GBS ……..CIDP **Issue. 84. September, 2013**

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)
26 Belmont Road, Glen Waverley, 3150. Victoria, Australia. www.ingroup.org.au email: info@ingroup.org.au.

THE ANNUAL CHRISTMAS LUNCHEON & DUTCH AUCTION
12.30PM SUNDAY, 17TH NOVEMBER AT THE
BALWYN LIBRARYMEETING ROOM
WHITEHORSE ROAD, BALWYN
RSVP to Margaret 9802 5319 or Melva 9707 3278 by Monday, 11th November.

Buffet Lunch : $15 per person.
The donation of a small wrapped gift for the Dutch Auction with a small tag to indicate value to aid the Auctioneer, would be greatly appreciated.

This is a special day with lots of fun and good company and an excellent buffet lunch.

Annual General Meeting

Welcome: Thank you all for coming and a special welcome to Assoc. Prof. Richard Stark who is going to speak to us later which will be very interesting I know.

Apologies: Irma Hanner, Valerie and Peter Simpson, John De Ravin, Peter Balis, Barbara and Tom Rivett and Barbara Clifford.

Minutes: Accepted.

President's Report:

It is my pleasure to present the 2013 President's Report.

The IN Group has enjoyed another year of meetings and two social occasions which whilst we all enjoy meeting together we are at the same time raising money for research. I am sure the members who attended the meetings gained knowledge from the speakers and also being able to discuss their problems with other members is very beneficial. Those who for various reasons are unable to attend have the excellent newsletters to read the details stemming from the meetings.

My thanks go to the wonderful committee who are most willing in any way to offer their services.

The Vice-President	John Burke. Thank you for your support John.
Secretary	Peter McInness who does a great job with the Minutes. Thank you.
The Treasurer	Doug Lawrence the keeper of the accounts and Website management.
Newsletter,	Melva and Joe Behr. Their excellent newsletters result in great feedback.
Committee Members:	Barbara Rivett who does the Library
	Peter Males. Donation of paper and help with the Christmas Auction
	Brian Boyd. Support and assistance to members
	Len Waters. Willingness and support on any job
	Gwen McInness for her craft work which results in a great cash flow.

In addition I would like to thank Ken Clark who although is not a committee member, but attends the mid-year and Christmas functions travelling from the country to assist with money collection and being our Auctioneer.

I also wish to give a special "**thank you" to Dorothy Brennan** who has for many years collected the library key for our meetings. Dorothy is retiring from the job and **we wish to thank her very much for all her help.**

I am also so **grateful to the many members for their generous donations** resulting in the group being able to present to **Assoc. Prof. Andrew Kornberg a cheque for $8,000 towards research**.

Thank you to our numerous speakers for their wonderful presentations.

The Committee will remain for the coming year and I look forward to their support. I also look forward to being able to assist our members as much as possible in the forthcoming year. Thank you.

Treasurer's Report:

Once again I am pleased to present the Treasurer's Report for the Financial Year 1/7/12 – 30/6/13. Financially we have had another good year with a total income of $12, 200 which is an increase over last year of $5325. Most of the increase results from a State Government Grant of $4000 which covers 2 years but both yearly payments were received in this financial year.

Donations were up by $1,624. Expenses were down $338 which was basically due to the purchase of a printer last year resulting in more in-house printing.

During the year we again made a donation to the Royal Children's Hospital Foundation of $8000 to be used by Assoc. Prof. Andrew Kornberg and his staff for research into GBS and CIDP. Our donations for the years 2006 to 2013 now total $62,000. This is a fabulous result as all donated funds come from our very generous members and the fundraising activities organized by the committee.

Finally on behalf of all members I say, "thank you" to our hard working committee who spend many hours organizing and donating to our various fund raising activities and to CSL for their continuing support towards the website. Thank you.

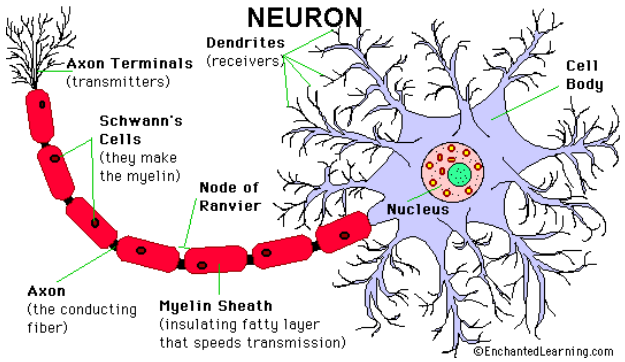
Meeting closed.

Assoc. Prof. Richard Stark. Neuropathy: An approach to diagnosis & management

What I want to do is talk a little about **neuropathy in general and the processes we go through in deciding that someone has an inflammatory neuropathy,** then a bit of why we are so keen to be sure we made a diagnosis of inflammatory neuropathy correctly. Why we ask that is because **treatment for it is specific to that condition**.

The point about **neuropathy: it is a disorder of the peripheral nerve** and peripheral nerves **are unique in the sense that they are so long.** The cells are so long. Most cells are just 10 microns across – 100^{th} of a millimetre - across and they have a nucleus where all the metabolic work is done and it is pumping out chemical things to nourish the rest of the cell. It doesn't have to get the chemicals very far. With **a peripheral nerve cell,** the cell body is sitting up close to the spinal cord, (in the spinal cord for the motor cell and just next to it for the sensory cell) and **the nerve has offshoots that then have to travel all the way to the rest of your body**. If you are going from the base of your spine down to your toes, it is **probably a metre long**. There is a huge difference between that and other cells and there are particular metabolic difficulties that these cells have to be able to do to get **all of the nourishment to the furthest end of the cells**.

There are lots of things that can go wrong with peripheral nerves and there are lots of things that relate to this disturbance of the metabolism and the disturbance of getting things to the far end of the cell. There are a couple of terms we talk about when we are talking about peripheral nerves. The two most important parts are the cell body which is sitting in the spinal cord or near it and there is this axon which is the long terminal that is going out to the periphery. The axon is covered by an insulating sheath, the myelin sheath. In fact the way the myelin works is, when you send a nerve impulse down the nerve, instead of actually having to run along fraction of a millimetre by a fraction of a millimetre, it actually leaps from each of these nodes, to node, to node, which makes the transmission of the impulse a lot quicker than it would be otherwise. If your myelin is not working well, that's bad for you. If the axon of the nerve fibre is not working well, that's not good for you either.



Having the myelin misbehave is actually not quite so bad because the myelin is produced by cells that are sitting next to the nerve fibre, that surround the nerve fibre. You can have those cells recover more easily and can **remyelinate the nerve,** whereas if you get **damage to the axon, that's very difficult to overcome.** It may not regrow, or it may actually have to regrow from where the damage is and if that's the case it re-grows quite slowly, approximately1mm a day. If we have someone with a traumatic injury to a nerve fibre (say having a knife through it and it is sewn together by a surgeon), the nerve will grow back, maybe not 100% efficiently, but it **grows back at about 1mm per day** (an inch a month) so it takes **a long time for recovery to occur.**

It is better not to have **damage** to your nerve at all, but if you are going to have damage it is **better to have it to your myelin than your axon.**

There is **a long list of things that can cause peripheral neuropathy**. These are: -

Diabetes mellitus, Uremia, Porphyria, Hypoglycemia, Vitamin deficiencies, Critical illness, Chronic liver disease, Primary biliary cirrhosis, Primary systemic amyloidosis, Hypothyroidism, Chronic obstructive lung disease, Acromegaly, Malabsorption, Carcinoma, HIV infection, Lyme disease, Lymphoma including Hodgkins, Polycythemia vera, Multiple myeloma lytic type, Multiple myeloma, osteosclerotic, Monoclonal gammopathy of undetermined significance.

That is to daunt you; to tell you there are lots of things and to demonstrate how clever we are to be able to sort out which is which. In fact if you just went through the list you would never do it. The approach we usually take is to look at a number of features at the patient's presentation and to see how they fit in to all of these. That is how we approach someone with any sort of neuropathy and of course the **inflammatory neuropathies have particular characteristics** and that's what alerts us to say **"This is an inflammatory neuropathy**, **like Guillain-Barre` Syndrome or Chronic Inflammatory Demyelinating Polyneuropathy or some of the other rarer types."**

Approach to diagnosis. Tempo
Fibre type
Polyneuropathy or Mononeuritis Multiplex
Proximal or Distal
Pathology.

We will go through each of those and tell you a little about them.

Tempo. The **tempo is probably the most important thing**. By tempo I mean, **how quickly it comes on?** Someone who goes from being perfectly well to being severely **affected within a few hours or days, that's an acute tempo** and it's likely to be **Guillain-Barre` Syndrome. GBS is the classic condition that produces an acute onset peripheral neuropathy**. Most neuropathies are chronic and **CIDP is usually a chronic condition, i.e. it takes weeks or months to develop** and **gradually progresses over that time.**

Some patients with CIDP have an unusual feature of having relapsing neuropathy. They have an episode and get better, then have another episode and get better and if we see that it suggests that it might be a form of CIDP. Again, the inflammatory neuropathies are interesting to you because that's what you have and that is why you are here, but the **inflammatory neuropathies are really important to neurologists** and in fact to all doctors, because **of all the sorts of neuropathies we see, they are the ones that have specific treatment** associated with them. If you make a mistake and you fail to diagnose someone who has a neuropathy that has no treatment it is bad, but the patient has been denied nothing because there is no treatment that works. With

inflammatory neuropathies where there really is treatment, it is important to be tuned in and to get the diagnosis right as quickly as you can. We are always very tuned in to these. There are **things in tempo that would make us think it is an inflammatory neuropathy.** If it's **acute you think Guillain-Barre`**: if it's **relapsing you think of relapsing CIDP** and if it is **chronic it could still be CIDP** but there are a lot of other sorts of neuropathies it might be as well.

Fibre Type. The next thing we think about is **which fibres are involved**. There is a slide later on that tells you the symptoms that are associated with these fibres but most of you would appreciate that the **peripheral nerves have motor fibres (that is fibres that make the muscles flick) and they have sensory fibres.** The sensory fibres in fact have a range of different sorts of fibres in the sensory system. The **large sensory fibres are responsible for things like joint position sense, vibration sense, a little bit of light touch, so if they are affected and you don't know where your joints are it tends to make you unsteady.** There are **smaller fibres** which are involved with **pain and temperatures** and such and it is true that **some neuropathies will affect mainly large sensory fibres and some will affect mainly small sensory fibres, so the symptoms you get from those are quite different.**

The **motor fibres are large fibres.** We sometimes talk about large fibre neuropathies and small fibre neuropathies. You will be aware that **when we examine patients with neuropathy we spend time testing power and physical sensations as well as testing reflexes. The reflexes tend to be significantly impaired in people who have a significant neuropathy.** It **relies on large fibre function** but we do sometimes see people who have, what we call, a **pure small fibre drop**. It is affecting these fibres but not the big motor sensory fibres and they can actually have **normal reflexes but still have a neuropathy**.

The **autonomic fibres, the ones that supply sweat glands, that involve supply to the bowel and to some extent the bladder functions and also affect heart rate and blood pressure control, they can be affected in some forms of neuropathy as well, although relatively uncommon. It is one of the things that happens in patients who have acute Guillain-Barre` Syndrome** as they can get **fluctuating blood pressure**; the blood pressure is going up and down and it is also quite common with people with many neuropathies that have alterations in their sweating, either **sweating too much or not sweating enough**.

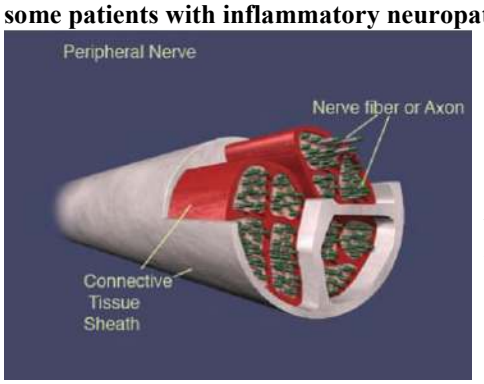
Polyneuropathy or mononeuritis multiplex. Most patients with neuropathy have symmetrical involvement. It involves both arms, both legs, usually the furthermost parts of the arms and legs, but there are some neuropathies where nerves are "picked off" in random. There is a process going on where you get patchy involvement. Mostly this isn't relevant in patients with inflammatory neuropathies. If we do see that we revert to a particular group of conditions that we think about. There is a condition called **Multi Focal Motor Neuropathy with induction block which is one of the forms of inflammatory neuropathies that can start off as a patchy neuropathy.** That would be one of the ones we would think of there.

Proximal or distal. Most neuropathies affect the bits of the body that are furthest from the central core and that's **because the longest nerve cells are the ones most vulnerable to damage.** For most neuropathies that's the case but for **some neuropathies the parts of the body close to the core are affected as well and Guillain-Barre` is an important one of those. People with GBS can within a very sort time develop weakness in their respiratory muscles and that is one of the things we are really tuned in to looking for because obviously if you are not able to breathe strongly it is not good for you.**

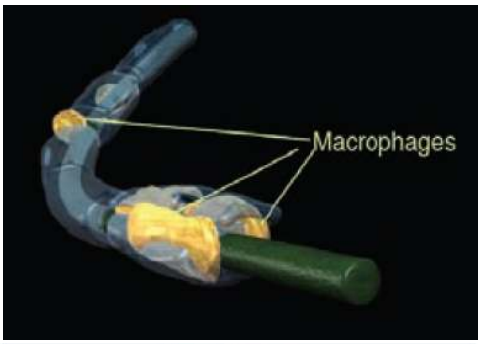
Many **patients with inflammatory neuropathies will have some involvement of the proximal muscles and some of the distal muscles;** proximal meaning closer and distal meaning the further ones.

Pathology. The real thing we are interested in here is **whether it is mainly a problem with the axon or mainly a problem with the myelin sheath**. For most **patients with inflammatory neuropathies it is a demyelinating neuropathy and that's one of the reasons that treatments are most likely to work** because if you can **actually stop the damage to the myelin before there has been associated axonal damage then there is a very good chance of getting a reasonable recovery**. A lot of the treatments we have are aimed at **turning off inflammatory processes, letting the myelin repair and hoping that will happen before there has been associated damage to the axon.** Generally **if the myelin is damaged severely and for a significant period the axon tends to degenerate.** There is a certain amount of **time pressure to get things sorted out quickly** because we really want to get things moving **before that damage happens**.

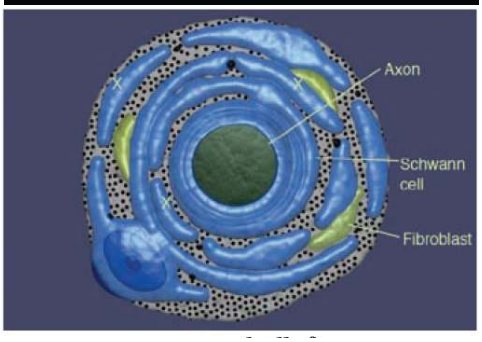
There is often **secondary axonal involvement in inflammatory neuropathy**. There are a couple of inflammatory neuropathies where the axon is affected directly. There is one **Acute Motor Axonal Neuropathy AMAN,** which is a rare one, but it seems to be more common in China for some reason.
Again, we are tuned in to what may make an inflammatory neuropathy and in **patients with CIDP they can actually have thick nerves.** Other demyelinating neuropathies can have thick nerves as well, so occasionally **we actually feel the nerves to feel if they are thickened**. If there is a lot of fluctuation from day to day, it suggests that there is a myelin problem which is causing problems sometimes perhaps less at other times and that would make you think of these inflammatory neuropathies as well. One of the things **you can see with some patients with inflammatory neuropathy is tremor associated with it.**



These are microscopic images of nerves sliced through so the axons are coming out of the screen towards us and they are coated by the myelin coat. So that's what it should look like, the axon in the middle with quite a nice thick surround of myelin around it.



In the nerves of CIDP patients, the myelin surrounding the motor and sensory nerves is attacked and stripped from the nerve by invading white blood cells called macrophages.



In this one, in a patient with a severe myelin neuropathy, you can see there is barely any myelin around and it is replaced by all these thickened nerve things sometimes called "**onion bulbs**". They look a bit like an onion bulb I guess.

When we try to work out whether **someone has an axonal demyelinating neuropathy** we don't really do a biopsy because we can usually **tell from the nerve conduction studies**. Many of you with neuropathy will have had **nerve conduction studies** where you are electrically zapped and we can actually **measure how long it takes the impulse to get from point A to point B**. Generally speaking if you have a **demyelinating neuropathy you get a slow conduction of the impulse but the size of the response is not too bad.** With an **axonal neuropathy** where there is damage to the axon **you tend to get relatively normal speed of conduction but much smaller signs of response**. That is how we tell the difference.

Occasionally we do a nerve biopsy if it is not clear from the electro physiology what sort of neuropathy we are dealing with as it is helpful at ruling out some specific types of neuropathy that are not really CIDP or GBS.

When we look at someone who has a peripheral neuropathy who has come to see us they will have symptoms. The symptoms are caused by the peripheral nerve not working as it should. Basically, **peripheral nerves can only do two things. They can not fire when they should or they can fire in an abnormal or distorted way**

when they shouldn't. We tend to get symptoms that are negative and positive. Negative symptoms of when they don't fire when they should and positive symptoms if you are getting distorted or abnormal firing. With the motor nerves it is **pretty obvious if it is not firing when it should as you are going to be weak and that is often the most telling problem. If it is there for quite a while the muscles waste away and are less bulky. We do sometimes see positive symptoms from motor neuropathy. You can get flickery, jumpy movements of the muscle because it is firing off when it shouldn't or you can get cramps. Cramps can be really quite troublesome in people with neuropathy.**

When you go to **the large sensory fibres, (they are the ones that mostly deal with joint position sense),** you can imagine if you are standing and you are not quite sure whether your ankles are flexed to 90 degrees or 110 degrees or 80 degrees, then **it is difficult to actually know how to adjust your position to stand**. If you are standing and you are becoming co-ordinated, but maybe you don't precisely put your foot where it should be when you walk because you are not quite sure where it is, **that lack of joint position sense tends to produce balance and co-ordination problems.**

The funny feelings that you can get from distorted large sensory fibre function are mainly **an odd feeling of a tight band around the leg or the arm or a tingling sensation.** They are features we sometimes see with **people with large fibre sensory dysfunction**. **Small fibre dysfunction** (obviously the small fibres don't work at all) so **you can't feel it if there is a pain sensation**. That's a bonus sometimes, but of course what it means is that **you may injure your hand and not realize, you might place your hand on a hot plate and you are slow to recognise it, so we do sometimes see people who end up with painless burns or other injuries that are caused by not actually withdrawing their fingers as quickly as they should.**

More often with the **small sensory fibres we get these positive symptoms which are really a feeling of pain when there shouldn't be pain.** In other words the nerve fibres firing off when there is nothing that should be triggering the pain at that time or a burning sensation. **It is very common with people with small fibre neuropathy.**

Autonomic impairment - when things are not working as they should - where you can get **impotence in men** and you can get **postural dizziness** when you stand up from lying. **One of the things that will put your blood pressure up is the autonomic input**. If that doesn't work quickly enough and you stand up, your **blood pressure** drops and you feel light-headed or dizzy. **Too much autonomic function or distorted autonomic function can produce sweating** when you shouldn't sweat or **diarrhoea** when you shouldn't have diarrhoea.

There's a whole lot of different symptoms and we actually listen and tune into what the symptoms are and they give us a clue as to where they might not be functioning as they should. **Patients with inflammatory neuropathies can get all sorts of variations.** The classic original description of an inflammatory neuropathy is **Guillain-Barre` Syndrome** which is **acute neuropathy which is predominantly motor**. In fact even with **Guillain-Barre` we often see people with sensory symptoms** which are quite troublesome.

With **CIDP**, I think it is fair to say that again the original thing that was described **involved weakness as the predominant symptom.** We would recognize there are many, many variations on the theme and basically you can take any combination of those, mix them all up together, and you have **200 different patterns** that you can produce from that and **everyone is different**.

When we are talking to our medical students we say **"If you go through this process then you should be able to recognise certain patterns and certainly one of the most important patterns is acute severe generalized polyneuropathy which is almost always Guillain-Barre`. CIDP is the other one because it is so important not to miss that in terms of treatment."**

There is this condition of **critical illness neuropathy** which **occurs when someone has been in the intensive care unit** but usually that is not diagnostically difficult. You know they have been in the intensive care unit so it is pretty fair where the problem might arise there. Some of these other things can produce reasonably acute neuropathies.

Chronic Demyelinating Neuropathies (CIDP is one of those) are **basically in two gro**ups. There is the **Inflammatory Group** which is **CIDP, Multi-Focal Motor Neuropathy** and there are some situations associated with blood cells that are producing anti-bodies as part of a cancer syndrome. There is a group of **Inherited Demyelinating Neuropathies** as well which typically **occur earlier in life** and they are not difficult to tell from CIDP usually.

With **GBS** the typical form is what was originally described by Guillain and Barre` as acute ascending motor neuropathies. Typically it starts in your legs and works its way up the body and involves a lot of different areas. In fact, as time has gone by, we recognised that there are **variants**. There is the one called the **Miller Fisher Variant** which **involves eye movements** particularly, so people are getting **double vision** as an early feature of it and there is what we call the **Axonal Variant** which occurs **particularly in China** and there is **Acute Motor Axonal Neuropathy (AMAN)** also **Critical Illness Neuropathy.**

The other thing that we sometimes see **is patients getting a localized area of inflammatory neuropathy**. The one we see **is Neuralgic Amyotrophy**. Somebody who has a **neuralgic amyotrophy** probably wouldn't regard themselves as having **something similar to CIDP or GBS but it really is**. It is actually **anti-bodies causing damage to the nerves, particularly in an arm**. With that condition patients typically have an onset of pain, usually **triggered by having a viral illness**, which causes anti-bodies to form and anti-bodies damage nerves in a particular area, usually one arm. They have **pain in the arm**, the pain is quite troublesome for a week or two and **then they get weakness in the muscles of the arm** which gradually **can recover**. It is a characteristic sort of condition.

End Section – Diagnosis. The Section on Treatment will be in our next newsletter.

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.

Support

A number of people have been given support over recent weeks. We were able to give parents who have children with recurring CIDP the opportunity to have contact with other parents in different States. This is the first time we have had multiple young patients to assist. Thank you to the mothers who are supporting each other and to member Beth in S.A. for your help.

Special thanks to Rebecca who is supporting a newly diagnosed patient with acute GBS. Our thoughts are with Vanessa and Stuart during this difficult time.

Also "thank you" to those who supplied information to assist a member moving to the Geelong area. It is great to be able to obtain information from members on such things as where they have their 'Intragam' etc. It was very helpful and our member was most grateful.

Sites to check out on the Web: www.gbs-cidp.org . Check out: Living with CIDP – Online Support Group, also our friends at www.gbsnsw.org.au

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.

THANK YOU TO THOSE WHO HAVE PAID THEIR SUBSCRIPTIONS.

FOR THOSEWHO HAVE NOT YET PAID, PLEASE NOTE THEY ARE NOW OVERDUE.

THE 'IN' GROUP

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 Subscription 1/7/13 to 30/6/14.

Annual Subscription		$ 15.00
Other Items		
Booklets- GBS	$3	$
CIDP	$3	$
After GBS	$3	$
The Road to Recovery A-Z	$6	$
- Boy, Is This Guy Sick	$2	$
Easy Recipes Book	$14	$
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

PRINT POST APPROVED SURFACE POSTAGE PAID
PP 335708/00020 MAIL AUSTRALIA
(ABN) 77 954 503 188

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)
Postal Address for Newsletter only: 44 Mavis Ave., Beaconsfield, 3807. Telephone: 03 9707 3278