

Bile Duct Cancer (Cholangiocarcinoma)

Bile duct cancer is a cancer which arises in the bile ducts of the liver. It is a relatively uncommon cancer in the USA. Most cases are sporadic. Surgery to remove the cancer and in most cases followed by chemotherapy and radiation therapy offers the best chance of a cure.

What is the bile duct ?

The bile ducts begin within the liver and join together to form larger bile ducts. The bile ducts are the tubes connecting the liver and gallbladder to the small bowel. The function of the bile duct is to carry bile from the liver to the gallbladder and then to the intestines. Bile is a digestive fluid that helps to break down fats. It is made by the liver and stored in the gallbladder. It then passes from the gallbladder into the intestine to help break down fatty food.

What is cancer?

Cancer is a disease of the cells in the body. The body is made up of millions of tiny cells. There are many different types of cells in the body, and there are many different types of cancer which arise from different types of cells. What all types of cancer have in common is that the cancer cells are abnormal.

A cancerous or malignant tumor is a 'lump' or 'growth' of tissue made up of cancer cells which continue to multiply. As they grow, malignant tumors can invade into nearby tissues and organs.

Malignant tumors may also spread to other parts of the body. This happens if some cells break off from the first (primary) tumor and are carried in the bloodstream or lymph channels to other parts of the body. These small groups of cells may then multiply to form 'secondary' tumors (metastases) in one or more parts of the body. These secondary tumours may then grow and invade nearby tissues, and spread again.

Some cancers are more serious than others, some are more easily treated than others (particularly if diagnosed at an early stage), some have a better outlook (prognosis) than others.

In each case it is important to know what type of cancer has developed, where it has developed, how large it has become, and whether it has spread. This will enable you to get information on treatment options.

What is bile duct cancer?

Bile duct cancer begins in the bile duct tissue and is known as adenocarcinoma. If the cancer starts in the part of the bile duct inside the liver, it's known as intra-hepatic bile duct cancer. This type of bile duct cancer typically will form a tumor in the liver and is discussed with the primary liver cancers. If, on the other hand, the cancer begins in the area of the bile ducts outside the liver, then it's known as extra-hepatic bile duct cancer.

What causes extra-hepatic bile duct cancer?

Although it is not fully understood why bile duct cancer develops, there are certain factors that make bile duct cancer more likely.

- Age - it's most common around age 65.
- Primary sclerosing cholangitis is a condition in which inflammation of the bile duct (cholangitis) leads to the formation of scar tissue (sclerosis) and an increased risk of bile duct cancer. The cause of the inflammation is not usually known, but it may or may not occur with ulcerative colitis.
- Choledochal cysts are dilations of the bile duct. The cells lining the dilations may have areas of pre-cancerous changes, which increase a person's risk for developing bile duct cancer.
- Chronic gallstones or bile duct stones which can cause inflammation.
- Exposure to certain chemicals used in the metal or rubber industry.
- Infection with a parasite called the liver fluke (*Clonorchis sinensis* or *Opisthorchis viverrini*) is associated with an increased risk of bile duct cancer. They are found in South Asia and Africa.
- Family history of bile duct cancer seems to increase a person's chance of developing this cancer, but the overall risk is still low. Despite this, most bile duct cancers are not found in people with a family history of the disease.
- Smoking.

What are the symptoms of extra-hepatic bile duct cancer?

As opposed to many cancers, extrahepatic bile duct cancers may cause symptoms early because of obstruction of the bile duct. The first symptoms to develop may include:

- Yellowing of the skin and the white of the eyes (known as jaundice)
- Dark yellow urine
- Pale colored stool
- Mild pain in the upper abdomen
- Loss of appetite or feeling sick
- Weight loss and tiredness
- Itchy skin

How is cancer of the extra-hepatic bile duct diagnosed ?

The extra-hepatic bile duct is located deep inside the body, so early tumors cannot be seen or felt by health care providers during routine physical exams. Most bile duct cancers are found only when the cancer has grown enough to cause symptoms. The most common symptom is jaundice, a yellowing of the skin and eyes, which is caused by a blocked bile duct

Thus, extra-hepatic bile duct cancer is typically diagnosed by a combination of physical examination and imaging tests.

- Imaging such as ultrasound scan, CT scan or MRI/MRCP. These can help to show the location and extent of the cancer.

- An endoscopic ultrasound scan (EUS) - an endoscope with an ultrasound probe is used to check the bile duct, pancreas and the surrounding tissues.
- Endoscopic retrograde cholangio-pancreatography (ERCP) - images of the pancreatic duct and the bile duct are taken using an endoscope (a camera attached to a flexible tube) passed down through the mouth to look inside.
- Percutaneous transhepatic cholangiography (PTC) - a needle is passed through the skin to the liver and a dye is injected into the bile duct and X-ray image is taken. For this procedure a local anesthetic is used.

In some cases, the doctor may also need to do a biopsy, but in general, a biopsy is not required before surgical removal if the lesion is resectable and the suspicion of cancer is high. Talk to your doctor.

What are the treatment options ?

Surgical resection (removal of the cancer) may be done to remove the cancer if it hasn't spread beyond the bile duct (resectable). If the tumor is resectable, surgery is main-stay treatment, as it offers the only reasonable chance to cure the cancer. The extent of surgery depends on the location of the tumor, but in general involves removal of the bile duct and reconstruction. Surgery may also include removal of a portion of liver.

Radiation therapy and/or **chemotherapy** may be added after surgery in some cases, or they may be used instead of surgery if the cancer cannot be entirely removed. In some centers, liver transplant may be an option.

Palliative : If it's not possible to remove the cancer, a good option is stent placement (a small hollow tube) in the bile duct to help relieve symptoms of jaundice. This allows the bile to flow from the liver to the small bowel. A stent is fitted into the bile duct to allow bile to drain away.

A **stent insertion** may be used to help without having to perform surgery. This can be done using ERCP, but more commonly, a PTC procedure may be performed to insert a stent through the skin into the bile duct.

What is the prognosis (outlook)?

The best chance of a cure is with surgery when the cancer is completely removed and has not spread from the primary site of the bile duct. However, if this is not possible, treatment with chemotherapy with or without radiation therapy is the next option.

Sources : www.ncbi.nlm.nih.gov/pubmedhealth
www.bupa.co.uk/individuals/health-information
www.cancer.org

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