

The Biliary Bulletin

Volume 3, Issue 4

Spring 2001

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 are all keeping as well and positive as possible.

Recently I have talked to many people in our group who are not doing too well right now, my thoughts are with all those people. I hope that life gets better soon.

My very best wishes to Doreen Cheong who received her gift of life recently at RPA in New South Wales. Wishing you continued good progress Doreen, your bright and positive attitude is an inspiration to us all.

We need to continue to promote organ donation. I have organ donor registration forms and car stickers, and I am happy to send them to anyone who would like to give them out.

It is three years since our group was formed. We have grown so much in that time, we have to date about 172 members but have been in contact with and sent out information to many more people diagnosed with PBC - somewhere around three hundred. There is a long way to go but it's good to know that all of those people were able to access information about PBC.

I usually ask people when they contact me how they found us - their answers can be amazing. It can be a doctor in St. Kilda giving out our brochure, to a doctor in a hospital in WA. Of course many find us through our web site at www.oz-pbc.org.au where for new members who have access to the Internet, all back issues of our Biliary Bulletin can be read.

We also ask people when they first contact us if they are taking Ursosalk, and we are very often surprised and disappointed that there are still some patients who are not being given information about it. At least if patients are given the information that Ursosalk is available they can then make the decision whether or not to take the medication.

To those who decide not to take it I would recommend that you listen to the tape *Long Term Effects of Ursodeoxycholic Acid Therapy in PBC* from the PBCers conference.

Jocelyn and I are happy to talk to people on the telephone but of course we cannot be available all the time. If you need to contact me and my answering service is on, please leave a message and I will get back to you.

There are days and evenings when my telephone rings

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constantly, so from time to time I need to switch off to get some rest myself, but once again if you leave a message I will return your call.

The countdown to Christmas has started once again, how time flies! Every year I intend to do my Christmas shopping early but never manage to get round to it. I envy people such as Jenny in Tasmania who is always organised for Christmas halfway through the year. What bliss that would be, maybe you could come over and get mine organised Jenny, but I guess I am like many who have good intentions but always run out of time.

Thank you once again to all who have sent in donations with their memberships.

Keep on keeping positive everyone, best wishes to all.

Rosemary.

Pre Christmas Get Together

Please join us, and bring your family on Saturday 17th November 2001 at Zampelis, Chadstone Shopping Centre, Chadstone, Victoria.

They have a marvellous array of food from sandwiches to full meals, and the desserts are Bellissimo. (Being told you have to put on weight is just one of the bonuses of being on the transplant waiting list.) And of course the company will be excellent!

We will raffle a Christmas hamper which we will put together on the day - so please bring a small contribution. If you would like to purchase raffle tickets please contact Rosemary. (Unfortunately limited to Victoria only). Hope to see as many as possible there!

Jocelyn.

Member Profile

Sue Spain

Hi, my name is Sue Spain (age 53) and I thought some of your readers may be interested in a profile of my PBC history.

I receive and read with interest many e-mails and the "Biliary Bulletin" and often think to myself, that's me, or "I can relate to that" as it's all happened to me over the years.

At the age of 22 I was put on the pill and shortly afterwards I presented to my Dr. with very itchy skin for no apparent reason, and jaundiced. I was quickly taken off the pill but the itchy skin persisted over the years in times of ill health or when my body was under any stress.

At 25 I married and as we'd married in South Africa and I wanted to return to NZ, we honeymooned on a ship to Auckland. By the time I got there (after what I thought was sea sickness) I had lost a lot of weight, was jaundiced and had itchy skin and was generally feeling very sick.

My Dr. incorrectly diagnosed my condition as Hepatitis. It took 3-4 months to recover from this bout and life went along quite smoothly for about 5 years until I fell pregnant. All the earlier symptoms returned and I lost that baby at 26 weeks.

The following year I was pregnant again and gave birth prematurely at 32 weeks. The baby was born healthy and today is a very healthy 23 year old.

By this time all the consulting Drs thought the problem was my gall bladder so scheduled me for surgery. During this they discovered my liver and spleen enlarged and "cloudy lungs" (I've never discovered the reason for the lungs being like that) but at the time this condition was labelled sarcoidosis. They did find some gallstones at that time so that was thought to be the problem.

I got back on my feet eventually and four years passed. We wanted another child although Drs warned that this could mean time in hospital. This pregnancy was up and down again and by the 6th month they admitted me to hospital for the duration. Kelly was born about 3 weeks early and then it was a case of getting back to normal life.

We shifted just after this from Sydney to Gladstone in Queensland where we now live and my GP thought it would be a good idea to go to a specialist to have a check up. The specialist told me I had oesophageal varices and in time would need a liver transplant.

About 5 years later I had a life threatening bleed from the varices and from then on until 1992 (10 years) I received regular endoscopies and sclerotherapy for them.

By 1992 I was seriously ill, losing weight, jaundiced, and with many other symptoms. I had my life saving liver transplant in December of 1992.

So now I am coming up to my 8th year post transplant and feeling fine. My biggest problem is keeping the weight off but compared to my condition before, that's nothing.

It wasn't until I had my transplant that my disease was finally given a name: Primary Biliary Cirrhosis.

Sue Spain.

Primary Biliary Cirrhosis and problems outside the liver. (I)

Dr. Katrina Watson

People with primary biliary cirrhosis (PBC) have an overactive immune system. Immune cells called T-lymphocytes attack the liver, and result in secondary damage to liver cells by bile acids. People with PBC seem to have a genetic tendency to this overactive immune system. Occasionally the overactive immunity can affect other parts of the body, away from the liver.

About 40% of people with PBC will have aches and pains in their joints and muscles. Some of these people (about 10%) may have an actual arthritis, called simply "the arthritis of PBC". A small number (5 to 10%) may have classical rheumatoid arthritis. This condition may need particular arthritis medication.

A syndrome of dry eyes and dry mouth is quite common in PBC. This syndrome is called "Sjogren's Syndrome", and can occur in about 50% of people with PBC. Quite often the symptoms come on before the diagnosis of PBC is made. Eye drops can be helpful to relieve the dryness in the eyes.

About 10% of people with PBC may have a version of another autoimmune condition called scleroderma. This condition causes tight skin. Fortunately, the scleroderma associated with PBC is usually mild. It may cause Raynaud's phenomenon (blue, red or painful reactions in the fingers when it is cold), skin changes in the fingers or face, or reflux oesophagitis (ulcers in the oesophagus). However, the scleroderma of PBC does not usually cause life-threatening problems such as severe hypertension or kidney disease (other forms of scleroderma can do this). The reflux oesophagitis responds well to anti-acid medications (proton pump inhibitors).

Another autoimmune problem which occurs with increased frequency in people with PBC is thyroid disease. It is quite common for people with PBC to suffer from an underactive or, less commonly, overactive thyroid gland. The overactive immune system attacks the thyroid gland, causing "thyroiditis". The problem is easy to diagnose, using a blood test, and sometimes a thyroid scan. Fortunately people respond very well to treatment with thyroid hormones (thyroxine, or Oroxine).

High cholesterol levels are another accompaniment of PBC. The good news is that these high cholesterol levels do not cause heart disease, vascular disease or strokes, as a

general rule. Fortunately, people with PBC often have high levels of a “good” lipid - the high density lipoprotein (HDL) - this seems to counteract the effect of the cholesterol, and the “bad” lipids (low density lipoproteins or LDL) are usually low. There is a special lipid called “lipoprotein X” which is elevated in PBC.

The functions and significance of lipoprotein X are unknown, and are the subject of ongoing research. People with PBC can develop “fatty deposits” around their eyes (xanthelasma) or under the skin elsewhere (xanthomas). Although people with PBC are not at increased risk of vascular disease, it is probably a good idea to keep an eye on the cholesterol and lipid profile by occasional blood tests, and follow a low-cholesterol diet if the tests are high. Some of the usual cholesterol-lowering drugs don't seem to be very effective in PBC.

What does all this mean to the average person with PBC? Although you may be at increased risk of other autoimmune conditions, the conditions are usually mild and fairly treatable. You should let your doctor know of any symptoms you may be having. Your doctor may wish to check blood tests for thyroid function and high cholesterol every so often. Your doctor may also suggest tests to watch for osteoporosis (the subject of another article, in the next Biliary Bulletin).

Book Review

Victoria Lyn

Dr. Melissa Palmer's Guide to Hepatitis and Liver Disease; What You Need to Know by Dr. Melissa Palmer

I have been amazed, pleased and daunted by Dr. Palmer's book. There are moments when I wish it had been handed to me along with my diagnosis of PBC in February 1998, and moments when I feel it would have been too much information too soon.

The first surprise for me was that PBC is strongly represented in this book, not only with a chapter devoted to PBC, but also in chapters about tests, symptoms, treatment – even in the section pertaining to nutrition and exercise she is specific about PBCers needs vs. the needs of a patient with hepatitis.

The second surprise is that she explains the reasons behind her recommendations, leaving me with a greater understanding of why certain things are suggested for our disease.

In the chapter on nutrition she addresses one reason many of us experience fatigue – “Converting foods other than carbohydrates into energy is stressful even to a normal liver. By eating an unbalanced diet that is low in complex carbohydrates, a person with liver disease will add to the stress that the disease has already caused the liver. In fact, this is one reason why so many people with liver disease feel fatigued. Simply put, their diets are working against them. A well balanced diet can help combat the fatigue

associated with liver disease. Eating multiple small meals throughout the day instead of three large meals is recommended.”

This certainly may not be new information for many PBCers, but the manner in which Dr. Palmer explains the mechanics of nutritional intake gave me a clearer picture of how best to care for my body.

Many of us have families and friends that have difficulty understanding all we are dealing with and this book may be of help. Because of its length others may not wish to read its entirety, but you could copy sections to share.

I do recommend this book as an in depth resource manual, as a support in moving more fully into being an informed patient and in being our own advocates.

Reprinted with kind permission from the September edition of the PBCers newsletter.

Letters

Hi,

My name is Liz Allan and I hesitated to write as I had PSC (Primary Sclerosing Cholangitis) not PBC, and even that diagnosis was uncertain.

I was ill for about 4 years and kept working and studying part time. Prior to that I was raising my family and working full time in child care. The only times I had ever been in hospital were when my children were born.


In the last few years I have had trouble with insomnia, tiredness, itching and pain, but still had some good days.

Then my liver went into failure. Fortunately I had a liver transplant and due to the skills and care of the wonderful transplant team at the Austin Hospital, I am facing life anew!

I have to remind myself now how far I have come in such a short time and I get new pleasure in the love of friends and family, good food, movies, music, walks along the beach, and just enjoying each day as precious.

The support group has been great, particularly Rosemary, Jocelyn and Maria who are a wealth of information and inspiration.

Liz Allan.



Dear Jocelyn,

Firstly, thank you most sincerely for your great effort in producing the newsletter, it has helped me enormously. Thank you also for the brochure – my daughter has been behaving very much like an ostrich when it comes to learning about PBC (if she ignores it perhaps it will go away) but she sat down and read the brochure and is finally being educated about it.

In hindsight, the onset of my PBC occurred in 1975 when I was just thirty, but it was only two years ago that I consented to a biopsy and the diagnosis was finally made.

I had been going to doctors and specialists for many years trying to find out why I was so tired but jacked up at having the biopsy. Finally it became impossible for me to continue working as I was so exhausted, so I had to agree to find out exactly what was wrong. It was only three years ago that it was mentioned that PBC might be the problem.

I am now on four tablets of Ursosalk a day, and it has made a huge difference to my life. Twelve months ago my specialist was considering putting me on the transplant list as my quality of life was so poor, and to be honest, I wouldn't have cared less if I had to shuffle off this mortal coil, but after increasing my medication, things are hugely improved.

I am able to walk most days now and that certainly helps the self-esteem. One thing I did find after increasing my medication was that I piled on two stone in weight in a matter of two months but I have managed to lose half of that now that I am walking and hope to clear it all up some day in the future. As long as I can keep on an even keel, life goes along quite nicely. Of course, I get tired easily, but I can cope with normal daily life now that I no longer have to work. I find the itch comes and goes, but it is never so bad that a good scratch doesn't work.

There are no PBC people from the Sunshine Coast in your contact list, so I asked my specialist if he had other patients with PBC and he said he has ten or twelve. He has taken note of your web site and address so I hope he passes them on to other patients as I know this has helped me hugely.

Thank you again for being such a help,

Sincerely,

Wendy Thorpe.

From the Editor

The last few months of being on the waiting list for transplant has been a busy and satisfying time for me. My children are all back in one place at last and I am gaining great pleasure out of being fairly involved in the happenings of their lives. John and the children are wonderfully supportive, and although there have been some difficult moments, life is really very good these days.

Under instruction from the dietician and the physiotherapist at the Austin Hospital I have been trying hard to gain some weight and become as fit as possible for the operation ahead. This was made more difficult by my feelings of disgust at any food that wasn't low-fat, and the extreme fatigue that had already forced me to cut back on walking, my only form of exercise.

However, going slowly, taking one small step at a time, I have managed to reintroduce some full cream milk products and even scoff the odd Danish pastry or a Devonshire tea.

I have also found a local aquaerobics group that is a great deal of fun with the added bonus of new friends there. This form of exercise seems to be very suitable - the water offers both support and resistance, making it possible to build and tone muscles without the strain that would be otherwise be involved. When I first started I had to work at a reduced pace because I tired quickly and it seemed to take days to recover if I overdid it, but now I find that I can tolerate a much more sustained session, and the recovery time is shorter.

So I have put on two kilos - not bad for some one struggling not to lose weight all the time, and I feel much more able to cope physically with the demands of life. If I get much better maybe I won't need a transplant!

You will notice that we have published a couple of letters this issue. Thank you to those contributors - I am so happy to hear from you and it certainly makes my job as editor much easier to have material for the next edition. So, bearing this in mind, perhaps YOU could sit down and write a letter or a member profile for our next issue - we are running very short of material at the moment. New members in particular, please tell us your story or give us some feedback.

Please make a note in your diary that there will be a talk at the Austin Hospital by Dr. Peter Angus or Dr. Paul Gow, and Physiotherapist Jacqui Lynch entitled "Living with PBC", on March 3rd, 2002. More details in the next issue.

Jocelyn.

The Biliary Bulletin is printed with the kind assistance of Orphan Australia



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