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mychcc.ca

## Cholangiocarcinoma (CCA) Bile Duct Cancer



### What do the bile ducts do?

The bile ducts are the tubes that connect the liver and gall bladder to the small intestine. Bile is produced in the liver and the bile ducts allow bile to be delivered from the liver and gall bladder to the small bowel to aid in fat digestion.

### What is cholangiocarcinoma?

This is a cancer of the bile ducts.

There are three different types of cholangiocarcinoma (CCA), based on location:

1. **Intrahepatic cholangiocarcinoma (iCCA)**
  - Found in bile ducts within the liver
2. **Extrahepatic or distal cholangiocarcinoma (dCCA)**
  - Found in the part of the bile ducts outside the liver
3. **Hilar cholangiocarcinoma or Klatskin's tumor (pCCA)**
  - Found where the right and left bile ducts join

## Signs and Symptoms

- ✓ Jaundice (yellowing of the skin and eyes)
- ✓ Dark urine
- ✓ Clay coloured stool
- ✓ Pain in the abdomen
- ✓ Itchy skin
- ✓ Fever
- ✓ Nausea and vomiting
- ✓ Fatigue
- ✓ Weight loss

## Risk Factors

- ✓ Most CCA occur without a distinguishing cause
- ✓ Primary sclerosing cholangitis (PSC)
  - An autoimmune disorder
- ✓ Primary biliary stones (gallstones)
- ✓ Chronic viral infection with Hepatitis B or C
- ✓ Ulcerative colitis
- ✓ Liver flukes (parasitic worms)
- ✓ Diabetes
- ✓ Chemical exposure
- ✓ Congenital conditions:
  - Abnormal joining of the bile and pancreatic ducts and Choledochal cysts
  - Abnormal dilations of the bile ducts outside the liver (rare)

## Associated with cholangiocarcinoma

- ✓ Obesity
- ✓ Alcohol consumption
- ✓ Tobacco smoking
- ✓ Cirrhosis of the liver
- ✓ MASLD (Metabolic Dysfunction-Associated Steatotic Liver Disease)
- ✓ Metabolic syndrome

## Clinical Trials

- ✓ CAPTUR website (Canadian)
  - <https://clinicaltrials.gov/ct2/show/NCT03297606>
- ✓ Website for clinical trials world wide
  - <https://www.clinicaltrials.gov>

## Prognosis

- ✓ Aggressive cancer with a median survival rate of 24 months
- ✓ The only cure at this time is totally resectable tumor
- ✓ Globally, the survival rate is 10%

## Prevention

- ✓ Stop smoking
- ✓ Reduce the risk of liver disease:
  - Drink alcohol in moderation (0-2 drinks per week)
- ✓ Maintain a healthy weight
- ✓ Cook meat thoroughly
- ✓ Follow directions when working with chemicals

## References

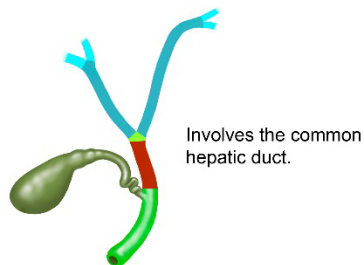
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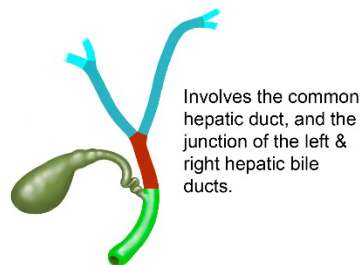
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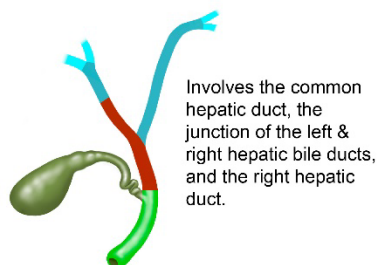
## Perihilar Cancers Bismuth-Corlette Classification of Types



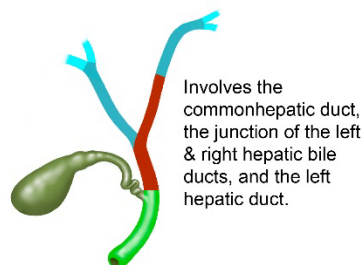
**Type I**



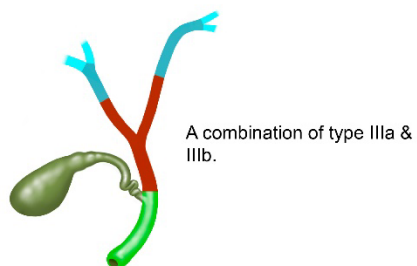
**Type II**



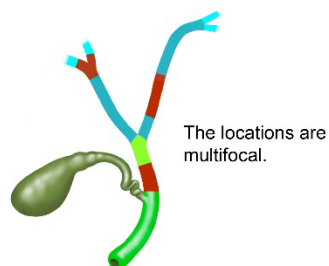
**Type IIIa**



**Type IIIb**



A combination of type IIIa & IIIb.



The locations are multifocal.

**Type IV**  
Can be either of these

## Diagnosis

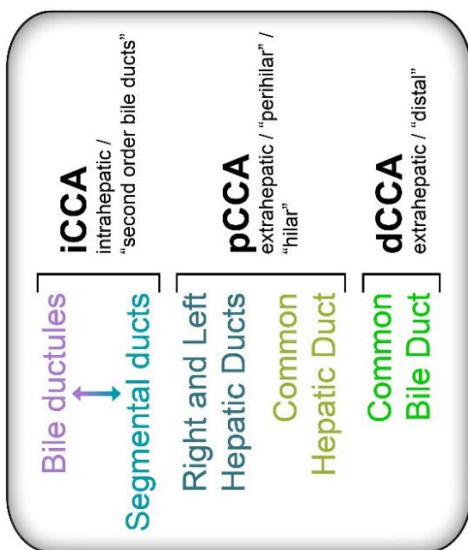
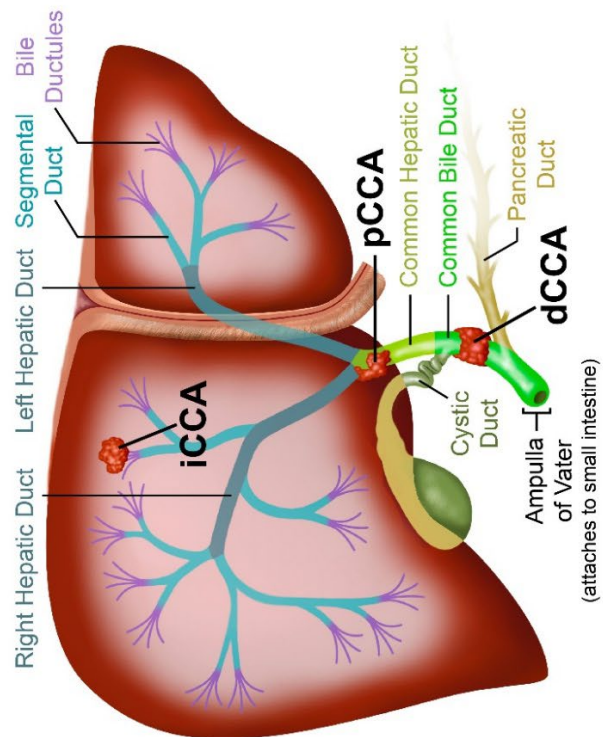
- ✓ Blood chemistry tests
- ✓ Tumor marker tests
  - CEA and CA-19-9
- ✓ Biopsy
- ✓ Ultrasound
- ✓ CT scan
- ✓ Laparoscopy
- ✓ Biomarkers
- ✓ ctDNA liquid biopsy

## Treatment

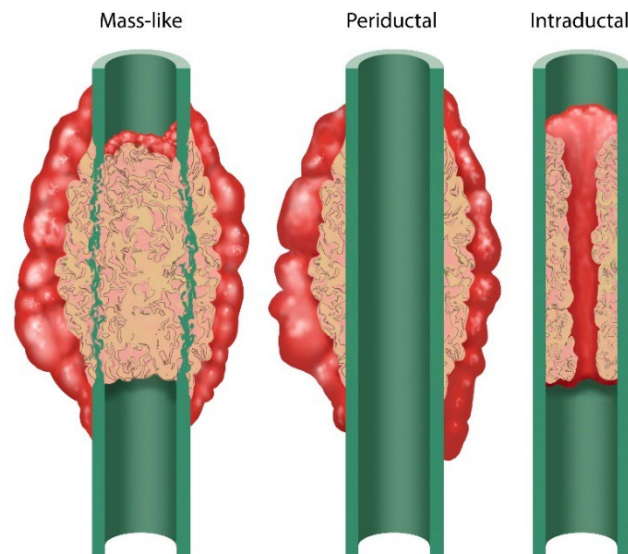
Discuss options with your physician.

- ✓ Surgery:
  - Removal of part of the bile duct if the tumor is small or partial hepatectomy or Whipple procedure
- ✓ Adjuvant therapy after surgery:
  - Chemotherapy or radiation therapy to lower risk of cancer returning
- ✓ Palliative surgery:
  - Biliary bypass or Endoscopic stent placement
- ✓ Radiation therapy
- ✓ Chemotherapy used drugs to stop the growth of cancer cells
- ✓ Regional chemotherapy
- ✓ Immunotherapy
- ✓ Targeted therapy
- ✓ Histotripsy

## Cholangiocarcinoma (CCA)



## Morphologic Classification of Biliary Tumors



## Biliary System

