Your journey with primary biliary cholangitis (PBC)

Intercept

Understanding PBC

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.

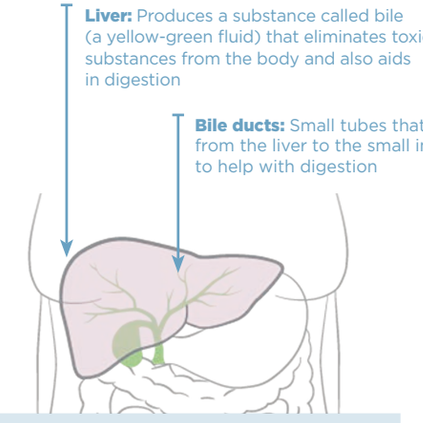
What's going on inside the body?

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.

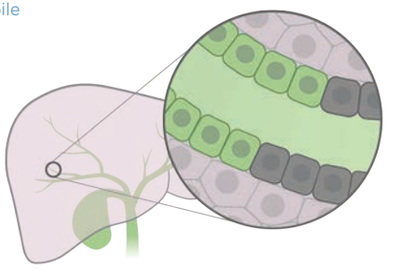
The liver is an important organ that has many functions, including:

- **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**

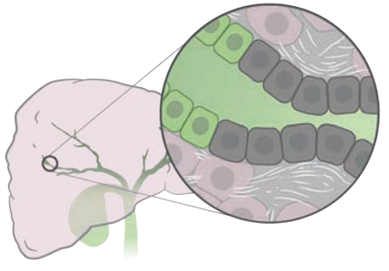
Understanding the progression of PBC



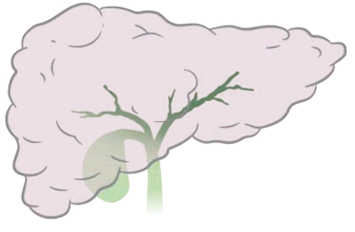
Autoimmune response: In PBC, the body's own immune system mistakes your bile duct cells as foreign objects and attacks them.



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.

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

How did this happen?

Your PBC wasn't caused by anything that you did.

The truth is, the cause or causes of PBC are currently unknown. It is thought that a combination of genetic and environmental factors can increase one's risk of developing PBC.

The following lists some of these risk factors:

POTENTIAL BIOLOGICAL RISK FACTORS	POTENTIAL ENVIRONMENTAL RISK FACTORS
• **Gender:** More common in women than men • **Age:** Most likely to occur in people between the ages of 40–60 years • **Family history of PBC** • **History of other autoimmune disorders:** More common in people who have other autoimmune conditions such as rheumatoid arthritis	• **Infection** • **Smoking** • **Environmental toxins**

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.**

How is PBC diagnosed?

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 progression varies from person to person

PBC is a chronic (long-lasting) disease, but don't let that discourage you.

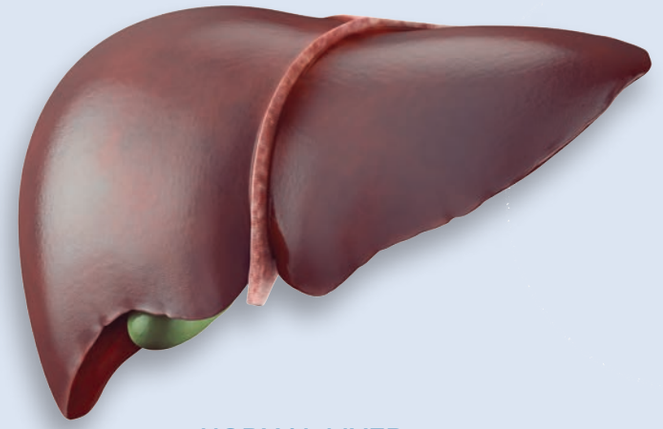
Although it can't be cured, PBC usually progresses slowly and can be controlled in most people. In fact, for many, the disease may remain silent (no symptoms) and may never become serious.

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

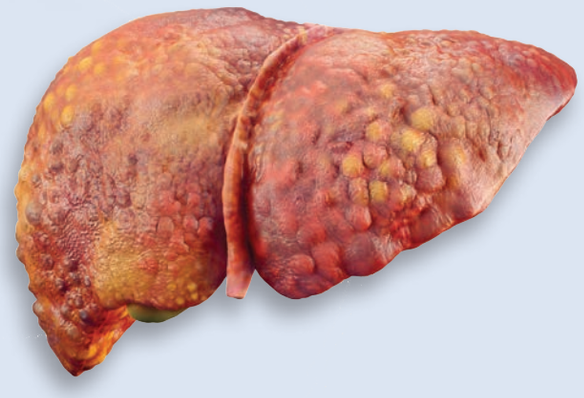
The severity of liver damage and function can be classified using a staging system. Staging provides information about the severity and location of the damage to the liver.

There are four stages of PBC:

STAGE	WHAT IT MEANS
1	Inflammation and/or abnormal liver tissue localized in one area
2	Inflammation and/or scarring localized in one or two areas of the liver
3	Liver scarring progression
4	Severe scarring: cirrhosis



NORMAL LIVER



LIVER WITH CIRRHOSIS

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?

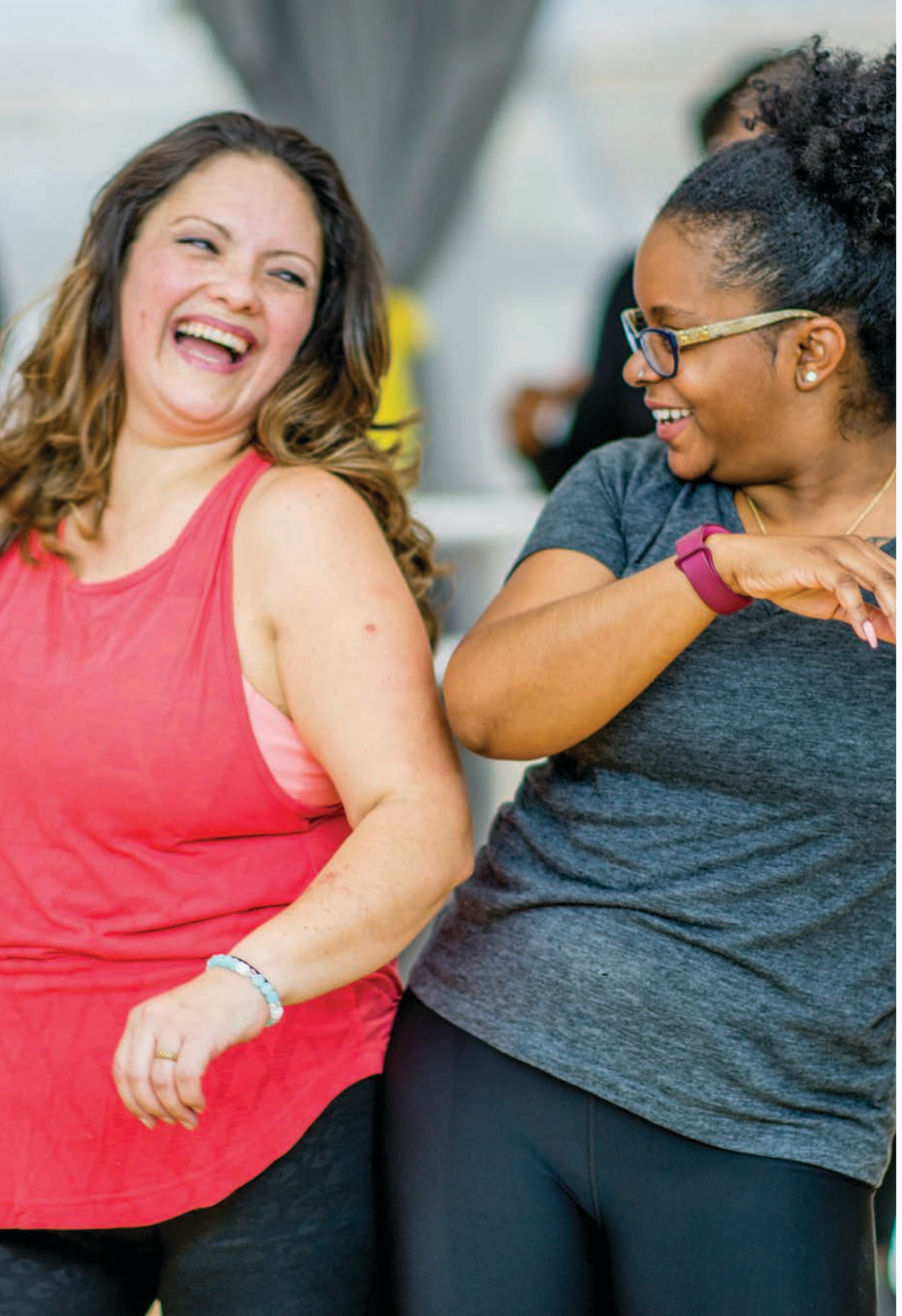
ALP operates as a disease barometer for PBC – Continually elevated ALP levels can mean that the disease is progressing and there is an increased risk of needing a liver transplant.

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

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.

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.



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

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

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

Take care of yourself

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:

PERSONAL HYGIENE

WHAT YOU SHOULD TRY

- ✔ Use skin moisturizer and oatmeal extract to improve dry and inflamed skin
- ✔ Use mild, hypoallergenic, dye-free, fragrance-free nonalkaline soaps and lukewarm water
- ✔ Hydrate your skin immediately after a bath or shower
- ✔ Cut your nails short to reduce scratching lesions
- ✔ Train yourself to stop the cycle of itch – try to find a distraction to forget about the itching

WHAT YOU SHOULD AVOID

- ✘ Heat, including hot showers and baths which may dry out your skin
- ✘ Excessive washing/bathing
- ✘ Rubbing your skin
- ✘ Products that irritate the skin such as alcohol-based creams or soaps
- ✘ Deodorant soaps
  - If too strong, they can irritate the skin and aggravate itching

DRY CONDITIONS

WHAT YOU SHOULD TRY

- ✔ Use wet, cold, or moist wraps or a compress and apply to the area of the skin that is irritated
- ✔ Use a humidifier in the house if the air is dry

WHAT YOU SHOULD AVOID

- ✘ Dry air

CLOTHING

WHAT YOU SHOULD TRY

- ✔ Wear gloves when cleaning and doing housework to protect your hands from bleach, dish soap and other cleaning products
- ✔ Wear loose clothing made of natural fibres to avoid irritation from friction
  - Cotton is the softest and coolest fabric to wear

WHAT YOU SHOULD AVOID

- ✘ Clothing that rubs the skin
- ✘ Overly scented detergents
- ✘ Wearing woolen or tight clothing

FRAME OF MIND

WHAT YOU SHOULD TRY

- ✔ Use relaxation techniques

WHAT YOU SHOULD AVOID

- ✘ Stress and anxiety
  - If you suffer from chronic stress or anxiety, talk to your doctor for strategies to help you

DIET

WHAT YOU SHOULD TRY

- ✔ Ensuring you have adequate water intake

WHAT YOU SHOULD AVOID

- ✘ Spicy food and stimulants (tobacco, alcohol, coffee, etc.)

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.

Living well with PBC

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
Phone number: 1-866-441-3643

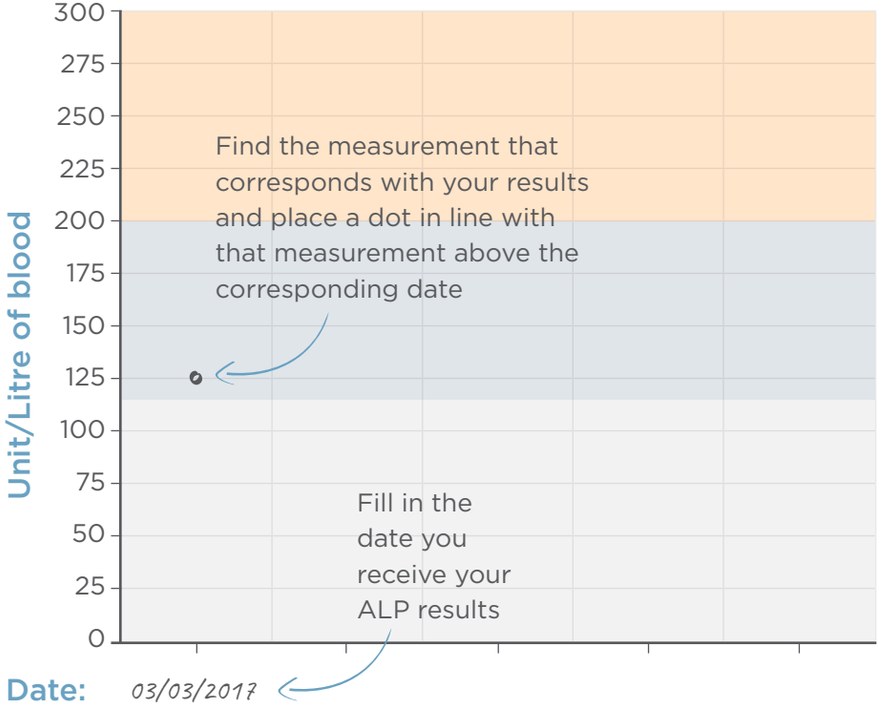
The Canadian Liver Foundation
https://www.liver.ca/patients-caregivers/liver-diseases/primary-biliary-cholangitis/
Phone number: 1-800-563-5483

Notes

Important information and questions for my doctor

Your ALP tracker

How to use this ALP tracker



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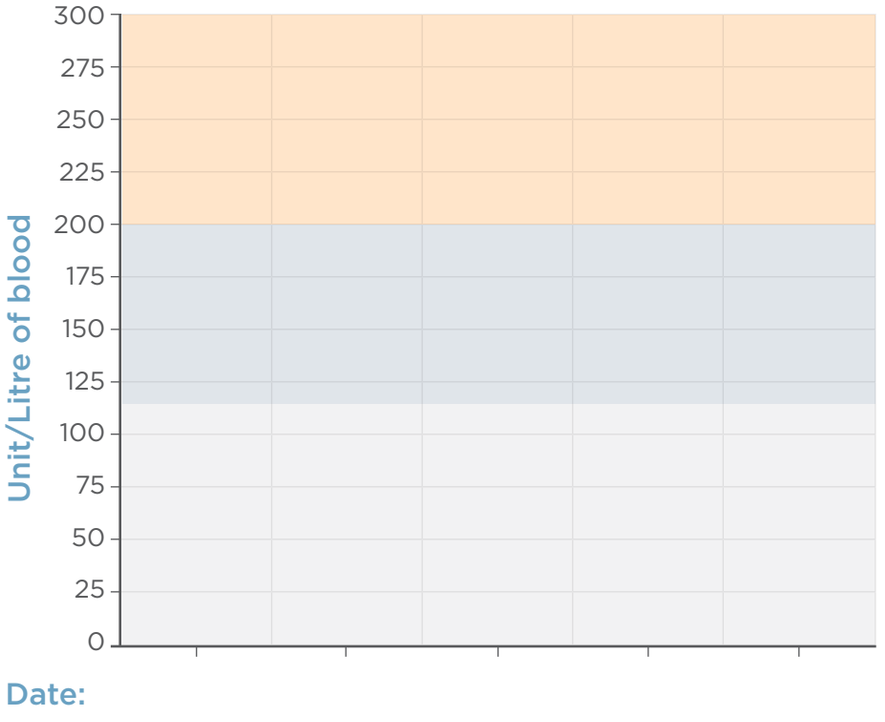
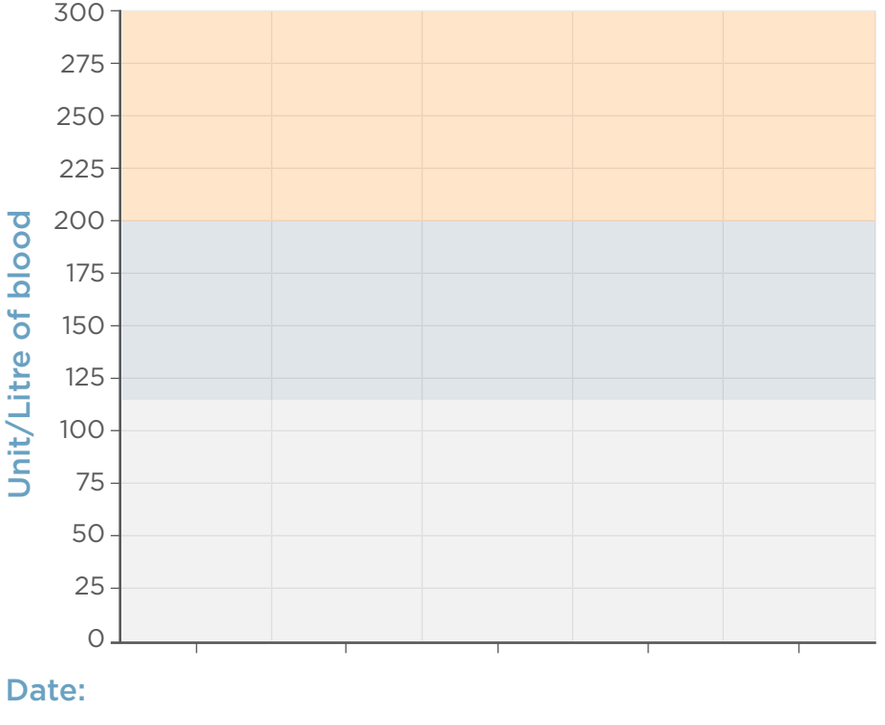
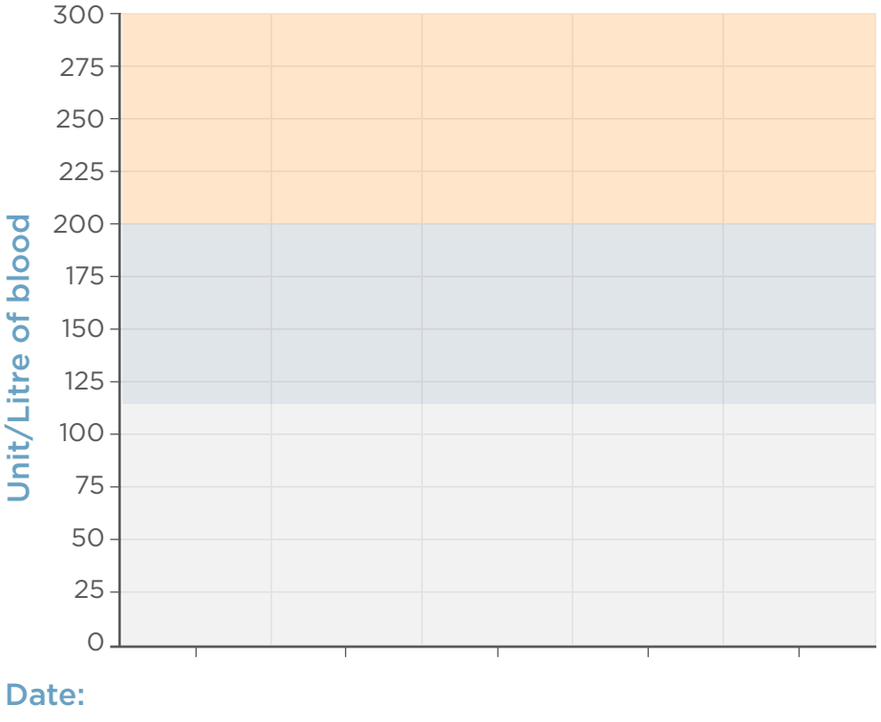
Talk to your doctor about your ALP level at each visit.







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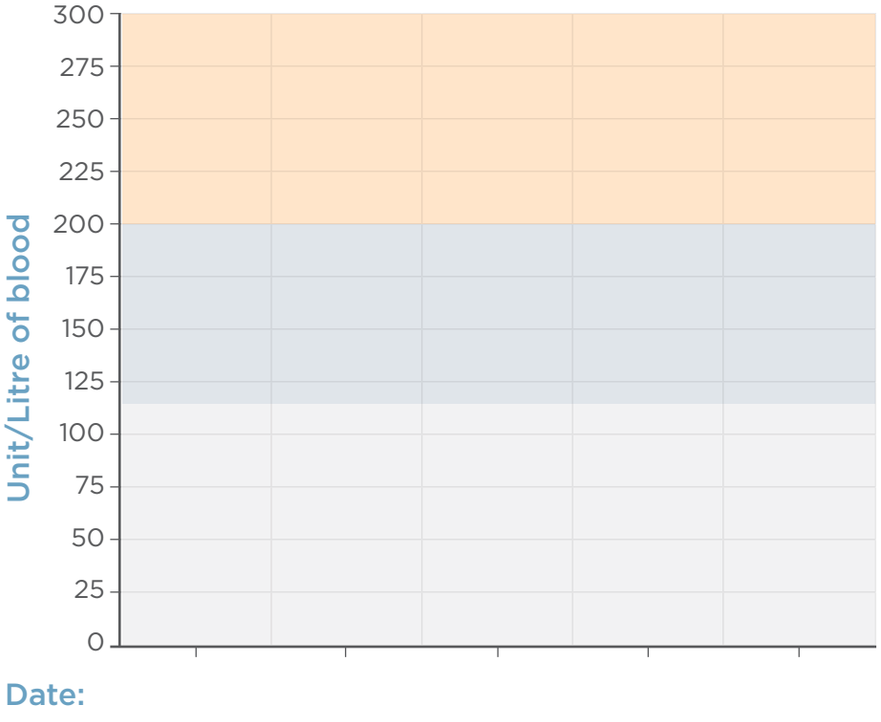

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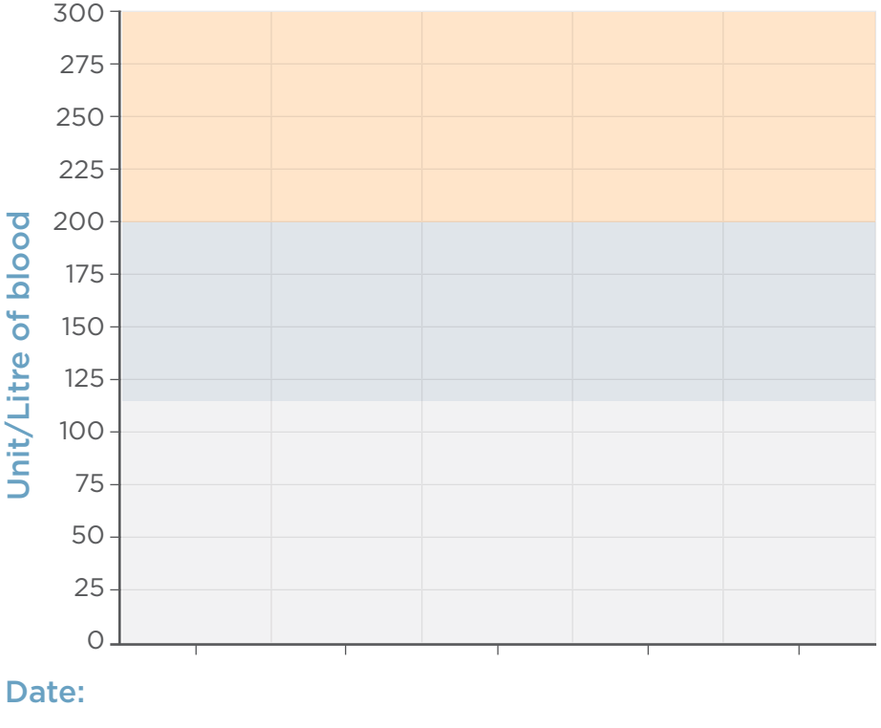

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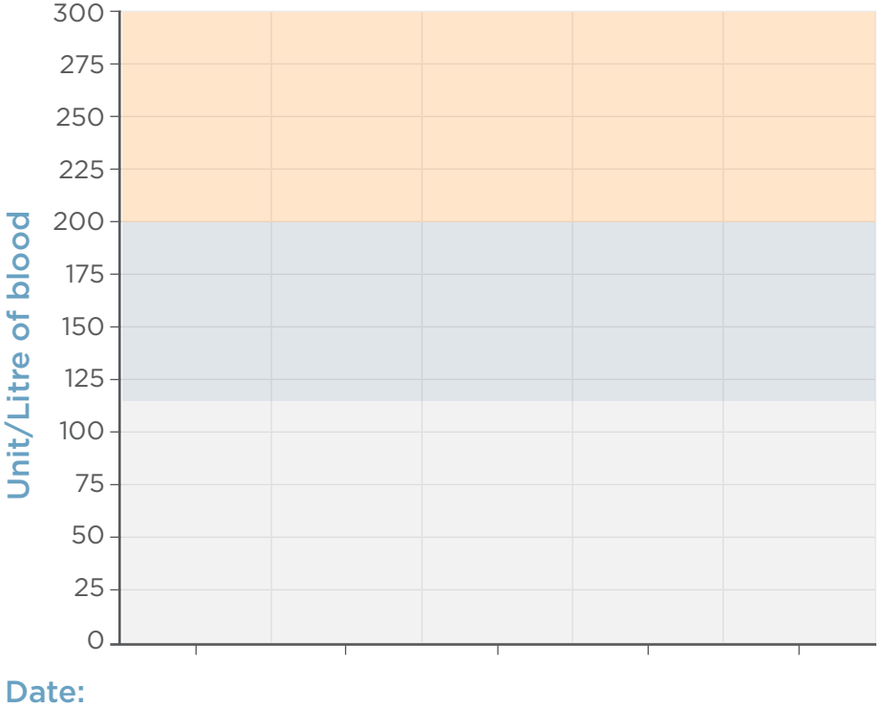

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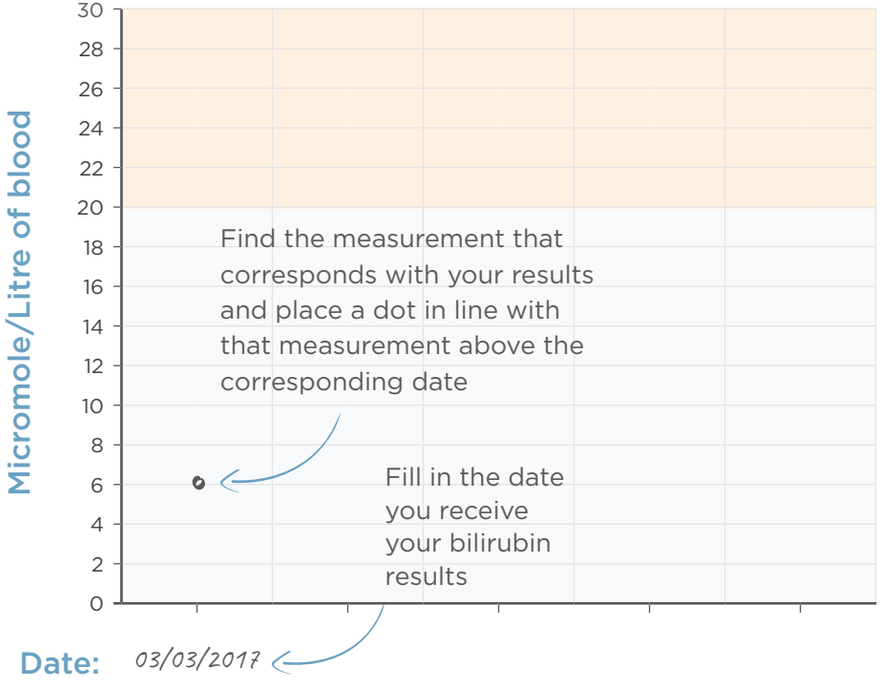
Your bilirubin tracker

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



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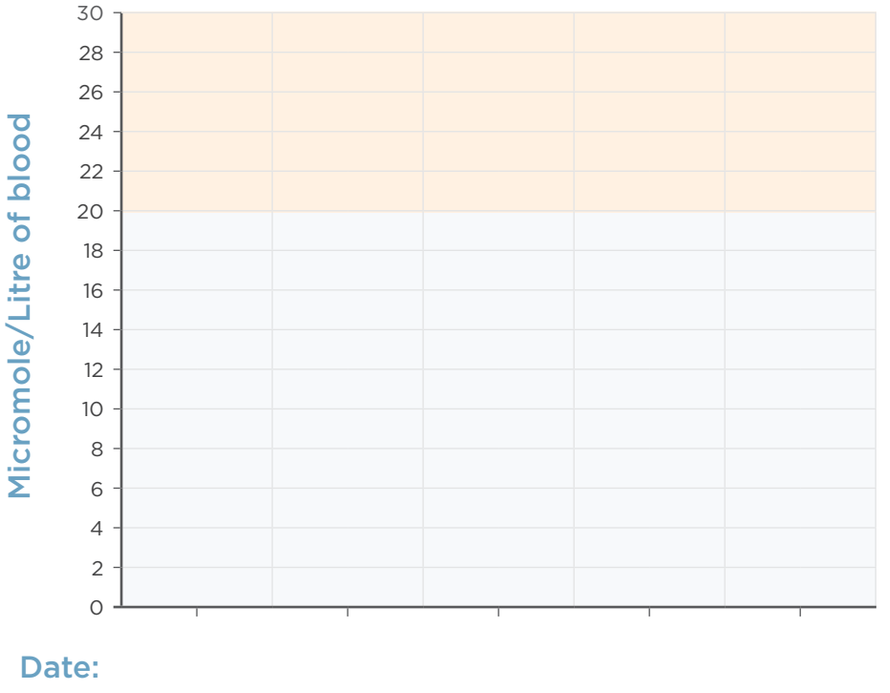
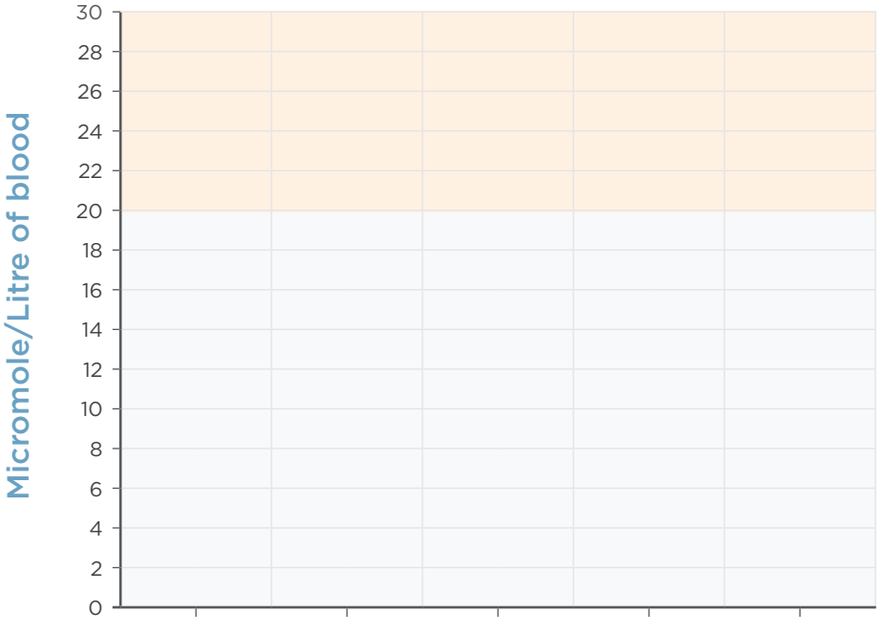
Talk to your doctor about your bilirubin level at each visit.





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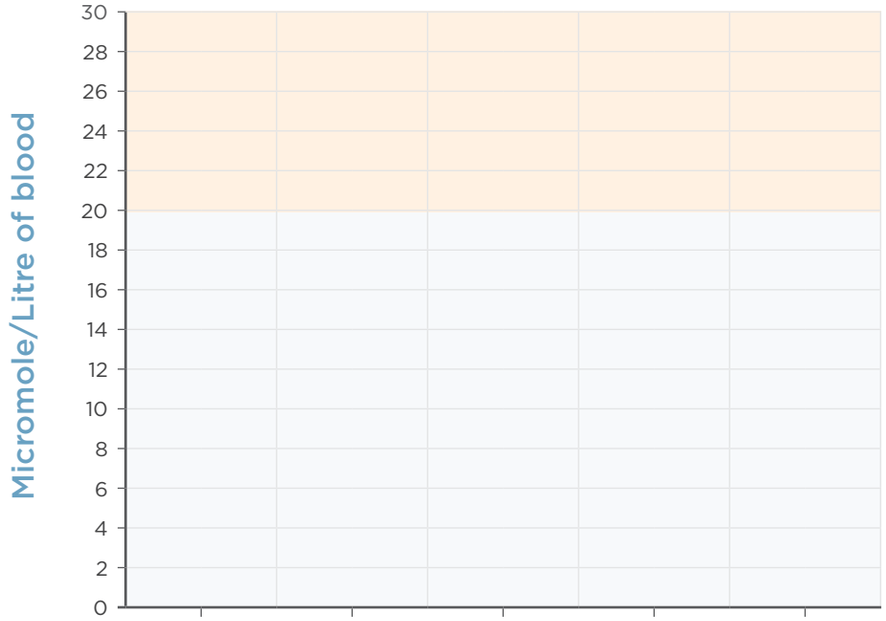

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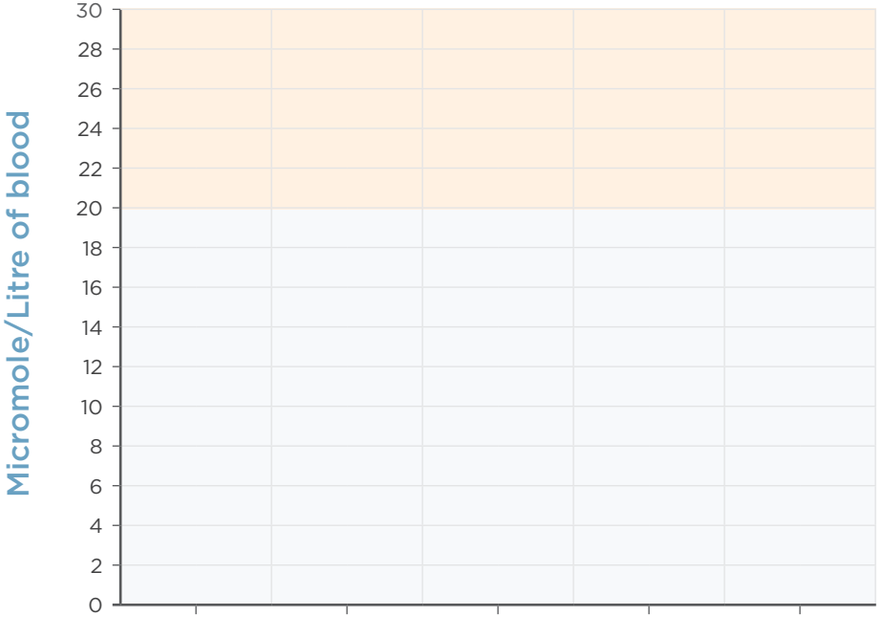

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