

Primary biliary cholangitis

PATIENT INFORMATION LEAFLET

Department of Medicine, University of Thessaly
University Hospital of Larissa

INTRODUCTION

The Department of Medicine, University of Thessaly at University Hospital of Larissa works to:

- support people with all kind of liver disease
- improve knowledge and understanding of the liver and related health issues
- encourage and fund research into new treatment
- lobby for better services

All our publications are reviewed by medical specialists and people living with liver disease. Our website provides information on all forms of adult liver disease. Calls us on 2413502436 or visit

<http://83.212.32.147/internalmedicine/index.php/el/>

THE LIVER

Your liver is your body's 'factory' carrying out hundreds of jobs that are vital to life. It is very tough and able to continue to function when most of it is damaged. It can also repair itself – even renewing large sections.

Your liver has around 500 different functions. Importantly it:

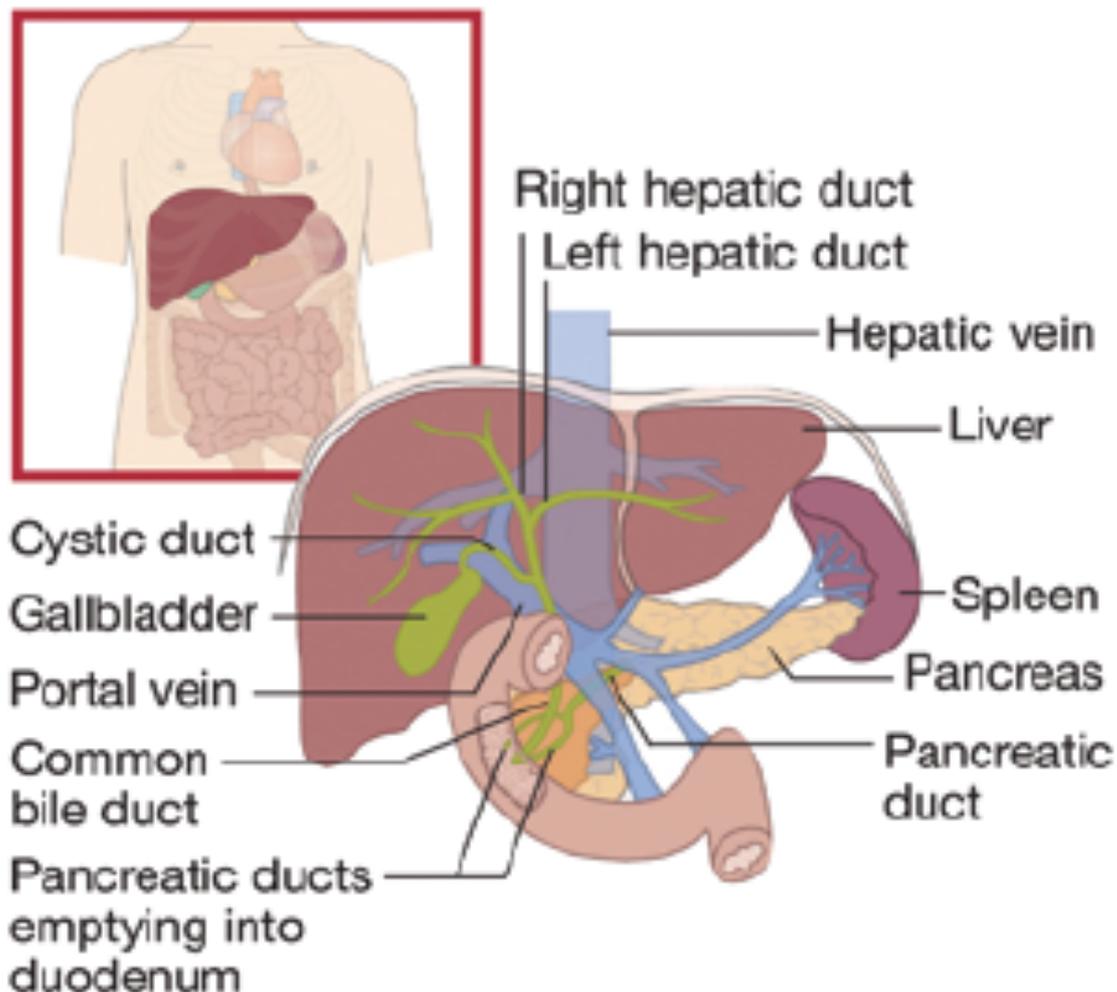
- fights infections and disease
- destroys and deals with poisons and drugs
- filters and cleans the blood
- controls the amount of cholesterol
- produces and maintains the balance of hormones
- produces chemicals – enzymes and other proteins – responsible for most of the chemical reactions in the body, for example, blood clotting and repairing tissue
- processes food once it has been digested
- produces bile to help break down food in the gut
- stores energy that can be used rapidly when the body needs it most
- stores sugars, vitamins and minerals, including iron
- repairs damage and renews itself.

HOW LIVER DISEASE DEVELOPS

Liver damage develops over time. Any inflammation of the liver is known as hepatitis, whether its cause is viral or not. A sudden inflammation of the liver is known as acute hepatitis. Where inflammation on the liver lasts longer than six months the condition is known as chronic hepatitis.

Fibrosis is where scar tissue is formed in the inflamed liver. Fibrosis can take a variable time to develop. Although scar tissue is present the liver keeps on

functioning quite well. Treating the cause of the inflammation may prevent the formation of further liver damage and may reserve some of the scarring.



Cirrhosis is where inflammation and fibrosis has spread throughout the liver and disrupts the shape and function of the liver. With cirrhosis, the scarring is more widespread and can show up on an ultrasound scan. Even at this stage, people can have no signs or symptoms of liver disease. Where the working capacity of liver cells has been badly impaired and they are unable to repair or renew the liver, permanent damage occurs.

This permanent cell damage can lead to liver failure or liver cancer. All the chemicals and waste products that the liver has to deal will build up in the body. The liver is now so damaged that the whole body becomes poisoned by

the waste products and this stage is known as end stage liver disease. In the final stages of liver disease, the building up of waste products affects many organs. This is known as multiple organ failure. Where many organs are affected, death is likely to follow.

WHAT IS PRIMARY BILIARY CHOLANGITIS (PBC)?

PBC is a chronic disease that can, little by little, destroy some of the tubes linking your liver to your gut. These tubes are called bile ducts.

The bile ducts carry a fluid called bile to your gut (also known as your intestine or bowels).

When PBC damages your bile ducts, bile can no longer flow through them. Instead it builds up in the liver, damaging the liver cells and causing inflammation and scarring.

Over the years this damage becomes widespread. The liver becomes less able to repair itself, leading to a condition known as cirrhosis. The damage caused to your liver cells by cirrhosis can result in the failure of your liver carry out all of its jobs properly.

WHAT IS BILE?

Bile is a yellow-green fluid produced by your liver which contains chemicals as well as waste products to aid digestion. It plays a central role in helping the body digest fat. It acts as a detergent, breaking the fat into very small droplets so that it can be absorbed from food in your gut. It also makes it possible for your body to take up the fat-soluble vitamins A, D, E and K from the food passing through the gut.

WHAT ARE THE SYMPTOMS OF PBC?

Some people with PBC will never get any symptoms of the disease. Clear symptoms of PBC are constant tiredness (for some people this can be severe)

and intense itching in any part of the body. Itching, also known as pruritus, may be a result of your liver's inability to process bile. It is thought that bile acids are not the cause of the itching but rather other chemicals that are retained in the body. As with tiredness, the severity of the itching will vary from person to person. Severity is not an indication of the amount of liver damage.

Other symptoms that may develop usually include the following:

- dry eyes and/or dry mouth
- constant or variable ache or discomfort in the upper right-hand side, below your ribs
- indigestion, nausea or poor appetite
- arthritis (inflammation of the joints)
- pain in the bones
- mottled palms with red or pink blotches
- diarrhea
- dark urine and/or pale stools
- jaundice – yellowing of the skin and whites of the eyes.

Tiredness and itching are generally the first symptoms to appear while jaundice is usually associated with the later stages of the disease.

WHO IS AT RISK OF PBC?

Nine out of ten people who get PBC are women. No one knows why this is. In particular the following women are most at risk:

- women who are middle aged or older

- women who have a family history of PBC.

There is no evidence that PBC can be passed on to others by simple contact, kissing, blood or sexual contact.

WHAT ARE THE CAUSES OF PBC?

PBC is an autoimmune disease. This is a disease where the body attacks itself. In this case, your body's defense against germs and infections – the immune system – mistakes the cells lining the bile ducts as 'foreign' and attacks them.

Why this happens is still not known. Some medical specialists believe that something may trigger your body into making this mistake. Possible causes of this 'trigger' could be:

- an infection
- some form of toxin (poison) taken in from the environment.

Occasionally PBC comes to light during or just after pregnancy. It is not clear whether pregnancy may spark it off or, because pregnant women are under close medical supervision, the symptoms of PBC are first noticed during pregnancy.

This external trigger probably starts PBC only in people who are already at risk because they have inherited certain genes from their parents.

TESTING FOR PBC

AUTOANTIBODIES

Most people with PBC have something in their blood called antimitochondrial antibody (AMA). An antibody is a chemical made by the body to attack an

'invader'. Although doctors are not really sure why, the presence of AMA in your blood is an important sign that you have PBC.

LIVER FUNCTION TESTS (LFTS)

This type of test is performed to gain an idea how the different parts of your liver are functioning. The liver function test is made up of a number of separate examinations, each looking at different properties of your blood.

In PBC, doctors will be looking for increased levels of both alkaline phosphatase (ALP), an enzyme released into the blood by damaged bile ducts, and the immunoglobulin IgM.

The liver enzymes alanine transaminase (ALT) and aspartate transaminase (AST) are also monitored, although these are a measure of any leakiness or damage relating primarily to liver cells rather than the bile duct cells.

ULTRASOUND SCAN

Ultrasound is the same technology used to confirm all is well in pregnancy. Gel will be applied to your skin, which may feel slightly cold. A probe, like a microphone, will be moved across your skin to send sound waves into the liver area.

The reflected sound waves, or echoes, are picked up through the probe and used to build a screen image of the liver's condition.

This painless test is used to check the condition of the bile ducts and to rule out the possibility that your symptoms could be a sign of a different liver problem.

LIVER BIOPSY

If tests show you have PBC you may need (not always) a liver biopsy to see how serious the condition is.

During a liver biopsy a tiny piece of the liver is taken for study. A fine hollow needle is passed through the skin into the liver and a small sample of tissue is withdrawn.

The test is usually done under local anaesthetic and mean an overnight stay in hospital. As the test can be uncomfortable and there is a very small risk of internal bleeding or bile leakage, a stay in bed of four hours is required. Ask your doctor for more information on this.

TREATMENT

There are a number of treatments for the symptoms of PBC. Some of them help with any unpleasant side effects, such as dry eyes, and others slow the progress of the disease.

MANAGING UNPLEASSED SYMPTOMS

ITCHING (SKIN):

A drug called cholestyramine (sold as Questran) may be prescribed by your doctor to help ease itching. Taken orally, cholestyramine works by preventing re-absorption of the chemicals that cause the itching. It can take days or even weeks before this becomes effective.

Some people taking cholestyramine have problems such as changed bowel habits and bloating. Your doctor may prescribe 'Questran light' to reduce these side effects.

If cholestyramine does not help, a hospital specialist may try other medicines such as rifampicin and naltrexone.

Itching is made worse by dry skin. It is very important to use plenty of moisturizer. If you're running a bath, don't make it too hot.

DRY EYES AND DRY MOUTH

The combination of dry eyes and a dry mouth (sicca syndrome) might be soothed by such treatments as artificial tears and saliva, lubricating gels and oestrogen creams. You may find that lozenges from your pharmacist will help with the dryness in your mouth.

CONTROLLING YOUR CONDITION

A medication called ursodeoxycholic acid (URSO, also referred to as UDCA) has helped some people. Made from naturally occurring bile acid, it seems to work through several mechanisms by protecting the liver from the harmful chemicals in the bile.

Although early studies failed to show any direct impact on survival rates, new evidence suggests that URSO may help to keep people alive for longer and delay liver transplantation.

A number of other medicines are also now being tried and your medical team may ask for your assistance in this. At present, however, a liver transplant is the only option for people who have advanced PBC.

WILL I NEED A LIVER TRANSPLANT?

A liver transplant is usually only recommended if other treatments are no longer helpful and your life is threatened by end stage liver disease. It is a major operation and you will need to plan it carefully with your medical team, family and friends.

However, liver transplantation works well for people with PBC. It is possible to get PBC in your new liver, but it may take up to 15 years before the disease becomes significant.

LOOKING AFTER YOURSELF

PBC affects people in very different ways. Because of this it is very difficult to say what impact PBC may have on your life.

For example, many people with PBC may live with very few problems for many years or even decades. On the other hand, PBC can have a major impact on people's day-to-day lives early on by causing unpleasant symptoms such as very itchy skin.

If your liver has become damaged it can have a major impact on your health. For example:

- it may reduce your body's ability to digest food properly, especially fats
- it may mean your body does not get enough energy, making you always feel tired (fatigue)
- you may not cope very well with poisons such as alcohol and some medicines.

DIET

Whether you have PBC yourself by eating a healthy diet. This means:

- plenty of fruit and vegetables (at least five portions a day)
- lots of carbohydrates (pasta, potatoes, whole meal bread and rice)
- only a little fat (as little saturated animal fat as possible).

Some people with PBC may need to eat a special diet. If you have any questions about your diet, talk to your doctor. You can ask to be referred to a

registered dietician for some personal advice.

If you have any of the following you should definitely get advised by your doctor and dietician:

- cirrhosis
- fluid building up in your abdomen (ascites) or leg (oedema)
- mental slowness or confusion (encephalopathy)
- diarrhoea where the stools are pale, bulky and difficult to flush away (steatorrhoea).

PROBLEMS WITH FAT

Some people with PBC have difficulty digesting fat. This leads to a type of diarrhoea called steatorrhoea (see above). Some people may also feel sick and bloated. If this happens to you, apart from seeing a registered dietician, you may find that a low fat diet helps to reduce the diarrhoea, abdominal pain and any other discomfort from the steatorrhoea.

Fat is essential and should not be cut out of your diet completely without proper advice from a dietician. Fat is an important source of energy, so if you do have to cut it out you may need to eat more carbohydrates for more energy.

HEARTBURN

PBC may cause some people to experience an unpleasant taste in their mouth, or heartburn – a severe pain in the chest that can be mistaken for heart problems. Stomach acids escaping into your food pipe (oesophagus) are the usual cause of this sensation.

If you get heartburn you may find that eating small amounts often helps. Avoid big meals and spicy food. It is a good idea to always carry something to eat, preferably food high in carbohydrate such as a barley sugar or boiled sweet.

ALCOHOL

Alcohol is actually a poison dealt with by the liver. If your liver is damaged it may mean that you have difficulty coping with alcohol and many people with PBC find they can no longer drink alcohol. Others may find they can only drink a little alcohol now and then, perhaps on a special occasion.

Sensible drinking advice varies from person to person. It will depend on many different things, such as how serious your liver problems are and on your general health. Your doctor will be able to advise you.

ENERGY LEVELS

Always feeling tired is caused by your liver being unable to provide enough energy for the rest of your body. This means that carrying out day-to-day tasks can be exhausting. You may find that making changes to your lifestyle can make a big difference, such as:

- pacing your activities
- doing gentle exercises such as walking and swimming to help make you fitter without becoming exhausted
- changing your hobbies and daily activities
- if you are able, giving up your work or doing it part-time.

CAUTION WITH MEDICINES

Talk to your doctor before taking any medicine or treatment. Many medicines may have unwanted side effects that are usually dealt with by your liver, which

may not be working very well. For example, it is best to take paracetamol (but never more than four 500mg tablets in any day) rather than aspirin for day-to-day aches and pains.

However, most drugs are well-handled by the liver in PBC even in end stage disease.

THINNING BONES (OSTEOPOROSIS)

As we get older our bones often become thinner and weaker. This is more common among women, especially after the menopause. PBC may make this worse. You can help to stop this problem by doing plenty of weight-bearing exercise and eating food with plenty of calcium, such as skimmed milk.

Sometimes, despite these efforts, medicine is needed. You may be advised to take calcium supplements, a 'bone enhancing' medicine called a bisphosphate or a course of hormone replacement therapy (HRT), for example.

Bone loss can be detected with a special type of X-ray called a 'Dexa Scan'.

HIGHER RISK OF BLEEDING

With PBC you may be more at risk of bleeding because your liver is less able to make the chemicals needed to clot blood. This means that before treatment you need to tell people such as your dentist that you have PBC so they can take special care. But remember, there is no evidence that PBC can be passed on from contact with blood.

WHO ELSE CAN HELP?

PROMETHEUS – HELLENIC LIVER PATIENT ASSOCIATION

Tel.: 211 0122102

Address: L. Alexandras 213B, 11523 Ampelokipi, Athens

email: info@helpa-prometheus.gr

<http://www.helpa-prometheus.gr>

HPARXO – HELLENIC SOCIETY OF LIVER TRANSPLANTED PATIENTS

Tel.: 2103800550

Address: Veranzerou 15, 10677, Athens

email: info@shpel.gr

<http://www.shpel.gr>

This patient information leaflet was compiled by the Department of Medicine, University of Thessaly at University Hospital of Larissa.

Department of Medicine, University of Thessaly

University Hospital of Larissa

Mezourlo (Biopolis), 41110, Larissa

Tel.: 2413502285

Fax: 2413501557

email: skapeti1@gmail.com

<http://83.212.32.147/internalmedicine/index.php/el/>

