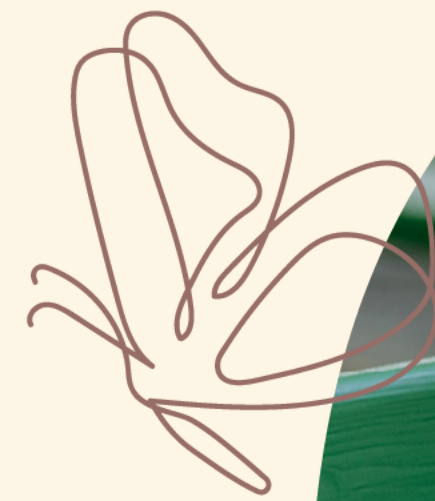


Understanding primary biliary cholangitis (PBC)

For people living with PBC



This leaflet has been given to you as you have been prescribed IQIRVO® (elafibranor) by your doctor.

Contents

Understanding PBC.....3

What happens in the liver?4

Who does PBC affect?5

How is PBC diagnosed?.....6

How is PBC managed?.....7

Risk assessment.....8

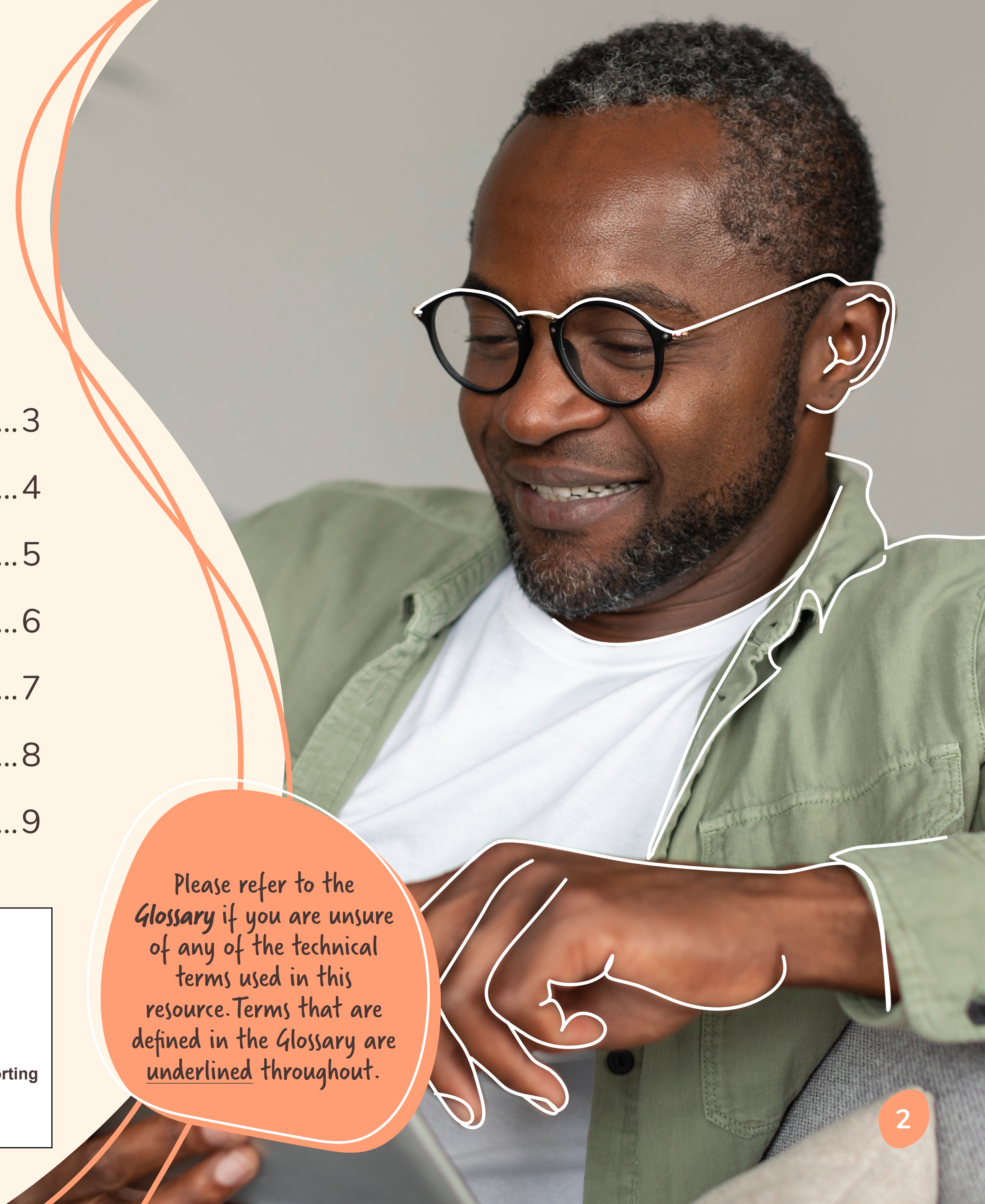
References.....9

Reporting of side effects: If you experience any side effects, talk to your doctor, pharmacist or nurse. This includes any possible side effects not listed in the package leaflet.

You can also report side effects directly via the Yellow Card Scheme at www.mhra.gov.uk/yellowcard/

By reporting side effects, you can help provide more information on the safety of this medicine. IQIRVO® is subject to additional monitoring. This will allow quick identification of new safety information. You can help by reporting any side effects you may experience. See www.mhra.gov.uk/yellowcard/ or how to report side effects.

Please refer to the Glossary if you are unsure of any of the technical terms used in this resource. Terms that are defined in the Glossary are underlined throughout.



Understanding PBC

Since your diagnosis of primary biliary cholangitis, or ‘P B C’, you might have questions or concerns about the condition, and how it could affect your day-to-day life.

This resource will help you to better understand PBC, including what causes it, how it might make you feel, and how PBC can progress over time.

What is PBC?

PBC is a fairly rare liver condition that can progress over time and lead to scarring of the liver (**fibrosis**), more advanced liver scarring (**cirrhosis**), and liver failure if left untreated.¹ Fortunately, there are treatments for PBC that can help how your liver works and help with your symptoms (and will be mentioned in more detail in further sections).

PBC is known as an **autoimmune disease**, meaning that the immune system (the body’s defence mechanism against illness) mistakes healthy cells, tissues, or organs for something dangerous for the body.^{1,2} As a result, the immune system falsely attacks and destroys a healthy part of the body, which can impair its ability to function correctly. In PBC, this happens in the liver — this is explained further below.²

A diagnosis of PBC can be a lonely and challenging time. Please explore our *Supporting you through your journey* guide for more support.



PBC fact

PBC was previously referred to as **primary biliary cirrhosis**, leading to the misconception that it could be caused by drinking alcohol. This is **not true**, and PBC is believed to be influenced by **multiple factors**. The new name, primary biliary **cholangitis**, helps to avoid the confusion.³

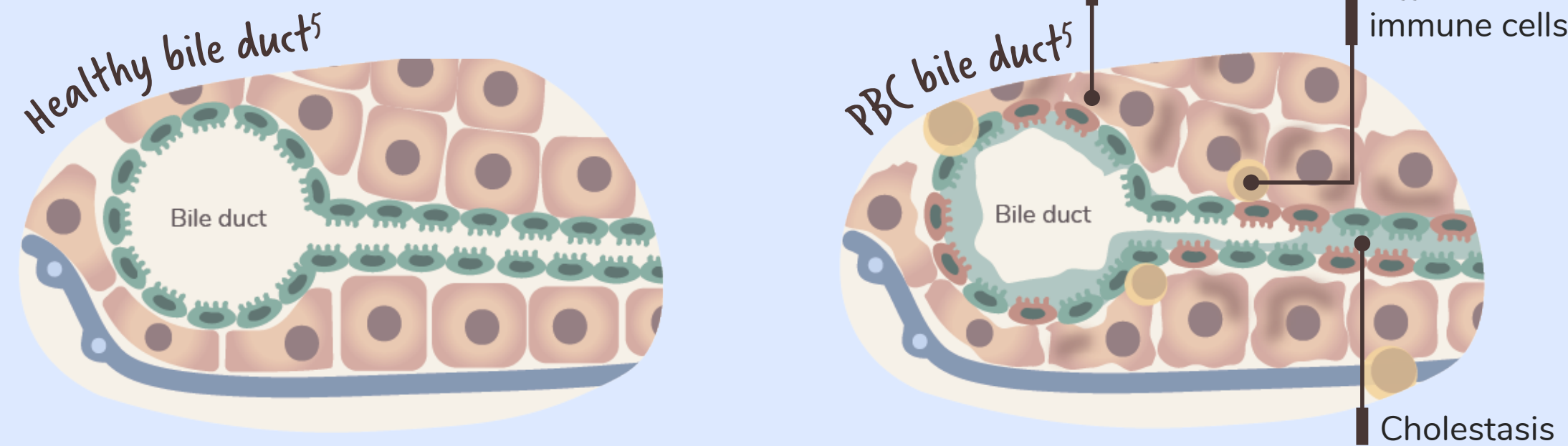


What happens in the liver?

The liver produces a liquid called **bile**, which helps us break down fats and eliminate waste products. To do so, bile must travel from the liver to other parts of the body, such as the small intestine, and is transported through structures called **bile ducts**.⁴

In people with PBC, their immune system mistakes bile ducts **in the liver** as harmful and begins to attack them.² When bile ducts get destroyed, this prevents the flow of bile out of the liver, leading to a condition known as **cholestasis**.⁴

These pictures show a liver bile duct up close



The left-hand image shows a healthy bile duct that allows bile to flow through and out of the liver to where it's needed. On the right-hand side, you can see how a damaged duct restricts the flow of bile.

As a result, bile and other substances build up in the liver, causing inflammation and further liver damage.² If PBC is left untreated, this can lead to liver fibrosis and, eventually, **cirrhosis**.^{1,2}



Who does PBC affect?

PBC is ten times more common in women than in men, and is usually diagnosed in people between the ages of 40 and 60.^{1,2}

Although there is a suggested link between having a family member with PBC and developing PBC yourself, it isn't considered a hereditary condition, meaning that there is no guarantee that PBC will be passed on to your children.⁶

The causes of PBC are not fully understood, but people with a history of smoking or those who experience recurrent urinary tract infections may be more likely to develop PBC.⁶ Some research has proposed that living in a polluted area might make you more susceptible to PBC.⁷

What are the main symptoms of PBC?

Symptoms of PBC can vary, and some people won't have any symptoms when they are diagnosed.^{1,2} Some common symptoms of PBC that you may experience, either now or in the future, include extreme tiredness (**fatigue**), itchy skin (**pruritus**), and impaired memory (**brain fog**).^{2,8}

For more information about the symptoms of PBC and recommendations on how to manage them, please refer to the **PBC symptom guide**.



How is PBC diagnosed?

PBC is usually diagnosed using a blood test, where a medical professional will look for a specific **protein** in the blood called **antimitochondrial antibody (AMA)**, which is linked to **PBC**. They will also investigate whether levels of liver **enzymes**, such as **alkaline phosphatase (ALP)**, in the blood are higher than usual.²

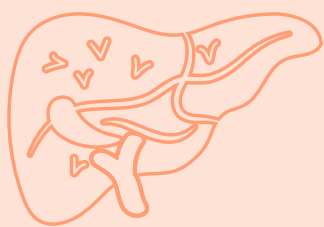
If a person’s blood test suggests PBC, they will need to have their liver looked at using imaging techniques such as ultrasound or magnetic resonance imaging (MRI). This will help confirm a PBC diagnosis and show how far along the condition is.^{2,9}

How does PBC progress over time?

In the initial stages of PBC, you may not notice anything different, but complications can develop as the condition progresses.² PBC generally progresses slowly, but the exact rate differs between people — some people may go decades without any progression, while others may experience progression within a few years.¹ No matter how fast or slow PBC progresses, medication can be used to slow it down and potentially limit its impact.¹

Progression of PBC¹⁰

1



The immune system begins to attack the bile ducts

2



Bile ducts become damaged, leading to cholestasis and the build up of bile

3



Cholestasis causes liver fibrosis

4



Liver fibrosis can progress to liver cirrhosis and end-stage liver disease



How is PBC managed?

The main goals of treatment for PBC are to **slow the damage** it causes to the **liver** and to **potentially reduce the impact of PBC symptoms**.^{1,2} It is important to note that the severity of your symptoms may not be reflective of the extent of liver damage caused by PBC, and there are different approaches for **managing liver disease progression and controlling symptoms**.^{2,9}

It is very important to bear in mind that, even though you may not feel any different after taking a PBC medication, research has shown that there is a positive long-term effect of treatment on liver health. Treatment can significantly delay the progression of liver scarring in people with PBC.¹¹

For these reasons, it's important to keep taking your medication unless your doctor tells you otherwise. Contact your doctor or someone from your healthcare team as soon as possible if your PBC medication is making you feel worse.

Managing liver disease progression

Certain medications can help to delay the progression of liver damage associated with PBC.^{1,2} A medicine called ursodeoxycholic acid, also called **UDCA** or **URSO**, is used first in people with a PBC diagnosis.^{1,2} UDCA facilitates the removal of bile from the liver and can slow down liver deterioration but usually does not improve the common symptoms of PBC.¹²

UDCA may not work for everyone, and some people experience side effects. In these cases, a second treatment can be offered in combination with, or in place of, UDCA.^{2,13}

PBC progresses differently in each person. While some people may experience a slow or no worsening of the disease, others might develop advanced liver disease more quickly. Researchers have identified a number of factors that are associated with how quickly PBC progresses:

How severe PBC was at diagnosis

It is easier to prevent PBC from progressing and causing damage to your liver the earlier it is diagnosed and treated. Unfortunately, not everybody is diagnosed with PBC early on. People with high levels of ALP and/or liver stiffness at diagnosis are more likely to be in the later stages of PBC and are at higher risk of severe disease.^{13,16}

Your age at diagnosis

People diagnosed with PBC that are under 45 years old are at an increased risk of PBC progression and a poorer response to treatment.¹³⁻¹⁵ The exact reason for this is unknown, but your doctor will still use this information to guide your treatment.

How well you respond to treatment

It is recommended that you try treatment for a period of time before being checked on how well it works.¹³ People who do not have a significant improvement in their liver function test (such as decreases in ALP or bilirubin) are at high risk of disease progression.¹³

Risk assessment

Your doctor can evaluate PBC on an individual basis using what's known as a '**risk assessment**'. Ask a member of your medical team for more information on the methods used for risk assessment in PBC.¹⁷

Having an increased risk of progression does not guarantee that it will happen. Understanding and measuring factors that put you at higher risk allows your doctor to create a personalised treatment plan.¹⁷

This guide lists risk factors to make sure that you are informed, but **only your doctor can advise on your risk profile**. If you are worried about any of these risk factors, please speak with your doctor.



Living with PBC can be challenging, and there's lots of information to take in. If you have any further questions about PBC, please talk to your doctor or another member of your healthcare team.



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For more information about Elafibranor, speak to your doctor.

