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# About Bile Duct Cancer

## 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.

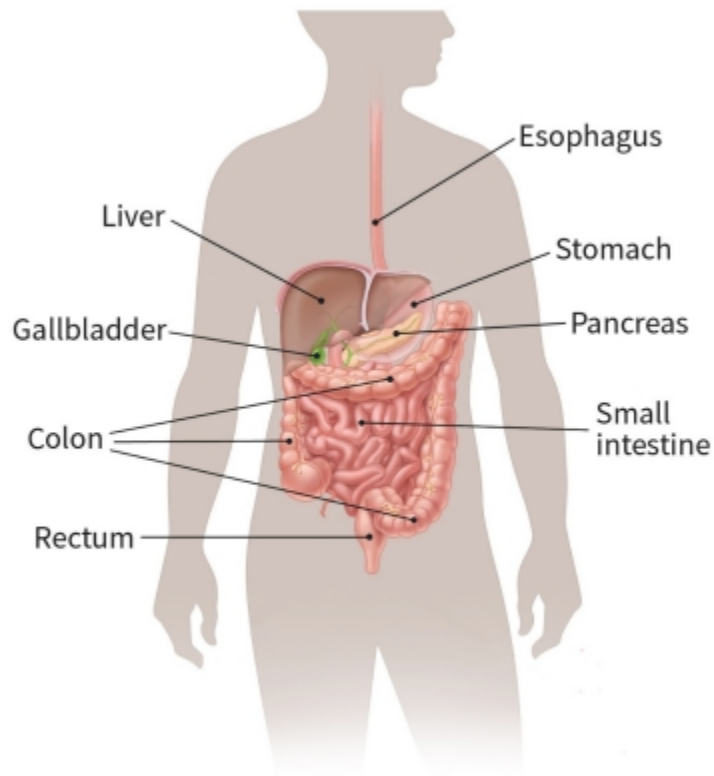
- [Key Statistics for Bile Duct Cancer](#)
  - [What's New in Bile Duct Cancer Research?](#)
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# What Is Bile Duct Cancer?

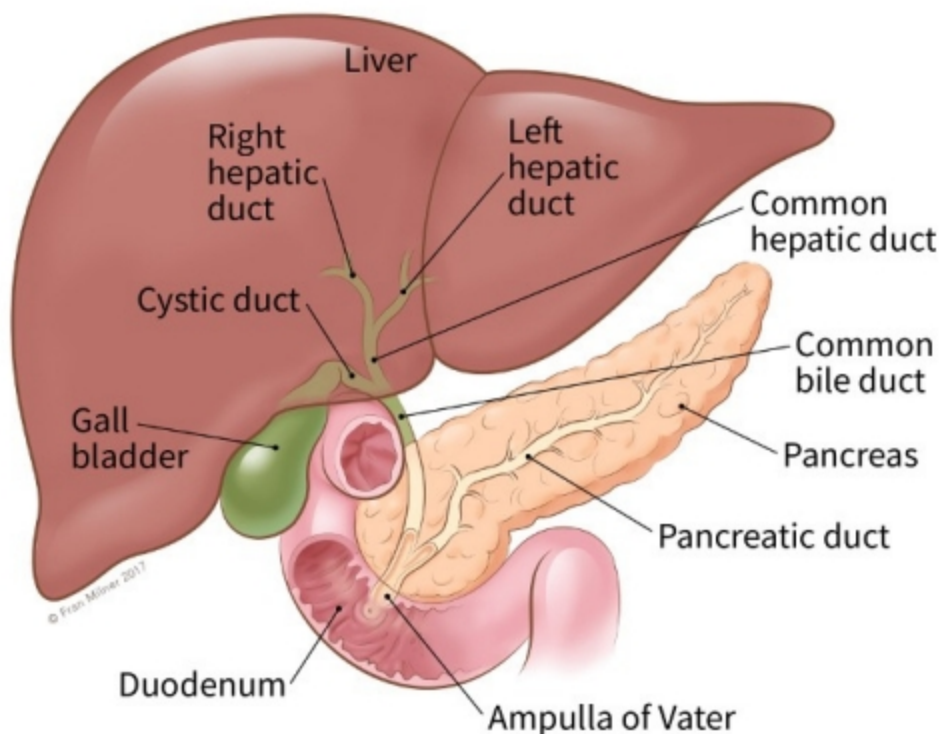
Cancer starts when cells in the body start to grow out of control. Cells in nearly any part of the body can become cancer, and can spread to other areas of the body. To learn more about how cancers start and spread, see [What Is Cancer?](#)<sup>1</sup>

Bile duct cancer starts in a bile duct. To understand this cancer, it helps to know about the bile ducts and what they normally do.

## About the bile ducts



The bile ducts are a series of thin tubes that go from the liver to the small intestine. Their major job is to move a fluid called **bile** from the liver and gallbladder into the small intestine, where it helps digest the fats in food.



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

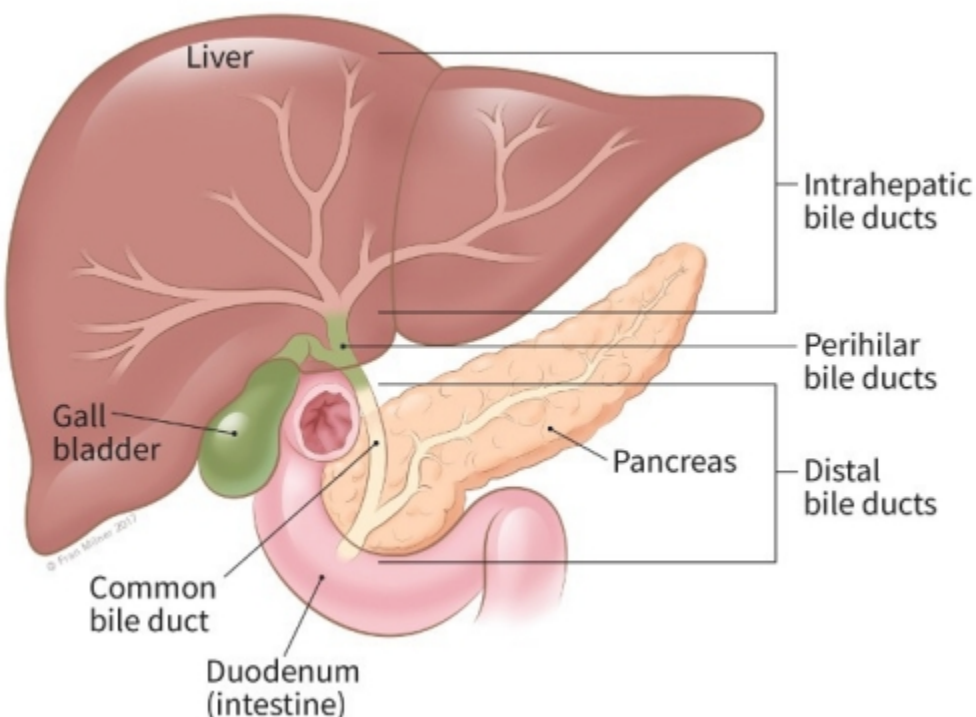
The left and right hepatic ducts exit the liver and join to form the common hepatic duct in an area called the **hilum**. Lower down, the gallbladder (a small organ that stores bile) is joined to the common hepatic duct by a small duct called the **cystic duct**. This combined duct is called the **common bile duct**. The common bile duct passes through part of the pancreas before it joins with the pancreatic duct and empties into the first part of the small intestine (the **duodenum**) at the ampulla of Vater.

## Types of bile duct cancers by location

Cancer can start in any part of the bile duct system. Based on where the cancers are (see the picture below), they're grouped into 3 types:

- Intrahepatic bile duct cancers
- Perihilar (also called *hilar*) bile duct cancers
- Distal bile duct cancers

**Another name for bile duct cancer is cholangiosarcoma.**



Cholangiosarcomas in these different groups cause different symptoms.

### **Intrahepatic bile duct cancers**

These cancers start in the smaller bile duct branches inside the liver. Sometimes they're confused with cancers that start in the liver cells, which are called hepatocellular carcinomas, which are often treated the same way.

### **Perihilar (also called hilar) bile duct cancers**

These cancers start at the hilum, where the left and right hepatic ducts have joined and are just leaving the liver. These are also called Klatskin tumors. These cancers are grouped with distal bile duct cancers as **extrahepatic bile duct cancers**.

### **Distal bile duct cancers**

These cancers are found further down the bile duct, closer to the small intestine. Like perihilar cancers, these are extrahepatic bile duct cancers because they start outside of

the liver.

## 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 or cholangiocarcinomas are adenocarcinomas, which are cancers that start in glandular cells. Bile duct adenocarcinomas start in the mucous 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 other types of bile duct cancer.

## Benign bile duct tumors

Not all bile duct tumors are cancer. Bile duct hamartomas and bile duct adenomas are examples of benign (non-cancer) tumors.

## Other cancers in the liver

The most common type of cancer that starts in the liver – much more common than cholangiosarcoma – is hepatocellular carcinoma, which starts in cells that form the [liver](#)<sup>2</sup>.

Cancers that start in other organs can spread to the liver. These are called liver metastases or metastatic cancer to the liver. Their outlook and treatment are not the same as cancer that starts in the liver (such as hepatocellular carcinoma) or bile ducts (like cholangiocarcinoma), but instead depend on where the cancer started. For this reason, it's important to know whether a tumor in the liver started in bile ducts (is a cholangiocarcinoma), or whether it's made up of cancer cells that started in another organ (like the colon) and spread to the liver (is metastatic cancer).

## Hyperlinks

1. [www.cancer.org/cancer/cancer-basics/what-is-cancer.html](http://www.cancer.org/cancer/cancer-basics/what-is-cancer.html)
2. [www.cancer.org/cancer/liver-cancer.html](http://www.cancer.org/cancer/liver-cancer.html)

## References

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See all references for Bile Duct Cancer ([www.cancer.org/cancer/bile-duct-cancer/references.html](http://www.cancer.org/cancer/bile-duct-cancer/references.html))

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## Key Statistics for 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, mostly because a [parasitic infection](#)<sup>1</sup> that can cause bile duct cancer is much more common there.

Bile duct cancer can occur at younger ages, but it's seen mainly in older people. The average age of people in the US diagnosed with cancer of the intrahepatic bile ducts is 70, and for cancer of the extrahepatic bile ducts it's 72.

The chances of survival for patients with bile duct cancer depend to a large extent on its location and how advanced it is when it's found. For more on this, see [Survival Statistics for Bile Duct Cancers](#)<sup>2</sup>.

Visit the American Cancer Society's [Cancer Statistics Center](#)<sup>3</sup> for more key statistics.

## Hyperlinks

1. [www.cancer.org/cancer/cancer-causes/infectious-agents/infections-that-can-lead-to-cancer/parasites.html](http://www.cancer.org/cancer/cancer-causes/infectious-agents/infections-that-can-lead-to-cancer/parasites.html)
2. [www.cancer.org/cancer/bile-duct-cancer/detection-diagnosis-staging/survival-by-stage.html](http://www.cancer.org/cancer/bile-duct-cancer/detection-diagnosis-staging/survival-by-stage.html)
3. <https://cancerstatisticscenter.cancer.org/>

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See all references for Bile Duct Cancer ([www.cancer.org/cancer/bile-duct-cancer/references.html](http://www.cancer.org/cancer/bile-duct-cancer/references.html))

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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 [clinical trial](#)<sup>1</sup> should be considered for any type or stage of bile duct cancer. This way people can get the best treatment available now and may also get the treatments that are thought to be even better.

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.

## Surgery

Doctors are constantly improving the [surgical techniques](#)<sup>2</sup> used to treat bile duct cancers and looking for ways to make surgery possible for more people.

For instance, sometimes surgery to remove the cancer might be possible, but it can't be done because it might not leave enough healthy liver behind. One option being studied is to cut off the blood supply to the part of the liver that's going to be removed (known as **portal vein embolization**). As this part of the liver shrinks, the other part of the liver grows to compensate. Over time, there might be enough healthy liver to go ahead with the operation to remove the part of the liver with the tumor.

Better ways to use laparoscopic surgery are also being tested and compared to open surgery. Adjuvant and neoadjuvant treatments, those 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 [radiation therapy](#)<sup>3</sup>. One example is using a different type of radiation called proton beam radiation therapy. This form of radiation uses proton beams instead of the usual photon or electron beams. Protons are parts of atoms that cause little damage to tissues they pass through but are very good at killing cells at the end of their path. 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.

Another approach being widely studied is the injection of tiny radioactive beads into the hepatic artery, which carries blood to the area of the cancer. The beads lodge in the



blood vessels near the tumor, where they give off small amounts of radiation. This is called **transarterial radioembolization** or **TARE**. So far, studies have shown good results with TARE, but more research is needed.

## 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 [chemotherapy](#)<sup>4</sup> and targeted therapy.

## Targeted therapy

Drugs have been developed that work differently from standard chemo drugs. These drugs can target specific changes in cancer cells that help them grow and survive. They can also change certain proteins made by the cancer cells to cause the cells to die. Targeted drugs can work with the immune system to help it find and kill cancer cells, too. Many of these drugs are being tested for use in treating bile duct cancer.

Many other kinds of cancers are already treated with targeted therapy. As researchers learn more about the changes in bile duct cells that cause them to become cancer, they're looking to use targeted drugs that focus on killing the cells with these changes.

For instance, some of these drugs target tumor blood vessels. Bile duct tumors need new blood vessels to grow beyond a certain size. Bevacizumab (Avastin<sup>®</sup>), erlotinib (Tarceva<sup>®</sup>), ramuciruman (Cyramza<sup>®</sup>), and regorafenib (Stivarga<sup>®</sup>) are examples of drugs that target blood vessel growth and are being studied against bile duct cancer.

Other drugs have different targets. For example, EGFR, a protein that helps cells grow, is found in high amounts on some cancer cells. Drugs that target EGFR have shown some benefit against many types of cancer. Some of these drugs, such as cetuximab (Erbix<sup>®</sup>) and panitumumab (Vectibix<sup>®</sup>) are now being studied for use in people with bile duct cancer, often in combination with chemotherapy or other targeted drugs.

Other types of targeted therapy, such as MEK inhibitors (like tivantinib ) and [anti-PD1](#)<sup>5</sup> drugs (like pembrolizumab [Keytruda<sup>®</sup>]), are also being studied for use against bile duct cancer.

Again, this is an active area of research, but a lot more research is needed to find out if

targeted therapy works and which drugs work best in treating bile duct cancer.

## Hyperlinks

1. [www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html](http://www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html)
2. [www.cancer.org/cancer/bile-duct-cancer/treating/surgery.html](http://www.cancer.org/cancer/bile-duct-cancer/treating/surgery.html)
3. [www.cancer.org/treatment/treatments-and-side-effects/treatment-types/radiation.html](http://www.cancer.org/treatment/treatments-and-side-effects/treatment-types/radiation.html)
4. [www.cancer.org/cancer/bile-duct-cancer/treating/chemotherapy.html](http://www.cancer.org/cancer/bile-duct-cancer/treating/chemotherapy.html)
5. [www.cancer.org/treatment/treatments-and-side-effects/treatment-types/immunotherapy/immune-checkpoint-inhibitors.html](http://www.cancer.org/treatment/treatments-and-side-effects/treatment-types/immunotherapy/immune-checkpoint-inhibitors.html)

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