Cholangiocarcinoma

What are Bile Ducts?
Bile is a liquid made in the liver. Bile ducts are tubes that carry this liquid out of the liver to the gallbladder and small intestine. These ducts start inside either side of the liver and connect to form a main tube 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.

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. Some tumors can become cancerous. Cancer cells in these tumors can grow or spread into nearby structures or 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 each 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, which is a disease that causes chronic swelling (inflammation) and ulcers or open wounds in the colon.
- Primary sclerosing cholangitis, which is a chronic disease that causes progressive inflammation and scarring of the bile ducts.
- History of infection with liver parasites, which rarely occurs in the United States.
- Birth defects involving the bile ducts.

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

What are the Symptoms?
Common symptoms include stomach pain, 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 is very early cancer that is confined to the bile ducts.
- Stage II is a tumor that may have grown into nearby tissue, but has not spread to the lymph nodes or other parts of the body.
- Stage III is cancer that has spread to nearby lymph nodes.
- Stage IV is an 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 patient's general health and whether or not the treatment benefits will outweigh its risks. The treatments listed below may be used alone or in combination to control both the cancer and the resulting symptoms.

Surgery

Surgically removing the tumor is the best chance to cure cholangiocarcinoma. However, over 90 percent of these cancers are *not* removable by surgery due to its size and location or if the tumor has spread. In order to 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 you are a candidate for surgery, then you may be seen by a surgeon, who will discuss this option in more detail.

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 a liver transplant, ask your doctor.

Radiation Treatment

Radiation treatment uses high-energy x-rays to kill cancer cells. It is designed to target the cancer in the area being treated. External beam radiation therapy uses a machine to deliver the radiation from outside the body. 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 discuss this in more detail, including risk factors.

Chemotherapy

Chemotherapy uses drugs to damage or kill cancer cells. It is typically given through a needle in a vein or taken by mouth. Chemotherapy works throughout the body and can treat cancer cells that have metastasized.

Chemotherapy can be combined with radiation therapy. 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 has its own side effects. This is because the drugs affect healthy cells in the body as well as those that are cancerous.

The most common side effects of chemotherapy are:

- Nausea.
- Appetite loss.
- Fatigue.
- Diarrhea.
- 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 medications to help manage these side effects and your quality of life.

Other Treatments

Other methods that may be used to treat cholangiocarcinoma or its complications include Y-90 radioembolization, endoscopic retrograde cholangiopancreatography (ERCP) and biliary drain.

Y-90 radioembolization involves the placement of tiny beads containing radioactive material 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.

During this ERCP, a doctor passes a scope (tube with a camera on the end) down the throat and through the stomach to examine the common bile duct. Often, when a tumor blocks a bile duct, the bile cannot drain and the patient becomes jaundiced. 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.

A biliary drain 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

You will be asked to come in for follow-up visits during and after treatment. During these visits, your doctor will examine you to see how your body is handling the treatment. He or she will modify 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 proceed. 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.