

The Biliary Bulletin

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Summer 2002

Primary Biliary Cirrhosis (PBC) is an autoimmune disease which affects the liver and for which there is no known cause or cure. This newsletter communicates news and information to members of the Australian PBC Support Group and other sufferers of PBC.

From Rosemary

Dear Friends,

I hope you all had a happy Christmas. I would like to wish everyone a happy and healthy New Year. My thanks to all who sent such lovely Christmas cards and good wishes.

My wish for 2002 has to be that we get some good news from the doctors around the world who are working so hard to find the cause of PBC. Along with so many other PBCers I will be watching the outcome of Dr Andrew Mason's pilot study of Combivir therapy which started in November 2001, and will take six months (see article included in this issue).

Looking back to November and the pre Christmas get-together at Chadstone, it was lovely to catch up with everyone and spend a couple of hours chatting. Our Christmas hamper was overflowing with goodies - thank you to all who donated to make it a lovely day. I'm sure a good time was enjoyed by all

We had many raffle prizes too: Wendy Brien very kindly knitted a beautiful jumper, there were gift bags donated by Chemistop Dandenong and Amcal Chemist Endeavour Hills, lovely gift packs donated by Glenys Tyzack and perfume donated by Juliet Aspden. A delicious dessert was provided by Anthoula Panagiotopoulos.

I must not forget a very dear kind lady, Mary McInerney who made a donation to our group because she could not make it to Chadstone. Mary lives in New South Wales (long drive to Chadstone). Although Mary suffers from many other conditions as well as PBC, I know that she gives much of her time to helping others in her community. Mary is such a happy and positive person to talk to, she has a heart of gold. Thank you Mary.

Thank you too once again to all who made donations to our group along with their membership subscriptions.

The year 2001 was not the best year for me, I now need to take the advice of my good doctor to slow down. The meeting at the Austin in March will now be organised by Debbie and Liz. I know they will do an excellent job and I wish them both a huge success.

A message finally to those members who subscribe to our email list at Yahoo-groups.

I receive many emails asking how to unsubscribe and re-subscribe when changing email addresses. To remove your old address send an email from your old address to oz-pbcers-unsubscribe@yahoogroups.com

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To add your new address to our list send an email from your new address to oz-pbcers-subscribe@yahoogroups.com

To post a message to the list send your email to oz-pbcers@yahoogroups.com

Keep on keeping positive everyone, best wishes to all

Rosemary

The Source of Ursofalk *Rob Chiarolli*

For some reason, I presume a TV or newspaper report, we have had a few inquiries from gastroenterologists as to whether Chinese Black Bears are actually harmed during the manufacture of Ursofalk.

I have made inquiries with the German manufacturer and am happy to report that Chinese Black Bears are NOT sacrificed for the manufacture of Ursofalk, although it was the original substance the active ingredient was identified in (hence the name "urso", latin for "bear").

The basic building block (chenodeoxycholic acid) is actually derived from the bile of cows, which is then synthetically modified to the final product. The cows are farmed and slaughtered for normal commercial use.

Interestingly the source of the cows includes USA, South America AND Australia and this is to avoid the potential for BSE ("Mad Cow Disease") -infected cattle from Europe and Asia.

Regards,

Rob Chiarolli,

Orphan Australia

Member profile

Glenys Tyzack

After some gentle persuasion and arm bending by Rosemary and Jocelyn I was volunteered to put pen to paper and give a bit of a story about myself and PBC. As you can imagine, how could I refuse with one on either side and hope to escape in one piece. I am sure you can relate to this but, the best part is, someone else is going to be nominated for the next issue – good luck.

My name is Glenys, I am a pretty young 52 year old living in Melbourne and was diagnosed with PBC August 2000. I had been unwell for about a year or so, constantly tired with all sorts of aches and pains.

For a while I put it down to ‘the job’ but really knowing deep down it was a little more than that. My bones ached and getting out of a chair after sitting for a while was a bit of a chore. It wasn’t just the sitting though and the way I could tell it was the bones and not the joints was because of the shape – sounds crazy but hey, I could tell you about the colour of the pain as well – that rates between mild and very bad ...anyway; as well as that there was the nausea – it seemed never ending, but worse really is the constant ache in the right side – again, from ache to pain – mild to b. awful.

I simply kept going back to my doctor, who; god love her, kept sending me for every test known to mankind. The aches/pains are still there, but a little more controlled now.

The bone ache is the greatest improvement – I don’t creak, groan or hobble anywhere nearly as much as I used to – but to my mind, the medication is making fluid retention a problem, the old feet are worse and extremely bad on the warmer days, the rest can be covered up but it is hard to get shoes to fit.

The nausea investigations showed stomach ulcers- they seem to come and go like old friends but I have also noticed mouth ulcers seem to be appearing far more frequently – the worst being on the tongue – mind you, there’s a great product like cement you paint on which is fantastic; my husband also likes it as it keeps my mouth shut; for a while at least.

Anyway, after a biopsy, the diagnosis was given. To say I didn’t understand is an understatement – but, at last I had a name; it wasn’t just in my head, relief flooded over me. Then as some explanations were being given to me I started to lose it. You see, I’m sure that just like many of you – I picked up on the words – auto immune – cirrhosis – transplant. I then thought – bugger – I shouldn’t have enjoyed a drink so much, at least the explanation that it wasn’t “that” cirrhosis was a relief. (temporarily anyway).

After leaving the surgery on that day, explaining to the family was a problem. I didn’t really understand or know what I was talking about. Then I went through the “why me” – “what does it mean” – “I don’t want to know” stages.

I rang Rosemary after 4 – 6 weeks and had a chat. I then received some literature from her and from family. At this point I began to stop feeling sorry for myself and started to read, but also, continue on as I had always been doing – the only difference for me was having a name to the problem – having pills to help slow all the ailments down, but also now being aware of just how much you need to listen to your body.

My family are trying to understand and help on the bad days, the pain and tiredness continue to bug me but I won’t let it get to me as I continue to remind myself there are others far less fortunate than me. My work (new job, less stress) and my family keep me going and support from the other end of the telephone when needed is fantastic. It is a great idea to be able to phone others with similar problems or to get together occasionally to swap notes and meet partners and families if you want to

I still don’t understand as much as some others in the network, but we all learn more every day – even the professionals.

I will sign off by wishing you all good day and good luck.. Maybe we’ll meet at another function or get together, the lunch in early July was lovely; thanks to Rosemary; hope to see more faces next time. Glenys.

P.S. My husband said I forgot to put “crabby” as a symptom – sorry folks.

Liver Transplantation and “Splits”-Two for the price of one

Dr. Paul Gow

Liver transplantation was developed in the 1960’s in North America. Initially it was considered a radical therapy with an uncertain future; it is now a widely available, readily accepted and successful treatment, with a majority of transplanted patients achieving a good outcome with a return to a high quality of life after the procedure.

In Australia the first liver transplant unit was established in Sydney in 1985 and subsequent units developed in all the Australian capital cities (with the exception of Hobart). Improvements in surgical techniques, anaesthesia and intensive care and refinement of immunosuppressive protocols have resulted in an expansion of the indications for transplantation.

This has resulted in an increasing number of patients being referred for and awaiting liver transplantation. Unfortunately the number of available liver donors has not increased at the same rate as the number of potential recipients, resulting in increased waiting lists for the procedure.

In Australia about 10% of all liver transplants are performed on children. Due to the small size of children, often a donor liver has to be “cut down” (reduced in size)

by the surgeon so it may fit into a child. The unused portion of the liver has historically been disposed of. Over the last 10 years surgical techniques have been developed such that, in some cases, the liver can be divided in two so that two recipients may benefit from the one donor liver. This is called "split liver transplantation".

Due to the disparity between waiting lists and available donors many units throughout the world have implemented the "split liver" technique so that as many recipients as possible may receive a donor organ. Usually a split liver is shared between a child and a small adult, or between two small adults. The split liver rapidly grows to a normal size in the recipient and functions normally.

Split liver transplantation is just one of the developments that have occurred in the area of liver transplantation over the last decade. Other improvements have occurred in surgical techniques, immune suppressant medication and intensive care of patients, so that as many patients as possible may be transplanted and achieve an optimal outcome.

Is Primary Biliary Cirrhosis caused by a virus? *Dr. Andrew Mason*

After many years of study, we still do not know what causes PBC. In patients who suffer from PBC, we know that the body's defence system appears to attack parts of liver. We also know that this immune response is directed to proteins that are part of the human body and this is referred to as an autoimmune response.

In this regard, PBC patients make Anti-Mitochondrial Antibodies, also known as AMA, that react with human mitochondria, which are small structures found within most cells. We do not know why this autoimmune response occurs in patients with PBC. Also, we don't know whether the autoimmune response and the AMA cause PBC.

If this were so, then treatments that decrease the immune response should work but these therapies do not seem to help patients with PBC. We know that AMA have an important role in making the diagnosis of PBC but they do not give an indication of how severe the disease is. Some patients without AMA have severe PBC, whereas others with AMA may have no disease at all.

There are data to suggest that PBC may occur as a result of an infection. PBC tends to occur in clusters and can be seen in non-related family members and caregivers. Also, there appears to be an increase in the number of people with PBC in Northern Europe. In liver transplant patients, we find that PBC can reoccur many years after transplantation, which suggests that an infection may stay in the body.

At Ochsner Clinic Foundation we have used many techniques to detect an agent in the liver samples from PBC patients. We narrowed down the search to a virus,

once we found serum reactivity to retroviruses in patients with PBC. In this study, we found that most patients with PBC had antibodies to a retrovirus isolated from patients with another related autoimmune disease, Sjogren's syndrome.

With James Neuberger's research group in Birmingham, UK, we have made a model to study whether an infection causes PBC. We performed these studies by growing normal bile duct cells in a dish and then adding lymph node samples, which are small glands attached to the liver that are removed during liver transplant. Only the bile duct cells mixed with PBC patients' samples developed a change found with PBC, which we thought was due to an infectious agent in the PBC patients' samples.

Once we believed that PBC might be caused by a virus infection, we used specific laboratory techniques on tissue samples from patients with PBC to isolate genes from a new virus. It is very difficult to prove that this new virus causes PBC. However, we believe that the virus that we have identified may cause the disease because most patients have some evidence of virus infection.

We have also found that virus can alter normal bile duct cells to appear like the damaged cells only seen in patients with PBC. In a pilot study, we have found that anti-viral treatment leads to improved findings on the liver biopsy as well as with the liver function tests. Although it is tempting to think that PBC may be caused by a virus infection that can be attacked by anti-viral treatment, these studies need to be confirmed in controlled trials.

We need to finish the lab studies and then our work has to be reviewed by other scientists and published. After this, other doctors will want to repeat the studies to see if they get the same findings as us. We need to repeat our pilot studies using anti-viral treatment in a controlled trial to assess whether the drugs actually work and are safe.

We are now starting a second study *A PILOT STUDY OF COMBIVIR THERAPY FOR PATIENTS WITH PRIMARY BILIARY CIRRHOSIS* to see if being on anti-viral treatment is better than not being on this treatment.

The study will start in November 2001 and patients will be treated 6 months and then need a 6 month follow up period. The study will be performed at the Ochsner Clinic, New Orleans, LA and the Mayo Clinic, Rochester, MN as well as Birmingham, UK.

To get into the study, patients must have increased liver tests for more than 6 months, serum AMA and a liver biopsy showing PBC. Patients should also be on standard ursodeoxycholic acid therapy for 6 or more months. Both the anti-viral and ursodeoxycholic acid treatment will be supplied free of charge for the study.

For further information about the study, please contact Dr. Andrew Mason at Ochsner Clinic Foundation.

E-mail amason@ochsner.org



Pre-Christmas Get-Together at Zampelis, Chadstone, VIC.

Meeting at the Austin Hospital

On Sunday March 3rd there will be a meeting in Zeltner Hall, at the Austin Campus of the Austin & Repatriation Medical Centre, from 2 pm – 5pm.

The theme is 'Living with PBC' and the speakers will be either Dr. Peter Angus or Dr. Paul Gow, and physiotherapist Jacqui Lynch.

Further information will be sent to Victorian members closer to the date of the meeting. For any queries please contact Liz (03) 9563 7190 or Debbie (03) 9571 3735.

Hope to see you there., Jocelyn.

HelpWanted

Due to my ever-decreasing energy levels and my problems with concentration etc.. I would love to have someone to assist with the production of the Biliary Bulletin.

A computer and access to e-mail would be required. The job mainly involves collecting material in between issues. I am still capable of quite a bit of input so it wouldn't be a case of having to step in and take over – unless of course, there is someone who would like to do that. The group has a double sided A3 printer (kindly donated by Orphan Australia) and I would be happy to keep doing the final editing and printing.

Sometimes the hardest part is “nagging” people to write or send in material they have promised! (See the profile from Glenys, and below) ✍

At the moment I am rather short of material for future newsletters. Everybody loves to read about the experiences of other members, however I do not have a Member Profile for the next edition and I really need several of you to put pen to paper and supply me. I have tried a more personal approach with some people, but that hasn't had the desired result!

Thank you to those people who alert myself and Rosemary to articles they discover in the press and on the Internet, and also to those of you who send us letters. Keep it up please!

Jocelyn.

Letters

Rosemary, Jocelyn and Friends,

Just a brief note to express my thanks for the concern, prayers and the many kindnesses extended to me through this significant period of my life. The lovely arrangement of flowers also cheered me up no end.

It must be so heartening for you both to see the “fruits” of your efforts as many of us on being diagnosed with PBC are at a loss to deal with the news and are “hungry” to learn more about the illness and to share the experience with other PBC sufferers. Your group has certainly met many of my needs.

Thanks again, Doreen Cheong

Dear Rosemary,

Thank you very much for the last newsletter. It came just at the right time as I was feeling very, very tired for a few weeks but I strongly believe in my saying “I'm lucky”. I can see, walk, talk and use my hands.

Then the bulletin arrived and I realised I was indeed lucky and it bucked me up no end – thank you.

Kindest regards, Mary McInerney

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