About Bile Duct Cancer

Overview and Types

If you have 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.

- What Are the Key Statistics About Bile Duct Cancer?
- What’s New in Bile Duct Cancer Research and Treatment?

What Is Bile Duct Cancer?

Cancer starts when cells in the body begin 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?

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

About the bile ducts
The bile ducts are a series of thin tubes that reach from the liver to the small intestine. The major function of the bile ducts is to move a fluid called *bile* from the liver and gallbladder to 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 small ducts, which then merge into larger ducts and eventually 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 from 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) joins the common hepatic duct through a small duct called the cystic duct. The 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**
Cancers can develop in any part of the bile duct system and, based on their location (see picture below), are classified into 3 types:

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

Cancers in these different areas can cause different symptoms.

**Intrahepatic bile duct cancers**

These cancers develop in the smaller bile duct branches inside the liver. They can sometimes be confused with cancers that start in the liver cells, which are called *hepatocellular carcinomas*, and are often treated the same way.
Perihilar (also called hilar) bile duct cancers

These cancers develop 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 are called cholangiocarcinomas. Most of these are adenocarcinomas, which are cancers that start in glandular cells. Bile duct adenocarcinomas develop from the mucous gland cells that line the inside of the duct.

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

The rest of this information refers only to cholangiocarcinomas.

Benign bile duct tumors

Not all bile duct tumors are cancerous. Bile duct hamartomas and bile duct adenomas are examples of benign (non-cancerous) tumors, which aren’t discussed further in this document.

Other cancers in the liver

The most common type of cancer that starts in the liver – much more common than intrahepatic bile duct cancer – is hepatocellular carcinoma, which develops from liver cells. Hepatocellular carcinoma is discussed in more detail in Liver Cancer.
Cancers that start in some other organs can spread to the liver. These are called secondary liver cancers or liver metastases. 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 an adenocarcinoma in the liver started in bile ducts (and is a cholangiocarcinoma), or whether it started in another organ (such as the colon) and then spread to the liver.

- References

See all references for Bile Duct Cancer

What Are the Key Statistics About Bile Duct Cancer?

Bile duct cancer (cholangiocarcinoma) is not common. About 8,000 people in the United States are diagnosed with bile duct cancer 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, as these cancers can be hard to diagnose, and some might be misclassified as other types of cancer.

Bile duct cancer is much more common in Southeast Asia, mostly because a parasitic infection that can cause bile duct cancer is much more common there.

Bile duct cancer can occur at younger ages, but it is 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 is 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 is found. For survival statistics, see “Survival statistics for bile duct cancers.”

Visit the American Cancer Society’s Cancer Statistics Center for more key statistics.
What’s New in Bile Duct Cancer Research and Treatment?

Bile duct cancer is an uncommon cancer, which in some ways makes it harder to study than more common cancer types. But research into the causes, diagnosis, and treatment of bile duct cancer is currently being done in many medical centers throughout the world.

Surgery

Doctors are constantly improving the surgical techniques used to treat bile duct cancers and looking for ways to make more people eligible for surgery. One potential option, a liver transplant, was discussed in the section “Surgery for bile duct cancer.”

Other options are also being explored. For example, sometimes surgery to remove the cancer might technically be possible, but it can’t be done because it might not leave enough healthy liver behind after the operation. One option might be 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. After several weeks, there might be enough healthy liver on this side to go ahead with the operation to remove the tumor.

Radiation therapy and chemotherapy

Researchers are looking at newer ways of increasing the effectiveness of radiation therapy. Some techniques, such as three-dimensional conformal radiation therapy (3D-CRT), intensity modulated radiation therapy (IMRT), and stereotactic body radiation
therapy (SBRT), are widely available and allow doctors to better aim radiation to affect only the tumor and to spare nearby normal tissues. Other radiation techniques such as intra-operative radiation therapy (IORT) and proton beam radiation therapy may be helpful but are not widely available.

Other forms of 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 studied is the injection of tiny radioactive beads into the hepatic artery (known as radioembolization). The beads lodge in the blood vessels near the tumor, where they give off small amounts of radiation.

Doctors are also testing different combinations of chemotherapy (chemo) drugs with radiation therapy, both on their own and before and after surgery.

In general, the effects of chemo against bile duct cancer have been found to be limited, but newer drugs and combinations of drugs are being tested. Newer ways to give chemo are also being studied. For example, trans-arterial chemoembolization (TACE) combines embolization (blocking off blood vessels supplying the tumor) with chemotherapy. Most often, this is done by using tiny beads that give off a chemo drug to plug up the hepatic artery.

**Targeted therapy**

Newer drugs are being developed that work differently from standard chemo drugs. These drugs target specific parts of cancer cells or their surrounding environments.

One target of several newer drugs is tumor blood vessels. Bile duct tumors need new blood vessels to grow beyond a certain size. Sorafenib (Nexavar®), bevacizumab (Avastin®), pazopanib (Votrient®), and regorafenib (Stivarga®) are examples of drugs that target blood vessel growth and are being studied against bile duct cancer.

Other new 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 several types of cancer. Some of these drugs, such as cetuximab (Erbitux®) and panitumumab (Vectibix®) are now being studied for use in people with bile duct cancer, usually in combination with chemotherapy or other targeted drugs.

Drugs known as MEK inhibitors, such as trametinib (Mekinist®), are also being studied for use against bile duct cancer.