

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,

First I would like to send my good wishes to those people I have spoken to recently who are not doing very well right now. To all who are going through really difficult times, my thoughts are with you.

As expected I get many telephone calls from people suffering all sorts of symptoms, some related to PBC, some not and others, well we just don't know of course.

It's good that we can be in contact with others to see if anyone else shares a particular symptom or problem we may have, this is the one of the good things about belonging to a self help group such as ours, but many of the questions put to me can only be and should be put to your doctor.

It's a good idea to write down your questions before your doctor's appointment, and if you do not understand the answers given to you, ask your doctor to explain in terms that you can understand.

I am amazed to find when talking to people that they still do not ask for copies of their Liver Function Test results and or do not understand them. I could not rest not knowing my test results, even though sometimes it can be an anxious few days waiting to get them. I make sure I have a copy so that I can follow the results myself.

For all those who wish to have contact with other members, you will have received a new contact list with your BB this time.

All of the people on this list have given permission for their details to be given out for contact so please take time to see if there is anyone near to you and think about getting in touch, as many of us already know it makes such a difference.

It is coming around to International PBC Day once again (May 30th), a day when we feel particularly close to other people around the world who suffer from PBC. How lucky we are to have access to wonderful groups online such as the PBCers Organization, where we can learn so much from others coping with PBC.

How lucky we are to be able to gain all the information available so that we can educate ourselves about this disease, and indeed how fortunate that there are good doctors willing to give their time to help us understand and who are working so hard to find the cause and cure.

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I would like to take this opportunity to thank Debbie Brent and Liz Allan for getting the meeting at the Austin hospital together on March 3rd, and to thank our guest speakers, Dr Paul Gow and physiotherapist Jacqui Luke for giving their valuable time.

Material for the Biliary Bulletin from group members is still needed, Jocelyn needs all the help she can get right now. I know from talking to so many people that there are stories to tell so please put pen to paper or easier still if you have a computer get your fingers moving and write in.

If you have a problem putting a story together (this we are often told) don't worry, just do your best and it will be edited for you, so please give us a little of your time, send in your stories or letters to Jocelyn. Your input would be appreciated.

Keep on keeping positive everyone, best wishes to all.

Rosemary.

Letters

Dear Rosemary,

The newsletter has been invaluable as I am not quite 'au fait' with internet and email. The article on overlap syndrome resulted in diagnosis of overlap and a change in medication. This in turn has resulted in a reduction in LFT results (for the first time). I have also begun Co-enzyme Q10.

I also agree with you on the benefits of walking. The more energy I use, the more I seem to have. I find that if I get up and walk when I want to doze off during the afternoon I am much better and I just love my first thing in the morning long walk. Thanks again to Jocelyn too.

Love,

Carolyn Warner

Member Profile

Tracy Brett

After receiving the Biliary Bulletin today I felt I should make an effort to contribute. You all do such a wonderful job and I'm sure it does take a huge effort to keep the support network running.

I like many others truly appreciate your efforts and find much comfort in reading the bulletin articles and all the emails. So on that note I thought I could share my profile with the group and feel that I am at least making a small contribution to the group.

Hi everyone! My name is Tracy, I am 36 years old and live in a semi rural area of Sydney called Glossodia. I have a wonderful husband and two beautiful children, aged 6 and 8. I was diagnosed with PBC in July 2000.

I had no symptoms only constant itchy skin. After enduring this for about a year the doctor did blood tests including LFT's. This led to a Liver biopsy which led to the result of PBC.

When I was told this news I felt my life had fallen apart, why, how, will I die?. After my first appointment with my doctor at RPA hospital I put things in perspective and I have not looked back.

I started on Urso straight away and within 6 months my LFT's and most other results were moving back down within normal range. I eat sensibly, exercise, don't drink or smoke any more and feel well overall. Tiredness is sometimes an issue but I put that down to leading a most enjoyable busy life with young children.

As I'm writing this I'm too aware that I have LFT's to be done next week, time flies between 6 monthly visits and I try to contain my worrying to these appointment time frames only...

I believe I am one of the younger sufferers and I am really hopeful that since the Urso was started in early stages I will live a healthy life and be able to do all I want to do with my children as they grow and journey through their lives.

I must say, which I'm sure others feel as well, being dealt this blow is like a jolt in life to say hey - life's good - get on with it !.

To Rosemary and her team who keep the support network running for all us PBCers, thank you !.

You need to know how much your efforts are appreciated and how comforting it is to have contact with other sufferers and have access to updated information on this disease which we are all still learning about, including the medical profession.

With love and good health to all,

Tracy xxx

PBC and Pruritus

Clinical Nurse Practitioner

Margot Miller,

As anyone who has PBC will know, pruritus, or itching, can be one of its most distressing symptoms. Unlike other itches, it is not relieved by scratching and can affect people in a variety of ways and with different severity. Some may experience only a mild intermittent irritation, while others are quite literally driven mad and will resort to all sorts of measures in an attempt to alleviate their suffering. Unfortunately, these measures often only make matters worse by causing wounds and infections to the skin.

What causes Pruritus? Well, if we had the answer to that we would be able to treat it a lot better than we can at present. The simple answer is that we don't really know for sure. Bile acids being deposited in the skin has always been the most popular theory, but some research has shown that the amount of bile acids in the skin does not necessarily correlate with the degree of itching felt. It may be that this is only part of the answer and research is being done on the role of the nervous system in the origin of pruritus.

What treatments are available for pruritus? They can be divided into two areas:

1) Medical Therapies and Interventions.

Cholestyramine, or Questran, is the most widely used medicine at present. This medicine works on the principle that the bile acids in the skin are responsible for pruritus. It prevents accumulation of bile acids in the body by combining with them in the bowel and preventing them from being reabsorbed. Unfortunately, it is not the most pleasant medicine to take as it is a bit like drinking sand in water and can cause changes in your bowel habit...usually constipation!

It can be made a bit more palatable by mixing it in fruit juice, and there is also a "light" version which you may find easier to take. Research has shown that Questran works best if you take a dose immediately before breakfast and then another immediately after. Your third (and fourth if you take four) can be taken later in the day.

Antihistamines are of questionable efficiency in pruritus, and many have the added drawback that they make you sleepy. Not a good idea if you drive, or operate machinery for a living! If the itch keeps you awake at night, then this may be the time for antihistamines if for no other reason than to help you sleep.

Opiate antagonists such as Naltrexone, are drugs that follow the theory of the nervous system being responsible for pruritus. It has been noted that patients who have been given morphine for medical reasons can develop associated itch. We produce our own morphine-like substances in our nervous systems, which some scientists

believe may be partly responsible for the itch of PBC. These drugs attempt to “switch off” the itch by blocking the receptor sites. However, like Questran, not everyone finds relief with these drugs and, as with the bile acids, the level of these opiate substances in the blood does not appear to correlate with the degree of itch experienced.

Ursodeoxycholic acid (Urso) is probably the most commonly-used drug in PBC. It is primarily given to delay disease progression, but some patients have reported an improvement in their itching. On the other hand it can, unfortunately, cause itch in some people. Urso is basically a bile acid and patients often ask why giving another bile acid should improve the situation. I like to think of Urso as a friendly bile acid which helps to protect your cell walls from the destructive actions of the less friendly bile acids.

Rifampicin is an antibiotic, which has shown in trials that it is fairly effective in treating pruritus. The exact mechanism for this is unclear, but it may be associated with the action it has on liver enzymes. Rifampicin is known in some cases to be toxic to the liver, and it would only be used under specialist guidance with extra checks on liver function. In addition, it can turn your urine red.

These are some of the drugs currently in use for treating pruritus and, as you have probably gathered, they all have limited efficiency. If these have been unsuccessful, then a specialist may recommend other more invasive treatments depending on the degree of distress that the patient is suffering. These include:

? Plasmapheresis

This is like a cleaning process of your blood. Your blood cells are separated from your plasma, and new plasma is added, presumably removing agents from your blood that are causing itch.

? External Biliary Diversion

A drain is placed through your abdominal wall into a bile duct, and bile is drained into a bag.

? Liver Transplantation

2) Non-Medical Interventions

These, basically, are the practical measures that you can initiate to minimise your itching. In some cases they may be enough to help and medications are not required, but usually they will be in addition to medical therapy.

Whatever the cause of PBC-related pruritus, your skin bears the brunt of it. During my career, I have heard horror stories of the extremes a patient will go to for a second's worth of relief and I have seen many damaged and mutilated skins that reflect the degree of distress felt. Obviously, open and infected sores and wounds only add to the anguish. It is of the utmost importance that you look after and protect your skin as much as possible.

Avoid extremes of heat

- ? In hot weather, wear loose-fitting natural fibres.
- ? Be very careful in the sun. Although ultraviolet rays can help itching, sunburn will not.
- ? Avoid very hot baths and showers.
- ? Avoid using electric blankets.
- ? Keep your bedroom well-ventilated.
- ? Avoid nylon sheets.
- ? Don't sit too close to fires and radiators.

Care for your skin

- ? Avoid using soaps that dry your skin.
- ? Use emollients in the bath or shower. These are preparations that soothe, smooth and hydrate your skin.
- ? Avoid heavily-perfumed lotions etc.
- ? Use only preparations to which you are not sensitive.
- ? Keep your nails short and well-filed.
- ? If you scratch at night, wear cotton gloves.
- ? If scratches become infected, consult your doctor immediately.

Look after your general health

- ? Eat a good healthy diet. You need vitamins and minerals for a good healthy skin.
- ? Try to find ways to relax and reduce stress. Stress makes us more aware of symptoms like itching.
- ? Try to exercise regularly.
- ? If necessary, try to lose some weight. Heavier people tend to sweat more, which can irritate your skin.

And finally, two old wives/nurses tales

As far as I know, there is no scientific evidence to back up the following tips, but I have recommended them over the years with varying levels of success. Certainly no-one has come to any harm by trying them.

- ? Firstly, following a low-fat diet has helped a few patients with pruritus. It has the added benefit of helping you to lose weight if you need to.
- ? Secondly, run a bath, not too hot. Mix in a half to a whole tub (50-100 grams) of bicarbonate of soda (not baking powder or you might “rise”) and soak in it for a while. This is especially helpful before going to bed.

This is an edited version of an article from Issue No. 17 of The Bear Facts Magazine, reproduced with kind permission of the PBC Foundation.

From the Editor

I found the article about PBC and itching to be one of the most practical I have read. For those of you who have a problem with itching, do try the low fat diet and see if it helps – it did for me. I also found the bi-carb bath was very soothing even if it didn't always stop the itching.

Another thing which also seems to help me is 'Natures Remedy' cream – available from the first aid section at Safeway. Sometimes a combination of things will work and I know that many of you are sometimes desperate enough to try the hairbrush scratch and feet in a bucket of cold water methods!

For those of you who have difficulty taking Questran remember that it is best shaken not stirred (like a James Bond martini)! Most chemists sell plastic shaker cups for children's vitamin drinks etc. and sometimes you can get a Milo cup with large tins of Milo.

I would like to say a huge Thank You to all my kind PBC friends, locally and overseas who send me messages and cards to let me know they are thinking of me as I notch up 9 months on the transplant list. It lifts my spirits each time I get a message of support from you. Joan Sharp has been waiting even longer and I gain great strength from her being there with me.

Jocelyn.

Stages of PBC Disease

These terms are used by pathologists to describe the results of liver biopsy examinations.

? Stage 1 - Portal Stage

Normal sized triads; portal inflammation, subtle duct damage

? Stage 2 - Periportal Stage

Enlarged triads; periportal fibrosis and/or inflammation

? Stage 3 - Septal Stage

Active and/or passive fibrous septae

? Stage 4 - Biliary Cirrhosis

Nodules present; garland or jigsaw pattern

Although 4 typical stages of evolution have been defined, the disease initially is focal with considerable "overlap" between stages in any one case.

First is inflammation of the medium-sized bile ducts and chronic inflammation of the portal tracts. Granulomas may be found. With progression of PBC, the portal tracts become distorted, inflammation spreads into the parenchyma, bile ducts proliferate intensely, and periportal fibrosis develops.

Progressive scarring continues with less bile duct proliferation and less inflammation. Fibrous bands link the portal tracts, and zone 1 cholestasis and Mallory hyaline

can become evident. The end product is a firm, regular, intensely bile-stained cirrhosis, difficult to distinguish from other cirrhotic processes in the absence of granulomas and the pathognomonic bile duct lesions.

Laboratory Findings

Early findings feature cholestasis with alkaline phosphatase elevated disproportionately greater than serum bilirubin and aminotransferases. In fact, the serum bilirubin is often normal early in the course of the disease. Serum bile acid concentration and gamma-glutamyl transpeptidase activity are elevated. Serum cholesterol concentration and total lipids usually are increased. Serum lipoproteins are increased, mainly because lipoprotein-X is present.

Serum albumin is normal early in the course of the disease, but the globulins usually increase the serum IgM often to very high values. Antibodies against a component of the inner membrane of mitochondria (in 85 to 95% of patients) are important diagnostically, but they can also be found in some patients with HBsAg-negative chronic active hepatitis, making this differentiation difficult.

Prognosis

The course of PBC varies greatly. It may not diminish the quality or the duration of life. Of patients who present without symptoms, 50% show evidence of liver disease over the ensuing 15 years. Slow progression suggests prolonged survival. A rising serum bilirubin, associated with autoimmune disorders, and advanced histologic changes indicate a poor prognosis. PBC is one of the best indications for liver transplantation.

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