

Understanding your cholangiocarcinoma

If you or a loved one has recently been diagnosed with cholangiocarcinoma (pronounced ko-LAN-jee-o-car-sin-O-ma), also called bile duct cancer or CCA for short, you probably have a lot of questions.

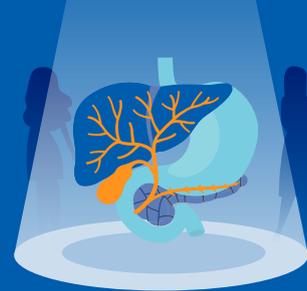
This booklet aims to answer some of the initial questions you may have about your cholangiocarcinoma. You should use this booklet as a prompt to think about the types of questions you could ask your doctor to help you make treatment decisions at your next appointment.

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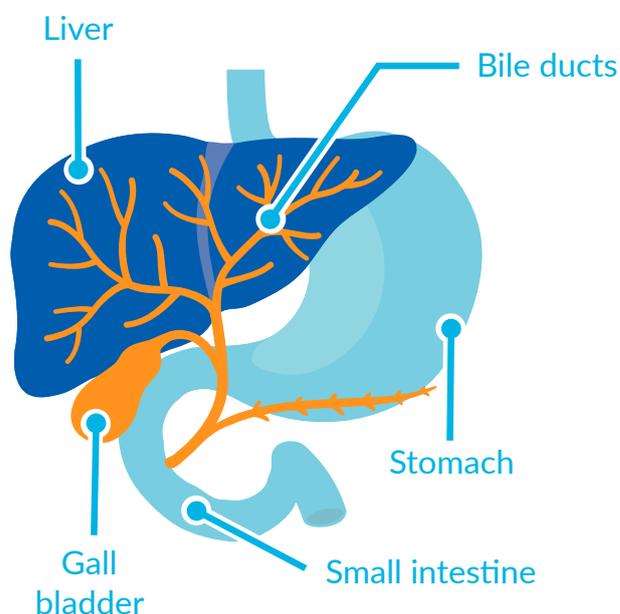
What is cholangiocarcinoma?



Cholangiocarcinoma is a rare type of liver cancer that develops in the bile ducts. Bile ducts are thin tubes that connect your liver, gallbladder, and small intestine. Their main job is to transport the digestive fluid known as bile, which is made in your liver and helps break down fats.^{1,2}

There are different types of cholangiocarcinoma, depending on where in the bile duct they develop. These are **intrahepatic** and **extrahepatic** cholangiocarcinoma.³

- Intrahepatic mean the cancer affects the bile ducts **inside** your liver³
- Extrahepatic means the cancer affects the bile ducts **outside** your liver, near the intestine³
- There are two forms of extrahepatic cholangiocarcinoma. These are called perihilar and distal cholangiocarcinoma³



The type of cholangiocarcinoma you have may determine how your doctor will treat your cancer.

How common is cholangiocarcinoma?

Overall, cholangiocarcinoma is not common and is considered to be a rare cancer; however, there are treatments available and specialists who can help. The number of people diagnosed with cholangiocarcinoma varies between countries.⁴ In most countries, for every 100,000 people in the population, less than 6 people will be diagnosed with cholangiocarcinoma.⁴ The number of cases in Europe and North America has increased in recent decades.^{3,5} Cholangiocarcinoma most often occurs in people who are over the age of 50 and is slightly more common in women than in men.^{3,5}

Are there any risk factors that may mean it is more likely to get cholangiocarcinoma?



Most cases of cholangiocarcinoma occur without any apparent cause. However, there are some risk factors, such as:^{3,5,6}

-  Age (cholangiocarcinoma is more common if you are over 65 years old)
-  Type 2 diabetes
-  High blood pressure (also known as hypertension)
-  Being overweight or obese
-  Tobacco and alcohol use
-  Exposure to certain chemicals (eg, those involved in production at a print factory)
-  Pre-existing liver conditions (such as primary sclerosing cholangitis or hepatitis)
-  Certain infections of the liver, such as those caused by parasites which are common in South East Asian countries



Having a risk factor does not necessarily mean it was the cause of your cholangiocarcinoma, and not all risk factors have the same effect. Some people will get cholangiocarcinoma without any of the known risk factors. Speak to your doctor if you have any concerns at all.

How is cholangiocarcinoma usually diagnosed?



Patients with cholangiocarcinoma may not always show symptoms until the later stages of the disease and the symptoms they do have can be general, such as nausea.^{4,5}

This makes the diagnosis of cholangiocarcinoma difficult, and it often requires the combined efforts of different types of experts. These experts are part of your multidisciplinary team (or MDT) and will be involved in making decisions about your treatment and care.^{4,5}

A sample of your tumour (called a biopsy) is often required to confirm the diagnosis of cholangiocarcinoma. Your doctor will also need to perform other tests such as blood tests, imaging scans (scans that can take images inside your body), and an assessment of your symptoms during a clinical examination.^{4,5,7}

Multidisciplinary teams



A **multidisciplinary team**, or MDT, is a group of experts, such as surgeons, gastroenterologists, radiologists, oncologists, pathologists, and nurses, who combine their expertise to provide you with the best possible care.

The MDT may order specific tests and will use the results to decide which treatment options are right for you.

Biopsy



A tissue **biopsy** is a procedure in which a small piece, or sample, of your tumour is taken to be studied. This allows your doctors to learn more about your cholangiocarcinoma, provide a more in-depth diagnosis, and determine the best treatment options for you.

Your biopsy may also be used for molecular profiling. Molecular profiling is a type of test which can identify any specific genomic alterations (changes in your DNA) that are unique to your tumour. More details on molecular profiling can be found on page 6.

Will surgery be an option for me?



Surgery to remove the cholangiocarcinoma may be an option for some patients, if their cancer has not spread to other parts of the body.⁸ This is also known as resection. Your doctor will be able to advise if this could be a suitable option for you.⁸ People who undergo surgery may also receive additional treatment before or after the surgery to help ensure all the cancer is removed.⁹

What are the other treatment options for cholangiocarcinoma?

As well as surgery, other treatment options for patients with cholangiocarcinoma may include radiotherapy, chemotherapy and targeted therapy.^{8,9}

Your doctor may perform tests to determine which treatment option is best for you. Performing blood tests or taking a small sample of your tumour (biopsy) to be used in molecular profiling may inform your doctor that you are eligible for targeted therapy.

Treatment options^{8,9}



Chemotherapy

A treatment using medicines or a combination of medicines that slow or stop the growth of rapidly dividing cells. Chemotherapy is not specific and will target both cancer cells as well as healthy cells.



Radiotherapy (or radiation therapy)

A treatment that uses high-energy X-rays or radiation to kill cancer cells or stop them from growing.



Targeted therapy

A more specific type of drug treatment that only targets the genes and proteins that have caused the cancer. Your doctor can use a technique called molecular profiling to learn more about your cholangiocarcinoma and assess whether targeted therapy is an option for you.



Best supportive care

This includes symptom and pain control management. For example, if the bile ducts are blocked, jaundice, pain, and infections can occur. Biliary draining using a catheter (a thin flexible tube) can help improve blockages and is considered a form of best supportive care.

What is molecular profiling?



Molecular profiling is a type of test used to look for faults or changes in the DNA of cancer cells. The results of molecular profiling can help your healthcare team understand more about what is driving the growth of your cancer and therefore which treatment option is best for you.

The cells in our body are constantly dividing. Sometimes faults can occur and this can lead to changes in the DNA of our cells. When this happens our cells behave differently and cancer can form.

Some of these DNA changes include:¹⁰⁻¹⁵



Mutations – small changes in the gene, which change its function



Gene amplifications – more copies of the gene than normal, which change its activity



Gene fusions – the joining of different genes, which change their function and/or activity



Your healthcare team is responsible for ordering your molecular profiling test. This is performed using a sample of your tumour (biopsy). Your doctor will then receive a report with the results of your molecular profiling, which can help decide the next steps in your treatment plan.

Molecular profiling can be done at any time in your treatment journey. However, the sooner it can be performed the better, as it can provide your doctor with important information to help guide their treatment decisions.^{16,17}

Are there any lifestyle changes I can make to support myself through treatment?



You may find that making lifestyle changes and taking up self-care routines has a positive impact on your treatment journey.

Self-care routines such as light exercise, relaxation, mindfulness, healthy eating and nutritional awareness can be beneficial to improving physical, mental and emotional health.^{18,19} Nutritional status is particularly important and where possible you should try to:²⁰



Eat a balanced diet



Talk to your doctor about nutritional therapy to avoid malnutrition



Maintain a healthy weight



For more information on how to improve nutritional status, you should talk to your doctor or read the accompanying booklet 'Living with cholangiocarcinoma: managing your nutrition needs'.

Or visit:

www.ammf.org.uk/nutrition



Remember, incorporating self-care routines into your lifestyle may enhance your wellbeing, reduce stress and treatment symptoms, and improve day-to-day life; however, these should always be discussed with your doctor beforehand.¹⁸⁻²⁰

Could a clinical trial be an option for me?



You can ask your doctor or a patient organisation if they know of any clinical trials that you may be eligible to join. You can also search for information online and register your interest in taking part in research. If you're eligible, being part of a clinical trial may offer certain benefits:²¹

- Potential access to new and innovative medicines that are being developed but aren't yet approved for doctors to prescribe
- You can help make a difference for other people in the future who have your specific type of cancer



Clinical trials

A **clinical trial** is a type of scientific research study that tests possible new medicines and evaluates their effects on the patients' health. Their purpose is to discover treatments which are more effective, have fewer side effects or improve the patients' quality of life, compared to existing treatments.

However, there are important things to consider for participating in clinical trials:²¹



As can happen with all treatments, the investigational approach being studied may not provide a benefit to you, the participant



As clinical trials investigate safety and side effects, sometimes these are not fully known at the time of study. Therefore, in some rare cases there may be an increased risk of harm



If you are interested in learning more about clinical trials you should speak to your doctor, who will be able to provide you with further advice.

How can I talk to my family and friends about my diagnosis?



Nobody can tell you how best to approach letting your family and friends know about your diagnosis, but there is advice available.

You may find it difficult to talk about, but being open and honest about your cholangiocarcinoma and how you are feeling will help those closest to you better understand what you may be going through.

Reading about cholangiocarcinoma with your family and friends may help reduce anxiety or confusion. In the next section, you will find some useful resources for cholangiocarcinoma. You may also find letting your loved ones read this booklet can help them to better understand cholangiocarcinoma and the journey ahead.



Finally, always remember you can talk to your doctor about anything, including your concerns or frustrations. You should always let them know about any further changes to your physical, emotional or mental health.



What other resources are available?



The 'AMMF – The Cholangiocarcinoma Charity' website provides all the latest news about cholangiocarcinoma and has lots of resources for you to learn more about your disease www.ammf.org.uk



The Penny Brohn UK website includes guides for managing your symptoms and improving your wellbeing www.pennybrohn.org.uk



The European Society for Medical Oncology – Biliary Tract Cancer: Guide for Patients provides further details on cholangiocarcinoma and the treatment journey www.ammf.org.uk/wp-content/uploads/2020/02/EN-Biliary-Tract-Cancer-Guide-for-Patients.pdf



The Cholangiocarcinoma Disease Awareness for Patients website is coming soon and will provide more details on the topics covered in this booklet

Coming soon!

What other resources are available?



The 'Cancer Research UK' website can provide you with the latest information about various cancers as well as tips for coping with your illness www.cancerresearchuk.org



The 'Macmillan Cancer Support' website can provide support to help you with the physical and emotional burdens of cancer. Information on various cancers and their treatments are also provided www.macmillan.org.uk



The 'Cholangiocarcinoma Foundation' website provides lots of useful resources and support for patients with cholangiocarcinoma www.cholangiocarcinoma.org



Use this space to take notes if needed. You can take this booklet with you to your next appointment with your doctor to help discussions about your cholangiocarcinoma.

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