BAPTIST MD Anderson Cancer Center
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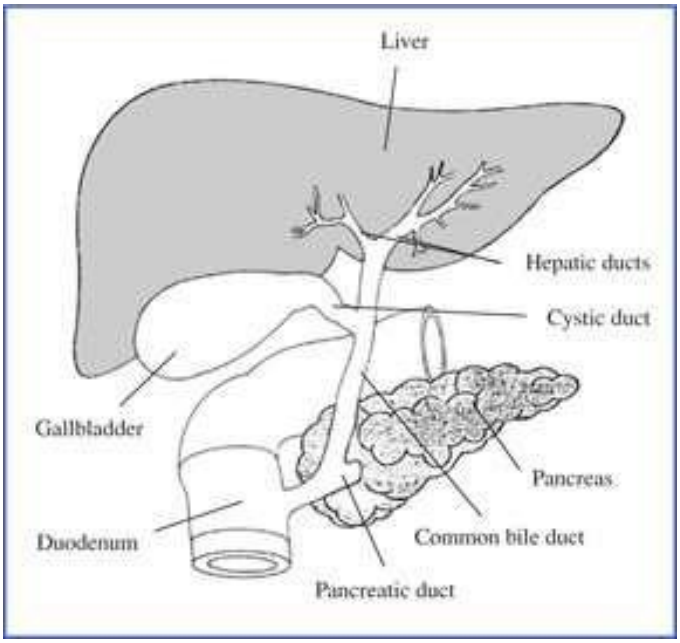
Cholangiocarcinoma

What are bile ducts?

Bile is a liquid made in the liver. Bile ducts are tubes that carry bile out of the liver to the gallbladder and small intestine. These ducts start inside the left and right sides of the liver and connect into a main duct called the common bile duct. The common bile duct is located outside the liver and connects to the small intestine. In the intestine, bile helps digest food.



The liver and bile ducts

What is cholangiocarcinoma?

Cells normally grow, divide and make new cells in order to keep the body functioning properly. When the cells are damaged, they can grow and divide out of control and form a tumor. Tumors can then form cancers. Cancer cells can grow into nearby structures or spread to other organs (metastasize). Cholangiocarcinoma is a rare cancer that starts in the bile ducts. It is estimated that about 2,500 people are diagnosed in the United States per year, but this number is rapidly rising. The exact cause is unknown, but there are certain risk factors for developing the disease. These include:

- Tobacco use
- Ulcerative colitis – This is a disease that causes chronic swelling (inflammation) and ulcers or open wounds in the colon.
- Primary sclerosing cholangitis (PSC) – This is a chronic disease that causes progressive inflammation and scarring of the bile ducts.
- History of infection with liver parasites – This disease rarely occurs in the United States.
- Birth defects involving the bile ducts

Many people who develop cholangiocarcinoma have none of these risk factors.

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What are the symptoms?

Common symptoms include pain in the belly, weight loss, fatigue, fever, jaundice (yellowing of the skin and eyes) and itching.

What happens after diagnosis?

After cancer is diagnosed, your doctor may order tests to determine the size and location of the tumor and if it has spread outside of the bile ducts. This is called staging. Staging is needed to help choose the best treatment for you. There are four stages of cholangiocarcinoma:

- Stage I: This is very early cancer that is confined to the bile ducts.
- Stage II: This tumor may have grown into nearby tissue, but has not spread to the lymph nodes are other parts of the body.
- Stage III: This cancer has spread to nearby lymph nodes.
- Stage IV: This advanced cancer has spread to other parts of the body, including organs or lymph nodes that are far away from the bile ducts.

How is cholangiocarcinoma treated?

Treatment for cholangiocarcinoma depends on the size and location of the tumor, as well as the general health of the patient and whether or not the treatment benefits will outweigh the risks of treatment. The treatments listed below may be used alone or in combination to control both the cancer and the symptoms it causes.

Surgery

Surgically removing the tumor is the best chance to cure cholangiocarcinoma. Over 90 percent of these cancers are **not** removable by surgery due to the size, location, or if the tumor has spread. In order for a patient to possibly be a candidate for surgery, these guidelines must be met:

- The cancer must be limited to one side of the liver, so it most often cannot involve both right and left bile ducts.
- The tumor cannot be invading or surrounding certain blood vessels that run through the liver.
- The patient cannot have cancer that has spread outside the liver.
- The surgeon must believe that **all** of the cancer can be removed. Otherwise, the surgeon will not try to remove it.

If your doctor feels that your cancer may be removed by surgery, then you may be seen by a surgeon. He or she will discuss the surgery that may be possible for you.

Liver Transplant

Many people with cholangiocarcinoma ask about the possibility of a liver transplant. Unfortunately, this is rarely an option because the screening process is very involved and is available only through specific clinical trials. Patients cannot have any cancer outside of the liver, including the lymph nodes, or have other medical conditions. If you have questions about

why you may or may not be a candidate for liver transplantation, ask your doctor.

Radiation Treatment

Radiation treatment uses high-energy x-rays to kill cancer cells. It only affects the cancer in the area being treated. External beam radiation therapy uses a machine to kill the cancer cells with radioactive rays. It may be used alone or in combination with chemotherapy. Patients who will have surgery sometimes receive radiation before or after surgery. Not everyone is a candidate, but if radiation is a treatment option for you, your doctor will talk to you in detail about the length of treatment and the possible side effects that it can cause.

Chemotherapy

Chemotherapy is the use of drugs to treat cancer. It is given in several different ways, such as through a vein or by mouth. Chemotherapy works throughout the body and can treat cancer cells that have metastasized to other parts of the body.

Chemotherapy can be combined with radiation therapy, or it may be given before and/or after surgery. Chemotherapy is the frontline treatment for tumors that cannot be removed by surgery. If given alone, chemotherapy does not cure cancer. Instead, the goal is to extend the length of life, provide quality of life and reduce the symptoms of cancer. However, chemotherapy often causes side effects. This is because chemotherapy affects healthy cells in the body as well as cancer cells. The most common side effects are:

- nausea,
- appetite loss,
- fatigue,
- diarrhea and
- low blood counts (this may increase risk for bleeding or infection).

Other side effects may occur depending on the type of chemotherapy you receive. Your doctor will give you prescriptions for medicine to manage these side effects and to help you maintain your quality of life.

Other Treatments

Other methods that may be used to treat cholangiocarcinoma or its complications include:

- **Y-90 radioembolization –** Tiny beads containing radioactive material are placed inside the tumor. This is done by threading a catheter through an artery in the groin to the main blood supply for the liver. Unlike external beam radiation, this procedure allows the radiation to act from inside the liver. It causes fewer side effects than chemotherapy because it affects only the surrounding tumor tissue. This technique is still an experimental treatment for cholangiocarcinoma. Talk to your doctor to learn more about the procedure.
- **Endoscopic retrograde cholangiopancreatography (ERCP)** – During this procedure, a doctor passes a scope (tube with a camera on the end) down the throat through the stomach to examine the common bile duct. Often, when a tumor blocks a bile duct, the bile cannot drain past the blockage and the patient becomes jaundiced from the backing up of bile. The doctor may place a stent to open the bile duct, which allows the bile to drain out of the liver. Before

treatment can begin, bilirubin levels in the body must be normal or very close to normal. Otherwise, chemotherapy treatments could damage the liver.

- **Biliary drain** – This is an external drain that may be needed if the stent placed during an ERCP does not help to reduce bilirubin levels and relieve jaundice. The tube is placed through the skin into the bile ducts. It allows bile that has been blocked to drain out of the liver and into a bag on the outside of the body.

Follow Up

If you are being treated at Baptist MD Anderson, you will be asked to come in for follow-up visits during and after treatment. During these visits, your doctor will examine you and check how your body is handling the treatment. He or she will adjust your treatment as needed.

After a certain number of treatments, your doctor will order more imaging tests to restage the cancer. In restaging, the doctor compares the new images to the original images. Comparing the two helps the doctor know if the treatment was effective and how to plan your next course of treatment. The next treatment may be to continue on the same chemotherapy regimen, change the chemotherapy, or switch to a different treatment such as radiation or surgery.

If you have any questions or concerns about this information, please contact your health care team.