

About Bile Duct Cancer

Get an overview of bile duct cancer and the latest key statistics in the US.

Overview and Types

If you've been diagnosed with bile duct cancer or are worried about it, you likely have a lot of questions. Learning some basics is a good place to start.

• What Is Bile Duct Cancer?

Research and Statistics

See the latest estimates for new cases of bile duct cancer and deaths in the US and what research is currently being done.

- Key Statistics for Bile Duct Cancer
- What's New in Bile Duct Cancer Research?

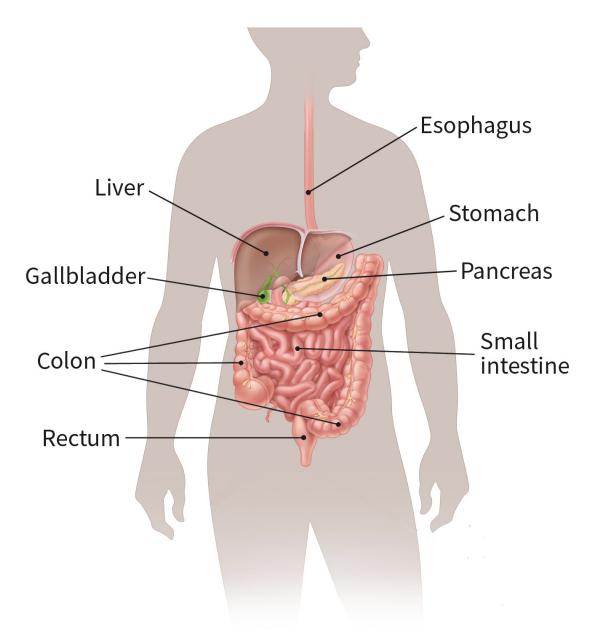
What Is Bile Duct Cancer?

Bile duct cancer (also known as **cholangiocarcinoma**) is a type of cancer that starts in a bile duct. It can start in any part of the bile duct system.

- About the bile ducts
- Types of bile duct cancers by location

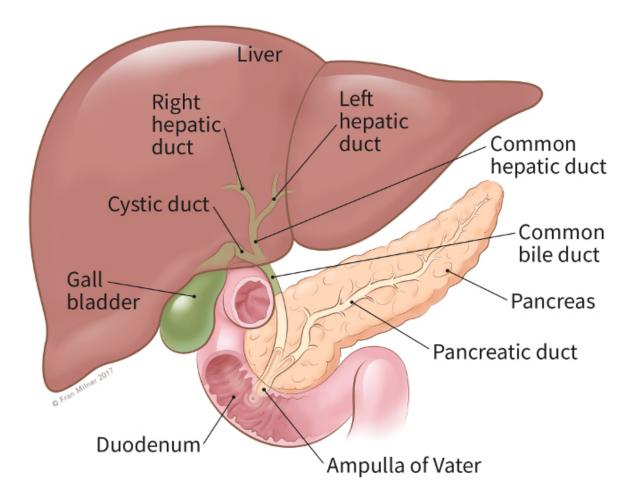
- Types of bile duct cancer by cell type
- Benign bile duct tumors
- Other cancers in the liver

About the bile ducts



To understand bile duct cancer, it helps to know about the bile ducts and what they normally do.

Bile is a fluid made in your liver and stored in your gallbladder. Your **bile ducts** are a series of thin tubes that carry bile from your liver and gallbladder into your small intestine, where the bile helps digest the fats in food. Bile also helps your body get rid of waste material that is filtered out of the bloodstream by your liver.



Different parts of the bile duct system have different names. In the liver, the bile duct system begins as many tiny tubes (called **ductules**). The ductules come together to form small tubes called **ducts**. These small ducts merge into larger ducts and then merge again to form the left and right hepatic ducts. All of these ducts within the liver are called **intrahepatic bile ducts**.

The **left** and **right hepatic ducts** exit the liver and join to form the **common hepatic duct**. Bile ducts located outside the liver are called **extrahepatic bile ducts**.

The **gallbladder** is a small organ that stores bile. It is located below the liver. It is connected to the common hepatic duct via the **cystic duct**. These come together to

form the **common bile duct**. The common bile duct passes through part of the pancreas before it joins with the **pancreatic duct** and empties into the **duodenum** (first part of the small intestine) at the **Ampulla of Vater**.

What Is Cancer? 1

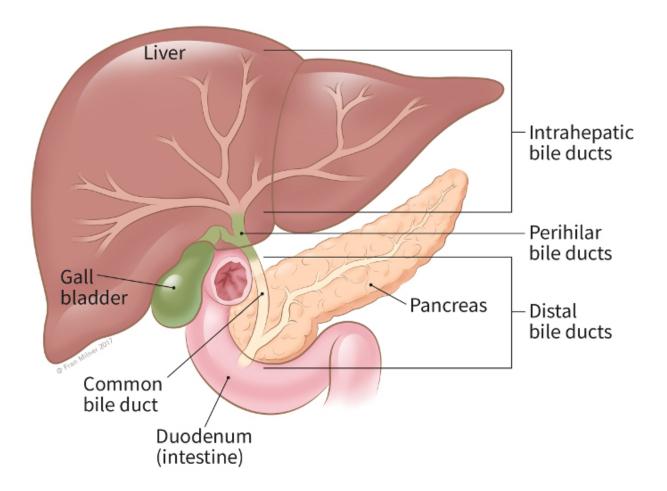
Cancer starts when cells in the body begin to grow out of control. Cells in nearly any part of the body can become cancer cells. Learn more here.

Anatomy Gallery: Digestive System²

Explore our 3D interactive tour of the digestive system.

Types of bile duct cancers by location

Bile duct cancer (cholangiocarcinoma) can start in any part of the bile duct system. Depending on where the cancer starts, it is either an **intrahepatic cholangiocarcinoma** or an **extrahepatic cholangiocarcinoma**.



Intrahepatic cholangiocarcinoma

Intrahepatic bile duct cancers start in the smaller bile duct branches within the liver. They make up about 10% of all bile duct cancers. Sometimes, intrahepatic bile duct cancers are confused with hepatocellular carcinoma, a cancer that starts in the liver cells. But these are two different types of cancer, and they are treated differently.

Extrahepatic cholangiocarcinoma

There are two types of extrahepatic cholangiocarcinoma: perihilar duct cancer and distal duct cancer.

Perihilar bile duct cancers are found in the hilar region of the bile duct system. The hilar region is where the left and right hepatic ducts leave the liver and join to form the common hepatic duct. Perihilar duct cancers are also called **Klatskin tumors**.

Distal duct cancers are found along the common bile duct. The common bile duct starts where the common hepatic duct joins with the cystic duct (from the gallbladder) and extends through the pancreas and into the small intestine at the Ampulla of Vater.

Cancers that start in different parts of the bile ducts can cause different symptoms³.

Types of bile duct cancer by cell type

Bile duct cancers can also be divided into types based on how the cancer cells look under the microscope.

Nearly all bile duct cancers are **cholangiocarcinomas** (a type of adenocarcinoma, which is a cancer that starts in gland cells). Cholangiocarcinomas start in the gland cells that line the inside of the ducts.

Other types of bile duct cancers are much less common. These include sarcomas, lymphomas, and small cell cancers. Our information does not cover these rare types of bile duct cancer.

Benign bile duct tumors

Not all bile duct tumors are cancer. For example, bile duct hamartomas and bile duct adenomas are types of benign (non-cancer) tumors.

Other cancers in the liver

The most common type of cancer that starts in the liver is <u>hepatocellular carcinoma</u>⁴. This type of cancer starts in the main cells that make up the liver. It is more common than cholangiocarcinoma.

Cancers that start in other organs, such as the colon or rectum, can sometimes spread (metastasize) to the liver. These **metastatic cancers** are not true liver cancers. For example, colorectal cancer that has spread to the liver is still colorectal cancer, not liver cancer.

The treatment and outlook for cancer that metastasizes to the liver is not the same as for a cancer that starts in the liver. For this reason, it's important to know whether a tumor in the liver started in bile ducts (cholangiocarcinoma), if it started in the liver tissue itself (hepatocellular carcinoma), or if it started in another organ and has spread to the liver.

Hyperlinks

- 1. www.cancer.org/cancer/understanding-cancer/what-is-cancer.html
- 2. <u>www.cancer.org/cancer/understanding-cancer/anatomy-gallery/digestive-</u> system.html
- 3. <u>www.cancer.org/cancer/types/bile-duct-cancer/detection-diagnosis-staging/signs-</u> <u>symptoms.html</u>
- 4. <u>www.cancer.org/cancer/types/liver-cancer.html</u>

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Key Statistics About Bile Duct Cancer

Bile duct cancer (cholangiocarcinoma) is rare in the United States, but more common in Southeast Asia. It occurs most often in older people.

- How common is bile duct cancer?
- Who gets bile duct cancer?

How common is bile duct cancer?

Bile duct cancer (cholangiocarcinoma) is rare. About 8,000 people in the United States are diagnosed with it each year. This includes both intrahepatic (inside the liver) and extrahepatic (outside the liver) bile duct cancers. But the actual number of cases is likely to be higher, because these cancers can be hard to diagnose, and some might be misclassified as other types of cancer.

Bile duct cancer is more common in Southeast Asia. This is mostly because liver flukes (a <u>parasitic infection</u>¹ that can cause bile duct cancer) are much more common there.

Who gets bile duct cancer?

Bile duct cancer is seen mainly in older people, but it can occur in younger people. The average age of people in the US diagnosed with bile duct cancer is in the 70s.

The chances of survival for people with bile duct cancer depend largely on its location and how advanced it is when it's found. For more on this, see <u>Survival Statistics for Bile</u> <u>Duct Cancers</u>².

Visit the American Cancer Society's <u>Cancer Statistics Center</u>³ for more key statistics.

Hyperlinks

- 1. <u>www.cancer.org/cancer/risk-prevention/infections/infections-that-can-lead-to-</u> <u>cancer/parasites.html</u>
- 2. <u>www.cancer.org/cancer/types/bile-duct-cancer/detection-diagnosis-</u> staging/survival-by-stage.html
- 3. cancerstatisticscenter.cancer.org/

References

American Cancer Society. *Cancer Facts & Figures 2024*. Atlanta, Ga: American Cancer Society; 2024.

Antwi SO, Mousa OY, Patel T. Racial, ethnic, and age disparities in incidence and survival of intrahepatic cholangiocarcinoma in the United States: 1995-2014. *Annals of Hepatology.* 2018;17(2):274-285.

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What's New in Bile Duct Cancer Research?

Because bile duct cancer is rare, it's been hard to study. Most experts agree that treatment in a <u>clinical trial</u>¹ should be considered for any type or stage of bile duct cancer.

Research on bile duct cancer is taking place in many university hospitals, medical centers, and other institutions around the world. Each year, scientists find out more

about what causes the disease, how to prevent it, and how to better treat it. The new and promising treatments discussed here tend to only be available in clinical trials.

- Early Detection
- Treatment

Early Detection

Most bile duct cancers are diagnosed when it's already progressed to late stage, when it is unresectable (cannot be removed with surgery) and incurable. But researchers are studying ways to screen for bile duct cancer so that it can be diagnosed at an earlier and curable stage. Tests currently being studied check a person's blood or bile sample to look for bile duct cancer <u>biomarkers</u>² or <u>circulating tumor DNA</u>³ that might mean cancer is present.

Treatment

Studies are also being done to find better ways of treating bile duct cancer.

Surgery

Doctors are constantly improving the <u>surgical techniques</u>⁴ used to treat bile duct cancers and looking for ways to make surgery possible for more people.

For instance, sometimes the tumor is resectable (removable with surgery), but the person or their liver might not be healthy enough for the surgery. Studies looking at options for minimally invasive surgery are ongoing.

Better ways to use **laparoscopic surgery** are also being tested and compared to open surgery. Adjuvant and neoadjuvant treatments (treatments used before and after surgery) are also active areas of research interest. Doctors are looking for ways to combine other treatments with surgery to improve outcomes.

Radiation therapy

Researchers are looking at better ways to use <u>radiation therapy</u>⁵. One example is using a different type of radiation called **proton beam radiation therapy**. Traditional radiation therapy uses photon or electron beams that release energy all along their path and can damage healthy tissues as well as cancer cells. Proton beams, however, release their energy at their destination (the tumor). This means that proton beam radiation may be

able to deliver more radiation to the tumor while reducing side effects on normal tissues.

Other ways to use radiation therapy are also being studied. For example, doctors are looking at whether radioactive stents placed inside bile ducts might help shrink tumors and keep the ducts open longer than standard stents.

Chemotherapy

In general, the effects of chemo against bile duct cancer have been found to be limited, but new drugs and new combinations of drugs are being tested. Studies are also looking for better ways to combine chemo with other treatments, like surgery, radiation, and liver transplant.

There's a lot of research interest in combining <u>chemotherapy</u>⁶ and<u>targeted therapy</u>⁷.

Targeted therapy

Many drugs that target a specific change (mutation) in cancer cells have been FDA approved and are in use today. These include drugs that target mutations, including *NTRK*, *RET*, *BRAF*, *FGFR2*, *IDH1*, *RET*, *KRAS*, and *HER2*. Studies that look at other targetable mutations are ongoing.

Another area of active research is looking at how combining targeted therapy with other forms of therapy, such as chemotherapy, can better help kill bile duct cancer cells. Different drug combinations are being tested for use in treating bile duct cancer in clinical trials.

Photodynamic therapy (PDT)

During photodynamic therapy (PDT), a doctor gives an inactive form of a drug and then directs a special light at the tumor in the bile duct. This is done using an endoscope in a procedure similar to endoscopic retrograde cholangiopancreatography (ERCP). It causes a chemical change in the drug, activating it to eliminate the tumor cells where the light is directed.

PDT is currently used to relieve pain and symptoms, not to eliminate bile duct cancer. Doctors are also looking at whether PDT can reduce the risk of a stent becoming blocked by stopping the tumor from growing into it.

Hyperlinks

- 1. <u>www.cancer.org/cancer/managing-cancer/making-treatment-decisions/clinical-</u> <u>trials.html</u>
- 2. www.cancer.org/cancer/diagnosis-staging/tests/biomarker-tests.html
- 3. <u>www.cancer.org/cancer/diagnosis-staging/tests/biopsy-and-cytology-tests/biopsy-types.html</u>
- 4. www.cancer.org/cancer/types/bile-duct-cancer/treating/surgery.html
- 5. www.cancer.org/cancer/managing-cancer/treatment-types/radiation.html
- 6. www.cancer.org/cancer/types/bile-duct-cancer/treating/chemotherapy.html
- 7. www.cancer.org/cancer/types/bile-duct-cancer/treating/targeted-therapy.html

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