

About Cholangiocarcinoma

Cholangiocarcinoma is a primary liver cancer that occurs within the bile ducts and, for many, it is a challenging diagnosis.

The facts:

- It's the second most common primary liver cancer in the world
- In 2019 it caused 2,754¹ deaths in England alone
- The incidence is increasing, and we don't know why
- It's difficult to diagnose clearly and accurately
- Most are diagnosed too late for potentially curative surgery
- In the western world the cause is generally unknown, most cases are sporadic
- Thailand has the world's highest incidence, caused by eating raw fish infected with liver fluke (*Opisthorchis viverrini*)

At AMMF we are working to:

- Raise awareness of cholangiocarcinoma
- Provide information to those who need it, either individually or via our website
- Encourage and support research, especially into causes, and finding ways to achieve better and clearer early diagnosis

¹ National Cancer Registration and Analysis Service (NCRAS) and NHS Digital

For more information on cholangiocarcinoma, the work of AMMF and ways to donate, see our website:

ammf.org.uk



**AMMF – The UK's only
cholangiocarcinoma charity**

Working across the UK and Europe,
and collaborating internationally to
improve the future for all those with
cholangiocarcinoma

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THE CHOLANGIOCARCINOMA CHARITY

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Registered charity nos 1198095 and 1091915

About AMMF

Founded in 2002, AMMF was the world's first charity dedicated solely to cholangiocarcinoma (CCA). Today we remain the UK's and Europe's only CCA charity.

AMMF works closely throughout the UK with patients and their carers, healthcare professionals, researchers, and policy makers, as well as actively collaborating across Europe, the USA, SE Asia – indeed globally.

Although CCA (bile duct cancer) is considered a rare cancer, worldwide it is the second most common primary cancer arising in the liver. However, because it is so difficult to diagnose accurately and early, CCA is often advanced by the time of diagnosis and so surgery, currently the only potential cure, is not an option.

Over recent years there has been an enormous and extremely worrying worldwide increase in the incidence of CCA. In 2019 it was the cause of 2,754¹ deaths in England alone, and the incidence appears to be increasing across all age groups, including younger people. The cause of this ongoing increase is currently unknown, and much more research is needed.

AMMF is dedicated to raising awareness, providing information to those who need it, either individually or via our informative UK and European websites, and supporting research into the causes, better diagnostic tests and more effective treatments for this challenging disease.

AMMF's work is totally CCA focused, and improving the future for those with this challenging cancer is always at the heart of everything we do.

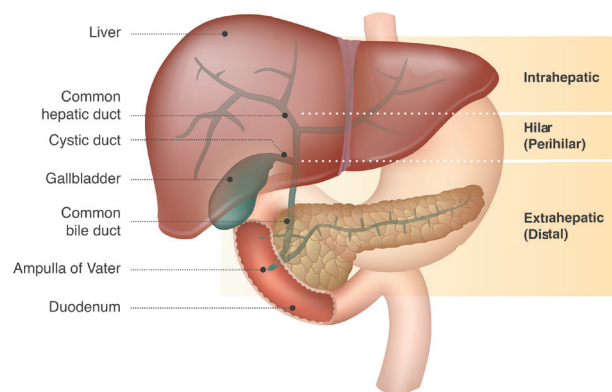
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What is cholangiocarcinoma?

Cholangiocarcinoma (CCA) is also known as bile duct cancer. This is a cancer that occurs in the bile ducts in or around the liver. Diagnoses fall into three main categories:

- **Intrahepatic CCA** – affects bile ducts within the liver.
- **Perihilar (or Hilar) CCA** – originates just outside the liver, where the left and right hepatic ducts join together.
- **Extrahepatic/Distal CCA** – originates anywhere in the common bile duct, which goes from the liver to the small intestine.



What are the risk factors?

In the western world the exact cause of CCA is unclear – most cases are sporadic. Established risk factors, including some illnesses that cause chronic damage to the liver and/or bile ducts, and possibly a small genetic predisposition, are thought to account for less than 30% of all cases. Primary Sclerosing Cholangitis (PSC) is the commonest known predisposing cause.

Other recognised risk factors include gallstones which have remained in the bile ducts for a long time; cysts in the bile ducts; and exposure to certain toxins. Newly discovered likely risk factors include obesity, diabetes and fatty liver disease. However, most people with CCA have none of these risk factors, so studies into the causes are a vital area of research.

What are the symptoms?

Cholangiocarcinoma (CCA) starts in the bile ducts inside or outside the liver, so commonest symptoms may be caused by blocked bile flow – jaundice with yellowing of eyes and skin, dark urine or pale stools and sometimes itching. Other possible symptoms include pain or ache under the right ribs, indigestion, unexplained weight loss, generally feeling unwell, and tiredness. Whilst several of these symptoms are non-specific and could be caused by benign conditions, they do require urgent assessment, especially if jaundice is present.

What treatments are available?

Surgery to totally remove the cancer is currently the only potentially curative treatment. Unfortunately, by the time most patients are diagnosed, their CCA is too advanced for surgery.

If surgery is not an option, there are several other types of treatment which, although not curative, can help control symptoms and may delay the cancer's progression. The gemcitabine/cisplatin chemotherapy combination has been standard of care for some time for those with inoperable CCA, but clinical trials looking for improvements have continued and are helping to find other treatments, including chemotherapy, radiotherapy, immunotherapy and targeted therapies.

And, with the growing understanding that no two tumours are exactly the same, new tests which analyse tumour DNA and proteins (molecular profiling) may allow patients to benefit from more 'personalised' treatments. For example, certain gene mutations, including IDH1 and FGFR2, are found in some intrahepatic CCAs and treatments targeting these mutations have been successfully trialled.

Full details on cholangiocarcinoma, its diagnosis and treatment can be found on AMMF's website, along with information on clinical trials, molecular profiling, and research the charity is currently supporting.