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Treating CIDP

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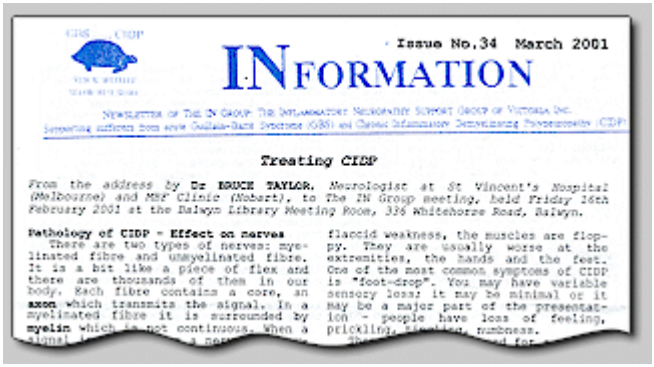
Issue No.34 March 2001

INFORMATION

Treating CIDP

From the address by Dr BRUCE TAYLOR, Neurologist at St Vincent's Hospital (Melbourne) and MBF Clinic (Hobart), to The IN Group meeting, held Friday 16th February 2001 at the Balwyn Library Meeting Room, 336 Whitehorse Road, Balwyn.

Pathology of CIDP - Effect on nerves

There are two types of nerves: myelinated fibre and unmyelinated fibre. It is a bit like a piece of flex and there are thousands of them in our body. Each fibre contains a core, an axon which transmits the signal. In a myelinated fibre it is surrounded by myelin which is not continuous. When a

flaccid weakness, the muscles are floppy. They are usually worse at the extremities, the hands and the feet. One of the most common symptoms of CIDP is "foot-drop". You may have variable sensory loss; it may be minimal or it may be a major part of the presentation - people have loss of feeling, prickling, numbness.

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Pathology of CIDP - Effect on nerves

There are two types of nerves: myelinated fibre and unmyelinated fibre. It is a bit like a piece of flex and there are thousands of them in our body. Each fibre contains a core, an **axon** which transmits the signal. In a myelinated fibre it is surrounded by **myelin** which is not continuous. When a signal is passed down a nerve it travels down the axon bouncing at 60 metres per second. This is pretty quick, needed, say, when you are commanding your hand to hit a tennis ball.

What happens in CIDP is that the myelin is destroyed or weakened, causing a fall in the rate of conduction. If it falls below 40 metres per second you start having symptoms. Some patients lose all their myelin with conduction falling to 15 metres per second.

Another thing happening with CIDP, which has not been emphasised, is that for some patients the axon is also attacked. If this happens recovery is not always complete because the axons may be damaged. The regrowth may not be in the right direction and it may take a very long time.

Another nasty factor with CIDP is it may not only affect the ends of nerves; it may affect the nerve anywhere along its course. This is why we use the term polyradiculo neuropathy. The radicules are the closest part of the nerve to the spinal cord. If this part is affected it is very much like having a bulging disk, pressing on the nerve. This can further cause slow recovery and can be why some patients do not recover fully.

The Swan cells can reduplicate the myelin in 7 to 14 days. With axons the recovery is slow and may not be complete.

Diagnosis of CIDP

This is often very difficult and may take time to determine correctly. The principle findings when you are examined by a neurologist are firstly a flaccid weakness, the muscles are floppy. They are usually worse at the extremities, the hands and the feet. One of the most common symptoms of CIDP is "foot-drop". You may have variable sensory loss; it may be minimal or it may be a major part of the presentation ? people have loss of feeling, prickling, tingling, numbness.

Then there is the need for a variety of blood tests, particularly looking for para-protein. This item is

important to know as it alters the treatment and the outcome. The tests include checking that the thyroid is working OK, for diabetes, for deficiency in vitamin B12 or folate, marking areas of inflammation.

Then there are electrical studies to determine the conduction velocity over various segments of the nerve. We also check for loss of nerve fibres. EMG may be carried out with needle examination of muscles affected by loss of nerve supply. This tells whether there has been axonal loss.

A lumbar puncture should be done be-cause the protein level of the spinal fluid will go up due to the inflammation at the radicular level. If you have this increase you are more likely to respond to treatment. People with normal CSF protein are a bit of a worry because they may have CIDP or may have purely axonal loss and so not respond to treatment.

Rarely do we do nerve biopsies because we can usually diagnose people with CIDP clinically through the above tests. A nerve biopsy is quite invasive. It can be helpful if you are not sure of the diagnosis but sometimes it does not provide any information. It can also cause side effects where you are left with a patch of permanent numbness on the leg or result in continuing pain at the site of the biopsy.

A major confusing factor in the diagnosis can be hereditary neuropathies. You can see a patient presenting with a neuropathy and you think this is not new, it has been here for years. You note the shape of the feet and ask if anybody in the family had difficulty in walking. The answer may be mother who had arthritis and you know that this is not an acquired neuropathy. Metabolic neuropathies such as diabetes, thyroid disease, are also sometimes confused with CIDP. A tumour, such as lymphoma of the glands, may look like CIDP. Some infections, such as HIV, may present like CIDP. Leprosy is a major cause of neuropathy worldwide. The acute form, GBS, is difficult to distinguish initially from CIDP. There are rarer neuropathies such as Multifocal Motor Neuropathy. Sometimes toxic neuropathies from drugs and poisons can be difficult to distinguish from CIDP.

Treatment of CIDP

Treatment is problematical and can be difficult. One of the first quest-ions to ask is, should this person be treated? Not everyone with CIDP needs to be treated. Some components do not respond well to treatment, particularly sensory symptoms; prickling, numbness, tingling of fingers and toes rarely get better with treatment. If these are the principal symptoms then it is often not worthwhile considering using heavy-duty therapy. Weakness is the symptom that responds best to therapy.

But you are often left with an incomplete response. This is often difficult for the patient and the doctor to adjust to. More or different treatment often will not make the patient better.

One of the useful rules is to consider treatment when a patient has difficulty in walking on his/her heels or toes, which indicates moderate weakness.

Initial Treatment

CIDP being chronic is a disorder which, at this stage, we don't think we can cure. It can go into remission.

The aim of treatment is to return to normal, acceptable levels of functioning at work and at home, maybe not the best level of running, playing sport, bushwalking. It will be treatment for a long time. It has to be acceptable as regards side effects, both immediate and long-term. It has to be easy to do and easily accessible, not involving great inconvenience to the patient.

There are only three treatments which have been shown to be truly effective in treating CIDP in

large clinical trials - **corticosteroids** or **predisolone**; **plasma exchange** (**plamapheresis**); **intravenous immunoglogulin** (intravenous drip of blood product **Intragam**). Other treatments that have been suggested as working include chemotherapy, interferons, cyclosporin.

The above three treatments are probably equivalent in their effectiveness in initial treatment of CIDP. They all have side-effects. A patient may be sensitive to one or more but not to all. Some patients are resistant to all therapies. **Predisolone** or **steroids** can induce diabetes, muscle weakness, thinning of the bones and/or skin, high blood pressure, increase in weight. **Plasma exchange** needs to be done in a hospital (not available in most country areas); needs venous access; there is a risk of introducing infection every time you stick a needle in a vein; can cause low blood pressure whilst being given; may make the patient feel dread-ful afterwards; the plasma removed has to be replaced with another fluid (albumen, another human blood product); it takes up to 4 hours to do; can be painful. But it is a very effective treatment. **Intravenous Immunoglobulin** (**IVIG** or **Intragam** drip) is relatively easy to give and does work. But there is a world-wide shortage because there are not enough blood donors. A significant number of patients will have an allergic reaction, it can cause head-aches, aseptic meningitis; there is a risk of transmitting infection because it is a human blood product, (the risk of transmitting Mad Cow Disease is a bit far-fetched).

The cost of the treatment is import-ant. Corticosteroids cost about 20 cents per day. Plasma exchange and IVIG cost about the same, up to about $1,500 per month.

I will review my initial chosen treatment after six weeks to see if there has been a response. If it has worked and is acceptable as a long-term therapy, then I try and find the minimum dosage which is required to maintain the patient at a near normal level of functioning.

If it did not work I will try another therapy. If none of the first three therapies works then I check to see if I have the diagnosis correct. If it is correct I consider a second line agent. Probably the most effective one is cyclophosphamide. It is a powerful chemo-therapeutic agent. An intravenous treatment monthly can be effective in stop-ping the disorder and significantly improving patients, refractory to the other three treatments. It is not free of side-effects though most patients tolerate it well. They may feel rather low, washed out, the next day. Rarely does a patient lose hair or vomit for days on end. It is relatively cheap and does not require a lot of input from nursing staff. The main complications are that it can drop the blood count to near zero, may run the risk of developing a tumour if used repeatedly.

The other treatment we consider using is interferon ? a naturally occurring substance within the body that turns on T-cells. We use it to treat Multiple Sclerosis, a demyelinating disease of the brain. There is some evidence that interferons will help a pro-portion of CIDP patients. It is very expensive and difficult to obtain.

Long-Term Therapy

Once you have found what works then you endeavour to determine the minimum dosage to maintain function. This will minimise side-effects, need to see the doctor, to go to hospital. Often one finds patients will be able to tolerate lower and lower dosages, or less and less frequency of treatment.

With long-term therapy the only three treatments proven to work are as mentioned above. If corticosteroids work, often this will be combined with a steroid sparing agent. This is a drug (such as Imuran) which allows you to gradually reduce your steroid level.

Exercise is vitally important for CIDP patients, to maintain physical fitness and muscle bulk and tone. Hydrotherapy is often very useful ? it doesn't matter if you fall over in the pool. It is important for the water temperature be not too high as this will slow down the rate of conduction. A review by a physiotherapist to design an appropriate exercise program is also very important. Walking aids ?

frames, walking sticks ? are helpful, mainly for balance, as are AFOs ? Ankle Foot Orthoptics.

People with chronic disorders can develop secondary problems such as depression. They can visit their doctor and complain of feeling worse but to the doctor everything seems the same. A simple treatment for depression may be better than increasing treatment for CIDP.

Treatment for pain is important. Normal pain relievers rarely have a major effect for neuropathic pain. We use pain modifying drugs which may be anti-epileptic drugs or even anti-depressants.

Anxiety can be a major problem. Education to allay fears, meeting other people with CIDP to understand what may happen is often good.

Fatigue is a common symptom. Treatment for this is to optimise the therapy. Steroids can cause some weakness. One looks for other causes of why the patient is not well. It is helpful to encourage the patient to get out and get on with their lives.

Curative Treatment?

Some patients will only require one treatment and then return to normal. But most of such patients will relapse but some have long remissions. Why this is so we don't know.

The best hope for curing CIDP is through high dosage early treatment. That is not something to be done as routine because it's working is questionable particularly as a cure. It has to be left to clinical trials.

Questions & Answers

Q. *Do GBS patients fully recover?*

A. *Many recover but not fully. They lose a percentage of their normal functioning. They can compensate for this except at times of stress - unwell, depressed, overtired, overworked, getting older - then symptoms will recur but it is not GBS which normally is a mono-basic illness, only occurring once.*

Q. *What are the prospects for stem cell research?*

A. *Some people with recalcitrant, difficult to treat CIDP in the USA have had bone-marrow transplants. The results have been encouraging but not great.*

Q. *Use of animal IVIG is being explored in the dairy industry. Is there any possibility of using such IVIG to over-come the shortage of human IVIG?*

A. *We have used IVIG specifically raised in horses to treat illness such as tetanus but the rate of allergic react-ion is so high that everybody would be-come allergic to it after a short time.*

Q. *Should an ex-GBS or ex-CIDP person have a flu injection?*

A. *I think it is a very good idea. The flu injection, being a live virus, can make symptoms worse but if you have a full-blown influenza it can be life-threatening, particularly for CIDP patients. So I do tend to recommend that such people have flu vaccinations.*

(I have given three of the four pages of this issue of "INformation" because of the value of the information that Dr Taylor has provided about this disorder. Editor)

The IN Group News

We give $3,500 to Research

The IN Group has given a further $3,500 to the medical research into GBS and CIDP being carried out by Dr Andrew Kornberg, of the Royal Children?s Hosp-ital. This brings the total of our donations over the past five years to $31,000.

The IN Group thanks the many members, families and friends for their continuing generous monetary contributions and personal activities.

Dr Korberg expressed his gratitude and reported on the International Work-shop on Childhood CIDP of the European Neuromuscular Centre he attended in The Netherlands 8-10th December. The main outcome was agreement to set up a formal childhood CIDP database to accumulate information regarding clinical characteristics, response to therapy and outcomes.

Continued shortage of Intragam

The shortage of Intragam continues with many patients receiving only 80% of prescribed amount (say 24gm instead of 30gm). The IN Group will continue to press the Federal and Victorian government to provide the $15 million recommended by the Blood Product Working Group as needed to overcome this shortfall.

The IN Group experiment of letter-boxing 1,000 Melbourne residents has produced only 3 blood donors so far in the Australian Red Cross Summer Cha-llenge. The trial would have had some publicity value. Our thanks to the ten who did this letter-boxing.

Support the name of our game

Hospital visit

JOHN POLLARD and **GINA MERNONE** have been very helpful in visiting a GBS patient, Chuck Lew, now recovering after being in Intensive Care at St Vincent?s Hospital. Chuck?s cousin, Jason Lourey, has also been most appreciative of John and Gina?s support.

Members? Stories

Two of our new members have produced histories of their experience of GBS. "**Richard?s Story**" by Richard Davies of Werribee and "**Another Saga of GBS**" by Guy Lauricella will be produced in booklet form and sold at a small fee, similar to June Cathcart?s "**A Road to Recovery ? A ? Z**" and John Pollard?s "**Boy, is this Guy Sick**", already avail-able.

Internet contactZ

The following have been sent information following request, usually by e-mail (names in bold now

members).

Mark l. Adams (GBS - North Carolina USA), Connie Emmons (husband CIDP ? Texas USA0, Michelle Patton (grandfather GBS/CIDP? ? Iowa USA), Carl Erikson (CIDP ? Minnesota USA), Hugh N Maddux (friend GBS ? Georgia USA, Sheri L Faiver (friend GBS ? North Carolina USA), Patty Steele (husband CIDP ? Whidbey Island WA), Hubert Bonis (client GBS ? East Devonport Tas), **Pauline Stanley** (CIDP ? Woody Point Qld), **June Black** (GBS Geelong West Vic), Mark Harris (CRS Werribee Vic), Lee-Anne Haigh (GBS ? Ashburton Vic), **John Keiger** (CIDP Geelong Vic), **Guy Lauricella** (GBS

C. jejuni infection and Link with GBS

This is an interesting 5 page article by David Acheson, M.D., Director of The Food and Safety Initiative at the New England Medical Centre, Boston USA. It was obtained from the Internet, being posted on gbs.etal@gbs.org

This clinical perspective concludes 30% to 40% (in the USA) are related to prior infection with Campylobacter jejuni C. jejuni bacteria are found in fowl and many wild and domestic animals. Most human infections probably result from contamination of milk and other food sources, particularly poultry.

A similar Australian study would be of interest.

Correction to e-mail address

The e-mail address, of committee member **VILMA CLARKE**, and now President of the Council of GBS/CIDP Support Groups of Australia (cogratulations Vilma) was incorrect in our September 2000 issue, it is kclarle@netc.net.au.

Guidelines for Support

The In Group

After having received a recent communication from another CIDP sufferer, I wish to make a plea for discretion in such contact. Support is the name of our game and networking is our strength, but to be furnished with a list of negative outcomes and unsuccessful treatments, at a time when my own condition was on a downturn, I found extremely depressing and counter-productive.

May I suggest some simple guidelines?

1. It is enough to state you are a GBS/CIDP sufferer, or are familiar with the conditions through a partner, family member etc.

2. Do not volunteer information. Ask what may be required.

3. Keep it brief, positive and supportive.

With best wishes for the Group?s continuing supportive role.

Annette de Courcy

Last Updated: 15 Oct 2007 17:49