e-Poster
ESGAR 2010, JUNE 2 – 5, DRESDEN/DE

Anatomic Variants, Congenital Anomalies and Pathology of the Extrahepatic Bile Ducts

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Congress: ESGAR 2010

Type: Educational Exhibit

Topic: Diagnostic / Bile Ducts

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Biliary Tract Diseases [C06.130]

Bile Ducts [A03.159.183]

Keywords: Bile ducts, Pathology, Congenital anomalies, Anatomic variants

1. Learning Objectives

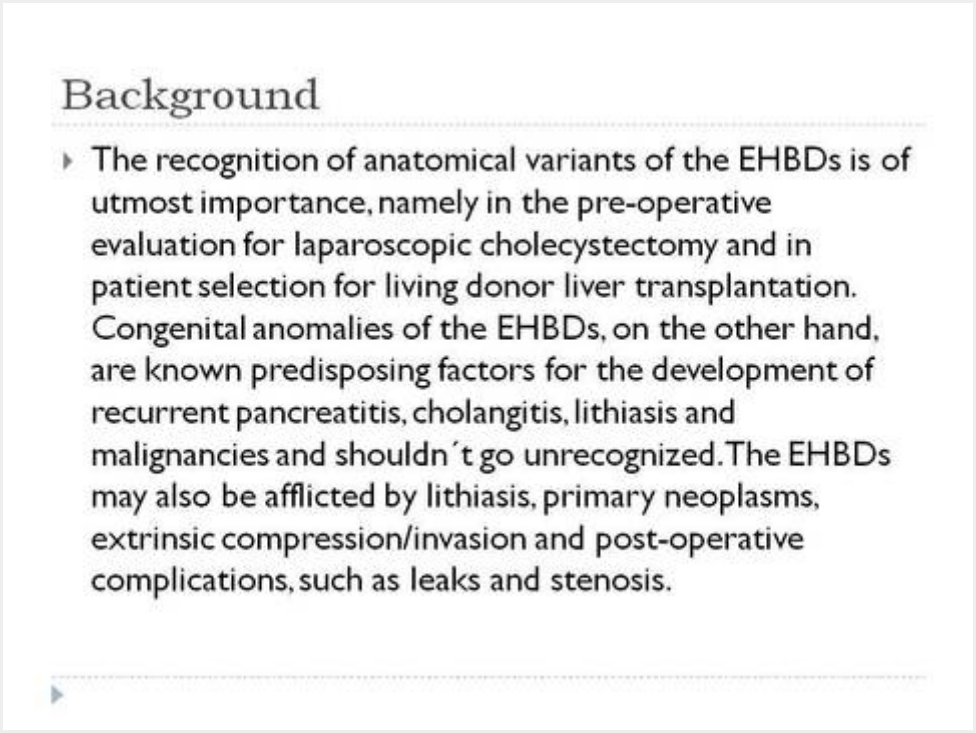
Learning Objectives

slide1.jpg

2. Background

Background

vb.jpg



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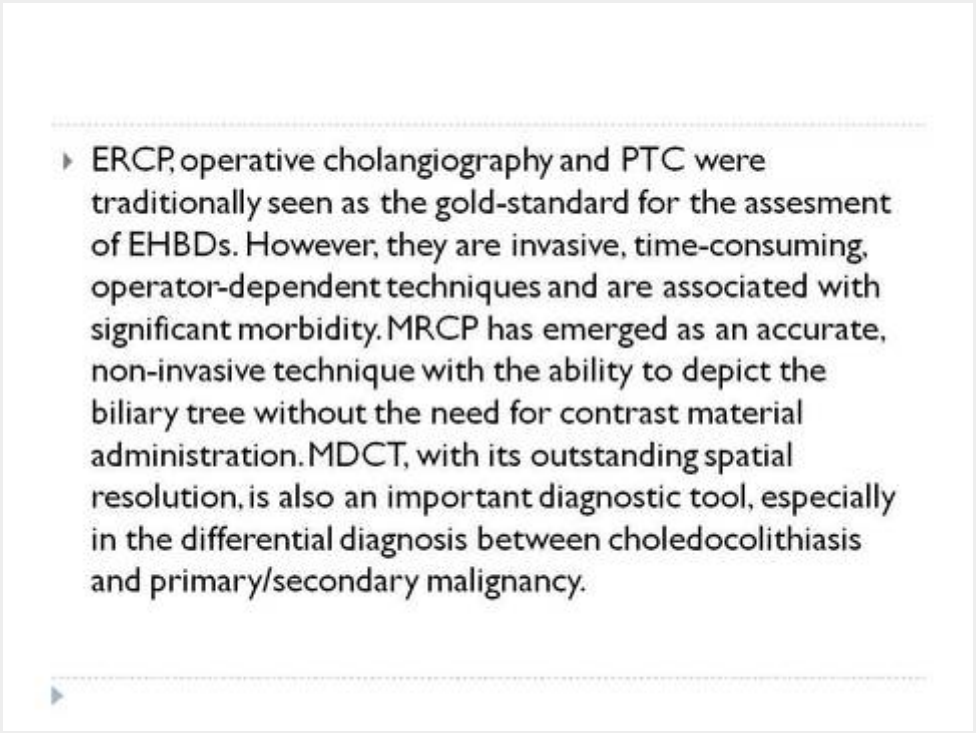


3. Imaging Findings/Procedure Details

Imaging Findings

slide2.jpg

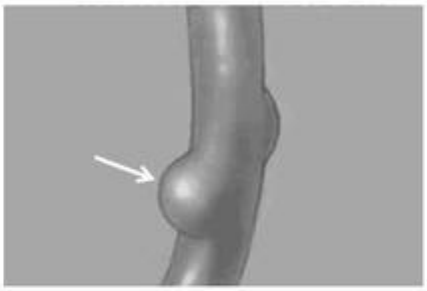
Content Organization

1. Embriology
2. Normal Anatomy and Anatomic Variants
3. Iatrogenic Complications
4. Congenital Anomalies
5. Pathology

slide3.jpg

1. Embriology

- At the beginning of the 4th embrionic week, an endodermal outgrowth of the ventral distal foregut arises – the hepatic diverticulum (→).



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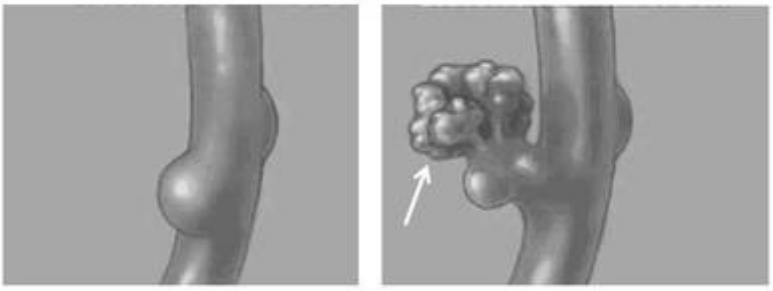
slide4.jpg

1. Embriology

- The cranial part of the hepatic diverticulum (*pars hepatica*) develops into the liver primordium (→).



Adapted from www.vesalius.com

slide5.jpg

1. Embriology

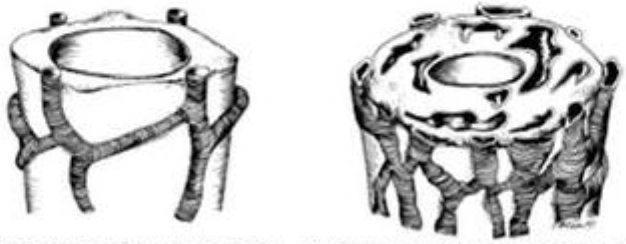
- The liver primordium originates liver cells and ductal plates (→), which develop in the mesenchyme adjacent to portal vein branches (→)
- The ductal plates later on remodel to form the intrahepatic bile ducts (→)

Illustration by Aletta Frazier

slide6.jpg

1. Embriology

- Ductal plate malformations result from abnormal persistence of embryonic bile duct structures, which may present as cystic, non-communicating lesions, or too numerous and ectatic bile ducts



NORMAL PORTAL TRACT DUCTAL PLATE MALFORMATION

Illustration by Aletta Frazier

slide7.jpg

1. Embriology

- The calibre of bile ducts affected determines the type of ductal plate malformation
  - Small intrahepatic bile ducts
    - Von Meyenburg Complexes a
    - Policystic Liver Disease b
    - Congenital Hepatic Fibrosis c
  - Large intrahepatic bile ducts
    - Caroli Disease c
  - Both
    - Caroli Syndrome c

a - failure of involution of embryonic ducts
b - non-communicating persistence of embryonic ducts
c - communicating persistence of embryonic ducts

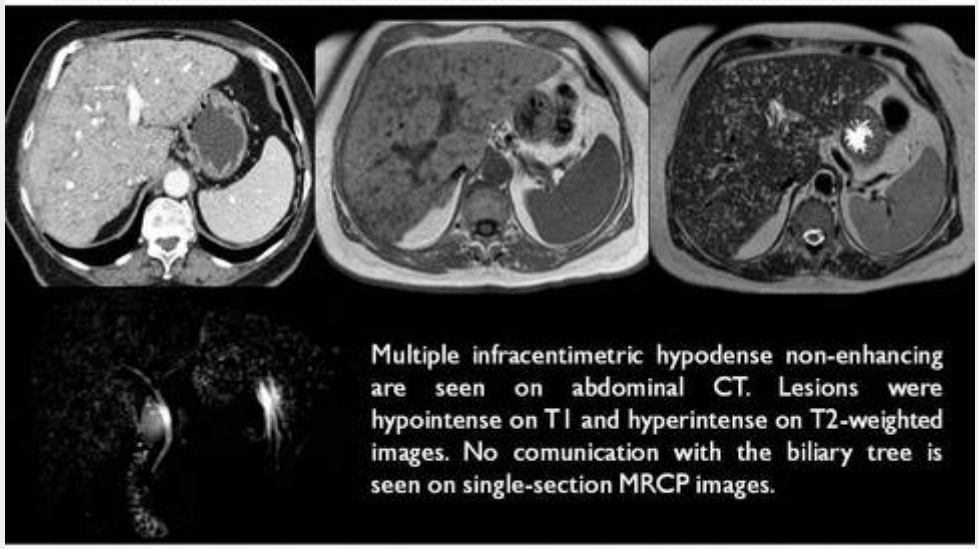
slide8.jpg

1. Embriology

- Von Meyenburg complexes or biliary hamartomas have a reported incidence of 0.69 to 2,8 % in autopsy series.
- Lesions are usually multiple, small (usually between 0,1 and 0,5 mm) and may be solid, cystic or mixed. When solid components are present (uncommon), sustained enhancement may be seen.

slide9.jpg

Von Meyenburg complexes



Multiple infracentimetric hypodense non-enhancing are seen on abdominal CT. Lesions were hypointense on T1 and hyperintense on T2-weighted images. No comunication with the biliary tree is seen on single-section MRCP images.

slide10.jpg

1. Embriology

- Polycystic liver disease may be associated with autossomic dominant polycystic kidney disease but also occurs as an isolated finding.
- The cysts arise from Von Meyenburg complexes lined with functional biliary epithelium. Both lesions may coexist.

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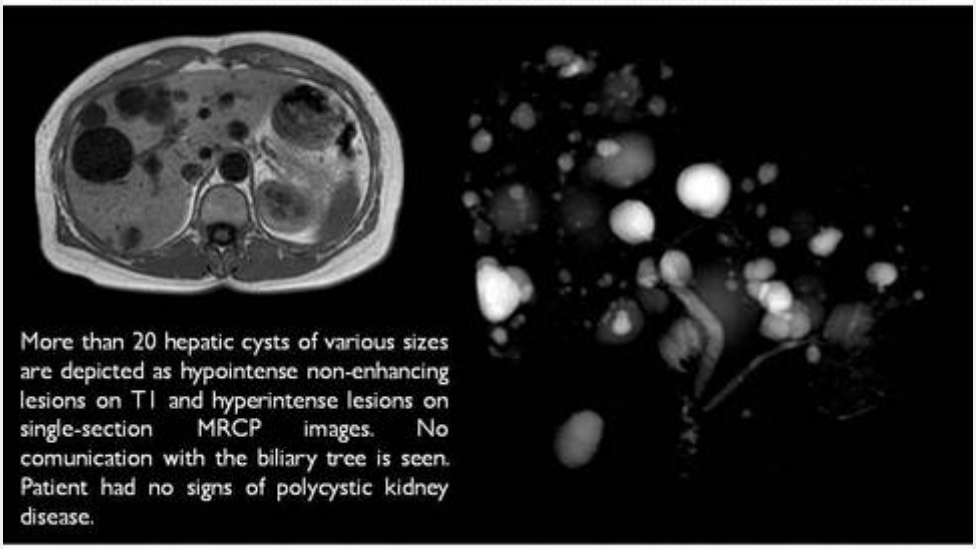
Polycystic liver disease



More than 20 hepatic cysts of various sizes are depicted as hypointense non-enhancing lesions on T1 and hyperintense lesions on single-section MRCP images. No comunication with the biliary tree is seen. Patient had no signs of polycystic kidney disease.

slide12.jpg

1. Embriology

- Caroli disease is a rare, autossomic recessive disease which is associated with kidney disease in up tp 60% of cases, namely medullary sponge kidney, ARPKD and nephronoptisis.
- It may be diffuse, lobar or segmental.
- The **central dot sign** is typical, due to cystic enveloping of portal or arterial branches.

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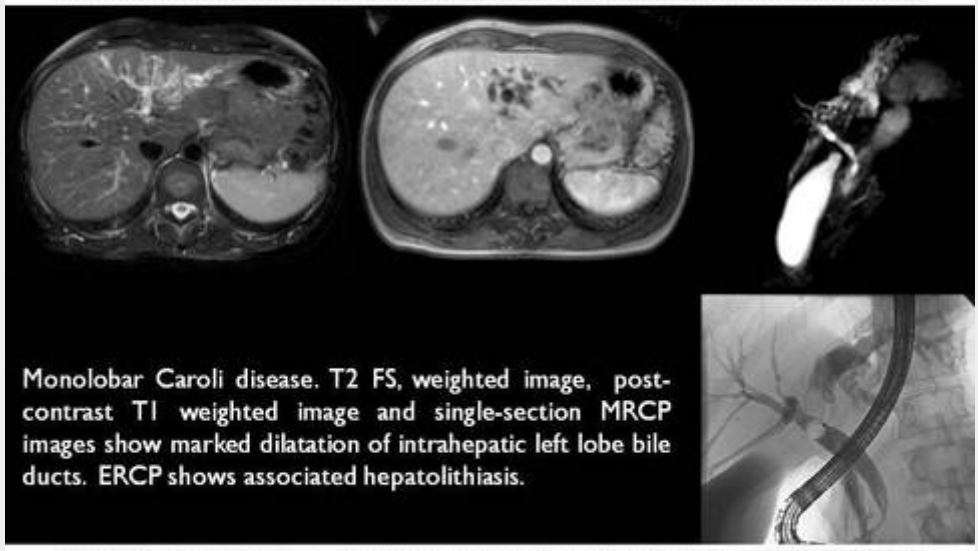
Caroli disease



Monolobar Caroli disease. T2 FS, weighted image, post-contrast T1 weighted image and single-section MRCP images show marked dilatation of intrahepatic left lobe bile ducts. ERCP shows associated hepatolithiasis.

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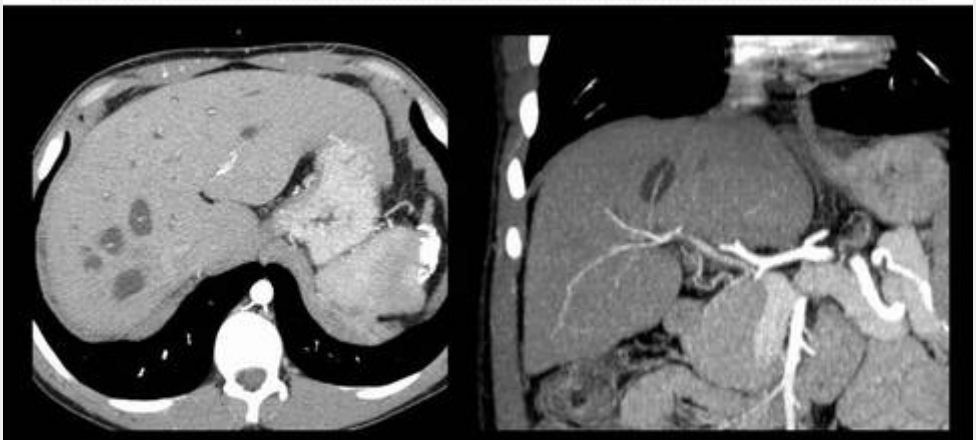
Caroli disease



Central dot sign here seen on post-constrast, portal phase, axial and coronal MIP CT images as multiple cystic lesions surrounding portal vein/arterial branches.

slide15.jpg

1. Embriology

- Caroli syndrome is said to be present when Caroli disease coexists with congenital hepatic fibrosis.
- Besides the complications of Caroli disease (cholangitis, hepatolithiasis, strictures and cholangiocarcinoma), portal hypertension and secondary biliary chirrosis are common late-stage findings.

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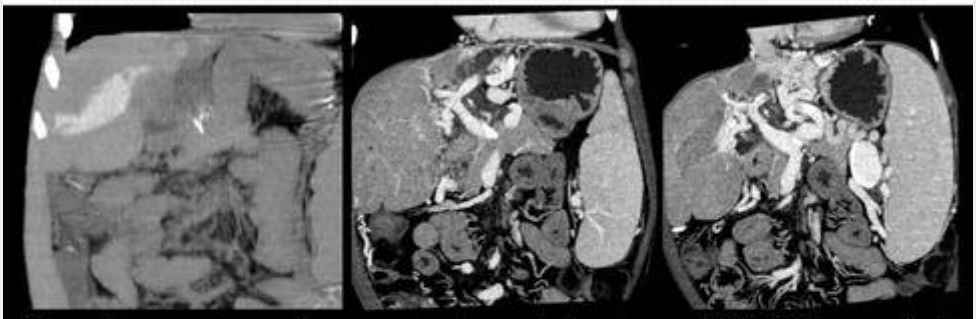
Caroli syndrome



Coronal, pre-contrast and post-contrast, portal phase, coronal MIP CT images depict marked dilatation of intrahepatic bile ducts on both lobes. One of the dilated right-lobe bile duct is completely filled with milimetric calculus. There are associated findings of cirrhosis and portal hypertention, namely liver dysmorphia, splenomegaly and tortuous perigastric and perisplenic colateral veins. Signs of chronic portal vein thrombosis are also seen (perypheral, partially calcified thrombus).

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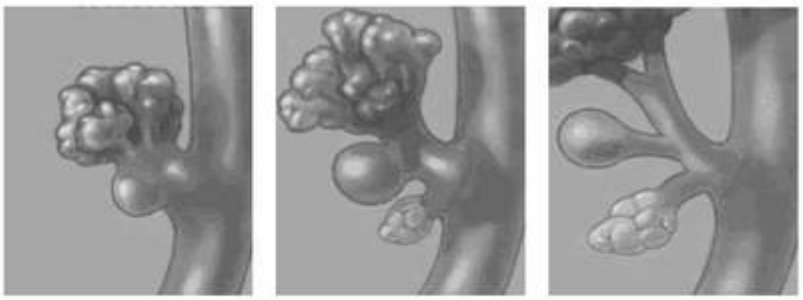
1. Embriology

- The ventral part of the hepatic diverticulum (*pars cystica*) develops into the gallbladder, cystic duct and choledocus.
- The common hepatic duct derives from the *pars hepatica*.



Adapted from www.vesalius.com

slide18.jpg

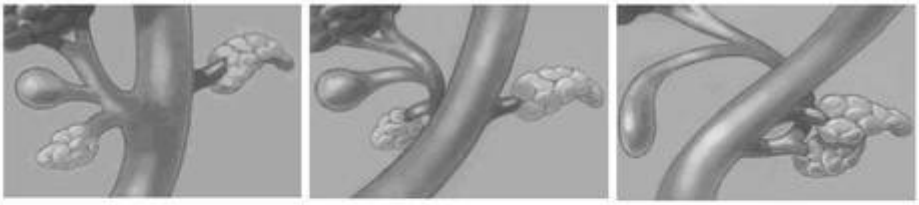
1. Embriology

- Intense remodeling of several channels present at the porta hepatis during the 5th week of embryonic life precedes the development of the left and right main hepatic ducts.
- Their proximal part derives from the first intrahepatic ductal plate and their distal part from the extrahepatic bile ducts.
- Alterations in this remodeling may be responsible for the several anatomic variants of the left-right hepatic duct junction.

slide19.jpg

1. Embriology

- By the end of the 6th week, the choledocus and ventral bud of the pancreas, so far draining distally into the choledocus, rotate posteriorly and come to lie it their definitive position.



Adapted from www.vesalius.com

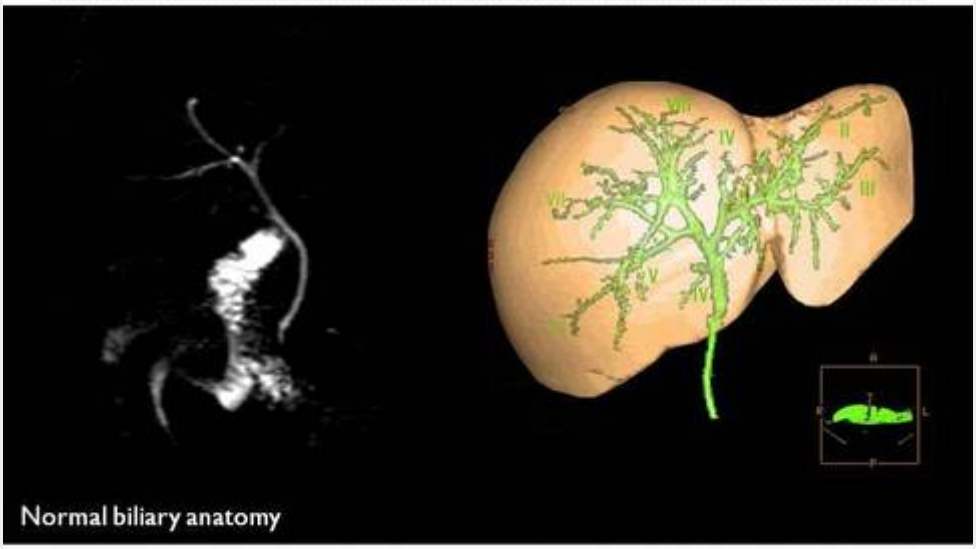
slide20.jpg

2. Normal Anatomy and Anatomic Variants

- The classic biliary anatomy is present in 58% of the population.
  - The **right main hepatic duct** results from the confluence of a horizontally-oriented **right posterior duct** (from segments VI and VII) and a more vertically-oriented **right anterior duct** (from segments V and VIII)
  - **Segment I** may drain to the left or right main ducts
  - The **cystic duct** drains into the lateral aspect of the common hepatic duct, midway between the hilum and the ampulla of Vater.

slide21.jpg

Normal biliary anatomy



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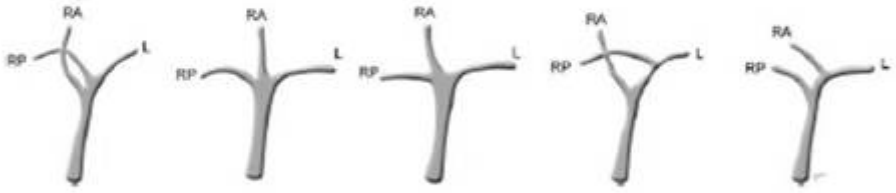
2. Normal Anatomy and Anatomic Variants

- There are several different anatomic variants of the hepatic duct confluence, the most important of which being depicted below.



RA right anterior hepatic duct
RP right porterior hepatic duct
L left hepatic duct

Radiographics March-April 2008 28:359-378

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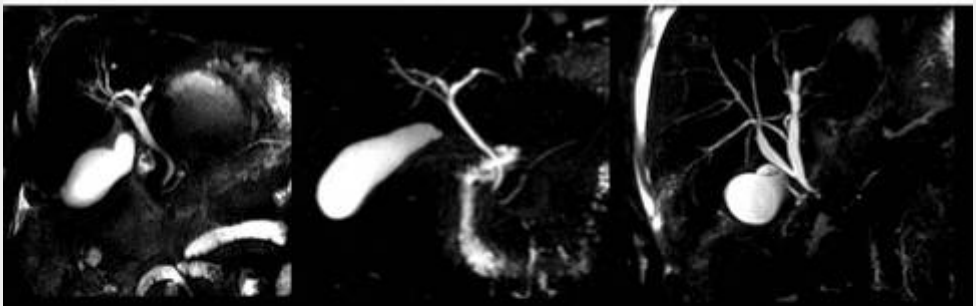
2. Normal Anatomy and Anatomic Variants

Drainage of the right posterior duct into the left main duct is the most common anatomic variant, present in 13 to 19% of the population.



Example on the right also shows accessory bile ducts draining to the left main bile duct and low confluence of left and right main ducts.

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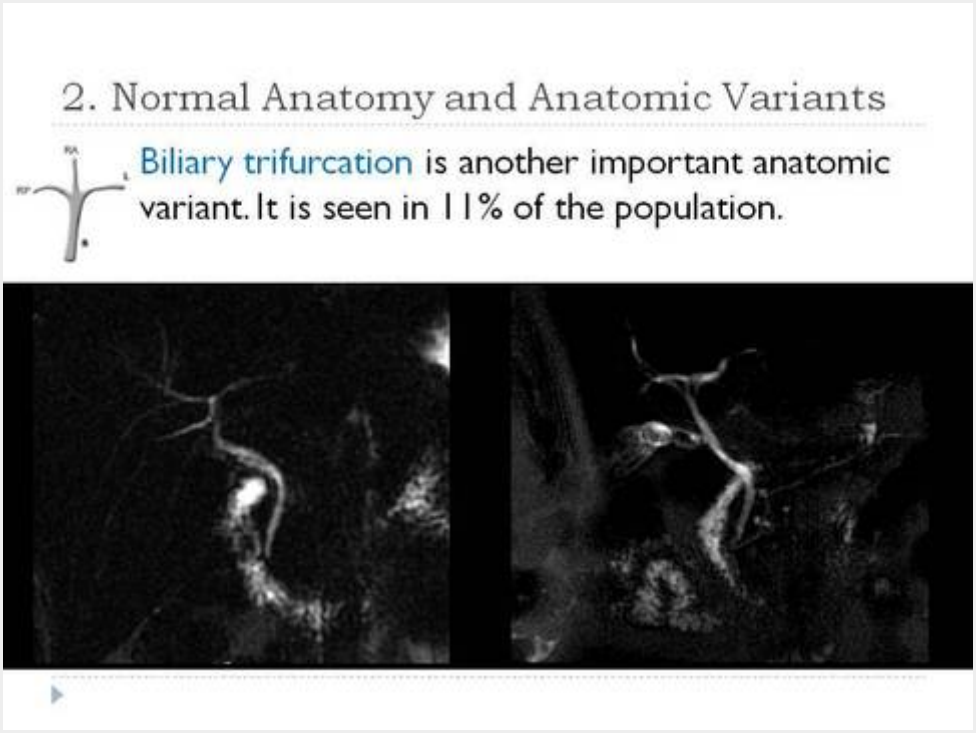

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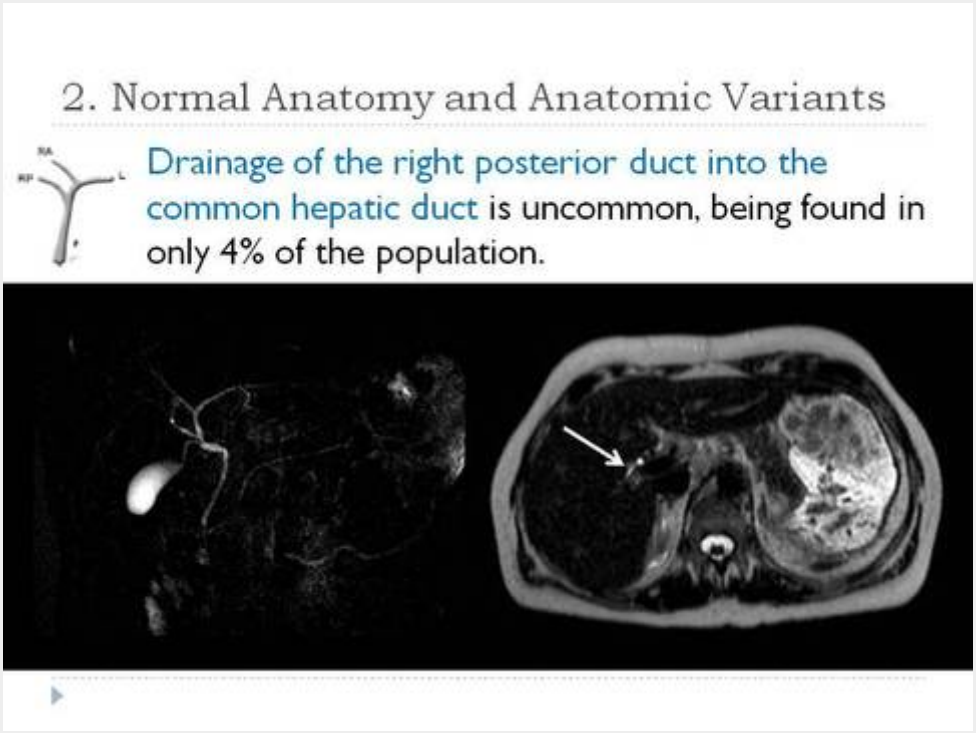


slide26.jpg

2. Normal Anatomy and Anatomic Variants

- Cystic duct anatomic variants are found in 50,1 % of the population. The most common variants are depicted below.

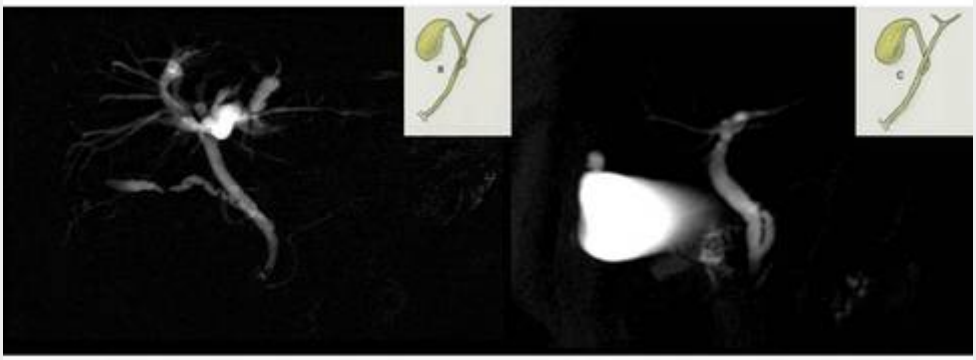
Right lateral insertion (*A*), anterior spiral insertion (*B*), posterior spiral insertion (*C*), low lateral insertion with a common sheath (*D*), proximal insertion (*E*), or low medial insertion (*F*).

January 2001 RadioGraphics, 21, 3-22

slide27.jpg

2. Normal Anatomy and Anatomic Variants

- Spiral insertion of the cystic duct is present in up to to 17% of the population.



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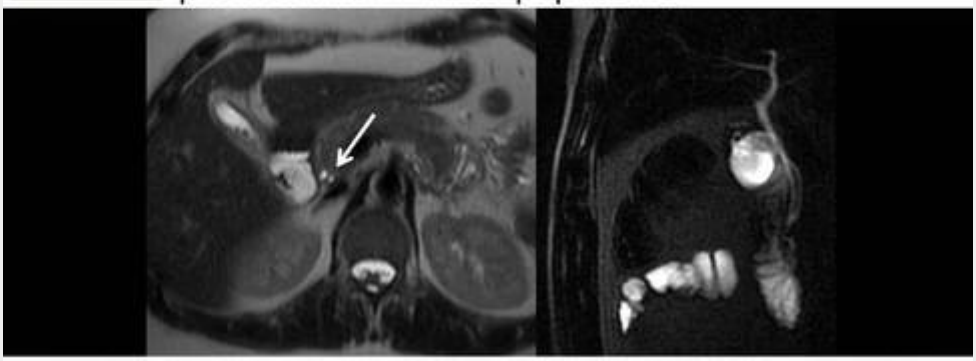
2. Normal Anatomy and Anatomic Variants

Cystic duct – common hepatic duct parallel course is said to be present when they run parallel and share a fibrous sheath along 2 cm or more. It is present in 10% of the population.



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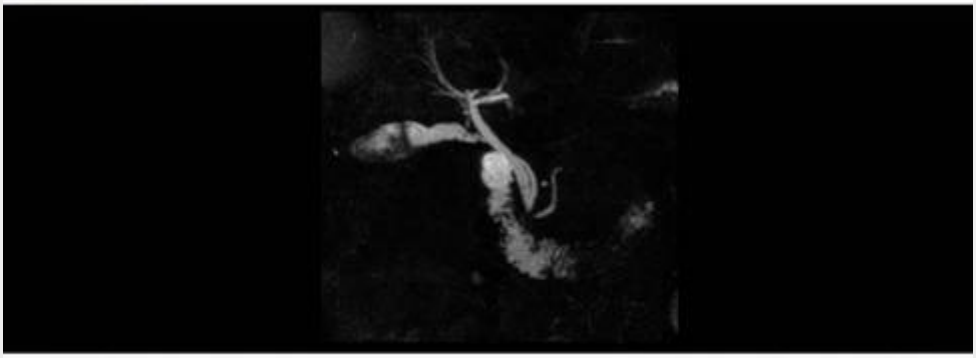
2. Normal Anatomy and Anatomic Variants

Low medial insertion of the cystic duct is present in 9% of the population.



slide30.jpg

2. Normal Anatomy and Anatomic Variants

- Aberrant ducts are the only draining ducts of a particular hepatic segment.
- Aberrant ducts draining part of the right hepatic lobe may
  - empty directly into the cystic duct (cystohepatic ducts) in 1 to 2 % of individuals.
  - Empty into the right or common hepatic duct (bile duct of Luschka) in one third of the population.

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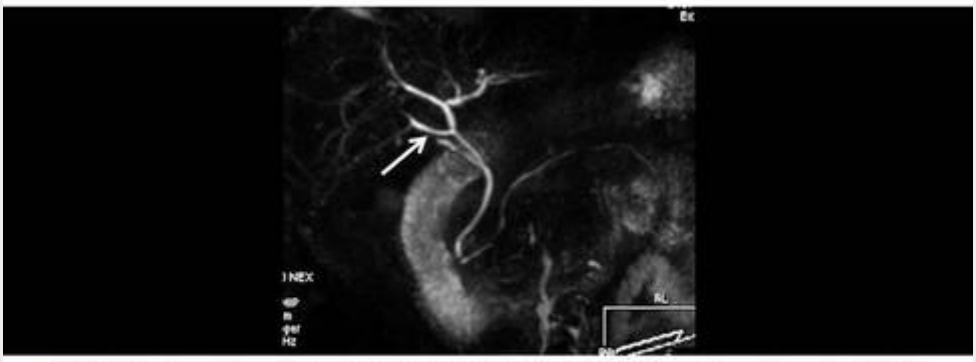
2. Normal Anatomy and Anatomic Variants

- An aberrant right hepatic duct (Luschka) drains part of the right lobe directly to the main hepatic duct.



slide32.jpg

2. Normal Anatomy and Anatomic Variants

- Accessory ducts are additional ducts draining a particular hepatic segment. They are said to be present in aproximately 2% of the population.
- They may also join the common hepatic duct at its junction with the cystic or drain directly into the cystic duct.

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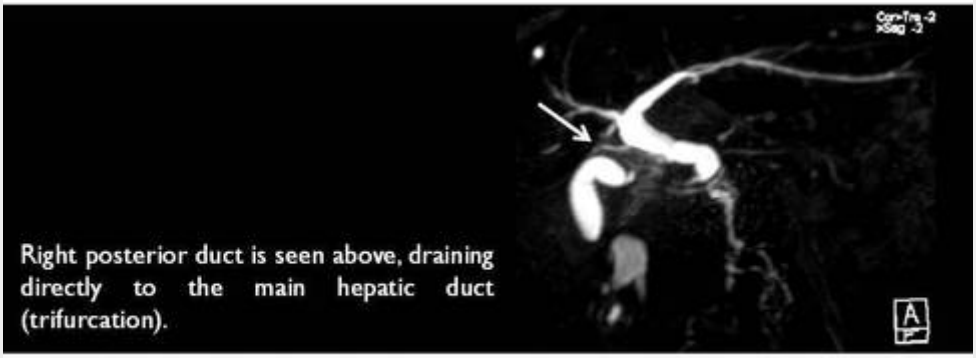
2. Normal Anatomy and Anatomic Variants

- An accessory right hepatic duct drains part of the right lobe directly to the main hepatic duct (→).



slide34.jpg

2. Normal Anatomy and Anatomic Variants

- The ampulla of Vater is another highly variable structure. The classic Y configuration of the union between the common bile duct and the pancreatic duct is observed in only 60% of the population.

Conventional Y configuration 60%

Double opening at the apex of the papilla 38%

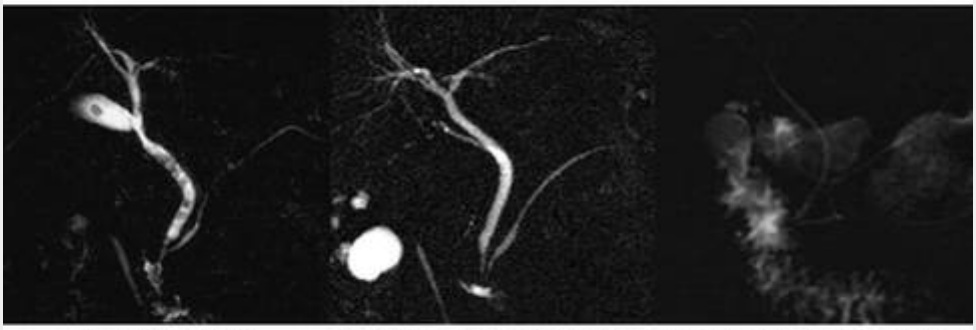
Separate duodenal openings for the CBD and Wirsung 2%

Long common channel with union proximal to the duodenal wall rare

RadioGraphics, 22, 1335-1352

slide35.jpg

Anatomic Variants of the Ampulla of Vater



Conventional Y configuration 60%

Double opening at the apex of the papilla 38%

Separate duodenal openings for the CBD and Wirsung 2%

slide36.jpg

3. Iatrogenic Complications

- Variants of normal biliary anatomy may determine bile duct injury during hepatic surgery, such as tumor resection or partial hepatectomy for living donor transplantation. It may also occur during simpler procedures such as cholecystectomy, particularly laparoscopic cholecystectomy.
- Resultant iatrogenic lesions include inadvertent ductal ligation, bile leaks and strictures.

slide37.jpg

3. Iatrogenic Complications

- Bile leakage after hepatic or biliary surgery may occur in different locations, but mainly occurs at the distal branches of the hilar convergence. It may also occur at the repair site of the hepatic duct or along parenchymal transection surface of the liver.
- MR Cholangiography after IV mangafodipir trisodium administration is useful for bile leak detection and quantification.

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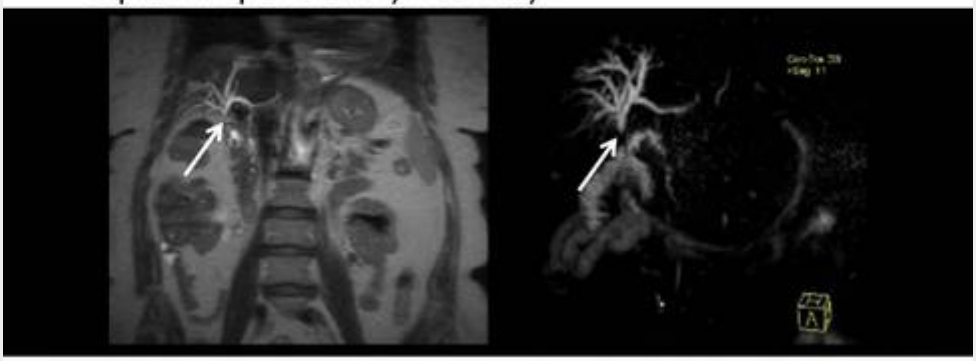
3. Iatrogenic Complications

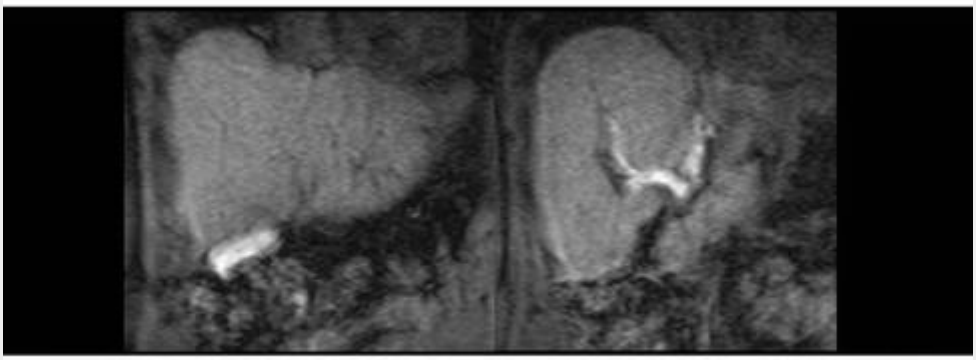
- Intrahepatic bile duct dilation due to inadvertent ligation of a right posterior bile duct with anomalous drainage to the left main hepatic duct during laparoscopic cholecystectomy.



slide39.jpg

3. Iatrogenic Complications

- A subhepatic with enhancement after IV Mangafodipir administration is seen, representing a biloma due to main hepatic duct leakage (→).

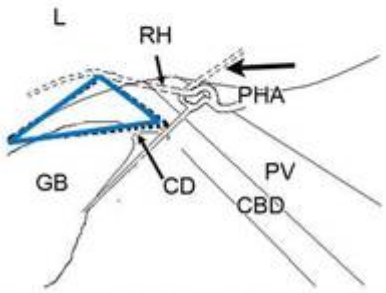


slide40.jpg

3. Iatrogenic Complications

- Particularly predisposing variants
  - Aberrant and accessory ducts which course in the triangle of Calot(-) and ducts that drain directly into the gallblader or cystic duct, are particularly susceptible to inadvertent ligation or transection during cholecystectomy.

CBD = Common bile duct
CD= Cystic duct
GB = Gallbladder
L = Liver
RH = Right hepatic artery
PHA = proper hepatic artery



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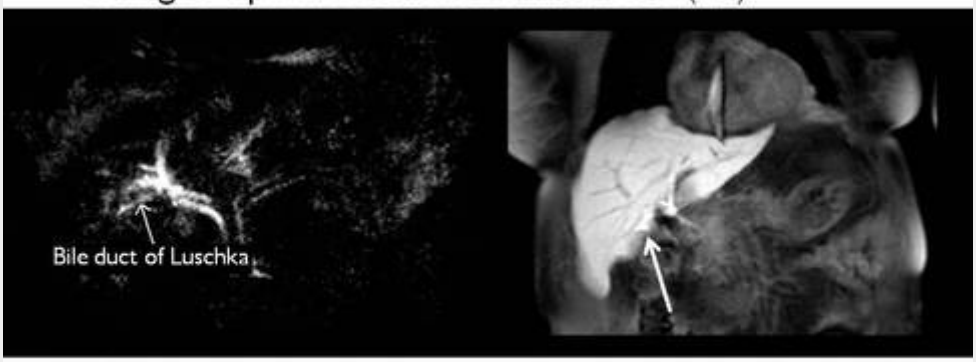
slide41.jpg

3. Iatrogenic Complications

- An accessory right hepatic duct (Luschka) was inadvertantly sectioned during laparoscopic cholecystectomy. Bile leak is depicted after IV mangafodipir trisodium administration (→).



slide42.jpg

3. Iatrogenic Complications

- Particularly predisposing variants
  - Cystic duct – common hepatic duct parallel course may result in surgical ligation too close to the common hepatic duct, with stricturing or common bile duct transection.
  - An abnormaly long remnant after cholecystectomy in patients with this variant is common and may also result in **postcholecystectomy syndrome** due to inflamation and calculi formation.

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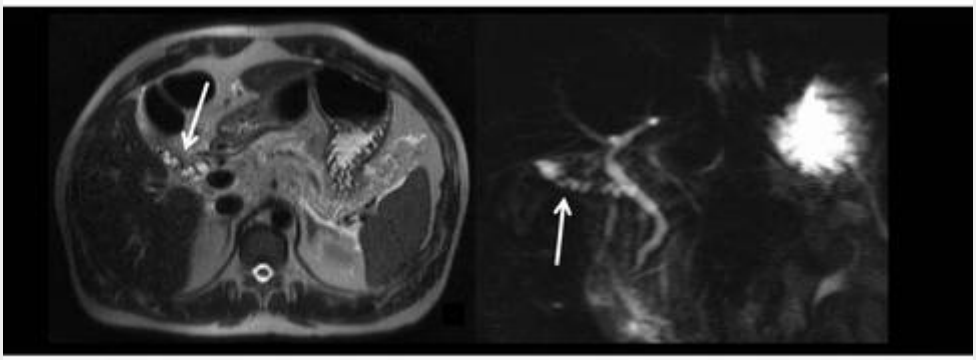
3. Iatrogenic Complications

- An unusually long and tortuous cystic duct remnant, within which are several gallstones, is seen in a symptomatic patient (→).



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3. Iatrogenic Complications

- Particularly predisposing variants
  - Low medial insertion of the cystic is particularly relevant because distal gallstone impaction may mimic choledocal obstruction and ERCP may result in inadvertent instrumentation of the cystic rather than the common bile duct.

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3. Iatrogenic Complications

- In the particular case of tumor resection
  - With left lobe resection
    - Right posterior duct draining directly into the left hepatic duct
    - Biliary trifurcation

    are particularly relevant
  - With right lobe resection
    - Left hepatic duct draining directly into the right hepatic duct
    - Biliary trifurcation

    are particularly relevant

slide46.jpg

3. Iatrogenic Complications

- In the particular case of living donor transplantation
  - Variants considered relevant in donors are
    - Right posterior duct draining directly into the left hepatic duct
    - Biliary trifurcation
    - Right posterior duct draining directly into the left hepatic duct
    - Accessory hepatic ducts
  - Variants considered relevant in recipients are
    - Left hepatic duct into the right anterior hepatic duct
    - Biliary trifurcation
    - Cystic duct draining into the right hepatic duct
    - Accessory hepatic ducts

slide47.jpg

4. Congenital Anomalies

- Biliary atresia is defined as lack of lumen in part of or all of the extrahepatic bile duct. It is the most common cause of obstructive jaundice in the first month of life.
- It is an acquired disease, occuring in 1/10000 live births (girls more frequently affected), and is associated with polysplenia in 11% of cases.

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4. Congenital Anomalies

- Biliary atresia of the extrahepatic bile ducts was classified by Kasai into three categories:
  - Type I: atresia of the common bile duct while bile duct segments adjacent to the liver are intact.
  - Type IIA: atresia of the common hepatic duct with or without atresia of the common bile duct.
  - Type IIB: atresia of all main branches of the extrahepatic system or lack of extrahepatic bile duct system.
  - Type III: atresia of the hepatic and cystic ducts with hilar ducts replaced by a fibrous core.
- Types I and II represent 10% of cases and are surgically correctable.
- Type III represents 90% of cases and is not correctable.

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4. Congenital Anomalies

- Choledocal cysts are uncommon anomalies of the biliary system manifested by cystic dilatation of the extra and/or intrahepatic biliary tree.
- Incidence is higher in East Asia (1/1000) than in western countries (1/100000).
- Presentation occurs during childhood in 60% of cases and females are affected in 80% of cases.
- Choledocal cysts predispose to cholangitis, lithiasis, obstruction, pancreatitis and malignancy.
- There are five subtypes of choledochal cysts, as defined by Todani´s modification of the Alonso – Lej classification

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4. Congenital Anomalies

- Todani´s modification of the Alonso – Lej classification

Type I	Solitary, extrahepatic cyst
Type II	Extrahepatic duodenal diverticulum
Type III	Intraduodenal cyst
Type IV	Extrahepatic and intrahepatic cysts
Type V	Multiple intrahepatic cysts

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4. Congenital Anomalies

Type I

Type I A	**Type I B**	**Type I C**
Cystic dilation	Focal dilation	Fusiform dilation

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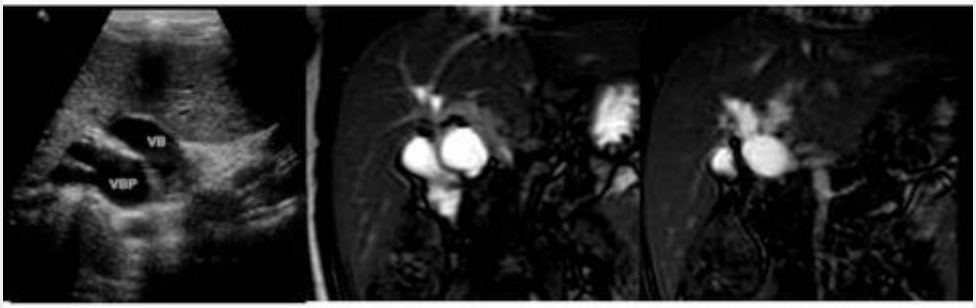
4. Congenital Anomalies

- **Type IC Choledochal Cyst** ♀, 6 Yo. Abdominal Ultrasound and MR SSFP sequences show a fusiform dilation of the common bile duct, continuous with the cystic duct. The right and left main hepatic ducts were also dilated.



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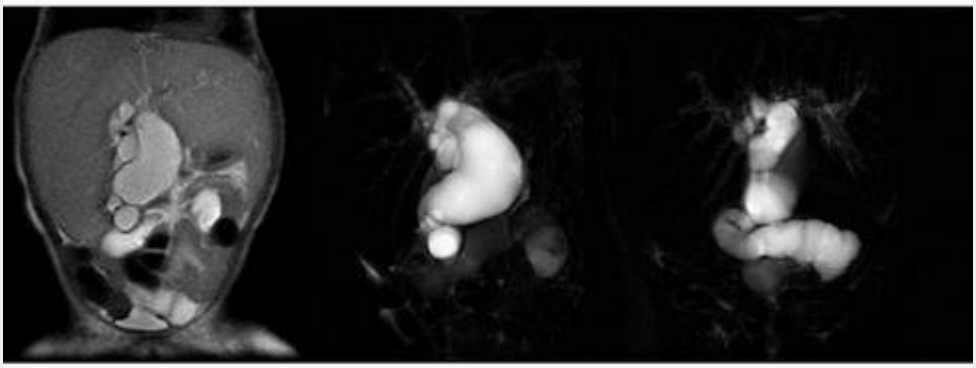
3. Congenital Anomalies

- **Type IC Choledochal Cyst** ♀, 4 Yo with jaundice and abdominal pain. MR images shows fusiform dilation of the common bile duct.

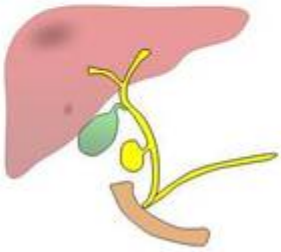


slide54.jpg

4. Congenital Anomalies

Type II

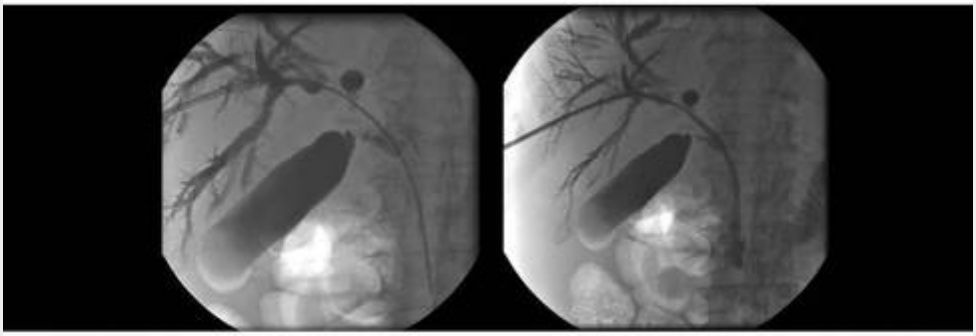
Diverticulum



slide55.jpg

4. Congenital Anomalies

- **Type II Choledochal Cyst - Diverticulum** ♂, 76 Yo. Cholangiocarcinoma – Klatskin tumor. Percutaneous transhepatic cholangiography (PTC) performed to paliate biliary obstruction showed a diverticulum of the common hepatic duct, located above the insertion of the cystic duct, draining to an area of neoplastic involvement.

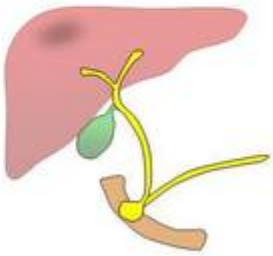


slide56.jpg

4. Congenital Anomalies

Type III

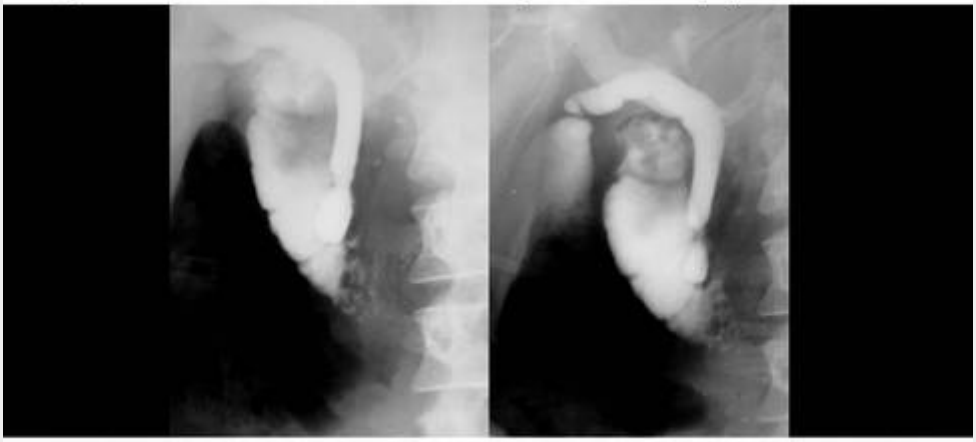
Choledochocele



slide57.jpg

4. Congenital Anomalies

- **Type III Choledochal Cyst – Choledochocele** PTC shows a diverticulum of the common bile duct located within the 2nd portion of the duodenum´s wall, adjacent to the papilla of Vater.



slide58.jpg

4. Congenital Anomalies

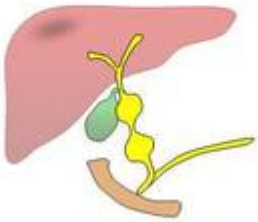
Type IV

Type IV A

Multiple intra and extrahepatic cysts

Type IV B

Multiple extrahepatic cysts



slide59.jpg

4. Congenital Anomalies

Type IVA choledochal cyst as seen on PTC. Massive dilation of the common bile duct and multiple intrahepatic bile duct dilations are apparent.

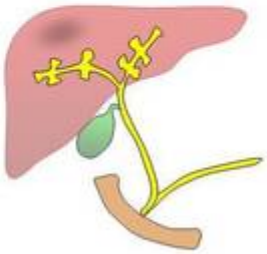
slide60.jpg

4. Congenital Anomalies

Type V

Caroli´s Disease*

Multiple intrahepatic cysts



*Already discussed – see section 1. Embryology

slide61.jpg

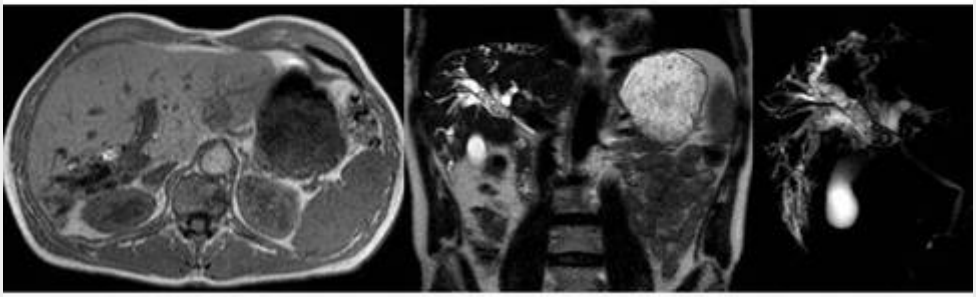
5. Pathology

- **Choledocholithiasis** is found in 10 to 15% of patients with cholelithiasis, and it´s incidence increases with increasing age.
- Undetected duct stones are left behind in 1 to 5% of patients after cholecystectomy.
- The majority of cases of choledocholithiasis are secondary (migrating cholesterol or mixed calculus from the gallbladder).
- Primary choledocholithiasis is uncommon and results from chronic hemolysis, hepatobiliary parasitism, chronic recurrent cholangitis, stenosis and strictures, congenital anomalies such as Caroli disease (*example above – Embriology section*).
- Common complications are biliary obstruction, cholangitis, pancreatitis and secondary biliary cirrhosis.

slide62.jpg

5. Pathology

- **Primary cholelithiasis** Patient with a history of trauma presents with obstructive ictericia. MR Colangiography shows right posterior segments bile duct ectasia due to a longstanding stenosis of the corresponding bile duct. Multiple hyperintense on T1-weighted and hypointense on T2-weighted images intraductal filling defects as depicted and result from primary bile-pigment-rich cholelithiasis.



slide63.jpg

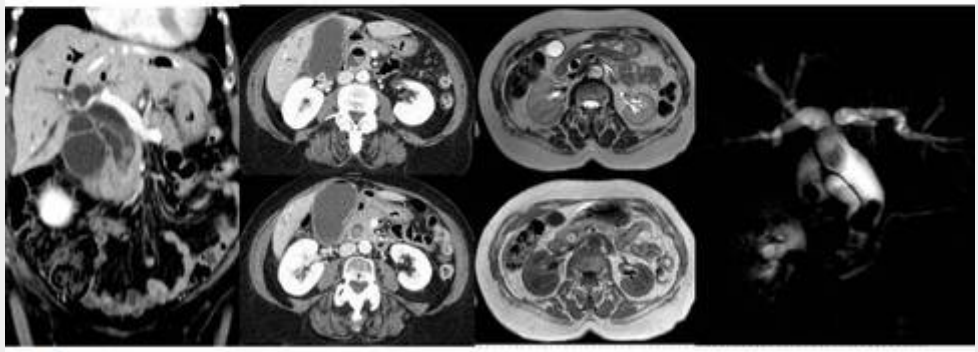
5. Pathology

- **Secondary cholelithiasis** Axial CT images after IV contrast administration show cholelithiasis and main hepatic duct and cystic duct dilation (notice main hepatic duct and cystic duct parallel course). Distally, a round filling defect is seen in the main bile duct, corresponding to a calculus(→).

slide64.jpg

5. Pathology

- **Secondary cholelithiasis** CT images after IV contrast shows intra and extrahepatic bile duct dilatation and two foci of choledocolithiasis. Signs of cholecystitis are also apparent. Axial MR images show the bile duct stone as a hypointense round image surrounded by hyperintense bile on T2 weighted and T2 SSH-TSE images. The stone is hyperintense on T1-weighted images.



slide65.jpg

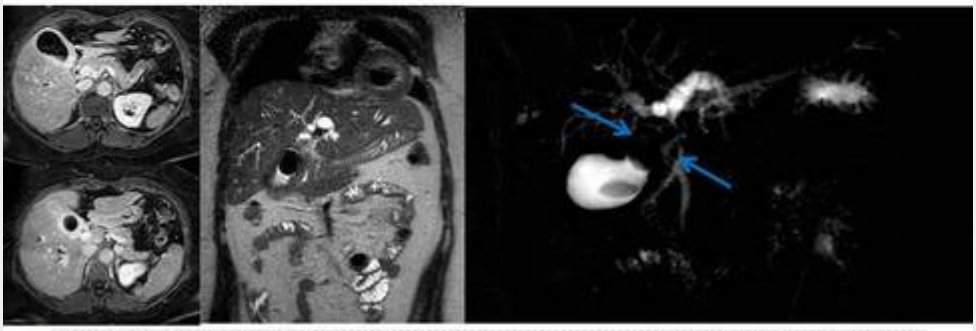
5. Pathology

- Mirizzi syndrome results from impaction of a calculus within the cystic duct and subsequent extrinsic compression of the common hepatic duct. Some authors also relate the obstruction to local extension of the inflammatory process (cholecystitis) to involve the common hepatic or common bile duct.
- Low medial insertion of the cystic duct and a parallel cystic-hepatic duct course are predisposing factors.

slide66.jpg

5. Pathology

- **Mirizzi Syndrome** T1 FS post-gadolinium images show thickening of the gallbladder's wall due to cholecystitis. Coronal T2-weighted images show a gallstone inpacted in the infundibulum – cystic duct. T2 SSH-TSE images show a luminal defect in the infundibular-cystic duct region, compressing the main hepatic duct and thus causing intrahepatic bile duct dilatation.



slide67.jpg

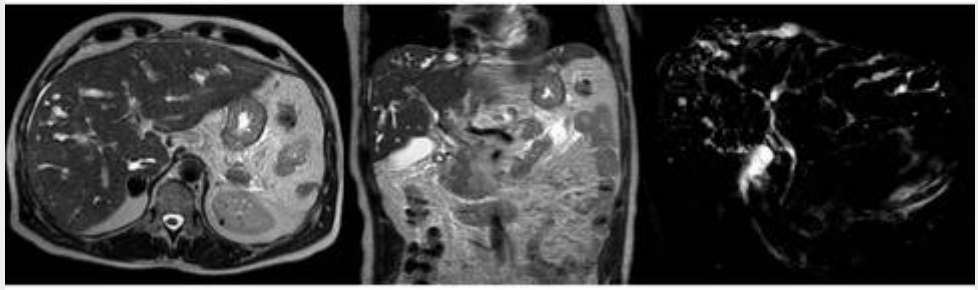
5. Pathology

- Sclerosing cholangitis results from chronic progressive inflammation and fibrosis of the intra and/or extrahepatic bile ducts.
- It may be primary (etiology not known), in which a strong association with inflammatory bowel disease (70%), particularly ulcerative colitis, is seen. A weaker association with multifocal fibrosclerosis syndromes such as mediastinal and retroperitoneal fibrosis has also been reported.
- Secondary sclerosing cholangitis may occur as a long-term complication of choledocholithiasis, cholangiocarcinoma, operative or traumatic biliary injury or contiguous inflammatory processes.
- Findings in all similar to sclerosing cholangitis are found in AIDS cholangiopathy, *Cryptosporidium*, MAI, CMV, *Microsporidia* and *Isospora* being associated infectious organisms.

slide68.jpg

5. Pathology

- **Primary sclerosing cholangitis** in patient with Chron disease. Multiple irregular intrahepatic and extrahepatic bile duct dilatations are seen on T2 FSE and e T2 SSH-TSE MR images.



slide69.jpg

5. Pathology

- The incidence of cholangiocarcinoma is increasing worldwide.
- Liver fluxes and hepatolithiasis are common risk factors in Asia, while primary sclerosing cholangitis, cirrhosis, alcohol and diabetes are common risk factors in western countries.
- Extrahepatic cholangiocarcinomas derive from the main bile duct (*hilar or Klatskin tumors are included in the intrahepatic subgroup*) They occur:
  - in the upper third in 50 to 75%,
  - in the middle third in 10 to 25%
  - in the lower third in 10 to 20% of cases.

slide70.jpg

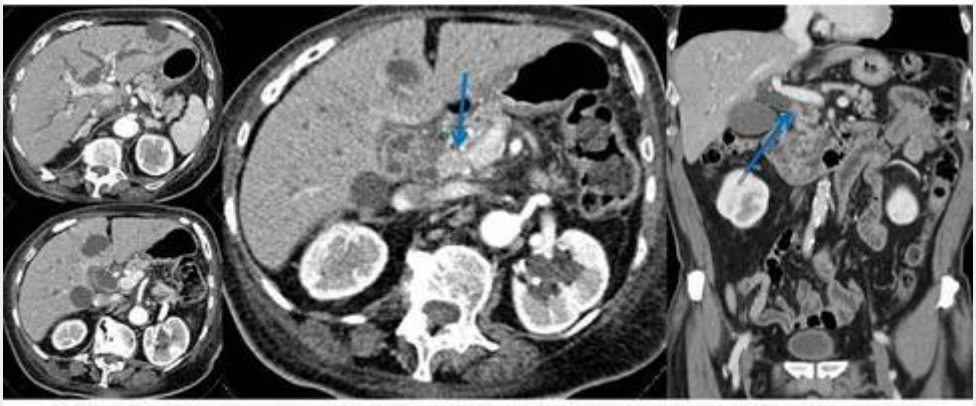
5. Pathology

- Extrahepatic cholangiocarcinoma may be classified morphologically as:
  - Mass-forming or nodular. A round, small mass is seen ocluding the main bile duct.
  - Periductal-infiltrating or sclerosing. Segmental or diffuse, desmoplastic thickening of the extrahepatic bile ducts without a discrete mass.
  - Intraductal-growing or papillary. One or multiple intraluminal friable polypoid lesions (cholangiocarcinomatosis),
- Periductal-infiltrating is by far the most common morphologic type observed.

slide71.jpg

5. Pathology

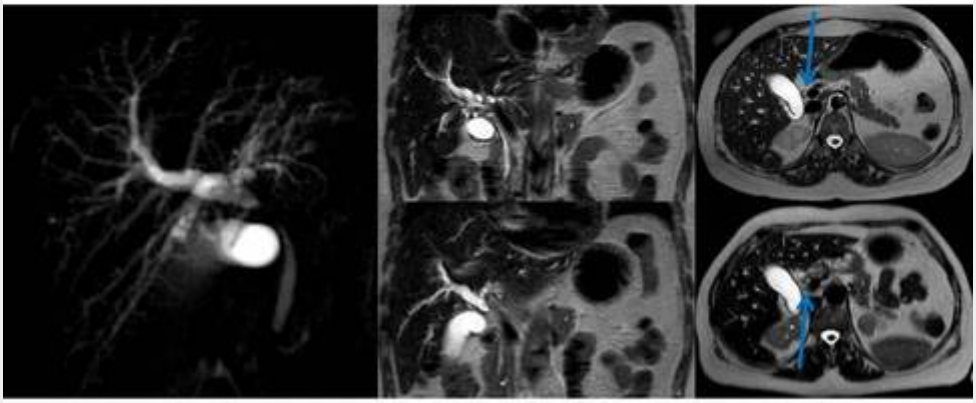
- Middle-third **cholangiocarcinoma**, sclerosing type. Post-contrast CT images show circunferential thickening of the common bile duct, resulting in marked narrowing of the lumen and intrahepatic bile duct dilatation.



slide72.jpg

5. Pathology

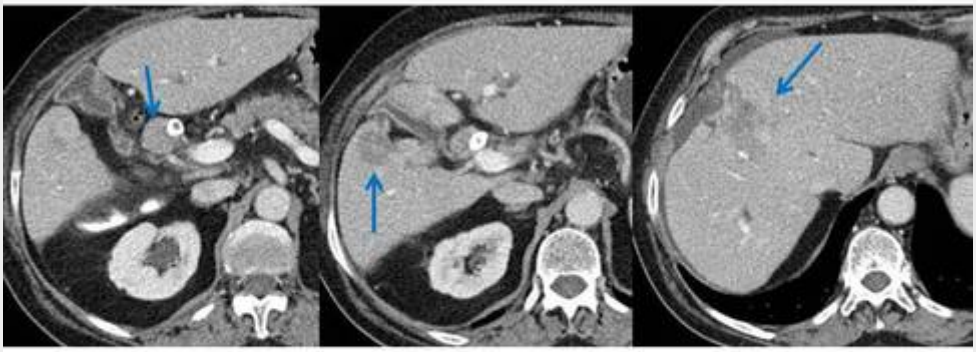
- Middle-third **cholangiocarcinoma**, sclerosing type. A similar example, here seen on T2-weighted images and SSFP MR images.



slide73.jpg

5. Pathology

- Multifocal, intrahepatic perypheral and extrahepatic, upper third, mass-forming type **cholangiocarcinoma** seen on post-contrast CT images A biliary prothesis had already been placed.



slide74.jpg

5. Pathology

- The periampullary carcinomas are those that arise within 2 cm of the major duodenal papilla and comprise four different malignancies:
  - ampullomas
  - distal cholangiocarcinomas
  - carcinomas of the pancreas
  - carcinomas of the duodenum
- Although their clinical features and surgical aproaches are similar, accurate determination of tumor origin is essential to adequately predict prognosis.

slide75.jpg

5. Pathology

- Although ampullary carcinoma is described by some as a CCC arising in the last centimeter of the CBD, it shares more features with duodenal carcinoma than with CCC, namely molecular features and clinical outcomes.
- It usually presents as a small (0.5 to 2 cm) nodule or as an irregular filling defect at the distal margin of the pancreaticobiliary junction. Less commonly, irregular periductal thickening may be seen. Although the double duct sign is typically found, the Wirsung may not be ectatic, possibly because, in 40% of individuals, the CBD and Wirsung drain separately into the duodenum.

RadioGraphics, 22, 1335-1352

slide76.jpg

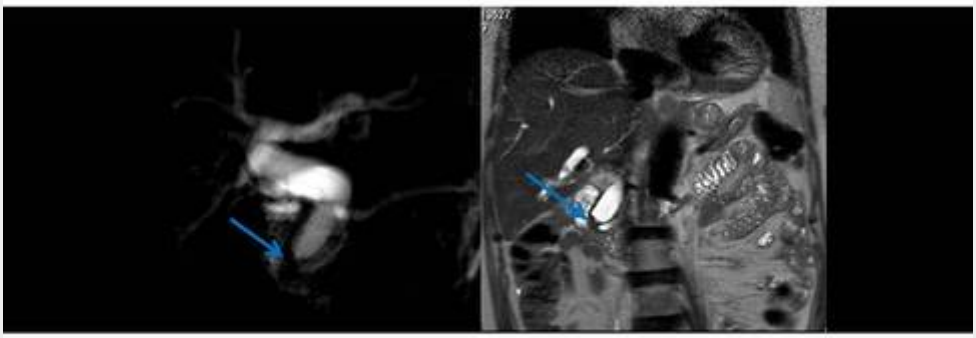
5. Pathology

- Marked abrupt duct dilatation in the absense of features of choledocolithiasis or pancreatitis should suggest the diagnosis of ampulloma.
- Differentiation of ampullary tumors from rare benign tumors in this location is virtually impossible with imaging.
- The morphologic type is usually polypoid and extraluminal extension, lymphatic spread and perineural invasion are infrequent. These features, along with early-onset of ictericia and thus early detection, contribute to it´s relatively good prognosis, with mean 5-year survival rates aproaching 40%.

slide77.jpg

5. Pathology

- Ampullary carcinoma Marked dilatation of the common bile duct with an abrupt cutoff at its intraparietal duodenal segment is seen on MR Colangiography T2 SSH- TSE images. Circunferential thickening of the ampulla is seen on coronal T2 TSE images.



slide78.jpg

5. Pathology

- Pancreatic carcinoma is the most frequent carcinoma in the periampullary region.
- It tends to be larger (1 – 2,5 cm) and more proximal (mean distance to the duodenal lumen of 25 mm) upon detection than ampulloma.
- Blunted, beak or rat tail terminations of the main bile and pancreatic ducts are typically seen in association with an uncinate process parenchymal mass.

slide79.jpg

5. Pathology

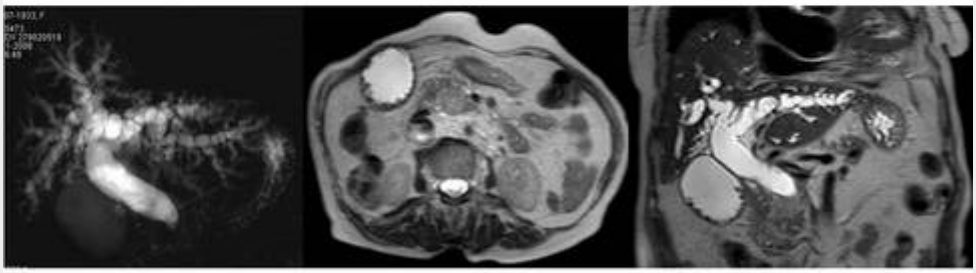
- The double duct sign is seen in 62% of patients with pancreatic carcinoma. Sometimes, the short distal duct segments may also be seen – four segment sign. This finding is very unusual in the other types of periampullary carcinomas. It may be seen in chronic pancreatitis but the narrowing tends to be long and incomplete.
- Dilatation of side branches of the Wirsung around the stenotic segment is not infrequent in pancreatic carcinomas and is also unusual with the other 3 types of periampulary carcinomas.

RadioGraphics, 22, 1335-1352

slide80.jpg

5. Pathology

- Carcinoma of the uncinate process of the pancreas Marked dilatation of bile ducts is seen on T2 SSH-TSE images, with an abrupt cutoff at the distal portion of the intrapancreatic segment of the common bile duct, due to a nodular mass, corresponding to a pancreatic adenocarcinoma. The mass is slightly hyperintense on T2 FSE images.



slide81.jpg

5. Pathology

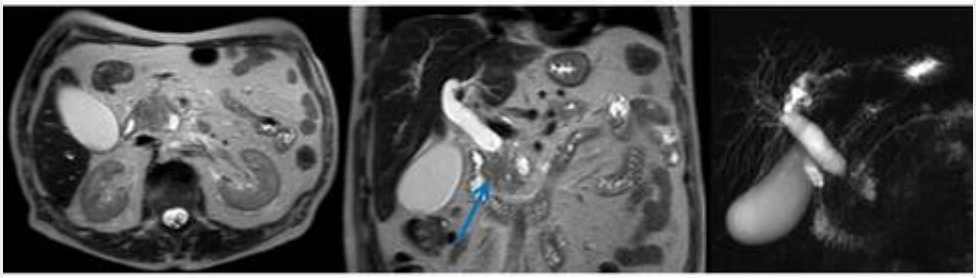
- Distal common bile duct carcinoma is of the periductal-infiltrating type in the large majority of cases. An intraductal-growing type may be found less frequently.
- Common bile duct dilatation is found in almost all patients. Not uncommonly, one can see both the distal and proximal CBD segment which, along with the pancreatic duct ,forms the three-segment sign.
- Extension to the pancreatic duct may cause dilatation and advanced stages may be impossible to differentiate from pancreatic carcinomas. In these cases, prognosis worsens accordingly.

RadioGraphics, 22, 1335-1352

slide82.jpg

5. Pathology

- Distal common bile duct carcinoma, sclerosing type. Marked dilatation of bile ducts is seen on T2 SSH-TSE images, with an abrupt cutoff at the distal portion of the intrapancreatic segment of the common bile duct, due to circunferential tumoral thickening. The mass is isointense to the pancreatic parenquima on T2 FSE images.



slide83.jpg

5. Pathology

- Duodenal carcinoma is the least common periampullary carcinoma.
- It may present either as a polypoid mass, a fungating mass or as a scirrhous carcinoma with eccentric wall thickening and duodenal luminal narrowing.
- Dilatation of the CBD and/or Wirsung, when present, tends to be inconspicuous. This finding may allow differentiation from the other types of periampullary carcinomas.
- Lymph node metastasis are frequent upon diagnosis but still, the mean 5-year survival rate is better than for all the other periampullary carcinomas, aproaching 50%.

RadioGraphics, 22, 1335-1352

slide84.jpg

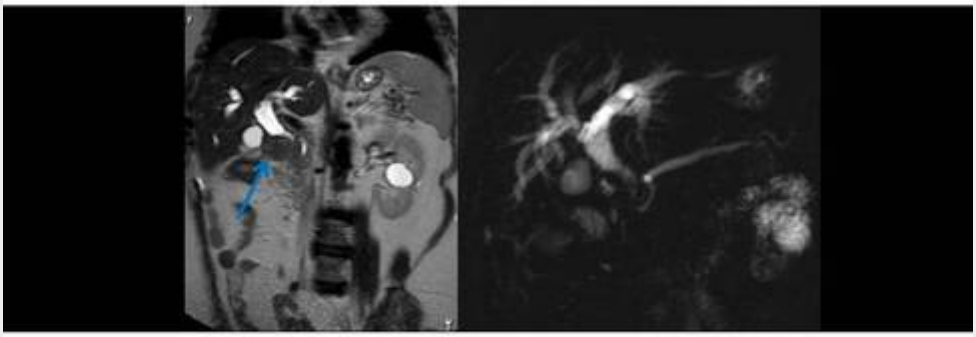
5. Pathology

- The extrahepatic bile ducts may also be secondarily invaded by non-periampullary tumors, such as stomach or gallbladder carcinoma.

slide85.jpg

5. Pathology

- A nodal metastasis from gastric carcinoma located in the pancreatic groove is seen on coronal T2 FSE and T2 SSH-TSE MR images, extrinsically compressing the common bile duct and causing proximal biliary dilatation.



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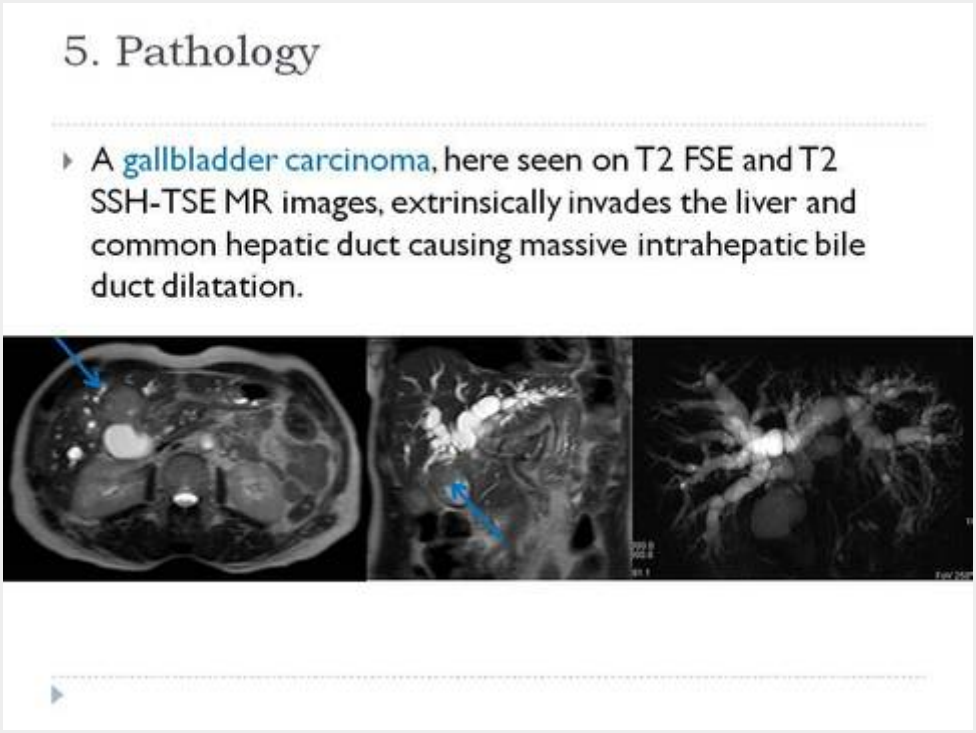
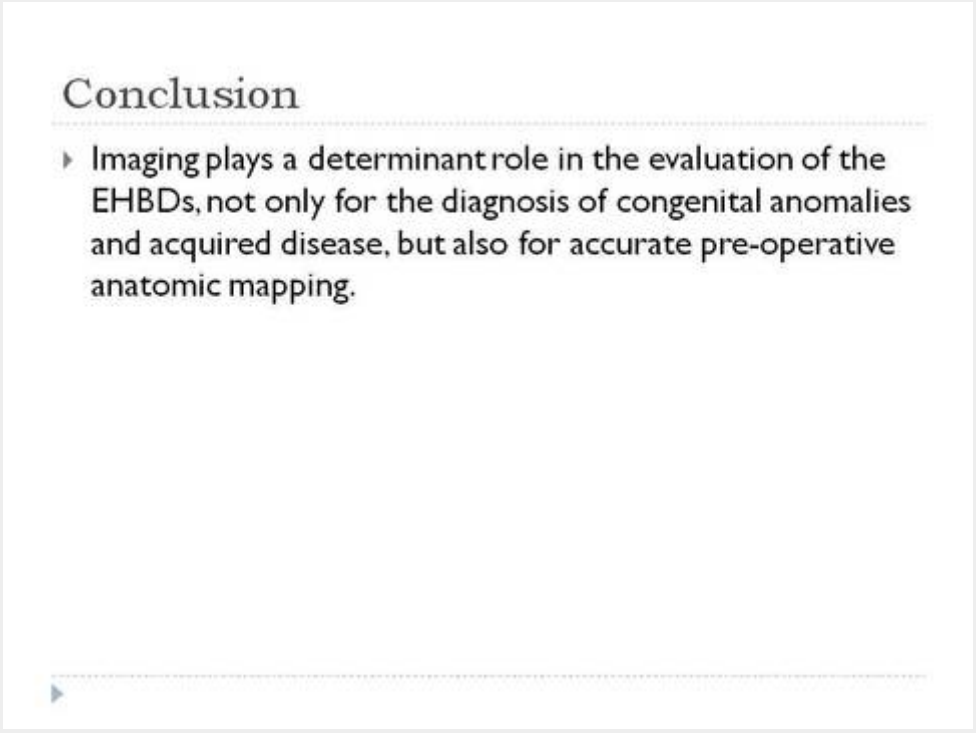


4. Conclusion

Conclusion

slide87.jpg



5. Author Information

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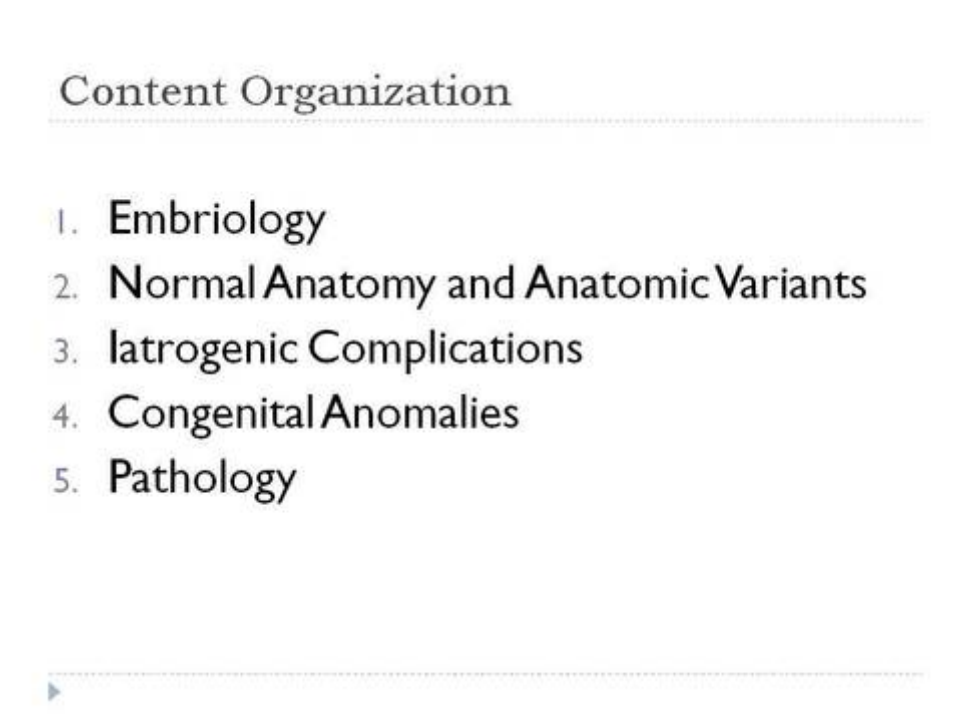
6. Mediafiles

slide1.jpg

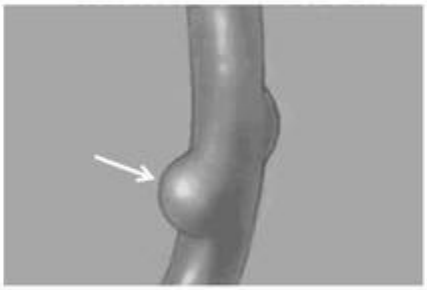
slide2.jpg



slide3.jpg

1. Embriology

- At the beginning of the 4th embrionic week, an endodermal outgrowth of the ventral distal foregut arises – the hepatic diverticulum (→).

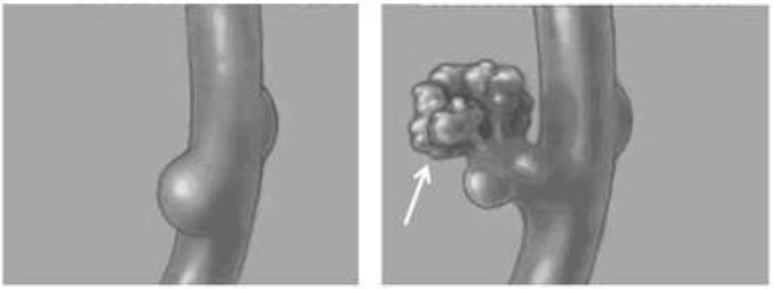


Adapted from www.vesalius.com

slide4.jpg

1. Embriology

- The cranial part of the hepatic diverticulum (*pars hepatica*) develops into the liver primordium (→).



Adapted from www.vesalius.com

slide5.jpg

1. Embriology

- The liver primordium originates liver cells and ductal plates (→), which develop in the mesenchyme adjacent to portal vein branches (→)
- The ductal plates later on remodel to form the intrahepatic bile ducts (→)

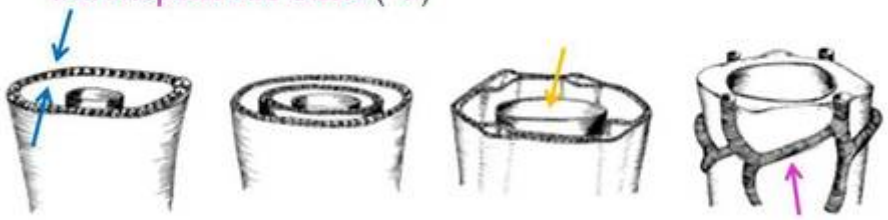


Illustration by Aletta Frazier

slide6.jpg

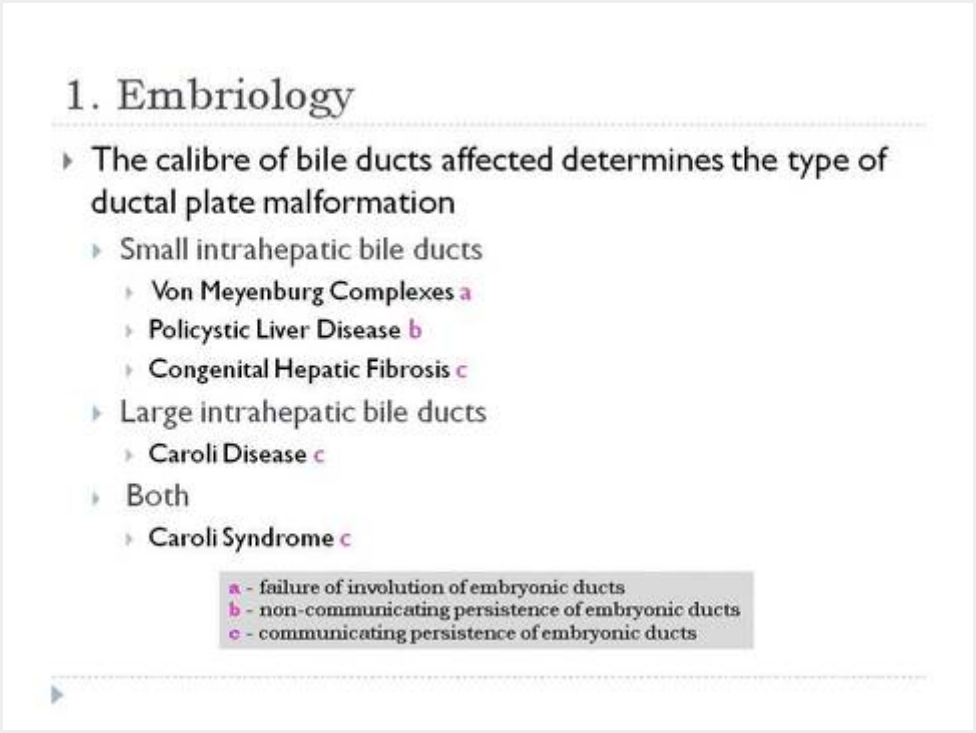
1. Embriology

- Ductal plate malformations result from abnormal persistence of embryonic bile duct structures, which may present as cystic, non-communicating lesions, or too numerous and ectatic bile ducts

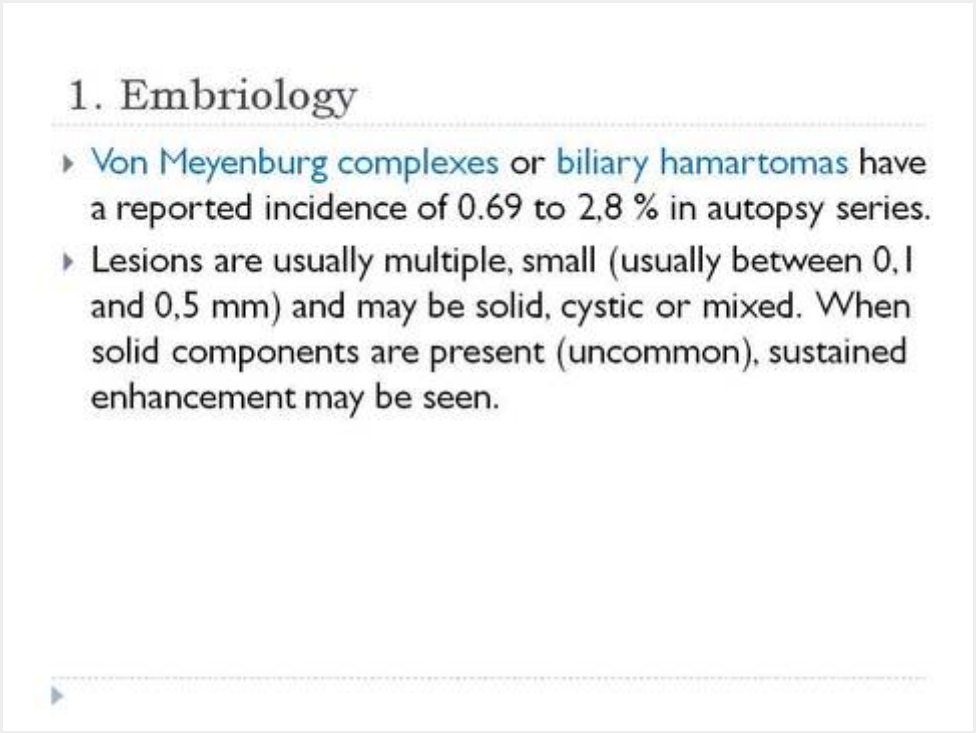
NORMAL PORTAL TRACT DUCTAL PLATE MALFORMATION

Illustration by Aletta Frazier

slide7.jpg

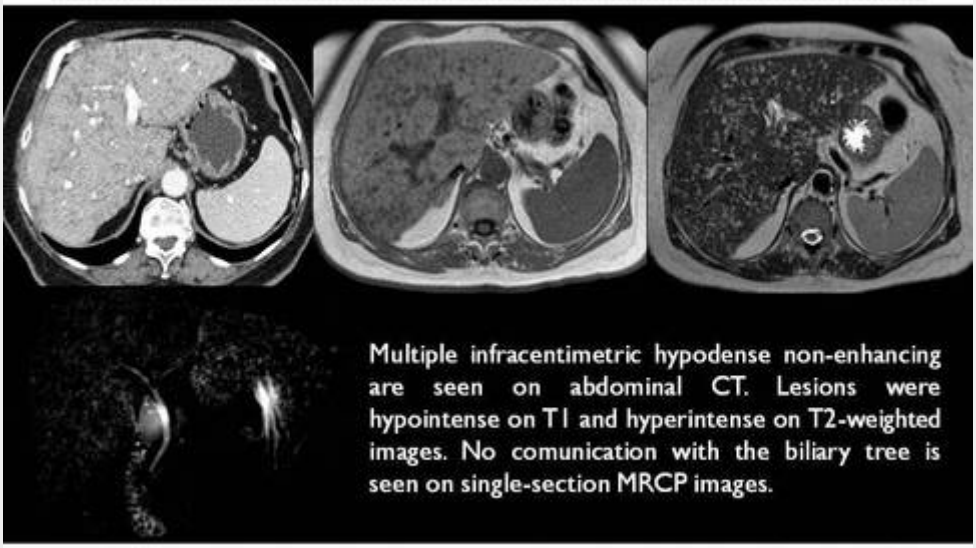


slide8.jpg



slide9.jpg

Von Meyenburg complexes



Multiple infracentimetric hypodense non-enhancing are seen on abdominal CT. Lesions were hypointense on T1 and hyperintense on T2-weighted images. No comunication with the biliary tree is seen on single-section MRCP images.

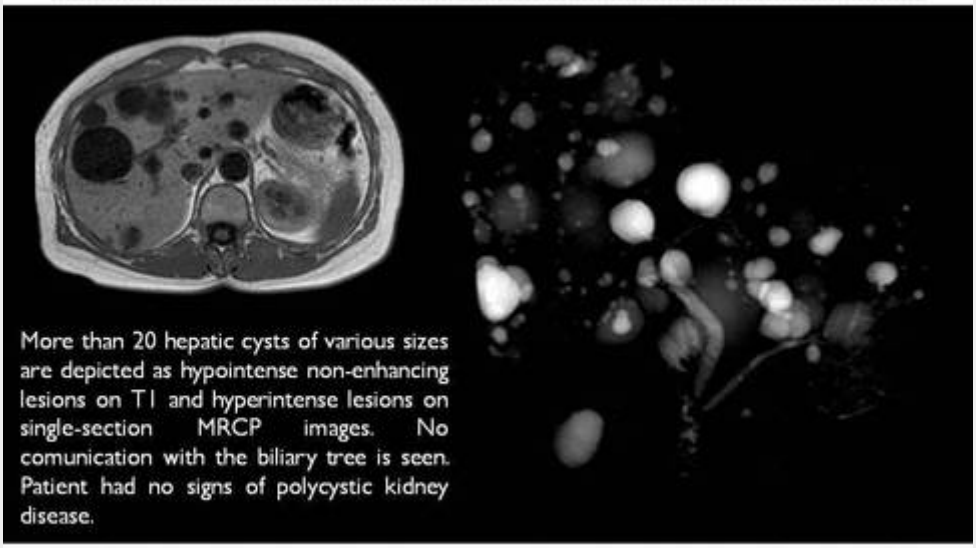
slide10.jpg

1. Embriology

- Polycystic liver disease may be associated with autossomic dominant polycystic kidney disease but also occurs as an isolated finding.
- The cysts arise from Von Meyenburg complexes lined with functional biliary epithelium. Both lesions may coexist.

slide11.jpg

Polycystic liver disease



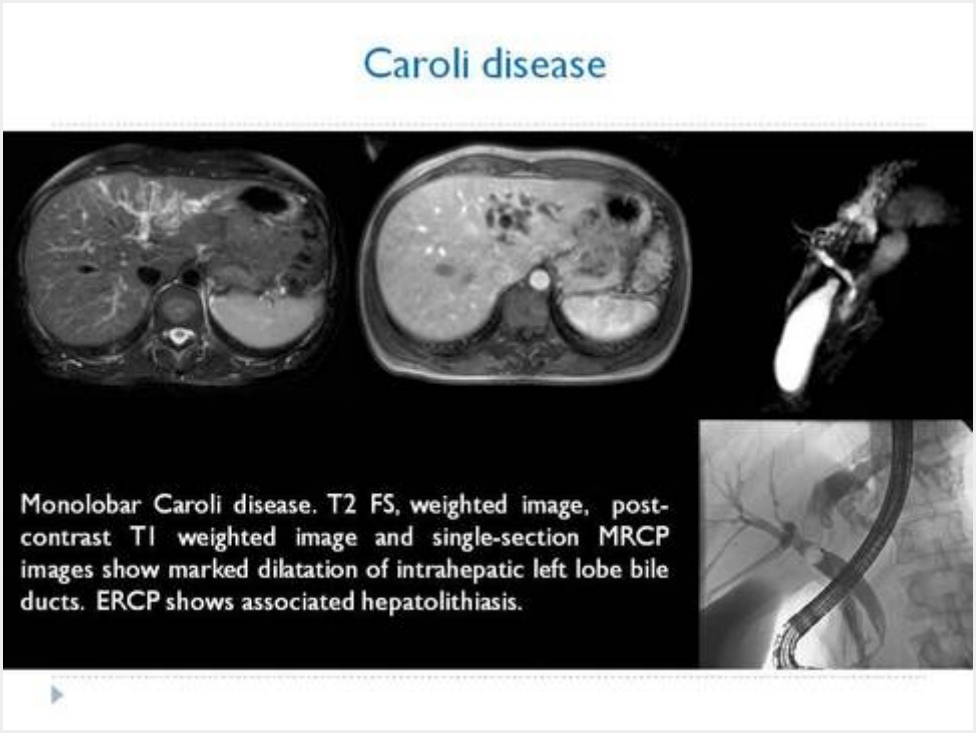
More than 20 hepatic cysts of various sizes are depicted as hypointense non-enhancing lesions on T1 and hyperintense lesions on single-section MRCP images. No comunication with the biliary tree is seen. Patient had no signs of polycystic kidney disease.

slide12.jpg

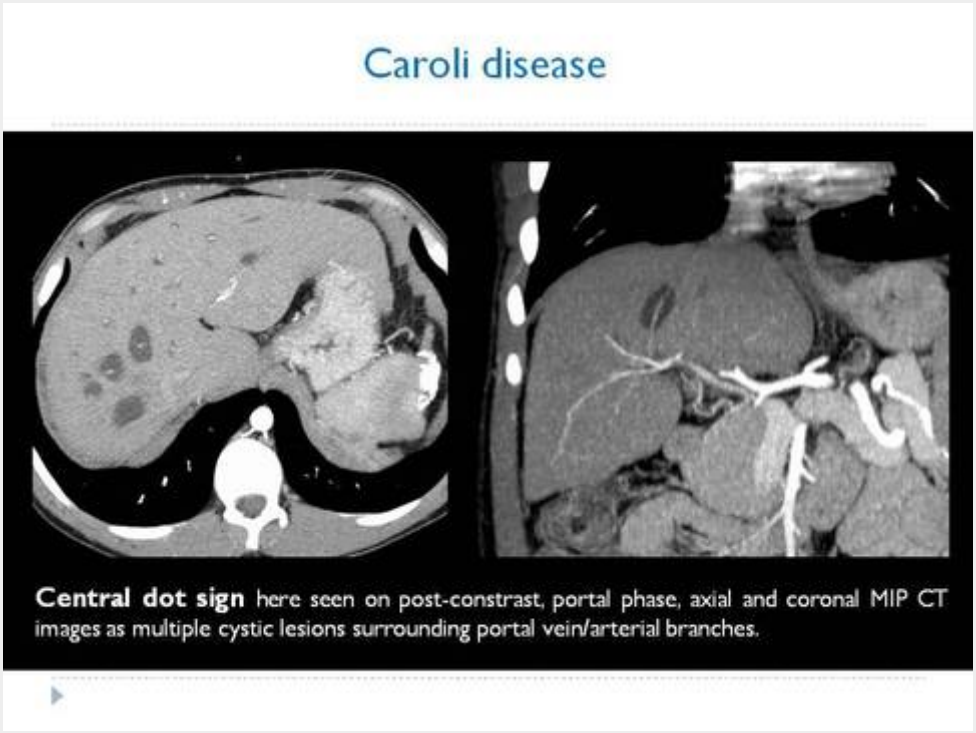
1. Embriology

- Caroli disease is a rare, autossomic recessive disease which is associated with kidney disease in up tp 60% of cases, namely medullary sponge kidney, ARPKD and nephronoptisis.
- It may be diffuse, lobar or segmental.
- The **central dot sign** is typical, due to cystic enveloping of portal or arterial branches.

slide13.jpg



slide14.jpg



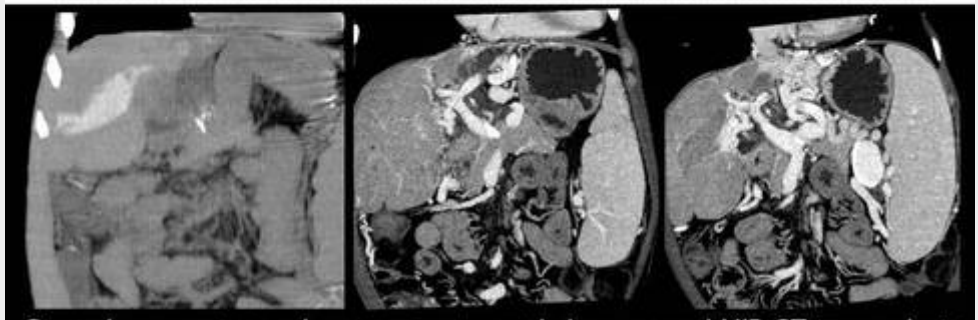
slide15.jpg

1. Embriology

- Caroli syndrome is said to be present when Caroli disease coexists with congenital hepatic fibrosis.
- Besides the complications of Caroli disease (cholangitis, hepatolithiasis, strictures and cholangiocarcinoma), portal hypertension and secondary biliary chirrosis are common late-stage findings.

slide16.jpg

Caroli syndrome

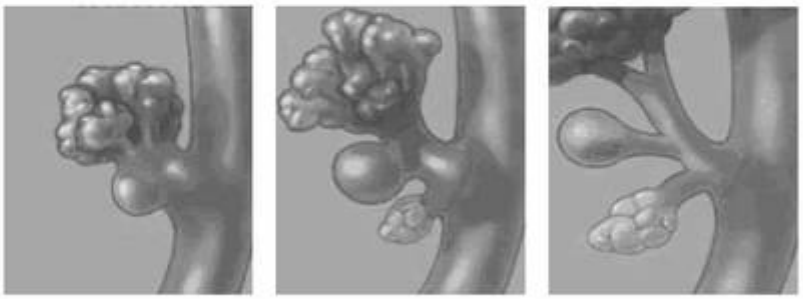


Coronal, pre-contrast and post-contrast, portal phase, coronal MIP CT images depict marked dilatation of intrahepatic bile ducts on both lobes. One of the dilated right-lobe bile duct is completely filled with milimetric calculus. There are associated findings of cirrhosis and portal hypertention, namely liver dysmorphia, splenomegaly and tortuous perigastric and perisplenic colateral veins. Signs of chronic portal vein thrombosis are also seen (perypheral, partially calcified thrombus).

slide17.jpg

1. Embriology

- The ventral part of the hepatic diverticulum (*pars cystica*) develops into the gallbladder, cystic duct and choledocus.
- The common hepatic duct derives from the *pars hepatica*.



Adapted from www.vesalius.com

slide18.jpg

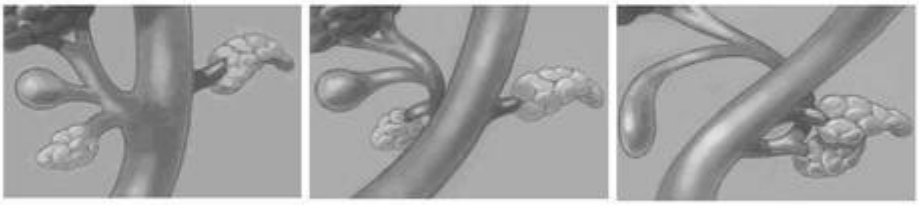
1. Embriology

- Intense remodeling of several channels present at the porta hepatis during the 5th week of embryonic life precedes the development of the left and right main hepatic ducts.
- Their proximal part derives from the first intrahepatic ductal plate and their distal part from the extrahepatic bile ducts.
- Alterations in this remodeling may be responsible for the several anatomic variants of the left-right hepatic duct junction.

slide19.jpg

1. Embriology

- By the end of the 6th week, the choledocus and ventral bud of the pancreas, so far draining distally into the choledocus, rotate posteriorly and come to lie it their definitive position.



Adapted from www.vesalius.com

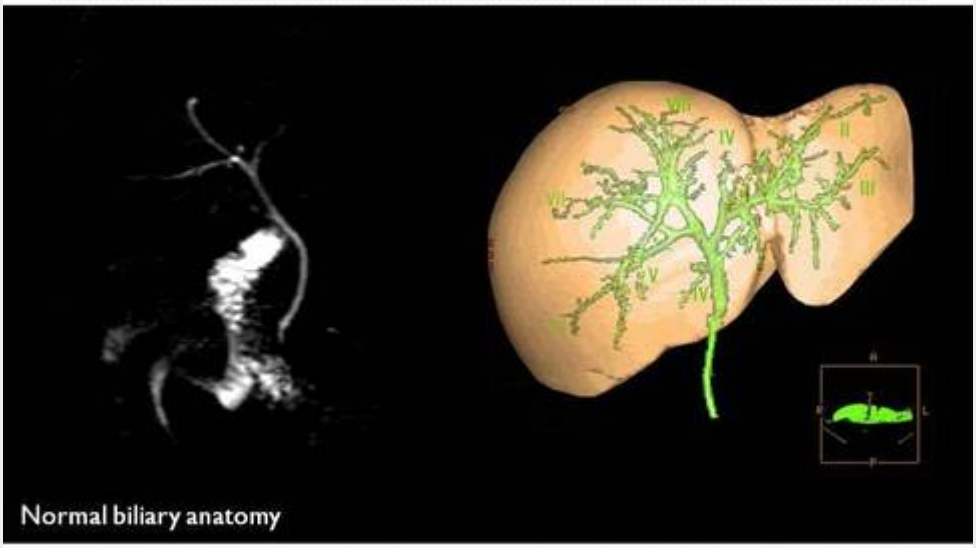
slide20.jpg

2. Normal Anatomy and Anatomic Variants

- The classic biliary anatomy is present in 58% of the population.
  - The **right main hepatic duct** results from the confluence of a horizontally-oriented **right posterior duct** (from segments VI and VII) and a more vertically-oriented **right anterior duct** (from segments V and VIII)
  - **Segment I** may drain to the left or right main ducts
  - The **cystic duct** drains into the lateral aspect of the common hepatic duct, midway between the hilum and the ampulla of Vater.

slide21.jpg

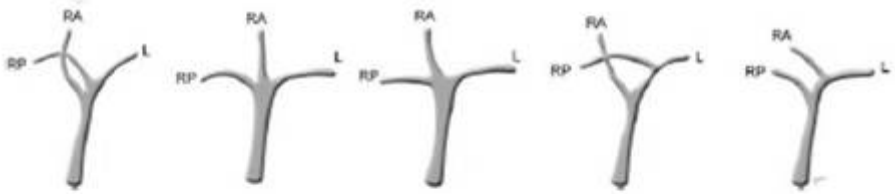
Normal biliary anatomy



slide22.jpg

2. Normal Anatomy and Anatomic Variants

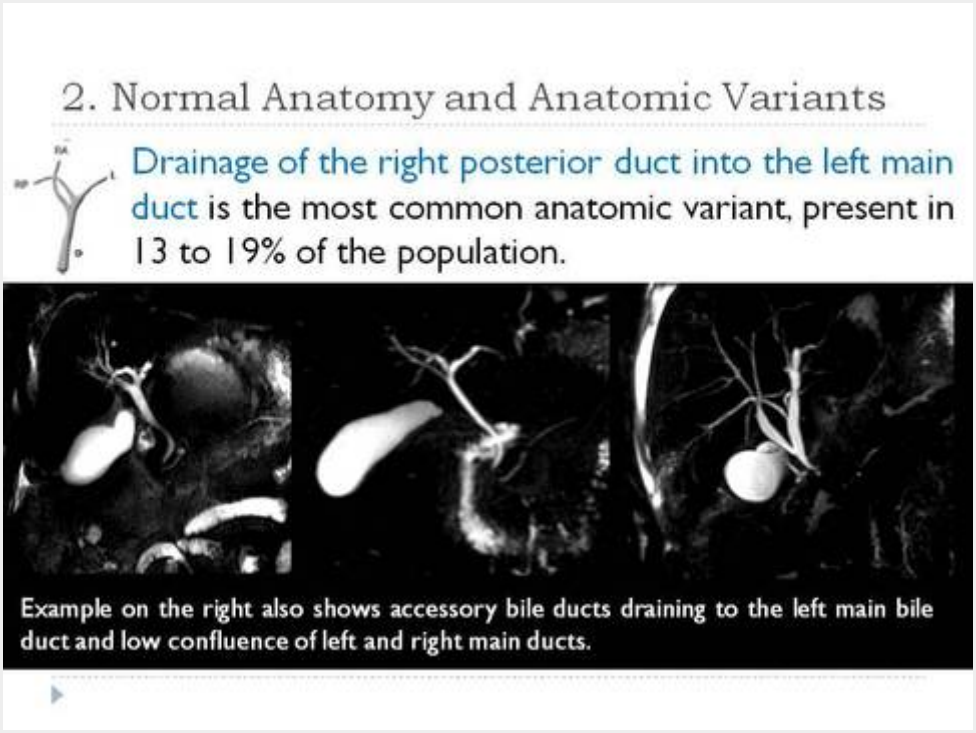
- There are several different anatomic variants of the hepatic duct confluence, the most important of which being depicted below.



RA right anterior hepatic duct
RP right porterior hepatic duct
L left hepatic duct

Radiographics March-April 2008 28:359-378

slide23.jpg



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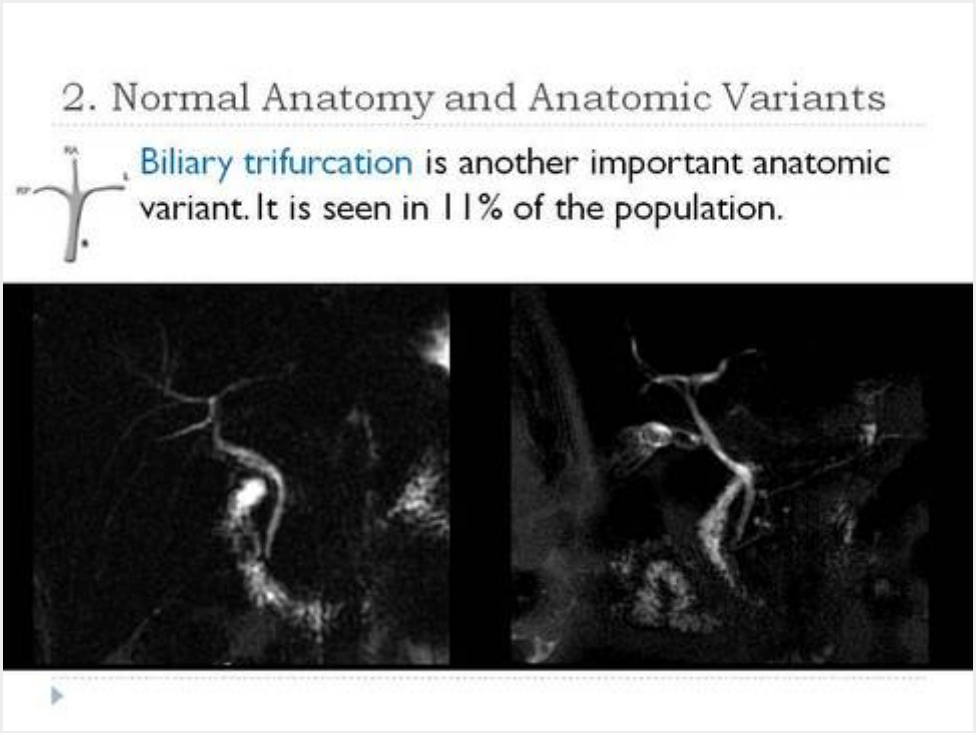
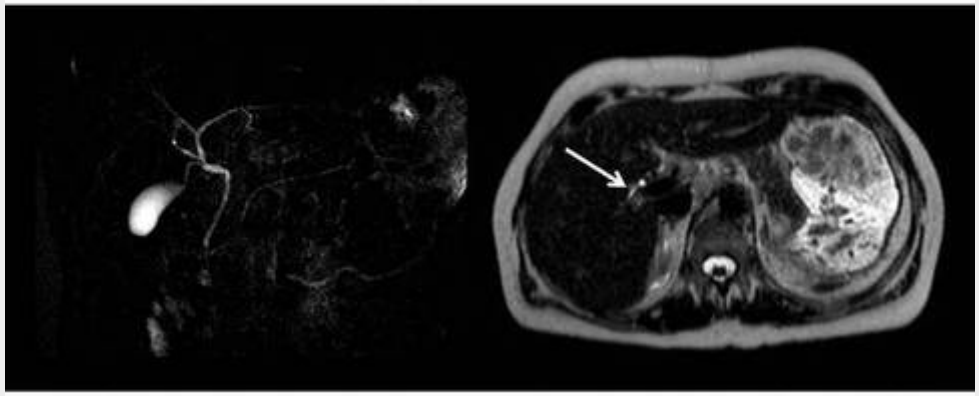


slide25.jpg

2. Normal Anatomy and Anatomic Variants

Drainage of the right posterior duct into the common hepatic duct is uncommon, being found in only 4% of the population.



slide26.jpg

2. Normal Anatomy and Anatomic Variants

- Cystic duct anatomic variants are found in 50.1 % of the population. The most common variants are depicted below.

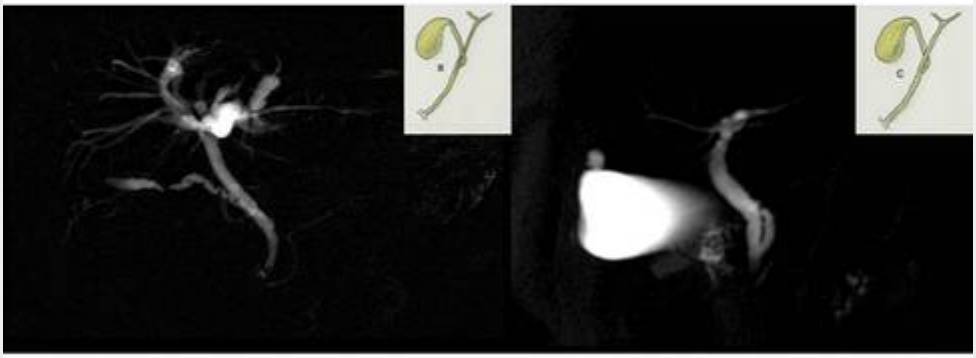
Right lateral insertion (*A*), anterior spiral insertion (*B*), posterior spiral insertion (*C*), low lateral insertion with a common sheath (*D*), proximal insertion (*E*), or low medial insertion (*F*).

January 2001 RadioGraphics, 21, 3-22

slide27.jpg

2. Normal Anatomy and Anatomic Variants

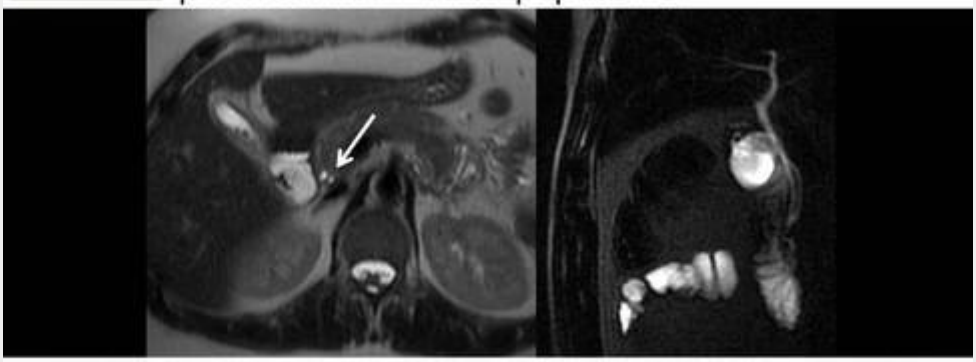
- Spiral insertion of the cystic duct is present in up to to 17% of the population.



slide28.jpg

2. Normal Anatomy and Anatomic Variants

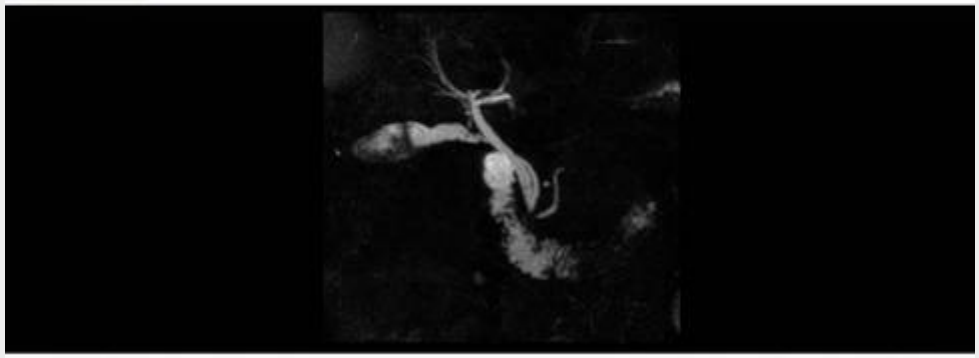
Cystic duct – common hepatic duct parallel course is said to be present when they run parallel and share a fibrous sheath along 2 cm or more. It is present in 10% of the population.



slide29.jpg

2. Normal Anatomy and Anatomic Variants

Low medial insertion of the cystic duct is present in 9% of the population.



slide30.jpg

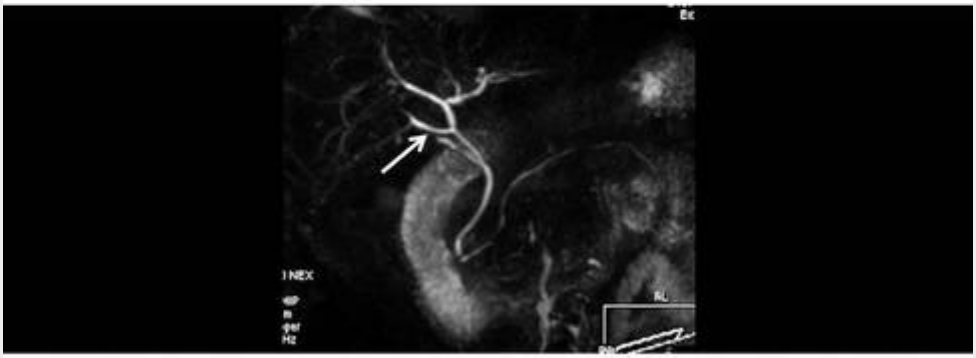
2. Normal Anatomy and Anatomic Variants

- Aberrant ducts are the only draining ducts of a particular hepatic segment.
- Aberrant ducts draining part of the right hepatic lobe may
  - empty directly into the cystic duct (cystohepatic ducts) in 1 to 2 % of individuals.
  - Empty into the right or common hepatic duct (bile duct of Luschka) in one third of the population.

slide31.jpg

2. Normal Anatomy and Anatomic Variants

- An aberrant right hepatic duct (Luschka) drains part of the right lobe directly to the main hepatic duct.



slide32.jpg

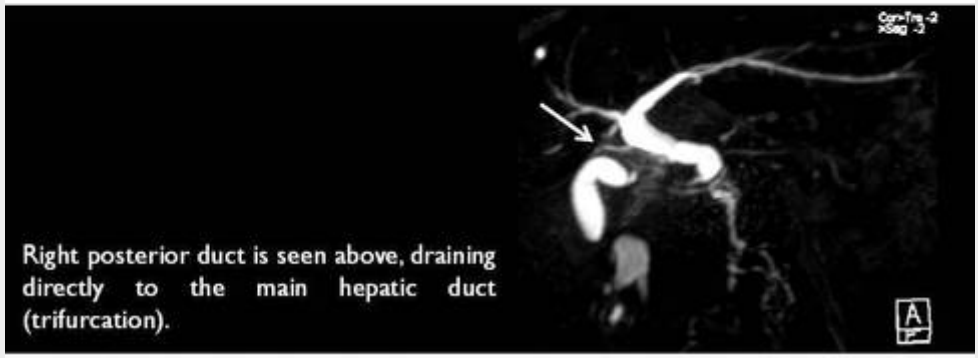
2. Normal Anatomy and Anatomic Variants

- Accessory ducts are additional ducts draining a particular hepatic segment. They are said to be present in aproximately 2% of the population.
- They may also join the common hepatic duct at its junction with the cystic or drain directly into the cystic duct.

slide33.jpg

2. Normal Anatomy and Anatomic Variants

- An accessory right hepatic duct drains part of the right lobe directly to the main hepatic duct (→).



slide34.jpg

2. Normal Anatomy and Anatomic Variants

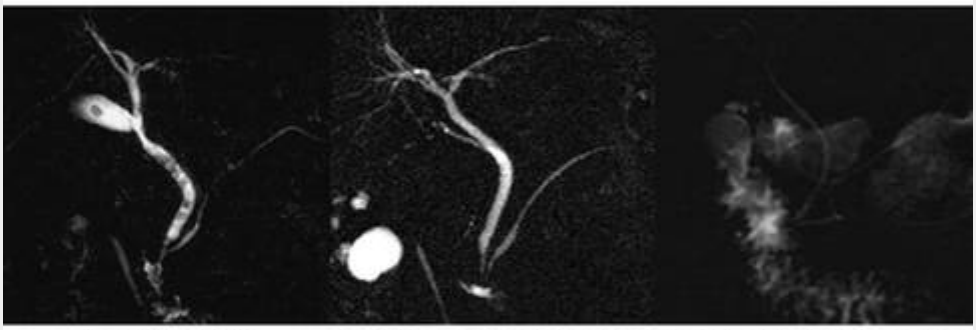
- The ampulla of Vater is another highly variable structure. The classic Y configuration of the union between the common bile duct and the pancreatic duct is observed in only 60% of the population.

Conventional Y configuration 60%	Double opening at the apex of the papilla 38%	Separate duodenal openings for the CBD and Wirsung 2%	Long common channel with union proximal to the duodenal wall rare

RadioGraphics, 22, 1335-1352

slide35.jpg

Anatomic Variants of the Ampulla of Vater



Conventional Y configuration 60%

Double opening at the apex of the papilla 38%

Separate duodenal openings for the CBD and Wirsung 2%

slide36.jpg

3. Iatrogenic Complications

- Variants of normal biliary anatomy may determine bile duct injury during hepatic surgery, such as tumor resection or partial hepatectomy for living donor transplantation. It may also occur during simpler procedures such as cholecystectomy, particularly laparoscopic cholecystectomy.
- Resultant iatrogenic lesions include inadvertent ductal ligation, bile leaks and strictures.

slide37.jpg

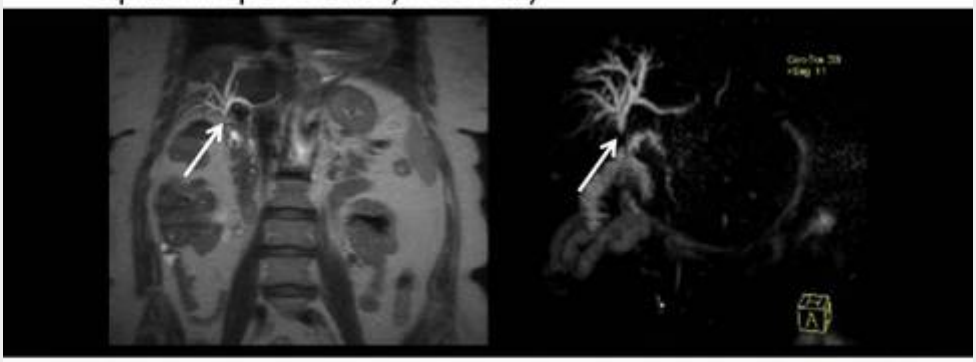
3. Iatrogenic Complications

- Bile leakage after hepatic or biliary surgery may occur in different locations, but mainly occurs at the distal branches of the hilar convergence. It may also occur at the repair site of the hepatic duct or along parenchymal transection surface of the liver.
- MR Cholangiography after IV mangafodipir trisodium administration is useful for bile leak detection and quantification.

slide38.jpg

3. Iatrogenic Complications

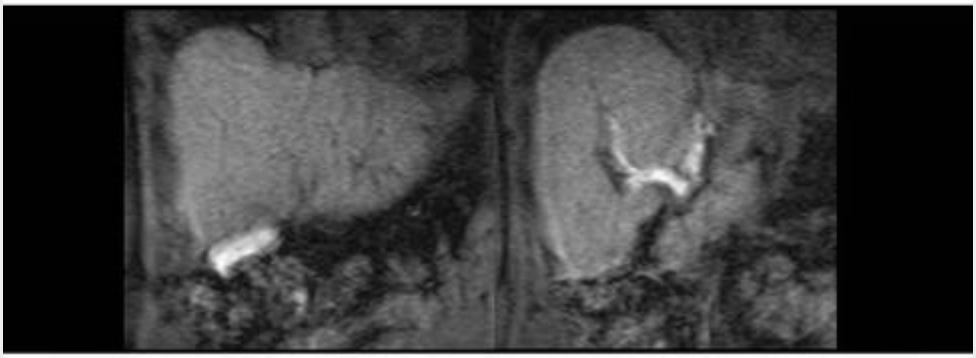
- Intrahepatic bile duct dilation due to inadvertent ligation of a right posterior bile duct with anomalous drainage to the left main hepatic duct during laparoscopic cholecystectomy.



slide39.jpg

3. Iatrogenic Complications

- A subhepatic with enhancement after IV Mangafodipir administration is seen, representing a biloma due to main hepatic duct leakage (→).

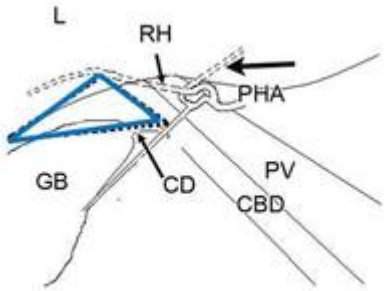


slide40.jpg

3. Iatrogenic Complications

- Particularly predisposing variants
  - Aberrant and accessory ducts which course in the triangle of Calot(-) and ducts that drain directly into the gallblader or cystic duct, are particularly susceptible to inadvertent ligation or transection during cholecystectomy.

CBD = Common bile duct
CD= Cystic duct
GB = Gallbladder
L = Liver
RH = Right hepatic artery
PHA = proper hepatic artery



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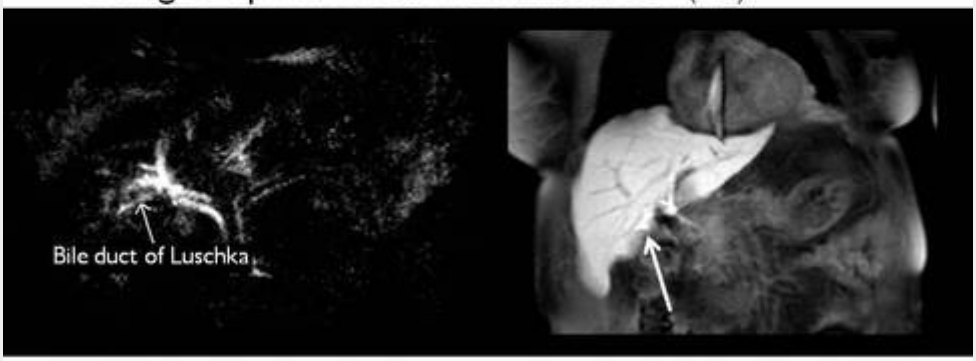
slide41.jpg

3. Iatrogenic Complications

- An accessory right hepatic duct (Luschka) was inadvertantly sectioned during laparoscopic cholecystectomy. Bile leak is depicted after IV mangafodipir trisodium administration (→).



slide42.jpg

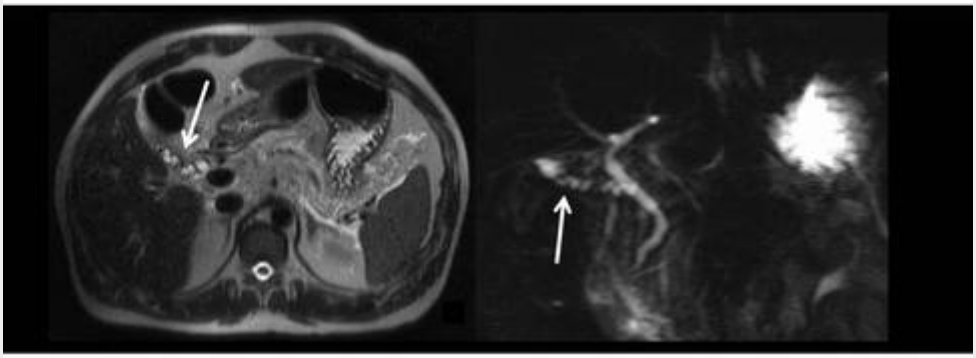
3. Iatrogenic Complications

- Particularly predisposing variants
  - Cystic duct – common hepatic duct parallel course may result in surgical ligation too close to the common hepatic duct, with stricturing or common bile duct transection.
  - An abnormaly long remnant after cholecystectomy in patients with this variant is common and may also result in **postcholecystectomy syndrome** due to inflamation and calculi formation.

slide43.jpg

3. Iatrogenic Complications

- An unusually long and tortuous cystic duct remnant, within which are several gallstones, is seen in a symptomatic patient (→).



slide44.jpg

3. Iatrogenic Complications

- Particularly predisposing variants
  - Low medial insertion of the cystic is particularly relevant because distal gallstone impaction may mimic choledocal obstruction and ERCP may result in inadvertent instrumentation of the cystic rather than the common bile duct.

slide45.jpg

3. Iatrogenic Complications

- In the particular case of tumor resection
  - With left lobe resection
    - Right posterior duct draining directly into the left hepatic duct
    - Biliary trifurcation

    are particularly relevant
  - With right lobe resection
    - Left hepatic duct draining directly into the right hepatic duct
    - Biliary trifurcation

    are particularly relevant

slide46.jpg

3. Iatrogenic Complications

- In the particular case of living donor transplantation
  - Variants considered relevant in donors are
    - Right posterior duct draining directly into the left hepatic duct
    - Biliary trifurcation
    - Right posterior duct draining directly into the left hepatic duct
    - Accessory hepatic ducts
  - Variants considered relevant in recipients are
    - Left hepatic duct into the right anterior hepatic duct
    - Biliary trifurcation
    - Cystic duct draining into the right hepatic duct
    - Accessory hepatic ducts

slide47.jpg

4. Congenital Anomalies

- Biliary atresia is defined as lack of lumen in part of or all of the extrahepatic bile duct. It is the most common cause of obstructive jaundice in the first month of life.
- It is an acquired disease, occuring in 1/10000 live births (girls more frequently affected), and is associated with polysplenia in 11% of cases.

slide48.jpg

4. Congenital Anomalies

- Biliary atresia of the extrahepatic bile ducts was classified by Kasai into three categories:
  - Type I: atresia of the common bile duct while bile duct segments adjacent to the liver are intact.
  - Type IIA: atresia of the common hepatic duct with or without atresia of the common bile duct.
  - Type IIB: atresia of all main branches of the extrahepatic system or lack of extrahepatic bile duct system.
  - Type III: atresia of the hepatic and cystic ducts with hilar ducts replaced by a fibrous core.
- Types I and II represent 10% of cases and are surgically correctable.
- Type III represents 90% of cases and is not correctable.

slide49.jpg

4. Congenital Anomalies

- Choledocal cysts are uncommon anomalies of the biliary system manifested by cystic dilatation of the extra and/or intrahepatic biliary tree.
- Incidence is higher in East Asia (1/1000) than in western countries (1/100000).
- Presentation occurs during childhood in 60% of cases and females are affected in 80% of cases.
- Choledocal cysts predispose to cholangitis, lithiasis, obstruction, pancreatitis and malignancy.
- There are five subtypes of choledochal cysts, as defined by Todani´s modification of the Alonso – Lej classification

slide50.jpg

4. Congenital Anomalies

- Todani´s modification of the Alonso – Lej classification

Type I	Solitary extrahepatic cyst
Type II	Extrahepatic duodenal diverticulum
Type III	Intraduodenal cyst
Type IV	Extrahepatic and intrahepatic cysts
Type V	Multiple intrahepatic cysts

slide51.jpg

4. Congenital Anomalies

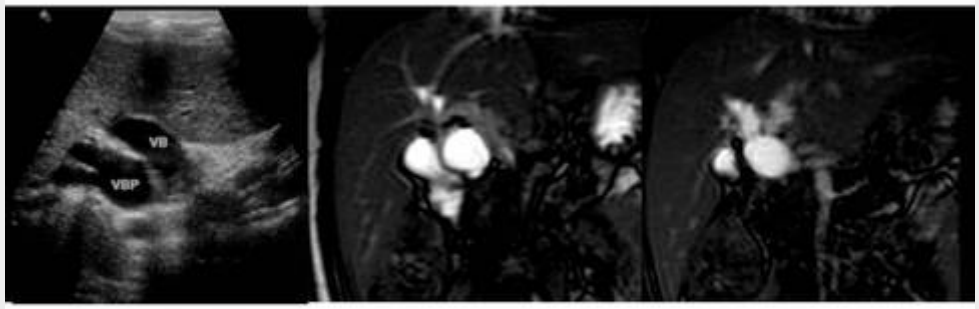
Type I

Type I A	Type I B	Type I C
Cystic dilation	Focal dilation	Fusiform dilation

slide52.jpg

4. Congenital Anomalies

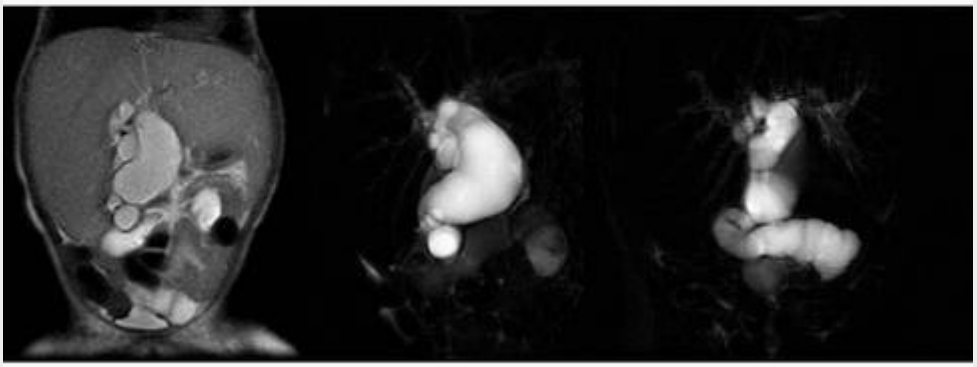
- **Type IC Choledochal Cyst** ♀, 6 Yo. Abdominal Ultrasound and MR SSFP sequences show a fusiform dilation of the common bile duct, continuous with the cystic duct. The right and left main hepatic ducts were also dilated.



slide53.jpg

3. Congenital Anomalies

- **Type IC Choledochal Cyst** ♀, 4 Yo with jaundice and abdominal pain. MR images shows fusiform dilation of the common bile duct.



slide54.jpg

4. Congenital Anomalies

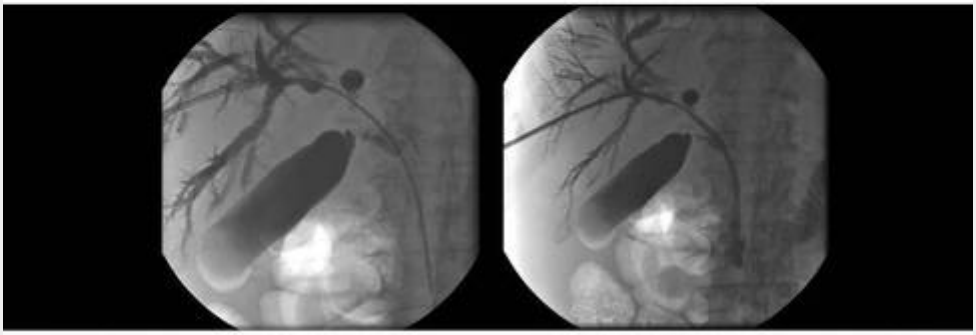
Type II

Diverticulum

slide55.jpg

4. Congenital Anomalies

- **Type II Choledochal Cyst - Diverticulum** ♂, 76 Yo. Cholangiocarcinoma – Klatskin tumor. Percutaneous transhepatic cholangiography (PTC) performed to paliate biliary obstruction showed a diverticulum of the common hepatic duct, located above the insertion of the cystic duct, draining to an area of neoplastic involvement.

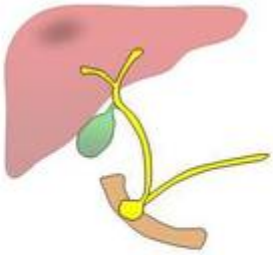


slide56.jpg

4. Congenital Anomalies

Type III

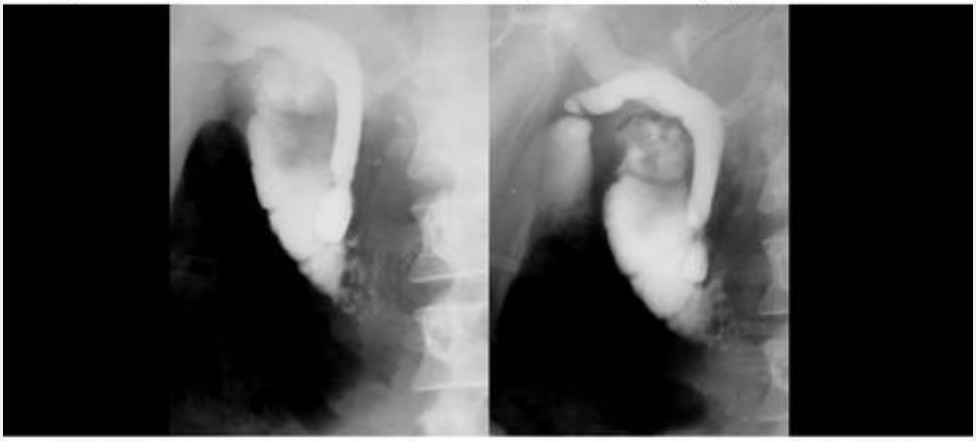
Choledochocele



slide57.jpg

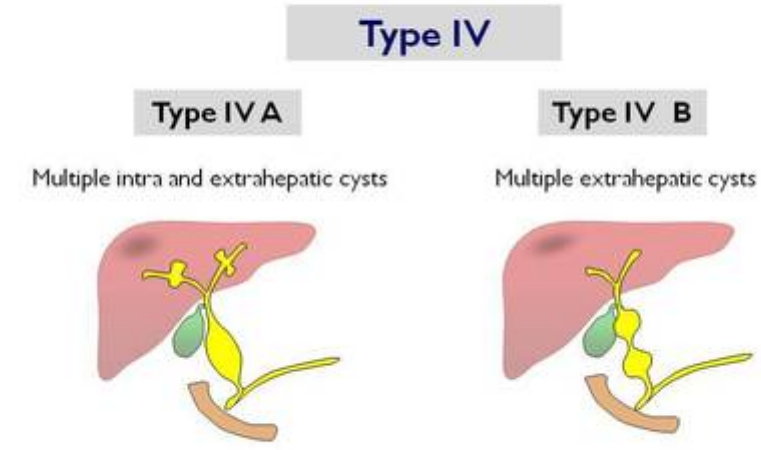
4. Congenital Anomalies

- **Type III Choledochal Cyst – Choledochocele** PTC shows a diverticulum of the common bile duct located within the 2nd portion of the duodenum´s wall, adjacent to the papilla of Vater.



slide58.jpg

4. Congenital Anomalies



slide59.jpg

4. Congenital Anomalies

Type IVA choledochal cyst as seen on PTC. Massive dilation of the common bile duct and multiple intrahepatic bile duct dilations are apparent.

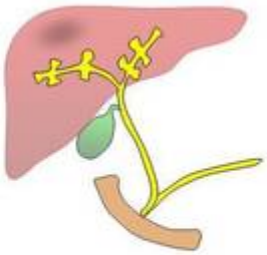
slide60.jpg

4. Congenital Anomalies

Type V

Caroli's Disease*

Multiple intrahepatic cysts



*Already discussed – see section I. Embryology

slide61.jpg

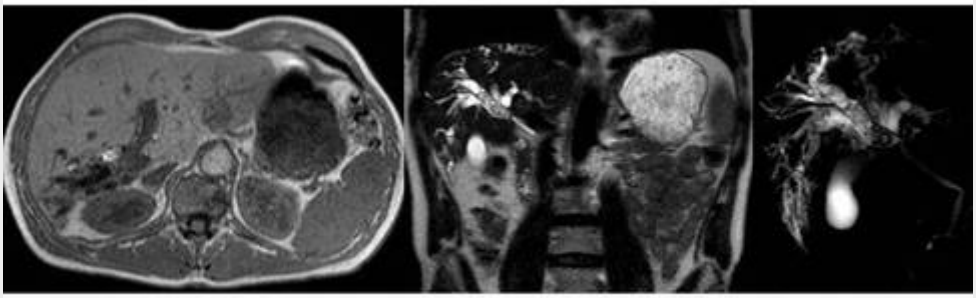
5. Pathology

- **Choledocholithiasis** is found in 10 to 15% of patients with cholelithiasis, and it´s incidence increases with increasing age.
- Undetected duct stones are left behind in 1 to 5% of patients after cholecystectomy.
- The majority of cases of choledocholithiasis are secondary (migrating cholesterol or mixed calculus from the gallbladder).
- Primary choledocholithiasis is uncommon and results from chronic hemolysis, hepatobiliary parasitism, chronic recurrent cholangitis, stenosis and strictures, congenital anomalies such as Caroli disease (*example above – Embriology section*).
- Common complications are biliary obstruction, cholangitis, pancreatitis and secondary biliary cirrhosis.

slide62.jpg

5. Pathology

- **Primary cholelithiasis** Patient with a history of trauma presents with obstructive ictericia. MR Colangiography shows right posterior segments bile duct ectasia due to a longstanding stenosis of the corresponding bile duct. Multiple hyperintense on T1-weighted and hypointense on T2-weighted images intraductal filling defects as depicted and result from primary bile-pigment-rich cholelithiasis.



slide63.jpg

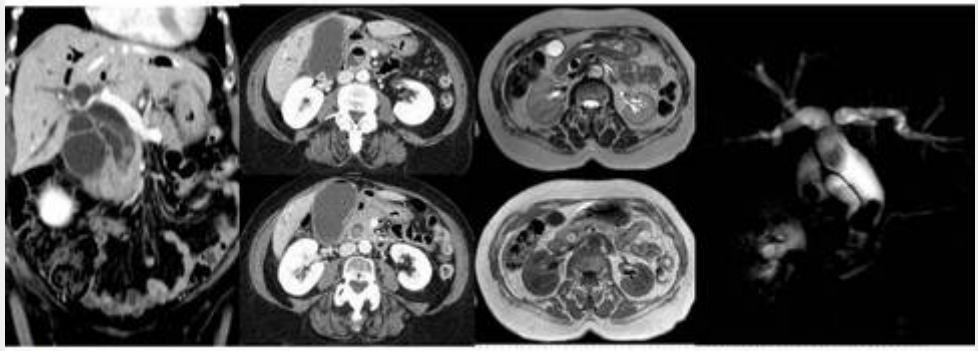
5. Pathology

- **Secondary cholelithiasis** Axial CT images after IV contrast administration show cholelithiasis and main hepatic duct and cystic duct dilation (notice main hepatic duct and cystic duct parallel course). Distally, a round filling defect is seen in the main bile duct, corresponding to a calculus(→).

slide64.jpg

5. Pathology

- **Secondary cholelithiasis** CT images after IV contrast shows intra and extrahepatic bile duct dilatation and two foci of choledocolithiasis. Signs of cholecystitis are also apparent. Axial MR images show the bile duct stone as a hypointense round image surrounded by hyperintense bile on T2 weighted and T2 SSH-TSE images. The stone is hyperintense on T1-weighted images.



slide65.jpg

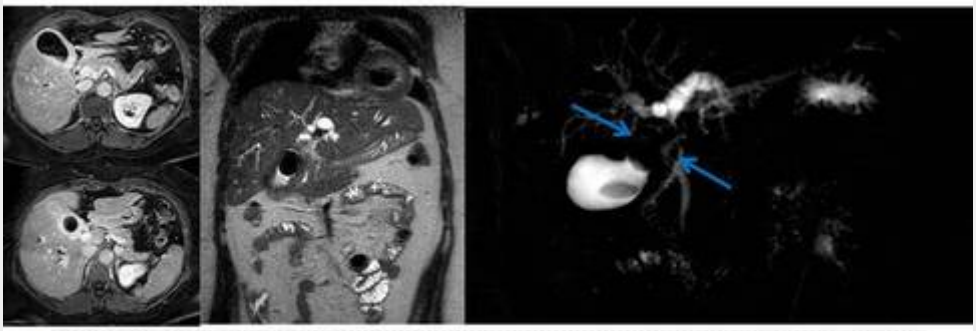
5. Pathology

- Mirizzi syndrome results from impaction of a calculus within the cystic duct and subsequent extrinsic compression of the common hepatic duct. Some authors also relate the obstruction to local extension of the inflammatory process (cholecystitis) to involve the common hepatic or common bile duct.
- Low medial insertion of the cystic duct and a parallel cystic-hepatic duct course are predisposing factors.

slide66.jpg

5. Pathology

- **Mirizzi Syndrome** T1 FS post-gadolinium images show thickening of the gallbladder's wall due to cholecystitis. Coronal T2-weighted images show a gallstone inpacted in the infundibulum – cystic duct. T2 SSH-TSE images show a luminal defect in the infundibular-cystic duct region, compressing the main hepatic duct and thus causing intrahepatic bile duct dilatation.



slide67.jpg

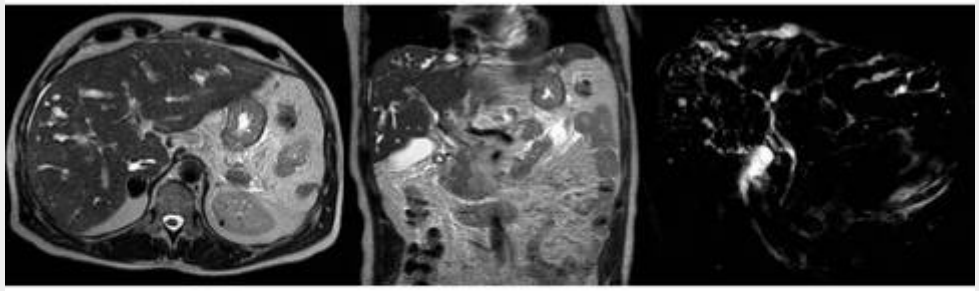
5. Pathology

- Sclerosing cholangitis results from chronic progressive inflammation and fibrosis of the intra and/or extrahepatic bile ducts.
- It may be primary (etiology not known), in which a strong association with inflammatory bowel disease (70%), particularly ulcerative colitis, is seen. A weaker association with multifocal fibrosclerosis syndromes such as mediastinal and retroperitoneal fibrosis has also been reported.
- Secondary sclerosing cholangitis may occur as a long-term complication of choledocholithiasis, cholangiocarcinoma, operative or traumatic biliary injury or contiguous inflammatory processes.
- Findings in all similar to sclerosing cholangitis are found in AIDS cholangiopathy, *Cryptosporidium*, MAI, CMV, *Microsporidia* and *Isospora* being associated infectious organisms.

slide68.jpg

5. Pathology

- **Primary sclerosing cholangitis** in patient with Chron disease. Multiple irregular intrahepatic and extrahepatic bile duct dilatations are seen on T2 FSE and e T2 SSH-TSE MR images.



slide69.jpg

5. Pathology

- The incidence of cholangiocarcinoma is increasing worldwide.
- Liver fluxes and hepatolithiasis are common risk factors in Asia, while primary sclerosing cholangitis, cirrhosis, alcohol and diabetes are common risk factors in western countries.
- Extrahepatic cholangiocarcinomas derive from the main bile duct (*hilar or Klatskin tumors are included in the intrahepatic subgroup*) They occur:
  - in the upper third in 50 to 75%,
  - in the middle third in 10 to 25%
  - in the lower third in 10 to 20% of cases.

slide70.jpg

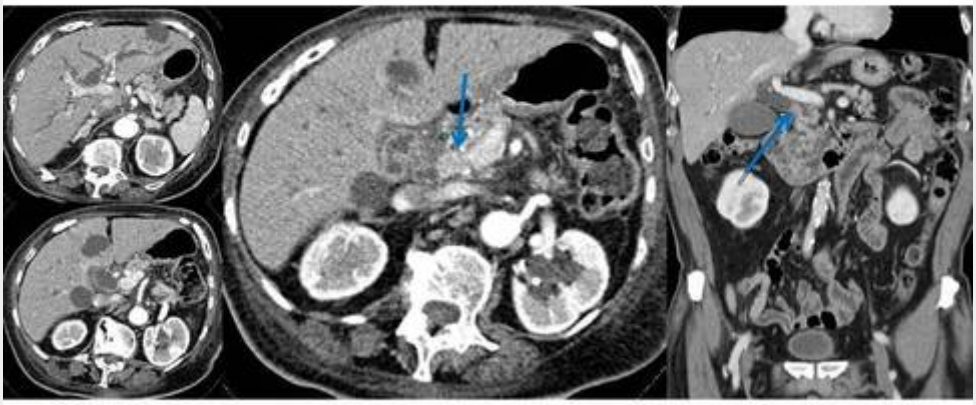
5. Pathology

- Extrahepatic cholangiocarcinoma may be classified morphologically as:
  - Mass-forming or nodular. A round, small mass is seen ocluding the main bile duct.
  - Periductal-infiltrating or sclerosing. Segmental or diffuse, desmoplastic thickening of the extrahepatic bile ducts without a discrete mass.
  - Intraductal-growing or papillary. One or multiple intraluminal friable polypoid lesions (cholangiocarcinomatosis),
- Periductal-infiltrating is by far the most common morphologic type observed.

slide71.jpg

5. Pathology

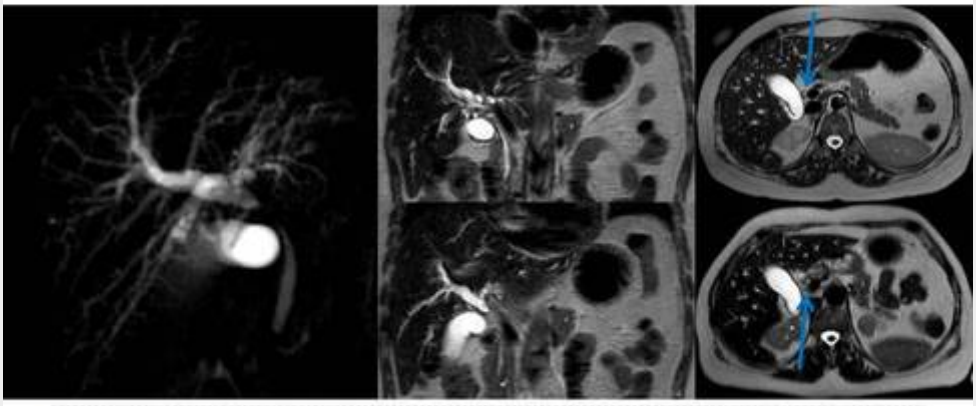
- Middle-third **cholangiocarcinoma**, sclerosing type. Post-contrast CT images show circunferential thickening of the common bile duct, resulting in marked narrowing of the lumen and intrahepatic bile duct dilatation.



slide72.jpg

5. Pathology

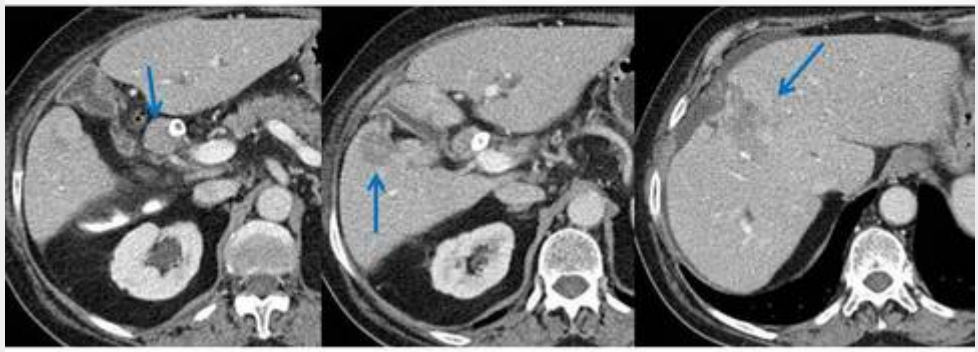
- Middle-third **cholangiocarcinoma**, sclerosing type. A similar example, here seen on T2-weighted images and SSFP MR images.



slide73.jpg

5. Pathology

- Multifocal, intrahepatic perypheral and extrahepatic, upper third, mass-forming type **cholangiocarcinoma** seen on post-contrast CT images A biliary prothesis had already been placed.



slide74.jpg

5. Pathology

- The periampullary carcinomas are those that arise within 2 cm of the major duodenal papilla and comprise four different malignancies:
  - ampullomas
  - distal cholangiocarcinomas
  - carcinomas of the pancreas
  - carcinomas of the duodenum
- Although their clinical features and surgical aproaches are similar, accurate determination of tumor origin is essential to adequately predict prognosis.

slide75.jpg

5. Pathology

- Although ampullary carcinoma is described by some as a CCC arising in the last centimeter of the CBD, it shares more features with duodenal carcinoma than with CCC, namely molecular features and clinical outcomes.
- It usually presents as a small (0.5 to 2 cm) nodule or as an irregular filling defect at the distal margin of the pancreaticobiliary junction. Less commonly, irregular periductal thickening may be seen. Although the double duct sign is typically found, the Wirsung may not be ectatic, possibly because, in 40% of individuals, the CBD and Wirsung drain separately into the duodenum.

RadioGraphics, 22, 1335-1352

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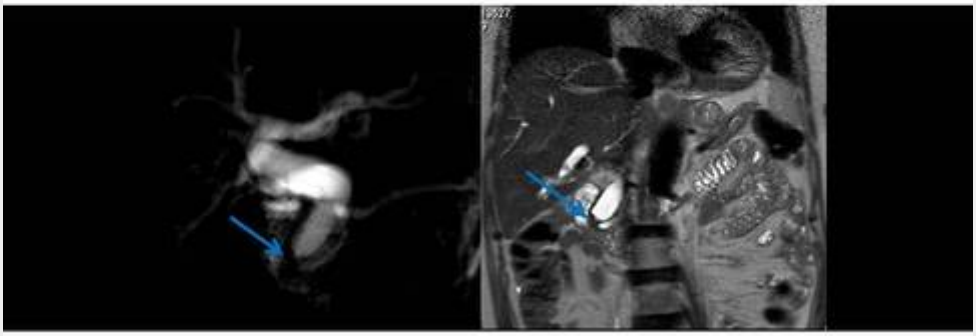
5. Pathology

- Marked abrupt duct dilatation in the absense of features of choledocolithiasis or pancreatitis should suggest the diagnosis of ampulloma.
- Differentiation of ampullary tumors from rare benign tumors in this location is virtually impossible with imaging.
- The morphologic type is usually polypoid and extraluminal extension, lymphatic spread and perineural invasion are infrequent. These features, along with early-onset of ictericia and thus early detection, contribute to it´s relatively good prognosis, with mean 5-year survival rates aproaching 40%.

slide77.jpg

5. Pathology

- Ampullary carcinoma Marked dilatation of the common bile duct with an abrupt cutoff at its intraparietal duodenal segment is seen on MR Colangiography T2 SSH-TSE images. Circunferential thickening of the ampulla is seen on coronal T2 TSE images.



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5. Pathology

- Pancreatic carcinoma is the most frequent carcinoma in the periampullary region.
- It tends to be larger (1 – 2,5 cm) and more proximal (mean distance to the duodenal lumen of 25 mm) upon detection than ampulloma.
- Blunted, beak or rat tail terminations of the main bile and pancreatic ducts are typically seen in association with an uncinate process parenchymal mass.

slide79.jpg

5. Pathology

- The double duct sign is seen in 62% of patients with pancreatic carcinoma. Sometimes, the short distal duct segments may also be seen – four segment sign. This finding is very unusual in the other types of periampullary carcinomas. It may be seen in chronic pancreatitis but the narrowing tends to be long and incomplete.
- Dilatation of side branches of the Wirsung around the stenotic segment is not infrequent in pancreatic carcinomas and is also unusual with the other 3 types of periampulary carcinomas.

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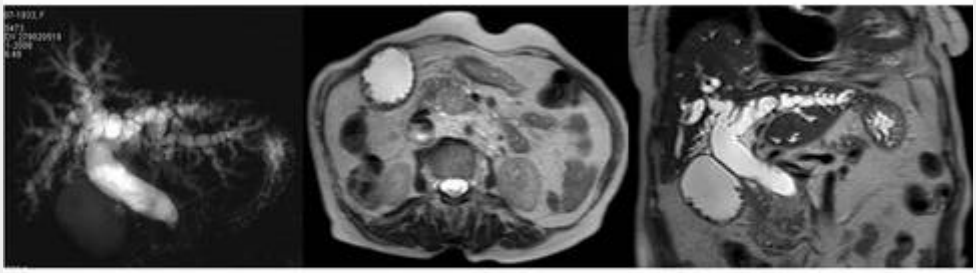
5. Pathology

- Carcinoma of the uncinate process of the pancreas Marked dilatation of bile ducts is seen on T2 SSH-TSE images, with an abrupt cutoff at the distal portion of the intrapancreatic segment of the common bile duct, due to a nodular mass, corresponding to a pancreatic adenocarcinoma. The mass is slightly hyperintense on T2 FSE images.



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5. Pathology

- Distal common bile duct carcinoma is of the periductal-infiltrating type in the large majority of cases. An intraductal-growing type may be found less frequently.
- Common bile duct dilatation is found in almost all patients. Not uncommonly, one can see both the distal and proximal CBD segment which, along with the pancreatic duct ,forms the three-segment sign.
- Extension to the pancreatic duct may cause dilatation and advanced stages may be impossible to differentiate from pancreatic carcinomas. In these cases, prognosis worsens accordingly.

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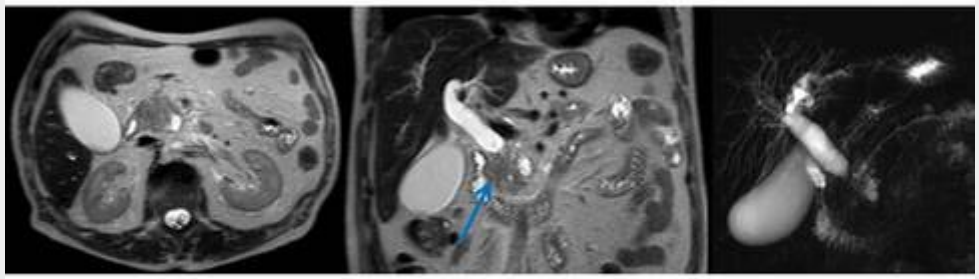
5. Pathology

- Distal common bile duct carcinoma, sclerosing type. Marked dilatation of bile ducts is seen on T2 SSH-TSE images, with an abrupt cutoff at the distal portion of the intrapancreatic segment of the common bile duct, due to circunferential tumoral thickening. The mass is isointense to the pancreatic parenquima on T2 FSE images.



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5. Pathology

- Duodenal carcinoma is the least common periampullary carcinoma.
- It may present either as a polypoid mass, a fungating mass or as a scirrhous carcinoma with eccentric wall thickening and duodenal luminal narrowing.
- Dilatation of the CBD and/or Wirsung, when present, tends to be inconspicuous. This finding may allow differentiation from the other types of periampullary carcinomas.
- Lymph node metastasis are frequent upon diagnosis but still, the mean 5-year survival rate is better than for all the other periampullary carcinomas, aproaching 50%.

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5. Pathology

- The extrahepatic bile ducts may also be secondarily invaded by non-periampullary tumors, such as stomach or gallbladder carcinoma.

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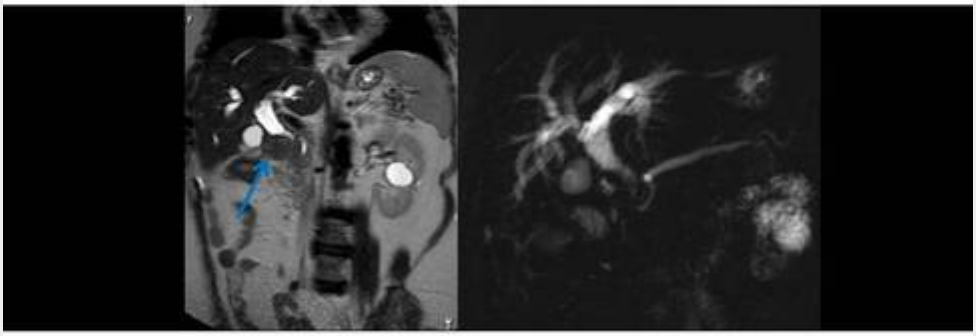
5. Pathology

- A nodal metastasis from gastric carcinoma located in the pancreatic groove is seen on coronal T2 FSE and T2 SSH-TSE MR images, extrinsically compressing the common bile duct and causing proximal biliary dilatation.



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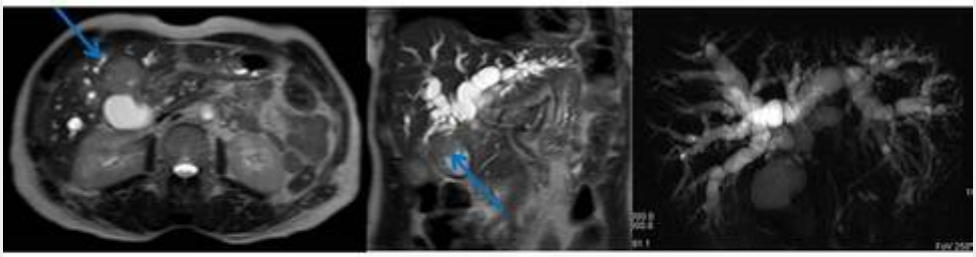
5. Pathology

- A gallbladder carcinoma, here seen on T2 FSE and T2 SSH-TSE MR images, extrinsically invades the liver and common hepatic duct causing massive intrahepatic bile duct dilatation.



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Conclusion

- Imaging plays a determinant role in the evaluation of the EHBDs, not only for the diagnosis of congenital anomalies and acquired disease, but also for accurate pre-operative anatomic mapping.

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Background

- The recognition of anatomical variants of the EHBDs is of utmost importance, namely in the pre-operative evaluation for laparoscopic cholecystectomy and in patient selection for living donor liver transplantation. Congenital anomalies of the EHBDs, on the other hand, are known predisposing factors for the development of recurrent pancreatitis, cholangitis, lithiasis and malignancies and shouldn´t go unrecognized. The EHBDs may also be afflicted by lithiasis, primary neoplasms, extrinsic compression/invasion and post-operative complications, such as leaks and stenosis.

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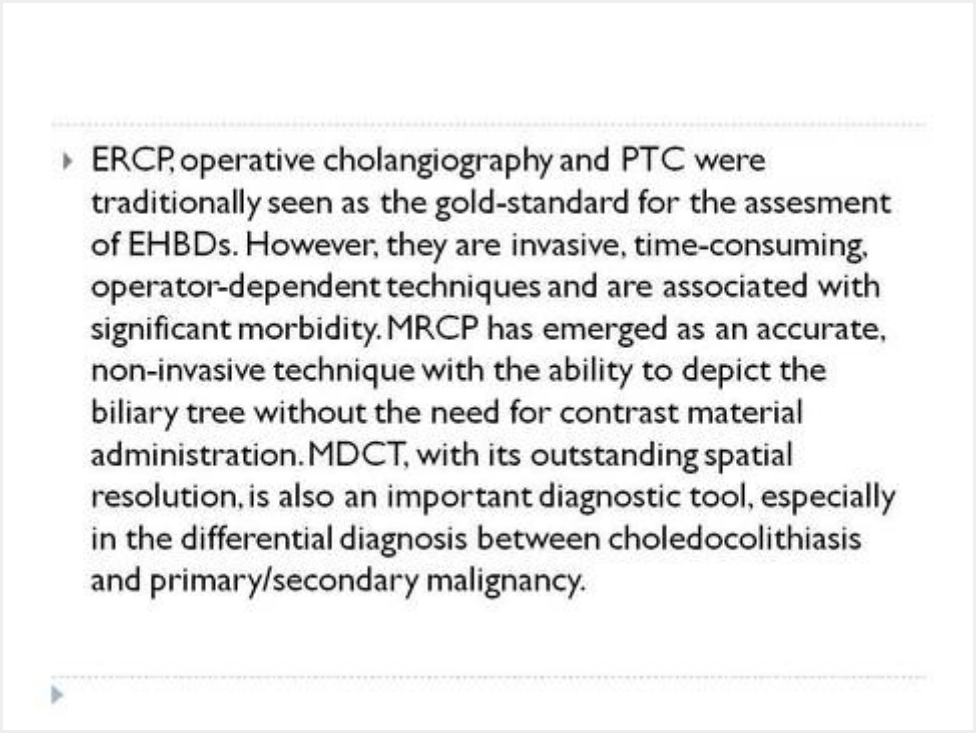
ERCP, operative cholangiography and PTC were traditionally seen as the gold-standard for the assesment of EHBDs. However, they are invasive, time-consuming, operator-dependent techniques and are associated with significant morbidity. MRCP has emerged as an accurate, non-invasive technique with the ability to depict the biliary tree without the need for contrast material administration. MDCT, with its outstanding spatial resolution, is also an important diagnostic tool, especially in the differential diagnosis between choledocolithiasis and primary/secondary malignancy.