

# The Biliary Bulletin

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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 communicates news and information to members of the Australian PBC Support Group and other sufferers of PBC.

## From Rosemary

Dear Friends,

Hope you are all making the most of the cooler weather. I love to walk and find it much more comfortable and enjoyable in Winter. I try to walk at least three or four times a week and much prefer to walk early in the morning. I feel so much better for it, more relaxed, refreshed and able to cope better with the day ahead.

The pouring rain did not stop some of us here meeting at Chadstone for lunch recently, it was good to catch up with everyone especially new members and of course Liz, looking so good only four weeks after her transplant.

Good news from the ladies in South Australia, their small group of five or six who meet for coffee and a chat have managed to arrange for a doctor to talk to them. Well done ladies, I hope that this will inspire other members interstate to get more involved with doctors. There are many doctors willing to help, you just have to reach out and ask. The ladies in South Australia are on the way to a better understanding of the condition they have.

A coffee morning was held recently for NSW pbcers at Chantilly's, Macquarie Centre. Thank you to Suzanne and mum Marlene for organising the get together.

They felt it was good to have a chance to talk generally about how they were all feeling and what they had in common, and it was decided that another get together will be held in about six weeks time. They will alternate between weekends and weekdays to cater for those who cannot get there during the week.

Well done Suzanne and Marlene!!

We were saddened to be told recently of the passing of Mary Baker in South Australia. Mary loved her quiet life in Sellicks Beach and said that she felt she had many friends because of our group. Our sincere condolences to Mary's family, she will be sadly missed.

My thoughts are with all those who are being evaluated or are on the transplant list. For those of us here in Victoria who have met Maria and Liz since they were given their gift of life, it is so inspiring for us to see how well they are doing. For me, being able to visit Maria and Liz in hospital so soon after transplant has been a learning experience which I know has helped to make me stronger.

I also believe as many do, that education about the condition you have takes away some of the fear.

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I would very much like to thank all members who send in donations to our group, your generosity and kindness is very much appreciated. A special thank you to:

Mary McInerney (NSW) Margaret McClunie (Vic) Neta Cooke (SA) Mary Walker (WA) Dorothy Furmage (Tas) K.Stewart (Vic) Daisy Bucklar (Cairns) Grace Chung (Malaysia) and Yolande Egyed (QLD)

We recently became the owners of a brand new photocopier, thank you to the Victorian Government Department of Human Services. Our application for a grant for self-help groups was successful, much to my surprise, and is accepted with appreciation.

Our brochures are being given out all around Australia, by doctors in hospitals, in medical centres and womens' health centres. Please continue to pass on our information by giving out the brochures included with this copy of the Biliary Bulletin.

Keep on keeping positive everyone, and best wishes to all.

Rosemary.

## Post Transplant Support Group

Last week Maria invited Liz and myself for lunch. As some of you will remember, Maria was transplanted 16 months ago, and Liz had her transplant op just 7 weeks ago. They seem to have appointed themselves my "guides" on this journey through the anxieties of the waiting list and all that lies beyond.

I am very grateful to have the support of those who have gone before. Just to be able to see how well Maria and Liz are and how normal their lives are becoming again, offers enormous encouragement and inspiration to me.

I have dubbed them the "Post Transplant Support Group" and hope to become an official member in the not too distant future.

It is amazing how our group has grown over the last few years. We now receive calls from all over Australia by people with PBC or their family members, wanting more information. We also receive many requests for our patient information brochures from clinics and hospitals, and rejoice in the knowledge that we are able to help to inform and educate others about PBC.

In the meantime, the three of us had a lovely lunch. There was good food, much hilarity and swapping of stories. There was also lots of talk about the Austin Hospital and the people who are so dedicated to the process of taking patients through the complex and difficult task of being evaluated and prepared, with such skill, openness, good humour and grace.

We all agreed that they are angels!

Jocelyn.

## Member Profile

*Evelyn Shaw*

Hi Rosemary, just thought I would drop you a line in response to your request for readers' stories. I have had PBC since August last year, and started taking ursol at the end of January this year. Every month after starting my Ursol my readings are coming down and I am flying high!

One of my first readings before starting the medication was 450! At the 1st test it was down to 140, then 120, 136. At this stage I was starting to lose weight. I asked the doctor about it and was told just to keep an eye on it. Then the reading went down to 68! Boy am I ever flying, I have lost on average a kilo a week! 13 kilos in 13 weeks and I'm up there with the clouds! Hahahaha.

I am 51 and I look and feel like I'm 25! I don't know if it's the medication or the time I spend on the net talking for 17 hours a day, or the weight loss! Whatever it is, I love it and there's no way I'm giving any or all of it up! Lol! My whole life has had one huge turn around!

Maybe this might help brighten up someone else's day. I only hope it does! The friends that I have made on the net only give me positive vibes which in turn just make me so happy! I am a completely different person since all this has started to fall into place! I hope this letter will inspire some of my fellow sufferers to have a think about the weight side of things or even think about getting on the net!

I have also been blessed by not having symptoms! I don't know how I'd cope if I did! My positive attitude to life and all that it offers me is what is keeping me free of them!

I hope this is a good enough reason for you to publish my letter and that somewhere out there somebody else can relate to me and my situation. Keep up the very excellent work for us.

Evelyn Shaw.

## Primary Biliary Cirrhosis, Mitochondria, Fatigue and Coenzyme Q10

*Dr. Ian McKay*

*The anti-mitochondrial antibody (AMA).*

Patients with primary biliary cirrhosis (PBC) will know about mitochondria because their blood serum contains an abnormal antibody protein, an autoantibody, that reacts with cellular mitochondria in laboratory tests.

In fact, this anti-mitochondrial antibody (AMA) reaction is one of the most important laboratory diagnostic criteria for PBC. But whether AMA is implicated in causing the disease is still undecided.

Researchers on PBC are faced with other puzzling questions as well, including how this autoantibody arises in the first place and given that mitochondria are present in all cells of the body, why is the pathology of PBC so sharply focussed on the small biliary ductules of the liver?

Well, what are mitochondria? They are discrete packages (organelles) mostly consisting of enzymes, in the cytoplasm of cells. There are about 700 mitochondria in almost every cell of the body.

It appears that far back in evolution, single cell organisms "pirated" bacteria and incorporated these into their own cellular structure to enhance their repertoire of enzymes and metabolic functions. As evidence for this, there is the fact that mitochondria, unlike any other organelle of the cell except the nucleus, contain their own "dedicated" DNA. In any event, single cell organisms, such as bakers' yeast, possess mitochondria with functional properties very like those of mammalian cells.

What do mitochondria do? The essential function of the mitochondrion is to provide cellular energy by enzymatic activities: it is the "powerhouse" of the cell.

Abbreviating the technical details the major energy-producing activity; called oxidative phosphorylation, depends on the co-ordinated and synchronized function of five subunit enzymes, complexes I, II, III, IV and V. These complexes constitute the "energy transport chain" of which the energy-rich end-product is adenosine triphosphate (ATP).

### *Fatigue and Coenzyme Q10*

The next part of the story concerns one of the troubling (and so far unexplained) symptoms of patients with PBC, which is fatigue.

Things would fall into place very nicely if the target antigen for the characteristic anti-mitochondrial antibodies in PBC were to be the particular enzyme that makes ATP which in fact was once suspected; but unfortunately for theory, this proved not to be the case.

Studies in my laboratory at the Hall Institute in 1985 led to the recognition that the target autoantigen was another

quite different multifunctional mitochondrial enzyme, pyruvate dehydrogenase complex. Nonetheless, there is some interest in tackling the fatigue problem in PBC by supplementing a key enzyme, complex I of the energy transport chain, with one of its constituent substances that is called Coenzyme Q10 or ubiquinone.

Q10 is a manufactured product identical to the natural counterpart and it is readily available (although not cheaply) in health food shops.

There have been some possibly extravagant therapeutic claims made for coenzyme Q10 including a restorative activity for declining energy levels in old age.

Also, since coenzyme Q10 is a non-prescription "natural" drug, it has not gone through the same degree of scrutiny and evaluative procedures that are applied to the usual medicinal drugs, and optimal doses and duration of treatment are uncertain. On the other hand, there are no known adverse effects. So, for a patient with PBC who is suffering from distressing fatigue, there would be no harm done (except to the purse!) by taking Q10.

A dose of 4-6 50 mg capsules daily for three months could be a suggested maximum. Whether it is deemed to work or not might have to depend on the opinion of the legendary "50,000 Frenchmen"! (with apologies to Gilbert and Sullivan).

## From the Editor

Well the PBCers Organisation Conference 2001 is over, the committee is already planning the next one for 2002. Please contact Rosemary or myself for any information.

We were very lucky to have had my internet friend Jaclyn Leuenhagen there to collect literature and an order form for tapes of the speakers. Thank you Jaclyn for sending us a 'showbag' chock-full of conference goodies!

Whilst reading the on line digest of the PBCers Organisation recently I came across this letter which I found rather lovely. It was written by Beverley Wiskow who has kindly allowed me to reprint it here...

"The response to PBCer Pete's question about liver spots in today's Digest got me to thinking...

Pete was wondering if the little dark spots on his hands were caused by or aggravated by our PBC. This is a matter about which I've done much research and I'll share this information with all of you. Though my parents and our neighbors who are in their seventies, and now, suddenly, almost all of my friends who are 50 something have liver spots on their hands, I'm convinced YOURS and MINE are caused by PBC!

In fact, sometimes it's a great comfort to know that every problem, issue, concern, and physical glitch which comes our way IS indeed because of our PBC.

And you are aware, I'm sure, that the time of our lives known as Middle Age has been changed from 35 to 45 to the more accurate 65 years to 75 years old (at least it has been officially changed in California!). So, hey, since I've not yet entered middle age my liver spots and all of the other changes in my life simply must be the result of PBC! There is no other logical scientific explanation.

For example, I've gained more weight in the last year than I care to discuss and now weigh more than I have since I was pregnant a thousand years ago. It's the PBC. The fact that I've had PBC for nearly 16 years is irrelevant. In fact, it is a well known by all of us in this group that weight gain caused by PBC begins about 14-16 years from onset.

I don't care what the doctors say; PBCers know more about our bodies than any researchers. The texture and thickness of my hair have changed - and not for the better. Yep, you're right: PBC. There is now a dusty hue to my otherwise dark hair color as I find the PBC is causing me to grey prematurely. No one knows exactly why.

Another matter I've researched is the quality of eyesight. PBC definitely causes us to require glasses for reading once we've had it for about 10 years and especially so if we are PBCers over age 50. Couldn't be anything but the PBC! Right?

The worst effect of PBC for me personally is that I have to get more sleep in order to function. Time was when I could go for 18 hours a day, sleep for six, bring home the bacon, fry it up in the pan, parent two teenagers, cook, clean, do laundry, volunteer in community organizations, paint, sew, take care of horses, be active in my church, and never feel tired. If it weren't for this darned PBC I'm sure I could still keep up that pace. The fact that I just turned 57 years old yesterday has absolutely nothing to do with it.

Also, it's an unfortunate certainty that we PBCers should not have babies after the age of 55. People with messed up livers like ours just can't handle the demands and stresses of a tiny baby 24 hours a day. On the other hand, having grandchildren at age 50 to 80 is a highly recommended therapy for PBCers! Why, we can get up in the middle of the night to change a newborn granddaughter's diaper without even exerting ourselves. Show me a grandmother without PBC who can do that one with such ease!

On the down side, there is a direct negative correlation between our PBC and some of life's daily challenges: having to drive in rush hour traffic, waiting in line at the express grocery checkout behind someone who has more than 10 items. I find being put on hold when I make a telephone call to the DMV and having to listen to Yanni has a very detrimental effect on my LFT's. Being on a rigid schedule of things I don't really want to do can push my PBC into the next stage in a heartbeat.

Another thing that is very bad for us PBCers is to be around people who have forgotten how to laugh. I

recommend you avoid that at all costs -- it'll make your liver spots turn a very dark brown!

Though not a great deal is known about PBC, the research proves that it is absolutely vital for PBCers to walk on a sunny beach, preferably in Hawaii, Tahiti, Jamaica, or Bermuda, at least one week annually, being careful to get lots of sand between our toes. And our Alk Phos and GGTP numbers are dramatically and mysteriously lowered when we watch a glorious sunset with someone we love.

Remembering a far away friend's birthday with a phone call while sipping on a cup of tea with honey and that indulgent teaspoon of half-and-half is highly recommended, but only for those of us who are in stages one through four of PBC. Massaging the hips of my thirteen year old black Lab so she can climb down off the porch and hunt for very slow moving rabbits on a spring morning is particularly good therapy for me.

The earlier our PBC is diagnosed the better because it gives us so many more years of opportunities to smell the roses, hug our children, reach out to someone who is hurting, express our gratitude for our abundant blessings, make more friends, grow more gardens, listen to more birds and crickets and critters on so many more spring evenings, accept more collect phone calls from our teenagers who are safely on the side of the road with a flat tire on their way home from a ball game in a distant town, go to Bob Dylan concerts, give haircuts to our loved ones in the back yard on a warm fall evening, teach our grandchildren how to make a kite from the Sunday funnies and fly it with a tail made of old socks.

When I have another of those uneventful checkups with my doctor and he says, "OK, see you in six months" because all my test results are totally boring.....well then, I am fully justified in treating myself to a large chocolate Blizzard from Dairy Queen!

Some of my symptoms, which I hope you'll experience as your PBC progresses include: a heightened sense of what is really important and what I can let go of with ease. And the infrequent days when I have more energy than I know what to do with, those are especially sweet. And celebrating the 10th, 11th, 15th, 16th anniversary of my PBC diagnosis with that once-a-year super-sized frozen Marguerita with salt all around the rim!

I find I almost always say "yes" to things I might otherwise have missed: a spontaneous picnic, stopping to pick flowers from the roadside, stepping on the brake and slowly, silently backing up the car so I can watch a red-winged blackbird on an old wooden gate, making a big bowl of popcorn to share while being trusted enough to watch a friend's old home movies, picking up a feather that finds me while I'm out walking.

My PBC is a reminder of the importance of saying how I really feel, speaking up for what I believe in, speaking out against what I know to be unkind, opening up to everyone

I meet, taking a chance when expressing an unpopular opinion, listening closely to the young people who bless my life with their trust, sewing a blue satin bridesmaid's dress for a friend who needs it in two days, taking a full half-hour to burp my granddaughter (even though she really burped four minutes into the process).

My mother is 84 years old and lives 3000 miles away in Florida. If I didn't have PBC, I wonder if I would be so conscious of the preciousness of every conversation we share, every letter I receive, every sweet gesture with which she graces my life. I adore being her little girl.

So Pete, about your question regarding liver spots. I embrace mine. They are my friends. They keep me alert to the preciousness of each moment. They whisper to me: "Go ahead. Yes! Do it! Why not? You deserve every good thing that comes your way."

Recently I saw an ad for a skin cream that was guaranteed to fade liver spots on your hands and arms. The before and after pictures showed the amazing results. It sells for about \$35 an ounce. It is available in major department and drug stores everywhere. Do you know how many long-distance phone calls can be enjoyed with \$35? Why I could buy 103 stamps to stick on letters to friends with \$35! For \$35 I could buy three blue hydrangeas and some marigold seed to plant along our front sidewalk.

So, as I am typing this I'm noticing my liver spots. It is because of this danged PBC that I have them long before I'm really old enough! Thanks for asking about them in the Digest - it reminded me to be grateful for their beauty and value. I wonder if yours are as pretty as mine!

Beverley."

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