Your journey with primary biliary cholangitis (PBC)

“I know PBC is chronic, but I am going to do what I can to manage it and keep living life on my terms.”
A diagnosis of primary biliary cholangitis (PBC) can be difficult for you and your family.

Your doctor may have told you that PBC is an autoimmune disease. In fact, it is the most common autoimmune disease of the liver. Approximately one in every 1000 women over 40 have received a diagnosis of PBC, which is nine times more common in women than men.

Yet not many people have heard of PBC, and you may be left wondering where to turn for more information about this disease. That is exactly where this booklet can help. It can answer some of the questions you may have in addition to providing some helpful lifestyle tips that you can use on a daily basis.

Keep in mind that this booklet is not intended to replace the advice of your doctor. But it is intended to give you the information you need to have informed discussions with your doctor and play an important role in how your disease is managed.
PBC is a progressive autoimmune liver disease. This means that the body’s own immune system, which is designed to protect you against infections, mistakes your bile duct cells as foreign objects and attacks them. This causes slow, progressive damage to the bile ducts in the liver.

The liver is an important organ that has many functions, including:

**Providing the body with energy:** Produces, stores and supplies glucose and fat to other parts of the body

**Helping the body fight infections:** Contains cells that target and eliminate microorganisms

**Excreting toxins from the body:** Metabolizes alcohol, drugs and other chemicals

**Helping the body stop bleeding:** Manufactures proteins that clot the blood

**Regulating hormones:** Helps balance sex, thyroid, cortisone and other adrenal hormones

**Regulating body cholesterol:** Produces and converts cholesterol to other essential substances

**Regulating the supply of essential vitamins and minerals**

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**Understanding the progression of PBC**

- **Liver:** Produces a substance called bile (a yellow-green fluid) that eliminates toxic substances from the body and also aids in digestion

- **Bile ducts:** Small tubes that carry bile from the liver to the small intestine to help with digestion

- **Autoimmune response:** In PBC, the body’s own immune system mistakes your bile duct cells as foreign objects and attacks them.

- **Cholestasis:** Damage to the bile ducts causes bile to be trapped in the liver. Bile acid buildup is toxic and leads to further damage to the liver, which is indicated by high levels of alkaline phosphatase (ALP), a liver enzyme.

- **Fibrosis:** Over time, the accumulation of bile acids leads to more damage causing scarring of the liver.

- **Cirrhosis:** Occurs when there is widespread scarring of the liver.

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**What does PBC mean?**

- **Primary:** Occurs without an obvious cause
- **Biliary:** Related to, or affecting the bile ducts in the liver
- **Cholangitis:** Inflammation of the bile ducts
It’s a myth!

PBC used to be called primary biliary cirrhosis. A common cause of cirrhosis is chronic alcohol abuse. However, it is important to know that alcohol is not a cause of PBC.

Because of this, and the fact that less than 50% of people with PBC progress to cirrhosis, the name of the disease was changed to primary biliary cholangitis.
Diagnosing PBC is typically done using specific blood tests.

ALP
Your doctor may have identified PBC by noticing increased levels of a liver enzyme called alkaline phosphatase (ALP). ALP is released into the blood by damaged bile ducts.

AMAs
Your doctor may have also done a blood test to check for signs of an autoimmune disease, specifically for anti-mitochondrial antibodies (AMAs). These antibodies almost never occur in people who do not have PBC, even if they have other liver disorders. A positive AMA test is considered to be a good indication of PBC.

ALT and AST
Your doctor may also monitor other liver enzyme levels called alanine transaminase (ALT) and aspartate transaminase (AST). These enzymes are an indication of liver inflammation/damage, but does not indicate damage specific to the bile duct.

Imaging tests may also be used in the diagnostic process, including:

- **Ultrasound:** Uses high-frequency sound waves to produce images of organs and other structures in the body
- **Fibroscan:** A type of ultrasound test that shows how much scarring is on the liver. Fibroscans can be done on a yearly basis to monitor any changes that may occur in the liver over time.
- **CT scan:** A special X-ray technique
- **MRI (magnetic resonance imaging):** Uses magnetic forces to create detailed images of organs and tissues
- **MRE (magnetic resonance elastography):** Combines MRI imaging with sound waves to create a visual map of internal organs. This is usually used to detect hardening of the liver – an indication of cirrhosis.

On rare occasions, if a diagnosis still cannot be confirmed, a liver biopsy may be performed. This process involves collecting a small sample of liver tissue to be examined in a laboratory.

Signs that something is wrong
If you didn’t experience any symptoms prior to being diagnosed, you’re not alone – approximately 60% of people are diagnosed with PBC without experiencing symptoms and many people with PBC can be symptom-free for years after diagnosis.

The most common symptoms that typically occur early in the disease include:
- Fatigue: Feeling tired
- Pruritus [proo-rahy-tuhs]: Itching all over the body

Be sure to talk to your doctor if you experience these or any other symptoms.
PBC is a chronic (long-lasting) disease, but don’t let that discourage you.

Although it can’t be cured, PBC usually progresses slowly and can be controlled in most people. In fact, for many, the disease may remain silent (no symptoms) and may never become serious.

In a smaller number of people, the liver can become severely damaged as a result of liver scarring leading to poor liver function.

The severity of liver damage and function can be classified using a staging system. Staging provides information about the severity and location of the damage to the liver.

There are four stages of PBC:

<table>
<thead>
<tr>
<th>STAGE</th>
<th>WHAT IT MEANS</th>
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<tbody>
<tr>
<td>1</td>
<td>Inflammation and/or abnormal liver tissue localized in one area</td>
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<tr>
<td>2</td>
<td>Inflammation and/or scarring localized in one or two areas of the liver</td>
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<tr>
<td>3</td>
<td>Liver scarring progression</td>
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<tr>
<td>4</td>
<td>Severe scarring: cirrhosis</td>
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</table>
ALP is the key
As previously mentioned, increased levels of a liver enzyme called alkaline phosphatase (ALP) is usually the first indicator of PBC. But did you know that continued elevated levels of ALP can also indicate disease progression?

This is why it is important to continually monitor your ALP levels. Your doctor can perform a simple blood test called a liver function test (LFT) to measure your ALP levels.

**ALP operates as a disease barometer for PBC**
- Continually elevated ALP levels can mean that the disease is progressing and there is an increased risk of needing a liver transplant.

Some people being treated for PBC may still have ALP levels that put them at risk.

**Ask your doctor to perform an ALP blood test every 3 to 6 months.**

Graph the results of your ALP blood tests using the ALP tracker at the end of this booklet. You can also track your bilirubin levels, another indicator of liver function. That way, you can keep track of your liver health along with your doctor.
Well-controlled PBC can have a positive impact on outcomes

People with PBC can expect to live just as long as a healthy person without PBC if:

• It is at an early stage
• ALP levels are lower than 200 U/L
• Bilirubin levels are normal after 1 year of therapy

What to look out for

Along with ALP, bilirubin and albumin are also markers of PBC that can be monitored by LFTs.

A change in their levels can highlight the potential risk of disease progression.

**ALP levels rise**

- Normal: <120 U/L
- Mildly abnormal: 120–200 U/L
- High: >200 U/L

**Bilirubin levels rise**

- Normal: 1.71–20.5 μmol/L
- High: >20.5 μmol/L

**Albumin levels drop**

- Abnormal: <35 g/L
- Normal: 35–55 g/L

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**Take control**

By Knowing Your Numbers, you can take a more active role in managing your PBC.

Visit http://pbc-society.ca/know-your-numbers/ to learn more about how you can make a difference.
Living with PBC can be challenging. There may be some days when you feel good and don’t experience any symptoms, and there may be other days when you don’t feel up to doing your regular day-to-day activities. But the good news is, there are steps you can take to help manage your symptoms and lifestyle changes you can make to help with your liver health.
Ways to manage fatigue

Fatigue is a common symptom of PBC, although the exact cause is unknown.

There may be days when you feel too tired to complete everyday tasks.
Here are some tips to help you manage fatigue:

Rest
Make time to rest during the day or week, but also before and after a tiring activity
- The duration of rest varies from person to person
- Although rest limits fatigue, prolonged lack of activity can result in additional fatigue

Keep naps short
Napping is great, but napping for too long or too often during the day can disrupt nighttime sleep and increase fatigue that does not give way to rest

Pace your activities
Don’t do too much at one time. Taking breaks between activities can help preserve your energy

Try daily aerobic exercises
Low-impact activities such as walking or swimming can help you increase your fitness without becoming exhausted

Adjust your hobbies or daily activities to match your energy level
Don’t force yourself to do anything that you don’t feel physically able to do

Relax
Learn to relax with activities that provide a sense of well-being and pleasure (massage, drawing, hands-on or creative activities, gardening, etc.) and therapies that allow you to let go (relaxation, meditations, etc.)

Talk to your friends and family
Help them understand what you are experiencing and how they can help
Ways to deal with itchy skin

Pruritus, or itchiness of the skin, is another common symptom of PBC. It may be a result of the accumulation of bile acids that have not been eliminated by the body.

The following are some tips to manage pruritus:

<table>
<thead>
<tr>
<th>PERSONAL HYGIENE</th>
<th>WHAT YOU SHOULD TRY</th>
<th>WHAT YOU SHOULD AVOID</th>
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<tbody>
<tr>
<td></td>
<td>✓ Use skin moisturizer and oatmeal extract to improve dry and inflamed skin</td>
<td>✓ Heat, including hot showers and baths which may dry out your skin</td>
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<tr>
<td></td>
<td>✓ Use mild, hypoallergenic, dye-free, fragrance-free nonalkaline soaps and lukewarm water</td>
<td>✓ Excessive washing/bathing</td>
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<tr>
<td></td>
<td>✓ Hydrate your skin immediately after a bath or shower</td>
<td>✓ Rubbing your skin</td>
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<tr>
<td></td>
<td>✓ Cut your nails short to reduce scratching lesions</td>
<td>✓ Products that irritate the skin such as alcohol-based creams or soaps</td>
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<tr>
<td></td>
<td>✓ Train yourself to stop the cycle of itch – try to find a distraction to forget about the itching</td>
<td>✓ Deodorant soaps ○ If too strong, they can irritate the skin and aggravate itching</td>
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<tr>
<th>DRY CONDITIONS</th>
<th>WHAT YOU SHOULD TRY</th>
<th>WHAT YOU SHOULD AVOID</th>
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<tbody>
<tr>
<td></td>
<td>✓ Use wet, cold, or moist wraps or a compress and apply to the area of the skin that is irritated</td>
<td>✓ Dry air</td>
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<tr>
<td></td>
<td>✓ Use a humidifier in the house if the air is dry</td>
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<tr>
<th>CLOTHING</th>
<th>WHAT YOU SHOULD TRY</th>
<th>WHAT YOU SHOULD AVOID</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>✓ Wear gloves when cleaning and doing housework to protect your hands from bleach, dish soap and other cleaning products</td>
<td>✓ Clothing that rubs the skin</td>
</tr>
<tr>
<td></td>
<td>✓ Wear loose clothing made of natural fibres to avoid irritation from friction ○ Cotton is the softest and coolest fabric to wear</td>
<td>✓ Overly scented detergents</td>
</tr>
<tr>
<td></td>
<td>✓ Hydrate your skin immediately after a bath or shower</td>
<td>✓ Wearing woolen or tight clothing</td>
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<tr>
<th>FRAME OF MIND</th>
<th>WHAT YOU SHOULD TRY</th>
<th>WHAT YOU SHOULD AVOID</th>
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<tbody>
<tr>
<td></td>
<td>✓ Use relaxation techniques</td>
<td>✓ Stress and anxiety ○ If you suffer from chronic stress or anxiety, talk to your doctor for strategies to help you</td>
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<tr>
<th>DIET</th>
<th>WHAT YOU SHOULD TRY</th>
<th>WHAT YOU SHOULD AVOID</th>
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<tbody>
<tr>
<td></td>
<td>✓ Ensuring you have adequate water intake</td>
<td>✓ Spicy food and stimulants (tobacco, alcohol, coffee, etc.)</td>
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</tbody>
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People with PBC may experience dry eyes, a dry mouth or swallowing problems. These are called “sicca syndrome.” It may be helpful to use artificial tears or saliva if you experience symptoms. Good oral hygiene is also important, such as regular brushing, flossing and visits to the dentist.
A healthy diet

Proper nutrition is important for your overall liver health. Remember the basics:

• Eat well: Make sure you include fruits and vegetables, whole grains/cereals and protein sources in your diet
• Minimize salty, sugary and fatty foods
• Avoid alcohol: Alcohol can cause damage to the liver. It is especially important for people with liver disease to avoid drinking alcohol
• Get your calcium and vitamin D: PBC may contribute to bones becoming thinner and more brittle. It is important to ensure that you eat foods that provide calcium (such as tofu, almonds and leafy green vegetables). Vitamin D is also important for bone health
• Drink plenty of water

Smoking is bad for the liver. People with PBC who smoke often have more liver scarring than those who don’t smoke.

Talk to your doctor if you need help to quit.
The effects of PBC vary from person to person, but many people lead active lives with few symptoms for 10 to 20 years. Keep in mind that PBC usually progresses slowly and can be controlled in most people.

It is also important to keep in mind that you are not alone. You can find support not only through your healthcare team, but also through friends and family. Don’t be afraid to ask for help or to accept help from those who are part of your support team, especially on days when you’re running low on energy.

You can also talk to your doctor about ways to manage symptoms so that you can stay as active as possible.

Getting a helping hand

For more information and to hear stories from other people with PBC, visit:

**The Canadian PBC Society**
[pbc-society.ca](http://pbc-society.ca)
Phone number: 1-866-441-3643

**The Canadian Liver Foundation**
[https://www.liver.ca/patients-caregivers/liver-diseases/primary-biliary-cholangitis/](https://www.liver.ca/patients-caregivers/liver-diseases/primary-biliary-cholangitis/)
Phone number: 1-800-563-5483
Important information and questions for my doctor
How to use this ALP tracker

Find the measurement that corresponds with your results and place a dot in line with that measurement above the corresponding date.

Fill in the date you receive your ALP results.

Over time, as you receive more results, you can connect the dots to identify any trends in your ALP levels.

Talk to your doctor about your ALP level at each visit.
Talk to your doctor about your bilirubin level at each visit.

Over time, as you receive more results, you can connect the dots to identify any trends in your bilirubin levels.

Bilirubin is a product of the breakdown of a substance called ‘heme’ found in red blood cells. It is usually excreted by the liver. There is usually a small amount of bilirubin in the blood, however, conditions such as liver dysfunction may result in an increase in the level of bilirubin in the blood.

How to use this bilirubin tracker

Find the measurement that corresponds with your results and place a dot in line with that measurement above the corresponding date.

Fill in the date you receive your bilirubin results.

Date: 03/03/2017