

Information for people with cancer

This information should not be used to diagnose yourself or in place of a qualified doctor's care.

The bile duct is a 10-13 cm (4-5 inch) long tube. It carries bile from the liver to the gallbladder and then to the small intestine.

Bile is a fluid made in the liver. It helps digest fats in the small intestine. Bile is stored in the gallbladder until it is needed in the small intestine.

Bile duct adenocarcinoma is also called cholangiocarcinoma.

## **Diagnosis and Staging**

### What are the signs and symptoms of bile duct cancer?

- Jaundice (when your skin and whites of your eyes turn yellow)
- Itchy skin
- Light or clay-coloured stools (poop)
- Weight loss you cannot explain
- Loss of appetite
- Fever
- Fatigue
- Abdominal (belly) pain
- Nausea (feeling queasy) or vomiting

If you have any signs or symptoms that you are worried about, please talk to your family doctor or nurse practitioner.



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## How is bile duct cancer diagnosed?

Tests that may help diagnose bile duct cancer include:

- **Physical examination:** your doctor may look for signs of jaundice. They may feel your abdomen for lumps, tenderness, fluid build-up or an enlarged liver.
- Imaging to check for bile duct cancer and to see if it has spread
  - Ultrasound
  - CT (computed tomography) scan
  - MRI (magnetic resonance imaging)
  - MRCP (magnetic resonance cholangiopancreatography): if your doctor thinks you have extrahepatic bile duct cancer.
- EUS (endoscopic ultrasound): a combination of an ultrasound and an endoscopy (when a long thin tube is put down your throat to look at your organs). This test can be used to do a biopsy of bile duct cells, check for bile duct stones or a tumour, and to check if cancer has spread to nearby lymph nodes.
- **Cholangiography:** dye is injected into your bile duct with a needle and x-ray pictures are taken. This can show where the cancer is.
- Endoscopic Retrograde Cholangiopancreatography (ERCP): used to check for extrahepatic bile duct cancer. A tube is put down your throat and into your bile duct. A dye is injected into the tube. The dye will show a narrowing or blockage of your bile duct. A brush can also be put into the tube to collect cells or tissue for a biopsy.
- Laparoscopy: surgery using a very small camera put into your body through your abdominal wall. The doctor will use the camera to look at your bile duct, liver and areas around them. This test can show how advanced the cancer is.
- Surgery or biopsy: may be needed to confirm cancer diagnosis.
- **Tumour marker tests:** A tumour marker is usually something made by a cancer cell or another cell when cancer is in your body. If you are diagnosed with bile duct cancer, your doctor may check for these tumour markers: CEA (carcinoembryonic antigen) and CA 19-9 (carbohydrate antigen 19-9).



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For more information on tests used to diagnose cancer, see our Screening and Diagnosis section: <u>bccancer.libguides.com/pathfinder-screening</u>

### What are the types of bile duct cancer?

Over 90% (90 out of 100) of bile duct cancers are adenocarcinomas. These cancers begin in the mucus glands lining the inside of the bile duct.

Bile duct adenocarcinoma is also called cholangiocarcinoma.

Other bile duct cancers include squamous cell carcinoma, lymphoma and sarcoma. The different types of cancer start in different cells.

Bile duct cancers either start in the liver or outside of the liver:

- **Extrahepatic** Cancer starts in the bile duct outside the liver. Extrahepatic bile duct cancers can be:
  - Perihilar Cancer starts in the hepatic duct where the bile ducts join just outside the liver. This is the most common place for bile duct cancer. It can also be called a hilar or Klatskin tumour.
  - **Distal** Starts in the area closest to the small intestine.
- Intrahepatic Cancer starts in the bile duct inside the liver. Only about 10% (10 out of 100) of bile duct cancers are intrahepatic.

### What are the stages of bile duct cancer?

Staging describes the cancer. Staging is based on how much cancer is in the body, where it was first diagnosed, if the cancer has spread and where it has spread to.

The stage of the cancer can help your health care team plan your treatment. It can also tell them how your cancer might respond to treatment and the chance that your cancer may come back (recur).



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#### Provincial Health Services Authority

The stage of the cancer is used to plan your treatment.

#### Bile duct cancer staging:

#### Perihilar extrahepatic bile duct cancer

Stage 0	The cancer is only in the innermost layer of the bile duct.
Stage 1	The tumour (cancer growth) is in the deeper layers of the bile duct.
Stage 2	The tumour has spread to surrounding fat or the liver.
Stage 3A	The tumour has spread into the portal vein or the hepatic artery on one side of the liver.
Stage 3B	The tumour has spread into the portal vein on both sides of the liver, the common hepatic artery, or other bile ducts on one side of the liver and a main blood vessel on the other side of the liver.
Stage 4A	The cancer has spread to 4 or more lymph nodes.
Stage 4B	The cancer has spread to other parts of the body.

#### Distal extrahepatic bile duct cancer

Stage 0	The tumour is only in the inner layer of the bile duct.
Stage 1	The tumour has grown less than 5 mm into the bile duct wall.
Stage 2A	The tumour has grown less than 5 mm into the bile duct wall and has spread to 1-3 nearby lymph nodes, or it has spread 5-12 mm into the bile duct wall.
Stage 2B	The tumour has grown 5-12 mm into the bile duct wall and has spread to 1-3 nearby lymph nodes, or it has spread more than 12 mm into the bile duct wall.



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Stage 3A	The tumour has spread into the bile duct wall and has spread to 4 or more nearby lymph nodes.
Stage 3B	The tumour has spread into the blood vessels in the abdomen.

**Stage 4** The cancer has spread to other parts of the body.

#### Intrahepatic bile duct cancer

Stage 1A	The tumour is only in the bile duct and is 5 cm or smaller.
Stage 1B	The tumour is only in the bile duct and is larger than 5 cm.
Stage 2	One or more tumours have grown into blood vessels in the liver.
Stage 3A	The tumour has spread through the membrane that covers and supports the liver (visceral peritoneum).
Stage 3B	The tumour has spread into surrounding tissues outside the liver or the cancer has spread to nearby lymph nodes.
Stage 4	The cancer has spread to other parts of the body.

### What are the grades of bile duct cancer?

The grade of the cancer describes how different the cancer cells look from normal cells and how fast the cancer cells are growing. A pathologist will give the cancer a grade after looking at the cells under a microscope.

Bile duct cancer can be grade 1, 2, 3 or 4. The lower the number, the lower the grade.

Low grade: cells are abnormal but look a lot like normal cells. Low grade cancers usually grow slowly and are less likely to spread.



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**High grade:** cells are abnormal and do not look like normal cells. High grade cancers usually grow more quickly and are more likely to spread.

The grade of the cancer helps your health care team plan your treatment.

## Treatment

#### What is the treatment for bile duct cancer?

Cancer treatment may be different for each person. It depends on your particular cancer. Your treatment may be different from what is listed here.

#### Surgery

- If possible, you will have surgery to remove the cancer.
- Surgery may include:
  - Removing the bile duct
  - Whipple procedure for distal bile duct tumours. This is a surgery that removes part of the pancreas, part of the small intestine, the gall bladder and the bile duct.
  - Hepatectomy (removing part of the liver) for cancer in or near the liver
  - Removing nearby lymph nodes
- Some patients may be able to have a liver transplant. This surgery is only for certain patients and not everyone can have this treatment.

**Radiation therapy** (high energy x-rays that kill or shrink cancer cells)

- Sometimes given with systemic therapy to kill any cancer cells that might be left after surgery. This is called adjuvant therapy.
- Can also help with symptoms such as pain, bleeding or obstruction (blockage).
- For more information about radiation therapy go to:
  bccancer.bc.ca/our-services/treatments/radiation-therapy



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#### Systemic therapy (chemotherapy)

- Sometimes given after surgery to kill any cancer cells that are left (adjuvant therapy).
- Can be used to shrink the tumour before surgery.
- For more information about systemic therapy go to: <u>bccancer.bc.ca/our-services/treatments/systemic-therapy-(chemotherapy)</u>

#### Advanced bile duct cancer

- Palliative systemic therapy can help some patients with advanced disease live longer. It may also help with symptoms.
- Please see the resources available for patients with advanced cancer: <u>www.bccancer.bc.ca/our-services/services/supportive-care/pain-symptom-management#Advanced--cancer</u>

### What is the follow-up after treatment?

- Follow-up testing and appointments are based on your type of cancer.
- Guidelines for follow-up after treatment are on our website: <u>www.bccancer.bc.ca/health-professionals/clinical-resources/cancer-management-manual/gastrointestinal/bile-duct</u>
- These are guidelines written for your doctor, nurse practitioner or specialist. You can look at them to see what appointments and tests you might need after treatment.
- After treatment, you may return to the care of your family doctor or specialist for regular follow-up. If you do not have a family doctor, please speak with your BC Cancer health care team.
- The BC Cancer Life after Cancer page has information on the issues that cancer survivors may face: <a href="https://www.bc.ca/lifeaftercancer">bc.ca/lifeaftercancer</a>



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## **More Information**

### What causes bile duct cancer and who gets it?

These are the risk factors for this cancer. Not all of these risk factors may cause this cancer, but they may help the cancer start growing.

- Being over 50 years of age.
- Having a disease such as primary sclerosing cholangitis, inflammatory bowel disease (e.g. ulcerative colitis), and choledochal cysts.
- Infection with liver flukes (parasites that live in the bile duct). Liver fluke infections are more common in some parts of Asia.
- Infection with hepatitis C virus, possibly hepatitis B virus.
- Exposure to hazardous materials, such as thorium dioxide and 1,2-dichloropropane.
- Diabetes
- Cirrhosis (damage to the liver)
- Being overweight
- Genetic disorders such as Lynch syndrome or cystic fibrosis

Other possible risk factors are:

- Drinking alcohol
- Smoking
- Chronic bile duct stones

#### Statistics

We do not have specific statistics on bile duct cancer. Bile duct cancer is included in "All Other Cancers" on our statistics page: <u>bccancer.bc.ca/health-info/disease-system-statistics/statistics-by-cancer-type</u>



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**Note:** Available statistics do not have information about the inclusion of transgender and gender diverse participants. It is unknown how these statistics apply to transgender and gender diverse people. Patients are advised to speak with their primary care provider or specialists about their individual considerations and recommendations.

### Can I help prevent bile duct cancer?

- Try not to drink too much alcohol: Not drinking at all will lower your cancer risk the most.
  - Visit the BC Cancer Alcohol and Cancer Prevention page: <u>www.bccancer.bc.ca/health-info/prevention/alcohol/reduce-your-risk</u>
- Hepatitis B may lead to cirrhosis: You should make sure your vaccinations are up to date. Learn more about Hepatitis B vaccination on the Immunize BC website: <u>immunizebc.ca/hepatitis-b</u>
- **Travel safely:** If you are travelling in parts of the world where liver flukes are common, drink only purified water and choose foods that have been well-cooked.
- **Stop smoking**: Do not smoke and try not to be around tobacco and cigarette smoke. Even if you have been using tobacco for many years, quitting now will lower your risk of getting cancer. Support is available to help you quit smoking.
  - Visit the BC Cancer Tobacco and Cancer Prevention page: <u>www.bccancer.bc.ca/health-info/prevention/tobacco</u>
- Maintain a healthy weight:
  - Visit the BC Cancer Body Weight and Cancer Prevention page: <u>www.bccancer.bc.ca/health-info/prevention/body-weight</u>
- Avoid exposure to hazardous chemicals: Follow the safety instructions on the container when working with chemicals.



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#### Is there screening for bile duct cancer?

There is no screening for this type of cancer.

### Where can I find more information?

- If you have questions about bile duct cancer, please talk to your health care team.
- Our librarians can help you find the information you need. Visit our Library page: <u>bccancer.bc.ca/our-services/services/library</u>
- BC Cancer Library Bile Duct Cancer pathfinder: <u>bccancer.libguides.com/pathfinder-bile</u>
- Managing Your Symptoms: <u>www.bccancer.bc.ca/health-info/coping-with-</u> <u>cancer/managing-symptoms-side-effects</u>
- Emotional Support: <u>www.bccancer.bc.ca/health-info/coping-with-cancer/emotional-support</u>