

# Cholangiocarcinoma

# Cholangiocarcinoma

## Pathology

Bile duct epithelial cells

## Anatomy:

Cholangiocarcinoma can occur

- 1) INTRA- hepatic
- 2) EXTRA- hepatic
- 3) Gallbladder
- 4) Peri-Hilar

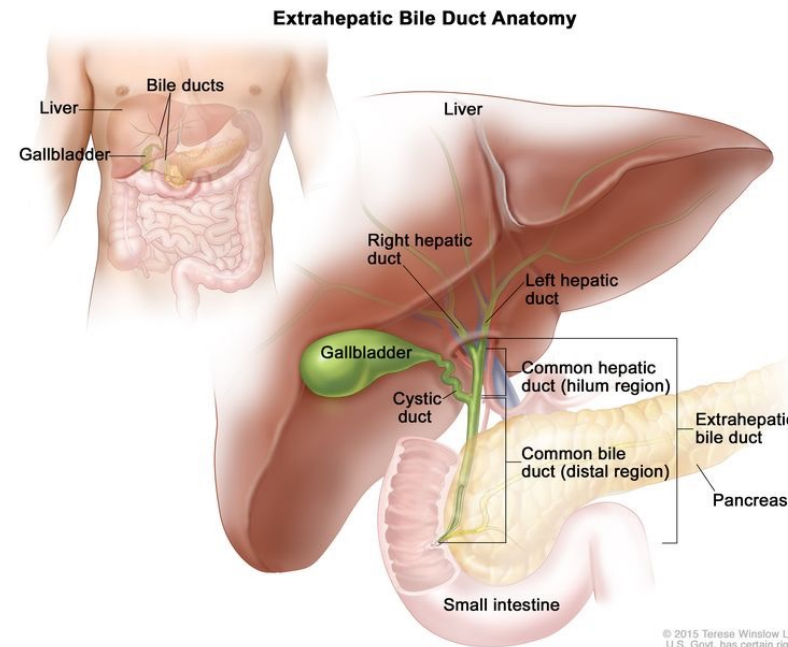
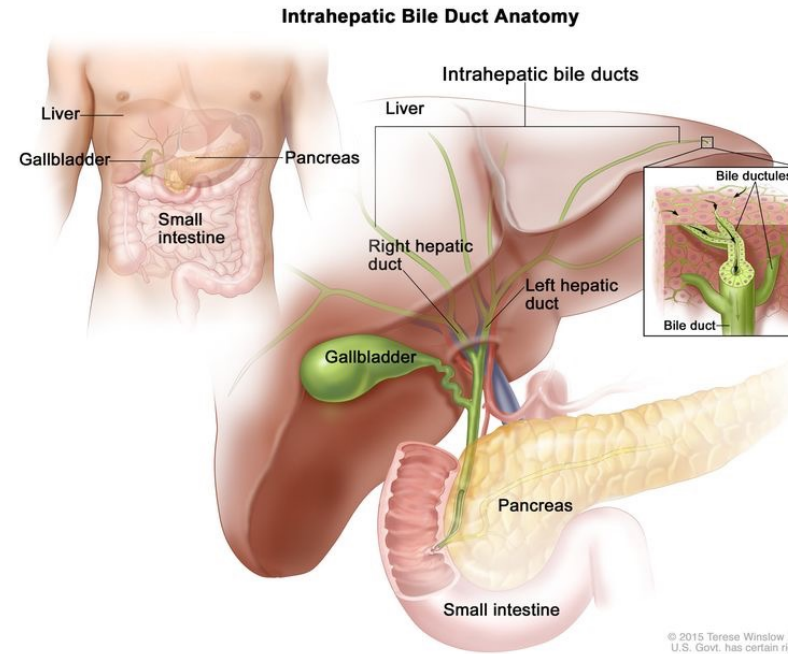
## Work Up:

### Imaging

Multiphasic CT/MRI  
MRCP (extrahepatic)  
CT Chest  
+/- EUS

### Labs

CA19-9, CEA  
AFP (intrahepatic)



# Cholangiocarcinoma

## Clinical Risk Factors

### Risk Factors

- Gallstones
- Porcelain gallbladder
- DM
- Obesity
- Smoking
- Women > Men
- Infections (HCV > HBV, Salmonella, Liver Fluke)

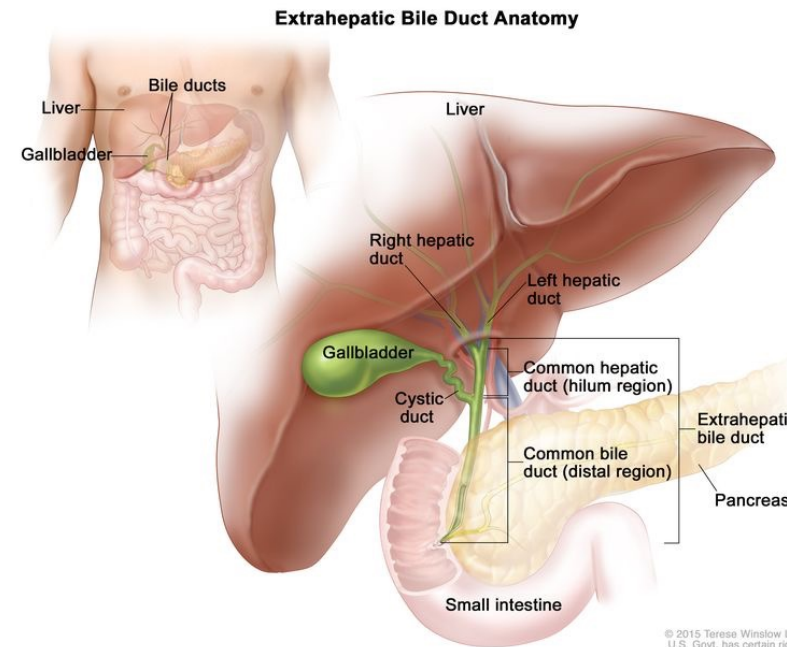
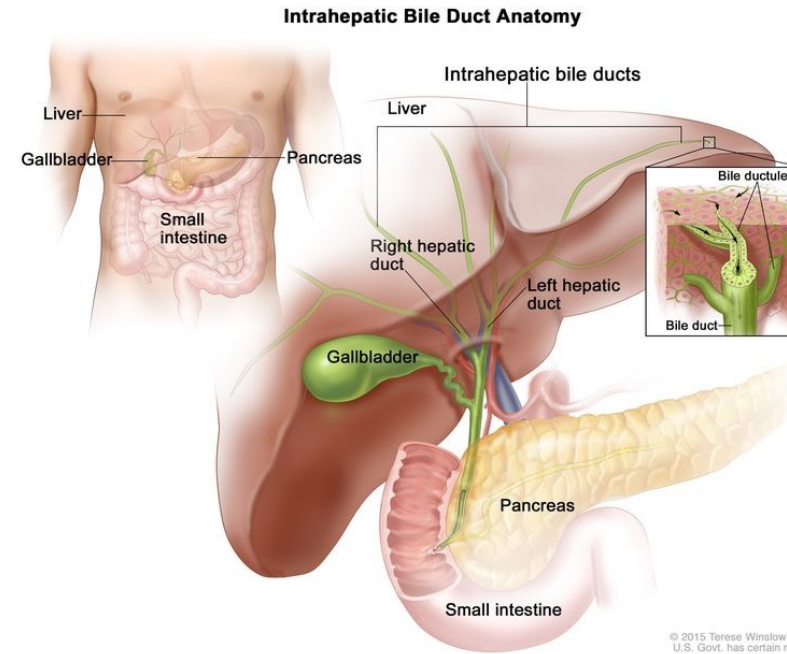
## Prognosis

Early Stage

5Y OS 25-65%

Stage IV

5Y OS <5%



# Cholangiocarcinoma

## Staging

### T1

- T0 = carcinoma in situ
- T1 = submucosa
- T2 = muscularis propria
- T3 = serosa
- T4 = major blood vessels: portal vein or hepatic artery, extra-hepatic structures

### N

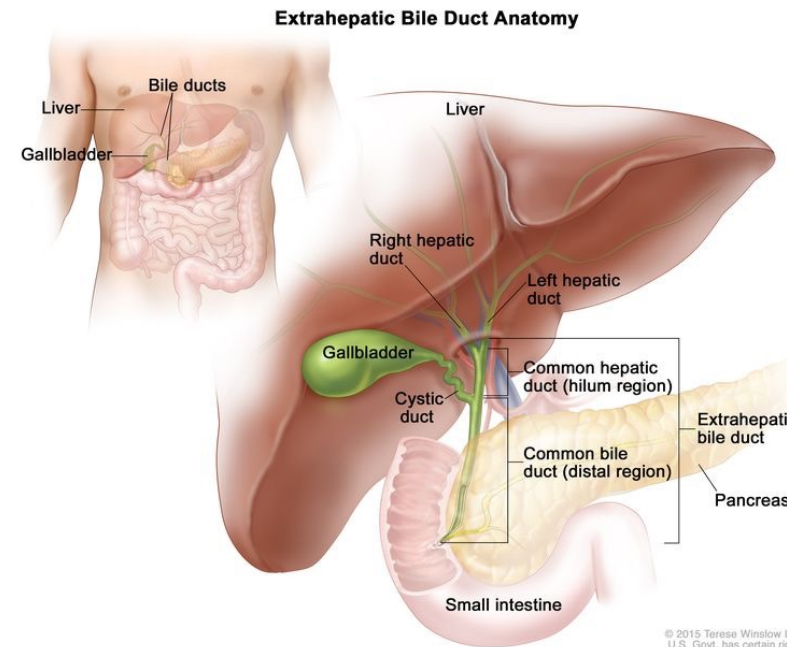
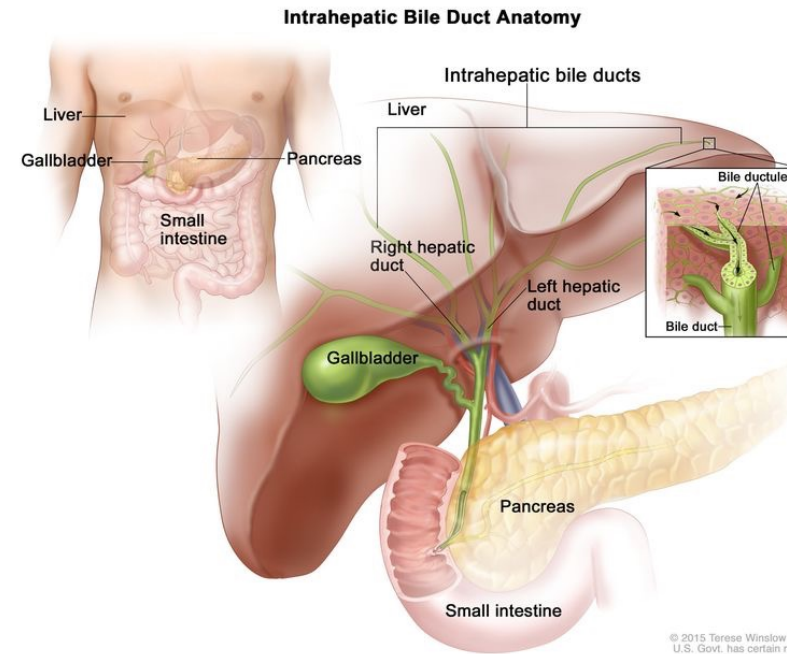
- N1 = 1-3 LN (includes “tumor deposits”)
- N2 = 4+ LN

### M

- M1 = distant mets, includes peritoneum

## Stage

- Stage I-II = T1-2N0
- Stage III = T3N0 or T1-3N1
- Stage IV = T4 or N2



# Cholangiocarcinoma: Early Stage Treatment

**Resectable**

## Surgery

**T0-T1a (carcinoma in situ, invasion into lamina propria alone)**

= cholecystectomy/partial hepatectomy alone is sufficient  
+ observation

**T1b+/T2 (invasion beyond lamina propria)**

= en bloc resection of primary gallbladder/liver tumor + bile duct excision + LND  
+ adjuvant therapy

\* gallbladder tumors require partial hepatic resection to ensure clear margins

# Cholangiocarcinoma: Early Stage Treatment

## Resectable

### Surgery

resection of primary gallbladder/liver tumor  
+ bile duct excision  
+ LND

### Hilar Cholangiocarcinoma, node negative

Consider neoadjuvant chemo → transplant

## Adjuvant Therapy or Unresectable

Consider in T2+, +LN, R1 resection

1. **Observe if:** < T2, N0, R0
2. **Adjuvant if:** > T2, N+, R+

Chemo

ChemoRT

Clinical Trial

### Options for Chemo/ChemoRT

Capecitabine +/- Gemcitabine

5FU +/- RT

# Cholangiocarcinoma: Metastatic Treatment

## Targeted Therapy for Special Populations

### Front Line

**Gemcitabine/Cisplatin**  
+/- Durvalumab or Pembrolizumab

### Second Line

**5FU or Gemcitabine-based chemo**  
FOLFOX/FOLFIRI  
GEMOX  
CAPOX

**Pembrolizumab + Lenvatinib**

#### **FGFR2**

Futibatinib  
Pemigatinib

#### **IDH1**

Ivosidenib

#### **MSI-High/dMMR**

Pembrolizumab (TMB >10 mut/mb)

#### **BRAF V600E**

Dabrafenib/Trametinib

#### **NTRK**

Entrectinib  
Larotrectinib

#### **HER2**

Trastuzumab/Pertuzumab  
Zanidatamab no FDA approval yet

# **Cholangiocarcinoma Reference Handout**



## Cholangiocarcinoma Dx

### Bile Duct Epithelial Cells

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### Work Up

Multiphasic MRI/CT  
CT Chest

+/- MRCP or EUS

Labs: CEA, CA19-9, AFP

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## Cholangiocarcinoma Early Stage Tx

### Resectable

#### Surgery

#### T0-T1

= cholecystectomy/partial hepatectomy

#### T2+

= en bloc resection of gallbladder/liver

+ bile duct excision

+ LND

### Adjuvant Therapy or Unresectable

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ChemoRT

Clinical Trial

#### Options for Chemo/ChemoRT

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5FU

## Cholangiocarcinoma Metastatic Tx

### Front Line

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+/- Durvalumab or Pembrolizumab

### Second Line

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Pembrolizumab + Lenvatinib

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