

Issue 67. June, 2009.

GBSCIDP

INFORMATION

STICK WITH IT SLOW BUT SURE

NEWSLETTER OF THE IN GROUP: THE INFLAMMATORY NEUROPATHY SUPPORT GROUP OF VICTORIA INC.
Supporting sufferers from acute Guillain-Barre Syndrome(GBS) & Chronic Inflammatory Demyelinating Polyneuropathy(CIDP)

WINTER LUNCHEON

Margaret and Doug Lawrence are again hosting the Winter Luncheon on July 26th at 12.30pm.
\$15 per head. Come along and enjoy good food, good company and good information.

We are having a guest who can answer your computer questions and also advise about "Voice Activated"
computer programs. All welcome. RSVP: Margaret on 9802 5833 or Melva on 9707 3278 by July 22nd.

**ANNUAL GENERAL MEETING: SUNDAY, 16th AUGUST, 2009, AT 1.30PM
BALWYN LIBRARY MEETING ROOM, WHITEHORSE ROAD, BALWYN
GUEST SPEAKER: BILL STEVENS, FITNESS INSTRUCTOR
AQUA AEROBICS AND HYDROTHERAPY**

Bill Stevens became a full time Fitness Instructor after a 40 year management career in the Australian wine industry.

When he retired in 1996, he made Fitness Instruction his second career later adding Aqua Aerobics and Older Adults Fitness to his qualifications. He is now teaching around 25 classes per week covering young and old, fit and not so fit participants, including working with people with disabilities.

Aqua classes led to working in hydro pools where the warmer water helps people recovering from injuries or suffering from a variety of conditions, such as arthritis. Movement of joints, painful on land, can be achieved by participants, leading to increased mobility. **Gentle water exercises can provide almost a "miracle"**. He is constantly amazed by the results and is rewarded with the happiness and smiles of his clients.

We look forward to meeting Bill and learning from his knowledge and experience.

Notes from May meeting.

After a small general meeting discussing topics such as guest speakers and computer access and information, we watched a video on GBS then were delighted by the video of the concert organized by **Anna Melville** to acknowledge the wonderful generosity of **blood donors**. What a wonderful night it was.

Over our usual cuppa the members then chatted and although there were not many members on the day those present enjoyed the afternoon. We welcomed member Marjorie.....

90TH BIRTHDAY CELEBRATIONS FOR JAMES AND BETTY GERRAND.

Prior to the meeting members Barbara Clifford and Joe and Melva Behr visited the home of **James and Betty Gerrand** where celebrations were underway for their **90th birthdays**. Both James and Betty were in great spirits in the company of their family and friends.

The 'IN' Group owes its very existence to James Gerrand and it is with sincere gratitude that we wish this wonderful couple many more years of happiness. Well done.

(This must be a tribute to Betty's cooking! Her nut loaf was always a favourite.) **All the best to you both.**

NEUROLOGISTS

Following the publication of Issue 66 we received a number of enquiries from members wanting to know of neurologists interested in GBS and CIDP. Members from country (and city) areas would like the name/s of neurologist/s who see **private patients**.

Our members are willing to travel for an appointment. Can you help? Do you see a neurologist who could help? Please phone Melva on 03 9707 3278. Thank you.

GEELONG SUPPORT GROUP GET TOGETHER

The next Geelong support group gathering will be at John & Margaret's home in Highton at 2pm on Saturday 15th August.

Newcomers can RSVP Margaret ph:5244 5311 or Dee ph: 5244 3382.

E-mail Mailing List

If you would like to be included on the IN Group email mailing list please send an email to John Burke at the following email address **jburke@contracts.com.au**

If you use *hotmail* or have junk mail filtering software running you will have to include the above email address in your "safe list" otherwise *hotmail* or your junk mail software is very likely to delete our emails.

Disclaimer Information presented in "INformation" the Newsletter of the Inflammatory Neuropathy Support Group of Victoria Inc., is intended for information only and should not be considered as advising or diagnosing or treatment of Guillain-Barre Syndrome, CIDP or any other medical condition. Views expressed in articles are those of the authors and do not necessarily reflect the opinions or Policy of The IN Group.

Telephone contacts over recent weeks.

Marlene from Queensland phoned asking for information as her 15 year old daughter was being tested for CIDP. This young girl has been deteriorating since February. Hopefully treatment will be given very soon.

Member **Peter** phoned and had a great chat and is making headway since being treated for POEMS. Peter is walking around the block with the aid of a frame and is doing well on his stationary bike as well as lifting weights which has helped build up his upper body.

Peter mentioned he takes a Parkinson's disease drug 'Siferol' for his legs. Previously his legs would not settle at night, requiring to be stretched, then cramping, then stretching, then trying to find a comfortable spot (many members would relate to this situation) but since taking this drug he can now get a good night's sleep.

June Cathcart phoned regarding an article she was doing for the local newspaper, as did **Jan and Bernie Pettit** who were being interviewed by a journalist for the Leader group of newspapers.

Both June and Jan and Bernie had articles published. They were very good. Thanks to all who sent out the information to their local papers. We may see further articles during the year. Again, thank you all for your support.

Pat phoned. She had read Bernie and Jan's article. Pat has peripheral neuropathy and hopes to attend our next meeting.

Trudy from Melbourne phoned to say her sister-in-law, only 17 years old, was totally paralysed with GBS in a Perth hospital. We spoke for some time and she later phoned to say the young lady was now being ventilated. At least they were prepared for this eventuation. Her husband has flown to Perth to be with his sister.

I phoned **Naomi** a young girl who had been in the same position with GBS approx. 18 months ago in S.A. and was told by her brother she had just moved to Perth. The world turns in amazing ways. Naomi was anxious to help and will visit as soon as possible. Thanks Naomi and it's great to learn you are getting on so well.

A gentleman phoned recently diagnosed with GBS who had been treated quickly by casualty in Melbourne, given 'Intragam' and was now home. He was experiencing a rash.

Headaches and rashes seem to be the most common side effects reported to me following IVIG treatment.

Tammy recently told me that prior to her treatment they give her a pain killer and now she doesn't experience the headaches.

It has been a busy but rewarding couple of weeks. No doubt what we hear about is only the tip of the iceberg as this season is a particularly difficult one.

AGENDA

ANNUAL GENERAL MEETING
Sunday, 16th August 2009 at 1.30pm
Balwyn Library Meeting Room
366 Whitehorse Road, Balwyn.

1. Confirmation of Minutes of 2008 AGM.
 2. Reports from President and Treasurer
 3. Election of Officers and Members of Committee.
 4. Any special business of which 21 days notice has been given.
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POSITIONS TO BE FILLED:

President, Vice-President, Secretary, Treasurer, Public Officer, General Committee Member/s.

NOMINATION FORM

Position:

Nominee:(Please print)

Nominated by:

Seconded by:

Signature of Nominee: Date:

To be returned to: The Secretary, The IN Group, 66 Eva St., Clayton, 3168
by 13th August, 2008.

All positions are up for re-election. If you have time to devote to The 'IN' Group, we would welcome your nomination.

All positions are voluntary but the rewards are great.

DATE TO REMEMBER: 29TH NOVEMBER, 2009 AT 12.30 PM.

**ANNUAL CHRISTMAS LUNCHEON AND DUTCH AUCTION
\$15 per head**

**Dr. Andrew Kornberg will be present to give us an update on his research.
If possible, a small wrapped gift for the Auction would be appreciated.**

NZ CONFERENCE

Following the New Zealand Conference, we were generously sent the following information which had been presented by Professor Gareth J. Parry, from the University of Minnesota, a world authority on GBS/CIDP, especially the associated pain that some patients experience.

GBS/CIDP IN CHILDREN

- GBS in children is rare but has been described as young as the first year of life.
- Some studies suggest that it tends to be milder in children than in adults.
- Treatment is the same as in adults but IVIg is preferred because of ease of administration.
- IVIg can be given subcutaneously if there is difficulty with venous access.
- Prognosis is excellent and residual disability is very rare.

- CIDP in children is rare but has been described as young as the first year of life.
 - The most common form resembles GBS because it tends to come on quickly, over 2-3 months.
 - If it comes on slowly, particularly in the very young (pre-school), it manifests as “failure to thrive”:
 - Loss of milestones or slowing of attainment of milestones.
 - Unable to keep up with peers in play.
 - Loss of sporting ability.
 - Sensory symptoms are typically absent, possibly because of reporting bias.

More likely (than in adults) to run a chronic monophasic course with resolution, with or without treatment, over months.

- No difference from adults in response to treatment but steroids are relatively contraindicated because of effects on growth.
- PLEX is difficult because of difficulties with venous access.
- IVIg becomes the basis of treatment in most children.
- Remission is common during adolescence.

- A 7 year old girl was noted by her parents to be unable to keep up with her siblings during play.
- She had several falls.
- Several trips to her pediatrician simply resulted in reassurance.
- Eventually (~1 year later) taken to the Emergency Room following a fall that resulted in a bad ankle sprain.
- She had no complaints but the ER doctor thought it was odd that she did not have any reflexes and called a neurologist.
- Neurological examination showed significant proximal and distal weakness.
- Nerve conduction studies showed typical changes of CIDP.
- CSF analysis showed high protein.
- Treated with IVIg for 5 days with initial excellent response but quickly relapsed.
- Treated with IVIg on 2 consecutive days every 2 weeks which maintained her strength but repeated attempts to reduce the dose failed.
- Pulsed steroids added at age 10 which allowed IVIg to be reduced to once a month.
- Slow withdrawal of all treatments over subsequent years.
- She is now 16 years old and is on no treatment. She functions completely normally and has normal height.

ACUTE MOTOR AXONAL NEUROPATHY

There are several different kinds of GBS:

- Acute inflammatory demyelinating polyneuropathy (AIDP)
 - Acute motor axonal neuropathy (AMAN).
 - Acute motor and sensory axonal neuropathy (AMSAN).
 - Miller Fisher syndrome (MFS).
 - Acute sensory neuronopathy.
 - Acute autonomic neuropathy.
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- The commonest form of GBS in developed countries is AIDP.
 - In under-developed countries (China, India, Central America) AMAN is a common form.
 - In AIDP the myelin sheath is the primary target.
 - In AMAN the axon is the primary target.
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- Some axonal damage is invariable in AIDP and may be severe even though the primary target is the myelin sheath.
 - Axons are injured in an “innocent bystander” reaction.
 - If axonal damage is minimal the prognosis is excellent because the myelin sheath can be reconstituted rapidly and completely.
 - If axonal degeneration is severe the prognosis is poor because the “scaffolding” of the myelin sheath is destroyed and there is no guide for the regenerating axons.
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- Although the primary target in AMAN is the axon, the myelin sheath degenerates because it needs the axon to survive.
 - Prognosis is good in AMAN because the “scaffolding” remains intact and provides a guide for the axons to regenerate.
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- AMAN more likely to follow diarrhea (C.jejuni).
 - AMAN more likely to have specific anti-nerve antibodies (GM1 ganglioside).
 - More severe cases have sensory nerve involvement (AMSAN).
 - Clinically indistinguishable from AIDP but the electrophysiological features are different.
 - No difference in the treatment.
 - No difference in prognosis.

PULSED STEROIDS IN CIDP – AN UPDATE

- We have now treated more than 40 CIDP patients with pulsed steroids (methylprednisolone 500 mg once weekly).
- The “permanent” remission rate is about 60%.
- Patients who remain on steroids are generally on lower doses (100-300 mg once weekly).
- Adverse effects remain annoying but manageable:
 - Insomnia (70%).
 - Irritability (60%).
 - Heartburn and indigestion (30%).
- Older patients showed a reduction in bone density during treatment, despite co-administration of calcium and vitamin D, and treatment with bisphosphonates should be considered.
- Only one patient has stopped treatment because of AE’s.
- Treatment is most effective if started early:
 - All but one of the patients started within 2 years of the onset of weakness is in remission.
- Treatment can be used to reduce dependence on IVIG in patients who have been started on that treatment first.
- We are currently preparing a study comparing different doses to see if a lower (250 mg) and therefore better tolerated dose would be equally effective.

A NEW TREATMENT FOR CIDP

- CIDP is an auto-immune disease in which antibodies and activated lymphocytes (white blood cells) attack the nerves.
- Rituximab is a monoclonal antibody that specifically targets a protein on the surface of the class of lymphocytes that produces antibodies and kills these cells.
- If the antibody-producing cells are eliminated the CIDP should improve.

RITUXIMAB IN CIDP

- We have treated 4 CIDP patients with Rituximab:
- All patients had been treated with IVIg, PLEX and pulsed steroids, alone and in various combinations.
 - 2 patients had been treated with cyclophosphamide.
 - All patients were continuing to respond to treatment but only if very high doses and frequent treatments were used.
 - All patients tolerated the rituximab treatment without adverse effects.
 - All patients have been able to reduce other treatments following treatment with rituximab.
- A 48 year old woman with diabetes was diagnosed with CIDP in 2003.
- Initially treated with IVIg with a good response but repeated attempts to reduce the dose were unsuccessful.
- Pulsed steroids were added in 2004, again with a good response, and the IVIg dose was reduced to one infusion every 2 weeks.
- She remained stable for more than a year but repeated attempts to reduce the steroid dose were unsuccessful.
- She then relapsed and steroids were increased to 500 mg twice weekly with some improvement but her diabetes became very difficult to manage.
- IVIg was increased to once every 2 weeks, then once a week and then twice a week, always with improvement but the increasing doses were of major concern.
- Despite twice weekly IVIg and twice weekly steroids she continued to lose function.
- PLEX was substituted for IVIG but did not result in further improvement.
- In 2006 she had a one year course of chemotherapy (cyclophosphamide) and during that time both steroids and IVIg were able to be reduced for 2007 and early 2008.
- By mid-2008 she requiring twice weekly steroids and PLEX to maintain function.
- In November 2008 she received 2 doses of rituximab 2 weeks apart.
- April 2009 she is receiving MP 250 mg once weekly and PLEX once every 2 weeks.
- She is working fulltime, has normal strength and her energy is good.
- Rituximab may prove to be an effective treatment for CIDP and warrants further study.
- It is an expensive treatment but much cheaper than IVIg and only needs to be administered every 6-12 months.
- Long term safety is a concern but it has been used in rheumatoid arthritis for many years with few problems.

A NEW TREATMENT FOR GBS

- In auto-immune diseases, antibodies bind to the target tissue and then recruit a variety of chemicals that damage that tissue.
- One of the chemicals that is activated during this process is *complement* and there is abundant evidence that activation of complement damages nerves in GBS.
- Eculizumab is a monoclonal antibody that inhibits activation of complement and should reduce tissue injury in GBS (and other auto-immune neuropathies).
- Human studies of eculizumab in a non-neurological auto-immune blood disease have shown a major protective effect and this drug is now approved for treatment of this disease.
- Human studies of eculizumab in myasthenia gravis (an auto-immune disease of muscle) have started in the US.
- In a laboratory model of GBS in mice, eculizumab had a major protective effect.
- Human studies of eculizumab in GBS are being planned.
- How do you study a new drug in humans with a disease for which effective, but imperfect, treatments already exist?
 - Combined treatment (IVIg + eculizumab) versus IVIg alone.
 - Eculizumab versus placebo as initial treatment followed by IVIg.

VACCINATIONS IN GBS/CIDP

- GBS is a disease that may be triggered by any event that stimulates the immune system.
- CIDP resembles GBS in so many ways that it may also be triggered in the same way.
- Vaccinations are specifically designed to stimulate the immune system to recognize an invading organism.
- Vaccinations may occasionally trigger GBS:
 - 1976 swine ‘flu vaccine resulted in a marked increase in GBS numbers.
 - No other ‘flu vaccines have been associated with an increased incidence of GBS.
 - One nationwide polio vaccination program was associated with a slight increase in GBS cases.
 - A recent meningitis vaccine may have triggered some cases of GBS.
 - Occasional individuals may develop GBS after the ‘flu vaccine as an idiosyncratic reaction.
- Anecdotes suggest that vaccinations may occasionally trigger a relapse in CIDP.

Should patients who have had GBS in the past or who have CIDP be vaccinated?

- If GBS was *clearly* associated with a vaccination there is no evidence that revaccination is contraindicated but it seems prudent to avoid the vaccine that triggered the initial event.
 - The GBS should have appeared 1-3 weeks after the vaccination.
 - There was no other antecedent event.
- If such an individual had a disease that would put him/her at high risk of serious complications of the 'flu (COPD, HIV, cancer, etc), the risk of triggering GBS should be weighed against the risk of being getting the 'flu.
- If GBS was not associated with a vaccination it is suggested that all vaccines be avoided during the first year following onset of disease.
- Vaccines are probably not contraindicated in CIDP:
 - The risk of triggering a relapse is small
 - CIDP does not usually cause such severe weakness as GBS.
 - Even if a relapse occurs it will respond to treatment.
 - Patients with severe CIDP, especially if it has run a relapsing course, might be wise to avoid vaccinations.
- *In all cases the risk of vaccination should be weighed against the risk of the disease for which the vaccine is being administered.*

GBS/CIDP UPDATE

Summary:

- GBS and CIDP can occur in children. It should be treated no differently from adults and has a good prognosis.
- AMAN is a form of GBS that is clinically indistinguishable from the more common form (AIDP), is treated in the same way and has a similar prognosis.
- Progressive inflammatory neuropathy resembles GBS or CIDP and is caused by exposure to aerosolized pig brains.
- "Pulsed" steroids remain the preferred treatment for most cases of CIDP and induce remission in ~60% of cases.
- New treatments are emerging for both GBS and CIDP.

Internet Contacts

With our Website www.ingroup.org.au now up and running we are receiving enquiries. One was from Suzanne from Queensland who has CIDP and other complications. Her neurologist is interested in using 'Mabthera', (Rituximab) and she wondered if any of our members with CIDP had used this drug. If you can help phone Melva 03 97073278 or email behrsden@optusnet.com.au.

THE 'IN' GROUP

Th Inflammatory Neuropathy Support Group of Victoria Inc.
Supporting sufferers from acute Guillain-Barre` Syndrome (GBS and
Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)
Registered No: A0025170R

Annual Subscription

Subscriptions due on the 1st July of each year.

1/07/09 – 30/06/10.

I am happy to help The 'IN' Group by my membership.

Annual Subscription \$ 15.00

Other Items

Booklets - **The Road to Recovery A-Z** \$6
- **Boy, Is This Guy Sick** \$2
- CIDP \$2
- GBS \$2 \$

Donation to support medical research \$
(Donations of \$2 or more are tax deductible) _____
(Tick if receipt required)

Total Payable: \$ _____

Enclosed is a cheque/money order (payable to The IN Group)

Membership Details

Name: _____

Address: _____

_____ Postcode _____

Telephone: (Home) _____ (Work) _____

Email Address: _____

Signed: _____ Date: _____

Thank you. Please forward this form along with your payment to:
The Treasurer, The IN Group, 26 Belmont Rd., GLEN WAVERLEY 3150