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# The Biliary Bulletin

Volume 1, Issue 4

September 1999

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

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## From Rosemary

### **Summary of the Meeting at St Vincent's Hospital Victoria June 20th 1999**

Guest speakers were :

- ◆ Dr Gideon Shaw who works in the Liver Clinic at St Vincent's.
- ◆ Dr Mark Patrick, Rheumatologist at Box Hill Hospital, lecturer at Monash University, who also has a practice in Dandenong.
- ◆ Alan McLean from the Transplant Promotion Council of Victoria.

**Dr Shaw** spoke about drugs and PBC.

Some points he made were that PBC is an immunological attack on the bile ducts which causes retention of bile in the body, it is not infectious, not diet related and it never occurs in children. It is rarely found under the age of thirty, the youngest known to be 22 and the oldest 93.

Dr Shaw mentioned the symptoms including pruritis, (the cause of which is unknown) the tiredness and lethargy, also that about 50% are asymptomatic. He then spoke about the symptoms of advanced liver disease such as bleeding from enlarged veins in the gullet (varices), and ascites (fluid accumulation) in the abdomen due to low protein levels.

He said that the Alk.Phos. levels rarely rise to over one thousand but when the level of bilirubin rises, this indicates that the disease is progressing. He also spoke about liver biopsies and the reasons for performing them, which are: to diagnose the disease, to identify the stage, or to check the progression.

Treatment of PBC using the oral preparation Ursodeoxycholic Acid was the next topic. Dr Shaws said it was a naturally occurring bile acid and a choleric – promoting bile excretion.

It was first trialed in the 1980's and found to be well tolerated with minimal side effects. There were fewer deaths in treated patients, an improvement in the itch, but no effect on liver histology findings. It may also delay the time to liver transplantation.

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## November Get-Together

Family Picnic Barbecue November 14th at Albert Park, from 11am onwards.

Meet at the Cormorant gazebo. BYO food and drinks and bring all the family for a fun day. This is your opportunity to stock up on our specially printed Christmas cards too. See the enclosed details for map and more information. See you there!

## Raffle Results

The raffle drawn at our June meeting had three happy winners:

- ◆ 1st Prize - Darren Thomas. NEC portable cordless telephone answering machine.
- ◆ 2nd Prize - Marie Abrahams. Glass bowl and jug set.
- ◆ 3rd Prize - Wilma Mitchell. Camera and tapes.

## Paper on PBC

Dr. Mackay is co-author of a paper:

*The Geopidemiology of Primary Biliary Cirrhosis: Contrasts and Comparisons with the Spectrum of Autoimmune Diseases* Arti Parikh-Patel, Ellen Gold, Ian R Mackay and Eric Gershwin

If you would like a copy, please contact Rosemary.

## Member Profile *Debbie Brent*

In October 1994 when I was first diagnosed with PBC I was relieved, rather than afraid. For one thing, it meant that I had an "Answer" to the unbearable itching I was experiencing, for another I was told there was medication available to assist. Most importantly however, was that I was just so happy that it was only PBC and nothing worse. My father had died some years before of pancreatic cancer - first symptom: ITCHING. My mother (in a million to one coincidence) had, only a few months prior to my diagnosis, also been diagnosed with pancreatic cancer - first symptom: ITCHING.

So that's how it started, for me, at the age of 38 and with three young children. I had visited the doctor after several weeks of itching because I had decided that if I did have cancer, I needed to know about it. My doctor told me that she was sure it was just something new in the shampoo I was using, but just to allay my worst fears she would send me for LFT's. No-one was more surprised than she when the test results came back the next day.....by the time she phoned me to come back and see her she had already spoken to a specialist who had confirmed the diagnosis over the phone.

I saw him the following day and was taking Urso (Destolit at the time) that afternoon already. My visit to the gastro was very interesting actually, because he confessed to not knowing a great deal about PBC himself, although he was treating several patients with the disease and seeing 4 new ones per year, he said. Prior to my visit to him, he had ordered an AMA test to absolutely confirm his telephone diagnosis. When I saw him he booked me in for a liver biopsy as well. All in all, he has been caring and careful in his treatment but as he rightly said at my last visit "You and your mates in that group probably still know more about this thing than I do!"

I feel quite confident that I am going to be OK, as my numbers have plateaued over the years and I feel well enough. I hardly itch at all (thanks to the Urso) and my bilirubin is pretty steady around 7 to 9.

My children are a little older now and that also makes me feel better, somehow. I remember one of my first reactions at diagnosis being "Will I see the children grow up?" but now it's more like "I can't wait to hold my grandchildren". I look forward to the future and I am confident of the experts wanting to find a cure. Whether they do or not, it's really largely a matter of money and time, I guess. I worry of course about my two daughters, given that there has been talk in the past of a genetic factor (female to female) but I hold on to the thought that nothing is certain and not enough is known about it to jump to any conclusions. In 1994 it was all gloom and doom and now PBC isn't seen in the same way.

I wish you all well. I think that keeping up your spirits and being good to yourself is important (especially trying to eat the "right" things - although I'm the worst offender!). So - be good to yourself!

Thanks for reading my story.

## Condolences

We were very saddened by the passing of one of our dear friends - June Abley in Tasmania recently. We send our love and deepest sympathy to her husband Sam and all her family.

Instead of flowers at her funeral, donations were made to our group. A beautiful legacy from June. The money has been put aside until some suitably special use can be found for it and suggestions are welcomed.

Wilma Mitchell's husband Jim passed away in August and Ann Clarke's husband passed away back in June.

Our thoughts and sympathy go out to Wilma and Ann.

## Addresses of resources for PBCers

### UK:

The PBC Foundation

The Dean, Longniddry, East Lothian, EH32 0PN, UK

**Web site:** <http://www.nhtech.demon.co.uk/pbc/>

**Email:** [pbc@nhtech.demon.co.uk](mailto:pbc@nhtech.demon.co.uk)

### USA:

American Group

**Web site:** <http://members.aol.com/pbcers/pbcers.htm>

**Email:** [pbcers@aol.com](mailto:pbcers@aol.com)

### Canada:

Patient Support Network

R.R.#5, Hwy. 43

Perth, Ontario, Canada K7H 3C7

**Web site:** <http://www.superaje.com/~pbc/index.htm>

**Email:** [pbc@superaje.com](mailto:pbc@superaje.com)

### Australian PBC Friendship Support Group

**Web site:** <http://www.vicnet.net.au/~ozpbc/>

(including back copies of the Biliary Bulletin)

**Email:** [granga@net2000.com.au](mailto:granga@net2000.com.au), [dordon@net2000.com.au](mailto:dordon@net2000.com.au),

[joc@eastax.com](mailto:joc@eastax.com)

## Christmas Cards

Due to the wonderful response to the Christmas cards we had printed last year, we are again offering them for sale. Profits go towards the cost of providing information and materials for PBCers, and help to keep the annual membership at \$5. (Have you paid yours?)

Please contact Doreen Donaldson for your early order:

Phone: 9706 2404 or email: dordon@net2000.com.au

Snail mail: 18 Throsby Court, Endeavour Hills. Vic. 3802

Cards will also be available at our Get-Together on November 14th.

## Thank You NEC

Thank you to NEC, Princes Highway, Noble Park for their generous donation of the cordless, portable telephone/ answering machine for our June raffle.

## From Rosemary

*continued from page 1*

Ursodeoxycholic acid is thought to be more effective when used in the early stages rather than later when the bilirubin is elevated, but Dr Shaw still thinks it is useful to continue to use urso in these patients. Drug interactions which might affect the absorption of Ursodeoxycholic Acid are Questran, Colestipol, charcoal and certain aluminium-based antacids.

### Some questions put to Dr Shaw were :

*1. When diagnosed is it best to start urso straight away?*

Yes but there is the problem of availability, mainly available through the Special Access Scheme in teaching hospitals. It is not available on PBS listing.

*2. When was PBC first identified as a problem?*

It has been known for about 40-50 years.

*3. Any evidence that PBC causes gynaecological problems?*

No documented evidence, there could be clotting problems in advanced liver disease which may indirectly lead to heavy periods, there are no links to hysterectomy in any literature.

*4. If the bile ducts are damaged how does urso increase bile secretion?*

Urso is more effective in the early stages because there are more bile ducts.

*5. Are there any data regarding the length of each stage?*

Variable, can stay in a stage for 10, 20, 30, 40 years.

*6. Can you stay in stage 4 for a long time?*

Yes but in stage 4 life expectancy decreases, increasing the dosage of urso will not help because there are not enough bile ducts.

A few people commented on liver pain and they thought that doctors did not believe it was associated with PBC. Dr Shaw said that while he was not denying it, it was not listed as a common symptom and that the cause of the pain should be identified, as it may be gall stones.

Dr Shaw concluded by saying that the majority of PBC sufferers would die from other causes.

**Dr Patrick** talked about Osteoporosis and other autoimmune conditions associated with PBC, such as rheumatologic and connective tissue disorders including: Sjogren's Syndrome, Rheumatoid Arthritis, Scleroderma and Raynaud's Phenomenon. He said that rheumatologists get involved because of the malabsorption of vitamin D which can lead to abnormal bone fractures in Osteomalacia and Osteoporosis.

He also described the tests for diagnosing Sjogren's Syndrome - the Rose Bengal Stain Test to determine the degree of corneal scratching and the Schirmer Test, where strips of filter paper are placed in the lower lid, to measure tear production.

The inflammation in Sjogren's Syndrome is mainly in the salivary and lachrymal glands. Artificial tears can help to lubricate the dry eyes and artificial salivas may provide relief for dry mouth.

Scleroderma and Raynaud's Phenomena were discussed quite widely. Scleroderma (systemic sclerosis) is a systemic connective tissue disease. Limited Scleroderma usually causes Raynaud's Phenomena and hardening of the skin in the hands, there may be some changes to facial skin and thickening of the skin on the forearm and lower leg.

Diffuse Scleroderma also affects the skin on the trunk, upper arms and thighs. Patients with this condition often have a more systemic illness with the scleroderma process affecting other organs and tissues.

Dr Patrick described Raynaud's as blanching of the fingers and hands changing from white to blue caused by vascular spasm which causes the blood vessels in the fingers and toes to constrict. CREST is the acronym given for the clinical combination of Calcinosis, Raynaud's Phenomena, Esophageal problems, Sclerodactyly (stiff fingers) and Telangiectasia (small dilated red blood vessels in the skin of the hands and face).

### Some of the questions Dr Patrick answered were:

*1. Can you get just dry eyes with Sjogren's?*

Usually you get both, generally involves salivary and lachrymal glands.

*2. Can ulceration in the mouth be associated with dry mouth of Sjogren's?*

It's more likely to be a gingival problem which can be related to gum hygiene.

*3. What is the difference between Osteoporosis, Rheumatoid Arthritis and Osteoarthritis?*

Osteoporosis is silent, you may not know you have it until there is a fracture. Rheumatoid Arthritis is an inflammatory arthritis. Osteoarthritis is the wearing out of cartilage.

*4. Is Osteoporosis hereditary?*

May be vitamin D deficiency, may be genetic, a lot is acquired, there is a greater risk if mother has it.

Dr Patrick concluded by saying he was not aware that caffeine is a major factor in Osteoporosis but some preventions are - regular weight bearing exercise, no smoking or alcohol and increased calcium intake.

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**Alan McLean** talked next about the importance of organ donation being discussed in the family, as this saves much heartache later if a situation arises.

Donors can donate one to six organs. The donation of a person's heart, lungs, liver, kidneys and pancreas can only take place if the person is declared brain dead, in intensive care and on a ventilator. However certain tissue including heart valves, corneas, bone and skin tissue can be taken from donors who have died because their heart has stopped and their blood is no longer circulating around the body.

At the conclusion of his talk Alan left folders and leaflets including registration forms which were quickly taken up by members of the group to distribute to many outlets.

## To our PBC Friends in Australia

*Letter from Collette Thain of The PBC Foundation*

I was delighted to hear that you are now coming together and forming your own Friendship Society. I was delighted also to hear that you were celebrating International PBC Day with lunch at the Park Royal Hotel in Melbourne. This was mentioned the evening before at our Annual Ball and received much applause. People were touched and moved by the fact that Australia and UK are coming together and sharing information and support and help with PBC.

My husband received a telephone call at his office after I had been very ill for some months and the telephone call revealed that I had Primary Biliary Cirrhosis. To demonstrate how ignorant my husband and I were about PBC he endeavoured to reassure me by saying well at least it's not in the secondary stages and is not yet Secondary Biliary Cirrhosis! We were thrown completely into a flat spin and we had no information at hand, you all know what I am talking about.

You just do not know what you have to come to terms with. It took many, many months before I did find some information albeit it was very paltry and what I had I simply just had to share. We put a small advert in the newspaper, a Scottish newspaper, the Sunday Post and we were bombarded with hundreds and hundreds of telephone calls. All people in the same position as myself who were very worried and desperate for information.

The doctors and consultants in the UK as you know Professor Oliver James, Professor James Neuberger and Dr.Niall

Finlayson all sympathised and empathised and were very keen to help supply the information we so desperately needed. Before we knew it within six months we were an ongoing and ever growing organisation. Like Topsy we just grew and grew!!

We became a nationwide and international charity in our own right almost three years ago and since then we have gone from strength to strength and I do believe that one of our major strengths is the fact that we all have PBC. We are all volunteers, we can do as much or as little depending on how we are at the time but because we have the condition we can give so much understanding to those in need.

We spend a large part of our time concentrating on those newly diagnosed because I will never forget the fear, loneliness and isolation I experienced myself. As you know you learn as time goes on to accept your condition and with the receipt of knowledge and information then you cease to become frightened of PBC and realise it is a condition that you can learn to live with.

I have met such wonderful people from all over the UK and indeed Australia. I receive such wonderful letters. I really do believe that a prerequisite to PBC is that you have to be such a lovely person.

This newsletter has been kindly sponsored by

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