



Choledochal Cysts - Spectrum of Imaging Findings

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MeSH:

Choledochal Cyst [C06.130.120.127]

Keyword: Choledochal Cysts

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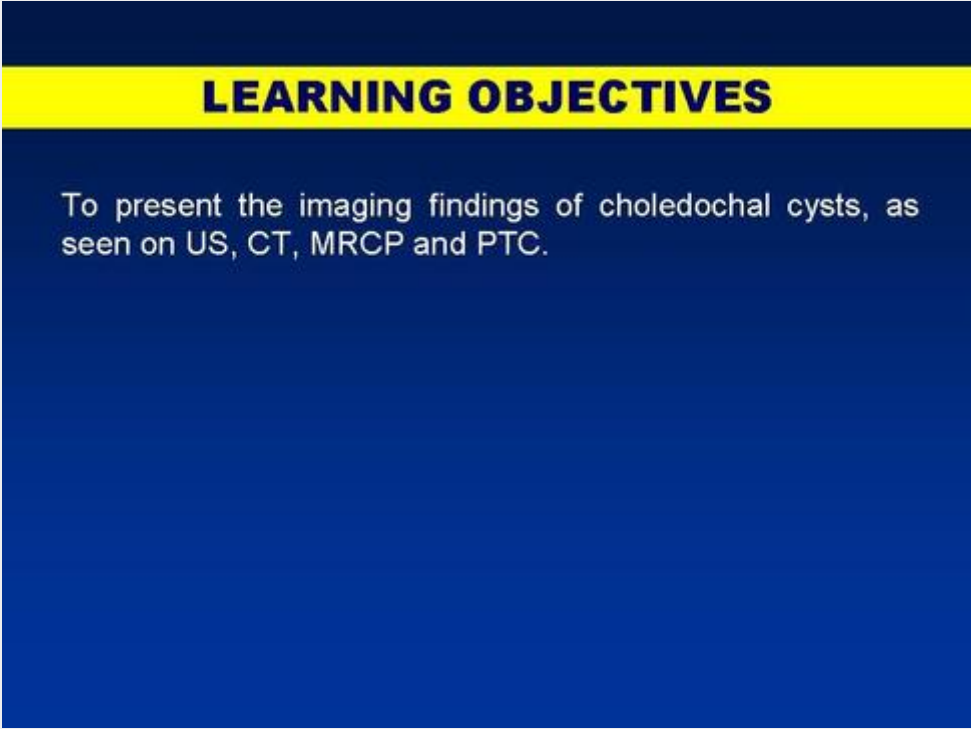
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1. Learning objectives

Learning Objectives

Learning objectives



LEARNING OBJECTIVES

To present the imaging findings of choledochal cysts, as seen on US, CT, MRCP and PTC.

2. Background

Background

Definition of choledochal cysts

BACKGROUND

Definition

- Uncommon anomalies of the biliary system manifested by cystic dilatation of the extra and/or intrahepatic biliary tree

diapositivo4.jpg

BACKGROUND

Origin

- Pancreatobiliary junction anomalies may promote reflux of pancreatic juice into the common bile duct, resulting in

inflammation



weakening of the bile duct wall



dilation

diapositivo5.jpg

BACKGROUND

Origin

Some speculate that the reflux may also happen the other way around – bile into the Wirsung channel -, predisposing to pancreatitis, which has a relatively high incidence in patients with cholelithiasis

diapositivo6.jpg

BACKGROUND

Origin

Other proposed mechanisms are:

- inherited/genetic factors
- infection
- congenital weakness in the walls of the biliary tract
- dysfunction of the sphincter of Oddi
- distal obstruction

diapositivo7.jpg

BACKGROUND

Epidemiology

- Estimated incidence:
 - 1/100000 in western countries
 - 1/1000 in Asia
- Higher prevalence in East Asia, particularly Japan
- Higher incidence in children – 60% in the 1st decade of life
- Higher incidence in ♀ - 80%
- 20% diagnosed in adults

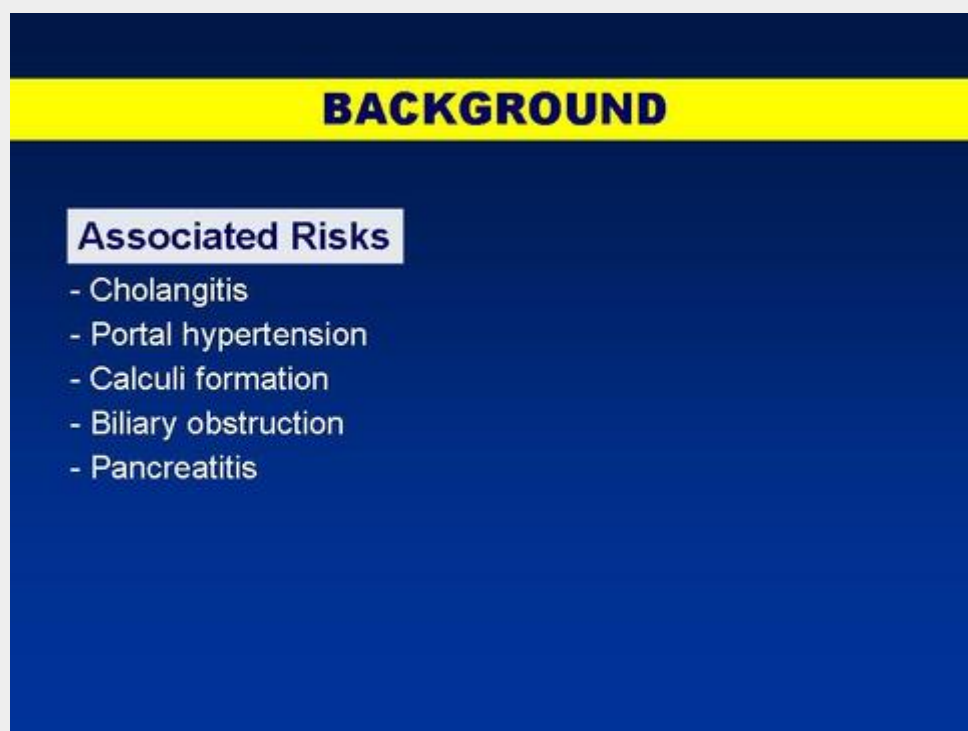
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BACKGROUND

Clinical Findings

- Classic presentation in a child:
 - jaundice
 - right upper quadrant pain
 - palpable right upper quadrant mass } 33%
- Presentation in adults:
 - right upper quadrant pain
 - pancreatitis
 - jaundice

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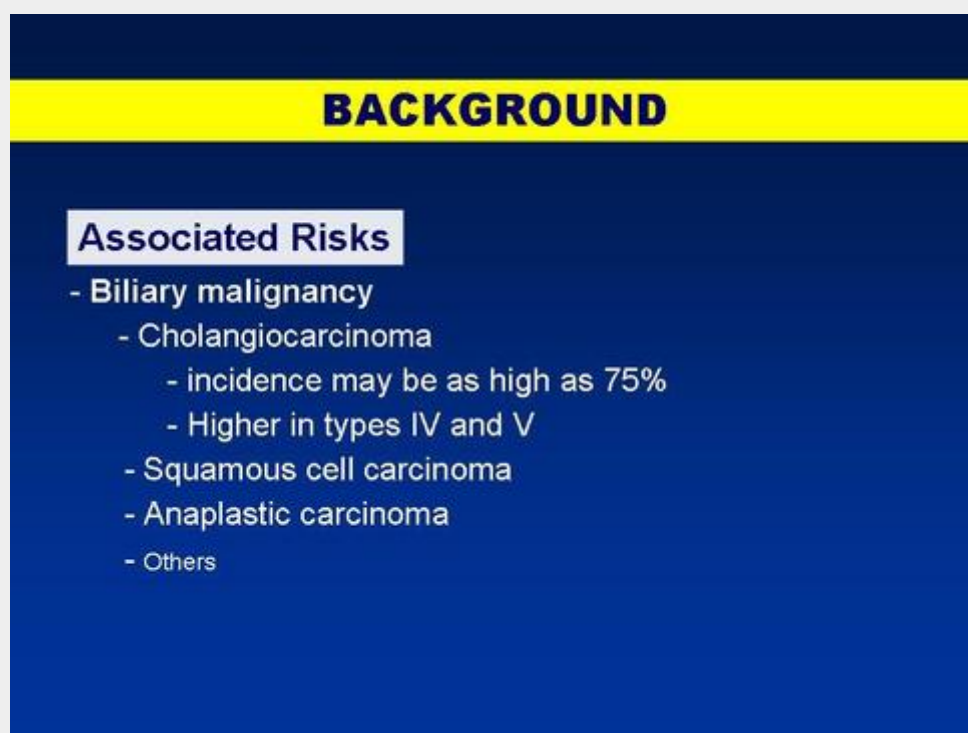


BACKGROUND

Associated Risks

- Cholangitis
- Portal hypertension
- Calculi formation
- Biliary obstruction
- Pancreatitis

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BACKGROUND

Associated Risks

- Biliary malignancy
 - Cholangiocarcinoma
 - incidence may be as high as 75%
 - Higher in types IV and V
 - Squamous cell carcinoma
 - Anaplastic carcinoma
 - Others

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BACKGROUND

Treatment

- Surgical resection with Roux – Y hepaticojejunostomy
- Partial hepatectomy for segmental intra-hepatic involvement
- Liver transplant for diffuse intra-hepatic involvement
- “Wait and see” for Type III choledochal cysts (duodenal epithelial lining does not predispose to biliary malignancy)

3. Imaging findings OR Procedure details

Imaging Findings

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IMAGING FINDINGS

- Choledochal cysts are characterized by biliary tree dilatation
- There are five subtypes of choledochal cysts, as defined by Todani's modification of the Alonso – Lej classification

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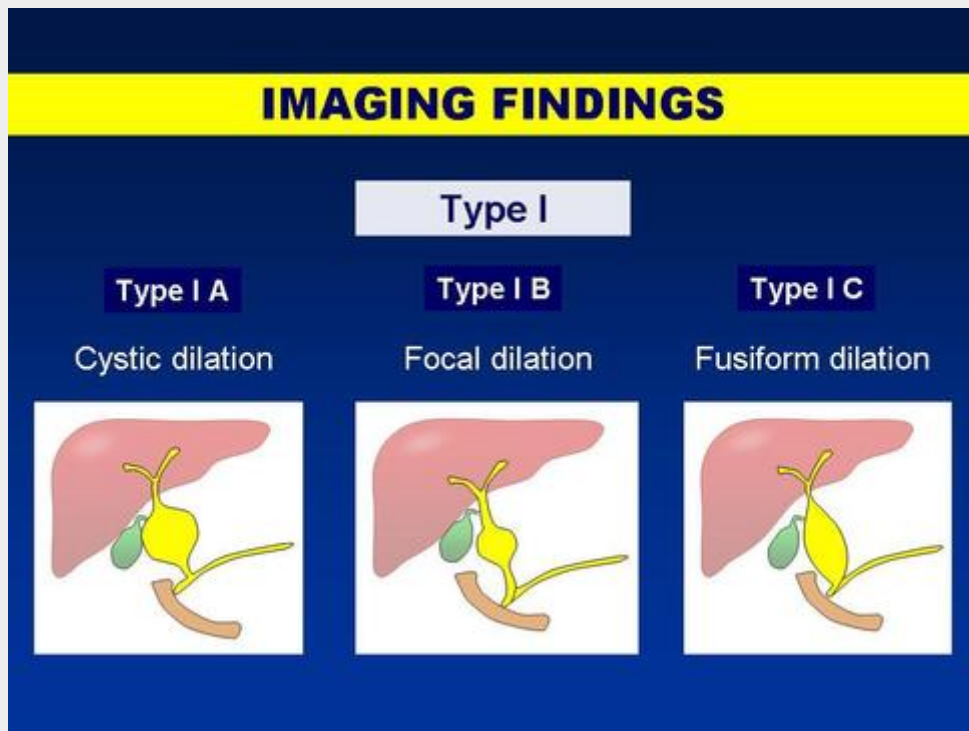
IMAGING FINDINGS

Classification

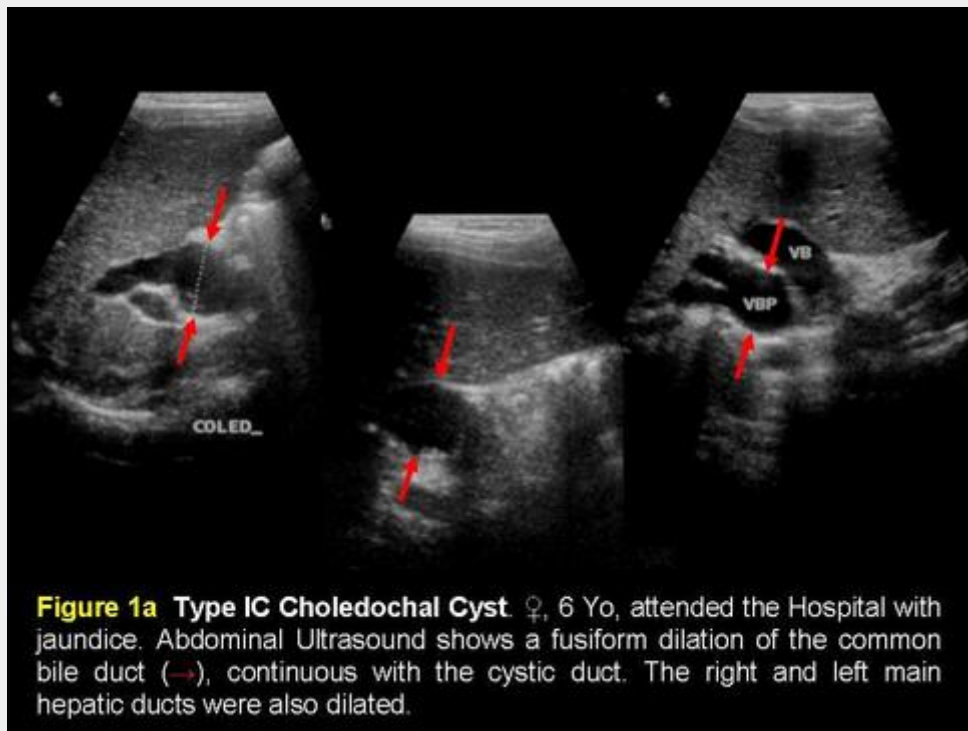
Todani Modification of the Alonso – Lej Classification

- | | |
|-----------------|-------------------------------------|
| 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 |

Type I



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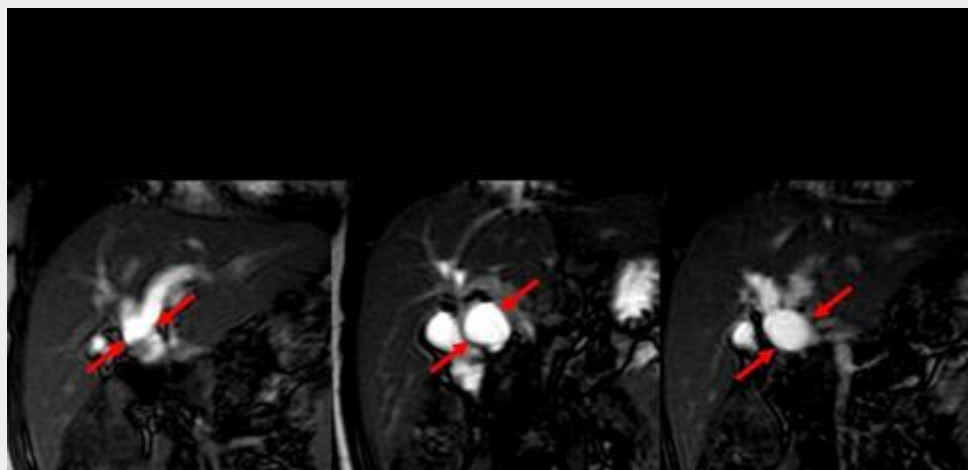


Figure 1b Type IC Choledochal Cyst. Magnetic Resonance SSFP sequences show a fusiform dilation of the common hepatic and common bile ducts. The cystic duct drains into the dilated extrahepatic bile duct.

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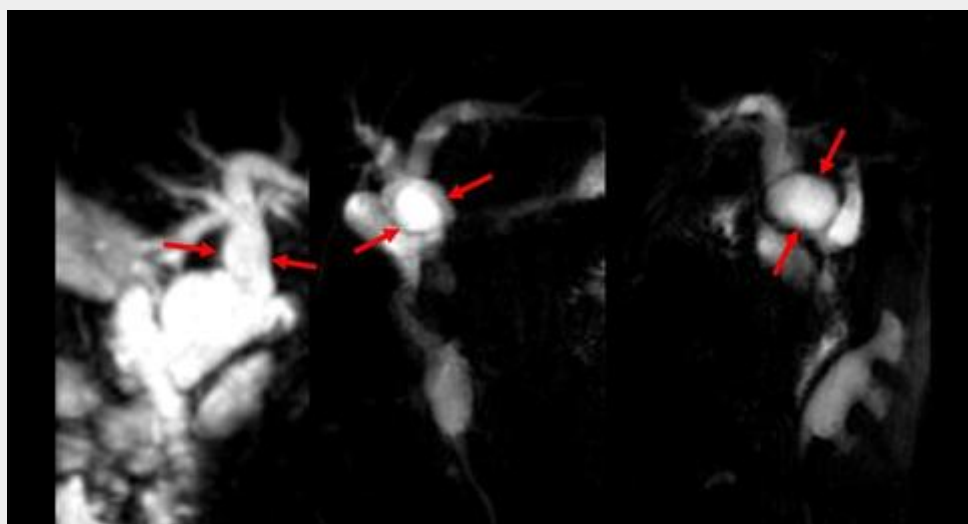


Figure 1c Type IC Choledochal Cyst. MR T2 reformatted images show a fusiform dilation of the common hepatic and common bile ducts. The cystic duct drains to the dilated extrahepatic bile duct. These findings correspond to a Type IC choledochal cyst, according to the Todani classification.

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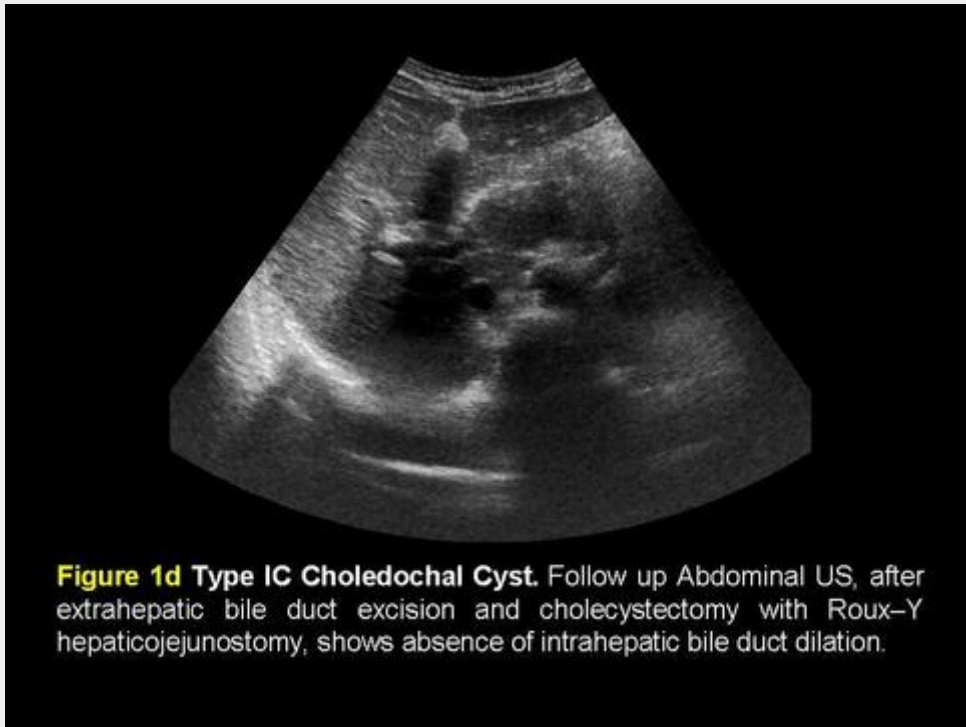


Figure 1d Type IC Choledochal Cyst. Follow up Abdominal US, after extrahepatic bile duct excision and cholecystectomy with Roux-Y hepaticojejunostomy, shows absence of intrahepatic bile duct dilation.

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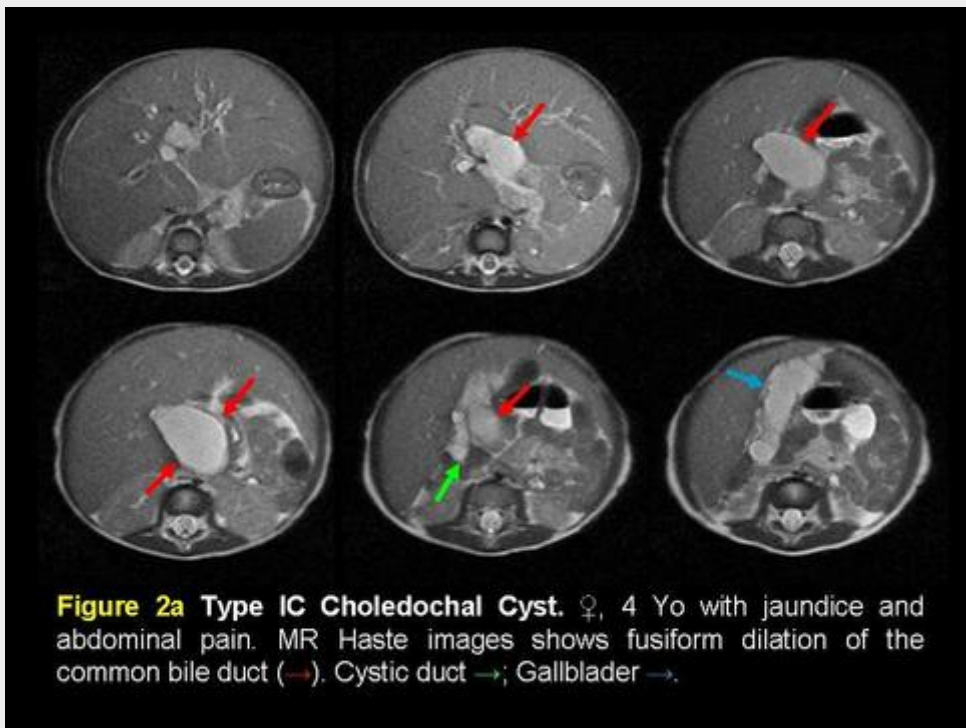
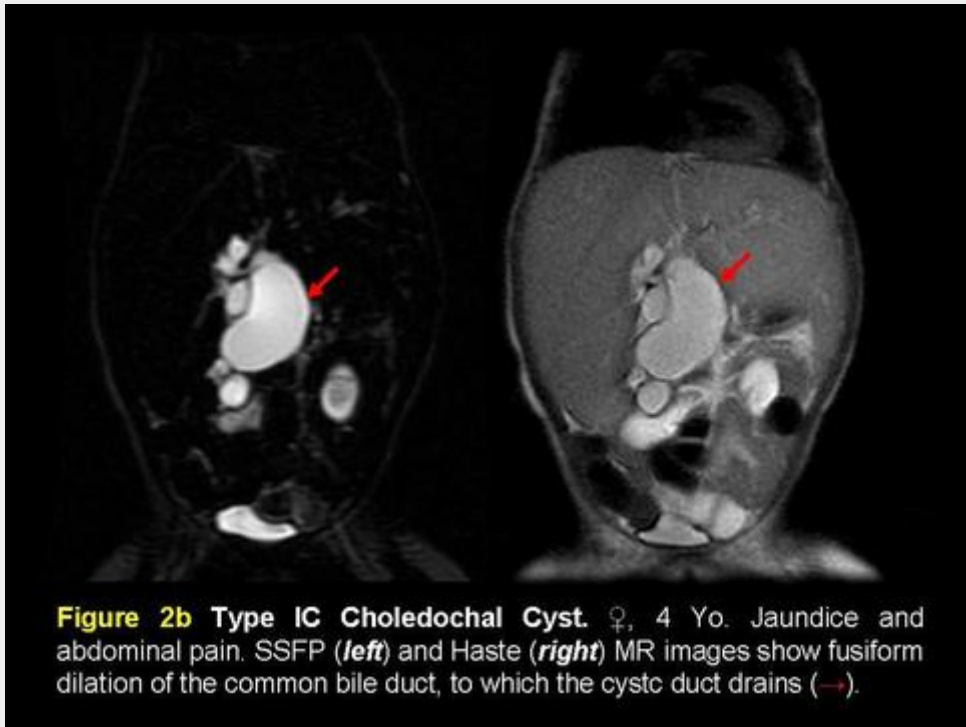


Figure 2a Type IC Choledochal Cyst. ♀, 4 Yo with jaundice and abdominal pain. MR Haste images shows fusiform dilation of the common bile duct (→). Cystic duct →; Gallblader →.

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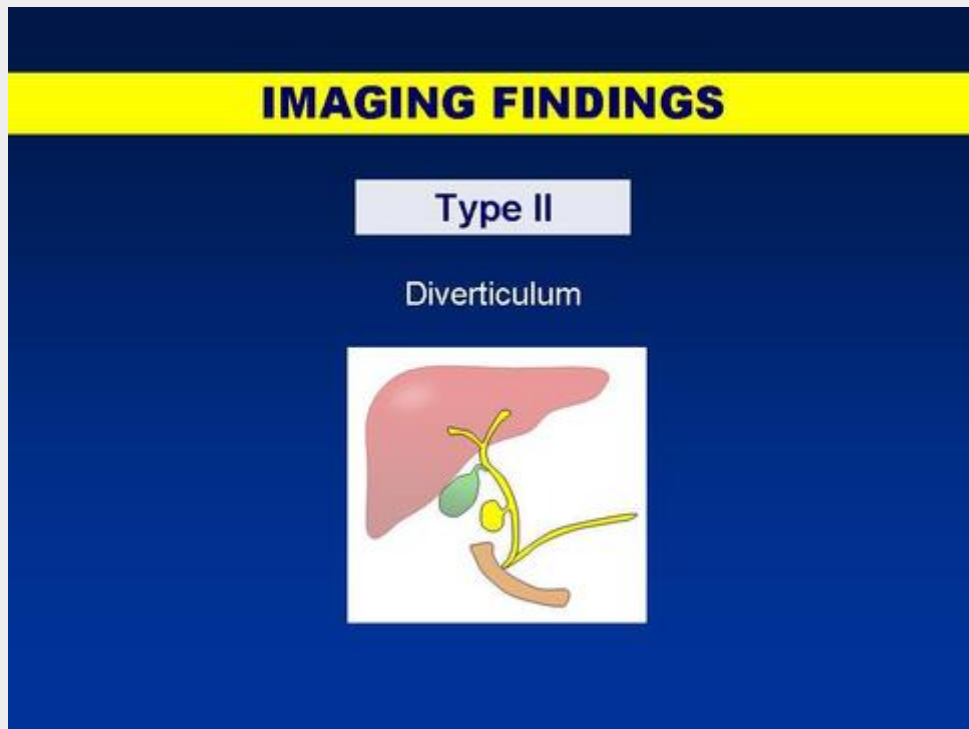
Type IC



[\[Type IC Choledochal Cyst\] Video 1](#) shows a Type IC Choledochal Cyst, as seen on MR Heavily

T2-weighted reformatted images.

Type II



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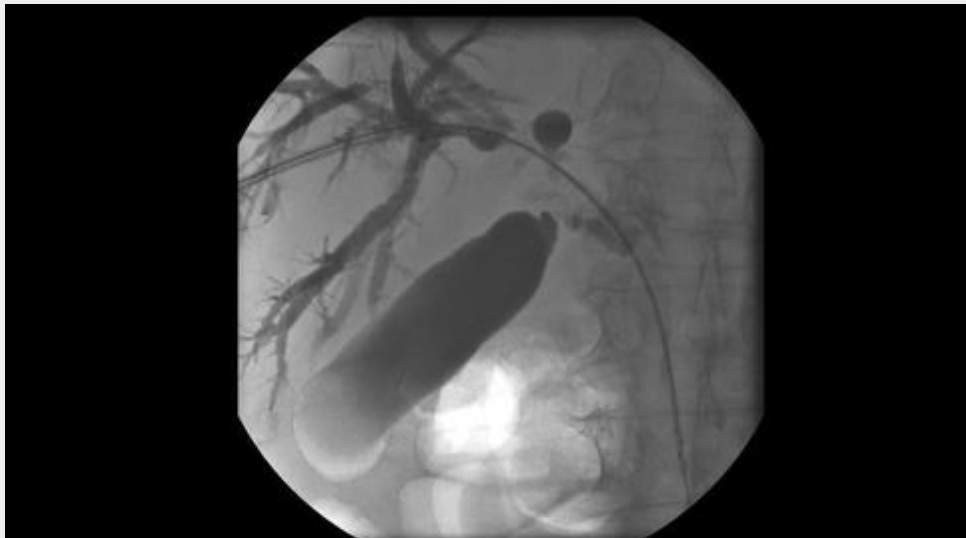
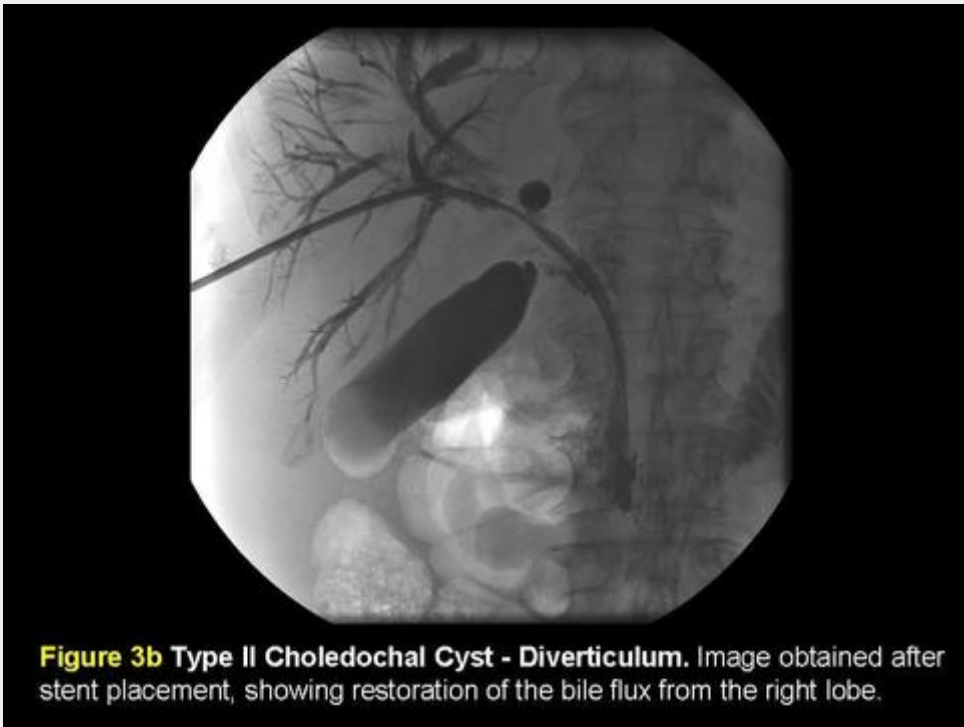


Figure 3a Type II Choledochal Cyst - Diverticulum. ♂, 76 Yo. Cholangiocarcinoma – Klatskin tumor. Percutaneous transhepatic cholangiography (PTC) performed to palliate biliary obstruction showed a diverticulum of the common hepatic duct, located above the insertion of the cystic duct, draining to an area of neoplastic involvement.

Diverticulum

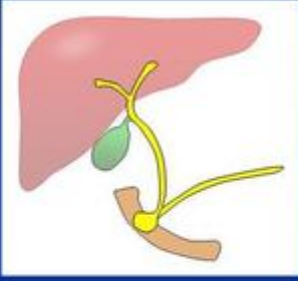


Type III

IMAGING FINDINGS

Type III

Choledochoceles



Choledochocele

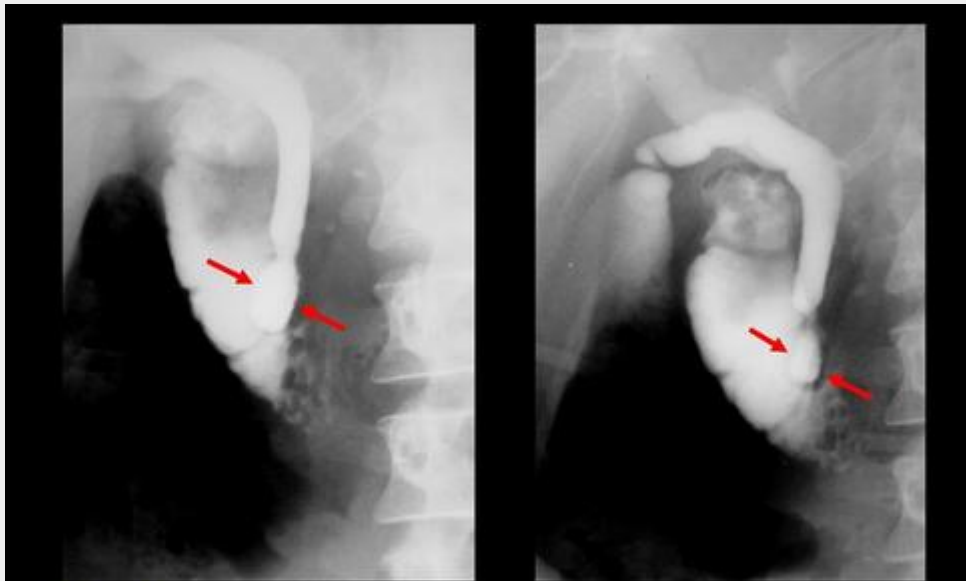


Figure 3 Type III Choledochal Cyst – Choledochocele. PTC shows a diverticulum of the common bile duct located within the 2nd portion of the duodenum's wall, adjacent to the papilla of Vater (→).

Type IV

IMAGING FINDINGS

Type IV

Type IV A

Multiple intra and extrahepatic cysts



Type IV B

Multiple extrahepatic cysts



Type IVA

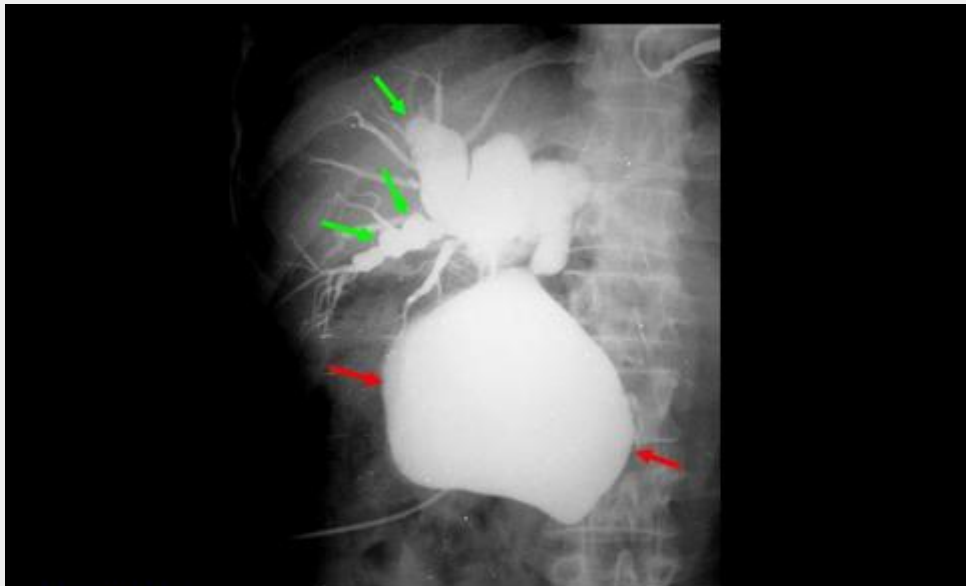


Figure 4 Type IVA choledochal cyst as seen on PTC. Massive dilation of the common bile duct (→) and multiple intrahepatic bile duct dilations are apparent (→).

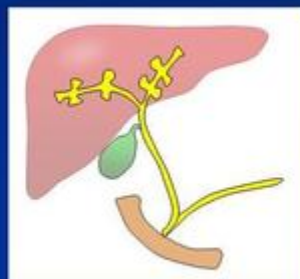
Type V

IMAGING FINDINGS

Type V

Caroli's Disease

Multiple intrahepatic cysts



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IMAGING FINDINGS

Intrahepatic Bile Duct Dilatation

- Autosomic recessive inheritance
- Multifocal segmental dilatation of intrahepatic bile ducts retaining communication with the biliary tree
- 2 types:
 - Caroli disease (pure form)
 - IHBD dilatations
 - Caroli syndrome
 - IHBD dilatations
 - Congenital hepatic fibrosis

diapositivo31.jpg

IMAGING FINDINGS

Caroli's Disease

Pathogenesis

- Neonatal occlusion of the hepatic artery, leading to bile duct ischemia and cystic dilatation
- Abnormal growth rate of the developing biliary epithelium and supporting connective tissue
- Lack of normal involution of ductal plates that surround the portal tracts, resulting in epithelium-lined cysts that surround the portal triads

diapositivo32.jpg

IMAGING FINDINGS

Caroli's Disease

Associated ductal plate abnormalities

- Congenital hepatic fibrosis
- Polycystic renal diseases
 - Medullary sponge kidney
 - ARPKD
 - Nephronoptosis

diapositivo33.jpg

IMAGING FINDINGS

Caroli's Disease

Differential Diagnosis

- Polycystic liver disease
- Biliary microhamartomas
- Primary sclerosing cholangitis
- Recurrent pyogenic cholangitis
(oriental cholangiohepatitis)

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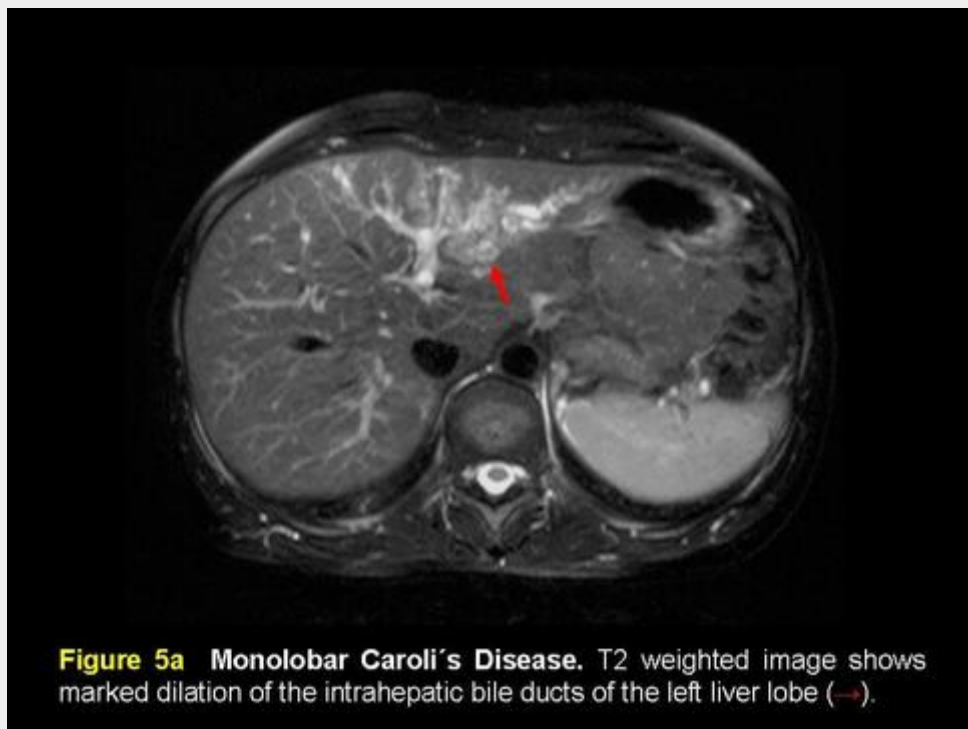


Figure 5a Monolobar Caroli's Disease. T2 weighted image shows marked dilation of the intrahepatic bile ducts of the left liver lobe (→).

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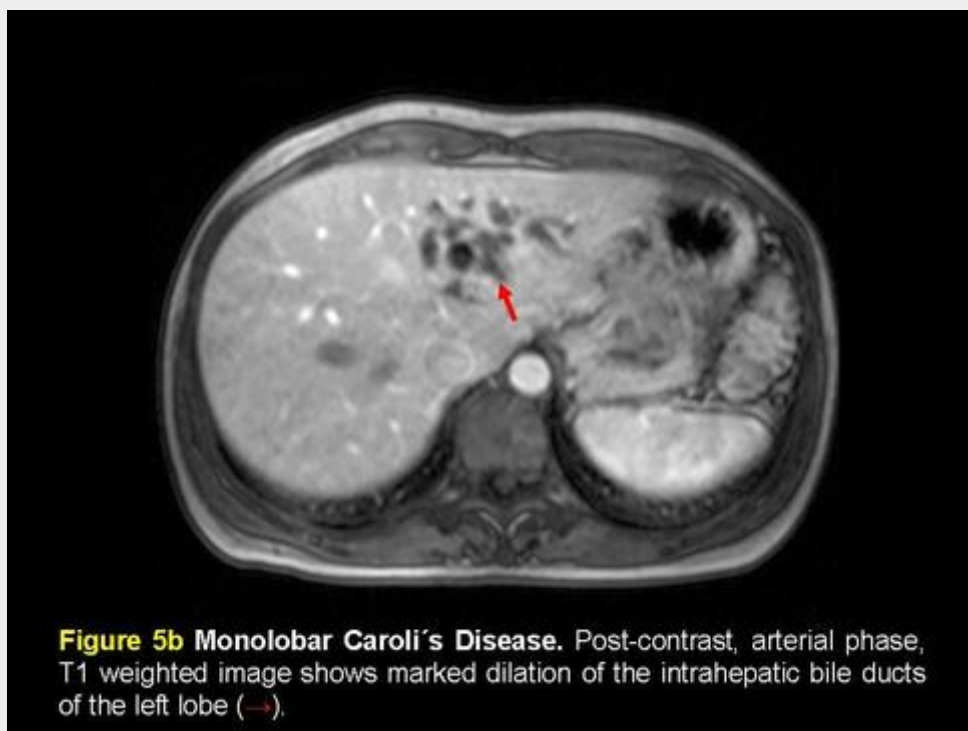


Figure 5b Monolobar Caroli's Disease. Post-contrast, arterial phase, T1 weighted image shows marked dilation of the intrahepatic bile ducts of the left lobe (→).

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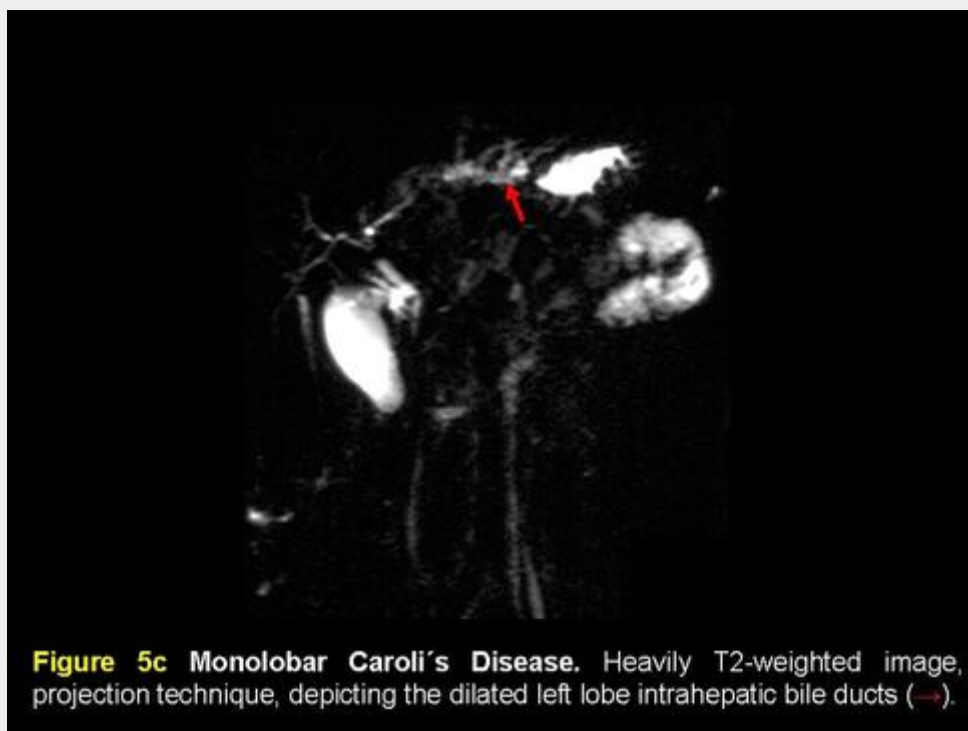


Figure 5c Monolobar Caroli's Disease. Heavily T2-weighted image, projection technique, depicting the dilated left lobe intrahepatic bile ducts (→).

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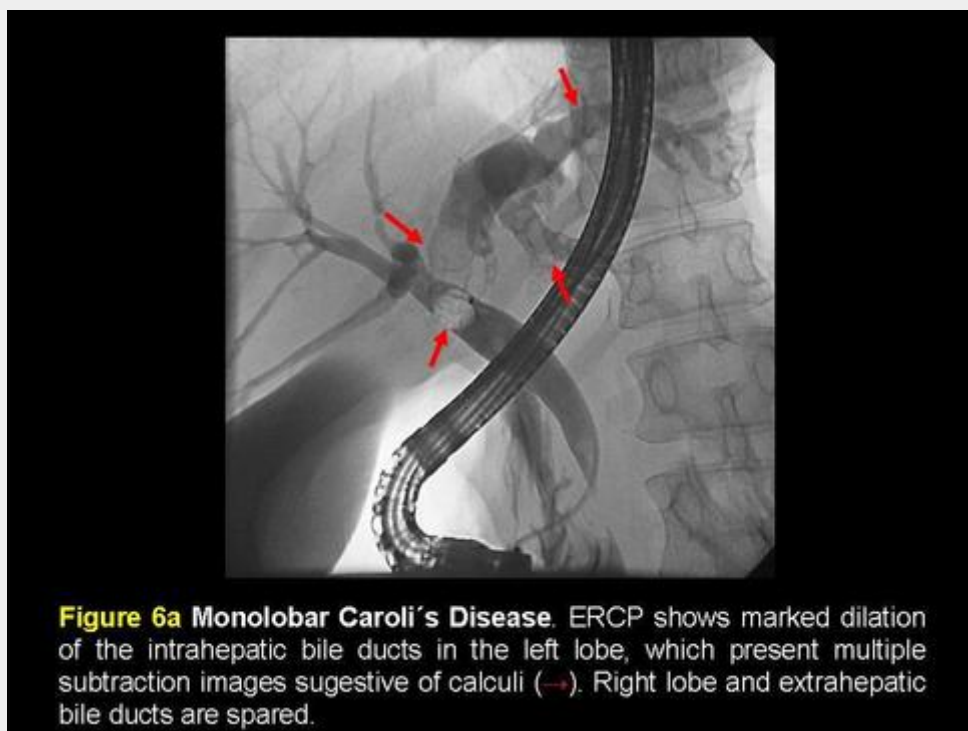


Figure 6a Monolobar Caroli's Disease. ERCP shows marked dilation of the intrahepatic bile ducts in the left lobe, which present multiple subtraction images suggestive of calculi (→). Right lobe and extrahepatic bile ducts are spared.

Caroli's Syndrome

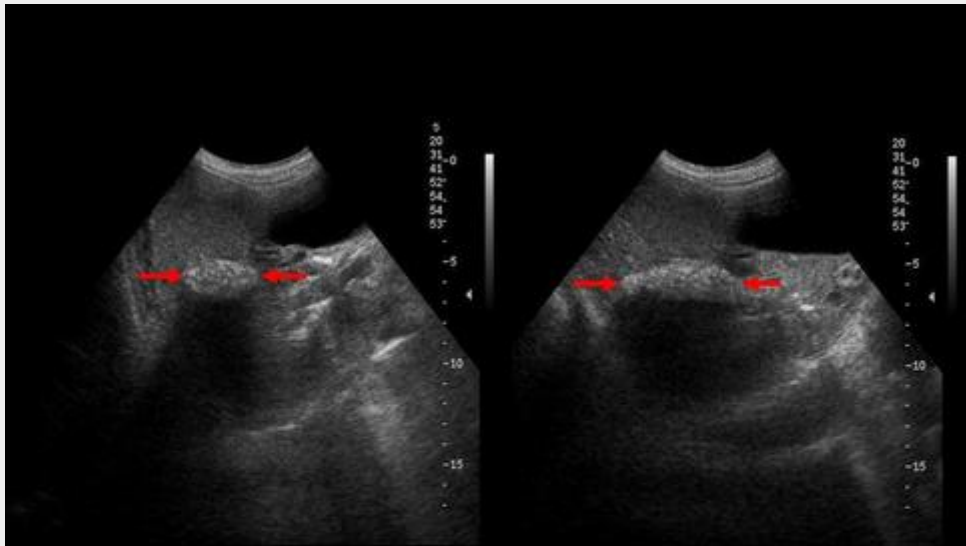


Figure 7a Caroli's Syndrome. Abdominal Ultrasound shows an oval hyperechogenic image with posterior acoustic shadowing in the right liver lobe (→), which corresponds to a markedly dilated intrahepatic bile duct, completely filled with calculi.

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Figure 7b Caroli's Syndrome. Abdominal Ultrasound also shows ectatic tortuous veins in the gallbladder's bed (→) representing varicosities due to portal hypertension. → calculi within dilated intrahepatic duct.

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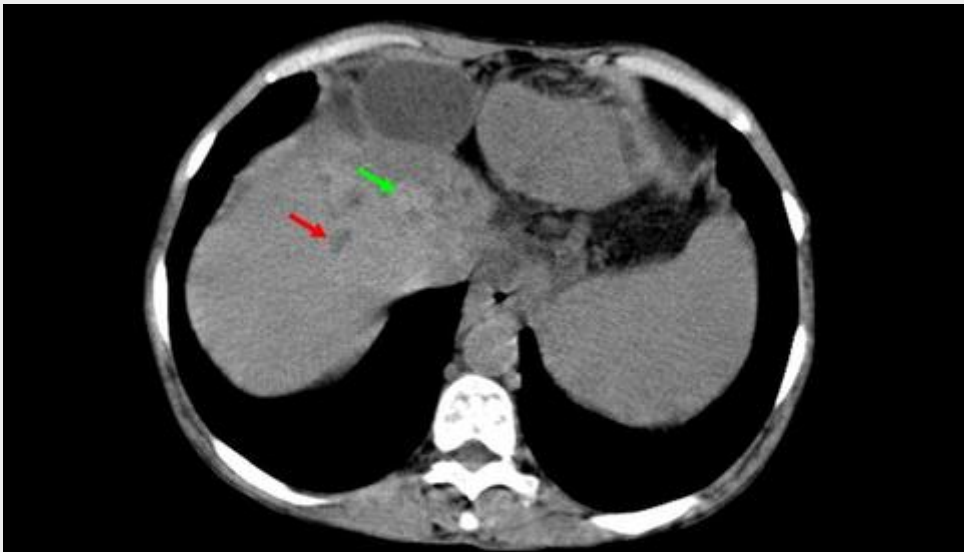


Figure 7c Caroli's Syndrome. Pre-contrast abdominal CT. There are dilated bile ducts in the left lobe (→), some of which filled with hyperdense material, corresponding to bile calculi (→).

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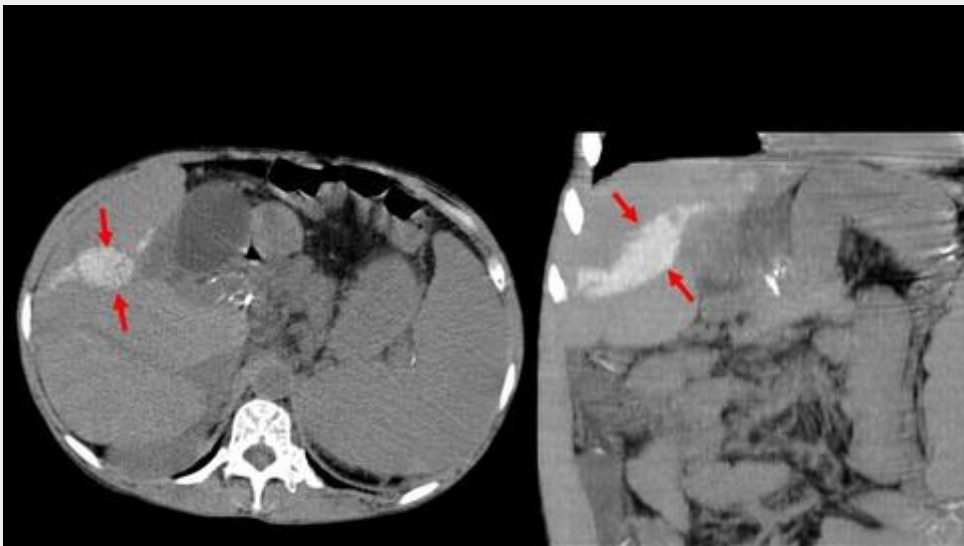


Figure 7d Caroli's Syndrome. Pre-contrast abdominal CT. Axial (*left*) and oblique reformatted (*right*) images. There is a markedly dilated bile duct in the right lobe, completely filled with hyperdense material, corresponding to bile calculi (→).

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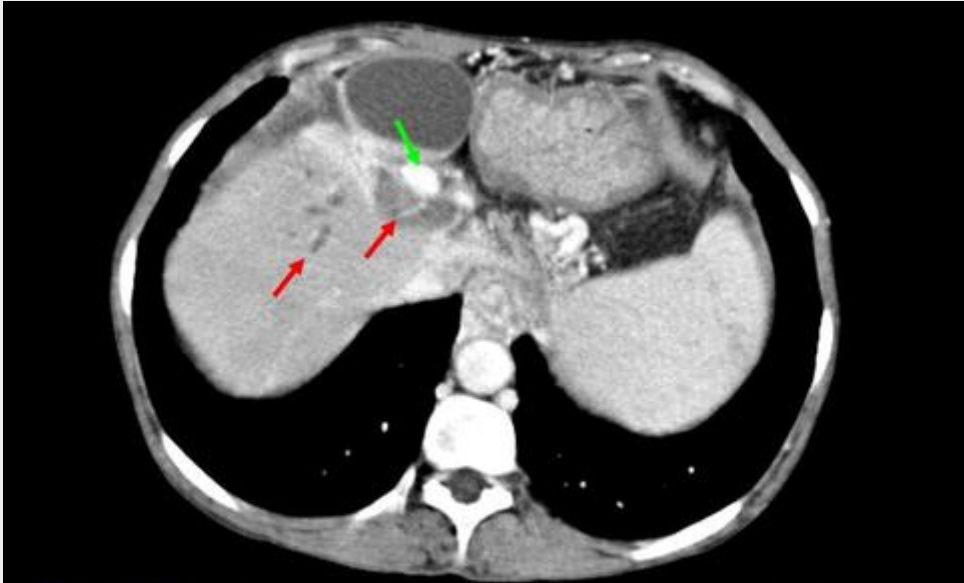


Figure 7e Caroli's Syndrome. Post-contrast abdominal CT better depicting the dilated bile ducts in the left lobe (→) surrounding a dilated patent umbilical vein (→) due to coexistent portal hypertension.

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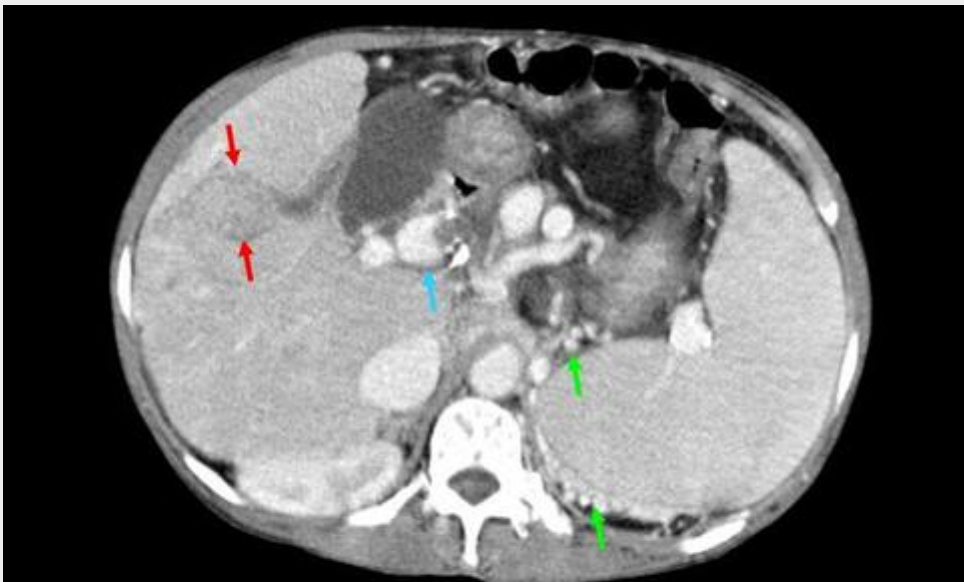


Figure 7f Caroli's Syndrome. Post-contrast abdominal CT depicting the dilated intra-hepatic bile duct filled with calculi (→). Note the splenomegaly, dilation of the portal vein (→) and perisplenic varicosities (→).

Caroli's Syndrome

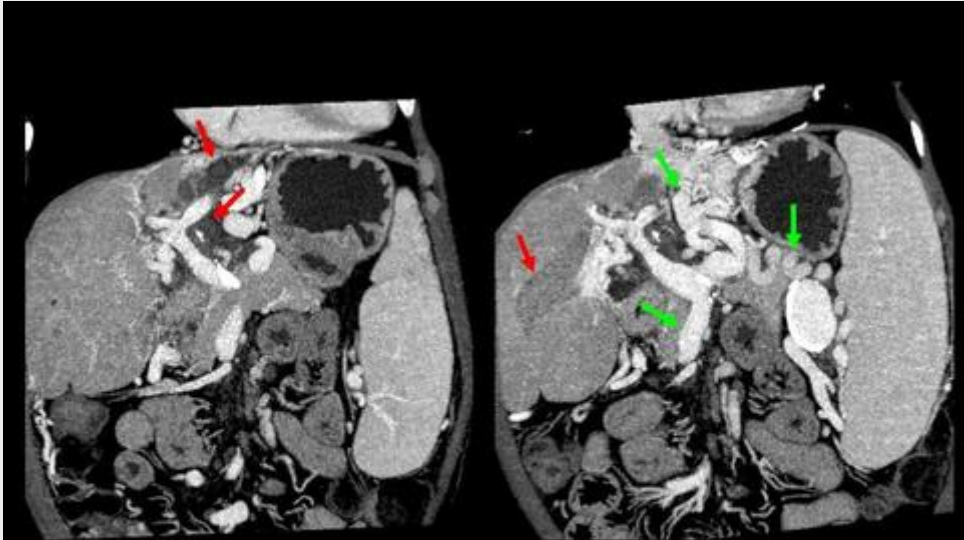


Figure 7g Caroli's Syndrome. Post-contrast abdominal CT. Reformed oblique MIP images depicting the dilated intra-hepatic bile ducts, some of which filled with calculi (→). Note the splenomegaly and the tortuosity and dilation of the portal, splenic and perigastric veins (→) due to portal hypertension.

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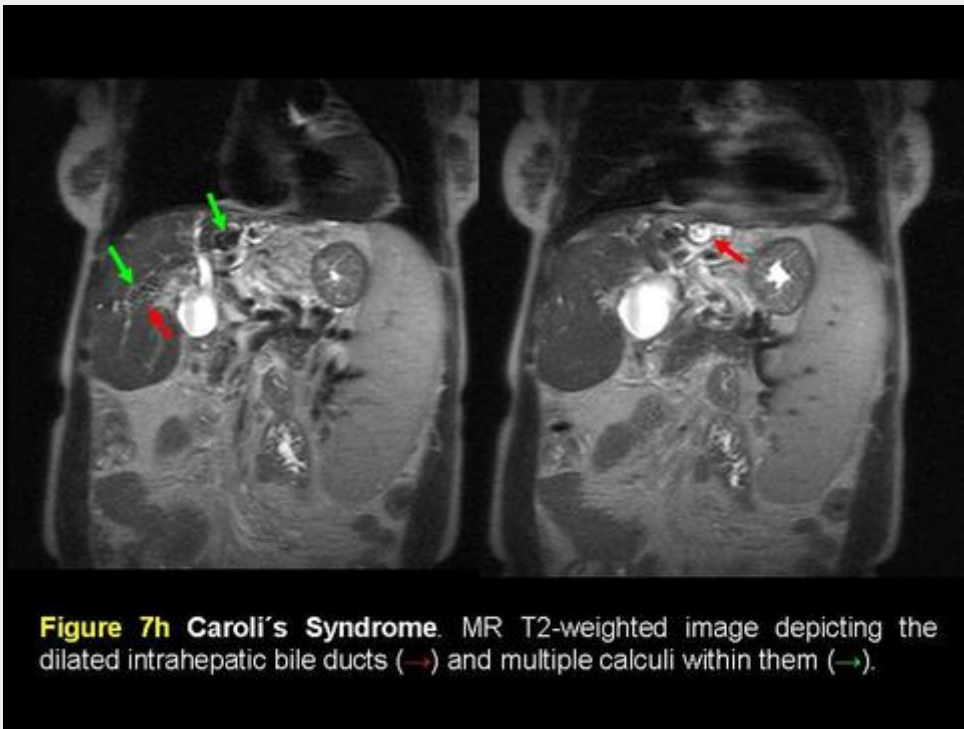


Figure 7h Caroli's Syndrome. MR T2-weighted image depicting the dilated intrahepatic bile ducts (→) and multiple calculi within them (→).

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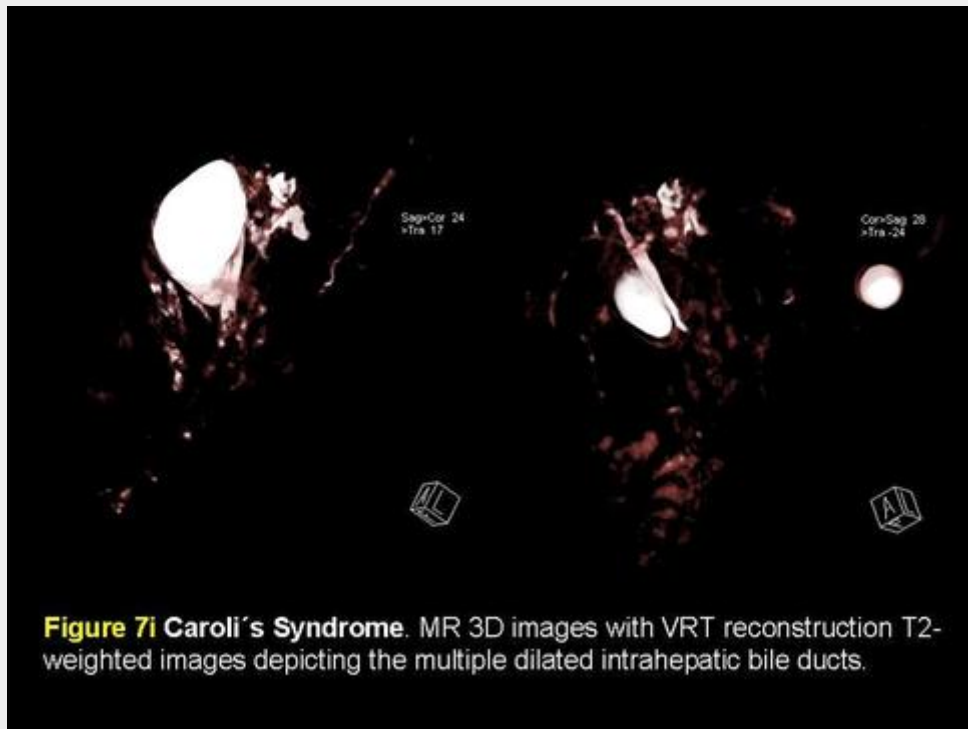


Figure 7i Caroli's Syndrome. MR 3D images with VRT reconstruction T2-weighted images depicting the multiple dilated intrahepatic bile ducts.

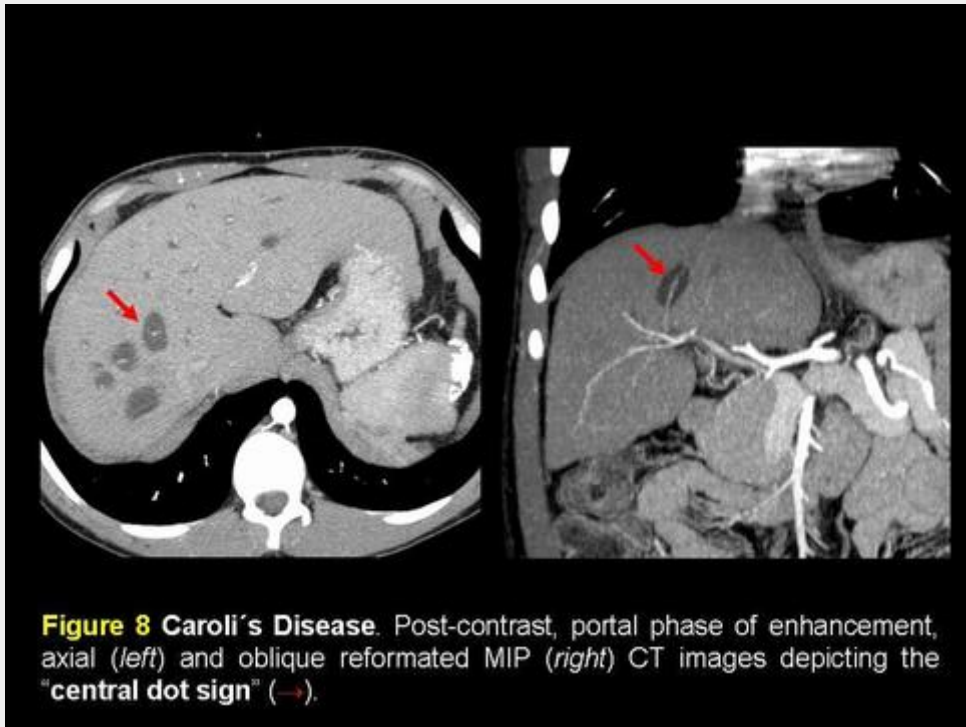
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IMAGING FINDINGS

Caroli's Disease

- Central Dot Sign
 - Solid "dot" within or at the periphery of a cystic liver lesion
 - Doppler signal
 - Continuous (portal vein branch)
 - Arterial waveform (hepatic artery branch)
 - Enhancement
 - CT, MRI

diapositivo51.jpg



diapositivo52.jpg

IMAGING FINDINGS

Caroli's Disease

- **Complications**
 - Cholangitis, stones
 - Strictures
 - Cholangiocarcinoma (7-14%)
- **Caroli's syndrome:**
 - Portal hypertension
 - Secondary biliary cirrhosis

4. Conclusion

Conclusion

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CONCLUSIONS

Choledochal cysts are uncommon entities easily depicted by the imaging modalities presented. Their recognition is very important because early intervention may avoid many of the possible unwanted complications.

5. References

References

diapositivo55.jpg

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Special thanks

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Nuno Neves, MD
Luís Semedo, MD

7. Mediafiles

Author information

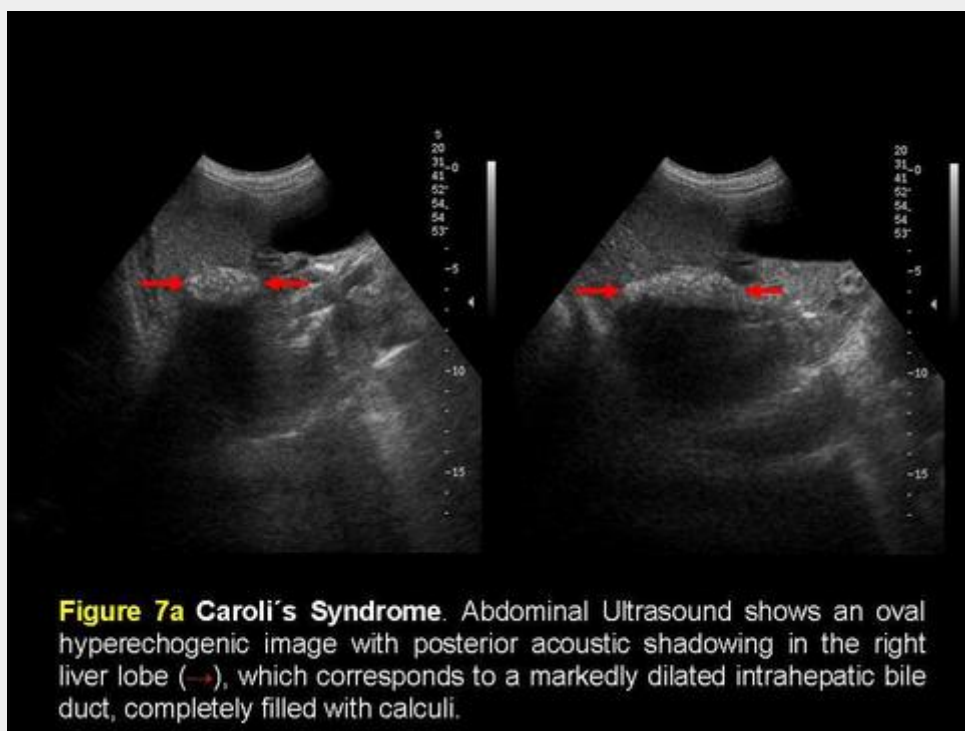
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Caroli's Syndrome



Caroli's Syndrome

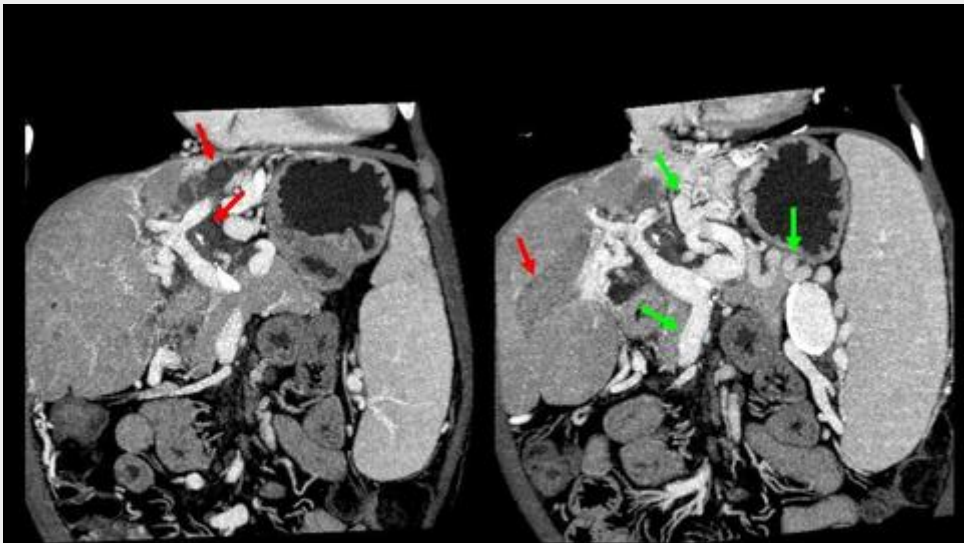


Figure 7g Caroli's Syndrome. Post-contrast abdominal CT. Reformatted oblique MIP images depicting the dilated intra-hepatic bile ducts, some of which filled with calculi (→). Note the splenomegaly and the tortuosity and dilation of the portal, splenic and perigastric veins (→) due to portal hypertension.

Choledochocele

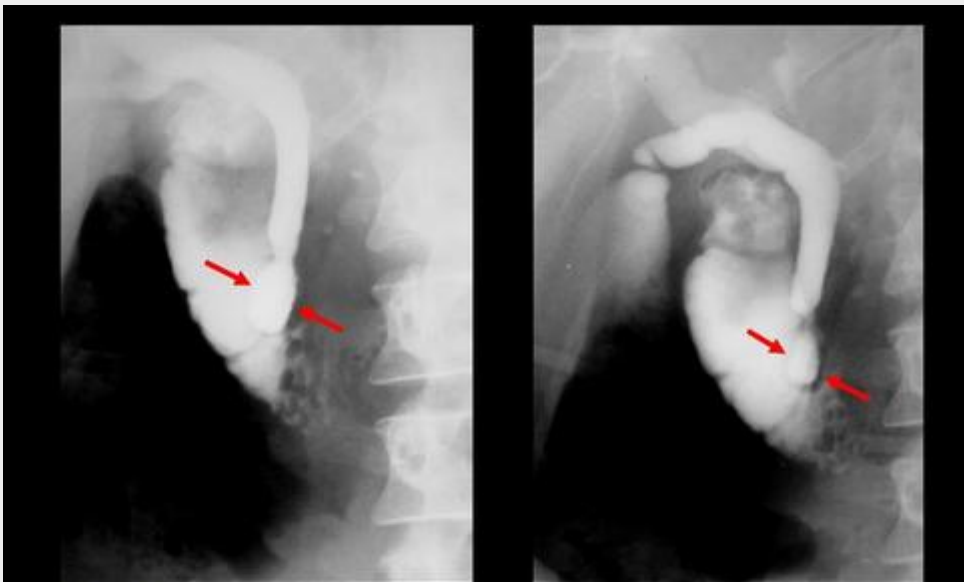


Figure 3 Type III Choledochal Cyst – Choledochocele. PTC shows a diverticulum of the common bile duct located within the 2nd portion of the duodenum's wall, adjacent to the papilla of Vater (→).

Definition of choledochal cysts

BACKGROUND

Definition

- Uncommon anomalies of the biliary system manifested by cystic dilatation of the extra and/or intrahepatic biliary tree

Diverticulum

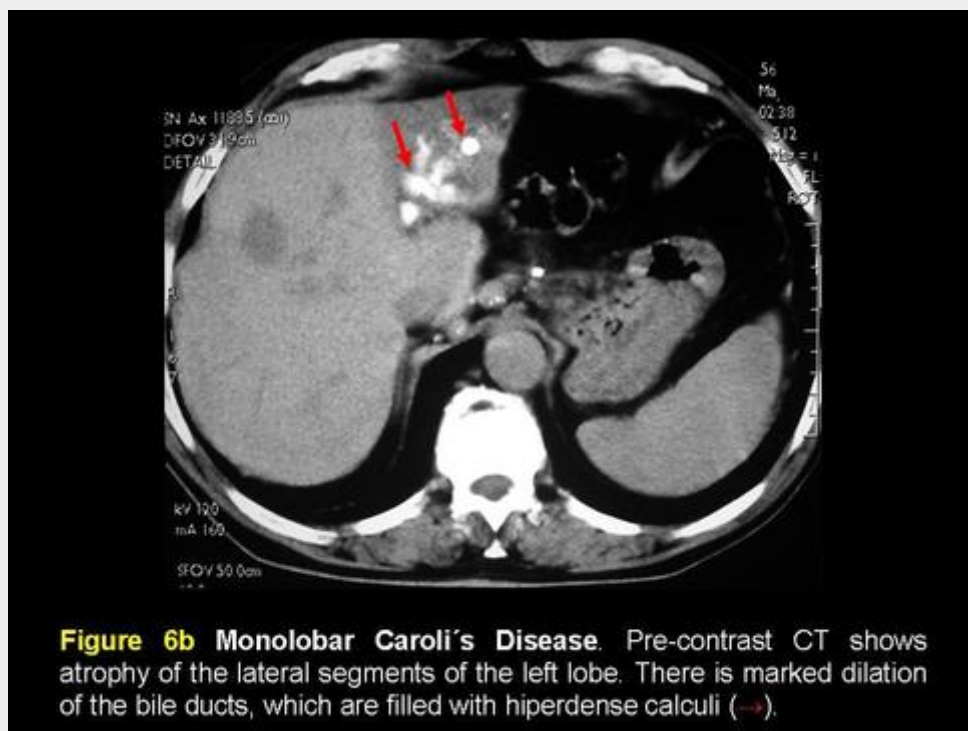


Learning objectives

LEARNING OBJECTIVES

To present the imaging findings of choledochal cysts, as seen on US, CT, MRCP and PTC.

Monolobar Caroli's Disease



Special thanks

SPECIAL THANKS

Nuno Neves, MD
Luís Semedo, MD

Type I

IMAGING FINDINGS

Type I

Type I A

Cystic dilation



Type I B

Focal dilation



Type I C

Fusiform dilation



Type IC



Figure 2c Type IC Choledochal Cyst. ♀, 4 Yo with jaundice and abdominal pain. MR Heavily T2-weighted reformatted images.

Type IC Choledochal Cyst

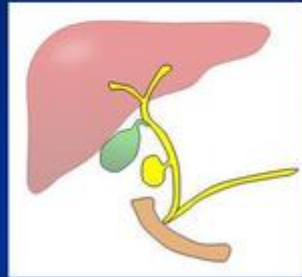


Type II

IMAGING FINDINGS

Type II

Diverticulum



Type III

IMAGING FINDINGS

Type III

Choledochocele



Type IV

IMAGING FINDINGS

Type IV

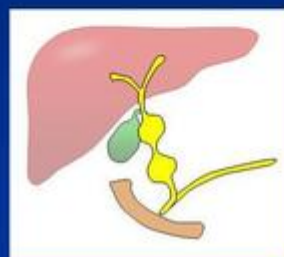
Type IV A

Multiple intra and extrahepatic cysts



Type IV B

Multiple extrahepatic cysts



Type IVA

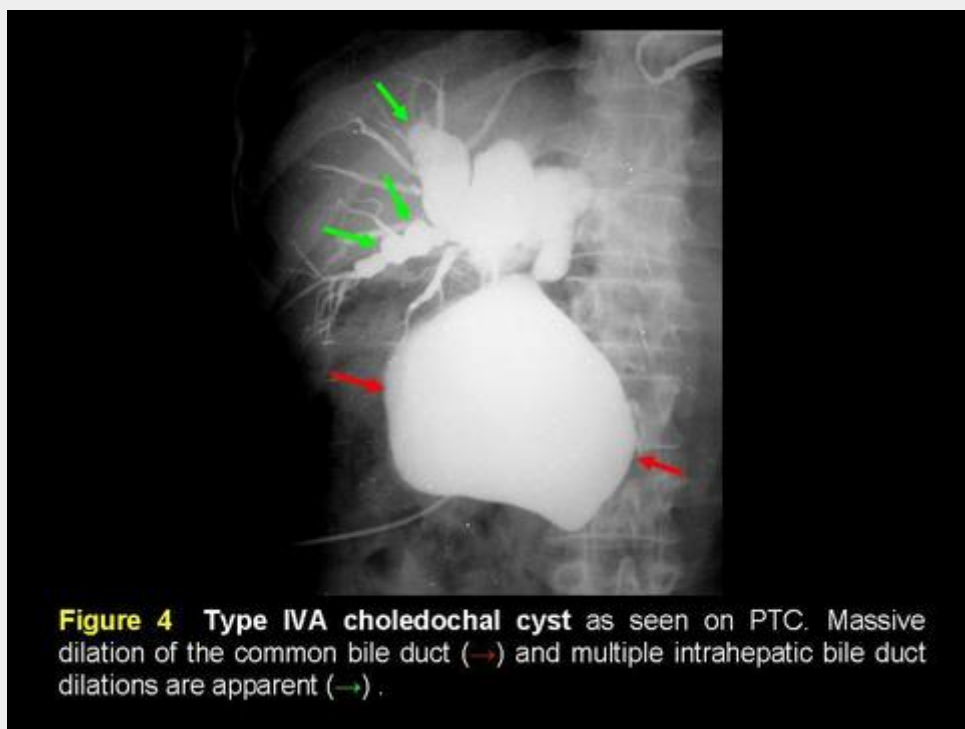


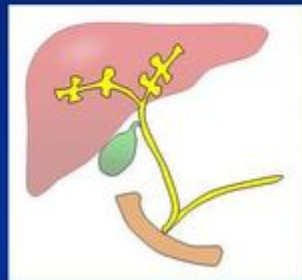
Figure 4 Type IVA choledochal cyst as seen on PTC. Massive dilation of the common bile duct (→) and multiple intrahepatic bile duct dilations are apparent (→).

Type V

IMAGING FINDINGS

Type V

Caroli's Disease
Multiple intrahepatic cysts



diapositivo4.jpg

BACKGROUND

Origin

- Pancreatobiliary junction anomalies may promote reflux of pancreatic juice into the common bile duct, resulting in

inflammation



weakening of the bile duct wall



dilation

diapositivo5.jpg

BACKGROUND

Origin

Some speculate that the reflux may also happen the other way around – bile into the Wirsung channel -, predisposing to pancreatitis, which has a relatively high incidence in patients with choledocal cyst disease

diapositivo6.jpg

BACKGROUND

Origin

Other proposed mechanisms are:

- inherited/genetic factors
- infection
- congenital weakness in the walls of the biliary tract
- dysfunction of the sphincter of Oddi
- distal obstruction

diapositivo7.jpg

BACKGROUND

Epidemiology

- Estimated incidence:
 - 1/100000 in western countries
 - 1/1000 in Asia
- Higher prevalence in East Asia, particularly Japan
- Higher incidence in children – 60% in the 1st decade of life
- Higher incidence in ♀ - 80%
- 20% diagnosed in adults

diapositivo8.jpg

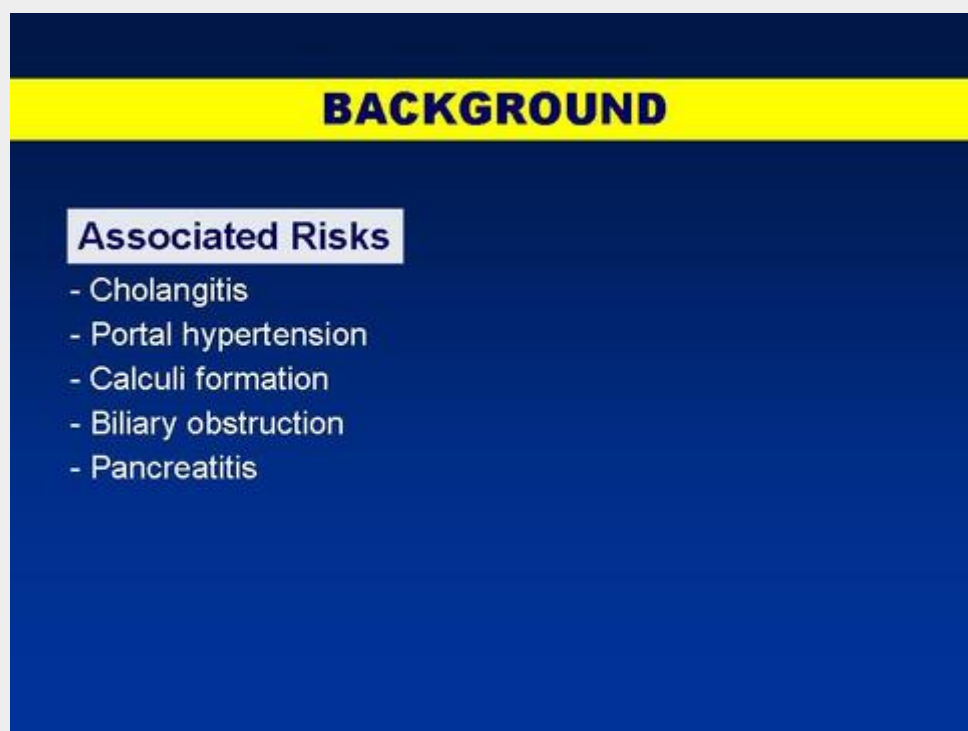
BACKGROUND

Clinical Findings

- Classic presentation in a child:
 - jaundice
 - right upper quadrant pain
 - palpable right upper quadrant mass

} 33%
- Presentation in adults:
 - right upper quadrant pain
 - pancreatitis
 - jaundice

diapositivo9.jpg

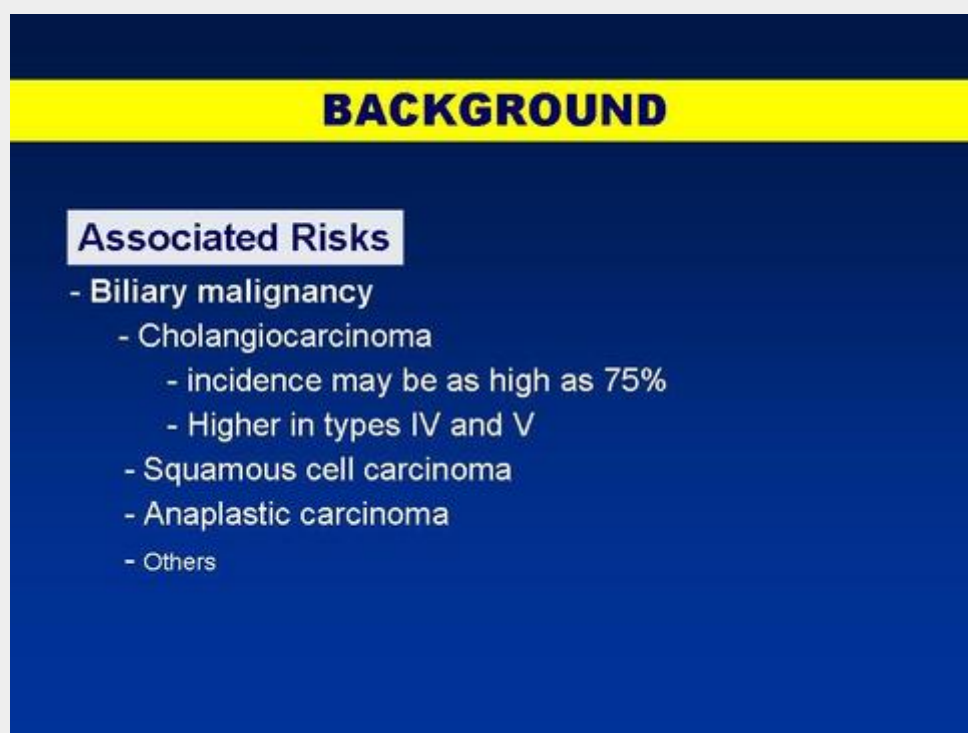


BACKGROUND

Associated Risks

- Cholangitis
- Portal hypertension
- Calculi formation
- Biliary obstruction
- Pancreatitis

diapositivo10.jpg



BACKGROUND

Associated Risks

- Biliary malignancy
 - Cholangiocarcinoma
 - incidence may be as high as 75%
 - Higher in types IV and V
 - Squamous cell carcinoma
 - Anaplastic carcinoma
 - Others

diapositivo11.jpg

BACKGROUND

Treatment

- Surgical resection with Roux – Y hepaticojejunostomy
- Partial hepatectomy for segmental intra-hepatic involvement
- Liver transplant for diffuse intra-hepatic involvement
- “Wait and see” for Type III choledochal cysts (duodenal epithelial lining does not predispose to biliary malignancy)

diapositivo12.jpg

IMAGING FINDINGS

- Choledochal cysts are characterized by biliary tree dilatation
- There are five subtypes of choledochal cysts, as defined by Todani’s modification of the Alonso – Lej classification

diapositivo13.jpg

IMAGING FINDINGS

Classification

Todani 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

diapositivo15.jpg

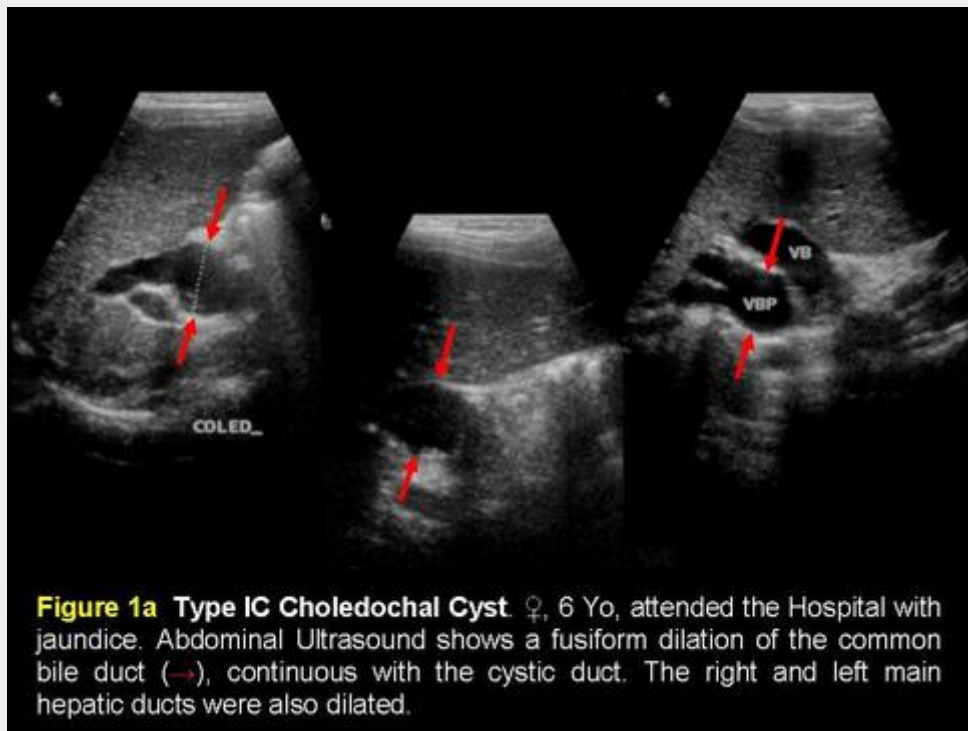


Figure 1a Type IC Choledochal Cyst. ♀, 6 Yo, attended the Hospital with jaundice. Abdominal Ultrasound shows a fusiform dilation of the common bile duct (→), continuous with the cystic duct. The right and left main hepatic ducts were also dilated.

diapositivo16.jpg

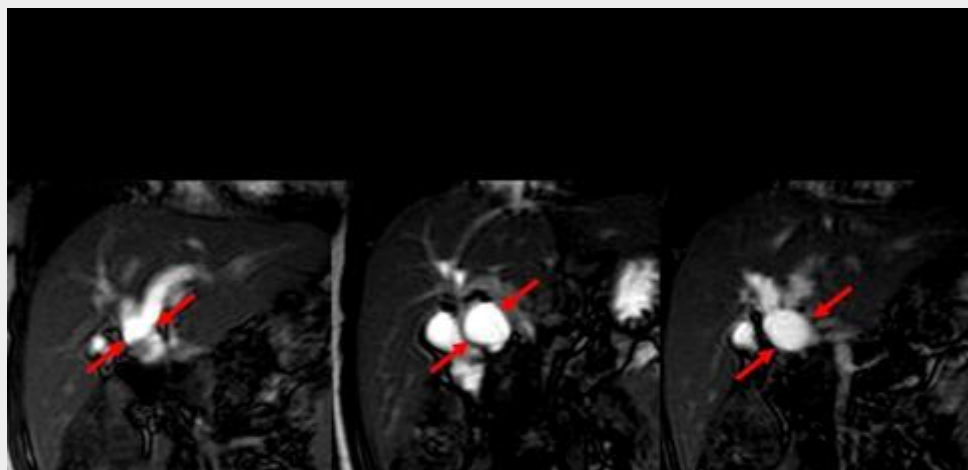


Figure 1b Type IC Choledochal Cyst. Magnetic Resonance SSFP sequences show a fusiform dilation of the common hepatic and common bile ducts. The cystic duct drains into the dilated extrahepatic bile duct.

diapositivo17.jpg

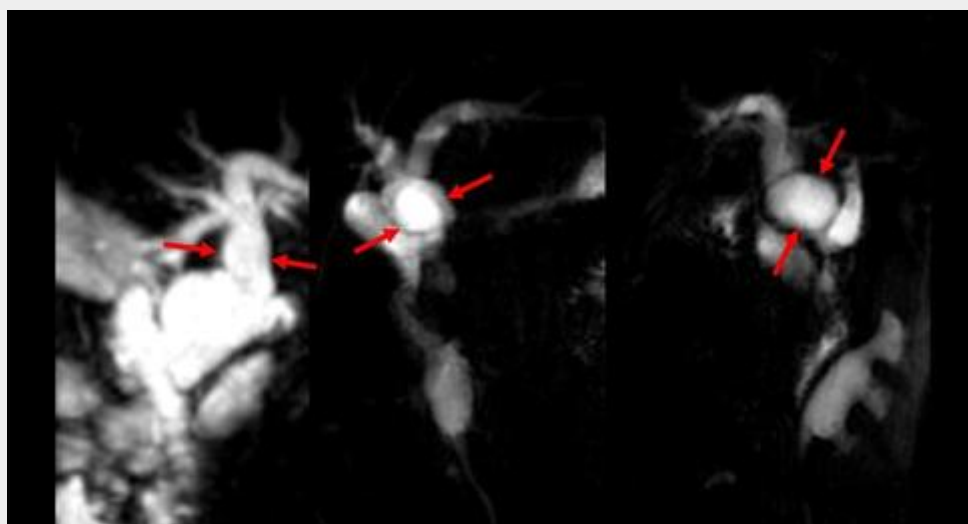


Figure 1c Type IC Choledochal Cyst. MR T2 reformatted images show a fusiform dilation of the common hepatic and common bile ducts. The cystic duct drains to the dilated extrahepatic bile duct. These findings correspond to a Type IC choledochal cyst, according to the Todani classification.

diapositivo18.jpg

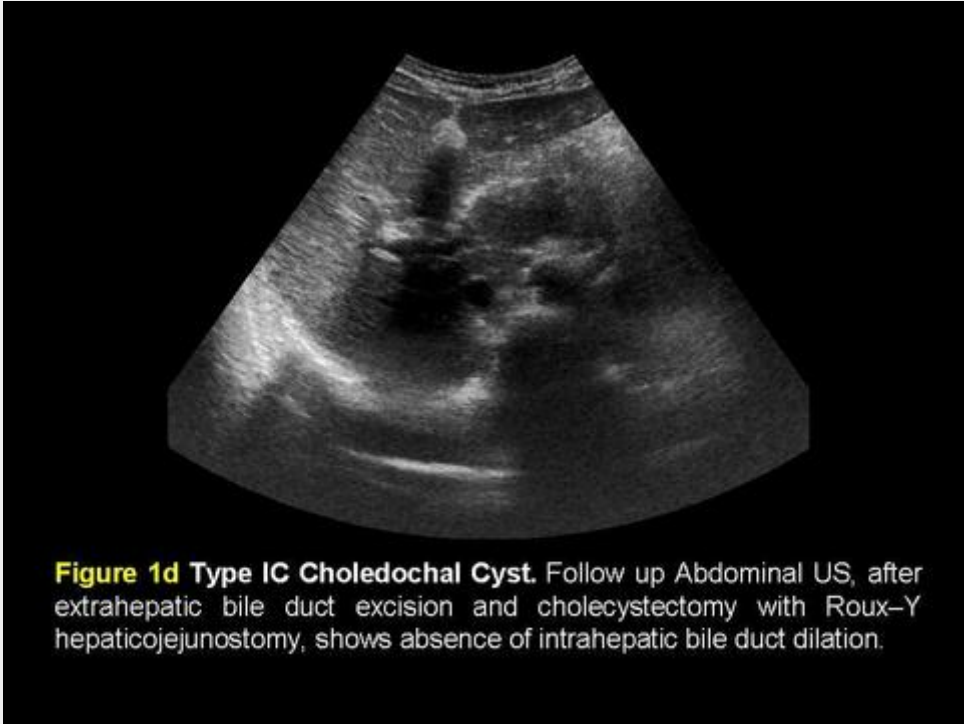


Figure 1d Type IC Choledochal Cyst. Follow up Abdominal US, after extrahepatic bile duct excision and cholecystectomy with Roux-Y hepaticojejunostomy, shows absence of intrahepatic bile duct dilation.

diapositivo19.jpg

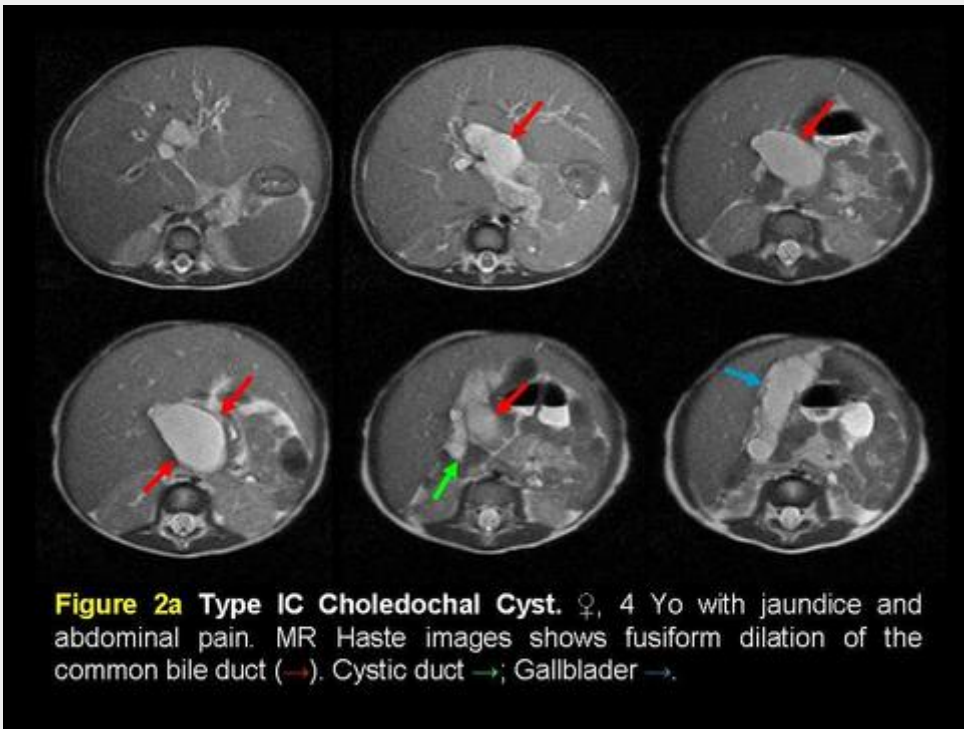
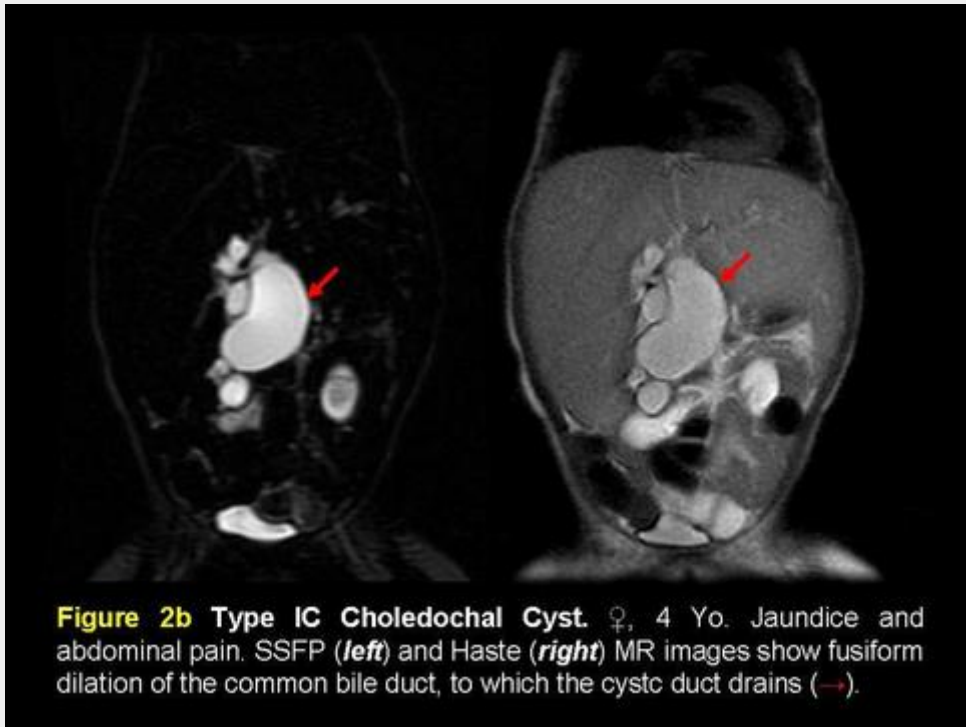
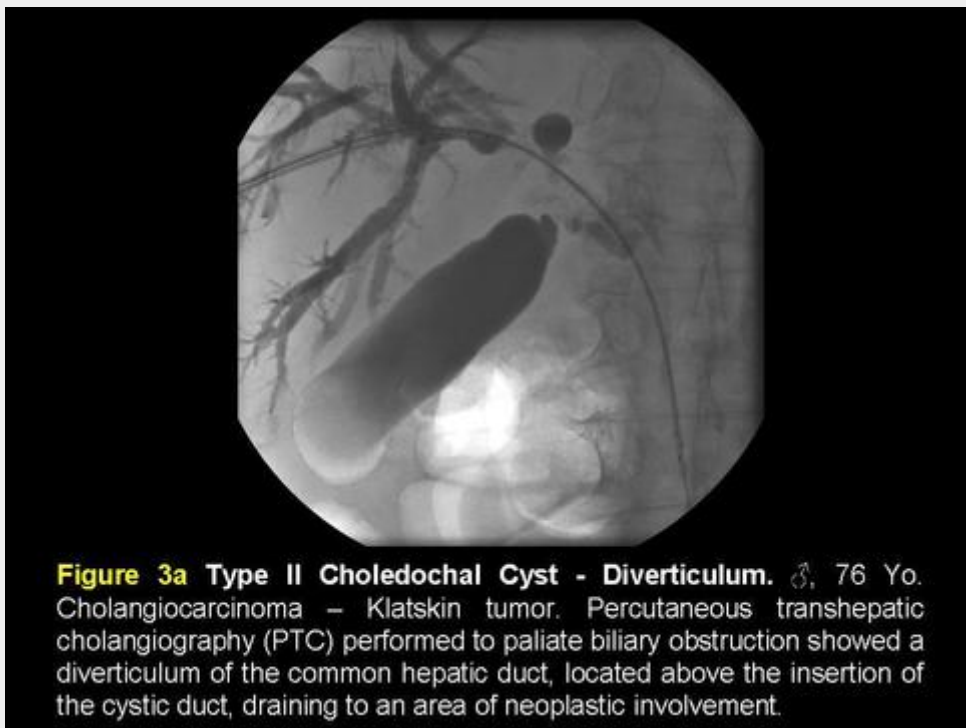


Figure 2a Type IC Choledochal Cyst. ♀, 4 Yo with jaundice and abdominal pain. MR Haste images shows fusiform dilation of the common bile duct (→). Cystic duct →; Gallblader →.

diapositivo20.jpg



diapositivo23.jpg



diapositivo30.jpg

IMAGING FINDINGS

Intrahepatic Bile Duct Dilatation

- Autosomic recessive inheritance
- Multifocal segmental dilatation of intrahepatic bile ducts retaining communication with the biliary tree
- 2 types:
 - Caroli disease (pure form)
 - IHBD dilatations
 - Caroli syndrome
 - IHBD dilatations
 - Congenital hepatic fibrosis

diapositivo31.jpg

IMAGING FINDINGS

Caroli's Disease

Pathogenesis

- Neonatal occlusion of the hepatic artery, leading to bile duct ischemia and cystic dilatation
- Abnormal growth rate of the developing biliary epithelium and supporting connective tissue
- Lack of normal involution of ductal plates that surround the portal tracts, resulting in epithelium-lined cysts that surround the portal triads

diapositivo32.jpg

IMAGING FINDINGS

Caroli's Disease

Associated ductal plate abnormalities

- Congenital hepatic fibrosis
- Polycystic renal diseases
 - Medullary sponge kidney
 - ARPKD
 - Nephronoptosis

diapositivo33.jpg

IMAGING FINDINGS

Caroli's Disease

Differential Diagnosis

- Polycystic liver disease
- Biliary microhamartomas
- Primary sclerosing cholangitis
- Recurrent pyogenic cholangitis
(oriental cholangiohepatitis)

diapositivo34.jpg

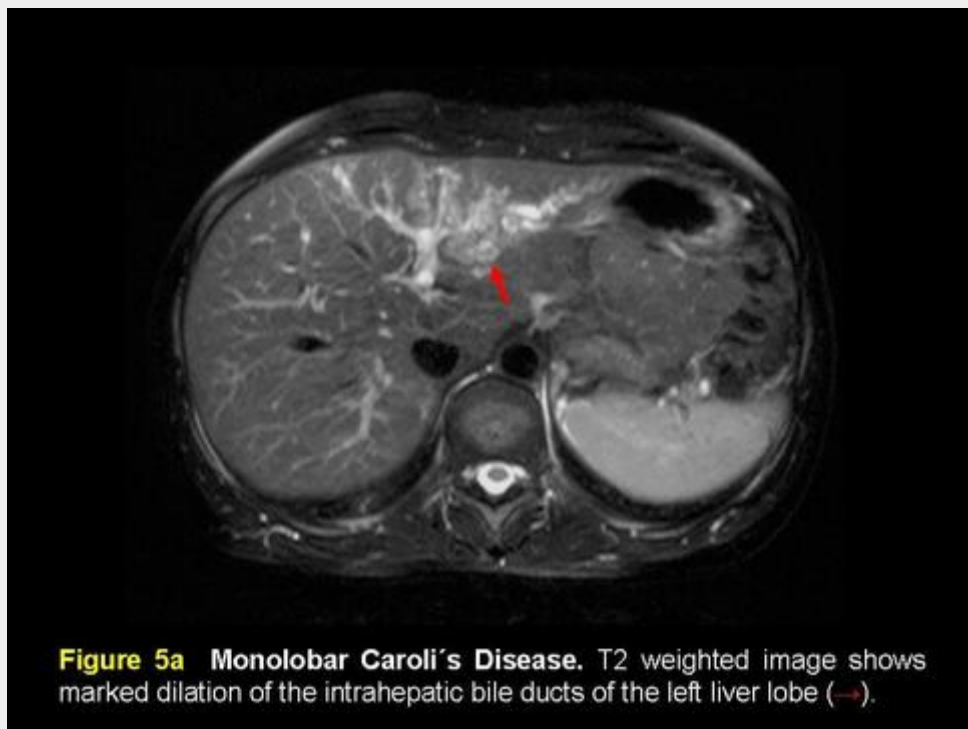


Figure 5a Monolobar Caroli's Disease. T2 weighted image shows marked dilation of the intrahepatic bile ducts of the left liver lobe (→).

diapositivo35.jpg

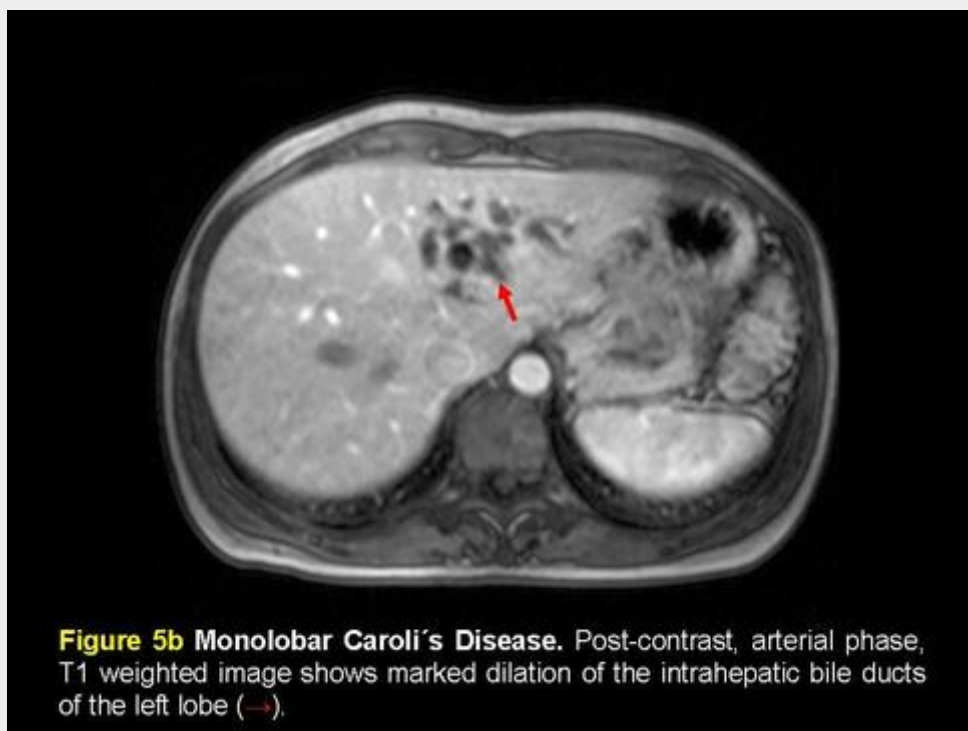


Figure 5b Monolobar Caroli's Disease. Post-contrast, arterial phase, T1 weighted image shows marked dilation of the intrahepatic bile ducts of the left lobe (→).

diapositivo36.jpg

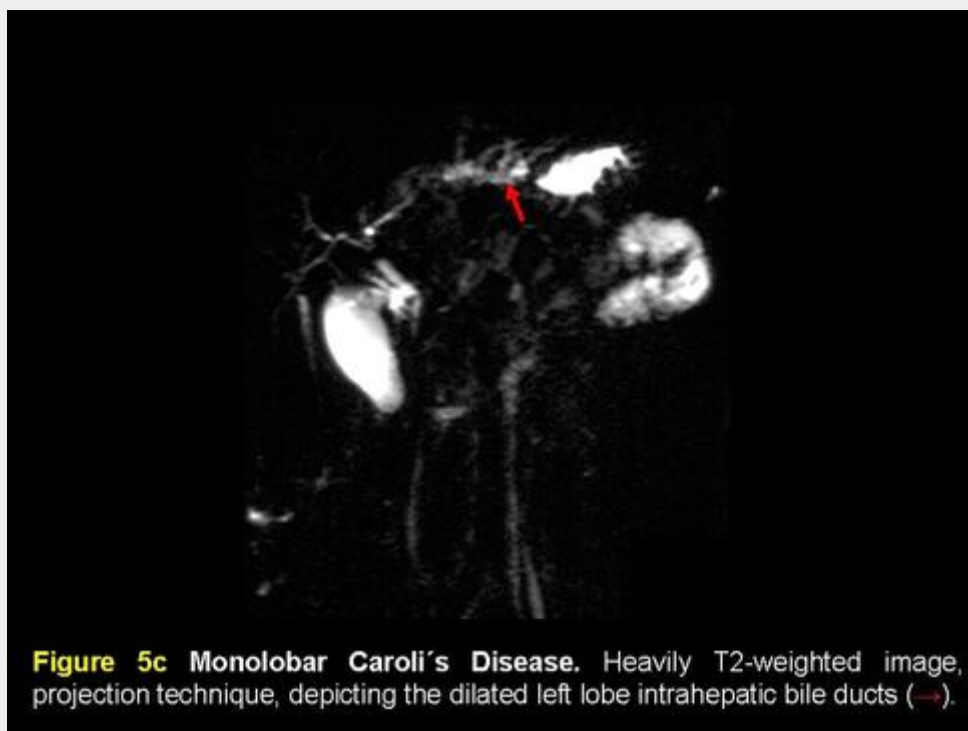


Figure 5c Monolobar Caroli's Disease. Heavily T2-weighted image, projection technique, depicting the dilated left lobe intrahepatic bile ducts (→).

diapositivo37.jpg

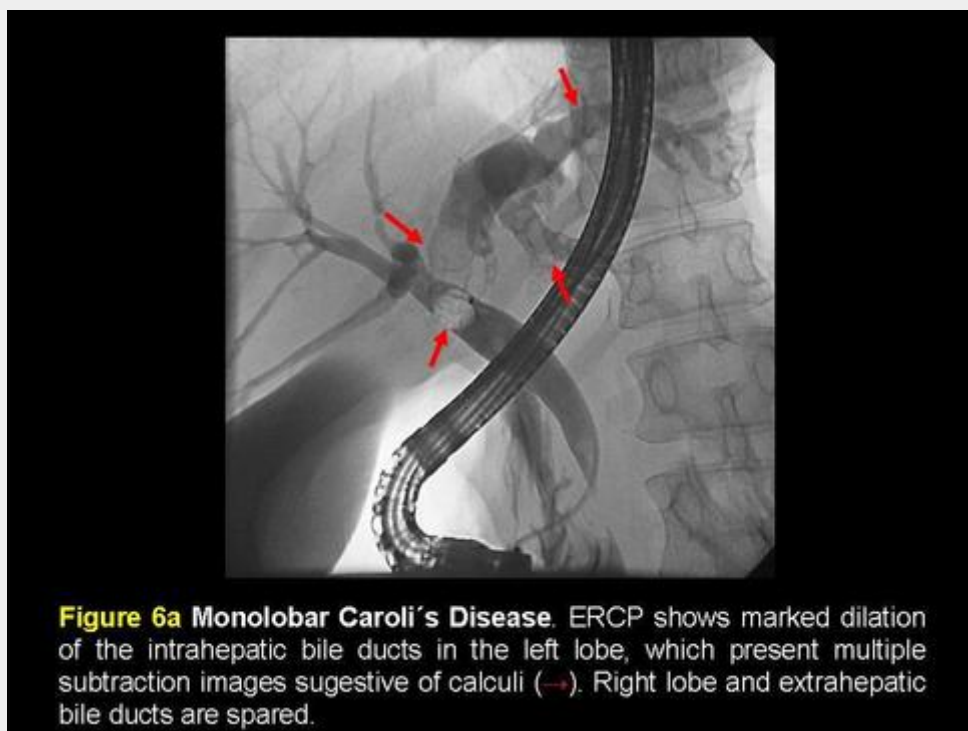


Figure 6a Monolobar Caroli's Disease. ERCP shows marked dilation of the intrahepatic bile ducts in the left lobe, which present multiple subtraction images suggestive of calculi (→). Right lobe and extrahepatic bile ducts are spared.

diapositivo39.jpg

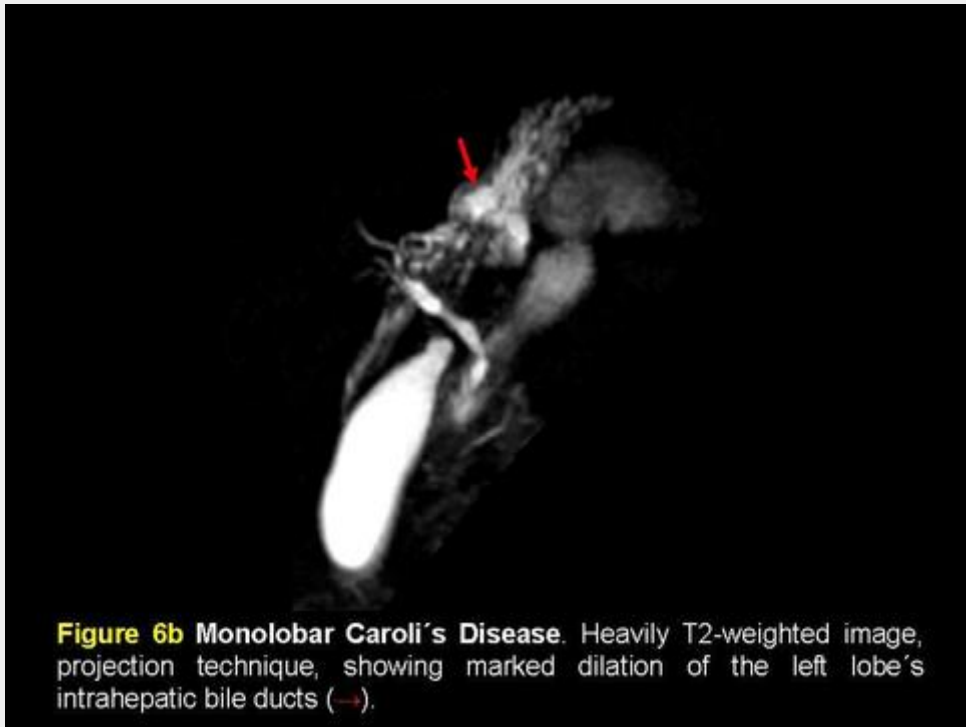


Figure 6b Monolobar Caroli's Disease. Heavily T2-weighted image, projection technique, showing marked dilation of the left lobe's intrahepatic bile ducts (→).

diapositivo41.jpg

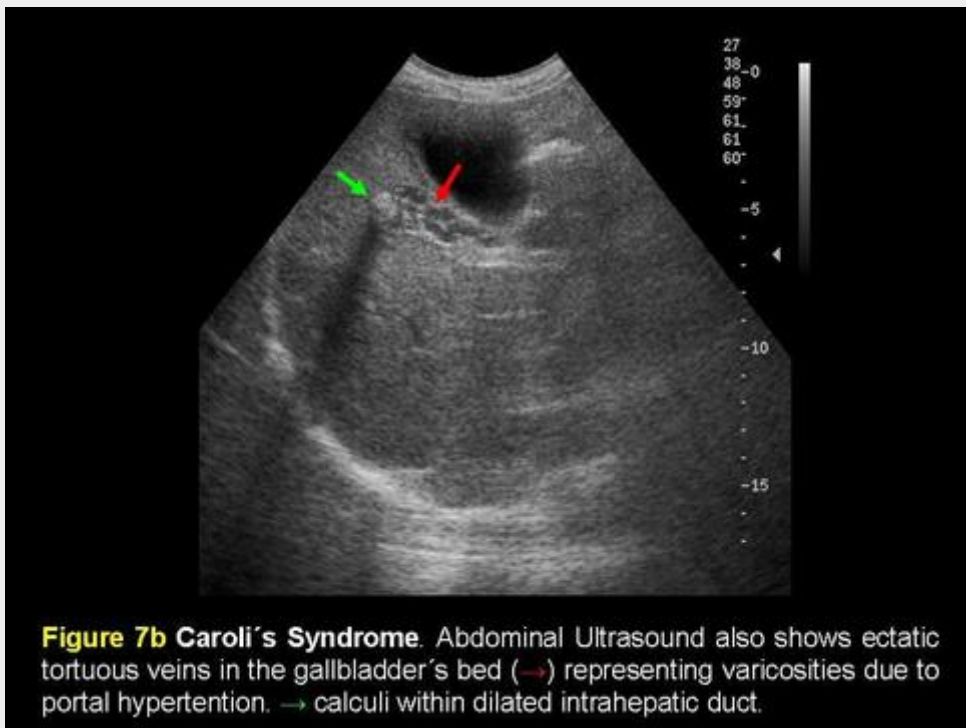


Figure 7b Caroli's Syndrome. Abdominal Ultrasound also shows ectatic tortuous veins in the gallbladder's bed (→) representing varicosities due to portal hypertension. → calculi within dilated intrahepatic duct.

diapositivo42.jpg

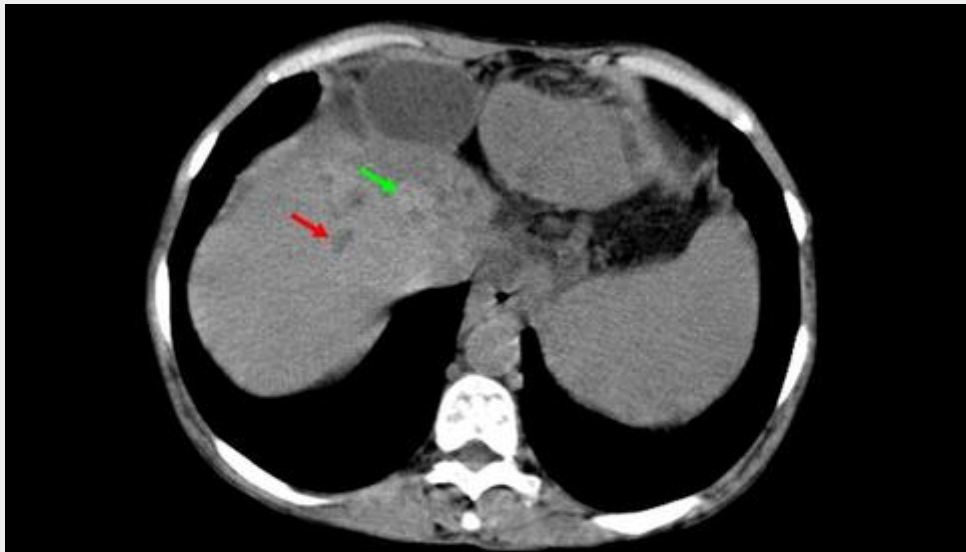


Figure 7c Caroli's Syndrome. Pre-contrast abdominal CT. There are dilated bile ducts in the left lobe (→), some of which filled with hyperdense material, corresponding to bile calculi (→).

diapositivo43.jpg

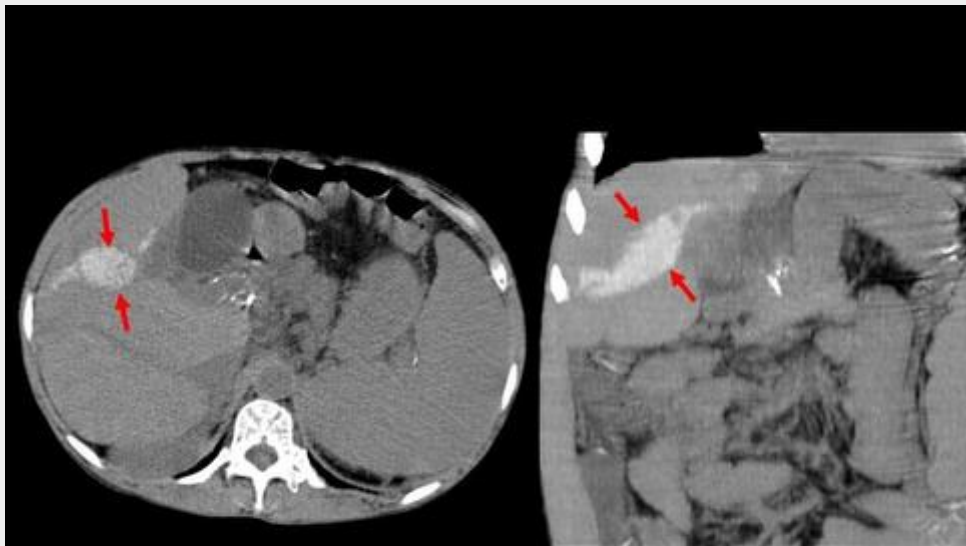


Figure 7d Caroli's Syndrome. Pre-contrast abdominal CT. Axial (*left*) and oblique reformed (*right*) images. There is a markedly dilated bile duct in the right lobe, completely filled with hyperdense material, corresponding to bile calculi (→).

diapositivo44.jpg

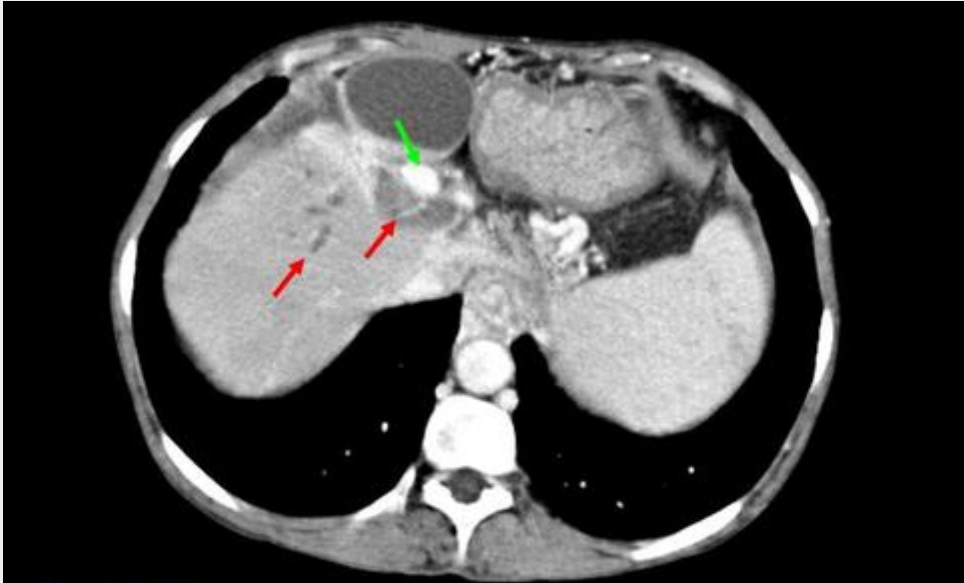


Figure 7e Caroli's Syndrome. Post-contrast abdominal CT better depicting the dilated bile ducts in the left lobe (→) surrounding a dilated patent umbilical vein (→) due to coexistent portal hypertension.

diapositivo45.jpg

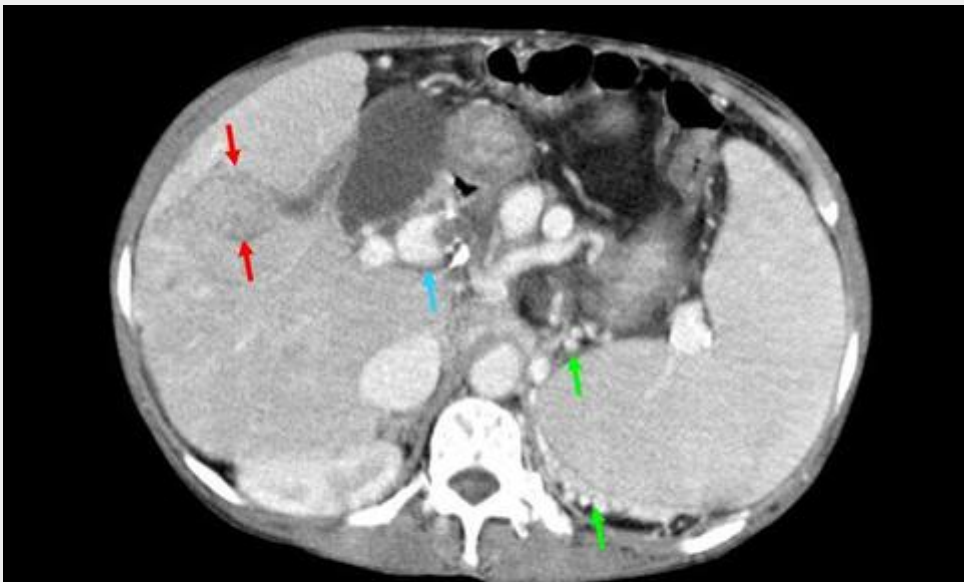


Figure 7f Caroli's Syndrome. Post-contrast abdominal CT depicting the dilated intra-hepatic bile duct filled with calculi (→). Note the splenomegaly, dilation of the portal vein (→) and perisplenic varicosities (→).

diapositivo47.jpg

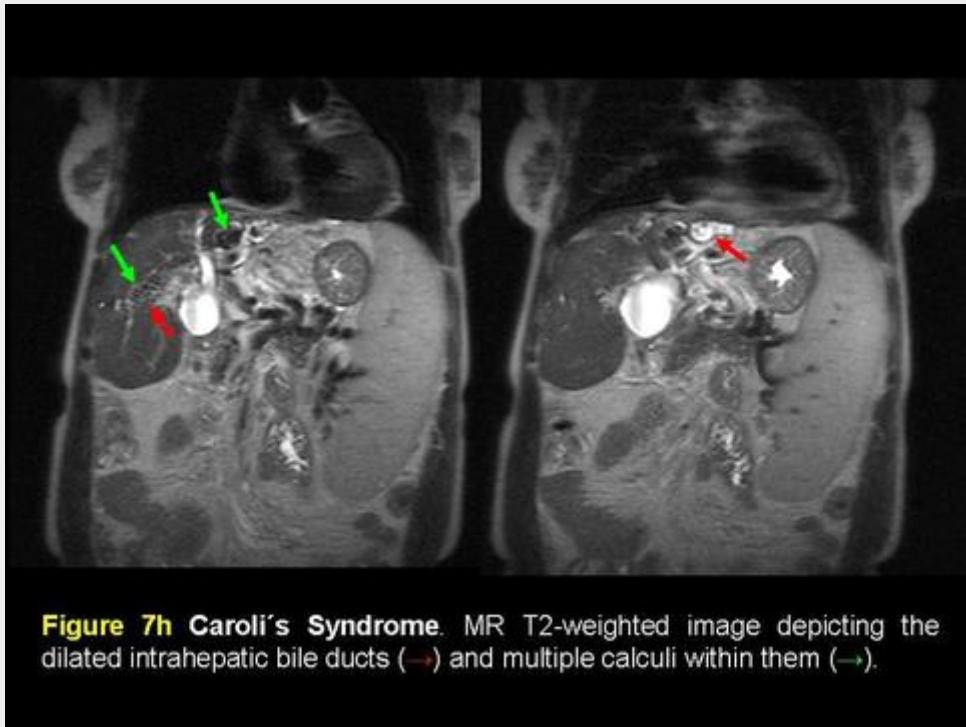


Figure 7h Caroli's Syndrome. MR T2-weighted image depicting the dilated intrahepatic bile ducts (→) and multiple calculi within them (→).

diapositivo48.jpg

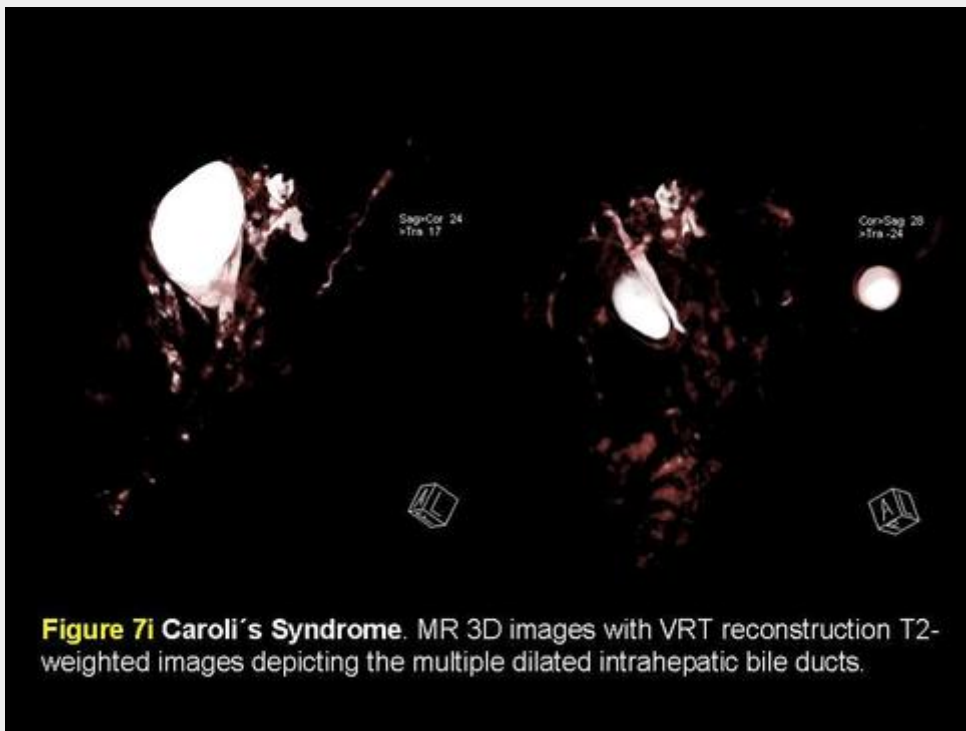


Figure 7i Caroli's Syndrome. MR 3D images with VRT reconstruction T2-weighted images depicting the multiple dilated intrahepatic bile ducts.

diapositivo50.jpg

IMAGING FINDINGS

Caroli's Disease

- Central Dot Sign
 - Solid "dot" within or at the periphery of a cystic liver lesion
 - Doppler signal
 - Continuous (portal vein branch)
 - Arterial waveform (hepatic artery branch)
 - Enhancement
 - CT, MRI

diapositivo51.jpg

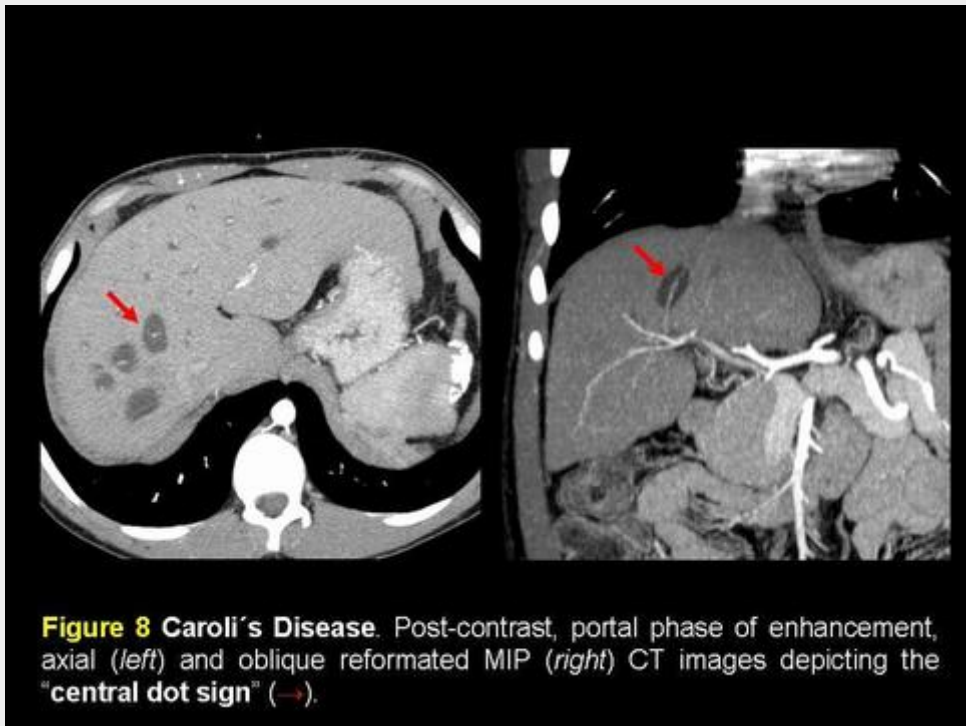


Figure 8 Caroli's Disease. Post-contrast, portal phase of enhancement, axial (*left*) and oblique reformed MIP (*right*) CT images depicting the "central dot sign" (→).

diapositivo52.jpg

IMAGING FINDINGS

Caroli's Disease

- **Complications**
 - Cholangitis, stones
 - Strictures
 - Cholangiocarcinoma (7-14%)
- **Caroli's syndrome:**
 - Portal hypertension
 - Secondary biliary cirrhosis

diapositivo53.jpg

CONCLUSIONS

Choledochal cysts are uncommon entities easily depicted by the imaging modalities presented. Their recognition is very important because early intervention may avoid many of the possible unwanted complications.

diapositivo55.jpg

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diapositivo56.jpg

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