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Primary Biliary Cholangitis

Liver NORTH
LIVER PATIENT SUPPORT

information leaflet 6
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What is Primary Biliary Cholangitis (PBC)?

PBC is a liver disease in which the small bile ducts of the liver are slowly damaged. These bile ducts are channels that normally drain bile from the liver. Bile is important for transporting digestive juices from the liver to the gut, and for removing poisons filtered by the liver from the blood into the gut. In PBC the body mistakenly identifies the cells lining the bile ducts within the liver as being foreign and attacks them. This damage leads to inflammation and swelling of the bile ducts, which makes it more difficult to drain the poisons away. This causes scarring which over time can become widespread. When there is extensive damage and scarring this is called cirrhosis.

If you have PBC will it always become cirrhosis?

NO! Many people with PBC never develop cirrhosis. Often the rate that PBC progresses can be very slow. PBC takes a long time to develop and it affects different people in many different ways. Most people have few, or no symptoms for many years and some live with a benign form of the disease for many years without experiencing any problems at all. Many people with PBC do have symptoms, but never reach the end stage PBC.

Cirrhosis. Does that mean it is caused by drinking alcohol?

NO! It is a popular misconception that the term cirrhosis refers to liver damage caused by drinking too much alcohol. This is not the case. The term cirrhosis refers to the combination of liver scarring caused by chronic liver damage and the attempts made by the liver to regenerate itself. Any cause of chronic liver damage (and there are probably well over 100) can result in cirrhosis.

What causes Primary Biliary Cholangitis?

The cause of PBC is currently unknown. It is thought to be an autoimmune problem, where your body's immune system does not work properly and (to a limited extent) turns on itself. Like many other autoimmune problems, PBC predominantly affects females (9:1), although it is thought that when PBC does occur in men it causes the same problems (or lack of problems) as it does in women. PBC does seem to run, to a limited extent, in families, with about 1 in 30 people with PBC having other people in the family who also have it. People with PBC tend to present to doctors with symptoms around the age of 60, although it can occur in any age group. Research has shown that people with PBC have several abnormalities of their immune system, and in the future these studies may help us to explain what is the cause of PBC, and why it occurs in some people and not others. Lots of studies have been done which suggest that an infection or toxin may act as a trigger for PBC.

How common is PBC?

PBC seems to be more common in Northern Europe and North America. In the North East of England 1 in 700 women over the

age of 40 have PBC.

What problems does Primary Biliary Cholangitis cause?

Patients with PBC develop two types of problems.

The first type of problem results from chronic damage to the liver resulting eventually in the development of cirrhosis. Whilst loss of hepatocytes (liver cells) initially causes little problem (the liver normally has a huge amount of spare capacity) eventually insufficient hepatocytes may remain to perform the normal functions of the liver. This can result in, amongst other things, the development of jaundice and problems with blood clotting (the clotting factors which are needed to make the blood clot are mostly made by the liver). Where cirrhosis develops as a result of chronic liver damage additional problems caused by an increase in the pressure of blood within the blood vessel connecting the bowel and the liver (the portal vein) can develop (portal hypertension). Problems caused by portal hypertension include development of blood vessel in the gullet with increased risk of bleeding (oesophageal varices) and accumulation of fluid in the abdominal cavities (ascites).

The second type of problems encountered by patients with PBC are related to the symptoms that are associated with the inflammatory process going on within the liver. These symptoms vary hugely between people, and their severity often bears no relationship to how severely the liver is damaged. The symptoms are important because they can affect your quality of life. The symptoms that people with PBC most frequently complain of are fatigue, problems with their sleep and memory, and sometimes muscle and joint pains. PBC can also make you itch and may lead to dry eyes and mouth and sometimes abdominal pain.

How is Primary Biliary Cholangitis diagnosed?

Where the condition is suspected (because of the presence of suggestive symptoms) the first test to be carried out will be the liver function tests (LFTs). This is a blood test, which assesses for the presence of damage to the cells of the liver. If abnormal it suggests that damage is occurring, but does not identify what is causing the damage. The next test to be carried out will be a blood test looking for the presence of specific antibodies in the blood, which are suggestive of the presence of Primary Biliary Cholangitis.

As physical obstruction is a common cause of jaundice, often doctors will need to visualise your liver. The easiest way to do this is with an ultrasound test. This is a simple test, which allows measurements of your liver and other organs in your abdomen. Sometimes the diagnosis may need to be confirmed by taking a tiny piece of liver during a liver biopsy (a procedure where, under local anaesthetic, a needle is used to remove a tiny piece of liver tissue, which is then examined under a microscope). In this day

and age most people do not need to have a biopsy. Where it is suggested it will be because the information that it can provide will make an important difference to your treatment.

Once the disease has been diagnosed and, where appropriate, treatment started, the response to treatment will be monitored primarily through the use of further blood tests. Very occasionally it will be necessary to repeat the liver biopsy to monitor response to treatment.

How is Primary Biliary Cholangitis treated?

At the moment there is no cure for PBC. People with PBC often take Ursodeoxycholic acid, which is licensed for treatment of PBC and is thought to slow down the progression of the disease. Other treatment may include Obeticholic Acid or Bezafibrate. Management of PBC is centred around control of the symptoms that are associated with it, as well as slowing down its progress.

Treating the symptoms of PBC

Itch is a common problem in people with PBC and it is easily treated with a range of oral medications. It is important to mention this symptom to your doctor.

Fatigue is described by those with PBC as the worst symptom of the disease. At the current time it is not clear what causes fatigue in PBC, and we cannot cure it. In the first instance it is important that causes of fatigue such as anaemia and thyroid problems are ruled out. Research has shown that fatigue is worse in those on certain medications and in those who have problems regulating their blood pressure. Again this is a symptom that it is well worth mentioning to your doctor as there may be things that can be done to help.

Dry eyes and dry mouth can be treated with lozenges and artificial tears.

Osteoporosis (or thinning of the bones) occurs more frequently in middle-aged females who have been through the menopause – just the population who typically suffer from PBC. It is therefore important to see whether osteoporosis is present by measuring whether your bones are thin every 5 years by a scan called a DEXA. This x-ray scan decides whether you have osteoporosis or not.

Liver transplantation in PBC

A liver transplant may be considered in a small proportion of people with PBC, this may be in those who present for the first time, or whose illness progresses. As PBC is slowly progressive, transplantation can usually be carefully planned well ahead. Those with PBC have very good results with transplantation, and it can dramatically improve people's quality of life. Liver transplantation is very effective in relieving many of the symptoms and

consequences of PBC, including itch and fatigue.

What can I do to help myself?

Medicines

In those who have a liver problem such as PBC, the liver is less able to carry out many functions; in particular the liver may be less able to break down alcohol and some medicines. It is therefore important to always tell doctors and the pharmacist that you have PBC.

Diet

It is important to eat healthily and to drink alcohol sensibly. If you are concerned about the amount of alcohol you can drink it is important to discuss this with your doctor. If you have cirrhosis or fluid retention (ascites) or have periods where you are confused (encephalopathy) then you will need to modify your diet, and your doctor will advise appropriately.

Activity

Fatigue can often mean that people do not feel like exercising. Sometimes people find that pacing activities allows them to do more or changing the type of exercise may help.

Mood

It is understandable (and not unexpected) that at times having PBC will get you down, and that the symptoms may make life harder than it used to be. This is something that often happens and it is important that people around you understand and are supportive. Having a friend to talk through how you feel with can sometimes help.

Acknowledgement:

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LIVErNORTH can help you. As well as having many of the Freeman's health professionals as members, we have access to extensive information on all liver diseases and treatments - contact details on the back cover.

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