
The Biliary Bulletin

Volume 3, Issue 2

Autumn 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

Welcome to all the new members who have joined us since the Summer edition of the Biliary Bulletin. It was good to meet some of you at the St Vincent's meeting, and I hope that you found the talks given by both doctors informative and helpful.

We were fortunate once again to have two doctors willing to give their time to talk to us. Dr Katrina Watson gave us an excellent overview of PBC and Dr Ian Kronborg talked to us about the management of PBC in a way that we could all understand.

Dr Kronborg was adamant that people who have PBC should not drink any alcohol at all. In the past we have heard that we should maybe only drink alcohol on special occasions or in moderation but as Dr Kronborg said, alcohol can damage the liver so why would you take the risk? I do not drink alcohol at all and would agree with Dr Kronborg on this - I am not prepared to take the risk.

My biggest problem right now is lack of sleep. I am sure that many of you have the same problem so if anyone has any good tips to help to get a good night's sleep I would be grateful to hear from you. Better still send your tips to our newsletter editor Jocelyn, then we can all read them.

The meetings we have had with doctors participating have played a great part in educating many of us about PBC. Education and meeting others who have the same illness I feel, makes us stronger and more able to accept and cope with this frustrating disease. Please make use of your contact lists and keep in touch with others. Share your thoughts and concerns, and be more educated about the disease you have.

It is very unfortunate that the meetings are only held in Victoria. They do take a great deal of time to set up and there is no way I could organise a meeting interstate. However I know that there are many good doctors in all states who would be willing to give their time to help educate people about PBC - they would only need to be asked.

If anyone would like to organise any type of get-together we can help. Just send us your details, place, date, time etc., and we would be happy to print out flyers for your event and post them out to all members in your state.

I am hearing from people who have been given our brochure, so they are getting around. Thank you to all who have put them out in many places - they have made a difference. If you would like more brochures just let us know.

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May 30th is International PBC Day; a day when we feel particularly close to PBC sufferers around the world, close to all those who share the same fears and concerns associated with PBC.

To try our very best to have positive thoughts and keep ourselves educated is the most important thing we can do for ourselves, so to this end let us continue to support each other by sharing information and promoting positive thinking to our PBC friends everywhere.

My very best wishes to all PBC sufferers and their families.

Rosemary

Our Library

We have a library of audio and video tapes that may be of interest to you. The tapes were recorded at the PBC Conference 2000, in Las Vegas. The titles are:

Diagnosis of PBC... Dr. Howard J. Worman

Ursodiol Therapy for PBC... Keith Lindor MD

Combination Therapy in PBC... Marshall Kaplan MD

Where Are We and Where Are We Going? (PBC Research) ...
M. Eric Gershwin MD

Diet and Menus for PBC... Norma J. Thiel RD LD

Love Your Liver and Live Longer... Sandra Cabot MD

Please feel free to borrow these, or we can record them for you for the cost of the tapes.

Member Profile

Pat Kohr

I am a retired teacher living in bayside Wellington Point (near Brisbane) with my husband Colin. We have two daughters living nearby and two wonderful grandchildren.

Over several years, routine blood tests showed elevated LFTs, but I had no other symptoms until 1997, when an intolerance for many foods, fatigue and the dreaded itch became a problem. My GP sent me to a very clever gastroenterologist, who immediately thought of PBC and confirmed this diagnosis with a liver biopsy shortly after, even though it showed I was still in Stage 1.

I was completely devastated by the news of this previously unknown (to me) chronic illness, and in spite of outstanding family support, felt very alone and depressed.

My son-in-law found plenty of information via the Internet, and I joined support groups in the USA and the UK, which helped me somewhat to come to terms with my illness. Then I received a few letters from people in Australia, one of whom was our very own Jocelyn, and for me that was the turning point. I realised that I was not alone in this part of the world, that life still had plenty to offer and that wasting time on regrets was very unproductive indeed.

I have now been taking URSO for some time and feel much better. It has made a great difference in my life. The itch is considerably improved, the fatigue much less and my LFTs are almost down to normal. I have learned to rest instead of pushing myself too far and life is good again.

I am very grateful to the group in Melbourne for their efforts in keeping us together and informed of any new developments, and only wish that Brisbane and Melbourne were closer geographically.

My very best wishes to all of you.

Pat.

From the Editor

Over the last several years I have met, talked to and compared notes with, many people with PBC in Australia, England, USA, Canada and Asia. Apart from great similarities in symptoms and patterns of the disease, there are certain common and recurrent emotional themes.

Most of us seem to be very shocked when we first hear the diagnosis and wonder what this really means – how it will affect our lives.

Information can be hard to find – not everyone has access to the Internet and medical text books can be very dry and scary. At this point it is very easy to play the ‘ostrich’ game and bury your head firmly in the sand!

This method of coping however, is usually only good for a relatively short time. For many of us the progression of

PBC brings symptoms which do affect the way we live and are difficult to ignore.

There are some things that you can do to make it easier:

- Try to deal with the diagnosis of a chronic, progressive auto-immune disease slowly – it takes time. Allow yourself to grieve for the ‘good health’ status you have lost.
- Find supports. If your family don’t understand or aren’t very supportive make contact with other support group members. Talking about your feelings and fears helps. Don’t forget your family may be having difficulty dealing with it too.
- Find a doctor who has other PBC patients or a good working knowledge of PBC and doesn’t mind answering your questions.
- Ask your doctor for copies of tests as they are done and ask her/him to explain what they mean.
- Read about PBC, ask those who know for more information. The more knowledge you have, the less scary it becomes.
- Make lifestyle changes that will make you feel better, like adopting healthier eating habits, giving up (or cutting down) smoking and alcohol, doing more exercise. Each of these things brings the short term result of making you feel better (and proud of your achievement!), and the long term result of making you healthier.

Yes, all of these things take time and energy – often hard to come by, and are sometimes difficult to achieve.

I have found that this approach has really helped me and enabled me to live life now, and enjoy it immensely without the fear of not knowing what the future holds.

Of course, everyone is different and what works for some does not work for all!

Please write and let us know how you have coped.

Jocelyn.

Recommended Reading:

- *Dr. Melissa Palmer’s Guide to Hepatitis and Liver Disease* – Melissa Palmer, MD.
- *The Liver Disorders Resource Book* – Dr. Howard Worman

State Organisers

Would you like to organise a meeting, get together or a social function in your state? We will help in any way we can. For example we can print advertising material and post it out for you, and we can offer advice based on our experience in Melbourne (No charge ☺).

International PBC Day *May 30th*

It would be good to get members together to celebrate this day. Are you interested? We would like to hear from anybody with ideas on what kind of get together we should have, and where. If you would like to host a get together or plan one somewhere, please contact me. I would be very happy to print invitations or flyers or assist in any way I can.

Jocelyn.

PBC Primer *Dr. Katrina Watson*

What do the words 'Primary Biliary Cirrhosis' mean?

'Primary' means that the condition has no external cause. For example if a bile duct is damaged by surgery, 'secondary biliary cirrhosis' can result. A gall stone stuck in the main bile duct for many years can block the bile outflow of the liver and cause 'secondary biliary cirrhosis'. However in primary biliary cirrhosis there is no external cause - the problem arises in the bile duct itself.

'Biliary' means that the condition arises in the bile duct - these are the tubes that drain bile from the liver. The liver makes bile for several reasons - to get rid of toxins from the body, and also to digest fat in the duodenum. The ducts which drain the bile into the duodenum are the bile ducts. In PBC these ducts become scarred and narrowed so that bile flow slows down (a bit like converting a two lane highway into one lane).

'Cirrhosis' means scarring of the liver itself. The slowing of bile flow causes back pressure in the liver, and the liver responds by growing scar tissue. The eventual result is that the liver loses its smooth, soft, shiny appearance and becomes knobbly and hardened. However it is important to remember that most people diagnosed with PBC do not actually have cirrhosis. In fact many may never develop it, with current treatment. The process of scarring in the liver is, fortunately, extremely slow.

What are the effects of having PBC?

Many people have no symptoms at all for years or decades. However, over time, symptoms can develop. The commonest are tiredness and itching. Later on other symptoms or signs can show up, including pigmentation of the skin or fatty deposits round the eye. Later still PBC patients could develop evidence of severe liver disease (swelling, bleeding, bruising) or vitamin deficiencies (the 'fat-soluble' vitamins A, D, E and K). Because vitamin D is important for bones and PBC patients have a tendency to osteoporosis, vitamin D levels should be checked even in early stages.

What is the cause of PBC?

No one knows the full story, but it is clear that there is a 'genetic component' to PBC - i.e. certain populations have a higher tendency to PBC (especially Anglo-Celtic populations). However it can occur in any ethnic, racial or

social group. Females are much more likely to develop PBC than males (ratio 9:1). This genetic tendency seems to lead to an overactive immune response to something in the environment. We don't know what! Maybe it is a common virus or bacteria, maybe a food component - we wish we knew. Whatever the 'triggering factor', PBC is an anti-immune disease - it comes from the patient's own immune responses.

How is it diagnosed?

Most often PBC is suspected after a blood test - liver function test - is performed. Sometimes this has been done for a routine health check up in someone with no symptoms. The diagnosis may then be strongly suspected if an antimitochondrial antibody test is done - this test is positive in 95% of patients with PBC. A liver biopsy is needed to completely confirm the diagnosis, and 'stage' the disease i.e. decide how much scarring is there.

What else do I need to know?

Anyone with PBC will want to know as much about treatment as possible, and this topic is going to be discussed by Dr Ian Kronborg.

Some other important topics to be covered in future newsletters are 'Other autoimmune conditions which can occur with PBC' and 'How do I look after my bones?'

Katrina Watson

Thank You

To the many people who enclose donations with their subscriptions. It is a real help with costs and lessens the need to have fund raising events all the time!

Thank you also to those people who have been sending stamps - it all helps.

Treatment of PBC *Dr. Ian Kronborg*

Summarised by John Holman

Dr. Ian Kronborg kindly outlined his personal view on the use of UDCA (e.g. UrsoFalk) in the treatment of PBC. For the impatient or inattentive, he suggested that it could be simply summed up by the rule: If you are not yellow, take UDCA; if you are, consider a liver transplant - and take UDCA anyway.

Dr. Kronborg outlined the history and natural history of the disease, and discussed the treatment therapies which have been used to date. He explained the mechanism through which the currently preferred therapy, UDCA, is thought to operate, and offered his own view as to why studies on the effectiveness of UDCA show conflicting results.

PBC was first described in 1851, and became known as a rapidly-progressing liver disease from which patients died after 3-5 years. In the 1950's PBC began to be diagnosed earlier by using liver function tests, and patient life -

expectancy increased to 10+ years after diagnosis. Finally in the 1960's AMA tests picked up the disease even earlier, up to 30+ years before end-stage disease.

During this period a number of different therapies were tried, including prednisolone, methotrexate, colchicine, azothiaprine and various combination therapies. Although some treatments improved test results, none showed a clear improvement in liver histology, and many had significant negative side effects. UDCA is currently the only treatment which has been shown in extended trials to improve survival by slowing the progression of the disease. It is not a cure for PBC.

Despite at least four major trials indicating improved survival, a recent meta-analysis of all trials to date has suggested that while biochemistry is improved by UDCA, there is no overall effect on long term survival. Dr. Kronborg believes this anomaly can be explained by the fact that some trials were carried out over too short a period, involved small numbers of patients, ceased treatment with UDCA before followup, or included patients with severe end-stage disease.

Because of the long course of this disease, studies extending over many years are required before significant conclusions can be drawn. Also, because liver function tests revert rapidly to pre-treatment levels if UDCA is stopped, it must be taken indefinitely in order to have a beneficial effect. It is also more effective in early-stage than in late-stage disease.

The beneficial effect of UDCA is thought to arise mostly from its displacement of toxic, apolar bile acids retained within the liver cells following destruction of microscopic bile ducts by the aberrant immunological reactions of PBC. These hydrophobic bile acids exacerbate the inflammatory reactions which probably cause the deterioration in liver function. UDCA also stabilises the cell membranes and may modulate abnormal immune responses. Fortunately UDCA has relatively few unwanted side-effects, and in many cases helps with symptoms such as itching.

In summary, while no current treatment cures PBC, Dr. Kronborg believes that UDCA therapy can slow the progression of the disease, particularly if commenced in the early stages. Some 'alternative' additional therapies such as Vitamin E in high (500-1000I.U.) doses may also have a non-specific beneficial effect, while agents such as Co-Enzyme Q10, said to help symptoms such as fatigue, deserve further investigation.

Canadian PBC Conference 2001

The Challenge - Convened by the PBC Patient Support Network.

To be held in Toronto, Canada on the 12th and 13th of October, 2001.

Please contact Jocelyn for further details of the Canadian or American Conferences.

PBC Conference 2001 "Progress through Partnerships"

Wednesday, June 20 - Saturday, June 23, 2001, Flamingo Hilton Hotel & Casino, Las Vegas, Nevada, USA

The PBCers Organization proudly presents the Primary Biliary Cirrhosis Conference 2001 "Progress through Partnerships." The conference is to provide the most up-to-date PBC information on research, treatments, transplantation, the growing PBCers Organization and other autoimmune liver diseases.

The conference is open to all those with PBC, other liver diseases, family members, friends and the medical community.

If you need any information, please visit the website at <http://pbcers.org>, e-mail Linie Moore at pbcers@aol.com or call 1-281-997-1516.

Best Wishes To....

Liz Allan in Victoria

Marlene Borlase in NSW

Doreen Cheong in NSW

Mary Marshall in NSW

Raylene Mounsey in WA

Lionel McCaskill in NSW

Joan Sharp in Victoria

All of whom are being evaluated for transplant, or are on the waiting list.

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