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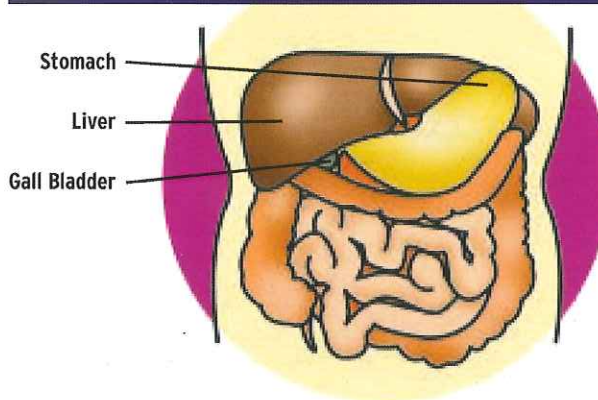
Primary Biliary Cholangitis

Patient Information Booklet

The Liver

The liver is the body's chemical factory and performs hundreds of complex functions, which are vital for life. Amongst other things, it:

- converts food into chemicals necessary for life and growth.
- produces quick energy when it is needed.
- manufactures new body proteins.
- prevents shortages in body fuel by storing sugars, vitamins and minerals.
- aids the digestive process by producing bile.
- helps digest fat.
- neutralises and destroys poisons.
- controls the production of cholesterol.
- maintains hormone balance.
- stores iron throughout life.
- helps the body resist infection by producing immune factors.
- regenerates its own tissue when healthy.



The liver is a very uncomplaining organ and has the capacity to continue its many functions with only a small portion in working order.

Since its discovery in 1851, names for what we now know as PBC have included "Primary Biliary Cirrhosis", "Xanthomatous Biliary Cirrhosis" and even "Chronic Destructive non-Suppurative Cholangitis". As early as 1959, it was known that Primary Biliary Cirrhosis was not a medically accurate reflection of the disease. In order to address this, in June 2015, it was agreed internationally that PBC will henceforth be known as Primary Biliary Cholangitis.

What Is Primary Biliary Cholangitis?

Primary Biliary Cholangitis (PBC) is a slow, chronic liver disease which causes progressive destruction of the small bile ducts within the liver. The bile ducts are the channels through which bile flows. The body attacks the cells lining the bile ducts within the liver as if they are foreign to the body itself, i.e., attacking its own cells. This damage causes poor drainage of bile acids, which leak outwards and damage the normal liver cells. This causes inflammation and scarring which may, after many years, become extensive. This widespread damage and scarring may result in cirrhosis.

Many people who have PBC never develop cirrhosis.

Cirrhosis is often assumed to mean damage to the liver caused by excessive alcohol consumption. This is not the case – cirrhosis simply means an advanced form of scarring, which changes the structure of the liver cells and can be the result of a wide range of liver and biliary diseases. Cirrhosis, whatever the cause, leads to a number of potential problems, requiring long-term monitoring.

What Causes Primary Biliary Cholangitis?

The cause of PBC is at present unknown, although it is thought to be an autoimmune disease whereby the body's immune system malfunctions and turns on itself.

PBC predominantly affects females at a ratio of approximately nine women to every one man, although PBC in men behaves in an identical way to that in women. PBC is more common in families where one member is known to be affected. There is a small number of recorded family cases – often a mother and daughter – and probably about one patient in thirty to seventy has a close relative with the disease.

Laboratory research has indicated that people with PBC display several abnormalities of the immune system, which may be an important factor in determining the cause. What initiates the bile duct damage is unknown. The trigger could be an infection, or a toxin. It cannot be spread by sexual intercourse or bodily contact. Generally, it presents in women aged between thirty and fifty-five, although it may be diagnosed at any age in adults and is rarely, if ever, seen in children.

How Common Is PBC?

It is thought that PBC is most common in Northern Europe and North America, although it is found in people all over the world. In high prevalence areas, it has been shown that as many as one woman in a thousand over the age of forty may have PBC although in many cases the illness may be silent (asymptomatic) and will never become serious.

What Are The Symptoms Of PBC?

These will vary between people. The severity of the symptoms often bears no correlation with the severity of liver damage.

The common symptoms associated with PBC include:

- itching ("pruritus")
- lethargy and chronic fatigue

Because of related conditions, some people with PBC may have:

- dry eyes and/or dry mouth (sicca syndrome)
- joint pains
- diarrhoea
- over or under-active thyroid

In later stages of the condition, people with PBC may develop:

- pale stools and dark urine
- bruising
- fluid accumulation in the abdomen (ascites)

Other conditions which are more common in people with PBC are:

- indigestion
- pain in the abdomen

Signs of PBC usually are present in those with advanced liver disease and are not specific to PBC.

These include:

- jaundice
- easy bruising
- reddened palms
- swelling of ankles
- swelling of abdomen

Some people may notice yellow plaques around the eyes and also some pigmentation of the skin.

How Is PBC Diagnosed?

The presence of liver disease is detected by liver function tests on a single sample of blood. People with PBC nearly always have the specific antibody, AMA (antimitochondrial antibody) and finding this in the blood is useful in reaching a diagnosis. Doctors will usually examine the bile ducts to make sure there is no physical obstruction to the flow of bile in the bile ducts, such as from gall stones and will use imaging techniques such as ultrasound, X-rays or MRI. In some cases, it is necessary to confirm (or refute) the diagnosis of PBC by taking a liver biopsy, which involves taking a very small sample of cells from the liver with a needle. This also helps to determine the extent of any liver damage.

Is There A Treatment For PBC?

Although there is presently no cure for PBC, there have been some major breakthroughs in the treatment of the illness in the last few years.

Ursodeoxycholic Acid is currently the only approved medical treatment for PBC. More information is available from either your family doctor or hospital specialist.

A range of treatments is available which have been found to ease the symptoms. Drugs that dampen the body's immune system and slow the progression of the disease are under investigation.

Like most chronic liver diseases, PBC can impair the capacity of the liver to break down toxic drugs and chemicals. This is usually a problem only in those with advanced disease. These include prescribed and over the counter medicines, alcohol and even some foods.

Proper advice on a healthy and balanced diet is important, as a good dietary intake is advisable for maintaining health for people with liver diseases. Eating little and often is a good guideline for those who have PBC as this ensures that the bile acids always have something to work on and this helps to alleviate digestive problems.

As the liver may not be creating the correct balance of minerals and vitamins, the diet should be rich in calcium, vitamins and proteins. As in virtually everybody, foods

containing unsaturated fats (such as oily fish) are probably preferable to fatty foods, which are high in saturated fats, but a normal fat intake must be continued. If there is steatorrhoea (fatty stools), specific advice must be sought from the doctor.

Medication In PBC

Just because you have PBC, it does not mean that you cannot have other problems! You may need, from time to time, treatment for other conditions. You should always remember to tell your doctor, or dentist, that you have PBC. There are few medicines that you should avoid because of PBC, but the dose may need to be modified in some instances for some people.

For simple aches and pains it is, however, best to avoid Aspirin and medicines containing Aspirin. If you have minor aches and pains, then Paracetamol, up to four tablets a day, is usually safe. If the pain persists, or you require stronger painkillers, then you should see your doctor. You may wish to take vitamin tablets and provided you follow the instructions on the manufacturer's label, these do not usually cause any problems. Do remember that some herbal remedies may interact with medicines and herbal medicines too may have side-effects.

Itching

Patients with PBC often have itching. The most common medicine used to relieve the itching is called Colestyramine. The trade name for this is Questran. Some people on Questran have side-effects such as altered bowel habit, or bloating. If this is a problem, ask your doctor whether Questran Light would be an alternative for you. This is usually associated with fewer side-effects. You must be careful when taking Questran that it does not interfere with absorption of other drugs. Again, you should ask your doctor about the best way of taking the Questran and any interactions with other medications. There are other treatments for itching and if Questran does not work for you, then you should consult your doctor. Remember it may take several weeks for Questran to work. There are several other medications that are effective for the treatment of itching and these should be tried if Colestyramine is ineffective or cannot be tolerated.

Lethargy

This can be a very troublesome feature of PBC in some patients. There is, at present, no specific treatment for

the lethargy. It is important to be sensible and try to adapt your life-style to suit the lethargy. Doctors are evaluating some medications for the treatment of lethargy. Exercise is also effective but not everyone has the strength to take this form of treatment.

Osteoporosis

There are many reasons why people may have osteoporosis (thinning of the bones). Being female, middle-aged and having liver disease are each a good reason for developing osteoporosis. You should talk to your doctor about the risks of developing osteoporosis and measures you can take to reduce the bone loss. Amongst the measures you may wish to consider are increasing exercise (but this should be weight-bearing so swimming, while good for your heart and general health is not effective in preventing bone loss) and taking additional calcium. Again, you should talk to your doctor about the best way of taking calcium, but it is often better to take it in tablet form at night. Some women may benefit from hormone replacement therapy (HRT). Whilst HRT does have benefits in reducing bone loss and in reducing the chance of a heart attack, there are additional risks from HRT and you should discuss with your doctor whether HRT is appropriate for you and, if so, how you should take it. There are other treatments for preventing/treating osteoporosis. You should discuss this with your doctor. Smoking and drinking alcohol both make osteoporosis worse.

Liver Transplantation In PBC

PBC takes a long time to progress and it affects different people in many different ways. Most have few, or no, symptoms for many years and some live with a benign form of the illness with little or no discomfort. Many do have symptoms, but never reach end-stage PBC. For those few who do reach this stage, liver transplantation may be considered, especially where quality of life is deteriorating.

If medical treatment no longer controls the disease and the person has reached end-stage disease, transplantation will be considered. Because PBC progresses slowly, transplantation can be planned carefully and those with PBC have very good results compared to all liver transplant patients. Over seven hundred successful liver transplants are performed annually in the UK for various liver diseases, which dramatically improve people's quality and length of life.

Liver transplantation is very effective in relieving many of the symptoms and consequences of PBC. Itching, in particular, resolves rapidly. While life after transplantation is usually very good, it is never totally normal. PBC usually recurs in the graft, but this rarely causes problems. Thus, transplantation should be considered a 'swap' rather than a 'cure'.

Self Care

Supportive care, an altered lifestyle and self-help can be beneficial. For some, changes may have to be considered, e.g., giving up work, or particular activities because of tiredness and/or lack of concentration. Tiredness is a physical symptom of PBC and you may find that pacing your daily activities will help to preserve stamina and energy. Exercise is to be encouraged whenever possible, although this should only be undertaken when energy permits. Do remember physical capabilities are very individual and vary widely from person to person. Gentle walking and/or swimming can be beneficial.

Where can I receive more information and support?

The PBC Foundation is a nationwide charity which offers information and support for those who have PBC and those close to them.

Services include:

- A nationwide helpline
- Information packs for newly diagnosed patients
- Compendium - "Living with PBC"
- A quarterly magazine for members and the medical professions, offering advice on quality of life issues
- Information leaflets for Healthcare Practitioners
- Website
- Facilitating research projects
- Raising awareness amongst the general public and throughout the medical professions
- Advice on applying for DLA
- Employment advice in relation to PBC as a potential disability.

Membership is free and open to all – Join us!
To become a member, please visit our website or call us on 0131 556 6811.

Information on any other liver disease is available from The British Liver Trust. Telephone: 01425 481320