

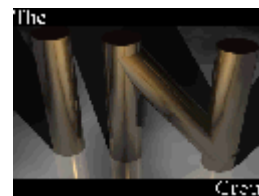
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Inflammatory Neuropathies other than GBS and CIDP

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*From the address by Dr RICHARD MACDONELL, Neurology Department Austin Hospital, to the quarterly meeting of **The IN Group** held 18/2/98, Balwyn Library Meeting Room, 336 Whitehorse Rd.*

Peripheral neuropathies are any disorder of peripheral nerves including disorders of the cell body, its axon (central conductor) or the myelin sheath. They may be diffuse (such as GBS and CIDP), multifocal (maybe a nerve in one leg and a nerve in one arm) or unifocal (affecting one nerve, mononeuropathy).

They may be acute (developing over several days or a week or two at the most) or chronic. The division between acute and chronic is quite arbitrary but the dividing line generally accepted is six weeks.

A) Mononeuropathies

This is the most common neuropathy encountered in clinical practice. Because only one nerve is affected all the symptoms are in the territory supplied by that nerve.

The most common cause is compression and its most common form is Carpel tunnel syndrome where the median nerve as it enters the hand is compressed by ligaments in the tunnel at the wrist, causing demyelination of the nerve.

Rarer causes are ischaemic (loss of blood supply to the nerve) often common in diabetes. Even less common is vasculitis (an inflammatory disorder of blood vessels producing narrowing of the vessels and loss of blood supply to the nerve). In much the same way, an obstruction to blood flow can cause a heart attack or stroke in other areas of the body.

Occasionally one can get an immune response, akin to GBS but restricted to certain areas. Brachial plexitis may affect a limb but unlike GBS it doesn't spread.

Infections such as shingles can cause acute neuropathic damage. Leprosy, whilst not common in Australia, is worldwide the commonest form of neuropathy.

B) Multifocal neuropathies

These are less common. A patient will present with weakness and loss of sensation associated with a median nerve. Then within a few days the patient may then develop symptoms in the opposite leg. They are patchy. They are almost always due to obstruction of blood supply to nerves with diabetes being its most common cause. Occasionally they may be due to infection eg AIDS, CMV, sarcoid, leprosy.

C) Polyneuropathies

They usually affect a number of nerves symmetrically in upper and lower limbs. Typically they will start in the lower limbs because their tolerance to damage, such as lack of blood supply, is less, possibly because they are furthest for the blood distribution.

The **Acute** is a condition causing over several days rapidly increasing weakness with or without alteration in sensation. The latter may be positive such as tingling, burning or uncomfortable feelings or negative such as numbness or other loss of sensation.

If it is not Guillain-Barre Syndrome what could it be? In the old days diphtheria was common but not so now due to inoculation. Diphtheria patients initially complain of difficulty in swallowing and speech before the weakness spreads to the limbs. Other conditions to consider include TOCP (organophosphate fertiliser used in farming) poisoning, cancers such as Hodgkins Lymphoma, AIDS in this day and age, most rarely porphyria (King George 111 suffered from this), vasculitis (inflammation of the blood vessel), ciguatera (a tropical fish) toxin and glue sniffers neuropathy.

As a group the **Chronic** is more common than the Acute. They typically affect the legs first, causing a loss of sensation described as a "glove and stocking" distribution - like having long socks on legs or gloves on the hands. Often there is a loss of reflexes. To distinguish a nerve disease from a muscle disease such as Myasthenia Gravis (severe muscle debility) one firstly checks for a loss of sensation and a pattern of weakness. In a neuropathy weakness normally affects first the further away parts, the feet and the hands, whereas muscle diseases for the most part affect closer to the body such as hips and shoulders. Another significant difference is the loss of reflexes in neuropathies.

If a neuropathy is indicated then inherited forms need to be considered. They are rare with Charcot-Marie-Tooth disease being commonest. There may not be a family history; it may present as a sporadic mutation.

The next group is the acquired for which there can be many causes:

- Toxic - chemicals and drugs, particularly alcohol and prescription drugs.
- Disease - thyroid disease, diabetes.
- Nutritional - Vitamin deficiency B1, B12, with or without alcohol.
- Associated with cancer.

- Immune mediated with a paraprotein.
- Ischaemic - diabetes, vasculitis.
- Heavy metals - Arsenic, Thallium.

In most cases they are due to axon damage. In a few cases the primary pathology is demyelination.

Demyelinating Chronic Neuropathies

The most common is CIDP.

In considering other than CIDP there is the paraprotein associated disease. The person is pumping out a lot of immunoglobulin in the form of antibody IgM. In a number of patients this paraprotein is binding to another protein on the myelin sheath, MAG. It is postulated that this is causing the neuropathy by damaging the myelin sheath. With this MAG neuropathy there is more loss of sensation. more numbness, as compared with CIDP.

There are other paraproteins which can be occasionally picked up in association with neuropathy but the cause/effect relationship is less firm.

Very occasionally there may be drug-induced cause but this is usually evident as there are only a few drugs that cause demyelination.

New Developments in Inherited Demyelinating Neuropathies

There are 3 main types of Charcot-Marie-Tooth (CMT).

Type 1 is the most common with a number of families affected in Melbourne.

Type 3 is a severe neuropathy with onset in childhood. Unfortunately the children generally will be in wheelchairs by 12 and have limited life span.

Type 4 is Refsum's disease. Here the problem is in an enzyme which breaks down fats in our body. Because of a defect in the enzyme there is a buildup of a compound that attacks nerves.

In most cases of CMT Type 1, the abnormality is on chromosome 17. This is on the gene that codes for peripheral myelin 22. There is a doubling of the gene leading to an excessive production of the protein which causes a fragility of the myelin.

There is a variant - liability to pressure palsies. If such affected persons lie on their elbow or on their arm in an odd way, this can cause an acute demyelination of the local nerve with resultant weakness. It will heal over several weeks but the pattern will repeat. This variant is interesting as the gene defect is in the identical spot for CMT Type 1 but a loss not a duplication, resulting in less protein production.

This tells us this protein PMP22 is extremely important in myelin integrity and myelin function. This information may lead to improving the understanding of demyelinating neuropathies.

Some Questions & Answers

Q. The IN Group received advice that some patients have been rediagnosed from Motor Neuron Disease to CIDP. What is the reason?

A. Motor Neuron Disease is a lethal condition where there is a damage to the cell body of the central nerve system causing progressive weakness and loss of function. In a very small number of patients the disease affects the nerve rather than the cell body. It is a separate entity, neither CIDP or MND.

Q. *What is Miller-Fisher Syndrome and Transverse Myelitis?*

A. MFS is a variant of GBS due to a specific antibody and affecting the cranial nerves. TM is not a neuropathy; it is more akin to Multiple Sclerosis.

Membership

The membership of The IN Group continues to grow, with the total now at 252.

Whilst our main support is to and from our Victorian members (181) it is pleasing to note the increasing numbers of interstate - NSW (10), Qld (10), SA (4), WA (2), Tas (4) - and overseas - Austria (1), Canada (2), Italy (1), Japan (1), NZ (4), Philippines (1), Slovakia (1), UK (3), USA (27) - members.

Shortage of Intragam

There is again a shortage of Intragam in Victoria. A number of our members have been given a reduced amount. For example I received only 24gm of Intragam instead of the prescribed 30gm treatment on my last two fortnightly visits to the Alfred Hospital.

Relating to a previous shortage last October, the Victorian Minister for Health, Rob Knowles, replied on 14/1/98 to our letter of 23/10/97 requesting his government provide funding to the Red Cross for a publicity campaign to attract more Victorian plasma donors. In his reply Mr Knowles advised that there is currently periodic shortages, that the Australian Red Cross Blood Service - Victoria are currently supplying Intragam to those patients who fall within the guidelines, that GBS and CIDP patients fall within these guidelines, and that his Department has funded a consultancy to analyse the causes of a general decline in blood donations.

I shall be writing to the Minister further to advise that the consultancy recommendations have apparently failed to increase the supply of Intragam.

JAMES GERRAND, Director

Council of GBS/CIDP Support Groups of Australia

The Council met by Conferlink on Sunday 8th February. Participants were President **Barbara Burzak-Stefanowski**, Secretary **Melva Behr**, Victorian Delegates **James Gerrand**, **Betty Gerrand**, South Australian Delegate **Heather Trenorden**, Tasmanian Delegate **John Stanley** and CSL observer **Werner Fiedler**.

During the hour's discussion by telephone linking parties at Melbourne (2 locations), Alice Springs and Devonport, the following were the major matters dealt with.

The draft Constitution was adopted with some minor corrections.

Noting that the GBS Association of NSW had advised that "this association would assist" "(the Council) "and co-operate where possible", the Council agreed to invite the GBS Association of NSW to be an Associate member of the Council.

It was agreed that June 1st should be the National GBS/CIDP Awareness Day. President Barbara had prepared a Press Advisory statement, Press Release, Public Service Announcement and Fact Sheet "to focus media attention on the" (GBS/CIDP) "conditions and our individual support groups".

James reported The IN Group had prepared a [questionnaire](#), based on that already in use by the GBS Association of NSW, to help arrange personal support to GBS and CIDP sufferers.

CSL Ltd was thanked for its generous financial support that made this telephone linkage possible.

Cake & Biscuit Stall at Maling Road

Thanks to the initiative and perseverance of committee member **Margaret Lawrence** The IN Group will be holding a **Cake & Biscuit Stall** at Maling Road on Saturday 2nd May.

This should be a good money spinner for the Group, raising more money to help the medical research being carried out by Dr **ANDREW KORNBERG** into GBS/CIDP at the Royal Childrens Hospital.

All that is needed now is for members and their friends to make the cakes and biscuits at this trendy location. The stall will also be a good publiciser for The IN Group.

Please contact **MARGARET LAWRENCE tel 9802 5319** and discuss with her what cakes and/or biscuits you and your friends will be able to make for this event. Contributions can be picked up.

Chronic Illness Alliance

The IN Group has joined this alliance of groups which include the AntiCancer Council, Cystic Fibrosis Association of Victoria, Asthma Foundation, Victorian Aids Council, Haemophilia Treatment Centre, Huntingtons Disease Association, Motor Neurone Disease Association.

At the Alliance meeting of 19th January there was general concern expressed at how hospitals were increasingly loath to keep patients diagnosed with chronic disorders because their cost upset their budgeting. Some doctors valiantly tried to keep treating their patients in hospital by rediagnosing the disorder under a new name.

I brought up the case of a member, slowly recovering from GBS in a rehab hospital. The staff were wanting to move him to a nursing home where he would no longer receive treatment. I advised the member to get an outside neurologist advice which resulted in the member being allowed to stay for another 6 weeks during which time he recovered sufficiently to return to his home to be looked after by his wife.

James Gerrand

Support is the Name of our Game

Some examples

Dear James,

Just a note to thank you for visiting me in intensive care some months back; it was encouraging to be connected to the group and talk with you about my condition.

I spent 3 1/2 months in hospital learning to breathe, eat, walk etc again and a further 2 months as an outpatient in Essendon Rehab. It is now 6 months since I first got GBS and I am going well, still get very tired and have numbness in my hands and feet and a straddle in my breathing from intubation. All in all I am progressing well and looking to return to part-time work in Feb.

Thanks again and if I can be of any help to others, I'd love to visit also.

JAN LANE, Sunbury

How the Internet helps

*Hay James I just got a message from a lady in Dubbo about her father. He has CIDP and she is pretty worried about him. I believe he lives in Melbourne. Thought maybe you could get her and him some support calls. Her address is mclean@crt.net.au Thanks, **Barbara (Beltz USA)**.*

I emailed (**Alison**) **McLean** and received her reply:

*Thank you for your offer to telephone my father. I spoke with him yesterday and he sounds very excited about the whole idea. He has been quite isolated I think, not knowing anyone else with CIDP or even much about it. A support group could have quite amazing healing powers!! ... **Rex Bone** is at Hampton Rehabilitation Hospital ... Thanks ...Alison McLean.*

I visited Rex at Hampton who is making a good if slow recovery and has expressed his gratitude at The IN Group support by becoming a member and generously donating \$100. Thanks, Rex.

CIDP booklet into Italian

ingroup@vicnet.net.au (James Gerrand)

I received the CIDP pamphlet and thank you very much. I gave it to my doctor who was very happy about it and told me they would like to have it translated into Italian. I am sending today my an

nual subscription to The IN Group. ...

Paoli Pioppi (ROMA ITALY)

I contacted **Andy Leitch** to get permission from the GBS Support Group of the UK for permission to translate their booklet. Andy advised OK but the booklet is being revised for a new edition.

Other Overseas contacts

Dear James,

Let me wish you a Merry Christmas and a Happy New Year. All the best wishes from Slovakia. ... Thank you for the newsletters. I am sending you US\$100 (=AUS\$150) as subscription + donation, perhaps you could kindly send me one pack of your Christmas cards. ... I wish you a lot of success with your great work for The IN Group.

George Pekkarovic (Bratislava, Slovakia)

Pamela Patchell, 46 year old mother from OHIO USA, wrote how her symptoms of CIDP developed in the fall (April?) of 1994 but was not finally diagnosed until November 1995. But confirmation was required before treatment which then only started in March 1996. The effect of eight weekly IVIG infusions was remarkable. Then her condition deteriorated and IVIG treatment was not restored until November 1996 which gave Pamela much improvement. In June 1997 all symptoms returned suddenly and forcefully. IVIG treatment was now resumed at three weekly intervals and has since continued.

Pamela's niece found The IN Group and its address on the Internet with the result Pamela has joined The IN Group. Pamela writes "... *The CIDP booklet is so helpful and informative. I am so pleased to receive this information and your newsletter and I appreciate the speed with which I received it. ...*

Committee member **Vilma Clarke** has been writing to a number of our overseas members who have suffered from CIDP, her complaint, with rewarding results. One such is from **Bram Smith** of New Zealand. "... *Had a delightful letter from Vilma Clarke. She seems in good heart and is doing a fine job*".

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