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Newsletter No.47 - June 2004

Council Care for the Disabled

Research Update into GBS/CIDP


INFORMATION

Research Update into GBS/CIDP

From the address by **A/Prof ANDREW KORNBERG**, *Neurologist, Royal Children?s Hospital, to The IN Group Public Meeting, held Wednesday 12th May 2004 at the Balwyn Library Meeting Room, 336 Whitehorse Road, Balwyn.*

Donation from The IN Group

Thank you all for the $5,000 cheque which will be going to our Neuromuscular Fellowship. The importance of this Fellowship cannot be stressed enough. There are only a few people interested in nerve and muscle disease in Melbourne. Through this Fellowship we will be training the future doctors interested in looking after people with nerve and muscle diseases including disorders like GBS and CIDP. Your help in continuing to sponsor this Fellowship is greatly appreciated.

See also: The next IN Group *Get Together* on 11th August 2004

Why does GBS and CIDP happen?

We are beginning to unravel why these disorders happen to people. We hope that with a better understanding of how a disorder occurs, we will be able to create better treatments. With the availability of better tests, people may be diagnosed earlier. Unfortunately, many people with CIDP may go from doctor after doctor until the diagnosis is made. Having better tests may help improve this.

Coming to see a Doctor

Many doctors may not be fami8liar with your condition. In the community as a whole it is rare. When you see a nerve and muscle specialist, they approach nerve disorders in a certain way. They use clinical patterns that are verified by tests to make a diagnosis. These tests may include nerve conduction studies or nerve biopsies, and recently, antibody tests from blood.

Clinical Patterns

These are very important. If you see a doctor and he or she doesn?t realise that your "footdrop" or your "tripping over" could be related to a nerve disorder then they won?t be able to diagnose the disease. When we see a patient, we must firstly suspect that the symptoms are of weakness, secondly we must then identify what type of weakness the person has. If it is a problem with mainly the muscles close to the pelvis, it tends to be a muscle problem rather than a nerve problem. If it is further away from the body such as "footdrop" then it tends to be a nerve disorder. The next steps

revolve around further testing which may include nerve conduction studies to work out whether the myelin, the outer lining of the nerves, is affected. We rarely then go on to nerve biopsies to help with diagnosis.

Since the late 1980s, we have started testing for "nerve" antibodies in blood. These antibodies may be the cause of some of these conditions. These immune-mediated nerve disorders include CIDP, GBS, Miller Fisher Syndrome (MFS), Multifocal Motor Neuropathy (MMN). They are conditions where the person?s own immune system by producing the antibodies may be causing the disease. There may be some trigger, antibodies are produced and these may attack nerve. When nerve is attacked, they don?t function normally and symptoms such as weakness or numbness can result.

We can now measure a variety of antibodies in the blood. These antibodies tests may help doctors diagnose a nerve disorder. It is possible that in time treatments can be fashioned for these disorders.

Some conditions such as CIDP respond very well to steroids, to gammaglobulin, or to other medications. But there are conditions that can look like CIDP that don?t respond to steroids, to plasma exchange, or such therapies. Therefore if the person has a particular antibody that is associated with that disorder, we would not use a therapy that would not help. I am sure there are some in the audience who have gone through lots of different treatments, some that have been very helpful and others that were not helpful and associated with side effects.

So using the information of serum antibodies may help us pick the best type of treatment.

List of Inflammatory Neuropathies

The list continues to grow. The first four on the slide are the most common types of immune-mediated neuropathies.

GBS usually becomes apparent in a previously well person. They may have an infection as a trigger, and they may get profoundly weak. Some individuals may need to be on a ventilator. The pattern of weakness and other tests may enable early diagnose and effective therapy.

CIDP is chronic and most of the time it takes many months to make a diagnosis.

MMN is much less common than CIDP but the important factor is that there is a condition ? Motor Neuron Disease (MND) - which looks like CIDP. However, patients with MND usually die a couple of years after diagnosis, whereas people with MMN live a normal lifespan. Some people have been told mistakenly they have MND when in fact they have a treatable condition such as MMN. Specific antibodies may be helpful in these circumstances. We have had a number of cases who have been given a diagnosis of MND, but the blood test suggests MMN. The person can be treated and improve.

Myelin associated glycoprotein (MAG) is one which once had no treatment but now there are effective therapies as we are understanding how they are caused.

Current Research in Inflammatory Neuropathies

Firstly we are trying to understand what causes the disease. By understanding we will be able to fashion better treatment. There are some new treatments available now because of this research.

Various triggers such as infectious agents: bacteria, viruses, have been identified. Research suggests

that individuals with some predisposition, which is infected with one of these triggers, may start to produce a particular type of antibody. That antibody may lead to a nerve problem if it attacks a nerve. This association between infectious agent and the development of antibody is termed molecular mimicry.

There is probably a genetic type of predisposition in some people. In families with CIDP there may be a history of somebody with diabetes, a history of multiple sclerosis, or some other condition that is auto-immune.

Q. About 5 years ago The IN Group carried out a survey of some 50 members, half diagnosed with GBS and half with CIDP. Those with GBS could identify some cause such as influenza but the best the CIDP ones could suggest was stress.

A. I think the reason for that is that the GBS people are perfectly well and then suddenly they are very weak and seen by a doctor. These people are going to remember the infections of the past few weeks. In CIDP, a person may have had a sort of rumbling symptoms for a number of weeks, months, years. When you try to recall when it started and whether there was an infection, you may not be able to recall a trigger. There are some cases where a viral or bacterial trigger is identified but this is in the minority.

Gangliosides

The antibodies that we do know are associated with many of these conditions are antibodies to ganglio-sides. Gangliosides are part of nerves. They have a variety of functions, some include nerve signal transmission. They are concentrated in areas where there are holes or nodes in the myelin, the outer layer of nerves. When you have an antibody to a ganglioside, those antibodies may stick to these areas and therefore cause nerve dysfunction. Myelin may be stripped away and damaged.

Most of the antibodies are against a variety of gangliosides. There are at least some 16 gangliosides which are associated with different conditions. Recent advances include a particular antibody associated with GBS, and MFS (a variant of GBS). The particular antibody associated with MFS occurs in 95% of individuals affected.

There is a whole variety of other antibodies we know that affect people. People may come to a doctor and say "I?ve got burning feet etc". In the old days the neurologist may say "I think you have a neuropathy but I can?t do anything about it". Now we can measure an antibody and say "Well, it looks like this is an immune problem and maybe we can use this medicine to try and make it better".

A variety of antibodies have been identified in GBS. Some antibodies are associated with particular symptoms..

What are the better treatments?

Essentially by understanding what causes these conditions and whether the antibodies are important can help us devise therapies. Understanding where they act may be helpful in treatment. If we have an animal model then the treatment can be used there

A variety of therapies have become available including rituximab (not readily available). This medication decreases antibody levels in the blood by binding to the cells that produce antibodies and stops such production. What happens then is the antibody levels are reduced and people improve. It is a very expensive medication but in time it will become more available.

There are other being developed that work in similar ways. They are mainly used for chemotherapy in cancer. We do use them in some nerve disorders.

So there is hope that new and effective therapies will be formulated. Thus, other than gammaglobulin or steroids; there are other medications in the pipeline.

For CIDP, predisolone is a standard treatment, plasma exchange is useful but is very tough to do and there is IVIG which may produce an improvement in strength in 25-60% of patients. Everybody is aware that there has been a shortage of Intragam so everybody has been getting less. But you should know we are out there trying to get as much as we can. Unfortunately our hands are tied because it is government that has to fund the extra. We are on a working group with the Australian Red Cross to get government to give us more money so that there is more gammaglobulin.

For GBS the main treatments are IVIG and plasma exchange. Steroids do not have a place. The most important factor is to support people in rehabilitation and in Intensive Care. Unfortunately in the last few weeks at the Royal Children?s Hospital we have had a huge number of children being admitted with GBS..

Q. What is the prospect for a child with CIDP?

A. CIDP in childhood is different to CIDP in adulthood. The prognosis for CIDP in childhood is better. Many children will outgrow their CIDP, some will need ongoing treatment like adults do. The mainstay is steroids in the first instance as gammaglobulin, say on a monthly basis, can be difficult. If steroids are not showing side effects then over time there is the hope that the process goes away and they won?t need the steroids. We found in a review that about 70% of children have a good response or they outgrew the CIDP.

In treatment with steroids you start off on a high dose and then when you get the children or adults under control, you wean down the dose. The rule of thumb is the lowest dose, as less frequently as possible. If they have side effects you use other medicines. The most common side effect with steroids is weight gain.

Q. How soon will diagnosis by identification of antibodies become standard procedure?

A. A few years ago it was standard. It is standard in the USA because it is readily available. The problem is that when there is a test that is readily available, it gets abused and used for a variety of conditions for which it should not be used. People may get confused because they see a mild elevation of an antibody and they put them then on treatment. In the end you have to interpret whether that person?s problems have any relationship to the antibodies measured.

When my laboratory started doing all the antibodies it became standard in Melbourne. We lost funding so I couldn?t do the antibodies any longer. We are now starting up again.

There are only a few antibodies associated with CIDP. Many people are trying to examine CIDP patients to see if there are certain antibodies that are associated. I believe CIDP is not one condition but many conditions that have similar symptoms. So we would not probably find as specific antibodies in CIDP as we in GBS. What the aim would be is to split out people with certain types of CIDP and say these people are going to respond to gammaglobulin, these to steroids, these to rituximab etc. By splitting people out we will be able to use the best treatment for that person. We have not the evidence at present but we are working on it.

Adult CIDP includes the adolescent. Childhood CIDP is probably in the first ten years of life. It is very rare for adults to outgrow CIDP. With treatment many can live a fairly normal life.

1974-2003 Study of GBS children at RCH

This major study was carried out by my Neuro-muscular Fellow, M. Victoria Rodriquez-Casero. Its main objectives were to look at the clinical plus electrodiagnostic features of all children with GBS over this period and then to evaluate whether treatment ? plasma exchange, steroids or IVIG ? was helpful. We looked at all their records (some-times very difficult!) and contacted many families.

We looked at how severe the condition rating was on a scale from 0 to 6 (0 being healthy and 5 being on a ventilator and 6 death). The mean score was 3.45 (able to walk about 5 metres with support). On discharge they just needed support, perhaps at rehabilitation. We found that the mean age was 7.5 years, youngest 14 months and oldest 15.5. We did not find any season that was particularly bad for GBS..

72% of the children had some sort of infection within 4 weeks prior to the onset of GBS. That may be the trigger which causes the GBS. Gastro (cam-phylobacter) infection was 20%. The mean days from the onset of symptoms, usually weakness, to admission was 6 days. We found half of our patients went to the Intensive Care Unit, duration ranging from 1 to 79 days, and 38 patients were ventilated from 4 to 78 days. Some patients needed a tracheostomy to help with their breathing. 100% of the children were weak and many had pain. Pain is very uncommon in children. 75% of the children returned to normal or only had minor signs.

Main Conclusion from Study

The comparison of the treatment options over this 28 year period showed that IVIG (Intravenous Immunogammaglobulin) appeared to be more beneficial to children diagnosed with GBS than PE (Plasma Exchange).

The patients spent less time in hospital, had less ventilation time, had a quicker recovery and there was a less complication rate.

However this conclusion needs to be confirmed by other studies as there are factors affecting these results. There can be bias in a retrospective study, the small number of patients may not be able to be statistically analysed.

Future Directions

We are developing a clinical tool ? "GBS Progress Form" - in order to ensure better documentation of clinical findings, get improved homogeneity of data. We shall kook at additional scales useful in the paediatric population.

We are considering asking CIDP patients to provide a blood sample to try and identify particular antibodies associated with CIDP.

Acknowledgement

We wish to thank CSL Bioplasma Ltd, Victoria for their Education Grant Neuromuscular Fellowship, RCH.

Donation of $5,000 to Neuromuscular Fund

As gratefully acknowledged by Dr Andrew Kornberg at the beginning of his talk, The IN Group has donated a further $5,000 to his research, making the total of our donations to date $60,500. These donations have come from a combination of many generous donations from our members plus our annual fund raising events of the Maling Road Cake Stall and the Summer and Winter Luncheons.

A very sincere thank you to the very many who have donated of their money and time to this great cause.

Winter Social Luncheon

MARGARET LAWRENCE will again be our hostess for The In Group **Winter Social Luncheon** to be held from noon on Sunday 20th June at her home 26 Belmont Street, Glen Waverley. $15 will provide a lovely luncheon as well as raising funds for medical research. Our young member **ANNA MELVILLE,** who has begun her music course this year at Melbourne University, will provide a pleasing recital.

Thanksgiving Celebration

Vilma Clarke 28/01/1935 ? 30/04/2004


Vilma Clarke 28/01/1935 – 30/04/2004

CLARKE.
Requiem Mass for the repose of the soul of the late Mrs Vilma Pita Clarke of Wangaratta, will be offered at St Patrick's Catholic Church, Ford St., Wangaratta on WEDNESDAY (May 5) at 10.30 a.m., followed by a Committal Service at the Crematorium Chapel, Union Rd, Albury at 2.30 p.m. In lieu of flowers donations may be made at the Service to the Inflammatory Neuropathy Support Group of Victoria.
GARLICK FAMILY FUNERALS
Mason St., Wangaratta
Tel: (03) 5721 5060

CLARKE, Vilma. – The Inflammatory Neuropathy Support Group of Victoria Inc. (The IN Group) mourns the loss of our outstanding committee member Vilma and extends our deepest sympathy to her family.

"HeraldSun" 3/5/04 "The Age" 3/5/04

A number of The In Group members attended the funeral of our outstanding and hardworking commit-tee member, held at Wangaratta.

The church was packed with her family and her many friends who mourned their loss and celebrated her many contributions to the well being of the community.

Some fifty of her acquaintances donated some $1,200 to The IN Group in lieu of flowers.

Thank you, once again, Vilma.

?HeraldSun"3/5/04 "The Age"3/5/04

GBS/CIDP Awareness Day 1st June 2004

The IN Group emailed a Media Release (copy enclosed) to the major papers ? "The Age", "Herald-Sun", "The Australian"; to three local papers ? "Progress Press", "Melbourne Weekly", "Toorak

Times"; and to Jon Faine of ABC Radio 3LO and Kerry O?Brien of ABC TV Ch 2 "7.30 Report".

Entertainment Books ? Last Chance

Nine of the 2004/2005 edition have so far been sold generating a profit of $108 (9x$12) to The In Group. Please send in your order, set out in the wrapper, by the 15/06/04.

A Temporary Fix to Gammaglobulin Shortage

A Shocking Cut to Intragam Supply

Many Victorians relying on treatment with gamma-globulin (Intragam P) for their GBS or CIDP diseases or their variants were shocked in late March to be advised that it was no longer available except for life-threatening cases.

Treatment necessary to live a normal life

"The Age" 26/03/04

Fortunately, through great media presentations of this shocking situation, particularly by **JON FAINE** 3LO Radio presenter, **KERRY O?BRIEN** ABC TV "The 7.30 Report" presenter and **CAROL NADER** journalist of "THE AGE", the Victorian Government found $1.7 million to enable a restoration of supply until the end of June this year, particularly through the importation of gammaglobulin from Switzerland, Sandoglobulin.

The reprint of Carol Nader?s article above ("THE AGE" 26/03/04), featuring three members of The IN Group needing gammaglobulin for a normal life, was a particularly telling pressure on the government.

During this temporary provision, current patients, who have gammaglobulin prescribed for their medical treatment, will continue to receive the Australian pro-duct, Intragam P, whilst new patients will receive the imported Sandoglobulin.

The Federal and Victorian governments have set up an **Intravenous Immunoglobulin (IVIG) Working Group** "to recommend guidelines and procedures for the appropriate allocation and distribution of the avail-able supply of IVIG in Victoria".

Membership of the Group includes representatives from: major clinical specialities which use IVIG, including medical, nursing and science; Australian Red Cross Blood Service (ARCBS), Victorian Department of Human Services (DHS); Patient/community groups. A representative of the Patient group is **JOHN BURKE,** an IN Group and now committee member, and of medical speciality is Dr **ANDREW KORNBERG**, of the Royal Children?s Hospital whose update on his research into GBS/CIDP is featured in this newsletter.

The Group is meeting monthly this financial year and then at least 3 or 4 times yearly.

The functions and roles of the Group are: to monitor IVIG usage (supply and demand) in Victoria; to advise ARCBS and DHS on the quantity of IVIG required to meet Victoria?s needs for the treatment of appropriate indications; to implement national guide-lines and to formulate local guidelines as necessary for the allocation and distribution of IVIG, according to current supply; to act as a forum for the evaluation and advice on new indications or treatment regimes for IVIG; to communicate recommendations to all stakeholders; to educate user, where appropriate, on appropriate use of IVIG; to consider research opportunities and applications; to receive reports from other relevant groups.

Need for a Permanent Solution

The IVIG Working Group should at least determine what is needed to make supply of IVIG meet the medical treatment. However it seems unlikely that the Group can make this finding by the end of this financial year, 30/06/04, noting the vast scale of information needed to cover this basic topic.

This means almost certainly that the Federal and Victorian Governments must at least provide the finance for enough IVIG for the first three months of the coming financial year 1904/05, ie probably another $1.7 million.

The IN Group will continue to agitate for a permanent solution and to make sure that never again will occur such a shocking shortage of treatment medication as presented last March. Having our representative **JOHN BURKE** on the Group will be a great help in pushing this need.

The IN Group will also continue to encourage the community to donate more blood and blood plasma to the Australian Red Cross Blood Service. Such donations are essential to provide for the manufacture by CSL Bioplasma Ltd of gammaglobulin Intragam P.

Report on IVIG Working Group

By JOHN BURKE

I attended the first meeting 19/04/04. How some of its activities are to be undertaken is not yet clear.

Sandoglobulin, a Swiss made IVIG has been made available to supplement supplies of the equivalent Australian Intragam P. It is expected that, if required, Sandoglobulin will continue to be made available beyond the present initial period ending 30/06/04.

In the neuropathy context Sandoglobulin is virtually equivalent to Intragam P. *(However A/Prof Alison Street of the Alfred Hospital stated at this meeting that the implementation of Sandoglobulin leaves a lot to be desired and is not working well between CSL/DHS/ARCBS and hospitals. In particular insufficient written information has been provided in advance and there has been no in-services provided for nurses administering the product.)* There are some differences in the way Sandoglobulin is prepared. However this will not affect patients. Patients currently receiving Intragam should continue to receive it and as far as possible only new patients, particularly patients only receiving a short treatment, will receive Sandoglobulin. The quantity of IVIG received by existing patients should not be less than what they were receiving prior to the March crisis.

An adequate supply of IVIG is critically important issue and we need to address the short, medium and long-term solutions.

It is not a simple fix and many issues are involved, such as increasing the number of people who donate blood and blood plasma. However the biggest initial problem is the level of Government funding for the ARCBS to collect blood plasma and for its production into Intragam by CSL Bioplasma Ltd.

If anybody wishes to speak with me I can be contacted as follows: John Burke, 15 Glen St. Ashburton 3147; email jburke@contracts.com.au; wk 98552899; a/h 98852377; mobile 0417885747; fax 98852866.

IN Group Letters to Health Ministers

The IN Group sent letters to the Federal Minister for Health, Tony Abbott, Shadow Federal Minister for Health, Julia Gillard and the Victorian Minister for Health, Bronwyn Pike seeking urgent action for a permanent solution to the shortage of gammaglobulin.

A spokesperson replying on behalf of Tony Abbott, advised that the Australian and State and Territory Governments have asked the National Blood Authority to immediately arrange for the purchase of additional imported IVIG (Sandoglobulin) but did not indicate whether this was a permanent commitment. The reply by Julia Gillard blames the National Blood Authority, responsible for blood and blood products since 1/07/03, for being poorly resourced and not effective due to it having to reach agreement with the Commonwealth and States. She ends with "a Labor Government will ensure that the NBA is able to undertake its important role so that no Australian is deprived of needed blood products". The best that Bronwyn Pike could promise in her reply was that "the Victorian and Australian Governments will continue to work together to ensure that patients requiring treatment with IVIG have access to the product." work together but no guarantee.

Another Successful Cake Stall Fund-Raiser

A total of $767.60, including $170 donations, was raised at the Maling Road Canterbury Cake Stall held Saturday morning 8th May. Many thanks to organisers **MARGARET LAWRENCE** and **BETTY GERR-AND,** helpers and contributors which included **BARBARA RIVETT, DOROTHY BRENNAN, BARBARA CLIFFORD** and **LUCY TIDBURY,** and donors which included **JOYCE MONTGOM-ERY, PHYL CAMERON, KEITH COLWILL, JUNE CATHCART** and **JEREMY GERRAND.**

The IN Group has a modern computer

GREG KEOGH, our computer wizard, who kindly updates The In Group webpage with our newsletters, noted that The IN Group?s computer, purchased in 1995 for $2,665, thanks to the donation of this amount by CSL, was slow and out of date. He arranged through a computer friend, to purchase the parts of a modern computer for the amazing modest sum of $500 which generous CSL again kindly donated. So now we have the Microsoft XP model.

Support through the Internet

Emails were received mainly seeking information about GBS or CIDP from Ben Lee (Washington USA), Bette Murchison (Chelmer Qld), Mandy Kretschmer (Blackwood SA), Gwen McInnes (Lysterfield Vic), Ewald Siedel (Templestowe Vic), Val Sewell (Patterson Lakes Vic), Leanne Eaton (Ormond Vic), Mary Batcher (Wisconson USA), Traci Lemons (Texas USA), Graham Wilson (Watsonia Vic), Debra Dunne (Virginia USA), Ian Kruse (Mulgrave Vic), Bernard Pettit (Narra Warren)

Pleasing email correspondence

The following is typical of much pleasing email correspondence.

To ingroup@vicnet.net.au

From Bette Murchison,

Have just been told I have CIDP and on looking at the Internet and I would like to join and receive newsletters. I know very little about this complaint but wish to do all I can to help my disability and would be pleased to hear from you. I am very ancient (88 in June) but care for my husband of 64 years and apart from this I am very healthy. CHELMER Qld.

Sent CIDP booklet, IN Group brochure and latest newsletter.

To ingroup@vicnet.net.au

From Bette Murchison

Very pleased to receive CIDP booklet and hope you have received my cheque for membership etc. ?

It is great to have contact with someone who knows about this rare disease. Do you know of any people in Brisbane with this disabling complaint?

I emailed Bette the names of two of our Brisbane members.

Ending on a Lighter Note!

Subject: Wit

A good pun is its own reword.
A man?s home is his castle, in a manor of speaking.
Practice safe eating ? always use condiments.
Shot gun wedding: a case of wife or death.
A man needs a mistress just to break the monogamy.
A hangover is the wrath of grapes.
Dancing cheek-to-cheek is a form of floor play.
Sea captains don?t like crew cuts.
Reading while sunbathing makes you well-red.
When two egotists meet, it?s an I for an I.

A bicycle can?t stand on its own because it is two-tired.
What?s the definition of a will? (It?s a dead giveaway.)
Time flies like an arrow. Flies like a banana.
In democracy your vote counts. In feudalism your count votes.
Acupuncture is a jab well done.
If you don?t pay your exorcist, you get repossessed.
Santa?s helpers are subordinate clauses.

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