Liver Foundation

Understanding Primary Biliary Cholangitis (PBC)

A guide to diagnosis and management

Learn more at liver.org.au

Liver Foundation acknowledges the Traditional Custodians of this land. We pay our respects to Elders past, present and emerging.

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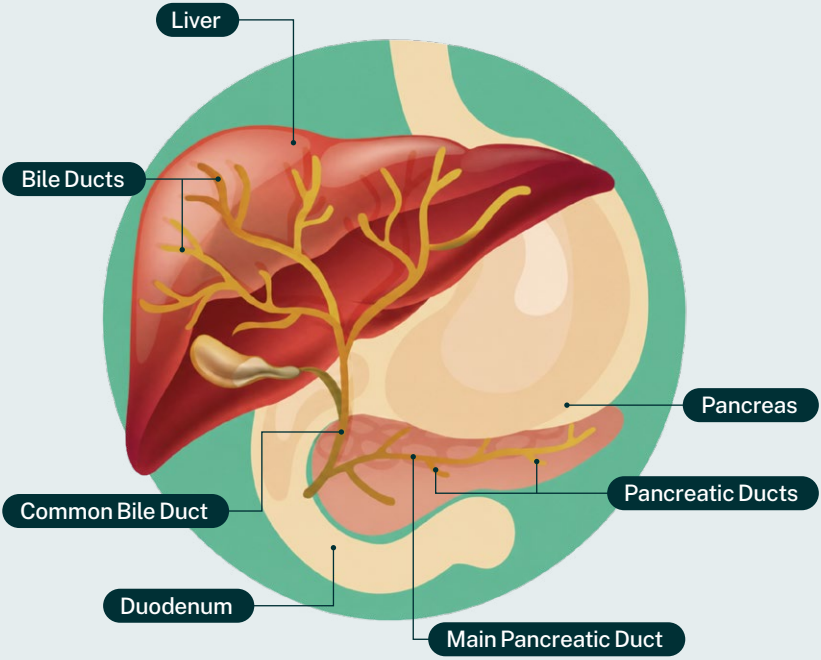
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WHAT IS PRIMARY BILIARY CHOLANGITIS?

Primary Biliary Cholangitis (PBC) is an autoimmune disease. Autoimmune diseases are where the body's own immune system attacks healthy cells and tissue.

If you have PBC, your immune system starts to attack the small bile ducts in the liver; these are a network of small tubes that carry bile in the liver. The ducts get inflamed (swollen) and blocked. Eventually, they can be destroyed. If this happens, bile builds up and causes liver damage.

THE LIVER AND BILIARY SYSTEM



PBC is a chronic disease (it is lifelong) and without treatment it gradually gets worse.

Although PBC is a very rare disease, the number of people who have it is increasing. More people in Australia are being diagnosed with PBC because better testing and increased awareness are helping doctors identify the condition earlier. Almost all cases of people with PBC (95%) are in women aged 35 to 70. It is most commonly diagnosed in middle age, between 45 and 65 years.

There has been a great deal of progress in our understanding of PBC, and most people will live a normal lifespan with good management of the disease and its symptoms. There is a lot of information about health online, but not all of it is accurate or reliable. Try to use trusted information from reputable health organisations. Information from unverified websites can sometimes be misleading or cause unnecessary worry.

This booklet will give you some useful information and support you to manage your care and talk to your GP and specialist. Sometimes, when people have a rare disease like PBC, it becomes even more important to prepare for your medical appointments to ensure your questions are answered.





WHAT ARE THE SYMPTOMS OF PBC?

PBC doesn't always cause symptoms. People might only find out they have PBC because they have an abnormal blood test for something else.

If you do have symptoms, they usually start with feeling very tired (fatigue) and/or having itchy skin.

Other symptoms that may be experienced by people living with PBC include:

- Dry eyes and mouth and/or vaginal dryness
- Brain fog (cloudiness that can lead to short term memory loss, forgetting a word midway through a sentence, forgetting what you went to a room for, difficulty making decisions)
- Running out of energy by the middle of the day
- Small yellow or white bumps around the eyes, eyelids, palms, soles, elbows or knees (these are deposits of cholesterol)
- Diarrhoea or oily poos because the body can't digest fat properly
- Pain in the upper right side of the belly
- Pain in the joints, muscles or bones
- Osteoporosis (brittle bones)
- Gallstones or bile duct stones
- High cholesterol
- Underactive thyroid
- Vitamin deficiency (your body can't absorb vitamins including A, D, E and K)
- Restless legs

HOW IS PBC DIAGNOSED?

If your doctor thinks you may have PBC, they will order some blood tests to check for signs of liver disease or injury to the bile ducts. The blood test will also check for antibodies that signal you may have PBC. These are called antimitochondrial antibodies (AMA).

Some people may also need an ultrasound or measurement of liver stiffness (e.g. FibroScan® or Shearwave Elastography (SWE) to check for any signs of fibrosis or cirrhosis (liver scarring). Other imaging to look at the bile ducts and rule out other diseases that have similar symptoms may also be needed.

If it's still not clear what's causing the disease, you may need a liver biopsy. This is when a small piece of tissue is removed from the liver to be examined under a microscope. Biopsies are used less frequently due to the accuracy of the AMA results along with abnormal liver results.

It's recommended that people with PBC are also checked for other autoimmune diseases such as coeliac disease, thyroid disease and Sjögren's syndrome.



WHY DID I GET PBC?

Doctors don't fully understand what triggers the body to turn against its own cells. It's possible that you inherit genes from your family, or that something in your life caused it to happen. Some of the triggers for PBC are thought to be:

- Smoking
- Exposure to some chemicals
- Urinary tract infections

As you are being diagnosed, your doctor might ask you about risk factors for other causes of liver disease such as viral hepatitis, metabolic or alcohol related liver diseases. This is a standard part of the diagnostic pathway and is not meant to be a judgement of you. It is important for your doctor to have a full picture so they can best support you.

CAN I PASS PBC ON TO MY CHILDREN?

PBC is not contagious, however research has shown that there is often a genetic link to other autoimmune conditions which can run in families. There is about a 5% chance your children will also develop PBC.



Treatment

HOW IS PBC TREATED?

There's no cure for PBC, but medications can be used to slow it down and prevent complications. Most of the medications are well tolerated but it is always important to discuss any concerns with your doctor. Your doctor will help you decide what is the best treatment for your personal circumstances.

Medicines you might be given include:

1 **First line treatment:** (this is the first treatment that will be tried):

Ursodeoxycholic acid (Urso) or UDCA capsules help the liver work properly by moving bile through the liver and into the small intestine. This medication can prevent the PBC developing into liver failure. You may need to take this medicine for around a year before your doctor can tell (by looking at your blood tests including the liver enzyme ALP) if you have responded to this treatment or not. You will need to take it every day for the rest of your life.

The dose of Urso is based on your weight, which means your doctor will need to weigh you to calculate an accurate dose (13-15mg/kg). You will need to inform your doctor if you lose or gain weight. It is very important to talk to your doctor if you are experiencing side-effects and unable to tolerate Urso. This medication is best taken with or after food. Your doctor will advise whether you take a single dose or split the dose throughout the day. You can check your dose using the diagram shown on pages 10-11 or see our full resource here:

https://liver.org.au/wp-content/uploads/2026/08/pbc-urso-calculator.pdf ↘





2 Second line treatments:

For people who don't improve with or can't tolerate Urso, second line treatments are available.

Elafibranor is a new medicine available on the PBS for second line treatment of PBC and is usually used in combination with Urso, but can be used by itself if Urso is not tolerated. It is not associated with worsening of itch. It should not be taken when pregnant so if you are planning pregnancy or become pregnant, please discuss with your doctor. A side effect to be aware of is muscle aches, if this develops, please discuss with your doctor.

Obeticholic acid is available on the PBS and can be used in combination with Urso if you haven't responded well to it, or used by itself if Urso is not tolerated. It may not be used as commonly as second line treatment since this medicine is contraindicated (not safe to take) in people with advanced cirrhosis. It can cause itching to get worse, so you should discuss with your doctor beforehand regarding any symptoms of itch or if it worsens on treatment.

Seladelpar is another new medicine you may hear about, but it is not currently available in Australia.

Fibrates (fenofibrate and bezafibrate) have been used to treat people with an inadequate response to UDCA and they are effective, however neither medicine is approved in Australia for the treatment of PBC.

There are also treatments for some of the complications of PBC. Your doctor might give you medicine to treat high cholesterol, osteoporosis or portal hypertension. You will need regular liver function blood tests. If you start to develop liver cirrhosis, this will also require regular follow up and management.

Treatment can slow PBC progression and protect your liver. Regular check-ups help keep your liver as healthy as possible.

EN

KNOW YOUR URSO DOSE

Your Ursodeoxycholic Acid dose is calculated on how much you weigh.

Use the Urso Dose Calculator to help you understand and manage your PBC treatment.



1. weigh yourself

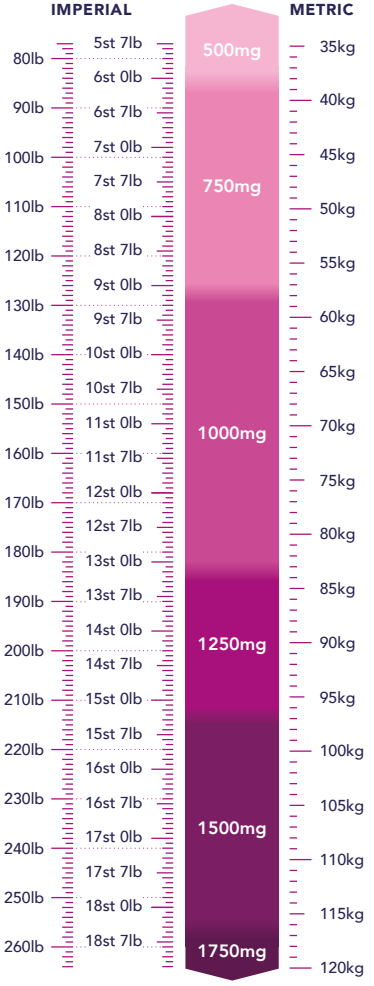


2. find your weight



3. know your dose

URSO DOSE CALCULATOR



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YOUR **URSO DOSE CALCULATOR** FOR PBC.

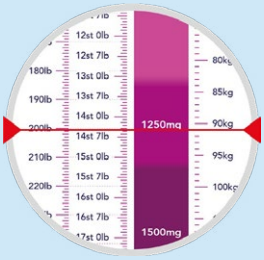
It's extremely important to know your weight as this has a direct relationship with your Urso dose. Remember, weight doesn't always stay the same, so if you gain or lose weight you may need to change how much Urso you take.

Use our 1, 2, 3 approach to find the right dose you.

1. weigh yourself | 2. find your weight | 3. know your dose

Ursodeoxycholic Acid (also known as Urso or UDCA) is the recommended first line treatment for people with **Primary Biliary Cholangitis** (PBC).

- Published research* has shown that for most patients, **responding to Urso** leads to a normal life expectancy.
- Data tells us that you are most likely to respond when taking the **optimal dose** (13-15mg/kg/day).
- Did you know, that patients who do not respond to Urso are also found to have **health benefits** from taking Urso every day?
- Unfortunately, there are still many PBC patients on too low a dose which can have **serious implications**.



Based on the optimal dose of 13-15mg/kg/day. A person who weighs **14st 5lb / 200lb / 91kg** has a recommended daily Urso dose of **1250mg**.

IMPORTANT INFORMATION:

Our **Urso Dose Calculator** has been designed as guidance for achieving the standard recommended dosage. In some clinical situations, your clinician may recommend a different dose. If you are not taking the optimal dose, we advise that you discuss this with your clinician so that you are on the right dose for you.

Do not stop or adjust your dose of Ursodeoxycholic Acid without discussing this first with your clinician. Urso is recommended as a lifetime treatment. Even if your liver results are normal you should not stop the treatment, this likely means it's working!

Keep this handy for future reference and/or when discussing your treatment with a healthcare professional.

*SOURCE: 'European Association for Study of the Liver (EASL), PBC Guideline 2017', 'American Association of Study of Liver Diseases (AASLD)', and 'PBC Guidance 2018, and British Society of Gastroenterology (BSG) PBC Guideline 2018'.

Adjusting to living with a long-term or chronic illness can sometimes feel overwhelming or challenging.

It is important to understand that there is nothing you could have done to prevent it, and that with the right treatment and monitoring you should have a normal lifespan. There is support available that can help you to come to terms with your diagnosis.

You will be referred to see a liver specialist (a hepatologist or gastroenterologist) who will monitor you regularly and guide your treatment along with your GP.

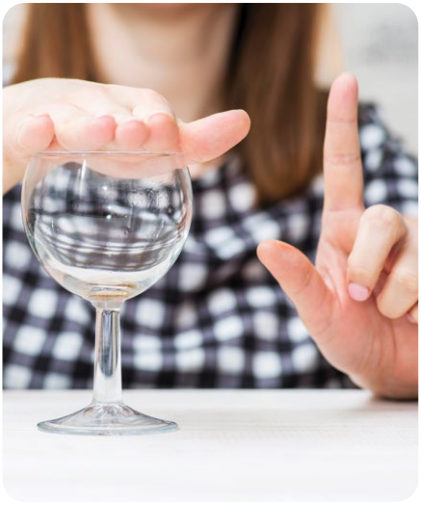
It is important to look after yourself as best you can. There are a few things which can help you to feel better and can also improve your overall health when you are living with PBC. These include:

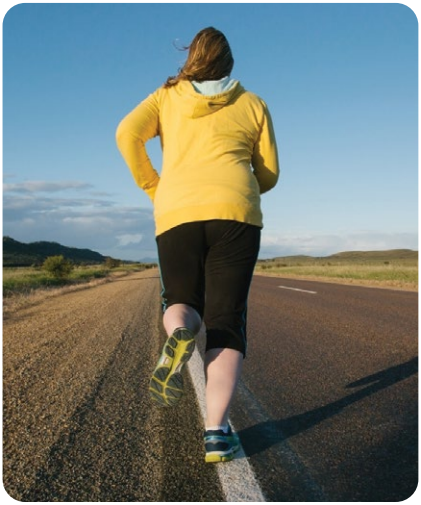
- Decreasing or cutting down alcohol – if you already have cirrhosis it is important you don't have any alcohol.
- Not taking any drugs or medicines that can harm the liver (including illicit drugs, alternative medicines and liver tonics).
- Continuing to exercise or finding ways to increase your daily exercise can help manage and improve fatigue. Strength and resistance training can also help protect your bones and prevent osteoporosis.
- De-stress if you can, practicing things like yoga, tai-chi, mindfulness and meditation can be helpful ways of relieving stress.
- Stop smoking.
- Look after your skin, especially if you are very itchy.
- Treat your dry eyes to keep the surface of your eye healthy and reduce symptoms of grittiness, itchiness and blurred vision.
- Manage symptoms of dry mouth and to avoid dental problems, get your teeth checked regularly.
- Seek support for your mental health if you need it, being diagnosed and adapting to living with PBC and its symptoms can be challenging.

There are things you can do to manage some of the more bothersome symptoms of PBC. Firstly, make sure your doctors know that you experience these symptoms. Your liver blood test levels are not always an indication of your symptoms, so unless you tell your doctor, they will not know.









FATIGUE

People with PBC often feel very tired. It's not a sign of how serious the disease is – sometimes people in the early stages feel very fatigued while others with more advanced disease don't feel tired at all. To treat tiredness, your doctor will talk to you about your diet, exercise and sleep. They may also test your thyroid as thyroid disease is more common in people with PBC and can also make you tired. Other causes of fatigue such as anaemia or sleep disturbances should also be ruled out.

ITCHING

Itching in PBC can be severe, the medical term is pruritus. There are several treatments available to help with itching. Medicines such as cholestyramine that binds bile acids to reduce itching, or rifampicin (used to treat some infections) may be effective in reducing itching by affecting the levels of bile acids. Other medicines including sedating antihistamines may help improve symptoms.

If you're itchy, try to keep your fingernails short. Avoid hot baths and showers, try to wear natural fibres and watch you don't get too much sun. Keeping the skin moisturised can also help - use simple water-based lotions or creams. Menthol based creams can also help with mild itch.

TIPS TO MANAGING ITCHING



- Use warm or cool water in the bath / shower
- Use mild, fragrance free soap and moisturisers
- Put a cool compress over the itchy area
- Wear loose fitting, cotton clothes
- Avoid heat and chemicals such as chlorine or spa water

DRY EYES AND MOUTH

You can buy artificial tears and saliva over the counter from a pharmacy. Chewing gum or sucking a lolly can help. If you experience vaginal dryness you could talk to your GP regarding topical vaginal oestrogen or using water-based lubricants.

DIET

It's important to eat a healthy, balanced diet to improve your overall health and help your liver work properly. Staying hydrated by drinking water is also very important. Eating a variety of whole foods including fruits and vegetables, lean protein sources, wholegrains, dairy and sources of healthy fats can ensure your liver can function at its best. Because your body may have trouble absorbing some vitamins, your doctor may recommend supplements such as A, D, E and K, calcium, folic acid or iron.

Liver Foundation has a great resource – the Fatty Liver Diet (https://liver.org.au/the-fatty-liver-diet/) that is suitable for people with PBC who do not have cirrhosis. It has loads of great recipes, exercise and mindset tips.

If you have been told that you have cirrhosis, your dietary needs may change, and it is important to speak with an accredited practicing dietitian for individually tailored advice.



WHAT MONITORING WILL I NEED TO HAVE?

You will most likely have regular blood tests which will guide your treatment. You may also need some other scans like an ultrasound or liver stiffness measurement at intervals decided by your treating team to monitor your liver health.

Over 15 to 20 years, some people with PBC might develop cirrhosis. In some cases, cirrhosis can lead to liver failure. Cirrhosis simply means scarring of the liver, and this can be caused by many different liver and biliary diseases. Most people with PBC will never develop cirrhosis. If you do, your doctor will talk to you about what ongoing care and monitoring you will need.

If you get cirrhosis, there are specific symptoms that you will need to monitor for. Many people will not experience side-effects or complications from cirrhosis but for some, complications related to cirrhosis can include:

- Swelling of the feet and ankles (oedema)
- Swollen belly due to build up of fluid (ascites)
- New veins in the oesophagus or stomach which can be at risk of bleeding (varices)
- Yellowness of the eyes (jaundice)
- Malnutrition which may cause muscle wastage
- Problems with memory and concentration (hepatic encephalopathy)
- Portal hypertension - when blood can't flow into the liver properly, pressure builds up in the portal vein system. To relieve pressure in the portal vein system, the blood takes detours around the liver in other veins in the oesophagus (food pipe) or stomach. The spleen may also get larger
- Liver cancer (HCC)

If you have cirrhosis, please visit Liver Foundation and search the term as we have a wide range of resources that can support you.

WILL I NEED A LIVER TRANSPLANT?

Most people will not require a liver transplant for PBC. If your doctor thinks you may require a liver transplant because your liver function starts getting worse, or you develop advanced cirrhosis, you may be referred to a transplant centre. Sometimes the PBC can come back, even after a liver transplant, but it rarely leads to liver cirrhosis.

A liver transplant may not always be an option. There can be many reasons that it is not the most suitable treatment for you and your treating team would talk to you about this.



Preparing for your appointments

TALKING TO YOUR DOCTOR

It's very common to feel overwhelmed during medical appointments and forget things you may have wanted to discuss. Spending just a few minutes preparing for your appointment, thinking about your symptoms, medicines and any questions you have can help ensure you get the most out of your time with your clinician.

There are 2 resources from the PBC Care Bundle developed by clinicians and patients in the United Kingdom to improve care and health outcomes for people living with PBC. Liver Foundation has adapted these for use in Australia. It is advised that these questions are discussed at each appointment you have with your doctor.

liver.org.au/wp-content/uploads/2026/08/ Patient-Guide -to-Appointment-Prep.pdf ↘

liver.org.au/wp-content/uploads/2026/08/ Patient-Guide -to-Discussing-PBC-with-your-Clinician.pdf ↘

TALKING ABOUT YOUR SYMPTOMS

PBC can cause symptoms that affect people in very different ways. Sometimes words such as fatigue, itch or brain fog do not fully describe what you are experiencing or how much a symptom affects your everyday life.

Before your appointment, it may help to think about what your symptoms feel like for you, how often they occur, and how they affect the things you usually do. Writing this down can make it easier to explain your symptoms to your clinician, particularly when appointment time is limited.

Itch (pruritus)

If you experience itching, think about how often you notice it and how much it bothers you.

How would you describe your itch?

- **Mild:** I notice it occasionally, but it does not usually bother me.
- **Moderate:** I notice it on and off during the day and it can be bothersome.
- **Severe:** It is present most or all of the time and causes significant discomfort.

You may also want to note whether your itch disturbs your sleep or affects your daily activities.

Fatigue

Fatigue can mean different things to different people. It is more than simply feeling tired, and there may be several ways that you experience it.

How does fatigue feel for you?

- Feeling tired, sleepy or low on energy
- Reduced stamina (I can't keep going as long as I normally would)
- Muscle tiredness, weakness or a heavy feeling in my body
- Physical or mental exhaustion, including difficulty concentrating
- I need more rest or recovery after activities
- Do I wake up feeling tired
- Do I have energy in the morning but by 2pm I have no energy
- Other .....

How does fatigue affect your day?

For example, think about whether it affects your ability to work, exercise, complete household activities, socialise, care for others or do the things you enjoy.



Preparing for your appointments

Brain fog

Brain fog is common and can also be difficult to put into words. It can affect how clearly or quickly you feel you are able to think.

What does brain fog feel like for you?

- Forgetfulness
- Difficulty concentrating or focusing
- Feeling confused
- Feeling vague or "foggy"
- Thinking or processing information more slowly
- Difficulty finding the right words
- Feeling mentally tired or exhausted
- Other .....

How does this affect your everyday life?

Dryness

PBC may be associated with symptoms of dryness, such as dry eyes or a dry mouth. If you experience dryness, think about where you experience it, how often it occurs and whether it affects your everyday life. Vaginal dryness may affect intimate relationships, and it is important to know that you can get help to manage this.

How does dryness affect you?





OTHER THINGS TO DISCUSS WITH YOUR CLINICIAN

Your appointment is also an opportunity to talk about changes in your overall health and make sure your PBC care is up to date.

Before your appointment, you may want to make a note of:

- **New or changing symptoms:** Have any symptoms started, improved or become worse since your last appointment?
- **Changes to your health:** Have you been diagnosed with another health condition, including another autoimmune or related condition?
- **Medicines:** Have you started, stopped or changed any medicines or supplements?
- **Prescriptions:** Do you need any repeat prescriptions? If you know the medicine and dose, write these down before your appointment.
- **Blood tests and scans:** Are there any recent results you would like your clinician to explain or discuss?
- **Other health checks:** Are you up to date with recommended checks, such as FibroScan or bone density testing, are your vaccinations up to date?
- **New PBC information:** You may want to ask, *"Have there been any new developments, treatments or research in PBC that might be relevant to me?"*

PRESCRIBED MEDICINES

Sometimes another healthcare professional, such as your GP, may want to prescribe a medicine for a health problem that is not related to your PBC.

If you or your healthcare professional is unsure whether a medicine is suitable for you, ask your PBC clinician or healthcare team for advice. Keeping an up-to-date list of all your medicines, including prescription medicines, over-the-counter medicines and supplements, can help your healthcare professionals make decisions about your care.

Support

WHERE CAN I GET FURTHER SUPPORT?

Liver Foundation provides free, trusted information and support for people living with liver disease or liver cancer, as well as their families and carers.

Visit our website at **liver.org.au** to find easy-to-understand information about liver conditions, treatment options, healthy living and ways to look after your liver.

For people living with Primary Biliary Cholangitis (PBC), visit our **PBC webpage** for information, resources and support to help you manage your condition.

Stay informed by signing up for our free newsletter at **liver.org.au** to receive updates about upcoming events, educational workshops, new patient resources, research and liver health information, delivered straight to your inbox.

Notes

Notes

About Liver Foundation

Liver Foundation is Australia's leading national organisation dedicated to improving liver health for all. Since 1995, we have been working to make liver cancer and liver disease a national health priority by raising awareness, supporting early detection, research and improving access to care.

Funded by the Australian Government, and staffed by expert hepatology nurses, the Liver Cancer Nurse-Led Support Line is an integral part of the Australian Cancer Nursing and Navigation Program that aims to ensure all people affected by cancer are empowered with education, resources, and the support they need to better navigate the health system.

Liver Foundation

liver.org.au/your-liver/liver-diseases/pbc