Perihilar Cholangiocarcinoma

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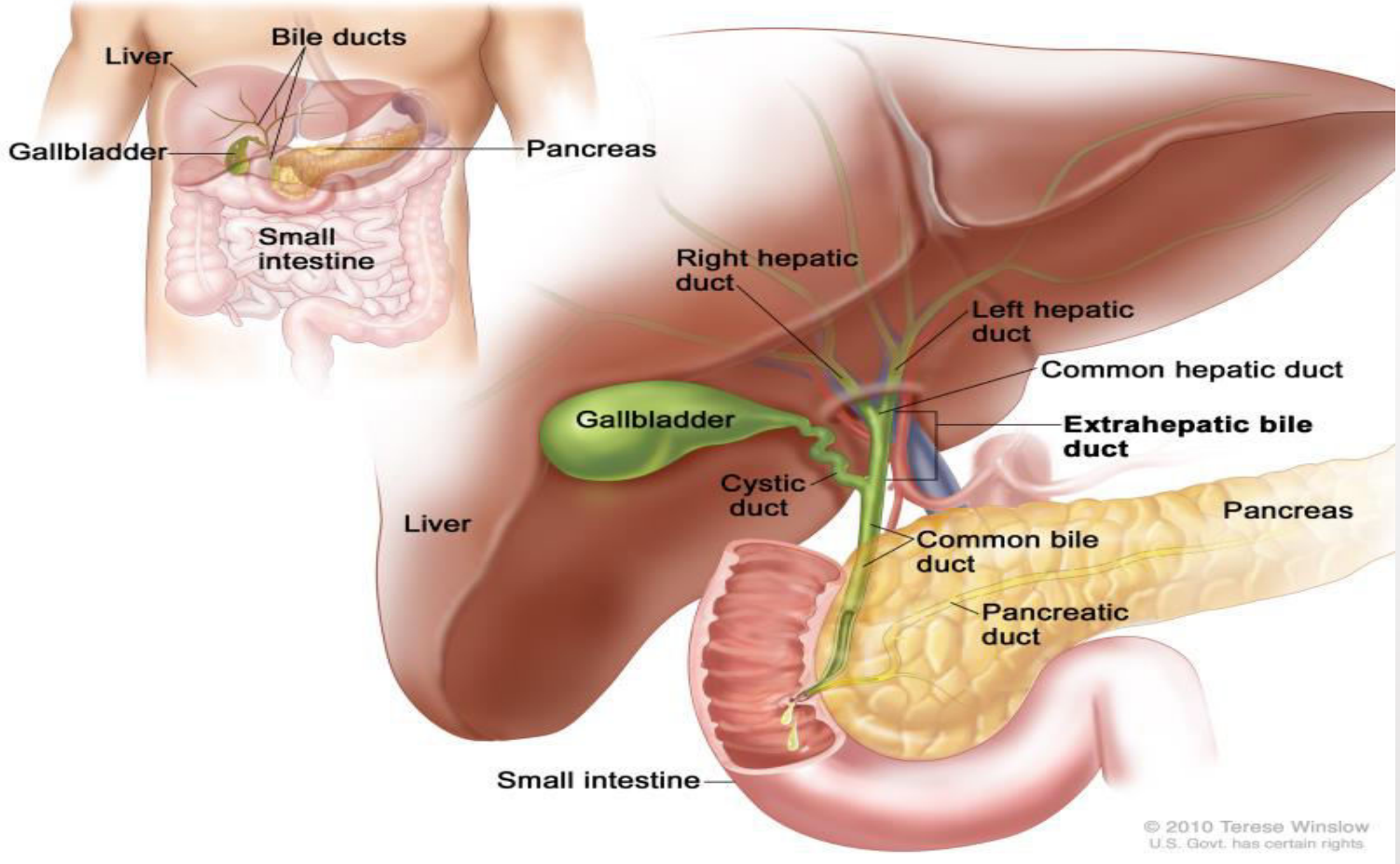
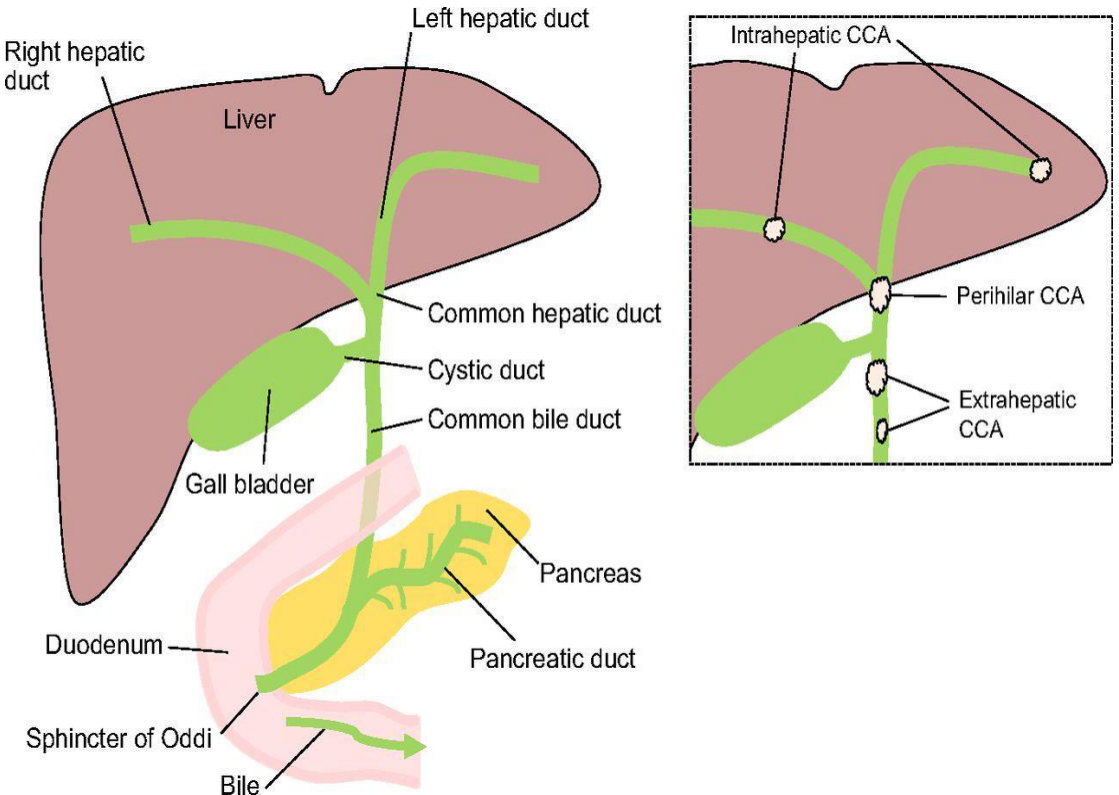
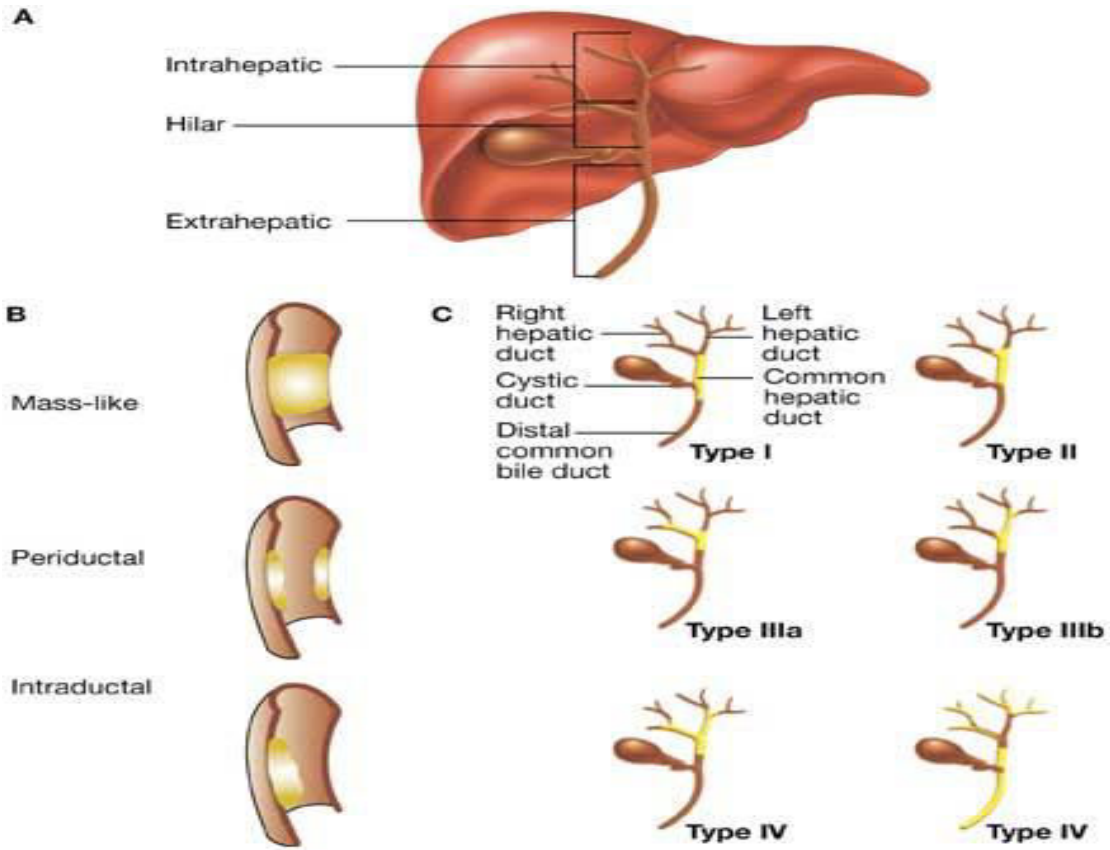


Definition of cholangiocarcioma

- are malignant tumors arising from the biliary tract
- the second most common primary malignancy of the liver after hepatocellular carcinoma
- it is classified as intrahepatic (IHCC), arising in the liver, or extrahepatic (EHCC), arising outside the liver.



IHCC is subcategorized morphologically into mass-forming, periductular-infiltrating or intraductal, the latter being the least common but with a more favourable prognosis; however, IHCC tumours can possess a combination of these characteristics



Incidence.

The incidence of bile duct carcinoma is about 0.3%. The overall incidence of cholangiocarcinoma in the United States is about 1.0 per 100,000 people per year,

With about 3000 new cases diagnosed annually.

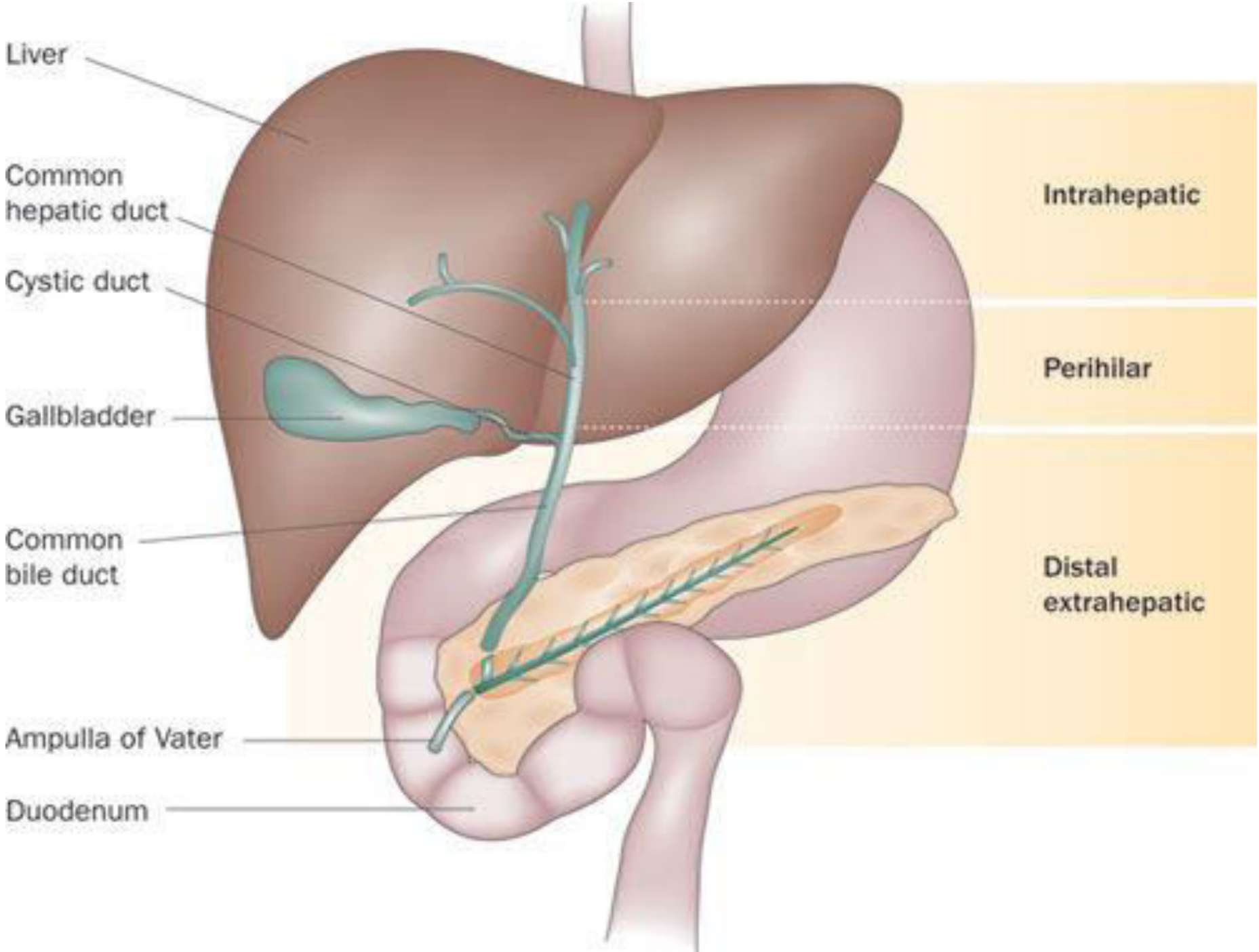
The male-to-female ratio is 1.3:1, and the average age of presentation is between 50 and 70 years.

Perihilar cholangiocarcinoma

It is a tumor of extrahepatic biliary tree occur at the confluence of right & left hepatic duct to the upper part of the pancreas.

Perihilar tumors are the most common and intrahepatic tumors are the least common.

Perihilar tumors, also called **Klatskin tumors**


Liver
Common hepatic duct
Cystic duct
Gallbladder
Common bile duct
Ampulla of Vater
Duodenum
Intrahepatic
Perihilar
Distal extrahepatic

Etiology.

Risk factors associated with cholangiocarcinoma Include

- primary sclerosing cholangitis,
- choledochal cysts,
- hepatolithiasis,
- biliary-enteric anastomosis,
- and biliary tract infections with *Clonorchis* or in chronic typhoid carriers.

Pathology :

Over 95% of bile duct cancers are adenocarcinomas.

Morphologically, they are divided into nodular (the most common type), scirrhous, diffusely

infiltrating, or papillary.

Perihilar cholangiocarcinomas, are further classified based on anatomic location by the Bismuth-Corlette classification.

Type I tumors are confined to the common hepatic duct

below the level of the confluence of the right and left hepatic ducts

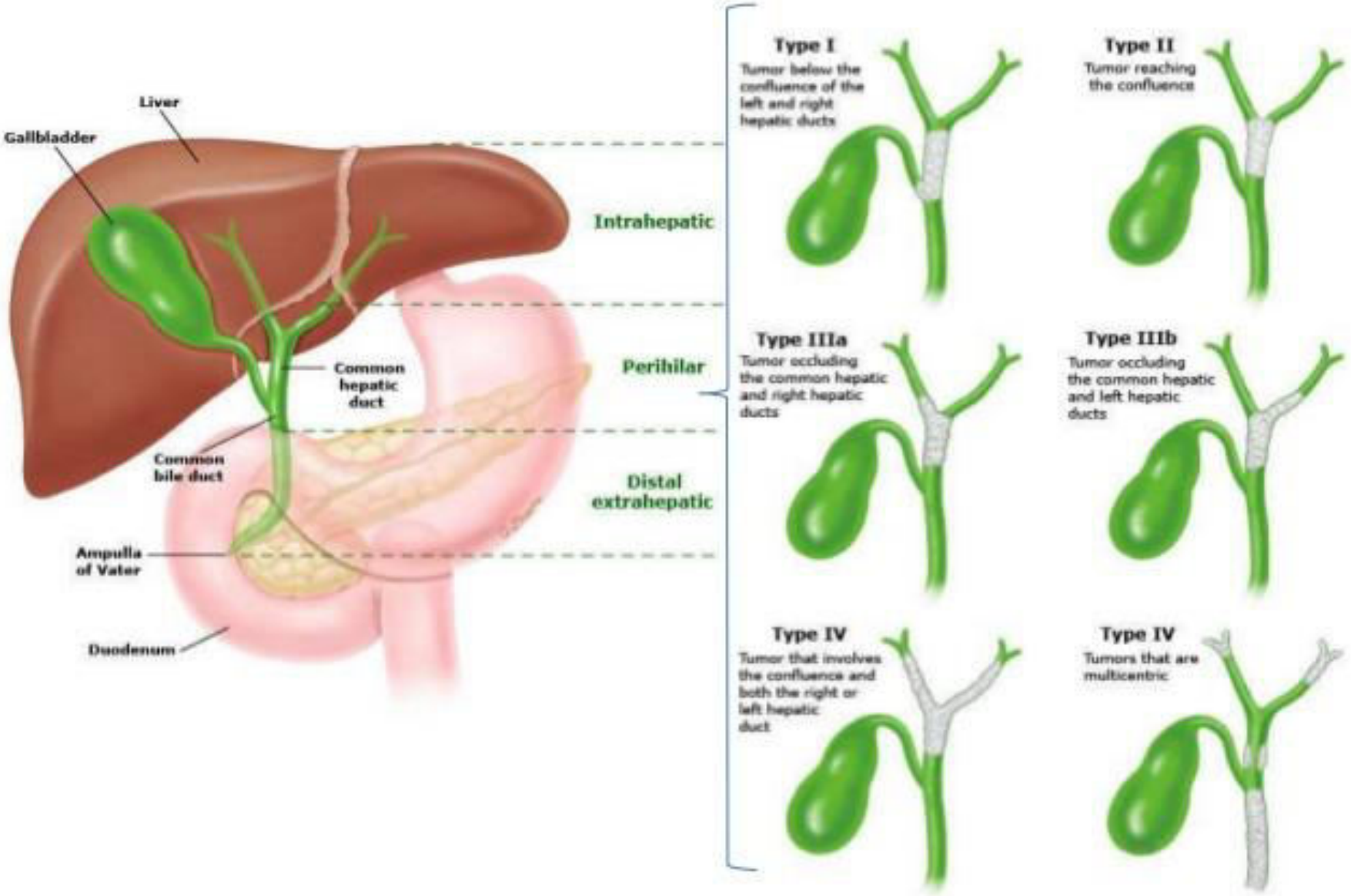
, but type II tumors involve the bifurcation without involvement of the secondary intrahepatic ducts.

Type IIIa type II and extends to the bifurcation of the right hepatic duct

and IIIb type II and extends to the bifurcation of the left hepatic duct

. Type IV tumors involve both the right and left secondary intrahepatic ducts

Bismuth-Corlette classification



Diagnosis :

Symptoms associated with cholangiocarcinoma are

- ❑ Asymptomatic :
- ❑ Obstructive jaundice :

Jaundice is the most common manifestation of bile duct cancer and, in general, is best detected in direct sunlight. The obstruction and subsequent cholestasis tend to occur early if the tumor is located in the common bile duct or common hepatic duct. Jaundice often occurs later in perihilar and is often a marker of advanced disease. The excess of conjugated bilirubin is associated with bilirubinuria and acholic stools.

- Clay-colored stools
- Bilirubinuria (dark urine)
- Pruritus

usually is preceded by jaundice, but itching may be the initial symptom of cholangiocarcinoma. Pruritus may be related to circulating bile acids

- Weight loss

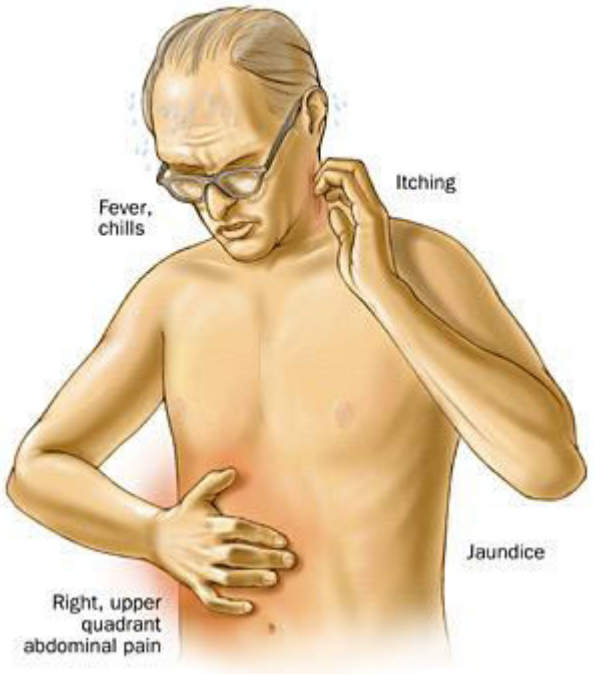
is a variable finding. It may be present in one third of patients at the time of diagnosis.

- Abdominal pain

is relatively common in advanced disease. It often is described as a dull ache in the right upper quadrant.

Signs :

- Jaundice
- Hepatomegaly
- Palpable upper quadrant mass (distended gall bladder)



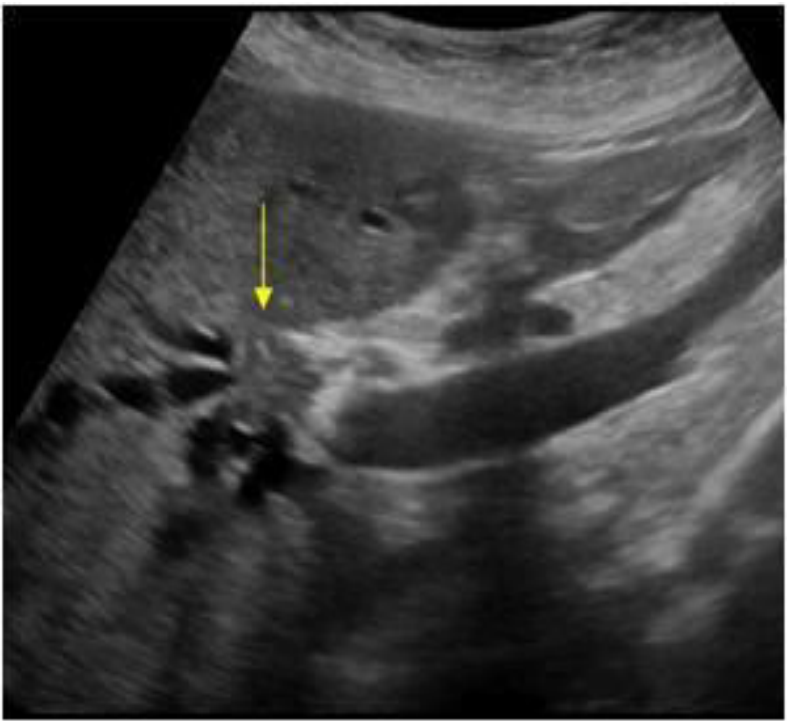
Investigations :

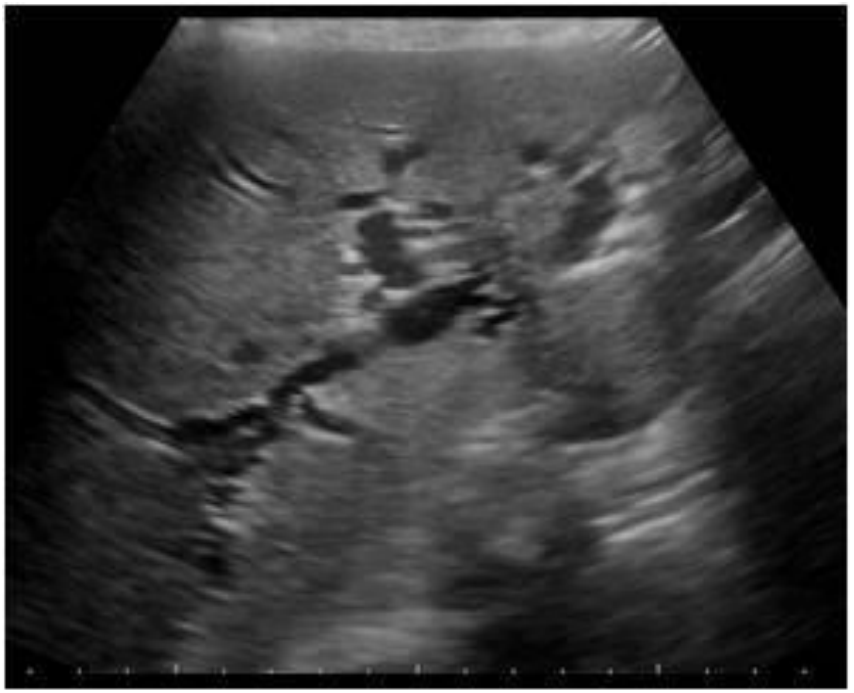
Radiologically :

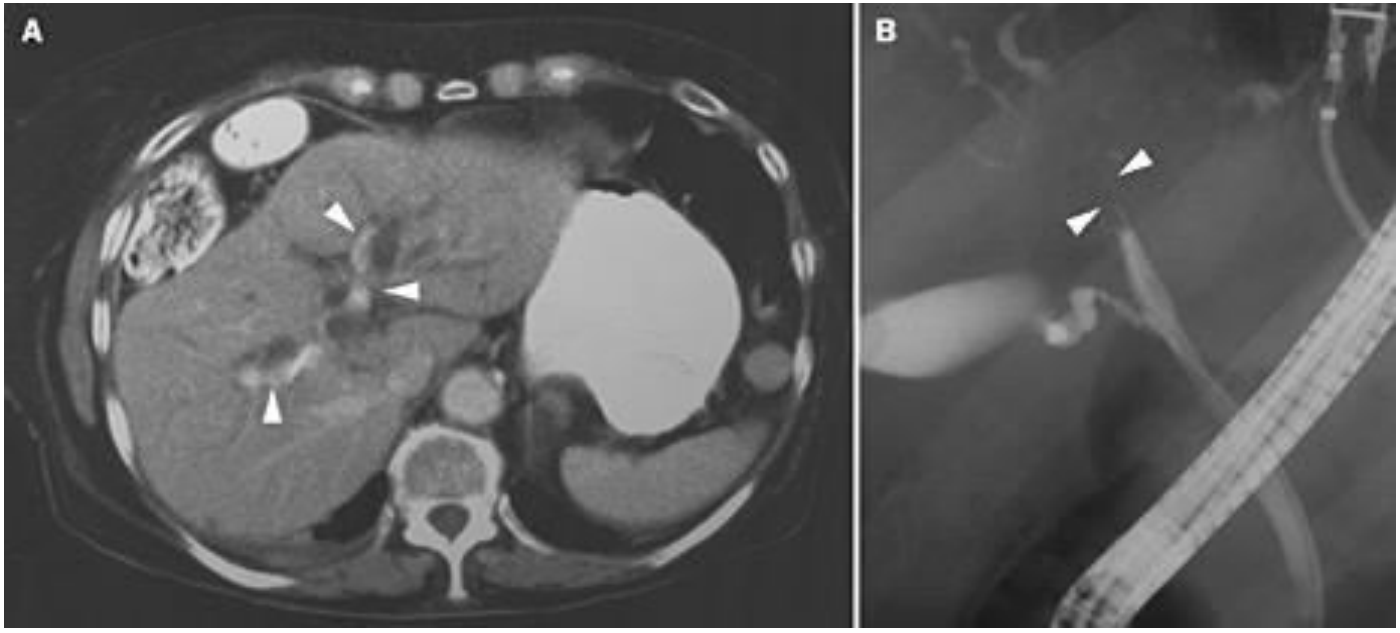
The initial tests are usually ultrasound or CT scan. A perihilar tumor causes dilatation of the intrahepatic biliary tree, but normal or collapsed gallbladder and extrahepatic bile ducts distal to the tumor

Ultrasound can establish the level of obstruction and rule out the presence of bile duct stones as the cause of the obstructive jaundice

CT scan makes it an excellent method to detect and stage bile tumors, both in vascular invasion and distant dissemination






A
B

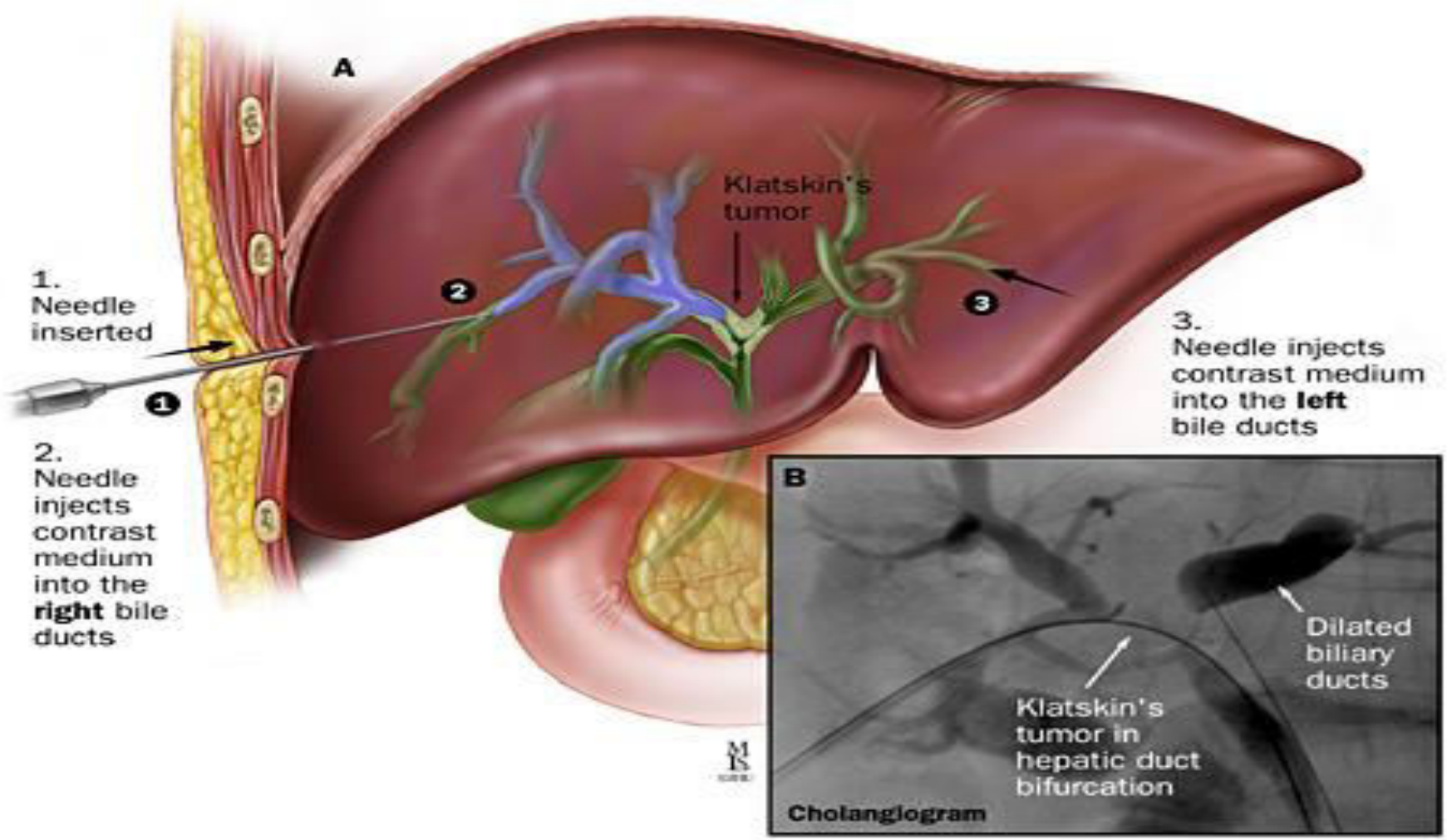
PTC (per cutaneous transhepatic cholangiography) defines the proximal extent of the tumor, which is the most important factor in determining resectability

ERCP(Endoscopic retrograde cholangio -pancreatography) is used, particularly in the evaluation of distal bile duct tumors.

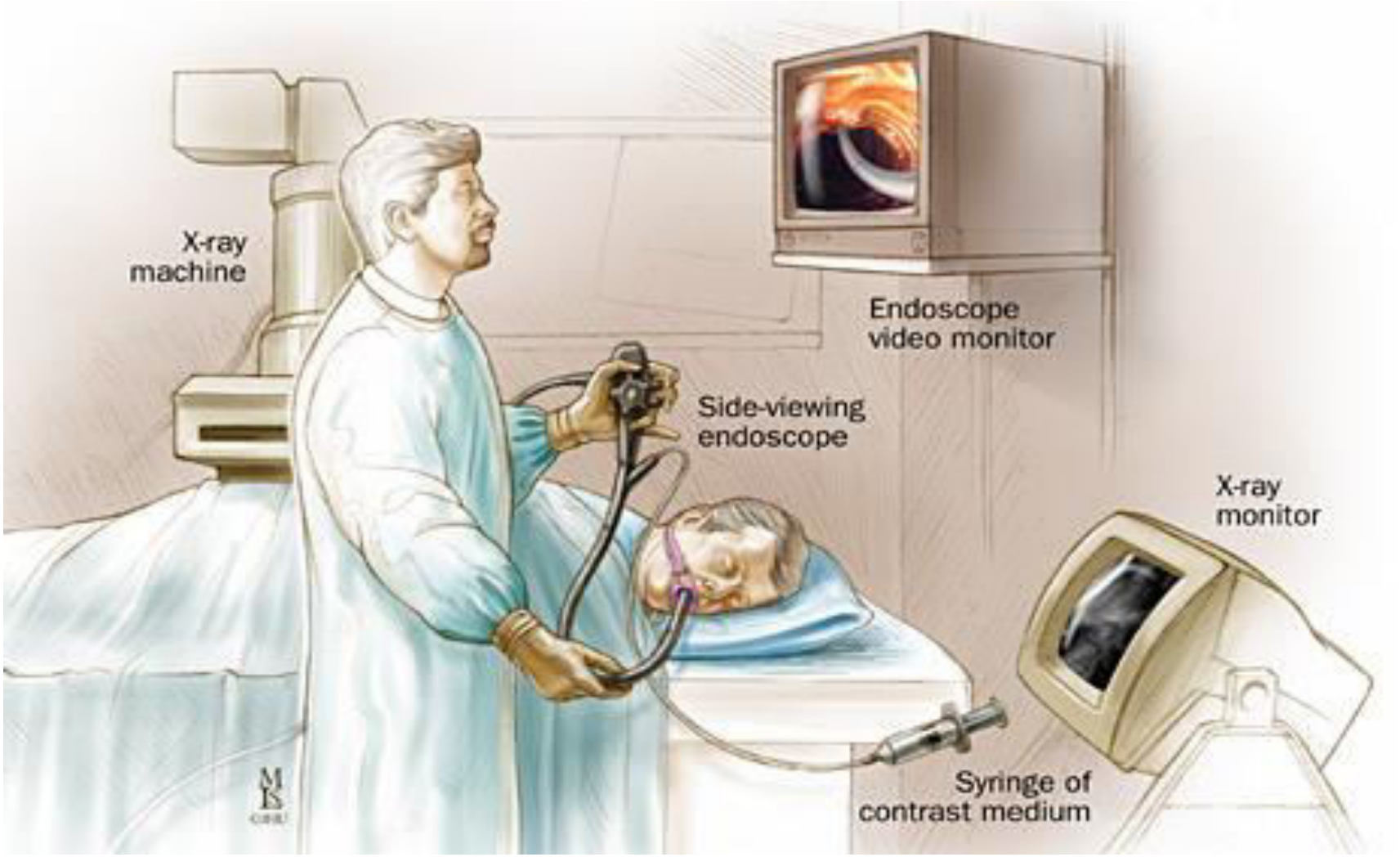
For the evaluation of vascular involvement, celiac angiography may be necessary.

With the newer types of MRI, a single noninvasive test has the potential of evaluating the biliary anatomy, lymph nodes, and vascular involvement, as well as the tumor growth itself.

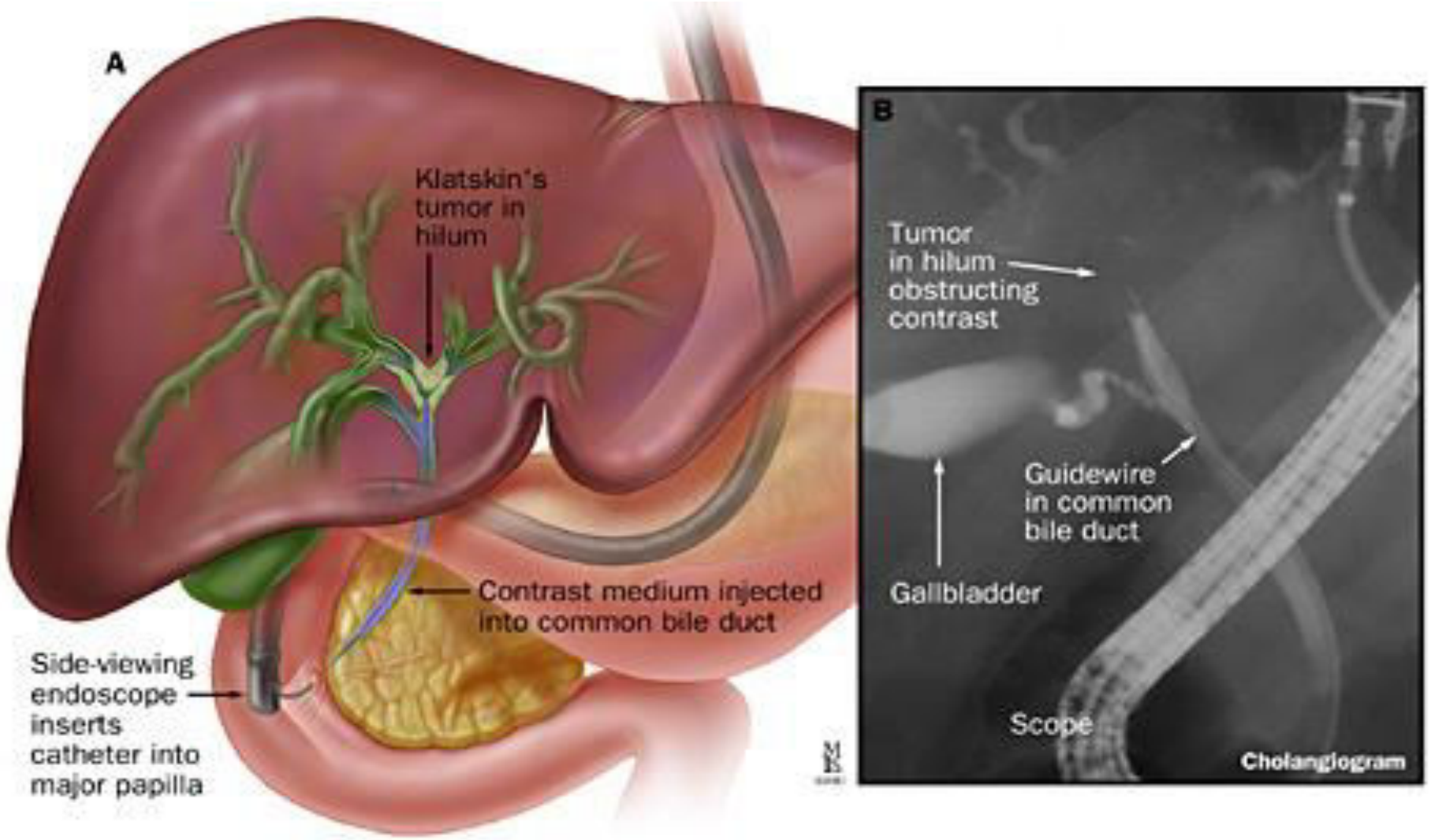
Technique of transhepatic percutaneous cholangiography;



Endoscopic Retrograde Cholangiopancreatography (ERCP)



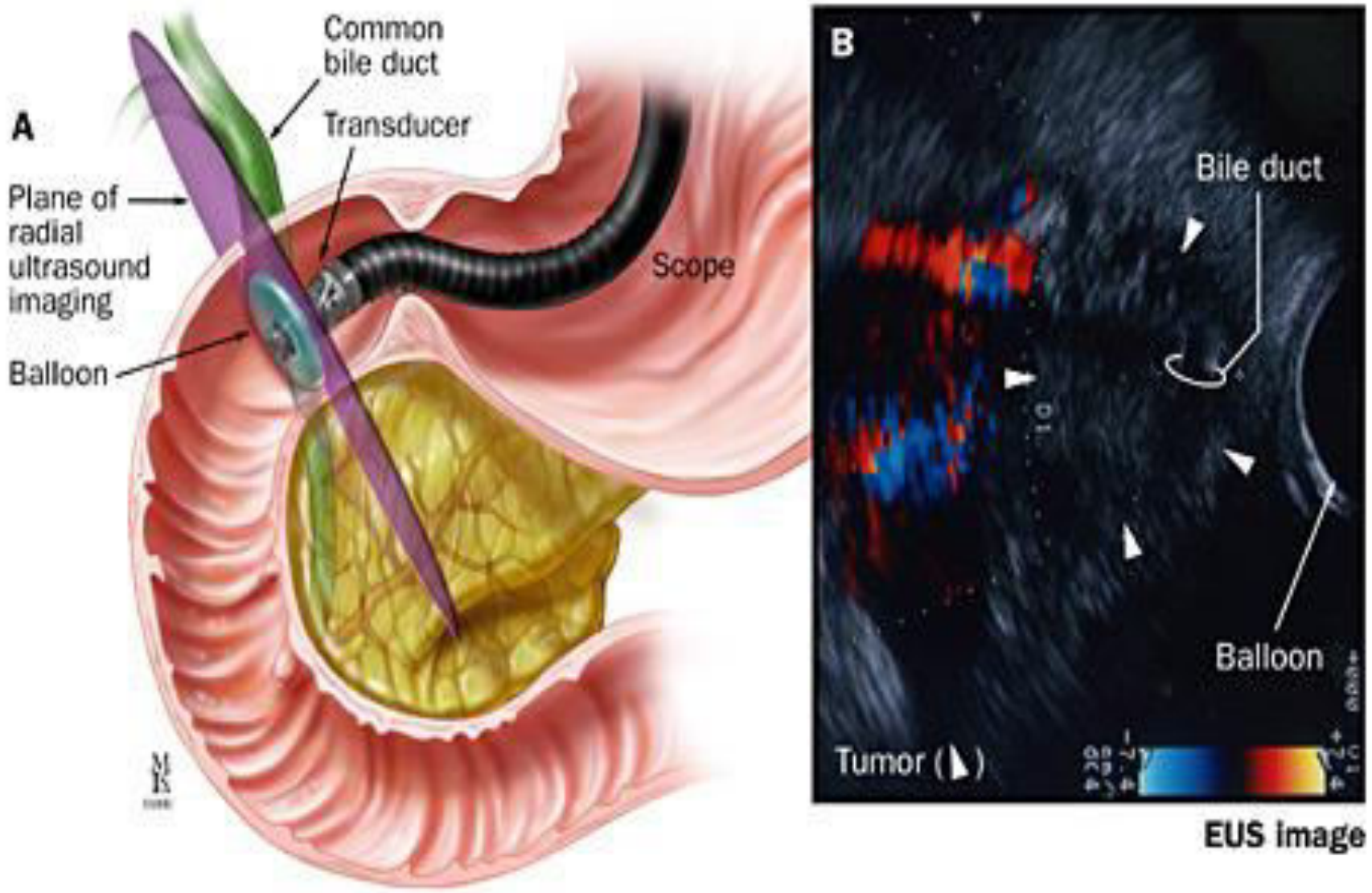
ERCP technique; A, endoscopic injection of contrast medium into biliary ducts with Klatskin's tumor; B, cholangiogram showing the tumor.



Endoscopic Ultrasound (EUS)

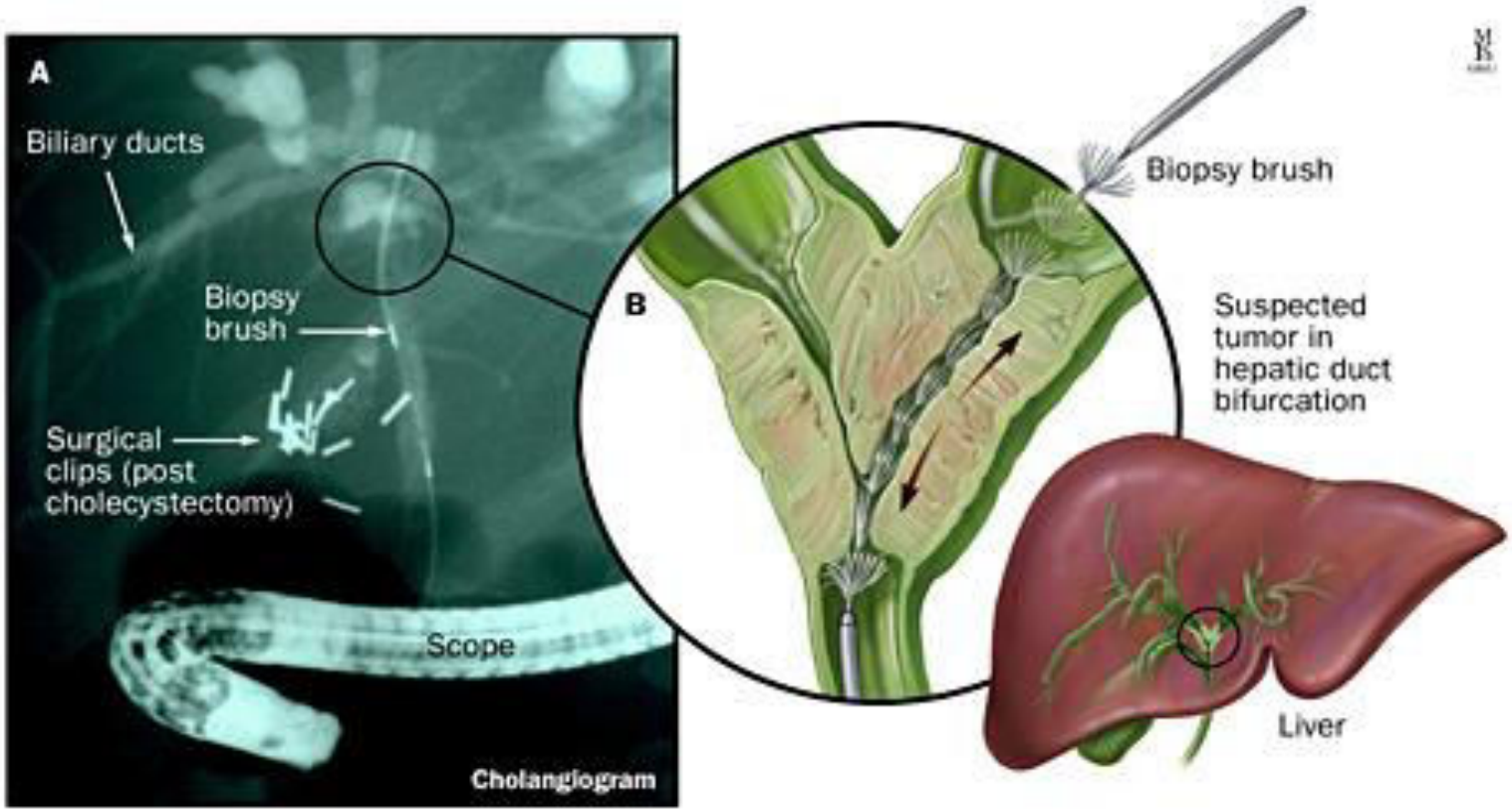
Endoscopic ultrasound is a combination of endoscopy with ultrasound to obtain images within the gastrointestinal tract. Endoscopic ultrasound has been used for the diagnosis of carcinomas of the bile duct.

Because the common bile duct and gallbladder are in close proximity to the duodenum and distal stomach, EUS has proven to be a useful tool for imaging these organs. This technique has been used to stage carcinomas of the bile duct and the gallbladder.


A
Common bile duct
Transducer
Plane of radial ultrasound imaging
Scope
Balloon
B
Bile duct
Balloon
Tumor

EUS image

Tissue diagnosis may be difficult to obtain nonoperatively except in advanced cases. Percutaneous fine-needle aspiration biopsy, biliary brush or scrape biopsy, and cytologic examination have a low sensitivity in detecting malignancy. Patients with potentially resectable disease should, therefore, be offered surgical exploration based on radiographic findings and clinical suspicion.



Laboratory :

Routine lab studies

- Extrahepatic cholestasis is reflected in elevated conjugated (ie, direct) bilirubin levels.
- Alkaline phosphatase levels usually rise in conjunction with bilirubin levels. Because alkaline phosphatase is of biliary origin,
- gamma-glutamyltransferase (GGT) also will be elevated.
- Aminotransferases (ie, aspartate aminotransferase [AST], alanine aminotransferase [ALT]) may be normal or minimally elevated.
- Biochemical tests of hepatic function (ie, albumin, prothrombin time [PT]) are normal in early disease.
- With prolonged obstruction, the prothrombin time (PT) can become elevated because of vitamin K malabsorption. Hypercalcemia may occur occasionally in the absence of osteolytic metastasis.

Tumour markers :

Tumor markers such CA 125 and carcinoembryonic antigen can be elevated in cholangiocarcinoma but tend to be nonspecific because they also increase in other GI and gynecologic malignancies or cholangiopathologies

Tumor marker carbohydrate antigen 19-9 (CA 19-9) can be evaluated in pancreatic and bile duct malignancies, as well as in benign cholestasis. A serum CA 19-9 level greater than 100 U/mL (normal < 40 U/mL) has 75% sensitivity and 80% specificity in identifying patients with PSC who have cholangiocarcinoma.

Treatment :

Surgical excision of biliary tract tumors is the treatment of choice in cholangiocarcinoma as it is the only therapeutic option that offers the potential for cure.

Surgical approaches have become increasingly aggressive over the last decade since it has become apparent that curative treatment is dependent upon aggressive excision. This often involves a major liver resection.

The objective is complete removal of the tumor and biliary drainage. Operative mortality in the hands of an experienced surgeon is extremely low (close to 0% for local resections and less than 10% for procedures with hepatic resection). Surgical management provides improved survival rates and quality of life.

Surgical treatment is dependent upon the localization of the mass.

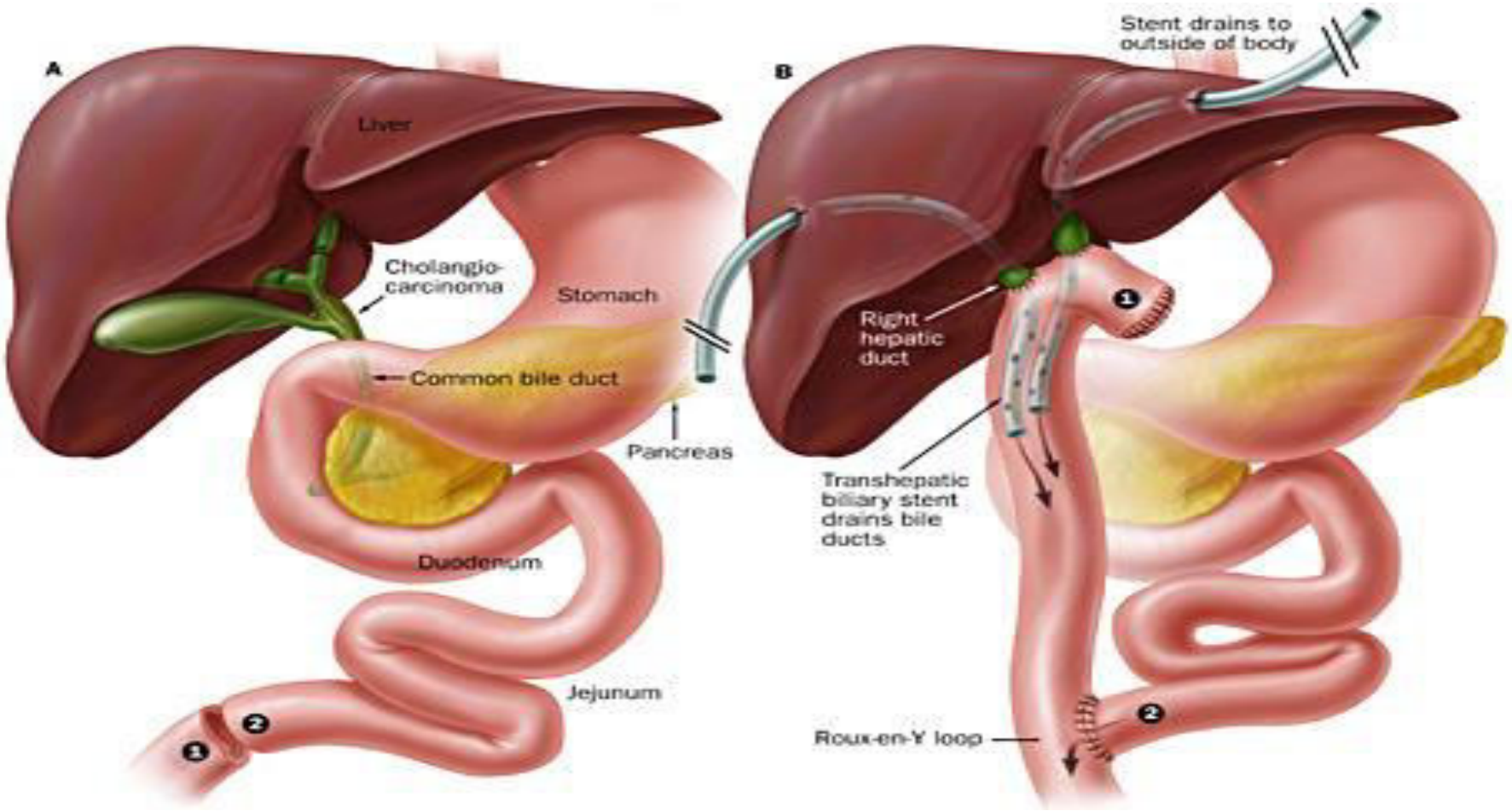
Treatment of hilar cholangiocarcinoma requires resection of the bifurcation of common hepatic duct.

The procedure starts with exploration of the peritoneal cavity to detect possible dissemination and resectability of the tumor. If the cholangiocarcinoma appears resectable, the gallbladder should be mobilized and the distal common bile duct divided.

Careful dissection continues proximally until right and left hepatic ducts are separated above the tumor. Biliary reconstruction is achieved through bilateral hepatojejunostomy on a Roux-en-Y intestinal loop above the transhepatic silicone biliary stents.

If cholangiocarcinoma extensively involves one lobe of the liver and relatively spares the other lobe, resection of the affected lobe or caudate lobe may be warranted with subsequent unilateral (in case of resection of right or left hepatic lobectomy) or bilateral (in case of caudate lobe resection) hepatojejunostomy

, Surgical technique for bilateral hepatojejunostomy with Roux-en-Y anastomosis for the removal of an extrahepatic tumor.



Endoscopic Therapy

Endoscopic biliary dilation may be used as a final palliative measure to relieve jaundice in patients who are poor surgical candidates, or as one of the steps prior to surgical intervention. This procedure requires use of a side-viewing endoscope to access the biliary duct and to introduce an inflatable balloon or series of endoscopic dilators over a guide wire. In many cases, a biliary sphincterotomy is performed prior to dilation and stent placement .

After successful dilation, plastic or self-expanding metal stents (endoprostheses) may be placed into the biliary ducts. Plastic stents should be replaced endoscopically at regular intervals (usually 8–12 weeks).

Right and left percutaneous self-expandable metal stents restore patency around a hilar tumor.