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INFORMATION

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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) and other Inflammatory neuropathies

Dates to Remember:

Sunday, August 13th 2.00pm Annual General Meeting
Guest Speaker: Neil McCoy – His Journey with CIDP/Using Social Media for The 'IN' Group
Sunday, December, 3rd 12 noon Annual Christmas Luncheon
Guest Speaker: Assoc. Prof. Andrew Kornberg

Notes from the June Meeting.

President Margaret Lawrence: Welcome everyone. I see we have some **new members with us today. A special welcome to Valerie, Peter and Ken** who are going to give us a talk on both **Valerie's Fifteen Years with CIDP and the New Zealand Group and the Conferences** there, so that will be great and very interesting. A couple of little items: **I would like to thank John Burke who sends out the notices on line to say what is going on. This is excellent and he is very happy to keep doing it for us. The other person is Neil McCoy who has started up our "Meet Up".** Our Meetup landing page is (people are encouraged to click the "Join us" link and "RSVP") - https://www.meetup.com/IN_Group_AU/. **I have looked at it and it is great because it is moving us on with the World today. So thank you Neil for all your work too.**

Doug: I second that as when I'm working on the Website Neil bales me out. It's great.

Finance. The finance is for the **quarter ending March**. It is a fairly quiet quarter seeing the **subs are due on the 1st July, so we do need to remind everyone that our year starts July 1.** We received just $45 in subs but we had against that **$140 in donations. That is a great** ratio still continuing on. Book sales of $16 and **Craft sales (again by Gwen** and her dolls) we collected another **$110** from the doll sales and this is ongoing. Total $365 against expenses of $345 with $19 in front for the quarter but in that time we paid $107 in Internet Costs and Newsletter of $138 for that quarter.

I will announce that **we have received a grant from CSL for the Website costs and that has gone into our account for the new quarter.**
At the moment we have $2844. We have a reasonable carry over considering all our inflow will commence from July 1st with the subs and the donations that come with it, so we are in quite a reasonable position to **hopefully repeat what we have been doing for the past 15 years, allowing us to make a donation to Assoc. Prof. Andrew Kornberg for research.**

Margaret: I must say that we had friends over the other day and they presented me with $50 as a donation to the group. They said they like that there is none syphoned off into other costs.

One thing I spoke about last time was how **difficult it is for us to get speakers**. I have actually been in touch with **Gerald Quigley** (you probably know him) he is a pharmacist who also has herbal medicines. He sent me back an email but unfortunately he flies to Sydney every Sunday to do radio programs. I feel he is interesting enough that it may be a good **idea to have a meeting on another day as he is a very good speaker.**

I tried others including Red Cross and Gilmore shoes but was unsuccessful. We keep on pounding away at these things and see how we go.

Talk by Valerie Simpson

I call my talk **FIFTEEN YEARS WITH CIDP**.

I hope it is relevant and of some interest. **I have long accepted that my CIDP is very atypical**. **Even when my neurologist Tim Day came to speak at the group he kept saying "Of course that doesn't apply to you Valerie". I wish something did,** but I'll tell you my story and hope that it resonates somewhere.

INITIAL DIAGNOSES AND HOSPITALISATION

I started experiencing symptoms at the beginning of 2002. **We lived in Tennant Creek, in the Northern Territory** at the time and **when I was sick** I used to go to the **Aboriginal Health Centre** because **all the best doctors** (like Richard Di Natale) worked there.

I remember the day I went it was 10 o'clock in the morning and **a few of my students (high school girls) were sitting in the waiting room. When the neurologist said "Valerie stand up and close your eyes and walk in a straight line", I staggered and they were shrieking "Our Mrs. She's drunk already!"**

The doctors at the Aboriginal Health Centre knew there was something very wrong so I came south to Melbourne. I was diagnosed in November after neurologists did a nerve conduction test. (We call it the "Cattle Prod Test").

The initial diagnosis was **GBS.** Subsequent clinical history (repeated episodes) led to a **revised diagnosis of CIDP.**

That was the start of **two years in Royal Melbourne Hospital**: two years moving between the ICU, the neurology or respiratory wards and the Rehab campus. **At my worst I was paralysed and on a ventilator**. What was **frustrating was the times I went backwards after I had made progress. This happened many times.**

Question: Were they giving you 'Intragam' then? **Val:** It didn't work.

When I was good I was at the rehab hospital stomping along between the parallel bars, but **at my worse I was totally paralysed 24/7 on a ventilator**. The **first intensive care was 7 months** and **they never heard my voice. I was on a ventilator all the time**.

The treatment I had during those 2 years in hospital included '**Intragam**' which made not a blind bit of difference, **Mycophenolate** which didn't work, **Methylprednisolone**, and **Cyclosporine.** None of them made any impact on the condition. **Something that did have a positive impact was** Plasma Exchange**. It never cured me but it did sometimes stop me getting worse and when I was really bad it was good not to get any worse.**

They eventually stopped the neuropathy in its tracks using Cyclophosphamide. When I came out of hospital **I would go and have regular Cyclophosphamid**e. I was finally **discharged at the end of 2005**.

LIVING AT HOME

At first we were living in a **charity house set up by Rotary**, but moved into our **own apartment in June 2005 and just last week Peter cooked me lobster (my favourite dinner) and champagne as it was 12 years of living in a community in our own apartment.**

IN REMISSION

Now when I'm in remission and living at home **I get around the house with a walking frame,** so I have a bit of mobility as I walk with the frame, or sort of stumble around with it.

I sleep on a ventilator because my respiratory muscles don't work when I go to sleep so Pete plugs me in every night to the ventilator.

I get help with wonderful carers who come every day to shower me, to dress me, and to help with the housework.

Peter looks after me absolutely splendidly and we have also found a wonderful Respite Facility where every carer has had to deal with ventilators. Peter can have a break or if he has to go to hospital, I have got a respite facility to go to.

CIDP is a chronic condition and since 2005 I have had seven more episodes, each of which has landed me back in hospital. Sometimes we know what has caused it **flu, flu shot, trauma of extended plastic surgery**, (I had an operation on my hand) or, possibly, the **stress** of watching someone close die a slow and painful death. Other times we don't know at all.

I know when it is happening when I experience loss of function. We have drawn up **a chart to monitor** this. Drawing on my Early Childhood teaching experience we have **three headings: Always, With Difficulty and Not At All**. We grade functions like "feeding myself" or "holding my head up". **As more ticks move to the "Not At All" column we know it is getting serious.**

My neurologists know me so well and trust me that **when I say "It's on again" they start the treatment.** They don't muck around doing tests etc.

WHAT HAPPENS WHEN I HAVE AN EPISODE

I get slowly **more paralysed** and often end up bedridden.

I am given Cyclophosphamide, which takes four weeks to start working. In that four weeks I just lie around waiting for my Lazarus moment (which, miraculously always comes). After 28 days I stand and walk. It is a miracle. It is just amazing.

If I get dangerously feeble, I have to have plasma exchange as well. This has its own problems: access needs to be created into a major blood vessel and the lines put in very often get infected. Often I'm in Intensive Care by then. As I said, the plasma exchange doesn't cure me, but it pulls me round for a bit and stops me deteriorating further.

Recovery has been harder after every episode. Cyclophosphamide is severe stuff and its side effects are hard to live with. My hair gets unattractively thin and I experience general feebleness. Harder to cope with is passing blood in my urine.

Just this year I have been referred to **Urologists and Gastroenterologists** to **follow up on some of these side effects**. Just two more speciality areas to add to **my list that now includes Neurologists, Respiratory doctors, Dermatologists and Intensivists. I fear that soon I shall have to add Ophthalmologists** to that list. Something funny is going on with my eyes.

Wonderfully, all these people have kept me going; all these **specialists** and **Peter.** While I am disabled and ventilator dependent, I plod (metaphorically) on. I don't really know if the health problems I have are related to the CIDP or to the treatment I have to have for it. Maybe it is just because I am getting old. **I'm certainly often tired and fatigued. This I know is a classic symptom of CIDP.**

I reckon I've had only four good months every year for the last six years: one month in hospital, a second month on Cyclophosphamide, six months recovering from the episode and the treatment, then my four months in the sun. I have come to love April, May, June and July.

MY NEW TREATMENT

After last year's admission **the neurology team at Royal Melbourne initiated a review of my case.** Peter and I ended up in front of **a lecture theatre with tiers of specialists** arranged in front of us at the Neurology Grand Round, while one of the **doctors presented an overview of my clinical history**. We felt a bit **like guinea pigs**. When the presenter got to **'Episode 12'** I felt hysterically giggly. It was ridiculous.

There was some consternation amongst the people in the Auditorium when one of the doctors said to the presenter **"How many doses of Cyclophosphamide has Valerie had?** And she said, "Oh I didn't count." I said, "Well I know." **So I gave Peter my piece of paper and he counted them and said "33" and they gasped because it is a terrible drug.**

Discussion with the whole neurology team led to the decision to start a new treatment.

The new treatment uses **a drug called 'Rituximab'**. It is described as **immunotherapy or immune therapy**. You just get hooked up and **they drip it into you**.

I have had two infusions, two weeks apart. I gather that the drug binds to some of the B cells, or leucocytes, rendering them ineffective. We want them ineffective because these are the cells that attack me. It does mean that, with my immune system is suppressed, I am vulnerable to infection. That, however, is not new. The wonderful thing about 'Rituximab' is that, unlike Cyclophosphamide, it is not a toxin. It didn't make me feel ill or nauseous. Not so far, anyway.

When they told me when the second one was I said "Oh. I can't be here for that. It is my husband's 70th Birthday and people will be coming from interstate." So they said, "Alright Valerie we will do it the day before."

I was **nervous about a new treatment** and when I told one of the doctors how worried I was she laughed and said, **'Valerie! Don't waste a moment worrying about 'Rituximab'. After what you've had before, 'Rituximab' is a dream!** People on it love it!'

Kind Peter stayed with me throughout the first infusion (I had even taken our Air Viva because I read that it could cause difficulty breathing). As it turned out, I suffered nothing worse than boredom.

We are very optimistic. If I can get through this winter without another episode of CIDP it will seem a miracle. I have already put the date of my next 'Rituximab' in my calendar: Thursday 16 November at 10.30. I am almost letting myself hope I'll make it!

I hope to stay strong enough to keep doing the things that make my life rich and rewarding. I love being a Grandma. I love swanning around to local coffee shops and cafes. I love the volunteer work I do at the Royal Melbourne Hospital. I love reading and talking about books, both at my Book club and informally with a wide group of bookish friends and I value the joy and happiness of my life with Peter and our marvellously supportive family and friends.

Peter Simpson: That's what it has been. It has been a long journey thus far and there is more to come with this 'Rituximab' and we will just have to wait and see if it works, which we want to believe very much it will.

INTRODUCTION FOR KEN DANIEL

Valerie: **We first met Ken in the New Zealand Support Group. When I got sick 15 years ago, my sister rushed off and joined the New Zealand Support Group because she wanted to learn about it and understand something of the condition.**

She is very important. **She has become the Patron of the New Zealand Society and in fact she worked with Ken to organize a recent Conference in Rotorua because she is now the Mayor of Rotorua.**

KEN'S STORY.

I have a nice story, probably similar to most of you. **9 years ago, almost to the day**, I was visiting grandchildren in Nelson. Actually I will go back one week. I was visiting other grandchildren in Christchurch and **I had campylobacter** which was a bit unpleasant. I got over it pretty quickly but a week later I was visiting other grandchildren and woke up one morning (we were at a motel) went to go to the toilet and **I couldn't open the door handle. I thought, "Something is funny; something is not right.** It must have been something I'd eaten or someone was trying to poison me". I called out to my wife and asked if she was still happy with the way I was managing and I was satisfied she didn't poison me.

My wife is a nurse and she actually realized by the way I was walking that something wasn't quite right. She said, "**We had better get on a plane back to Wellington**" which was where we were living. Within a matter of hours as I went to get on the plane and when at the top of the steps I fell backwards over all the passengers behind me. That caused a bit of a stir.

A very helpful hostess came out and said, "Are you alright?" I said, **"I think there is something wrong with my leg".** My wife was pushing me up and by the time we arrived in Wellington airport **I couldn't walk at all. She managed to get me to the hospital and by that stage I was totally paralysed**. It all happened within 24 hours. I could still talk, which my wife was not pleased about, because one of the **side effects I had was extreme pain** so I think the rest of the Wellington Hospital knew about it as well.

My lucky break was that it was very quickly diagnosed. There was a neurologist out from Canada who was working there at the time and she put her finger on it right away and said, "**I think he has GBS**." I don't remember a lot about it after that because I was put on quite heavy doses of morphine and other stuff but apparently I had a lumber puncture, didn't have the cattle prod, but **I was immediately put on, we call it immunoglobulin which I believe is the same as your 'Intragam' and they pumped in gallons of this highly precious liquid. They were satisfied at that stage that the attack had finished** and it was just a matter of waiting and rehab after that.

I was shipped off to a neighbouring hospital **for rehab and was there for 3 months. A lot of that time still very paralysed. In some ways looking back on it that was somehow very good for me because I had never been dependent on other people since I was a baby and it does things afterwards for your humility. If you can't feed yourself and you can't toilet yourself you really can't be proud anymore. Looking back it had a positive effect.**

It was a difficult time for me in hospital. My mother died so they had to ship me up to Auckland for the funeral. That was an adventure in itself. Air New Zealand were brilliant using all sorts of equipment I'd never seen before.

Got out of hospital after 3 months but I was very much confined to a wheelchair. I couldn't walk at all and it was about another 3 months before I could start to walk, so 6 months and then I got better and I suppose I am 95% better. I still have a tremor because of the nerve damage. You know, the axon damage on the inside of the nerve and extremes was damaged, so I can't run, can't stand on one foot. I dream about running. It's funny.

While I was in hospital I had a visit from someone from the GBS support group in New Zealand and that was really good because for me and my wife it was very frightening. I was really quite frightened. I had never felt that paralyses before. It had quite an affect on my son who at the time was 14. We realized after a while that he started skipping school. He started just going downhill and he started suffering from anxiety. We had been very close as a family but the biggest effect was on him. So the effect on families can be quite extreme.

To cut a long story short, **I had these visits, a lot of material, a box given to us with pamphlets, etc., and personal visits in hospital from members of the group, so when I got out I joined the support group and went on the Committee and for the last 6 or 7 years I was chairing the Board of the New Zealand group.**

I also chaired the Foundation. The New Zealand group has a Foundation we have managed to set up by salting away some funds from donations and bequests and things and the organization in New Zealand now is that there is a Board. We only meet every 6 months, but are in constant touch, either by video link or emails and phone.

We also have a Medical Advisory Board which we found to be really, really helpful. There is a group led by **Dr. Parry who is a world renowned person in various neuropathies, especially GBS and CIDP.** He spends half his time in Minnesota at the University lecturing then half his time back in New Zealand. **He has been a wealth of information.**

Valerie: He has written a wonderful book. The best book I have ever read on CIDP.

Ken: At every Conference (we have Conferences every 2 years) this
Medical Advisory Board made up of neurologists, psychologists, physiotherapists and various disciplines are all gathered together by Dr. Gareth Parry. **They present papers at these Conferences on latest developments**, causes, what they have found. We have guest speakers.

At the last Conference I went to they had **a guy from the Ministry of Health in New Zealand** who had traced some of the causes. **Why was it that New Zealand has got a very high incidence of GBS? The reason is it starts out with the poultry industry. It has a high concentration of campylobacter (or did have) and they couldn't figure out why.**

Question from new member: Is it a stomach thing?
Ken: Yes. Cramps in the stomach and diarrhoea. It is horrible. It doesn't last long.
Member: You said you had that prior to getting GBS.

Ken: Well some of the research they figured out was first of all why campylobacter was in **New Zealand. It has the highest rate of campylobacter of every country**. They showed charts of Romania and all these other countries **and New Zealand is off the chart**. **The reason** and you can blame this on **the British (**any Brits here – laughter) was once upon a time they **decided they would introduce Starlings to New Zealand** because they were nice birds they thought. There was **a high rate of campylobacter bug in the Starling population and they could track it. They could actually see what the incidence of campylobacter was with the introduction of the Starlings and it went to the chooks.**

Valerie: When we flew back from the New Zealand Conference where we learned that, the hostess came along and said, "Chicken or Fish" and we both said in unison "Fish".

Ken: But **it is great news, because once they figured out what the pathway was. they were able to do something and now the poultry industry in New Zealand, the farms, the processing, the shops that sell chicken, have all started to get their act together in a big way, so the incidence of the bug is incredibly reduced.**

The Medical Advisory Group in New Zealand is really brilliant because **all the members can write to them.** It is all filtered through the Chairman but they are always available for questions and answers like, what do I do about this? Is this normal? What's the story with this? My neighbour has tingly feet. He's just stopped smoking or whatever it is. They filter all these medical questions through.

They put applications in to the Foundation for funding. **All the neurologists, doctors, physiotherapists, whoever it is, will put in an application for research funding programs.** They then sort it out so that **funds** are **allotted according to what they think is the most valuable pathway to get some progress.**

We have got a Website Face-book Support Group as well which is very active. It is a closed group so if you are invited in you can discuss things. There are also in New Zealand a number of centres which have Coffee Meetings where people in say Rotorua, Auckland, Hamilton, and various places might meet just informally say once a month or so. Have coffee and keep in touch with each other.

There is a Newsletter and the Conference every couple of years. Altogether there is from memory, about **250 members that pay a sub of $10-15 a year. There are always donations like you have and additional funds from bequests. We look for people who have got ideas or can support us in any way at all.** We have got publications, some of which have been borrowed from England. **The GBS group in England is quite active** so **some of the publications we have their permission to transfer the material over to ours.**

Margaret: Can I ask how you were able to get all those doctors and link them together.

Ken: It was a chain thing really. It started from the Board as one of the members of the Board was Dr. Gareth Parry. He was the one that knew the neurologists around. He knew what the latest research was. Realize it wasn't just strictly medical. It was psychological as well. The lead nurses in the hospitals, in the intensive care wards and we have had talks about the treatment of GBS and CIDP. Then you realise you have all these people and then the Board said, "Why don't you get together as an advisory Board so that if we have questions we can go to you? We can ask a psychologist, What's the story about Intensive Care? How do the people in Intensive Care get in touch with a support group?"

Question: Is there an immunologist on the Board?
Ken: No, but there is on the Advisory Board. I put Dr. Gareth Parry in that position too as he is in research and immunology.

Len: It seems to be more about immunology.
Ken: Yes it is all to do with the immune system isn't it at the end of the day.
One of the things we have realized is that one of the key factors is early diagnosis.

One of the people on the Medical Advisory Board is a doctor who has had GBS. She actually had Miller Fisher Syndrome where the eyes are affected etc. She has produced a DVD of her time in ICU and the actual physical effects, her treatment and path to recovery. It is absolutely horrible, but great.

There are a whole range of people and they are all flown in free of charge to the Conferences so that they can be there. **There is a lot of advice given at the Conference**. You know you go up to someone and say "Am I getting better? Why have I got pain in my left knee? Is that related", or whatever it is. It is a good mixing of people.

Margaret: You don't raise money for Research do you?

Ken: Not directly. We do get people to donate and to make bequests to the support group itself, but although the Board keeps some funds, things like bequests are really put into a Foundation so that it is separated out and it is that fund that we use for research.

We don't say we are doing research because people have to apply for that as it is accountable. They have to present a paper as to what they want it for, what they are going to do. They have a way they justify their research. **The latest one that Dr. Gareth Parry has been stirring up is having a look at a plant in New Zealand which is a form of stinging nettle. You know how it affects your nerves or it makes you go numb and it hurts but it has something that takes away the pain. It actually has an effect on the nervous system. They have found there is a compound that actually decreases the nerve pain that you can get with GBS. It is being researched. It goes through a long process.**

Well that's me and New Zealand. **Have you any questions re New Zealand?**

Question: Is any 'Rituximab' being used in New Zealand?
Ken: For me it was just trying to flood my system with IVIg.

Question: Do you know anyone else that used it?
Ken: No I don't sorry. They may well do as I have heard that name mentioned at Conferences as one of the range of things being used and tested.

John: Who are mainly the attendees of the Conference? Medical?
Ken: Three sorts. Just counting the people there for the **Medical Advisory Board** and presenting papers and things. There are a group of "victims" the **actual patients who have had GBS or CIDP, their families and their care givers which are very much involved.**

Valerie: When we were invited to speak at their Conference they didn't just want to hear from me, they wanted to hear from Peter as to what it is like to be a carer and what the carer experienced, being both the carer and the advocate. I was mute for months and Peter stood by my bed and asked the questions.

Ken: There is quite a thing that a support group is not just for the patients with GBS or CIDP but supporting the families, the children and also some work with employers to help them understand what the affects of CIDP may be in the future. The third group who come to the Conferences are the staff who is sent from hospitals, medical practices, nursing staff from neurology departments and from intensive care. There is quite a range. It is a lot of fun.

Margaret: Would you know how much it costs to put on the Conference?
Ken: I did**. It is quite expensive but we charge to try and cover the cost. Being every 2 years we have to try and carry a profit through to the next year in case it falls short. The cost of going to the Conference is about $100 for the attendees. They would also have to pay their hotel expenses. Normally we have the Conference within a hotel and meals are provided within that $100. Meals are provided except for a Dinner, often on the Saturday night with some drinks and a guest speaker. We try to make it as attractive as we can because people in New Zealand have to travel a distance.**

Melbourne has the population of the whole of New Zealand so you haven't got the travel expenses but in New Zealand it is awfully long and skinny so people have to travel from the North to the bottom of the South Island. That's an expense they have to bear.

Part of the Foundation which I mentioned also provides financial assistance for people who would like to come and can't afford to. There is money that goes out to people who can't afford to go. That's not open to people from Australia who want to come over for the Conference.

Valerie: The funding isn't but the people could go.
KEN: That's what makes it fun. People from several overseas countries apply, eg. from America who have a very active group of GBS support groups, but people from Australia are always welcome.

Ken: Do you ever have Conferences here or in other parts of Australia?
Val: No because we do have very good medical people as our guest speakers.

Ken: Can I ask how many members you have?
Melva: About 220. We have lost a few because some had been members for 30 years or more and are no longer with us, but we keep at that approximate figure with new people joining and coming along. Most of our members are in Country Victoria and Interstate with a few overseas.

Ken: How do you contact them? Do you have a hospital visiting service?
Melva: We did have a hospital visiting service. We still do have people who go to the hospitals but they usually go when a family member gets in touch with us. These special members meet the family at the hospital and go into ICU with them. Other members talk to others whilst having infusions and pass on information about our meetings and offer support.

Val: It is very hard. I would never be a hospital visitor. Imagine if I stumped up they would think "Oh God. That's what is going to happen to me."

Melva: Our hospital visitors are members who have had GBS and are like Ken, practically completely okay. They are really wonderful and have never not attended when asked. Other times we talk to the family members by phone, sometimes for months and of course send out information.

Ken: We actually have a separate group of accredited visitors. Everyone on that hospital visiting group is accredited so you can go into the hospital and speak to the people on ICU or whatever if somebody contacts you. We have training sessions every Conference for about 4 hours we set aside just for hospital visits – things to do and things not to do. How to approach hospital staff. Information about GBS. What materials are available to actually give them. How can you support them. How can they get in touch. Do your family want to know more information. There is a separate group within GBS New Zealand for that.

Question: Would that be within both public and private hospitals?
Ken: Yes, anywhere but **maybe people with GBS tend to migrate to the public hospitals**. In New Zealand, although we have some very good private hospitals, **the top neurologists seem to be in the main centres.**

WE ALL THEN CHATTED WHILST ENYOYING ANOTHER LOVELY AFTERNOON TEA.

NZ CONFERENCE

Following another successful New Zealand Conference we were generously sent the following information which had been given as a powerpoint presentation by Gareth J. Parry. Gareth Parry, MB, ChB, FRACP, is Professor in the Department of Neurology at the University of Minnesota.

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.

- 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.

- 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.

- 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.

- 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 cyclophopsphamide.
  - 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.

PROGRESSIVE INFLAMMATORY NEUROPATHY ("pig" neuropathy)

- In 2007 several workers in a pork processing plant in southern Minnesota became ill with a neurological illness.
- Symptoms came on acutely (over days) or subacutely (over a few months).
- Weakness and sensory loss mainly affected the legs.
- Reflexes were absent.
- CSF analysis showed high protein levels with no inflammatory cells (albuminocytologic dissociation).
- Nerve conduction testing showed evidence of demyelination with some associated axonal loss.
- i.e.; the cases closely resembled GBS (when acute) or CIDP (subacute).

- All affected individuals worked in the "head room" where they were exposed to aerosolized brain tissue.
- Workers handling the brains or working in adjacent rooms were not affected.
- In 2008 additional cases were identified in Indiana.
- Patients who were removed from exposure improved but some have relapsed when re-exposed.
- Treatment with PLEX or IVIg seemed to hasten recovery

- PIN is a human version of Experimental Allergic Neuritis (EAN), a disorder created in animals to enable us to better understand CIDP and GBS.
- The occurrence of PIN validates the use of EAN to study GBS and CIDP.

STICK WITH IT
SLOW BUT SURE

IN Group

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The 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 SUBSCRITION

Item	Each	Payable
Joining Fee	$10	
Annual Subscription	$15	
(Due 1 July each year)		
GBS Booklet	$3	
CIDP Booklet	$3	
After GBS Booklet	$3	
The Road to Recovery Booklet	$6	
Boy, Is this Guy Sick Booklet	$2	
Recipe Book	$16	
Donation to support Medical Research (Donations of $2 or more are tax deductible) Tick if a receipt is required		
TOTAL Enclosed a cheque/money order (payable to The IN Group)		

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

BSB / Account: **063142 / 10006285**
Account Name: **The IN Group**
(Include Your Name in "Description / Reference")

MEMBERSHIP DETAILS (please Print)

Name:	
Address:	
Suburb:	
Postcode:	
Mobile:	
email	
To receive your Newsletter by email sent an email to John@bal.net.au	

ANNUAL GENERAL MEETING
The Inflammatory Neuropathy Support Group of Victoria, Incorporated
Ashburton Library Meeting Room, High Street, Ashburton. At 2pm on 13/8/17.

Agenda

1. Confirmation of Minutes of 2016 AGM.
2. Reports from President and Treasurer
3. Election of Officers and Members of Committee.
4. Any special business of which notice has been given.

Positions to be filled are: President, Vice-President, Secretary, Treasurer,
Public Officer, Membership/Newsletter Co-ordinator, General Committee Member/s

Nomination form for Committee

Position: ……..

Nominee: ……..

Nominated by: …….

Seconded by: …….

Accepted by: ……

Date: …….

To be returned to: The Secretary,
The IN Group, 26 Belmont Rd., GLEN WAVERLEY 3150 by 5th August, 2017.

Signature of Nominee: …… Date: ……..

Please be advised that notice has to be given on any matter to be raised at the Annual General Meeting.

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.

INFORMATION

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)
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