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 2,754[1] 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 England*

AMMF
THE CHOLANGIOCARCINOMA CHARITY

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

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

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About AMMF

Founded in 2002, AMMF was the world's first charity dedicated solely to cholangiocarcinoma (CCA).

AMMF works closely patients and their carers, healthcare professionals, researchers, and policy makers, across the UK and Europe, and collaborates 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 2,754[1] 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.

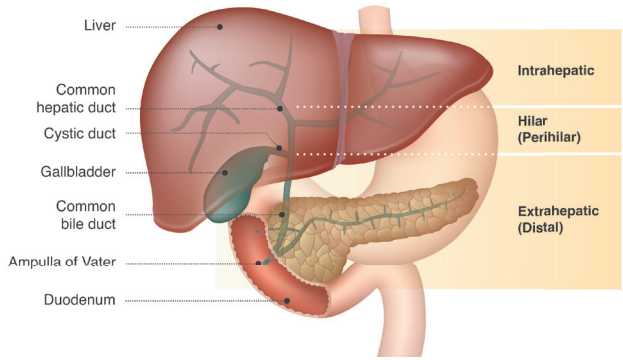
[1] *National Cancer Registration and Analysis Service (NCRAS) and NHS England*

About Cholangiocarcinoma

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.[2]

[2] *Cholangiocarcinoma: Epidemiology and risk factors: https://doi.org/10.1111/liv.14095*

What are the symptoms?

Cholangiocarcinoma (CCA) starts in the bile ducts in or outside the liver, with the commonest symptoms caused by blocking of the flow of bile – jaundice (yellowing of eyes and skin, although this may be less obvious in people with darker skin types), dark urine or pale stools, a pain or ache under the right ribs, indigestion and sometimes itching. There may also be unexplained weight loss, generally feeling unwell, and tiredness. Whilst several of these symptoms may be non-specific and could be caused by benign conditions, they require urgent assessment, especially where jaundice is present.[3]

What treatments are available?

Surgery to totally remove the cancer is currently the only potentially curative treatment. If surgery is not possible, there are several other types of treatment which, although not curative, can help control symptoms and delay the cancer's progression and its effects on the body. Gemcitabine/Cisplatin chemotherapies with the immunotherapy durvalumab is now standard of care first treatment for those with inoperable CCA (approved by NICE January 2024). And molecular profiling (tests which analyse tumour DNA and proteins) is becoming increasingly important. This test may show certain mutations which can be specifically targeted, allowing more 'personalised' treatments. For example, the mutations IDH1 and FGFR2 are found in some intrahepatic CCAs, and treatments targeting these are now available.

Clinical trials looking for further improvements continue, and it is expected that liver transplantation for CCA will be an option for some under stringent criteria soon.

Full details on cholangiocarcinoma, its diagnosis and treatment, molecular profiling, current clinical trials and AMMF-funded research, are available on AMMF's website: **www.ammf.org.uk**

[3] *BSG guidelines for the diagnosis and management of CCA: https://doi.org/10.1136/gutjnl-2023-330029*