



EXTRAHEPATIC BILE DUCT CANCER

General Information

Extrahepatic bile duct cancer is a rare disease in which malignant (cancer) cells form in the part of bile duct that is outside the liver.

A network of bile ducts (tubes) connects the liver and the gallbladder to the small intestine. This network begins in the liver where many small ducts collect bile, a fluid made by the liver to break down fats during digestion. The small ducts come together to form the right and left hepatic bile ducts, which lead out of the liver. The two ducts join outside the liver to become the common hepatic duct. The part of the common hepatic duct that is outside the liver is called the extrahepatic bile duct. The extrahepatic bile duct is joined by a duct from the gallbladder (which stores bile) to form the common bile duct. Bile is released from the gallbladder through the common bile duct into the small intestine when food is being digested.

Having colitis or certain liver diseases can increase the risk of developing extrahepatic bile duct cancer.

Anything that increases your risk of getting a disease is called a risk factor. Having a risk factor does not mean that you will get cancer; not having risk factors doesn't mean that you will not get cancer. People who think they may be at risk should discuss this with their doctor. Risk factors include having any of the following disorders:

- Primary sclerosing cholangitis.
- Chronic ulcerative colitis.
- Choledochal cysts.
- Infection with a Chinese liver fluke parasite.

Possible signs of extrahepatic bile duct cancer include jaundice and pain.

These and other symptoms may be caused by extrahepatic bile duct cancer or by other conditions. A doctor should be consulted if any of the following problems occur:

- Jaundice (yellowing of the skin or whites of the eyes).
- Pain in the abdomen.
- Fever.
- Itchy skin

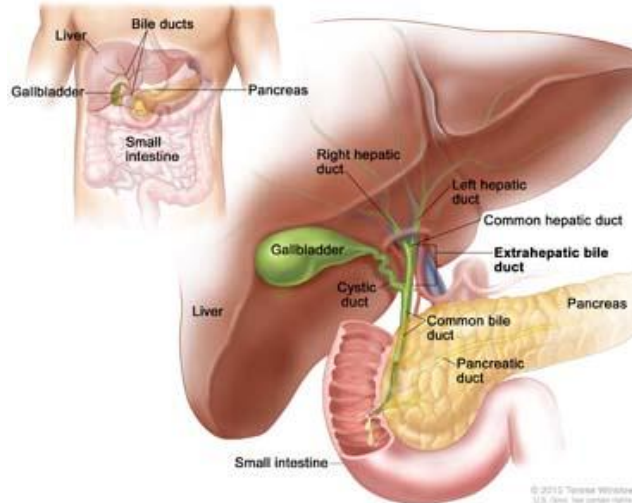
Anatomy of the Extrahepatic Bile Duct

The extrahepatic bile duct is part of a network of small ducts in the liver that collects bile. Bile flows into the right and left hepatic ducts which join together to form the common hepatic duct. The extrahepatic bile duct is the part of the common hepatic duct that is outside the liver. Bile flows through the extrahepatic bile duct and into the cystic duct to the gallbladder where it is stored. When food is being digested, bile flows back through the cystic duct and into the common bile duct where it is released into the small intestine.



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Tests that examine the bile duct and liver are used to detect (find) and diagnose extrahepatic bile duct cancer. The following tests and procedures may be used:

- **Physical Examination:** An exam of the body to check general signs of health, including checking for signs of disease, such as lumps or anything else that seems unusual. A history of the patient's health habits and past illnesses and treatments will also be taken.
- **Ultrasound exam:** A procedure in which high-energy sound waves (ultrasound) are bounced off internal tissues or organs and make echoes. The echoes form a picture of body tissues called a sonogram. The picture can be printed to be looked at later.
- **CT scan (CAT scan):** A procedure that makes a series of detailed pictures of areas inside the body, taken from different angles. The pictures are made by a computer linked to an x-ray machine. A dye may be injected into a vein or swallowed to help the organs or tissues show up more clearly. This procedure is also called computed tomography, computerized tomography, or computerized axial tomography. A spiral or helical CT scan makes detailed pictures of areas inside the body using an x-ray machine that scans the body in a spiral path.
- **MRI (magnetic resonance imaging):** A procedure that uses a magnet, radio waves, and a computer to make a series of detailed pictures of areas inside the body. This procedure is also called nuclear magnetic resonance imaging (NMRI).
- **ERCP (endoscopic retrograde cholangiopancreatography):** A procedure used to x-ray the ducts (tubes) that carry bile from the liver to the gallbladder and from the gallbladder to the small intestine. Sometimes bile duct cancer causes these ducts to narrow and block or slow the flow of bile, causing jaundice. An endoscope is passed through the mouth, esophagus, and stomach into the first part of the small intestine. An endoscope is a thin, tube-like instrument with a light and a lens for viewing. A catheter (a smaller tube) is then inserted through the endoscope into the pancreatic ducts. A dye is injected through the catheter into the ducts and an x-ray is taken. If the ducts are blocked by a tumor, a fine tube may be inserted into the duct to unblock it. This tube (or stent) may be left in place to keep the duct open. Tissue samples may also be taken and checked under a microscope for signs of cancer.
- **PTC (percutaneous transhepatic cholangiography):** A procedure used to x-ray the liver and bile ducts. A thin needle is inserted through the skin below the ribs and into the liver. Dye is injected into the liver or bile ducts and an x-ray is taken. If a blockage is found, a thin, flexible tube called a stent is sometimes left in the liver to drain bile into the small intestine or a collection bag outside the body.



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- **Biopsy:** The removal of cells or tissues so they can be viewed under a microscope to check for signs of cancer. The sample may be taken using a thin needle inserted into the duct during an x-ray or ultrasound. This is called a fine-needle aspiration (FNA) biopsy. The biopsy is usually done during PTC or ERCP. Tissue may also be removed during surgery.
- **Liver function tests:** A procedure in which a blood sample is checked to measure the amounts of certain substances released into the blood by the liver. A higher than normal amount of a substance can be a sign of liver disease that may be caused by extrahepatic bile duct cancer.

Certain factors affect prognosis (chance of recovery) and treatment options.

The prognosis (chance of recovery) and treatment options depend on the following:

- The stage of the cancer (whether it affects only the bile duct or has spread to other places in the body).
- Whether the tumor can be completely removed by surgery.
- Whether the tumor is in the upper or lower part of the duct.
- Whether the cancer has just been diagnosed or has recurred (come back).

Treatment options may also depend on the symptoms caused by the tumor. Extrahepatic bile duct cancer is usually found after it has spread and can rarely be removed completely by surgery. Palliative therapy may relieve symptoms and improve the patient's quality of life.

Stages of Extrahepatic Bile Duct Cancer

After extrahepatic bile duct cancer has been diagnosed, tests are done to find out if cancer cells have spread within the bile duct or to other parts of the body.

The process used to find out if cancer has spread within the extrahepatic bile duct or to other parts of the body is called staging. The information gathered from the staging process determines the stage of the disease. It is important to know the stage in order to plan treatment.

Extrahepatic bile duct cancer is usually staged following a laparotomy. A surgical incision is made in the wall of the abdomen to check the inside of the abdomen for signs of disease and to remove tissue and fluid for examination under a microscope. The results of the diagnostic imaging tests, laparotomy, and biopsy are viewed together to determine the stage of the cancer. Sometimes, a laparoscopy will be done before the laparotomy to see if the cancer has spread. If the cancer has spread and cannot be removed by surgery, the surgeon may decide not to do a laparotomy.

There are three ways that cancer spreads in the body.

- Through tissue. Cancer invades the surrounding normal tissue.
- Through the lymph system. Cancer invades the lymph system and travels through the lymph vessels to other places in the body.
- Through the blood. Cancer invades the veins and capillaries and travels through the blood to other places in the body.

When cancer cells break away from the primary (original) tumor and travel through the lymph or blood to other places in the body, another (secondary) tumor may form. This process is called metastasis. The secondary (metastatic) tumor is the same type of cancer as



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the primary tumor. For example, if breast cancer spreads to the bones, the cancer cells in the bones are actually breast cancer cells. The disease is metastatic breast cancer, not bone cancer.

The following stages are used for extrahepatic bile duct cancer:

- Stage 0 (Carcinoma in Situ) - In stage 0, abnormal cells are found in the innermost layer of tissue lining the extrahepatic bile duct. These abnormal cells may become cancer and spread into nearby normal tissue. Stage 0 is also called carcinoma in situ.
- Stage I - In stage I, cancer has formed. Stage I is divided into stage IA and stage IB.
 - Stage IA: Cancer is found in the bile duct only.
 - Stage IB: Cancer has spread through the wall of the bile duct.
- Stage II - Stage II is divided into stage IIA and stage IIB.
 - Stage IIA: Cancer has spread to the liver, gallbladder, pancreas, and/or to either the right or left branch of the hepatic artery or to the right or left branch of the portal vein.
 - Stage IIB: Cancer has spread to nearby lymph nodes and: is found in the bile duct; or has spread through the wall of the bile duct; or has spread to the liver, gallbladder, pancreas, and/or the right or left branches of the hepatic artery or portal vein.
- Stage III - In stage III, cancer has spread:
 - to the main portal vein or to both right and left branches of the portal vein; or
 - to the hepatic artery; or
 - to other nearby organs or tissues, such as the colon, stomach, small intestine, or abdominal wall. Cancer may have spread to nearby lymph nodes also.
- Stage IV - In stage IV, cancer has spread to lymph nodes and/or organs far away from the extrahepatic bile duct

Extrahepatic bile duct cancer can also be grouped according to how the cancer may be treated. There are two treatment groups:

Localized (and resectable)

The cancer is in an area where it can be removed completely by surgery.

Unresectable, recurrent, or metastatic

Unresectable cancer cannot be removed completely by surgery. Most patients with extrahepatic bile duct cancer have unresectable cancer.

Recurrent cancer is cancer that has recurred (come back) after it has been treated. Extrahepatic bile duct cancer may come back in the bile duct or in other parts of the body.

Metastasis is the spread of cancer from the primary site (place where it started) to other places in the body. Metastatic extrahepatic bile duct cancer may have spread to nearby blood vessels, the liver, the common bile duct, nearby lymph nodes, other parts of the abdominal cavity, or to distant parts of the body.

Treatment Option Overview

There are different types of treatment for patients with extrahepatic bile duct cancer.

Different types of treatment are available for patients with extrahepatic bile duct cancer. Some treatments are standard (the currently used treatment), and some are being tested in clinical trials. A treatment clinical trial is a research study meant to help improve current treatments or obtain information on new treatments for patients with cancer. When clinical trials show that a new treatment is better



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than the standard treatment, the new treatment may become the standard treatment. Patients may want to think about taking part in a clinical trial. Some clinical trials are open only to patients who have not started treatment.

Three types of standard treatment are used:

Surgery

The following types of surgery are used to treat extrahepatic bile duct cancer:

- **Removal of the bile duct:** If the tumor is small and only in the bile duct, the entire bile duct may be removed. A new duct is made by connecting the duct openings in the liver to the intestine. Lymph nodes are removed and viewed under a microscope to see if they contain cancer.
- **Partial hepatectomy:** Removal of the part of the liver where cancer is found. The part removed may be a wedge of tissue, an entire lobe, or a larger part of the liver, along with some normal tissue around it.
- **Whipple procedure:** A surgical procedure in which the head of the pancreas, the gallbladder, part of the stomach, part of the small intestine, and the bile duct are removed. Enough of the pancreas is left to make digestive juices and insulin.
- **Surgical biliary bypass:** If the tumor cannot be removed but is blocking the small intestine and causing bile to build up in the gallbladder, a biliary bypass may be done. During this operation, the gallbladder or bile duct will be cut and sewn to the small intestine to create a new pathway around the blocked area. This procedure helps to relieve jaundice caused by the build-up of bile.
- **Stent placement:** If the tumor is blocking the bile duct, a stent (a thin tube) may be placed in the duct to drain bile that has built up in the area. The stent may drain to the outside of the body or it may go around the blocked area and drain the bile into the small intestine. The doctor may place the stent during surgery or PTC, or with an endoscope.

Radiation therapy

Radiation therapy is a cancer treatment that uses high-energy x-rays or other types of radiation to kill cancer cells or keep them from growing. There are two types of radiation therapy. External radiation therapy uses a machine outside the body to send radiation toward the cancer. Internal radiation therapy uses a radioactive substance sealed in needles, seeds, wires, or catheters that are placed directly into or near the cancer. The way the radiation therapy is given depends on the type and stage of the cancer being treated.

Chemotherapy

Chemotherapy is a cancer treatment that uses drugs to stop the growth of cancer cells, either by killing the cells or by stopping them from dividing. When chemotherapy is taken by mouth or injected into a vein or muscle, the drugs enter the bloodstream and can reach cancer cells throughout the body (systemic chemotherapy). When chemotherapy is placed directly into the cerebrospinal fluid, an organ, or a body cavity such as the abdomen, the drugs mainly affect cancer cells in those areas (regional chemotherapy). The way the chemotherapy is given depends on the type and stage of the cancer being treated.

Additional Information

New types of treatment are being tested in clinical trials. This summary section describes treatments that are being studied in clinical trials. It may not mention every new treatment being studied. Information about clinical trials is available from the NCI Web site.



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Radiation sensitizers

Clinical trials are studying ways to improve the effect of radiation therapy on tumor cells, including the following:

- **Hyperthermia therapy:** A treatment in which body tissue is exposed to high temperatures to damage and kill cancer cells or to make cancer cells more sensitive to the effects of radiation therapy and certain anticancer drugs.
- **Radiosensitizers:** Drugs that make tumor cells more sensitive to radiation therapy. Combining radiation therapy with radiosensitizers may kill more tumor cells.

Patients may want to think about taking part in a clinical trial.

For some patients, taking part in a clinical trial may be the best treatment choice. Clinical trials are part of the cancer research process. Clinical trials are done to find out if new cancer treatments are safe and effective or better than the standard treatment. Many of today's standard treatments for cancer are based on earlier clinical trials. Patients who take part in a clinical trial may receive the standard treatment or be among the first to receive a new treatment. Patients who take part in clinical trials also help improve the way cancer will be treated in the future. Even when clinical trials do not lead to effective new treatments, they often answer important questions and help move research forward. Patients can enter clinical trials before, during, or after starting their cancer treatment. Some clinical trials only include patients who have not yet received treatment. Other trials test treatments for patients whose cancer has not gotten better. There are also clinical trials that test new ways to stop cancer from recurring (coming back) or reduce the side effects of cancer treatment.

Clinical trials are taking place in many parts of the country. See the Treatment Options section that follows for links to current treatment clinical trials. These have been retrieved from NCI's listing of clinical trials.

Follow-up tests may be needed. Some of the tests that were done to diagnose the cancer or to find out the stage of the cancer may be repeated. Some tests will be repeated in order to see how well the treatment is working. Decisions about whether to continue change, or stop treatment may be based on the results of these tests. This is sometimes called re-staging.

Some of the tests will continue to be done from time to time after treatment has ended. The results of these tests can show if your condition has changed or if the cancer has recurred (come back). These tests are sometimes called follow-up tests or check-ups.

Treatment Options for Extrahepatic Bile Duct Cancer

A link to a list of current clinical trials is included for each treatment section. For some types or stages of cancer, there may not be any trials listed. Check with your doctor for clinical trials that are not listed here but may be right for you.

Localized Extrahepatic Bile Duct Cancer

Treatment of localized extrahepatic bile duct cancer may include the following:

- Stent placement or biliary bypass to relieve blockage of the bile duct may be done before surgery to relieve jaundice.
- Surgery, with or without external-beam radiation therapy.

Check for U.S. clinical trials from NCI's list of cancer clinical trials that are now accepting patients with localized extrahepatic bile duct cancer. For more specific results, refine the search by using other search features, such as the location of the trial, the type of treatment, or the name of the drug. General information about clinical trials is available from the NCI Web site.



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Unresectable, Recurrent, or Metastatic Extrahepatic Bile Duct Cancer

Treatment of unresectable, recurrent, or metastatic extrahepatic bile duct cancer is usually within a clinical trial. Treatment may include the following:

- Stent placement or biliary bypass with or without internal or external radiation therapy, as palliative treatment to relieve symptoms and improve the quality of life.
- Chemotherapy.
- A clinical trial of new ways to give palliative radiation therapy, such as combining it with hyperthermia therapy, radiosensitizers, or chemotherapy.
- A clinical trial of new drugs and drug combinations.

Check for U.S. clinical trials from NCI's list of cancer clinical trials that are now accepting patients with unresectable extrahepatic bile duct cancer, recurrent extrahepatic bile duct cancer and metastatic extrahepatic bile duct cancer. For more specific results, refine the search by using other search features, such as the location of the trial, the type of treatment, or the name of the drug. General information about clinical trials is available from the NCI Web site.