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# Primary Biliary Cholangitis

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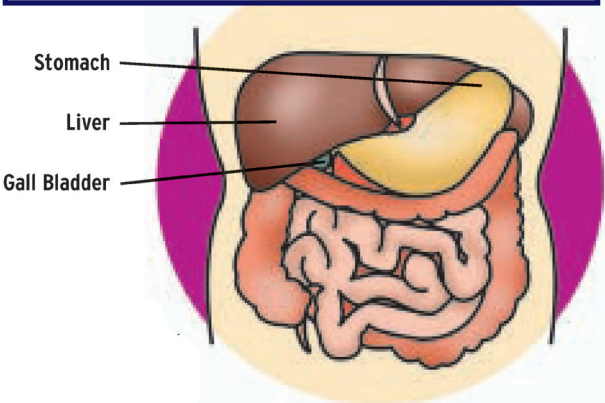
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**Patient Information Booklet**

## 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 many proteins.
- Stores sugars, vitamins and minerals including iron.
- Aids the digestive process by producing bile.
- Helps digest fat.
- Neutralises and destroys many chemicals and toxins.
- Controls the production of cholesterol.
- Maintains hormone balance.
- Regenerates its own tissue when healthy.
- Helps the body resist infection by producing some immune factors.



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

## 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?

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 pale yellow plaques around the eyes (xanthomas) and also some pigmentation of the skin.

Many people with PBC will live without symptoms. Some patients will experience one or two symptoms and some patients will experience more symptoms. Even though the symptoms of advanced liver disease are listed above, it is important to realise that relatively few patients with PBC reach the stage of advanced liver disease.

The severity of the symptoms often bears no correlation with the severity of liver damage or with liver biochemistry markers.

## How Is PBC Diagnosed?

A diagnosis of PBC is usually dependent on two pieces of information being present:

- Abnormal liver biochemistry (also known as LFTs, bloods or Liver Function Tests) usually involving raised levels of Alkaline Phosphates and GGT
- Positive results in PBC specific antibodies (AMA or in some cases ANA)
- Liver biopsy findings consistent with PBC (bile duct damage)

In most cases, liver biochemistry and positive antibodies are enough to diagnose PBC. If there is any doubt about diagnosis, then a liver biopsy may be done.

Around 95% of those affected by PBC will have positive AMA (Antimitochondrial Antibodies). A smaller number of patients will have positive ANA (Antinuclear Antibodies).

## **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 (UDCA) is currently the “Standard of care” in PBC. Every patient with PBC should be offered UDCA in a daily dosage of 13-15mg per kg. The majority of patients “respond” to UDCA which is usually indicated by a lowering of liver biochemistry markers. There are some data which suggest that even those who do not “respond” fully to UDCA still derive some benefit.

For those who do not respond to UDCA (usually measured by measurement of liver biochemistry over a period of 12 months), there is a secondary treatment available in many countries known as Obeticholic Acid (OCA).

For those who would not or cannot benefit from OCA, there may be opportunities to be involved in clinical trials. This can be discussed with your clinician.

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. These include prescribed and over the counter medicines, alcohol and even some foods. This is usually a problem only in those with advanced disease. If in doubt, you should consult your doctor or pharmacist.

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.

The diet should be balanced and contain adequate amounts of calcium, vitamins and protein. 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 a 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, dentist, nurse or pharmacist, 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/Fatigue

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 lifestyle to suit the lethargy. Doctors are evaluating some medications for the treatment of lethargy. Exercise can be very effective but this needs to be personalised to each patient. Unlike itching, the lethargy does not improve after transplant.

### 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 and 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, but certainly not for all, 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. Do remember physical capabilities are very individual and vary widely from person to person. Gentle walking, swimming, singing or even laughing can be beneficial.

In our experience, active self-management has a very positive impact upon living with PBC. Once people come to terms with the diagnosis of PBC, they are often in a position to make the best decisions possible to help their own situation.

## **Where can I receive more information and support?**

This leaflet is designed for those newly diagnosed who are looking for their first information about PBC. The PBC Foundation is a UK-based charity which offers information and support globally to those who have PBC and to those close to them.

### **Services include:**

- An international 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 GPs and consultants
- Website (in 19 languages)
- Self-management app (iOS and android)
- A UK-wide Volunteer Network
- Self-management workshops and webinars
- Facilitating research projects
- Raising awareness amongst the general public and throughout the medical professions
- Advice on applying for DLA
- HealthUnlocked peer-to-peer support forum
- International advocacy on behalf of patients
- Employment advice in relation to PBC

Studies have shown that accessing the Foundation's services improves management of PBC, of its symptoms and reduces isolation.

**Registration is free and open to all – Join us!**

To become a member, please visit our website or call us on +44 (0) 131 556 6811.