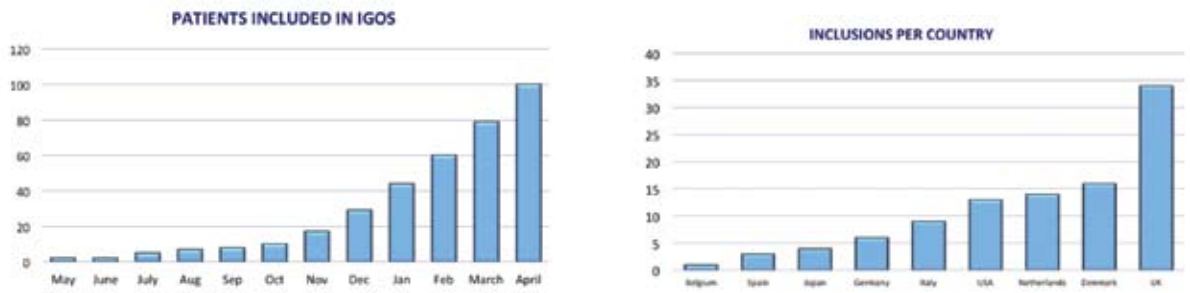


Figure legend: Information on the patient recruitment into IGOS up to April 2013. The countries involved are listed in the upper figure, alongside the patient recruitment numbers. The lower figure is the total number of cases recruited into IGOS worldwide, between May 2012 and April 2013

My Blog

Andrew Markham

Andrew outlined the findings of his personal survey on GBS and CIDP. He has a blog which will give more information www.mycidp.blogspot.co.uk