Biliary duct pathology

Dr Elham Eghbali

Biliary duct pathology

- Sonographic evaluation of the biliary tract is one of the most appropriate and efficacious uses of the ultrasound examination.
- Currently, sonography remains the modality of choice for the detection of gallstones, assessment of acute right upper quadrant pain, and for the initial evaluation of the patient with jaundice or elevated liver function tests.
- In conjunction with MRI/MRCP and contrast-enhanced CT scan, sonography also plays a key role in the multimodality evaluation of more complex biliary problems.

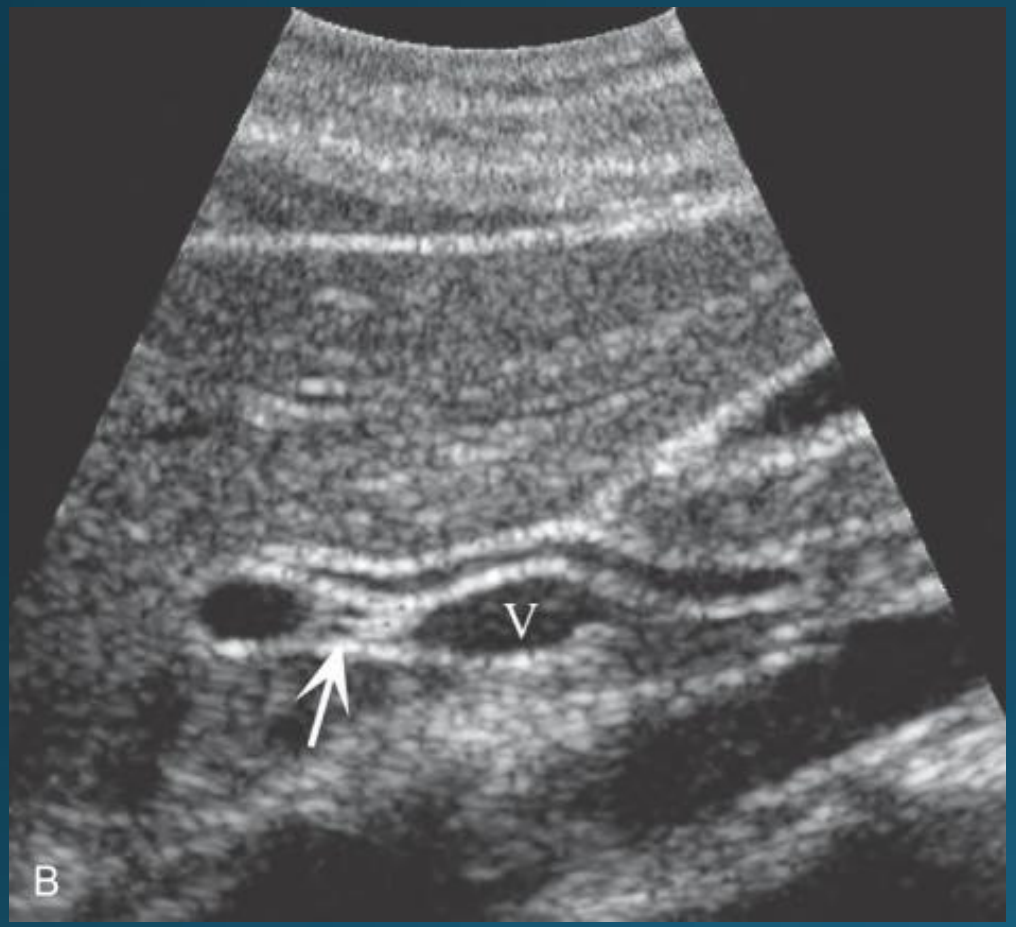
Biliary duct pathology

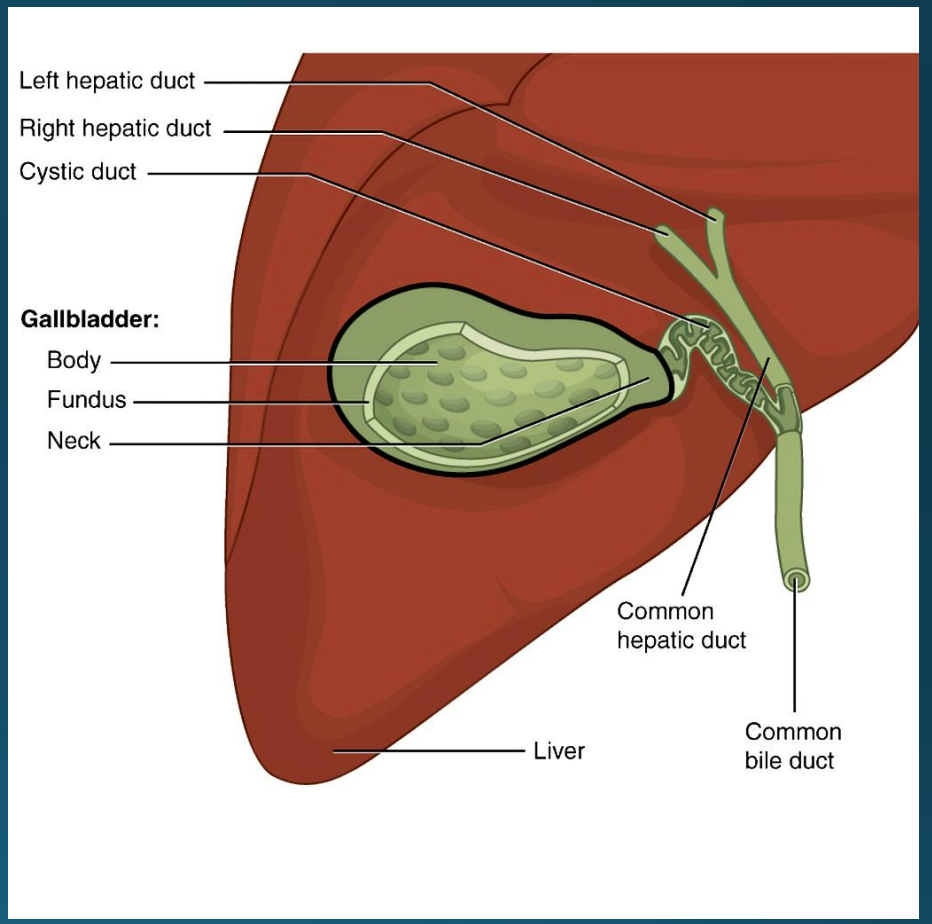
- The common bile duct (CBD), which is sometimes simply known as the bile duct, is formed by the union of the cystic duct and common hepatic duct (CHD).
- Of note, for decades, what had been labeled CBD in much radiology literature is now known actually, usually to have been the CHD.
- Thus, the of quoted normal value of < 6 mm (measured inner aspect of wall to inner aspect of the wall, typically by ultrasound) in adults actually refers to the CHD in most cases.

Biliary duct pathology

- In recent years, 7 mm has been proposed as a better cut-off.
- CBD could increase as much as 4 mm after cholecystectomy
- by age as much as 1 mm per decade after age 60.
- This has not been supported by subsequent studies, which indicate that the CHD diameter may increase only by 0.1-0.2 mm per decade and increases only about 1 mm after cholecystectomy.

Biliary duct pathology





Common hepatic/common bile ducts of normal caliber in sagittal view lying in the typical position anterior to the portal vein (V) and hepatic artery (arrow).

Biliary duct pathology

- Bile duct dilatation

- Bile duct dilatation refers to the dilatation of intrahepatic or extrahepatic bile ducts.

intrahepatic bile ducts

- >2 mm
- >40% of adjacent portal vein

extrahepatic bile ducts (common hepatic duct and common bile duct)

- usually measured in the proximal duct, near the proper hepatic artery
- diameter measured from inner wall to inner wall
- >6 mm +1 mm per decade above 60 years of age
- >10 mm post-cholecystectomy 2

Biliary duct pathology

bile duct dilatation

Obstruction

stricture

Neoplasm
- Cholangiocarcinoma
- gallbladder adenocarcinoma
- pancreatic adenocarcinoma
- Metastasis

Post inflammatory
- Pancreatitis
- Post radiation or chemotherapy

Inflammatory
- Aids cholangiopathy
- Biliary parasites
- Primary sclerosing cholangitis

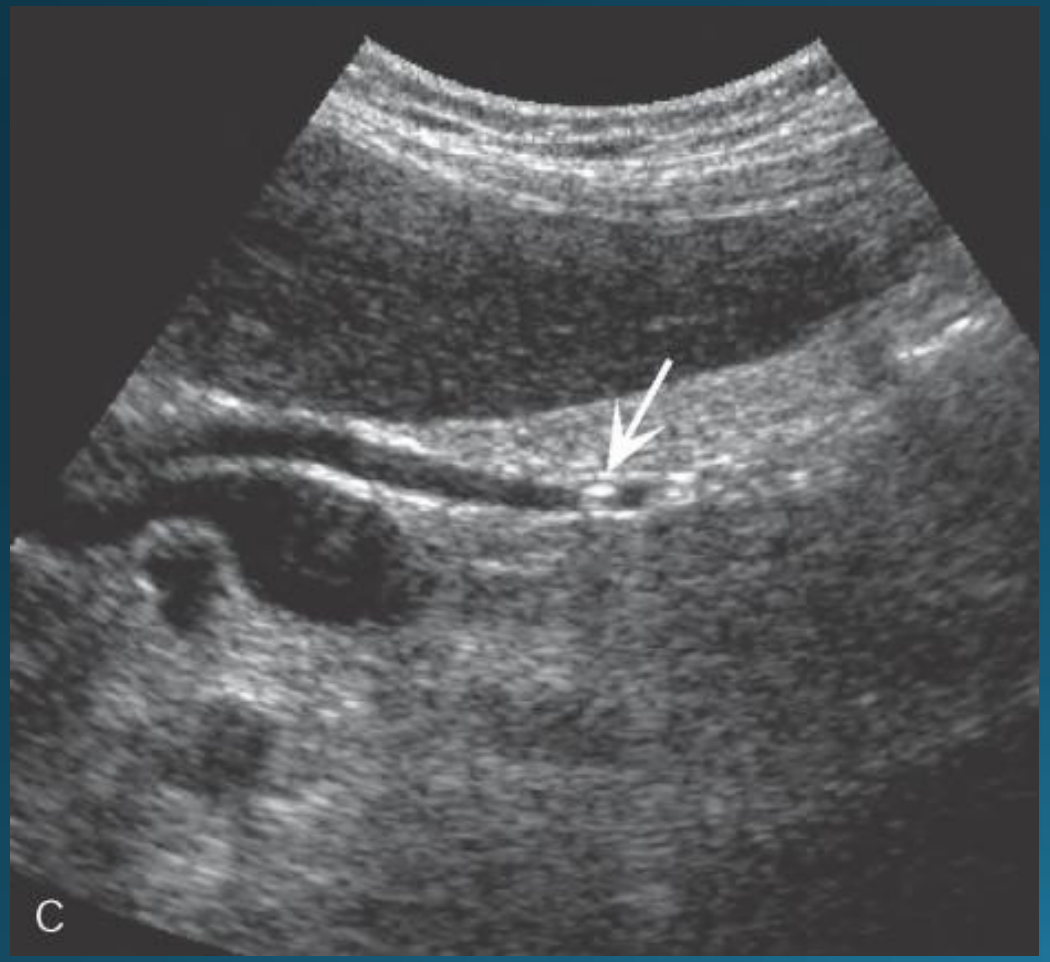
stones

No Obstruction

- Caroli disease
- Choledochal cyst
- Recurrent pyogenic cholangitis
- Primary sclerosing cholangitis

Bile duct dilatation

- Obstruction
- If there is an obstruction, we should first look for gallstones in the bile duct.

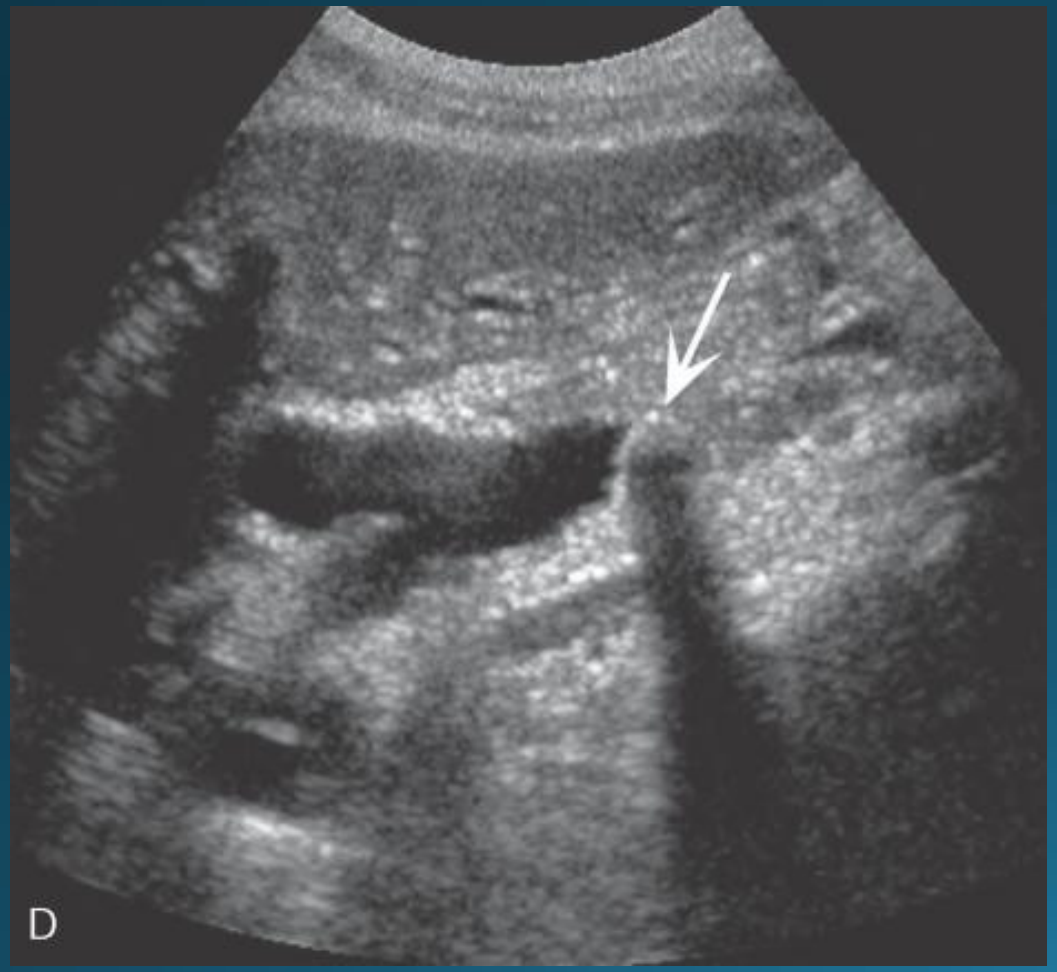


Common bile duct (CBD) stones.

C, Small stone (arrow) may not show shadowing.

Bile duct dilatation

- Obstruction
- If there is an obstruction, we should first look for gallstones in the bile duct.



Common bile duct (CBD) stones.

Large stone (arrow) has classic findings within a dilated CBD

Bile duct dilatation

- Obstruction

- If there are no gallstones involved, we should then look for strictures.

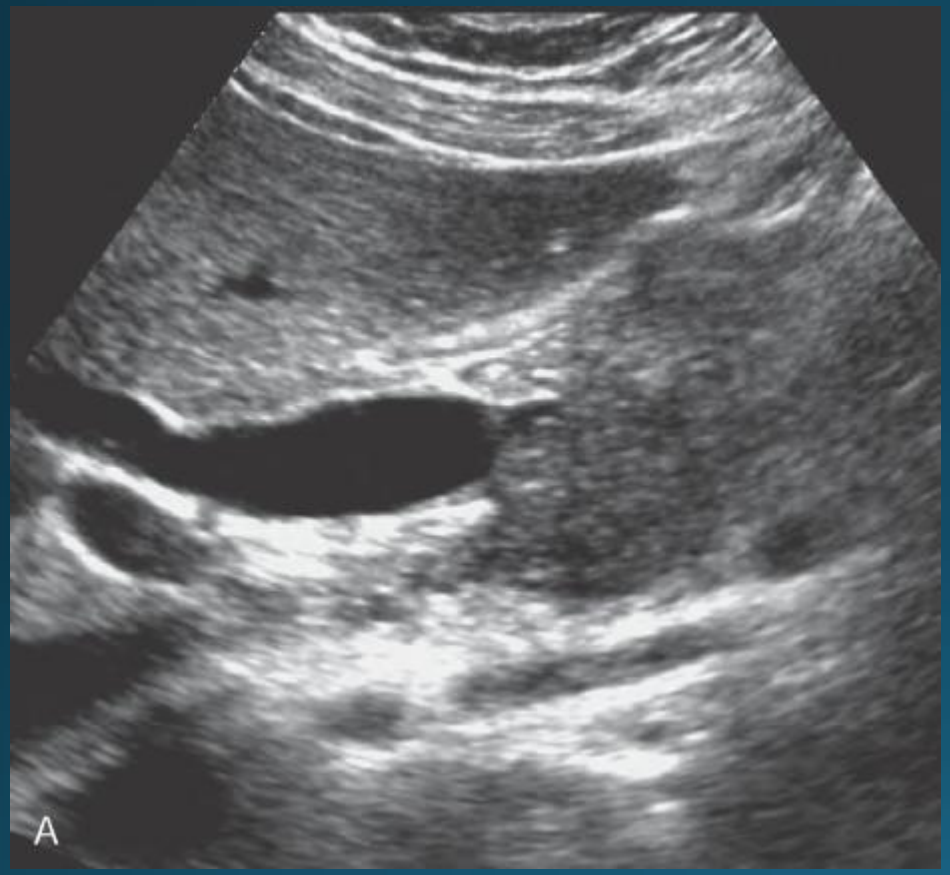
The differential diagnosis for a stricture is based on the location.

A distal stricture is most likely the result of :

- pancreatic carcinoma
- pancreatitis.
- distal cholangiocarcinoma

Bile duct dilatation

- Obstruction

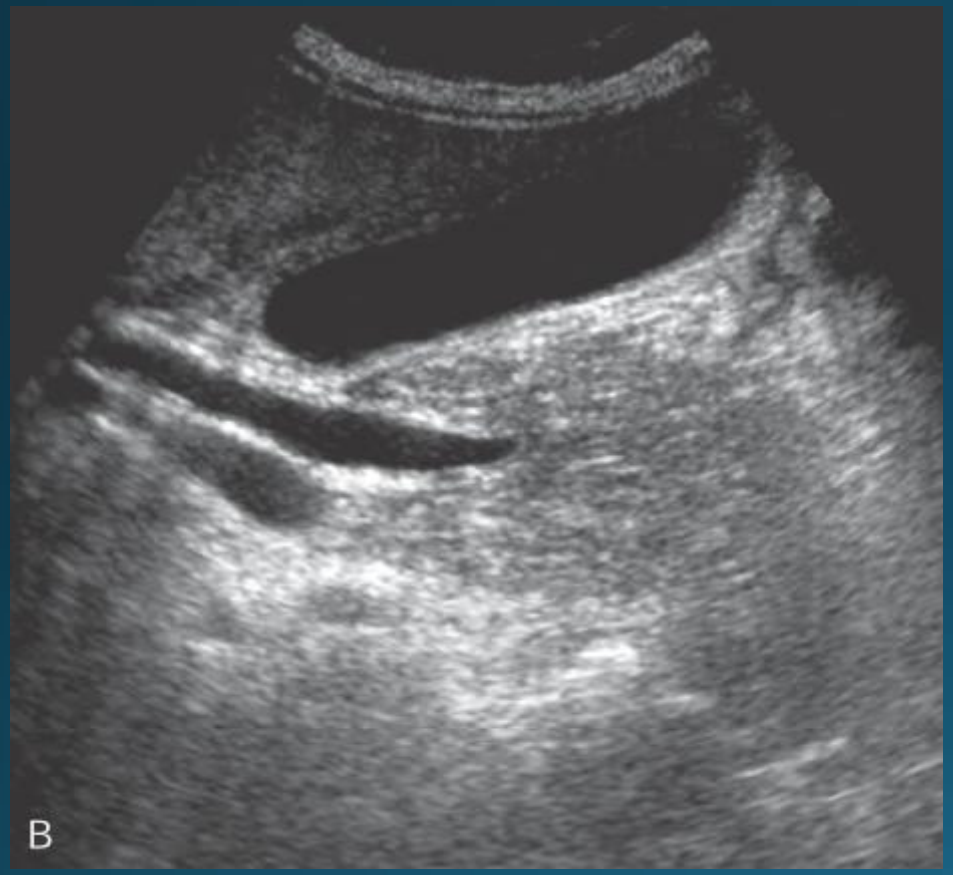


Common bile duct obstruction caused by extrinsic factors.

A, Pancreatic adenocarcinoma. Short transition zone with shouldering, large duct caliber, along with an obstructive mass are typical findings in malignant obstruction.

Bile duct dilatation

- Obstruction



B, Pancreatitis.

Elongated tapering of the duct suggests a benign cause.

Note mild sympathetic gallbladder wall thickening caused by adjacent inflammation.

Bile duct dilatation

- Obstruction

- Lemmel syndrome:

is defined as obstructive jaundice caused by a periampullary duodenal diverticulum compressing the intrapancreatic common bile duct with resultant bile duct dilatation.

Duodenal diverticulum of the second part of duodenum compressing the intrapancreatic part of the common bile duct (CBD) with resultant upstream dilatation of the extra- and intrahepatic bile ducts.

Bile duct dilatation

- Obstruction
- Lemmel syndrome:

CT

- Focal outpouching of the duodenum adjacent to the papilla (usually 2nd part of the duodenum) causing compression of the CBD.

MRCP

- Focal outpouching of the duodenum adjacent to the papilla (usually 2nd part of the duodenum) causing compression of the CBD.

.

Bile duct dilatation

- Obstruction
- Lemmel syndrome:

.

Bile duct dilatation

- Obstruction
- If there are no gallstones involved, we should then look for strictures.

The differential diagnosis for a stricture is based on the location.

A stricture within the liver is likely due to

- gallbladder carcinoma
- inflammatory strictures like PSC (Primary Sclerosing Cholangitis)
- AIDS cholangiopathy.

Metastatic disease can occur anywhere within the bilairy system.

Bile duct dilatation

- No Obstruction

- Once we have excluded obstruction, we have to think about nonobstructive biliary diseases like:

- Caroli disease
- Choledochal cyst
- Recurrent pyogenic cholangitis
- Primary sclerosing cholangitis

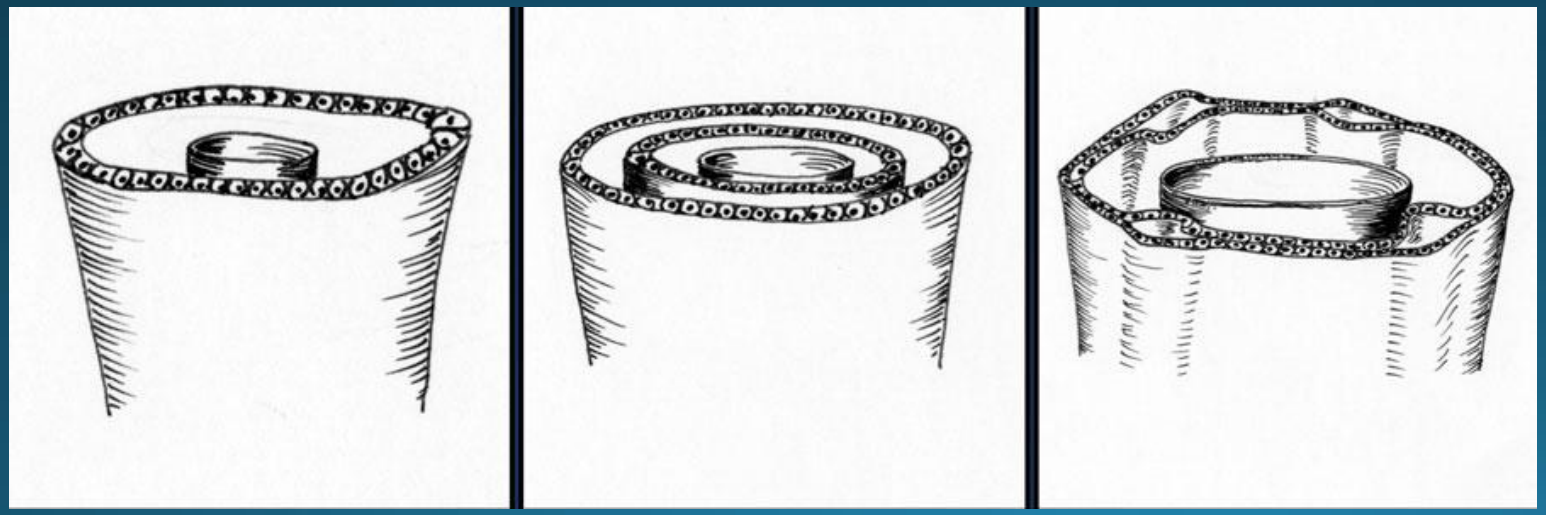
Caroli disease

- Caroli disease is an autosomal recessive disease secondary to the ductal plate malformation.
- It is associated with polycystic kidney disease, medullary sponge kidney and medullary cystic disease.
- So looking at the kidneys can sometimes help you make this diagnosis.
- The hallmark of Caroli disease is intrahepatic duct dilatation.
- The dilatation can be very large and saccular or it can be very linear.

Bile duct dilatation No Obstruction

Caroli disease

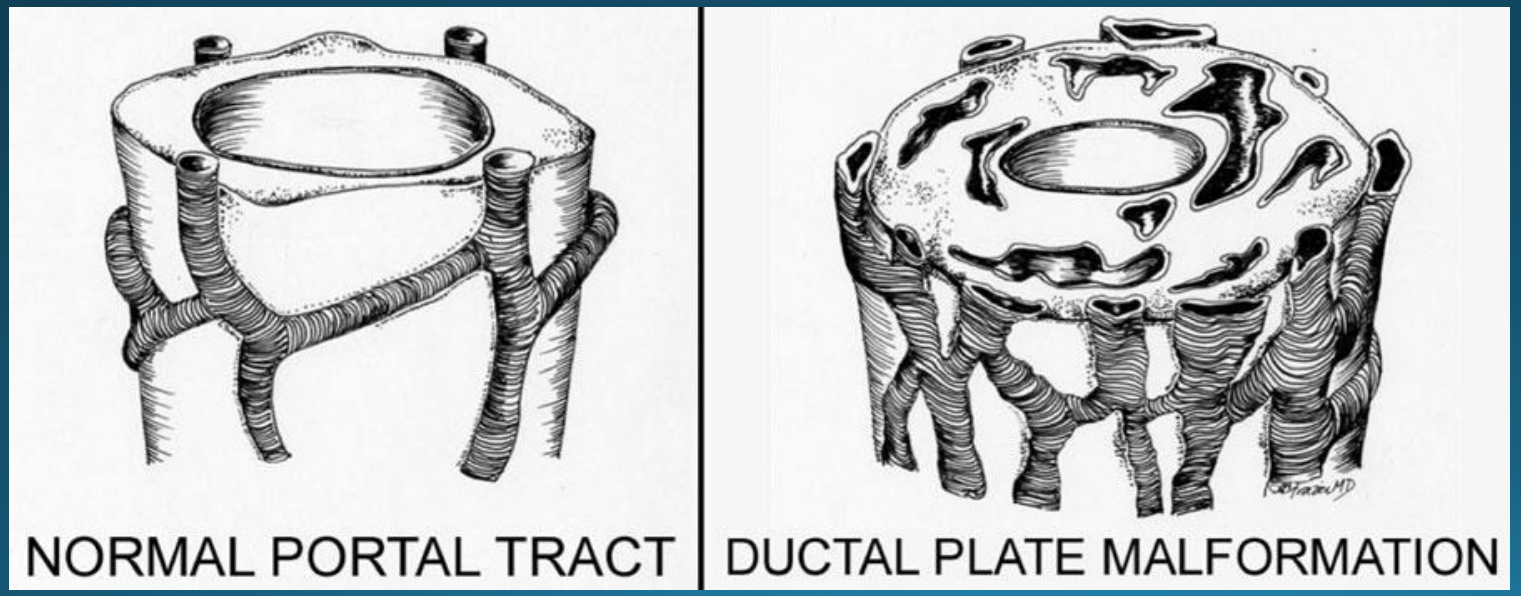
- ✓ The duct dilatation in Caroli disease is due to a congenital malformation of the ductal plate, which is the precursor of the intrahepatic bile ducts.
- ✓ Embryologically each bile duct begins as a single layer of cells that surrounds a portal vein.
- ✓ This layer then duplicates.
- ✓ Portions of this double layer fuse and resorb leaving unfused portions that become the bile ducts.



Bile duct dilatation No Obstruction

Caroli disease

- So in the normal situation each portal vein is surrounded by interconnecting bile ducts.
- However if the patient has ductal plate malformation, the bile ducts are too numerous and they are ectatic .
- Whether or not we see this on imaging depends on which portion of the bile ducts is affected.
- If the large ducts are involved, we see this as Caroli disease.
- However if only the very small ducts are involved, the result is congenital hepatic fibrosis.

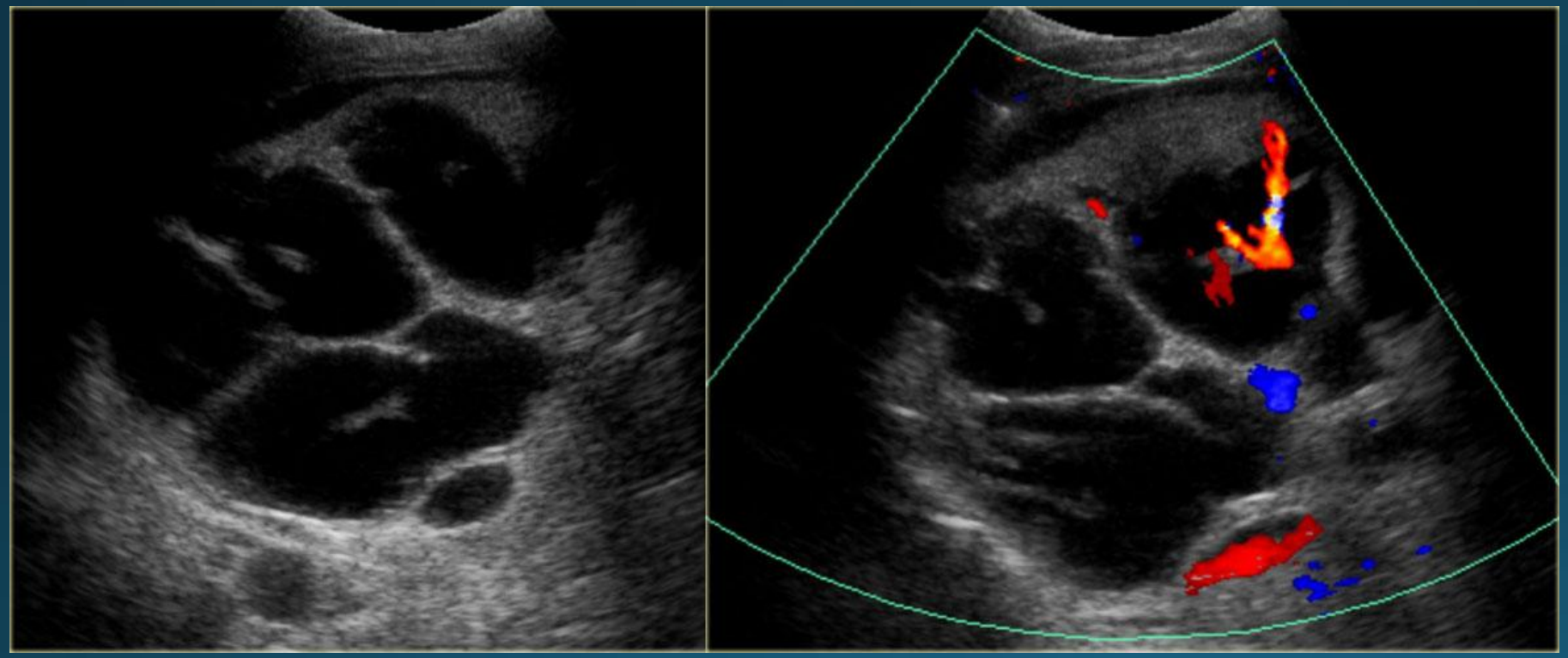


Caroli disease

- Most commonly the intrahepatic duct dilatation is segmental (83%) in distribution.
- The diffuse form is less common (17%).
- The shape of the dilatation is saccular in 76% or fusiform in 24% of the cases.
- A very important sign is the central dot sign.
- The central dot corresponds to the portal vein that is surrounded by dilated bile ducts.

Bile duct dilatation No Obstruction

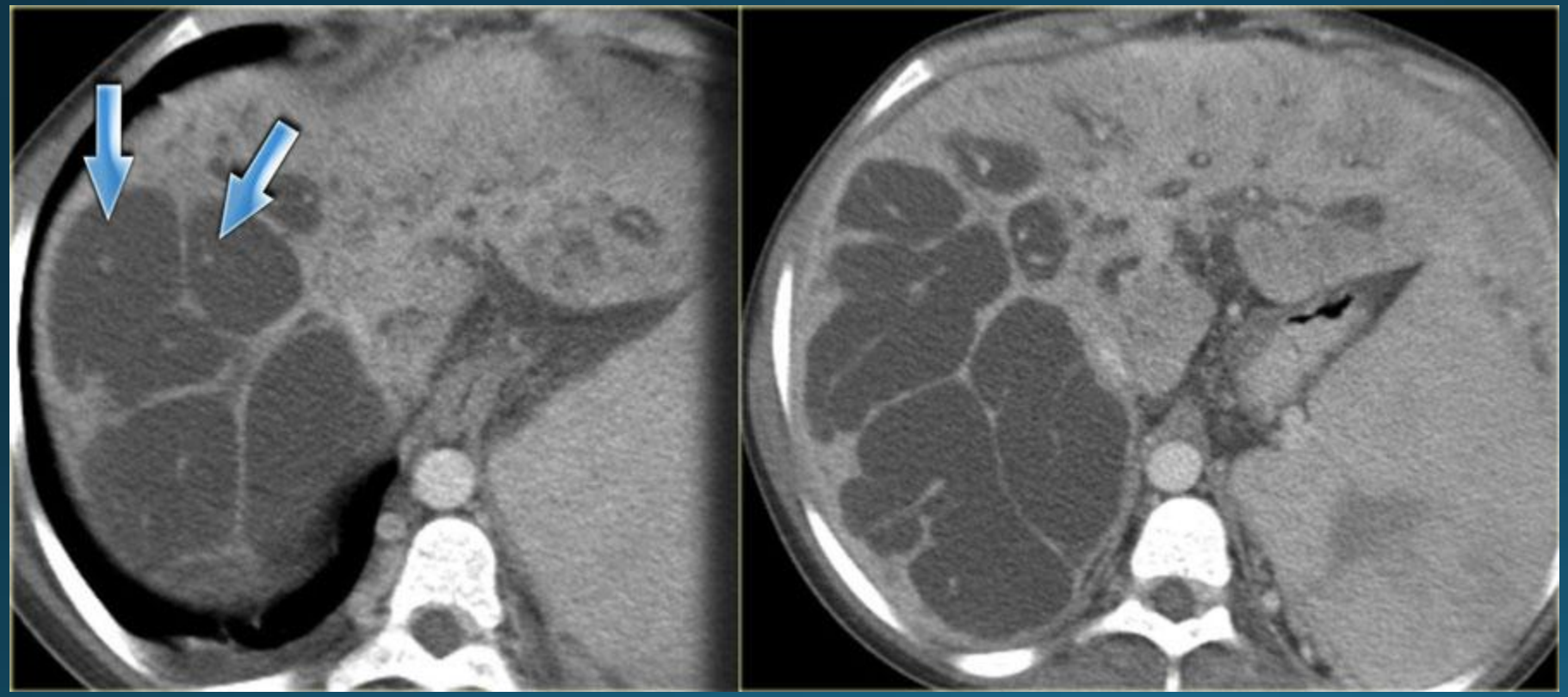
Caroli disease



Central dot sign in Caroli disease

Bile duct dilatation No Obstruction

Caroli disease

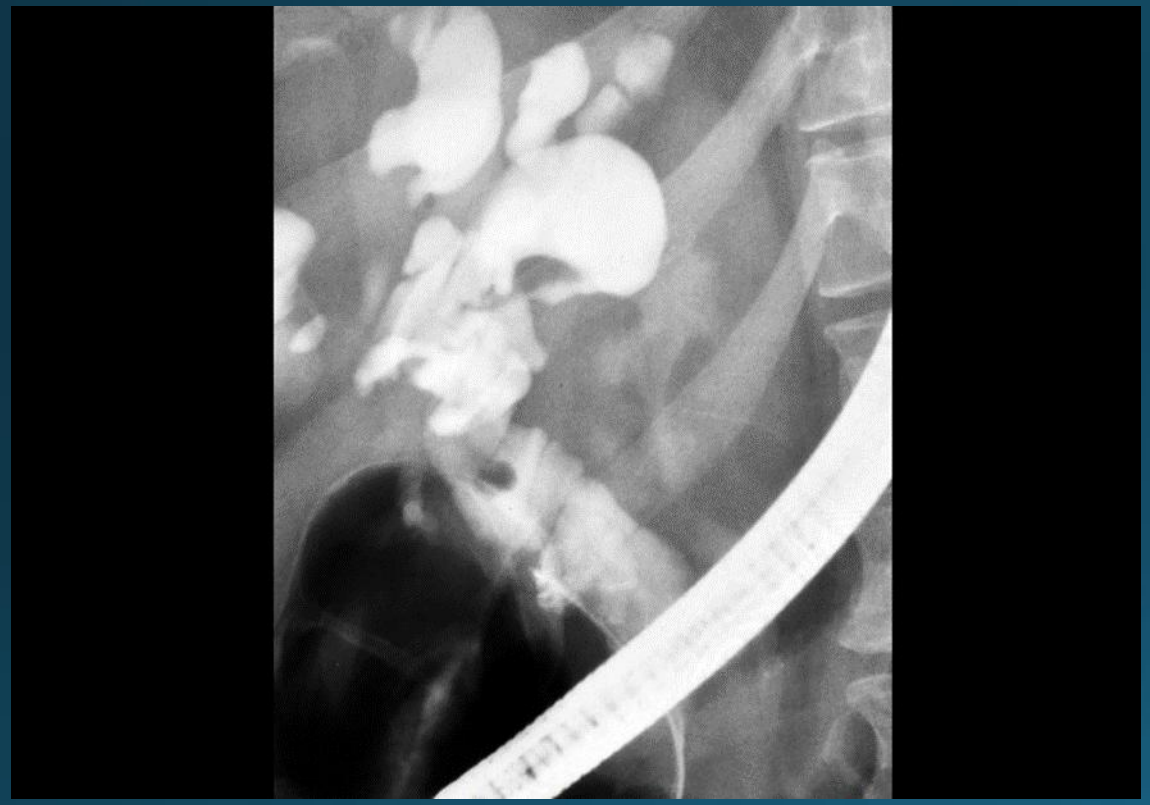


Notice the central dot sign and the segmental involvement.
This patient has cirrhosis with splenomegaly due to portal hypertension.

Extrahepatic duct dilatation is present in 53% of cases, secondary to cholangitis and stone or sludge passage.
These are secondary findings, that are not part of the primary disease.

Bile duct dilatation No Obstruction

Caroli disease

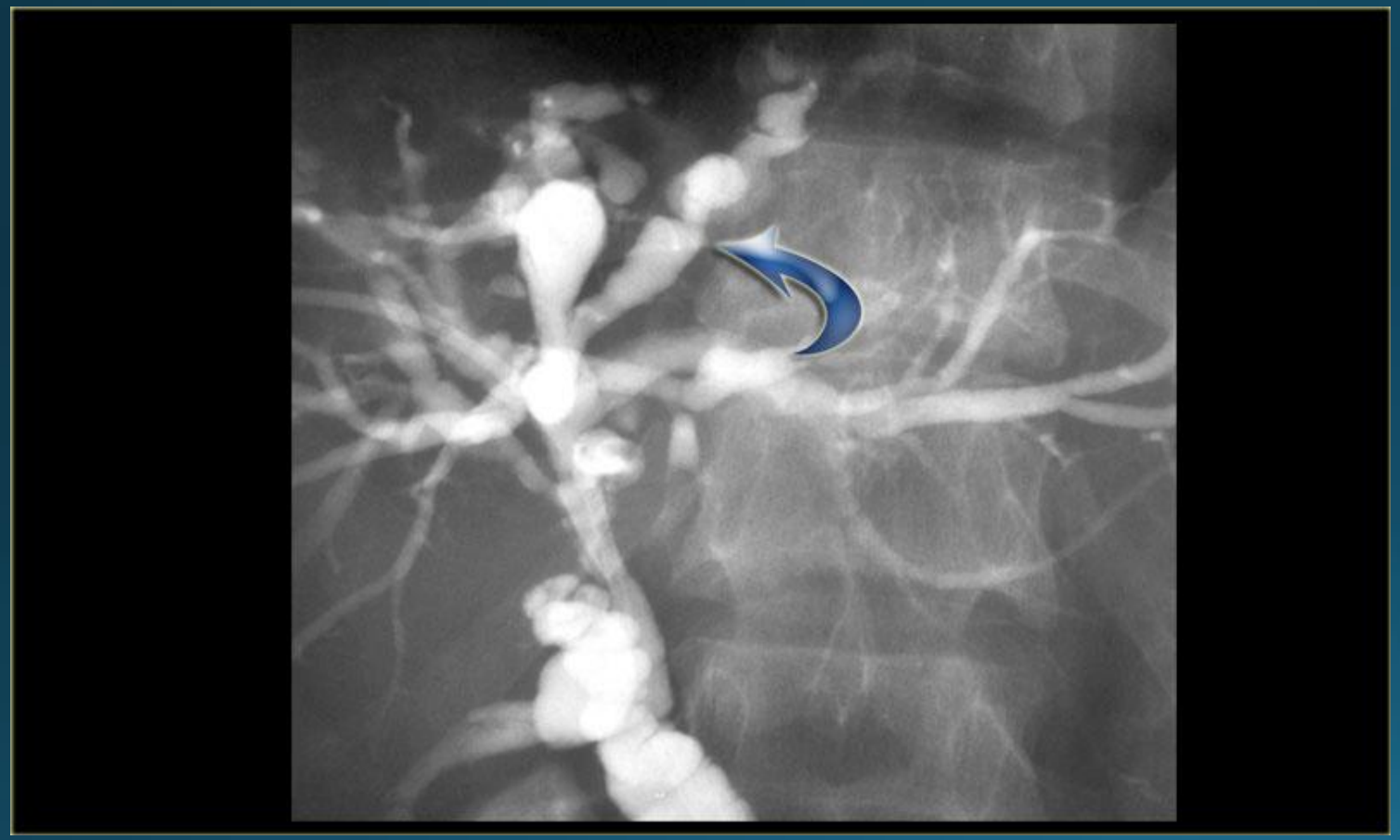


ERCP: Caroli disease with severe intrahepatic duct dilatation. No obstruction. Mild dilatation of the choledochal duct due to cholangitis

- The cholangiogram is important in the work up of these patients, because obstruction has to be excluded.
- This can be done with MRCP or ERCP, as is shown on the left.
- There was no sign of obstruction.
- The mild dilatation of the choledochal duct was the result of cholangitis.

Bile duct dilatation No Obstruction

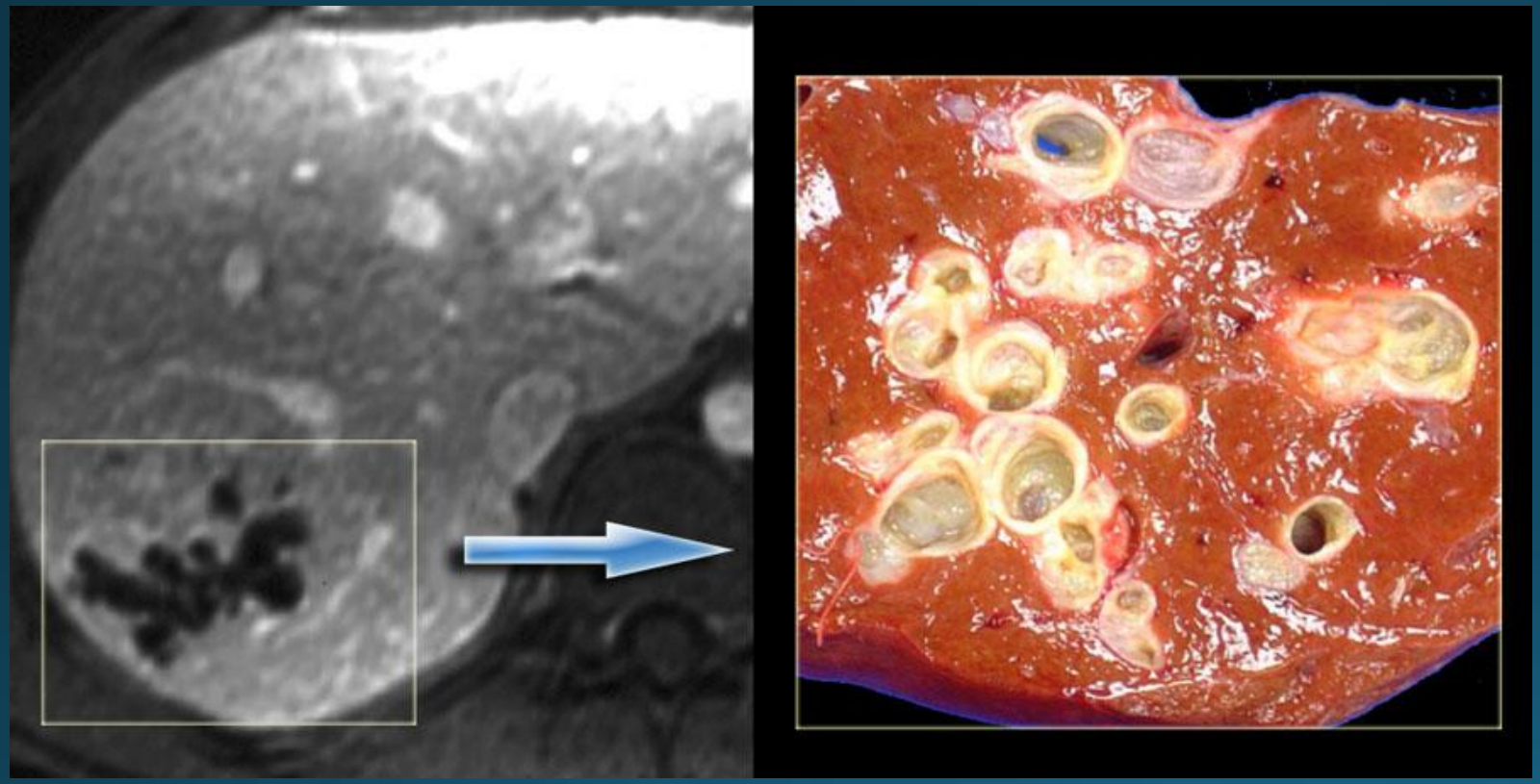
Caroli disease



There is focal dilatation with intermixing strictures of the bile ducts in segment IV (arrow).
The other bile ducts and the choledochal duct are normal.

Bile duct dilatation No Obstruction

Caroli disease



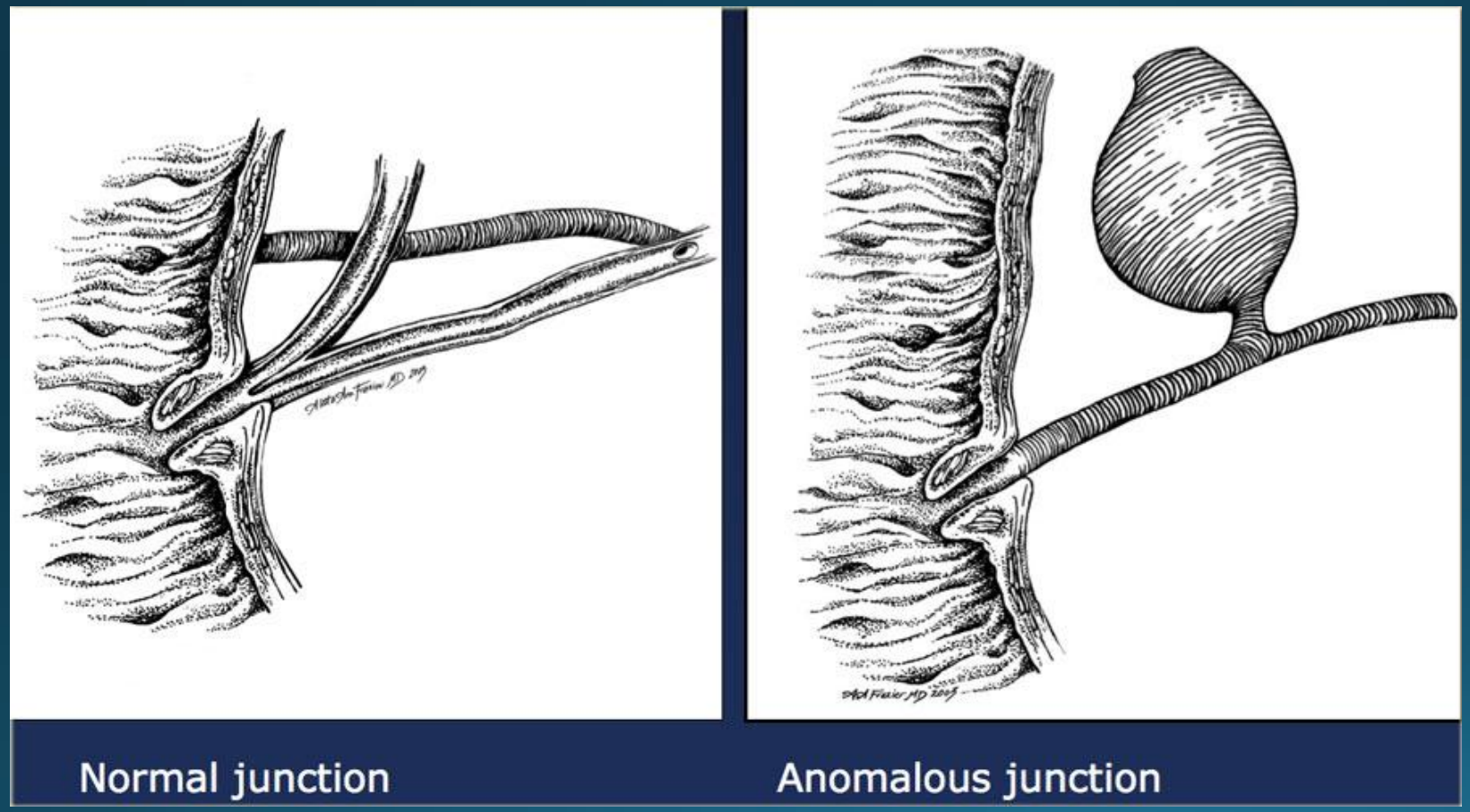
- In some of the cases of Caroli disease the imaging findings may simulate a cystic neoplasm as is seen in the case on the left.
- This case was originally diagnosed as a biliary cystadenoma.
- However, the gross specimen demonstrates dilated bile ducts and ductal plate malformation was present microscopically.

Choledochal cyst

- A choledochal cyst is a congenital dilatation of the extrahepatic bile duct.
- These patients do not have a ductal plate abnormality.
- The most common theory for the development of a choledochal cyst is that the dilatation is due to an underlying anomalous pancreatico-biliary junction.
- In the anomalous junction the biliary and pancreatic duct join proximal to the sphincter of Oddi.
- In these patients there is a long common channel.
- The theory is that when the sphincter of Oddi contracts, pancreatic enzymes will flow into the bile duct and causing dilatation and in some cases, narrowing of the distal duct.

Bile duct dilatation No Obstruction

Choledochal cyst



Bile duct dilatation No Obstruction

Choledochal cyst

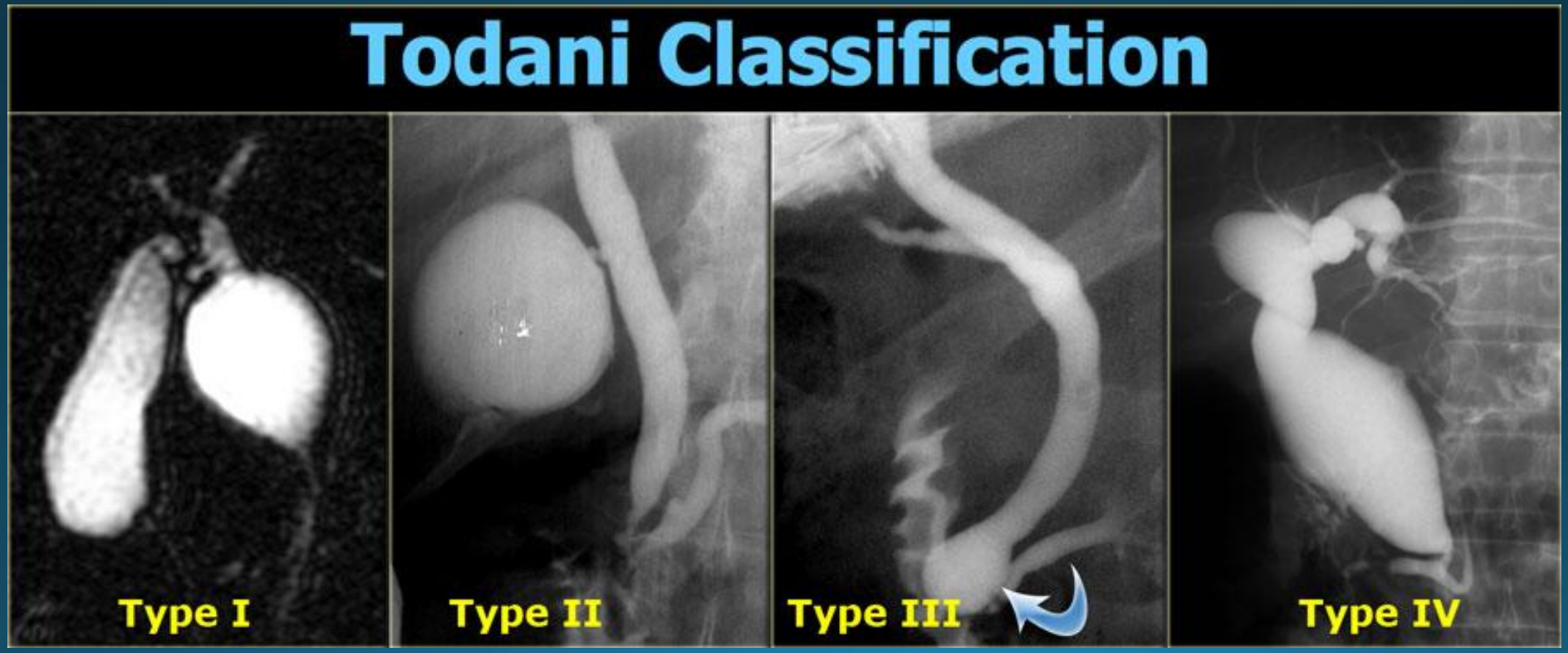
Todani Classification

- This classification classifies the choledochal cysts into 5 cathegories.
- Type V, is Caroli disease.We now know, that Caroli is a different disease.
- Type I is a true choledichal cyst with focal dilatation of the extrahepatic duct. This is the most frequent type (90-95% of the cases).
- Type IV is also a true choledichal cyst with dilatation of the entire extrahepatic duct with involvement of portions of the intrahepatic ducts.
  The intrahepatic ducts taper normally to the periphery, indicating that there is no obstruction.
- Type II and III are extremely rare and it is debatable whether or not these are true choledochal cysts.
- Type II is a diverticulum of the extrahepatic duct and many believe that this entity is not related to an anomalous pancreatico-biliary junction.
- Type III is a choledochocele, where there is dilatation of the distal part of the bile duct.
  These patients also have a normal pancreaticobiliary junction.

Bile duct dilatation No Obstruction

Choledochal cyst

Todani Classification



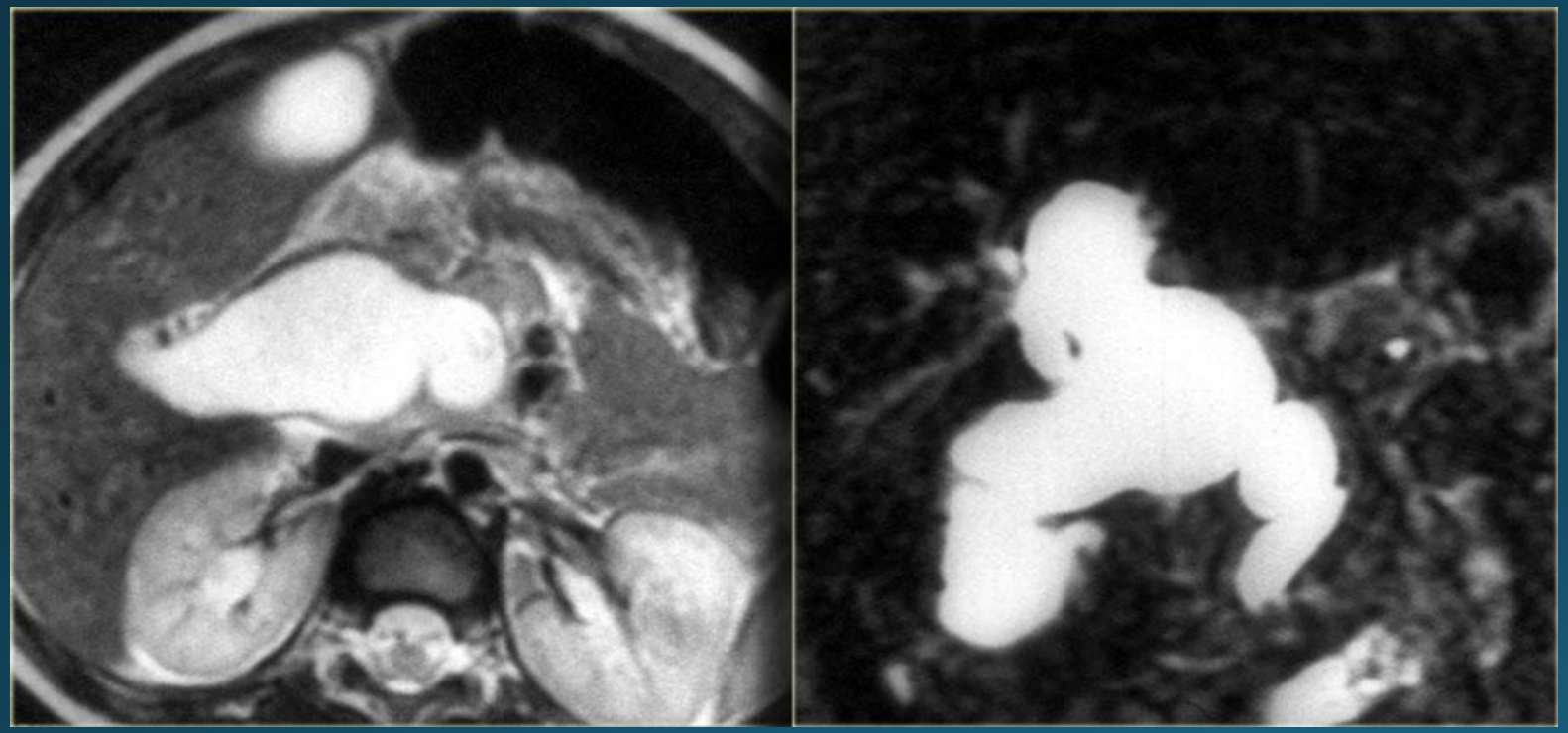
Bile duct dilatation No Obstruction

Choledochal cyst

Type IV choledochal cyst
with dilatation of both the extrahepatic duct and part of the intrahepatic ducts.
So this is a type IV.
Notice that the peripheral ducts are normal, so this is not an obstructive pattern.

Bile duct dilatation No Obstruction

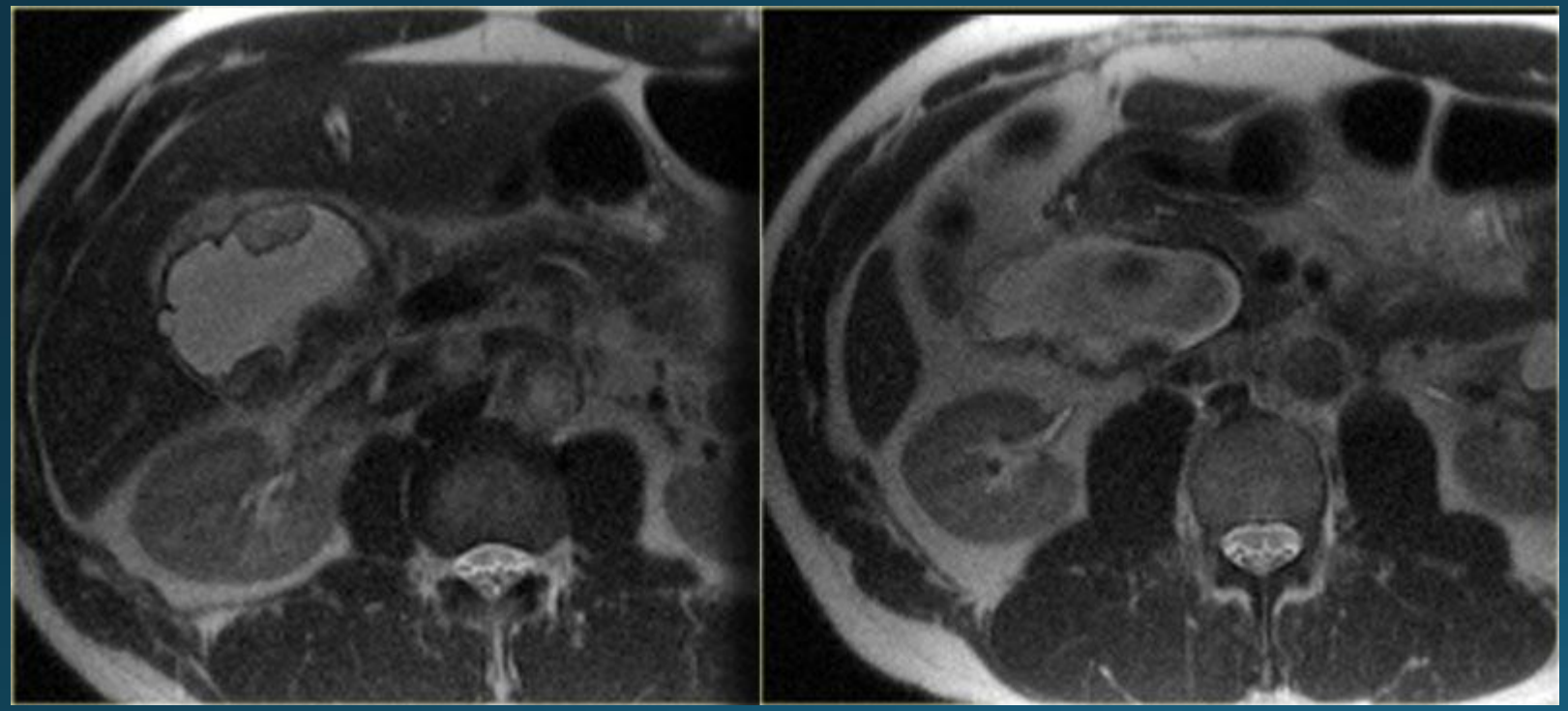
Choledochal cyst



Type IV choledochal cyst
There is dilatation of the extrahepatic duct, cystic duct and a small portion of the left hepatic duct.
There is no intrahepatic dilatation.

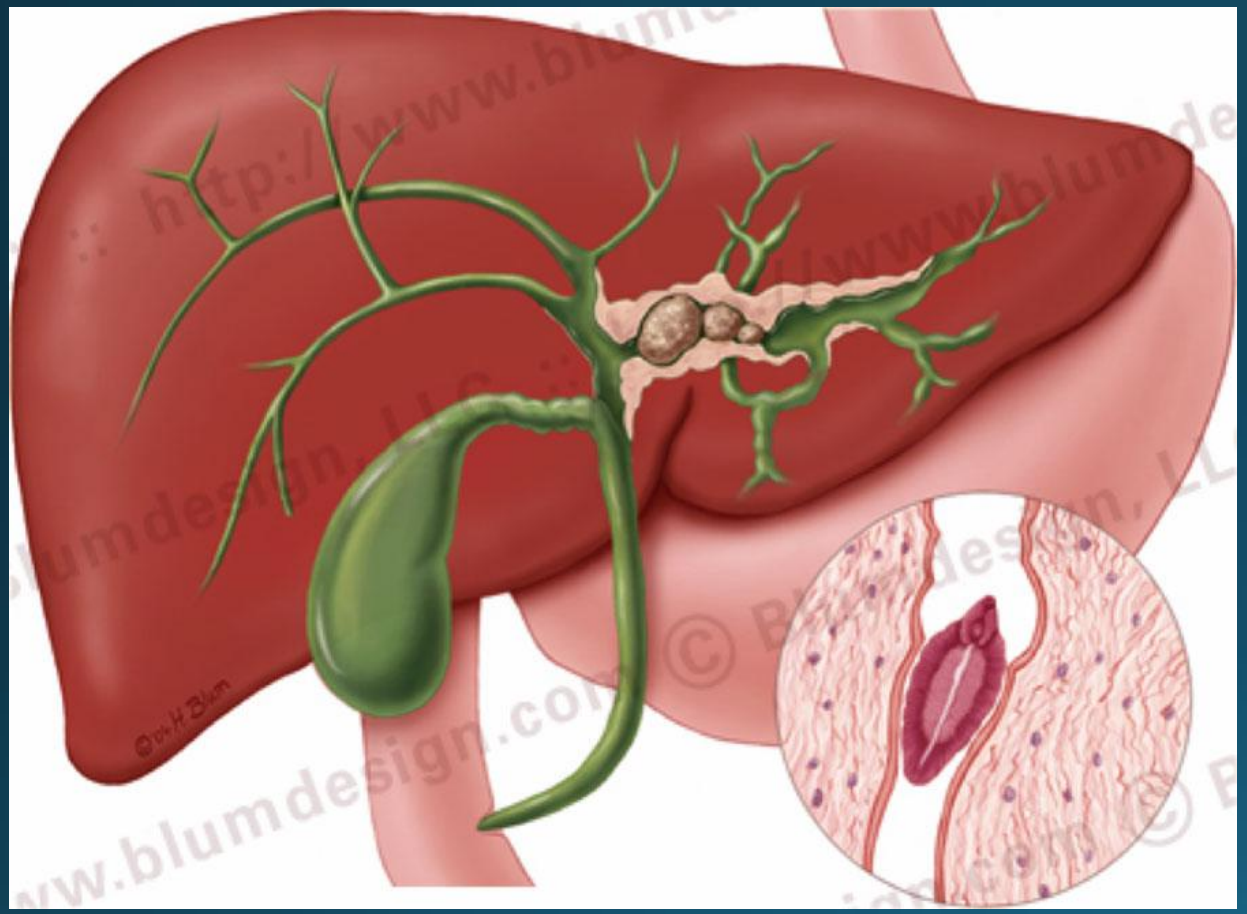
Bile duct dilatation No Obstruction

Choledochal cyst



There is an association of bile duct adenocarcinoma and choledochal cysts.
These carcinomas can occur within the choledochal cyst, or in the gallbladder or anywhere else in the biliary ducts.
In the bile ducts they can present as classic peripheral cholangiocarcinoma, Klatskin tumor or distal cholangiocarcinoma.

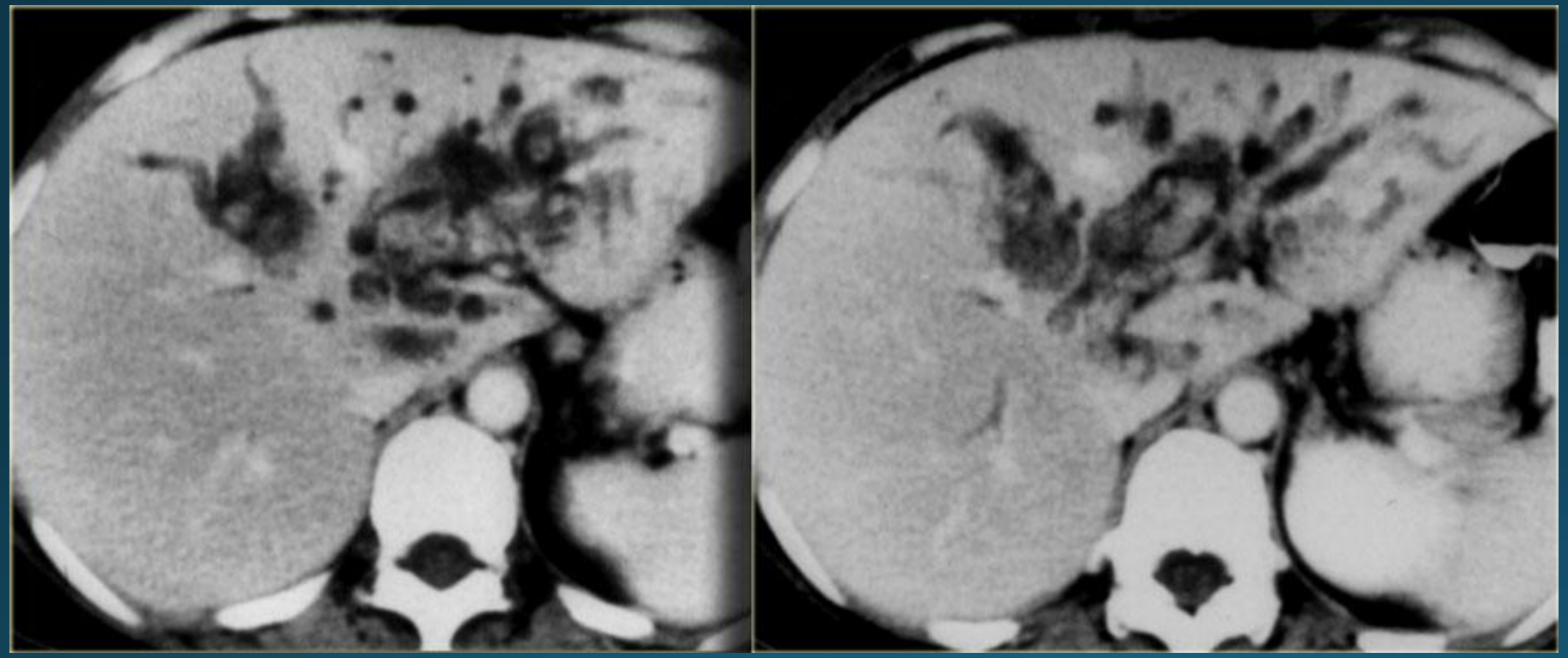
Bile duct dilatation No Obstruction



- Recurrent Pyogenic Cholangitis (RPC)
- Recurrent pyogenic cholangitis is an uncommon disease in the western world.
- Most of these cases are seen in Asian countries.
- The etiology is unknown, although some of these patients have biliary parasites.
- The disease is characterized by the presence of intrahepatic pigmented stones and recurrent infection.
- These patients are also at risk of developing biliary cirrhosis and cholangiocarcinoma.

Bile duct dilatation No Obstruction

- Recurrent pyogenic cholangitis

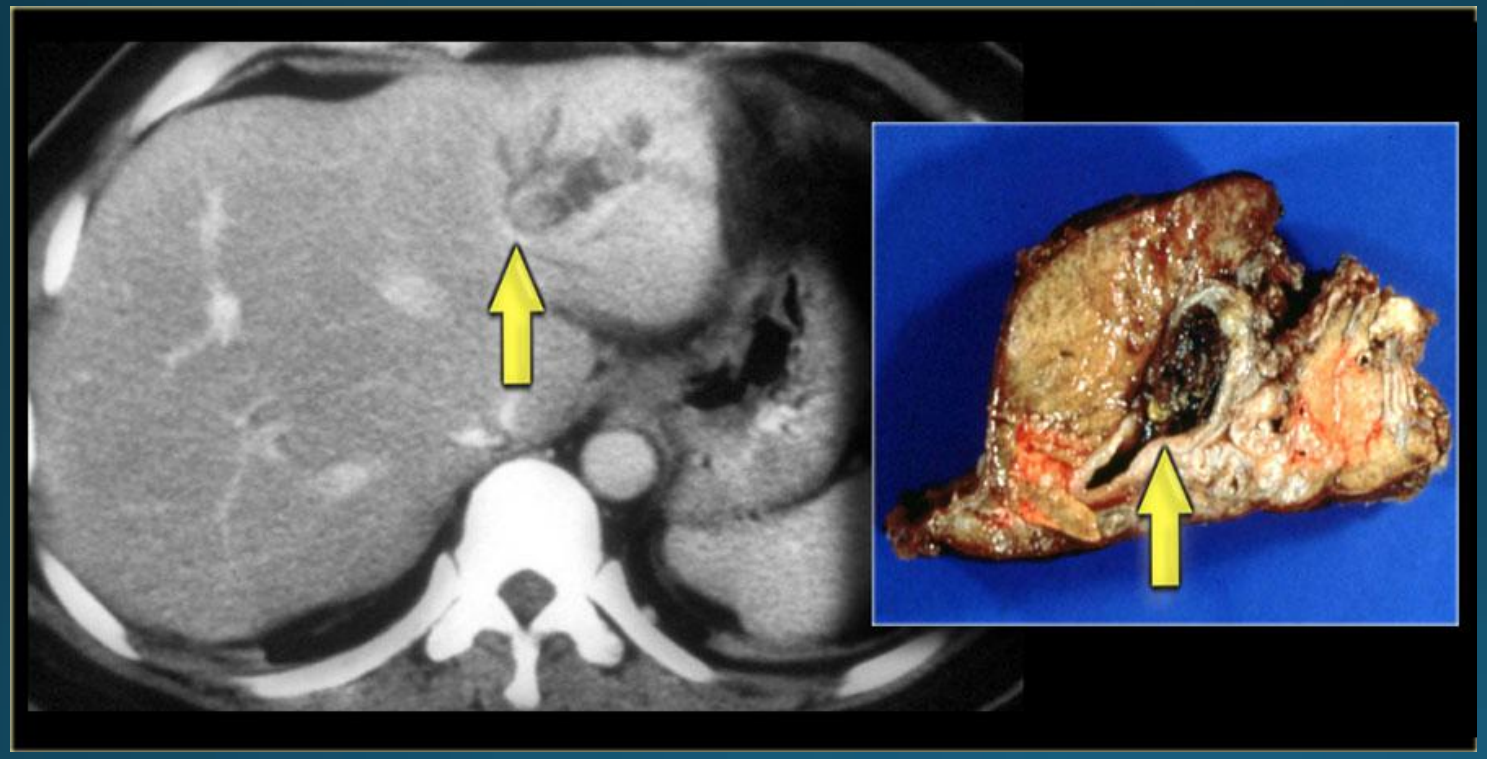


The left lobe is the most common location of the disease due to the delayed drainage of the left system.

a typical case.
There is focal dilatation of the bile ducts in the left lobe with stones.

Bile duct dilatation No Obstruction

- Recurrent pyogenic cholangitis



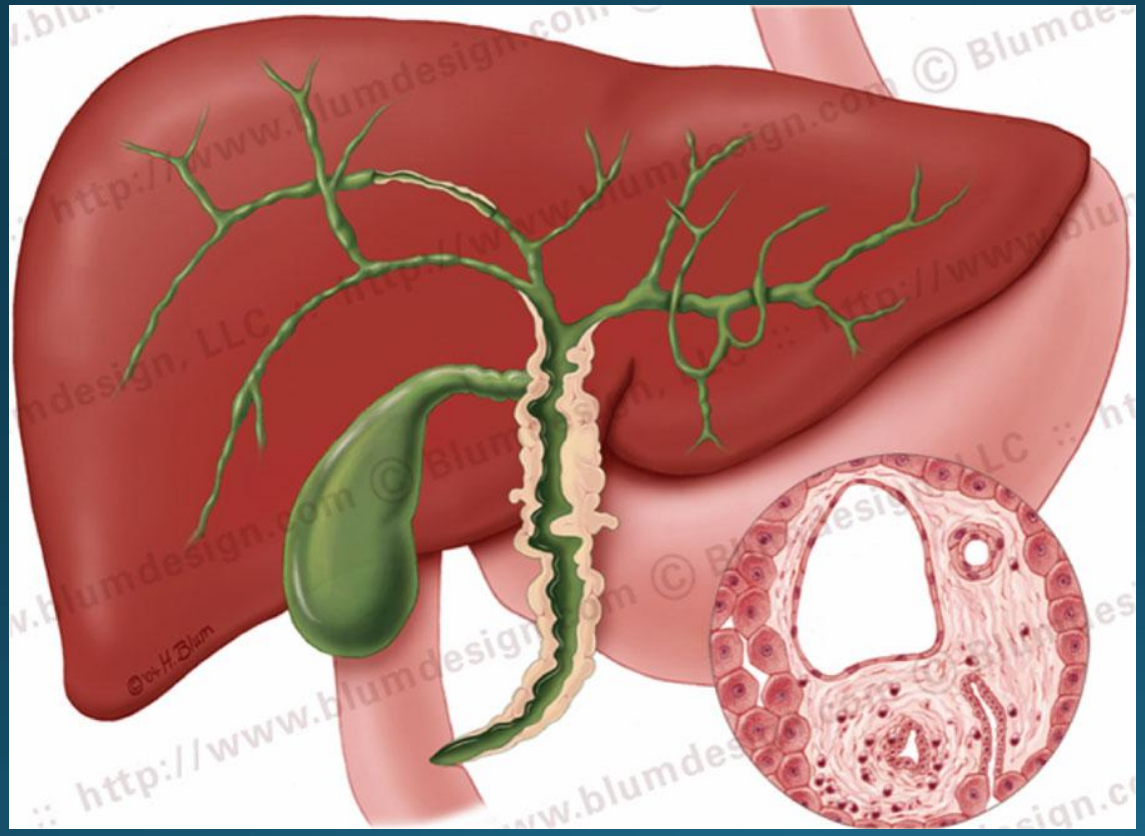
There is intrahepatic lithiasis with focal diatation.
A case like this is indistinguishable from focal Caroli disease with secundary stone formation.

Bile duct dilatation No Obstruction

- Primary Sclerosing Cholangitis
- The hallmark of PSC is strictures, but early on in the disease the strictures can be difficult to appreciate.
- The underlying abnormality in PSC is fibrosis, which is of unknown etiology.
- PSC is strongly associated with ulcerative colitis in up to 70% of patients, but it can also be associated with Crohn's disease of the large intestine.
- The reason for the association with IBD is unknown, but it is thought to be the result of an immune response.

Bile duct dilatation No Obstruction

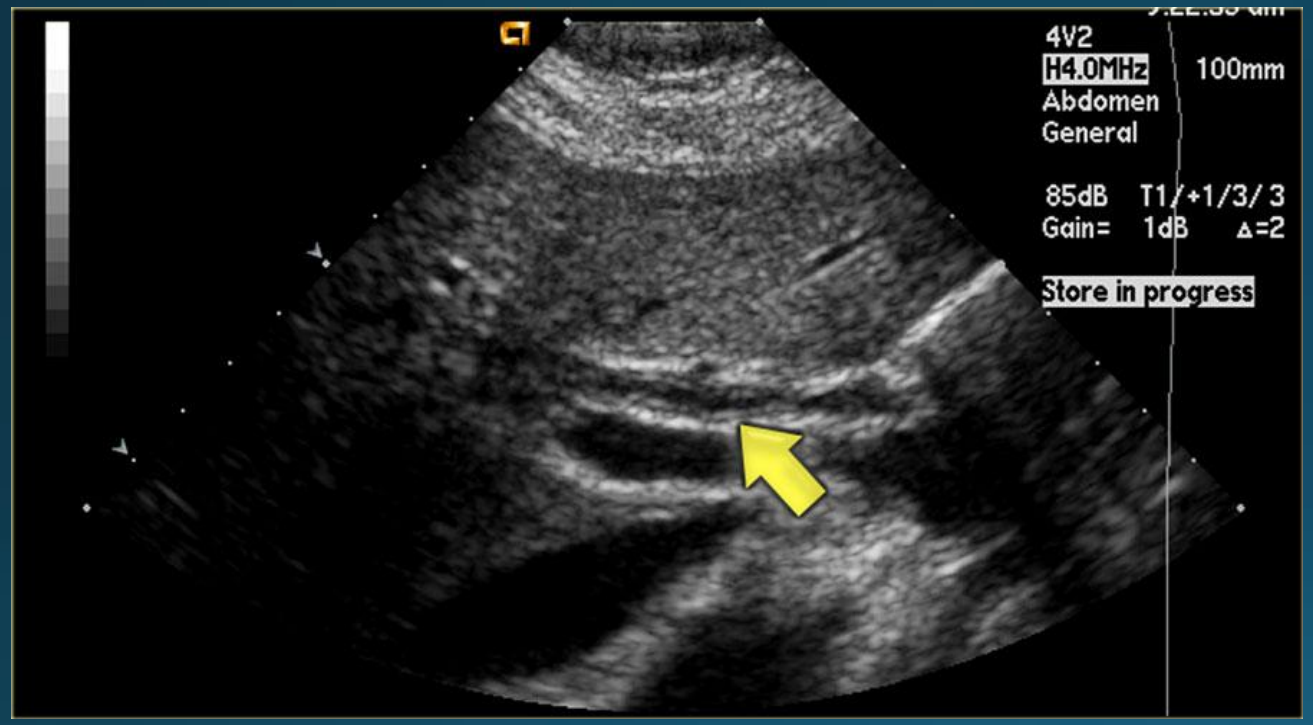
- Primary Sclerosing Cholangitis



Primary sclerosing cholangitis with strictures both in the intra- and extrahepatic bile ducts.

Bile duct dilatation No Obstruction

- Primary Sclerosing Cholangitis



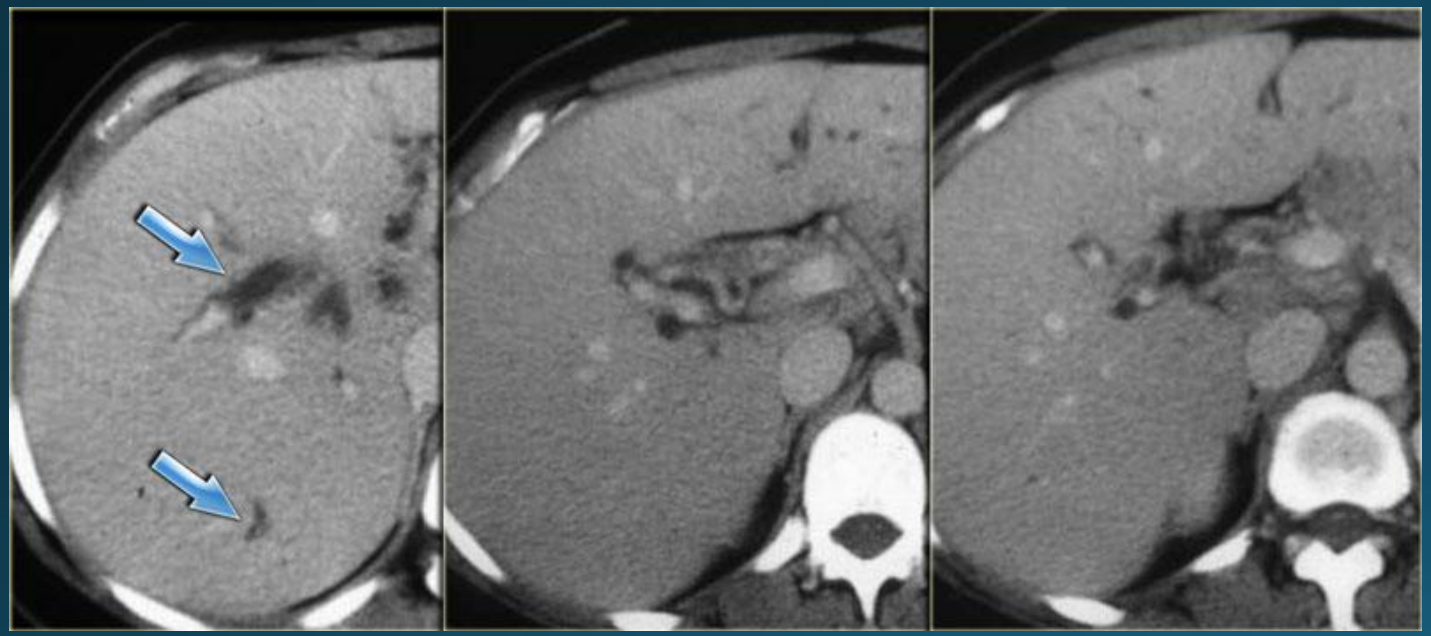
PSC with thickening of the wall of the bile duct (arrow)

Ultrasound findings

- One of the earliest features in PSC is on sonography where we see thickening of the wall of the bile duct as is seen in the image on the left.
- This patient came for an ultrasound examination to rule out gallstones.
- Notice that the intrahepatic ducts are normal.
- The differential diagnosis would include PSC, AIDS-cholangitis and cholangiocarcinoma.
- A cholangiocarcinoma would be rather unlikely, because there is no obstruction.

Bile duct dilatation No Obstruction

- Primary Sclerosing Cholangiti



Primary sclerosing cholangitis. CT findings

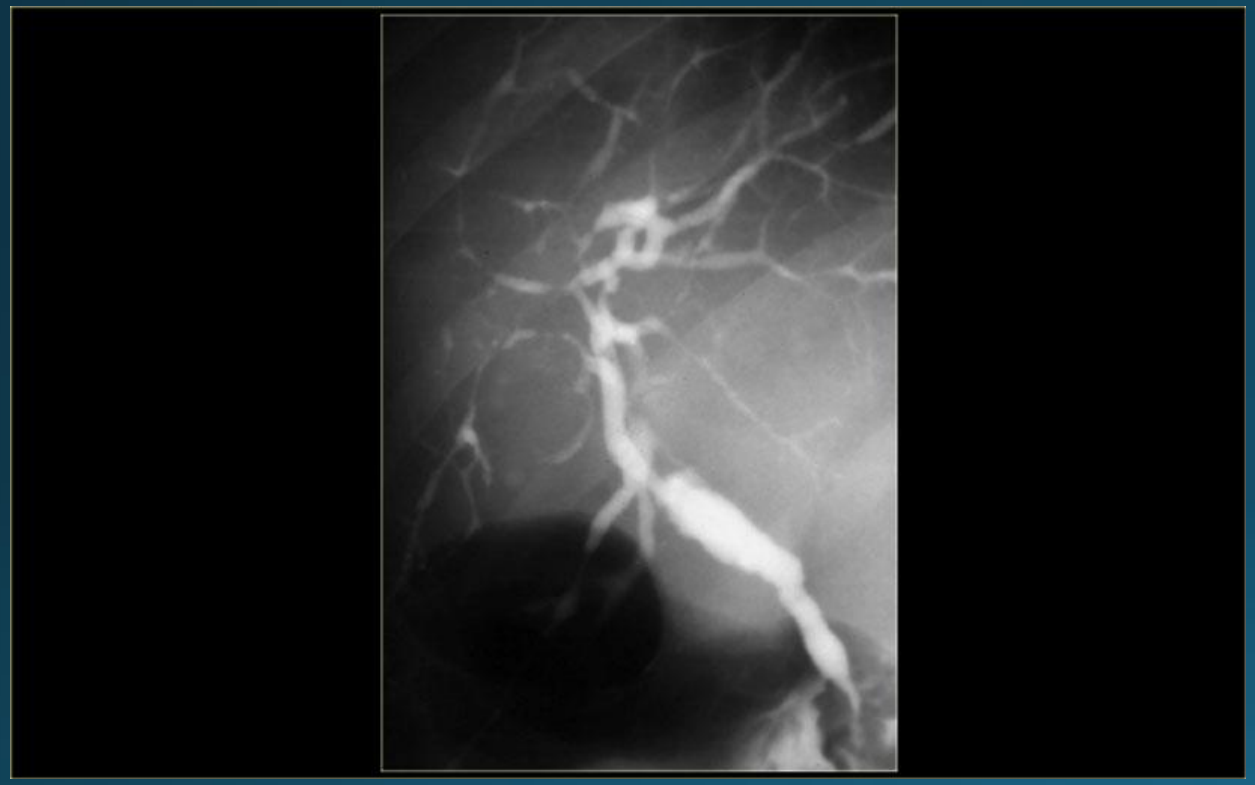
a patient with more pronounced CT findings.

The findings are:

Discontinuous dilatation
Bile wall thickening at the level of the porta hepatis
Lymphadenopathy

Bile duct dilatation No Obstruction

- Primary Sclerosing Cholangit



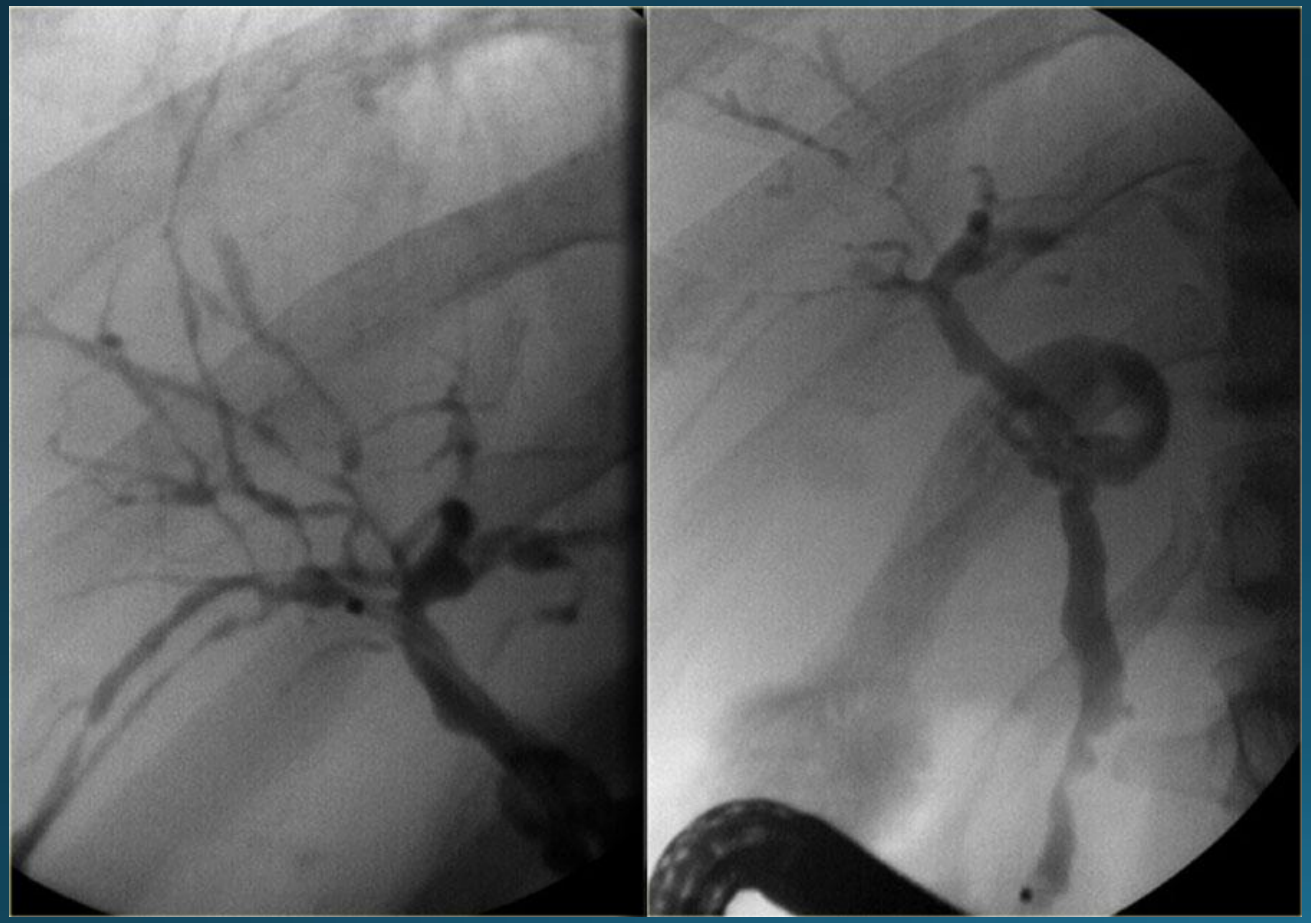
Cholangiography is used in the initial diagnosis of the disease, when there are only subtle strictures and in patients known with PSC to look for new strictures that are suspicious for carcinoma.

On cholangiography we can see:

- Beading: alternating pattern of strictures and normal or slightly dilated ducts
- Pruned-tree: distal bile ducts are narrowed and difficult to see
- Mural irregularity: irregular luminal margin (best seen on the left in the extrahepatic duct)
- Diverticula

Bile duct dilatation No Obstruction

- Primary Sclerosing Cholangit



typical findings in PSC.

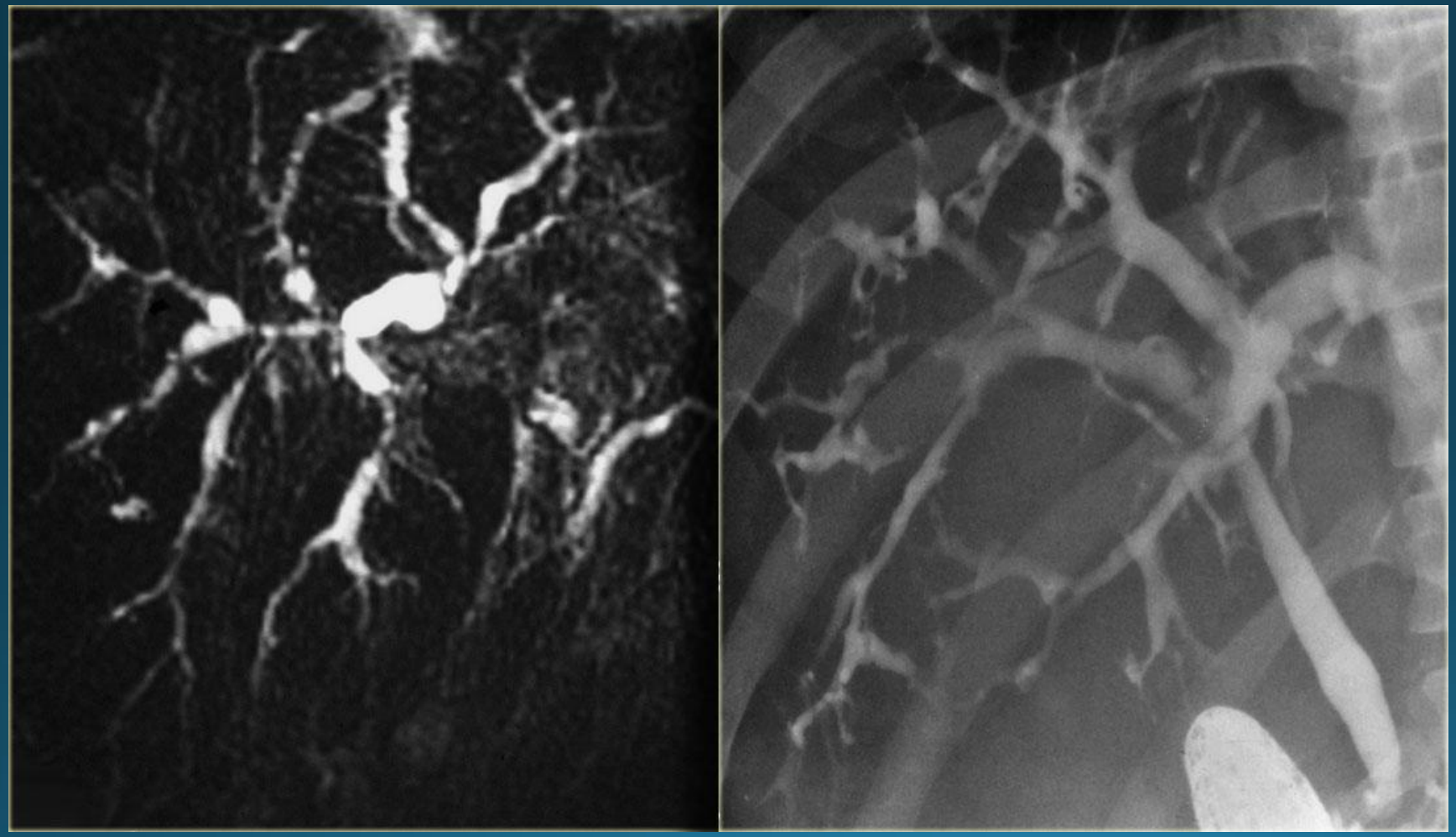
Notice the diverticula on the image on the right.

Diverticula are very specific for the diagnosis PSC.

So when you see these diverticula, you should immediately search for subtle strictures in the intrahepatic ducts.

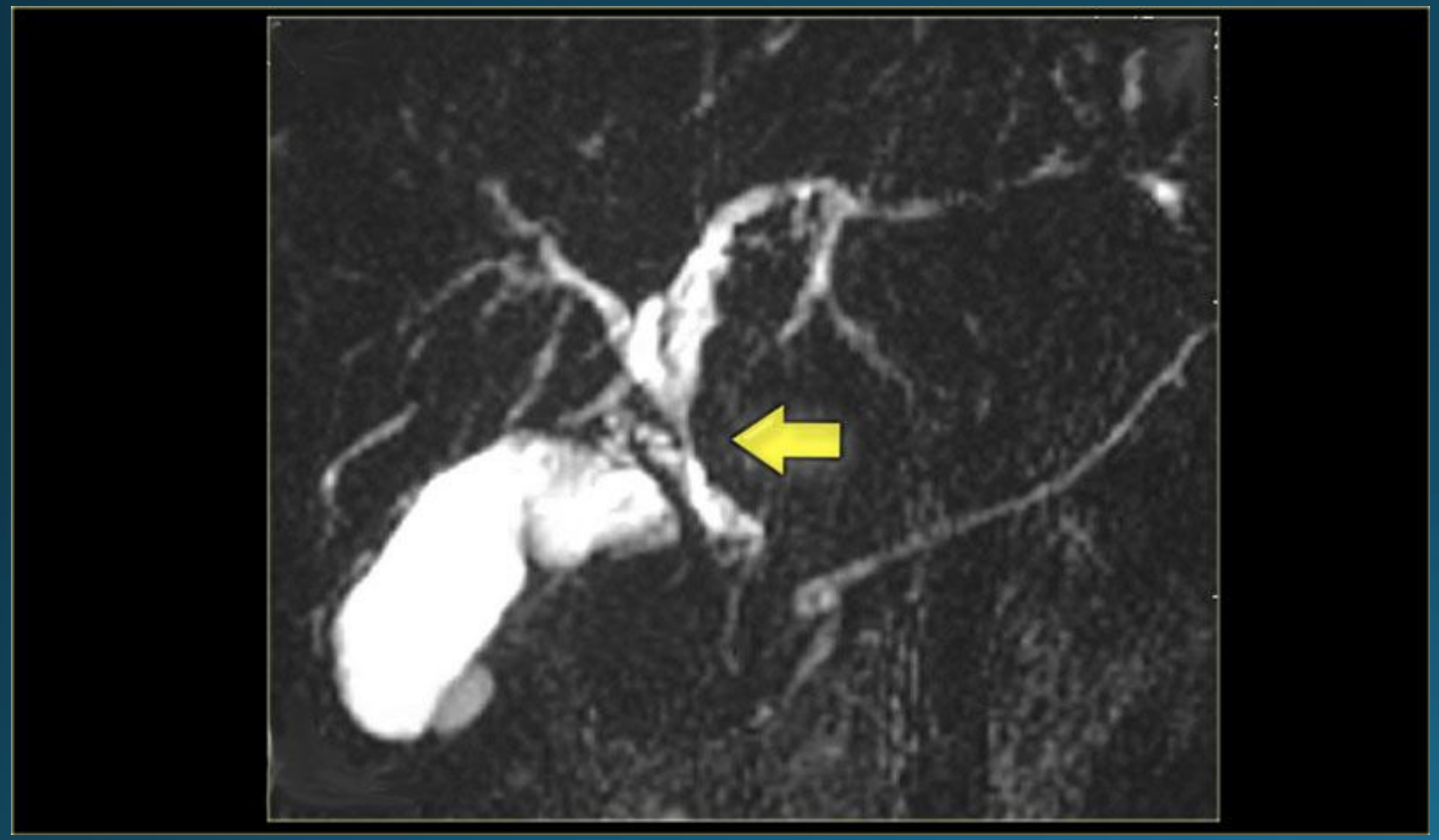
Bile duct dilatation No Obstruction

- Primary Sclerosing Cholangit



Bile duct dilatation No Obstruction

- Primary Sclerosing Cholangit



a MRCP in a pateint with PSC.

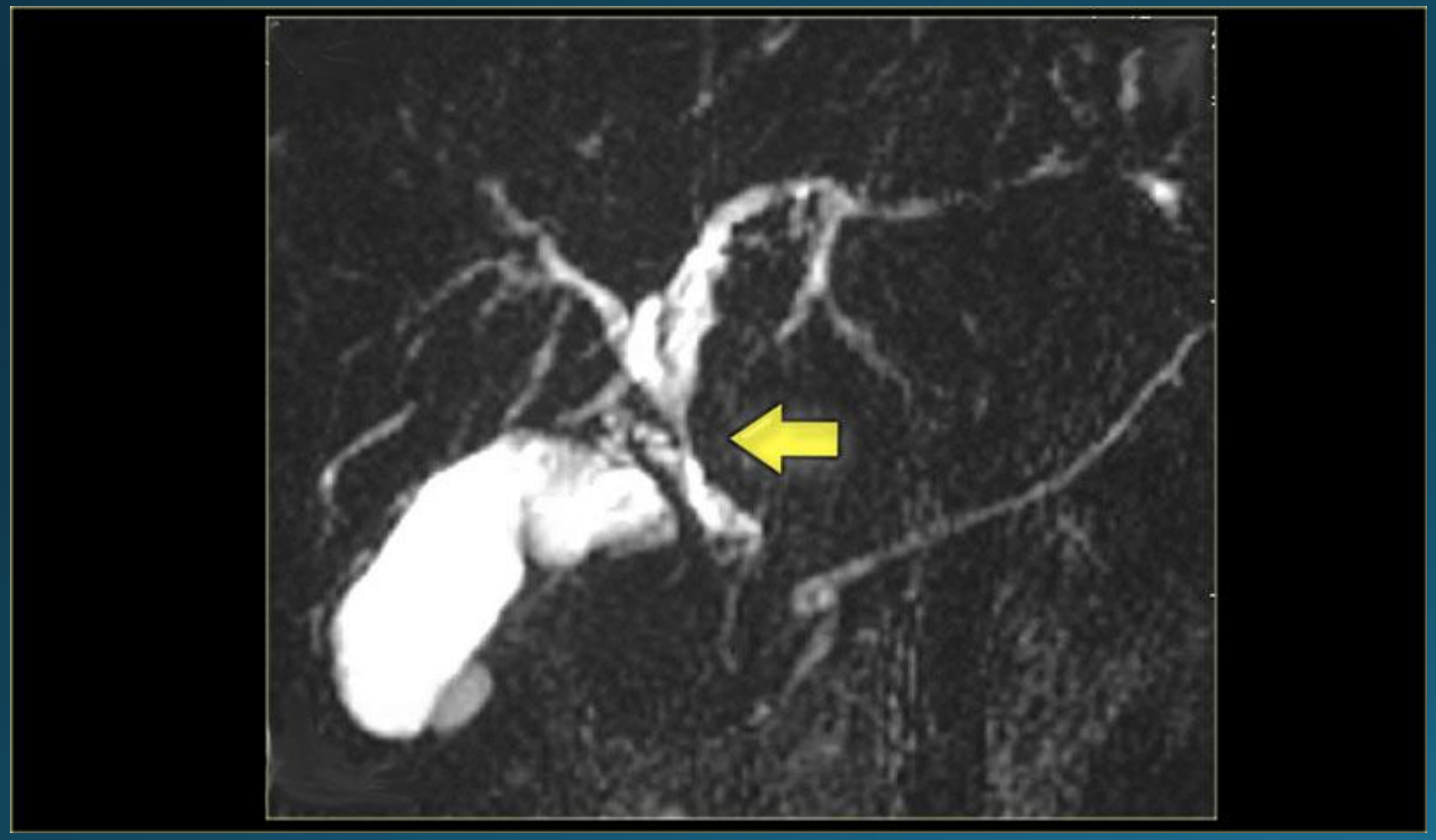
Notice the large stricture, which is quite worriesome for cholagiocarcinoma (arrow).

The strictures in PSC show an abrupt transission, while here we see 'shouldering', which indicates massefect.

In addition there is intrahepatic dilatation proximal to this stenotic area.

Bile duct dilatation No Obstruction

- Primary Sclerosing Cholangit



a MRCP in a pateint with PSC.

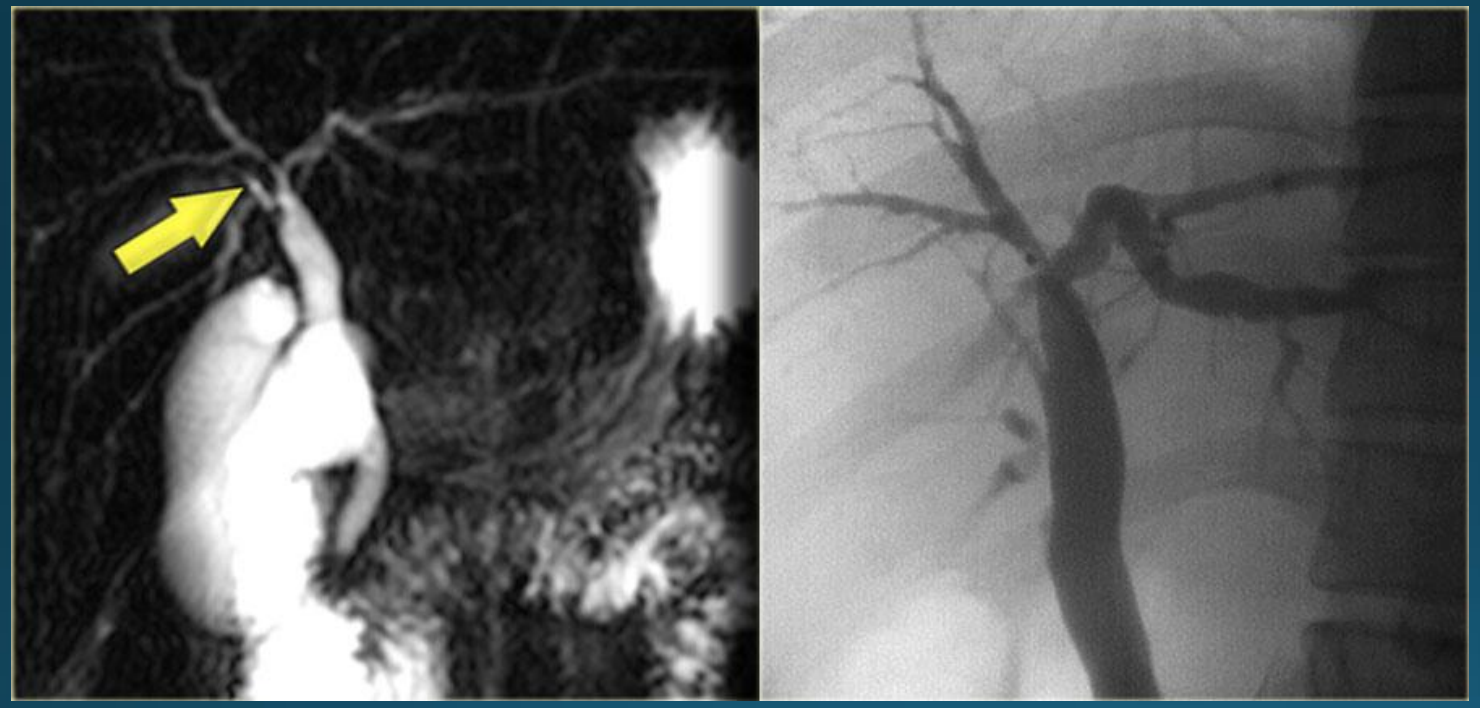
Notice the large stricture, which is quite worriesome for cholagiocarcinoma (arrow).

The strictures in PSC show an abrupt transission, while here we see 'shouldering', which indicates massefect.

In addition there is intrahepatic dilatation proximal to this stenotic area.

Bile duct dilatation No Obstruction

- Primary Sclerosing Cholangit



a MRCP in a patient demonstrating a stricture at the level of the hilum.
On MRCP this stricture looked long and worriesome for cholangiocarcinoma.
However, on the ERCP, the ducts have been distended with contrast and we can see that this is a short stricture compatible with the diagnosis of PSC.
During follow up this proved to be just PSC.

Bile duct dilatation Obstruction

- Cholangiocarcinoma
- Cholangiocarcinoma (i.e., adenocarcinoma of the bile duct) arises from the columnar epithelium of the bile duct.
- It is characterized by malignant glands within a desmoplastic stroma.
- These tumors have an infiltrative growth pattern and do not have a capsule.
- Cholangiocarcinoma is an uncommon tumor, that is mostly seen in patients with underlying benign bilairy disease.
- The incidence in the U.S. is 2000 to 2500 cases per year (coloncancer 150.000 per year).
- In Asian countries the incidence is ten times greater due to more chronic biliary infection.

Bile duct dilatation Obstruction

- Cholangiocarcinoma
- High risk groups are patients with:

Autoimmune diseases

- PSC, ulcerative colitis, primary biliary cirrhosis

Congenital anatomic anomalies

- Caroli, choledochal cyst, anomalous pancreaticobiliary junction
- Abnormal tumor suppressor genes, FAP, NF1

Infection

- Biliary parasites, recurrent pyogenic cholangitis

Clinical presentation:

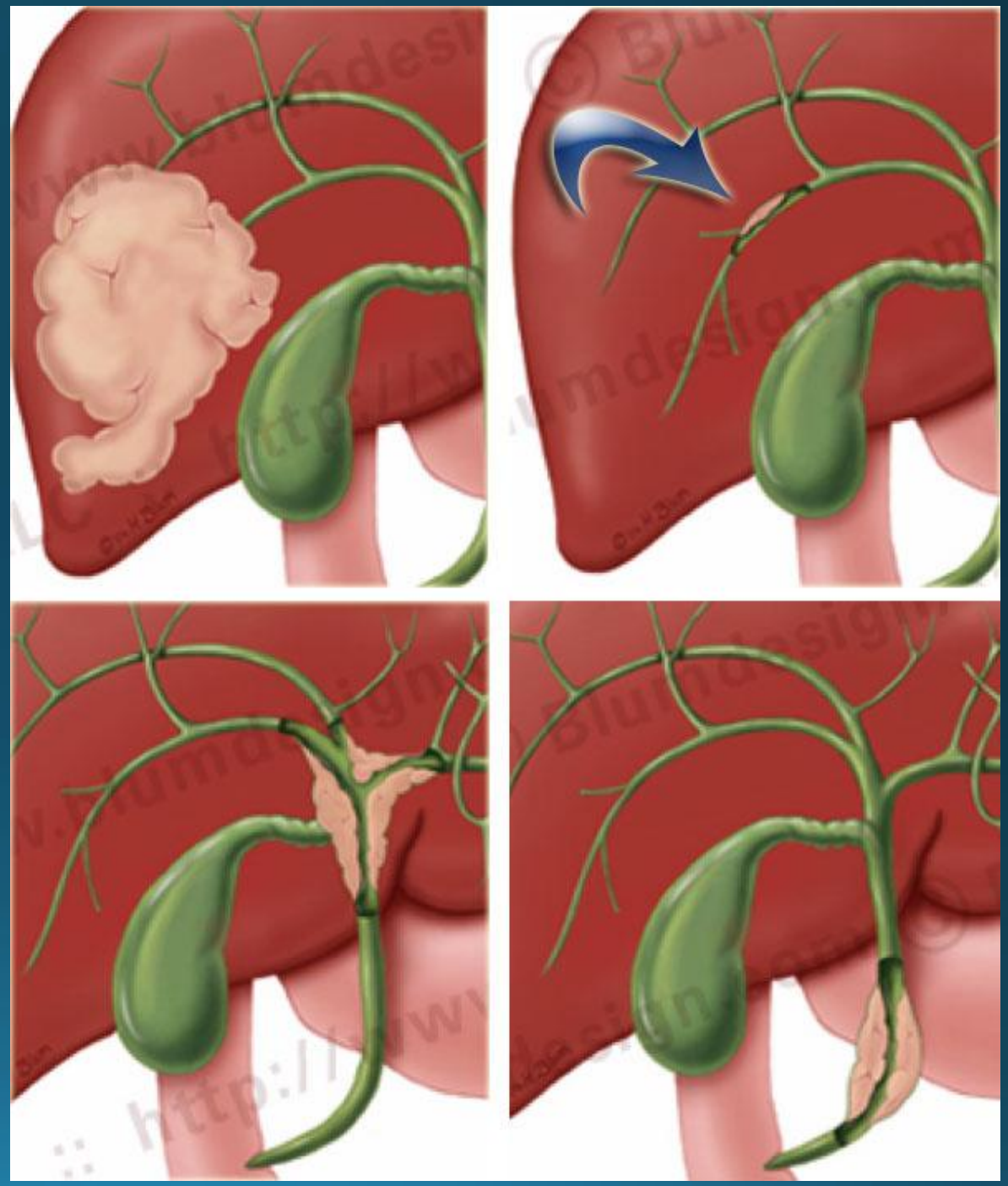
- Jaundice
- Pain
- Fever if secondary cholangitis

Bile duct dilatation Obstruction

Cholangiocarcinoma

There are four basic patterns of chlangiocarcinoma

- Intrahepatic cholangiocarcinoma
- Intraductal Cholangiocarcinoma
- Klatskin Tumor - Hilar Cholangiocarcinoma

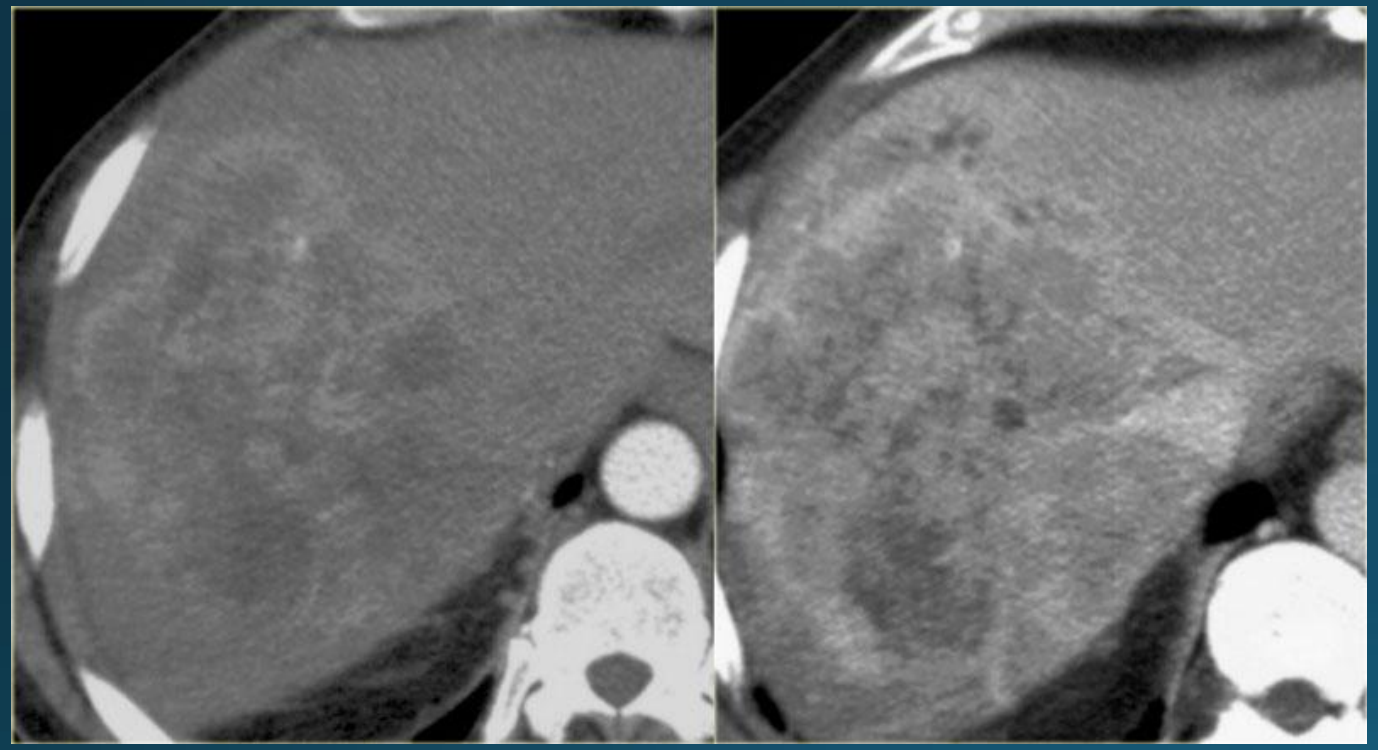


Cholangiocarcinoma

Intrahepatic cholangiocarcinoma

- These arise in the very small peripheral ducts.
- These tumors have abundant fibrous stroma that can cause retraction of the liver capsule.
- The tumor typically enhances in the equilibrium and delayed phases (5-10 minutes).

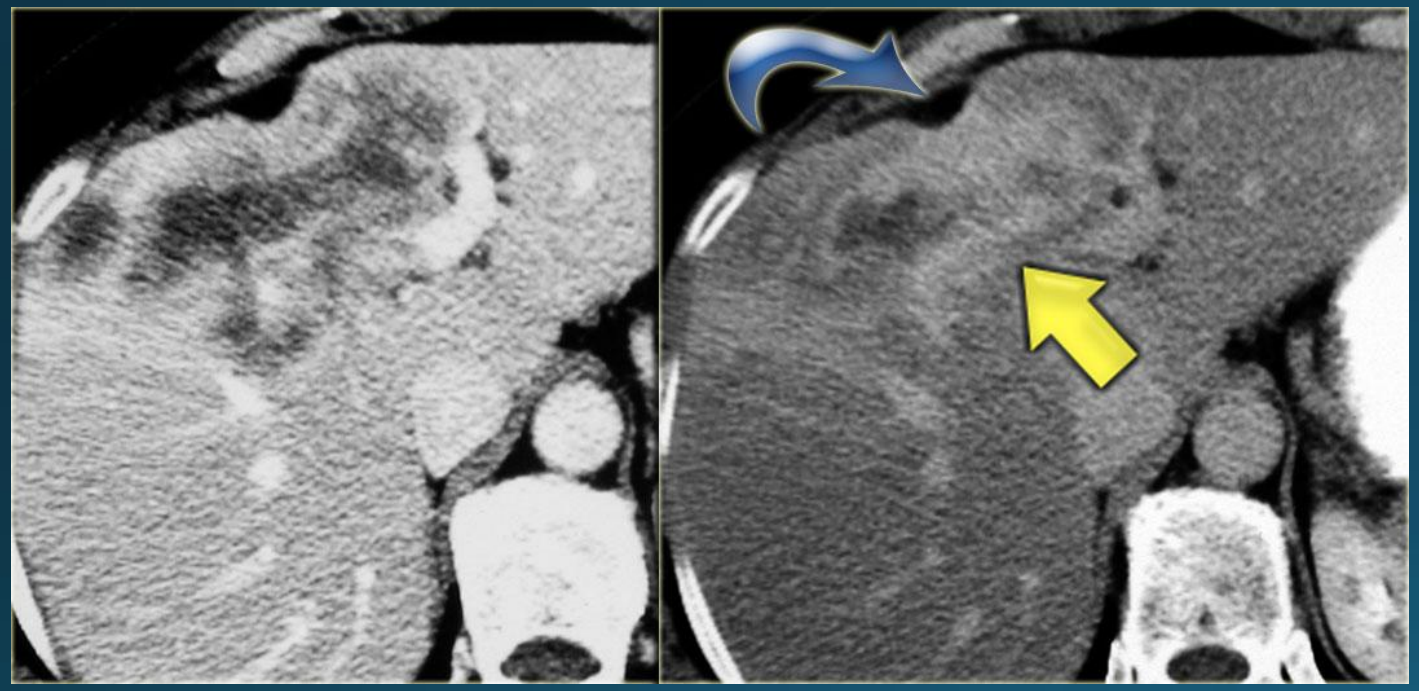
Cholangiocarcinoma



Intrahepatic cholangiocarcinoma

Although these tumors are usually quite heterogeneous because the contrast uptake is delayed and can be irregular.

Cholangiocarcinoma



Intrahepatic cholangiocarcinoma

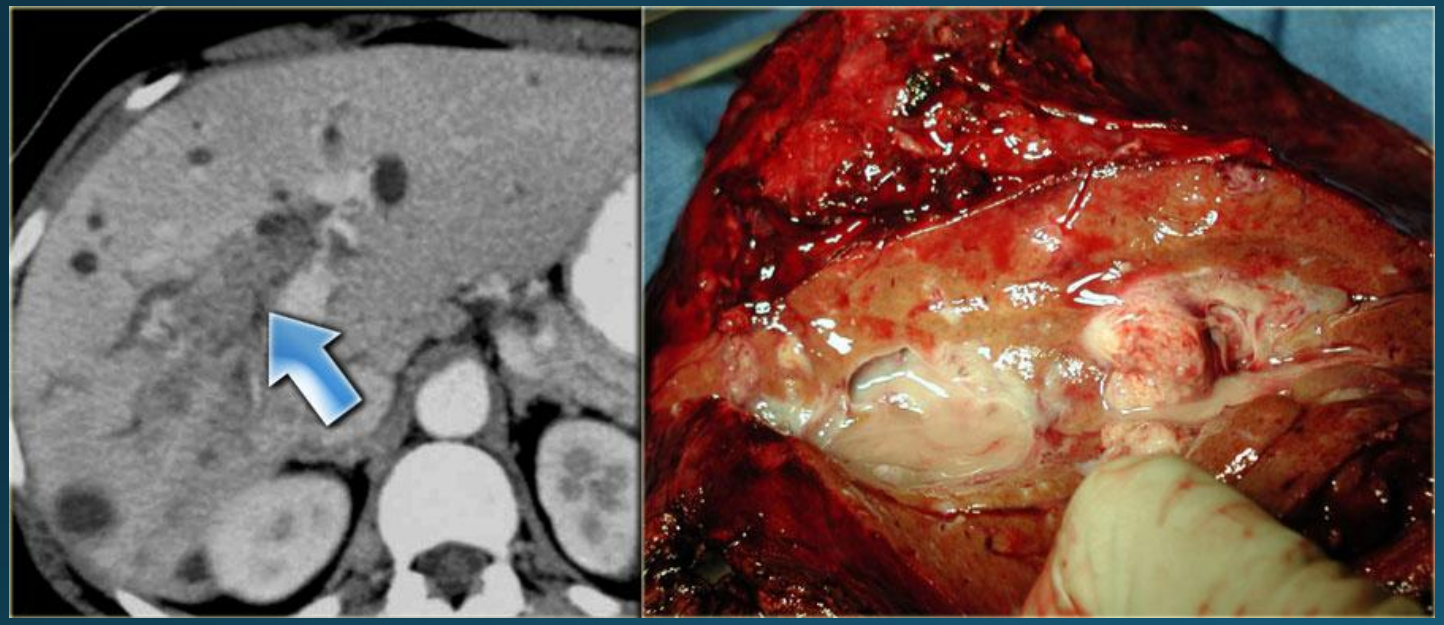
The key findings to look for are:

- Delayed enhancement
- Peripheral biliary dilatation
- Capsular contraction

On the left a typical case.

Notice the capsular retraction (blue arrow) and the late enhancement (yellow arrow).

Cholangiocarcinoma

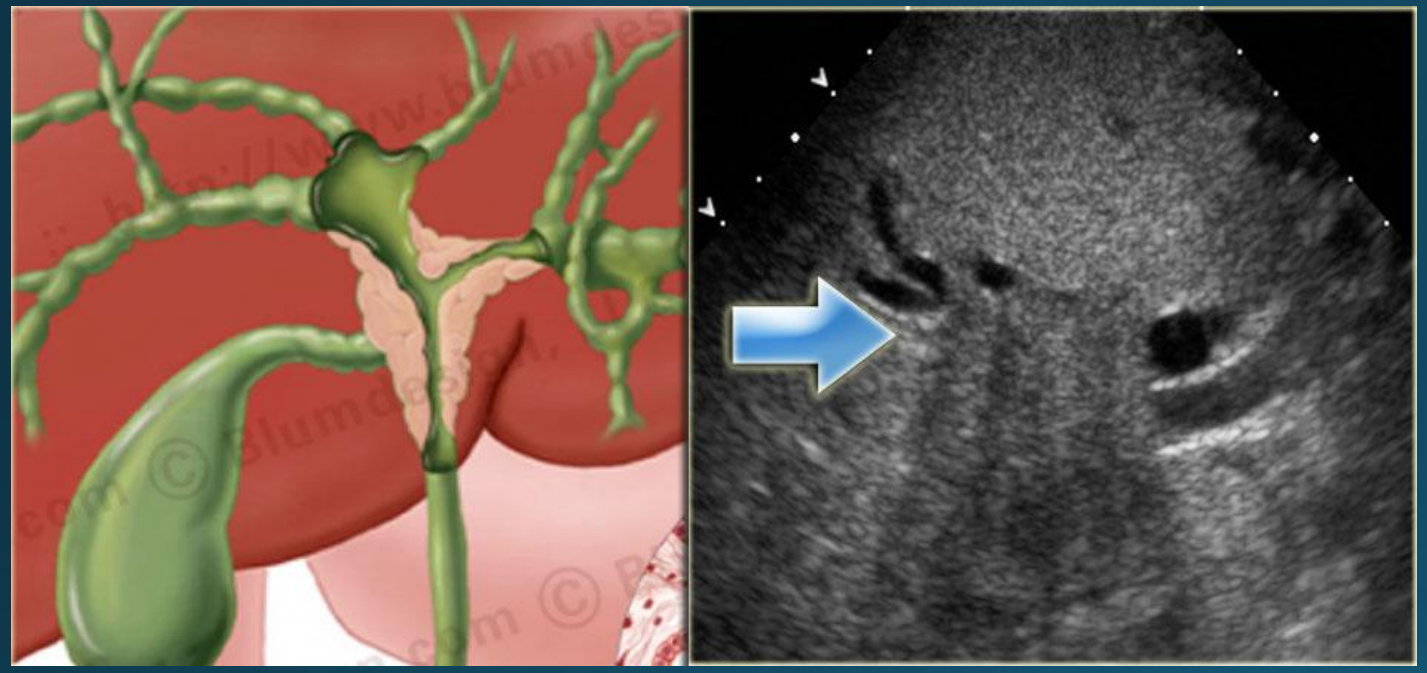


Intraductal Cholangiocarcinoma

These are very rare tumors.

They present as a intrabiliary mass with biliary dilatation peripheral to the mass.

Cholangiocarcinoma



Klatskin Tumor with dilatation of bile ducts in the right and left lobe of the liver
a nice correlation between an illustration and a sonographic image of a Klatskin tumor.
Notice how ill-defined the tumor is.

Klatskin Tumor - Hilar Cholangiocarcinoma

The most common site of biliary adenocarcinoma is at or near the confluence of the right and left hepatic ducts.

These tumors are also known as Klatskin tumors Klatskin tumors have an aggressive biologic behavior.

Imaging features:

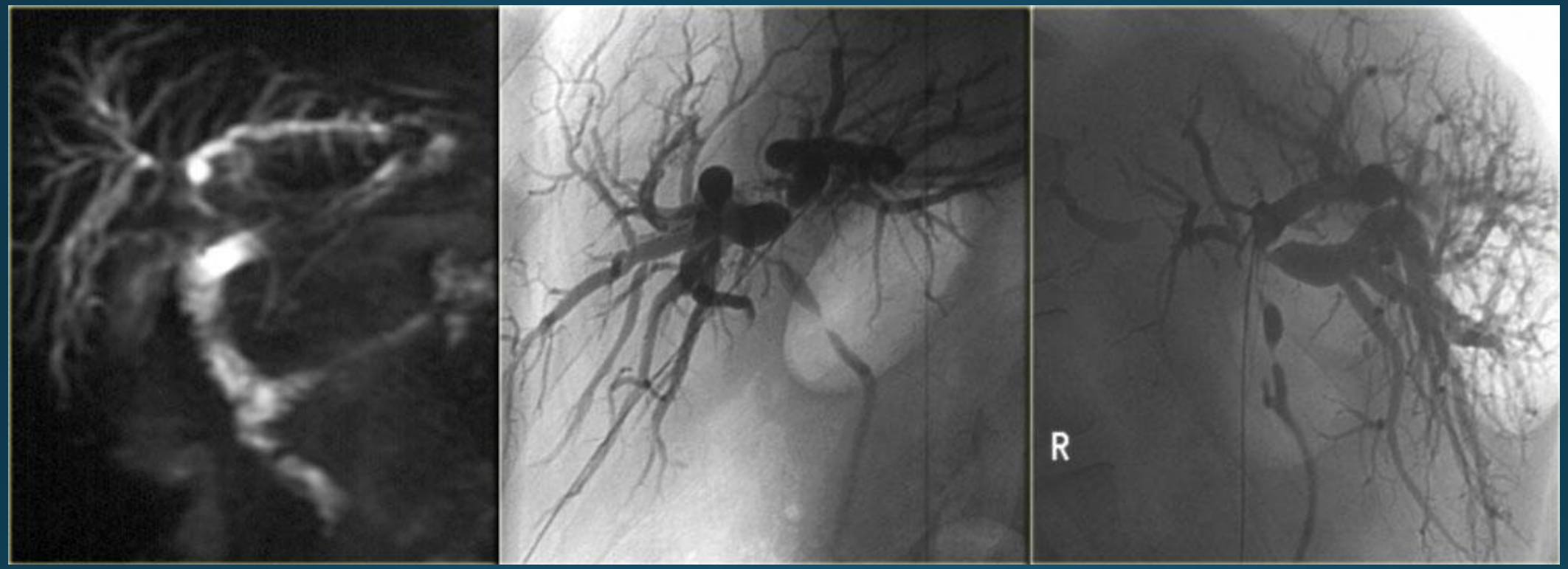
Duct dilatation

Ill-defined mass

Lobar atrophy

Vascular invasion

Cholangiocarcinoma



Klatskin Tumor - Hilar Cholangiocarcinoma

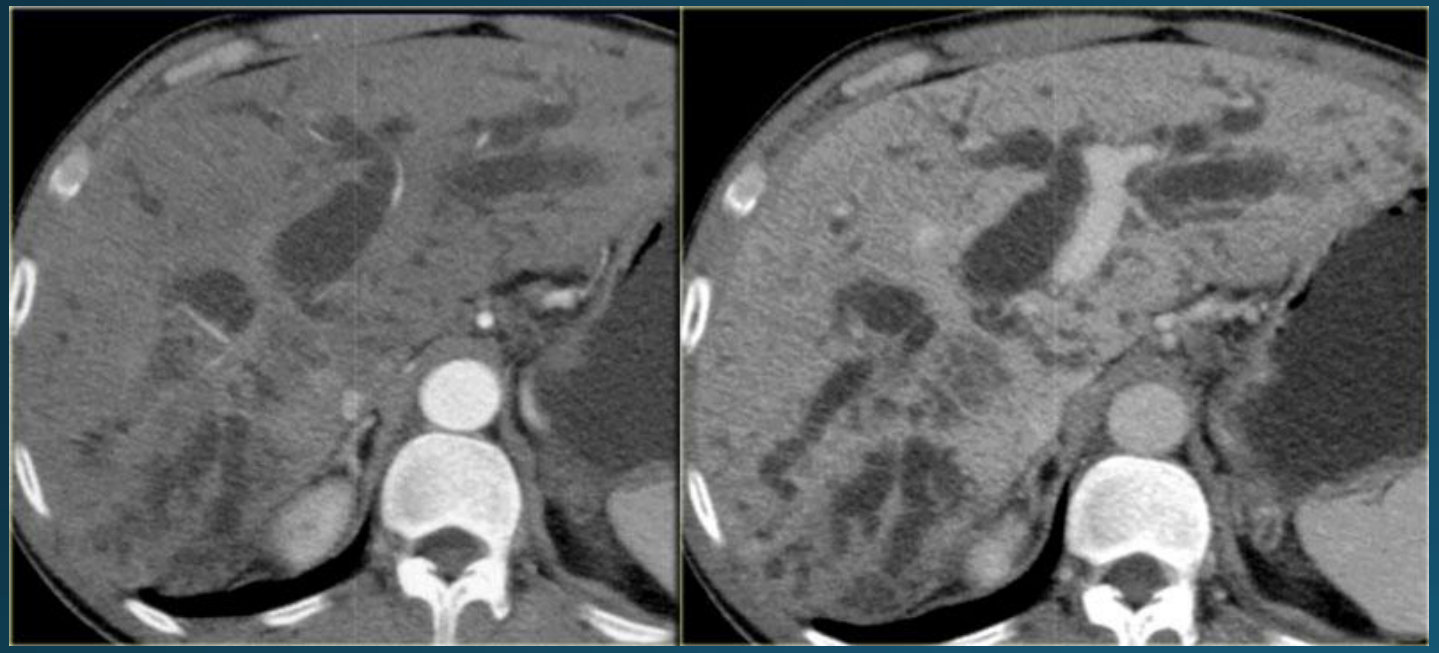
In these tumors it may be difficult to get a definitive diagnosis pre-operatively.Biopsy is almost impossible and results of endoscopic brushing are commonly negative.

The staging is done with cholangiography and is based on the finding of mass effect (shouldering), irregular margins and abrupt tapering at the obstruction.

The limitations of MRCP in staging are the spatial resolution and the inability in the evaluation of the secondary ducts.

ERCP is superior to MRCP (figure)

Cholangiocarcinoma



Klatskin Tumor: arterial and portal venous phase

Klatskin Tumor - Hilar Cholangiocarcinoma

In the case on the left we can identify the tumor at the confluens of the left and right hepatic duct.

The margins of the tumor however are imperceptible because of the infiltrative growth.

Based on the CT it is not possible to stage the tumor correctly.

Cholangiocarcinoma

Klatskin Tumor - Hilar Cholangiocarcinoma

In these tumors it may be difficult to get a definitive diagnosis pre-operatively.

Biopsy is almost impossible and results of endoscopic brushing are commonly negative.

The staging is done with cholangiography and is based on the finding of mass effect (shouldering), irregular margins and abrupt tapering at the obstruction.

The limitations of MRCP in staging are the spatial resolution and the inability in the evaluation of the secondary ducts.

ERCP is superior to MRCP (figure)

Cholangiocarcinoma

Klatskin Tumor - Hilar Cholangiocarcinoma

Resectability

These tumors are unresectable when:

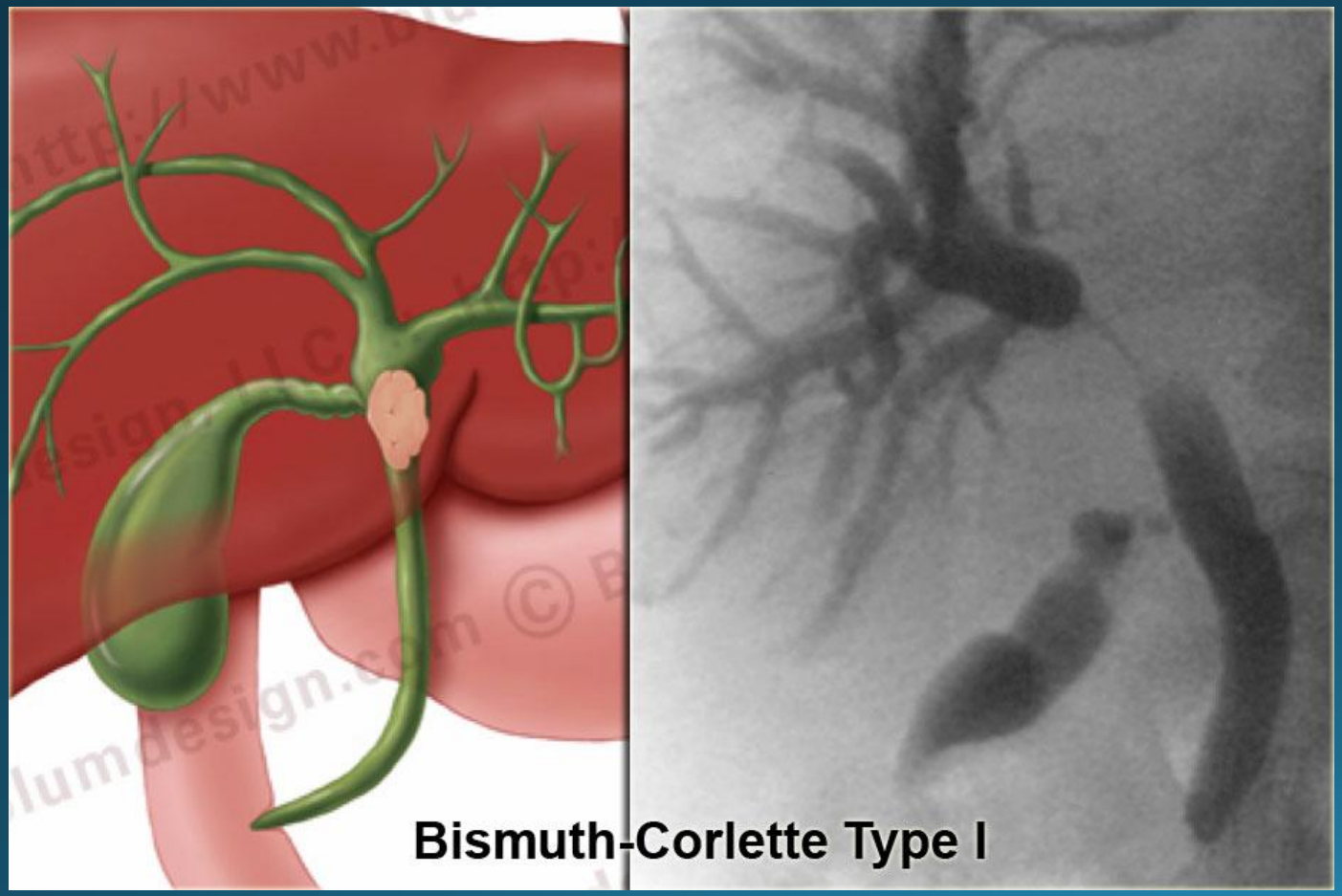
- Bilateral tumor extension
    - Into secondary ducts
    - Into hepatic parenchyma
    - Hepatic artery or portal vein
- Occlusion main portal vein
- N2 nodes (nodes around the pancreas)
- Distant metastases

Klatskin Tumor - Bismuth-classification

Klatskin Tumor - Bismuth-classification

Bismuth-Corlette type I

- A type I tumor is a lesion limited of the common hepatic duct, i.e. below the confluence.
- These patients can undergo resection with bile duct recontruction because the confluence is normal.

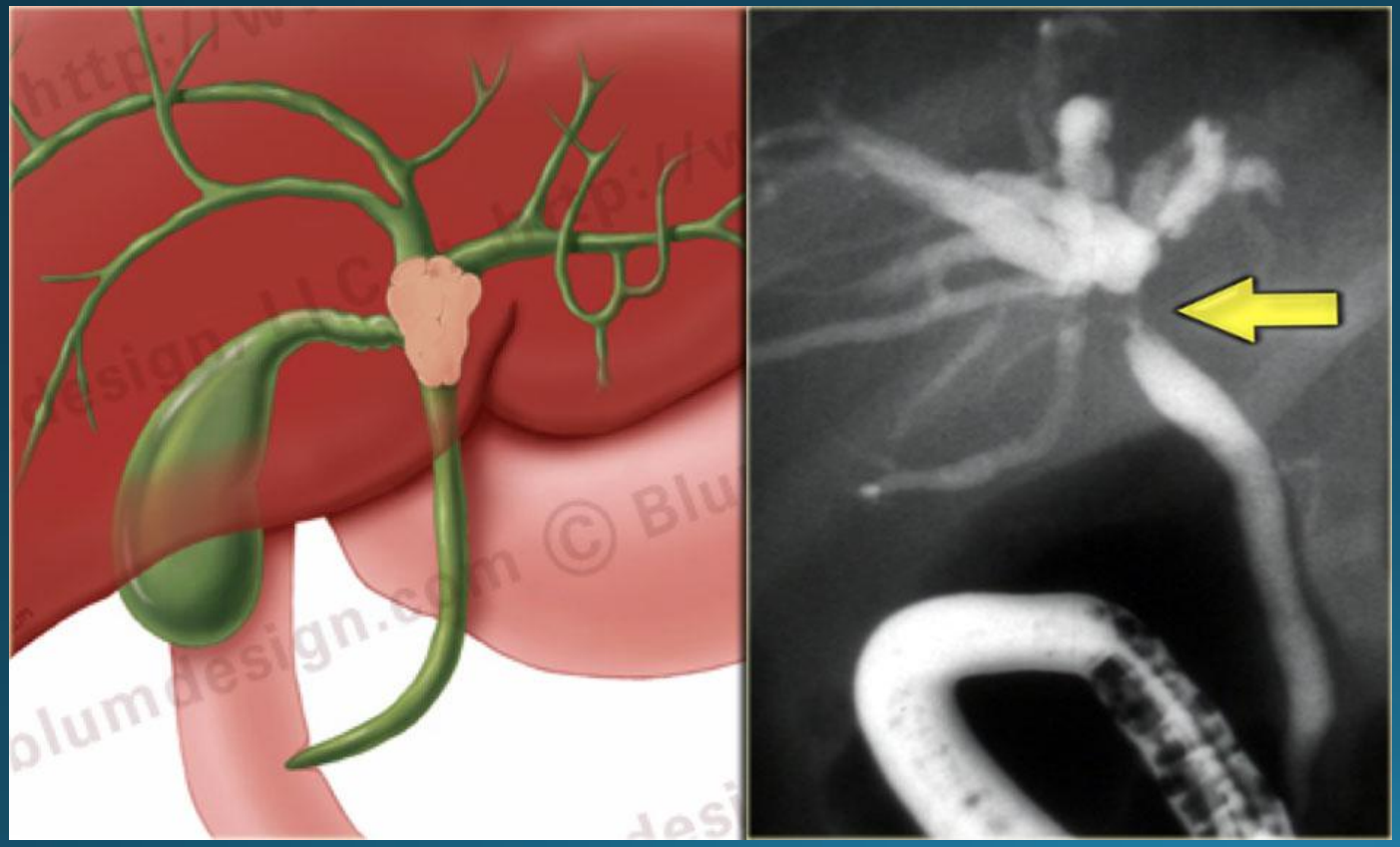


Bismuth-Corlette type I :
tumor with abrupt stricture and shouldering below the confluens

Klatskin Tumor - Bismuth-classification

Bismuth-Corlette type II

- A type II tumor is a lesion that extends to the confluence.
- These tumors are potentially resectable

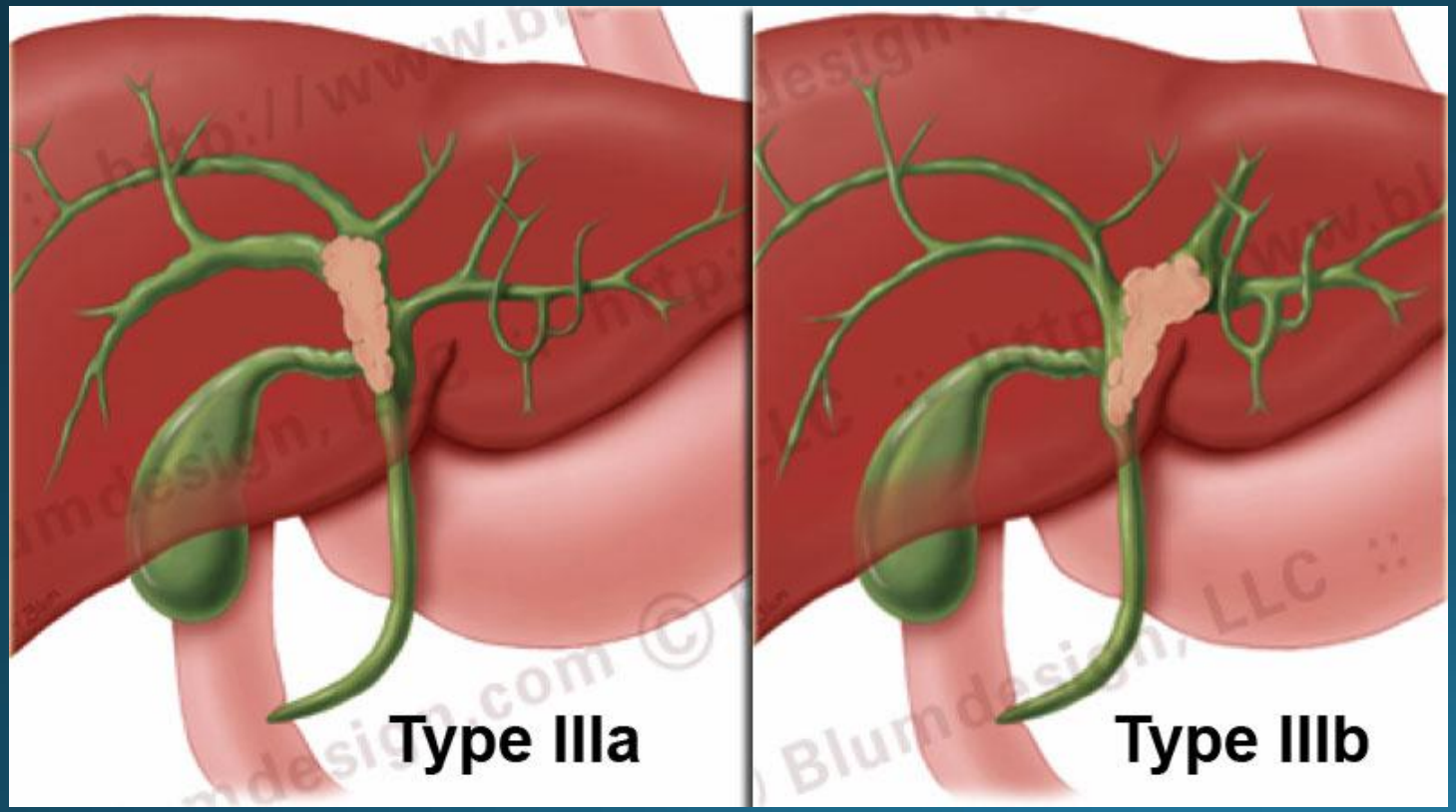


Bismuth-Corlette type II :
tumor with extention into the origin of the right and left hepatic duct.

Klatskin Tumor - Bismuth-classification

Bismuth-Corlette type IIIa and IIIb

- A IIIa-tumor extends into the right and a IIIb-tumor extends into the left hepatic duct.

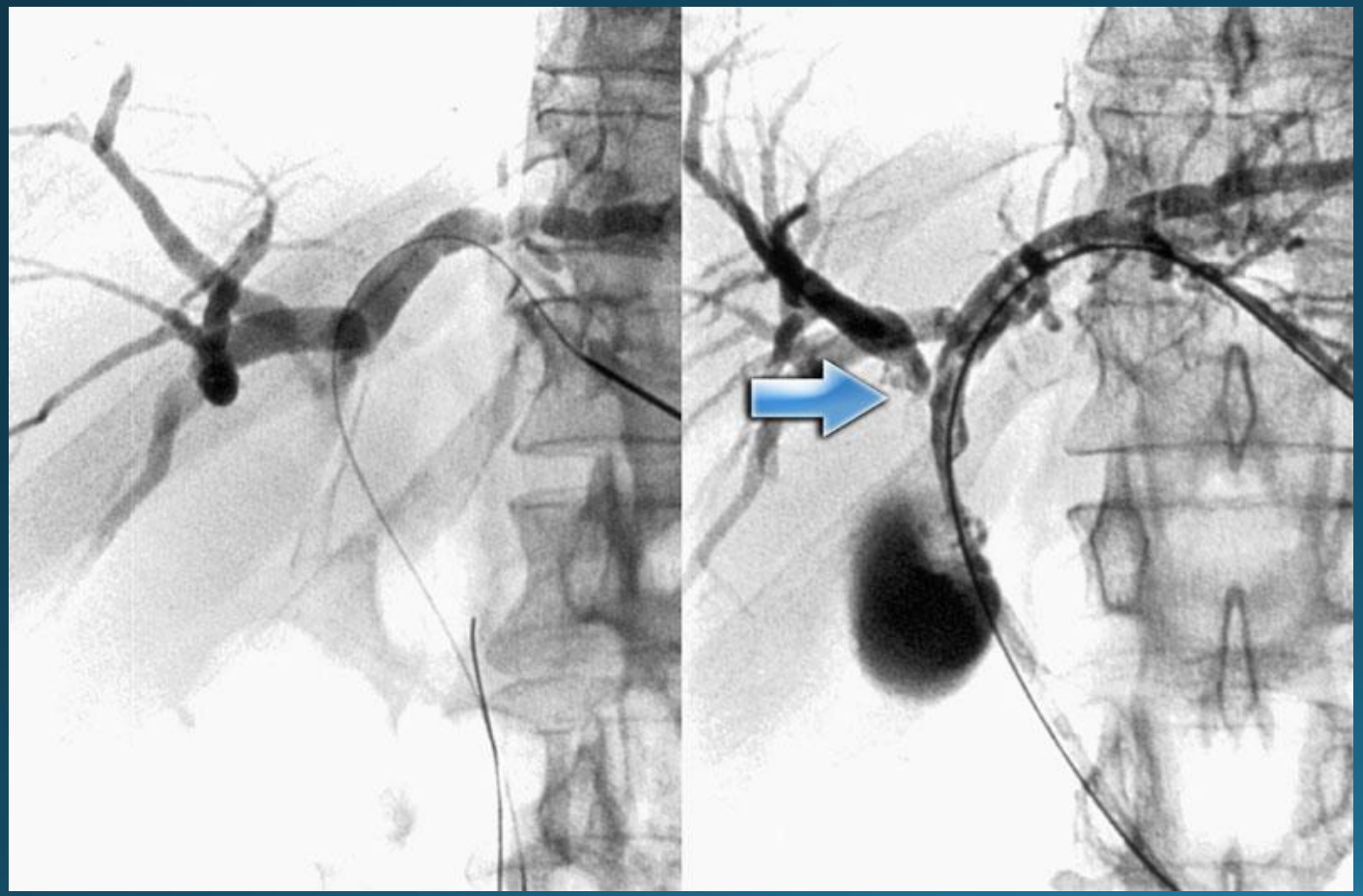


Bismuth-Corlette type III

Klatskin Tumor - Bismuth-classification

Bismuth-Corlette type IIIa and IIIb

- A IIIa-tumor extends into the right and a IIIb-tumor extends into the left hepatic duct

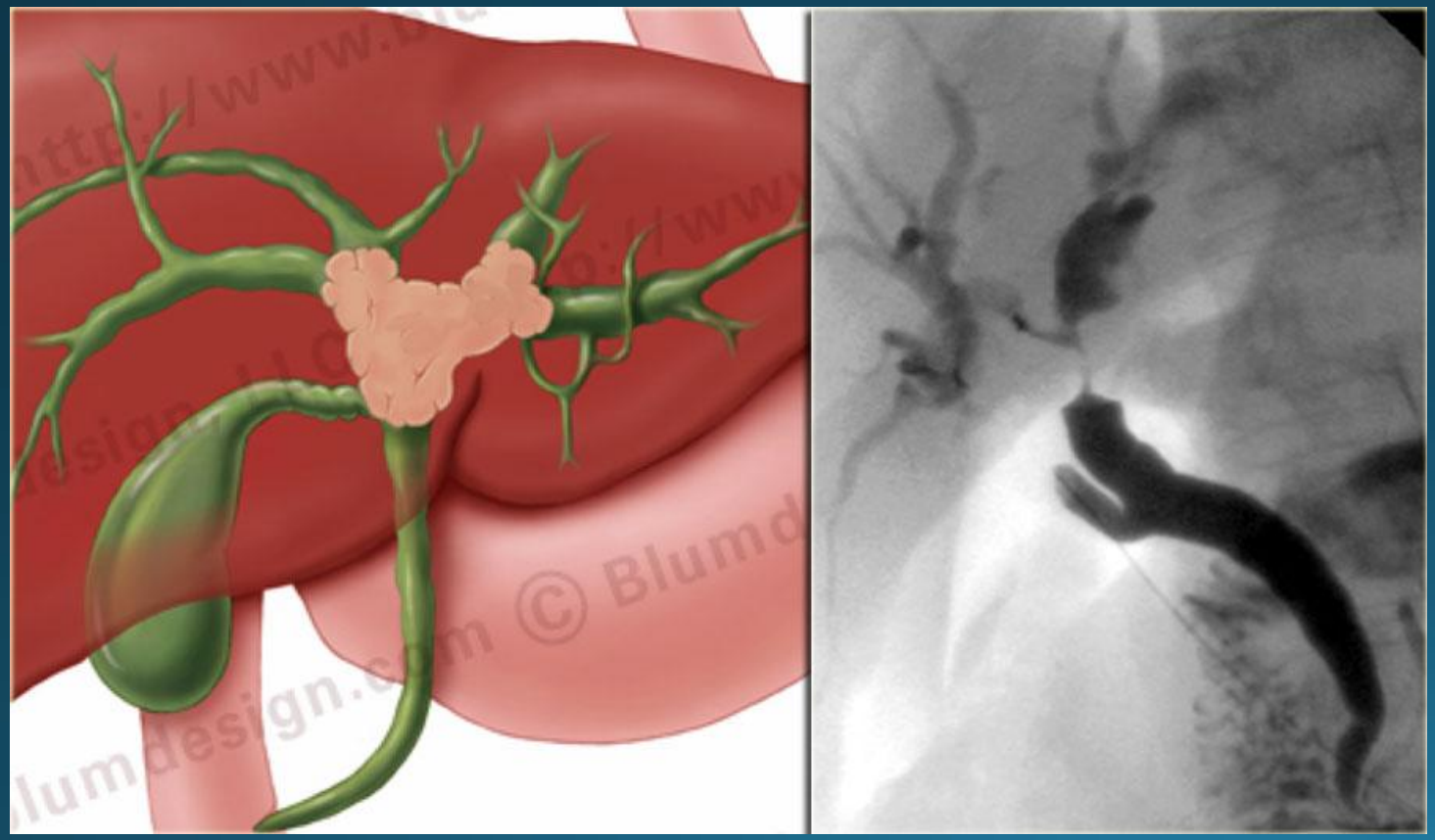


PTC-images of a type IIIa-tumor.
The arrow indicates the extention into the right hepatic duct.
The left duct is normal.
This patient can undergo a resection of the right lobe of the liver.

Klatskin Tumor - Bismuth-classification

Bismuth-Corlette type IV

- an illustration and ERCP of a type IV-tumor with extention into the right and left duct.
- A type IV tumor is unresectable.



Bismuth-Corlette type IV

- Thank for attention