Cholangiocarcinoma (Bile Duct Cancer)

The Bile Duct System (Biliary Tract)

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. Ducts within the liver are called intrahepatic ducts. 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.
Bile Duct Cancer (Cholangiocarcinoma)

Bile duct cancer, a rare cancer in the United States, starts in the lining of the bile ducts (epithelial cells). Bile duct cancers can start in the bile ducts within the liver. These are called intrahepatic cholangiocarcinomas. Or they can start in the bile ducts outside of the liver. These are called extrahepatic cholangiocarcinomas.

Risk Factors

The following disorders are risk factors for developing bile duct cancer:

- Primary sclerosing cholangitis
- Chronic ulcerative colitis
- Choledochal (common bile duct) cysts
- Infection with a Chinese liver fluke parasite

Symptoms

Bile duct cancers usually cause symptoms when the cancer blocks (obstructs) the biliary drainage system causing yellow skin or whites of the eyes (jaundice). Itching (pruritis) often occurs due to the deposits of bile salts in the skin. Other symptoms include pain, often in the right upper abdomen, fever, clay-colored stools, and dark urine.

Prognosis

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

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

Treatment options may also depend on the symptoms caused by the cancer. Bile duct cancer found after it has spread may not be completely removed by surgery (unresectable). If the cancer has spread, palliative treatment may improve the patient’s quality of life by controlling symptoms and complications of the disease.

Diagnosis and Staging

These tests and procedures may be used to diagnose bile duct cancers and determine the stage of disease (extent of the cancer). The stage of the disease is important to know in order to make a treatment plan.

- Physical exam and complete history of health habits, past illnesses, and treatments.
- Ultrasound – a radiology procedure that bounces high-energy sound waves (ultrasound) off tissues or organs to form a picture called a sonogram
- CT scan (CAT scan) – detailed picture of the inside of the body taken by a special x-ray machine that is attached to a computer
- MRI (magnetic resonance imaging) – a radiology procedure that uses a magnet, radio waves, and a computer to make detailed pictures of the inside of the body
- **ERCP** (endoscopic retrograde cholangiopancreatography) – a procedure performed by a gastroenterologist (a doctor who specializes in digestive tract diseases) where a small lighted tube (scope) is passed through the mouth, esophagus, stomach, and first part of the small intestine. A smaller tube or catheter is passed into the ducts, a dye is injected and x-rays taken. If the ducts are blocked, a small flexible tube (stent) may be inserted into the duct to unblock it. Tissue samples (biopsies) may be taken.

- **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 x-rays are taken. If a blockage is found, a flexible tube or stent is sometimes left in the liver to drain bile into either the small intestine or to a collection bag outside the body. Tissue samples or biopsies may also be taken.

- **Biopsy** – the removal of cells or tissues to be examined under the microscope to check for cancer. Tissue can be removed during an ERCP, a PTC, or surgery.

- **Liver function tests** – blood tests that measure the amounts of certain substances released into the blood by the liver. Higher than normal amounts can be a sign of liver disease that may be caused by bile duct cancer.

- **Laparoscopy** – surgery to look at the organs inside the abdomen to check for signs of disease. A small lighted tube (scope) is inserted through a small incision in the abdomen. Tissue samples (biopsies) may be taken through the scope. A laparoscopy helps determine if the cancer can be removed surgically or if it has spread to other organs in the abdomen.

**Stages of Bile Duct Cancer**

**Stage 0 (Carcinoma in situ)** – cancer is found only in the innermost layer of cells lining the bile duct.

**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** 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 branches of the hepatic artery or to the right or left branches 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** – cancer has spread:
- to the 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; or
- perhaps to nearby lymph nodes.

**Stage IV** – cancer has spread to lymph nodes and/or organs far away from the bile duct.

**Methods of Treatment**

**Surgery**

These types of surgery are used to treat 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 examined under the 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 (margin) around it.

- **Whipple procedure** - A surgery 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 produce 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.

**Treatment Groups**

- **Localized (and resectable)** – the cancer is in a limited area and can be completely removed by surgery.

- **Unresectable** – the cancer has spread to nearby lymph nodes, blood vessels or other organs and cannot be removed completely by surgery.
• **Stent placement** – If the tumor is blocking the bile duct, a stent (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. A stent may be placed during an ERCP, a PTC, or surgery.

**Radiation Therapy**

Radiation therapy is a cancer treatment that uses high-energy x-rays or other types of radiation to kill cancer cells. There are two types of radiation therapy. **External beam** radiation uses a machine outside the body to send radiation toward the cancer. **Internal beam** radiation 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 uses drugs to stop the growth of cancer cells, either by killing the cells or by stopping the cells from dividing. Unlike surgery and radiation therapy, chemotherapy is a systemic treatment that can reach cancer cells throughout the body. Chemotherapy is sometimes used along with radiation therapy to make the radiation therapy more effective.

**Clinical Trials**

Clinical trials, exploring ways of improving local control may be available using chemotherapy with or without radiation.

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