

What is Cholangiocarcinoma (CCA)?

CCA, also known as bile duct cancer, is a rare and aggressive type of cancer that affects the bile ducts, which carry digestive fluid (bile) and connect your liver, gallbladder and small intestine.¹

Approximately 8,000 people in the U.S. are diagnosed with CCA each year, but the actual number of cases may be higher. CCA can be hard to diagnose, leading to misclassification as other types of cancers²

There are three different ways in which bile duct cancers are categorized, depending on where the cancer starts:³

- 1 Intrahepatic cholangiocarcinoma:** occurs in the smaller bile ducts inside the liver
- 2 Perihilar cholangiocarcinoma:** occurs in the bile ducts just outside of the liver
- 3 Distal bile cholangiocarcinoma:** occurs further down the bile duct, closer to the small intestine



Risk Factors

There are a variety of factors that can increase your chance of developing CCA, including:^{1,4}

- Primary sclerosing cholangitis
- Liver infection or chronic liver disease
- Choledochal cyst disease
- Bile duct stones
- Cirrhosis
- Hepatitis B or C
- Abnormalities where the bile duct and pancreatic duct meet
- Smoking
- Older age



Symptoms

Signs and symptoms of CCA are usually general, non-specific and do not present until a more advanced stage of the disease. The symptoms often depend on where the cancer is located in the bile duct. These symptoms include:⁵

- Jaundice
- Light colored/greasy stools
- Itchy skin
- Abdominal pain
- Dark urine
- Weight loss or loss of appetite
- Fever
- Nausea and/or vomiting



Treatment

CCA is known as a difficult to treat cancer with few effective treatment options. However, genetic sequencing has identified new potential therapeutic targets for CCA. One genetic alteration of interest is isocitrate dehydrogenase-1 (IDH1). IDH mutations occur in up to 20% of cholangiocarcinoma cases in the U.S. and are not associated with prognosis.⁶

Mutations, like an IDH1 mutation, are changes in the DNA (deoxyribonucleic acid) of a cell, which can play a role in the development of a disease. That's why testing for mutations can help your healthcare team understand which targeted therapies and treatments may work for you.

Currently, there are three standard treatment options for people diagnosed with CCA:⁷

- Surgery
- Radiation therapy
- Chemotherapy

Additionally, gemcitabine-based chemotherapy is often recommended for newly diagnosed patients advanced or metastatic disease.⁷ Patients living with IDH1-mutated CCA, especially those whose disease progresses following chemotherapy, are in need of new treatment options. Speak to your oncologist about a new treatment option for previously treated IDH-mutated CCA.

Survivorship

While treatment may completely destroy or remove the cancer for some people, others may need to maintain regular treatment to control the cancer. For all CCA survivors, it's important to attend follow-up appointments with your oncologist and to keep your health care team informed of any new symptoms or problems that may arise.⁸

Always remember to speak with your oncologist and other members of your health care team about resources and tools that can help you navigate your cancer journey.

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3. American Cancer Society. What Is Bile Duct Cancer? <https://www.cancer.org/cancer/bile-duct-cancer/about/what-is-bile-duct-cancer.html>. Accessed on March 8, 2021.
4. American Cancer Society. Bile Duct Risk Factors. <https://www.cancer.org/cancer/bile-duct-cancer/causes-risks-prevention/risk-factors.html>. Accessed on March 8, 2021.
5. American Society. Signs and Symptoms of Bile Duct Cancer. <https://www.cancer.org/cancer/bile-duct-cancer/detection-diagnosis-staging/signs-symptoms.html>. Accessed on March 8, 2021.

6. Boscoe, A., Rolland, C., & Kelley, R. (2019). Frequency and prognostic significance of isocitrate dehydrogenase 1 mutations in cholangiocarcinoma: a systematic literature review. *Journal Of Gastrointestinal Oncology*, 10(4), 751-765. <https://jgo.amegroups.com/article/view/28868>
7. National Cancer Institute. Bile Duct Cancer (Cholangiocarcinoma) Treatment (PDQ®)-Patient Version. https://www.cancer.gov/types/liver/patient/bile-duct-treatment-pdq#_51. Accessed on March 8, 2021.
8. American Cancer Society. Living as a Bile Duct Cancer Survivor. <https://www.cancer.org/cancer/bile-duct-cancer/after-treatment/follow-up.html>. Accessed on March 8, 2021.