



## **Choledochal Cysts - Spectrum of Imaging Findings**

e-Poster: P-026

Congress: ESGAR 2009

Type: Educational Exhibit

Topic: Diagnostic / Bile Ducts

Authors: I. Santiago<sup>1</sup>, J. Maciel<sup>1</sup>, R.M.G. Loureiro<sup>2</sup>, C. Ruivo<sup>3</sup>, M.A. Portilha<sup>3</sup>, B.J.A.M. Gonçalves<sup>3</sup>, N.J.L. Dias<sup>3</sup>, M.A. Cipriano<sup>3</sup>, F. Caseiro Alves<sup>3</sup>; <sup>1</sup>Aveiro/PT, <sup>2</sup>Funchal/PT, <sup>3</sup>Coimbra/PT

MeSH:

Choledochal Cyst [C06.130.120.127]

Keyword: Choledochal Cysts

Any information contained in this pdf file is automatically generated from digital material submitted to EPOS by third parties in the form of scientific presentations. References to any names, marks, products, or services of third parties or hypertext links to third-party sites or information are provided solely as a convenience to you and do not in any way constitute or imply ESGAR's endorsement, sponsorship or recommendation of the third party, information, product, or service. ESGAR is not responsible for the content of these pages and does not make any representations regarding the content or accuracy of material in this file.

As per copyright regulations, any unauthorised use of the material or parts thereof as well as commercial reproduction or multiple distribution by any traditional or electronically based reproduction/publication method is strictly prohibited.

You agree to defend, indemnify, and hold ESGAR harmless from and against any and all claims, damages, costs, and expenses, including attorneys' fees, arising from or related to your use of these pages.

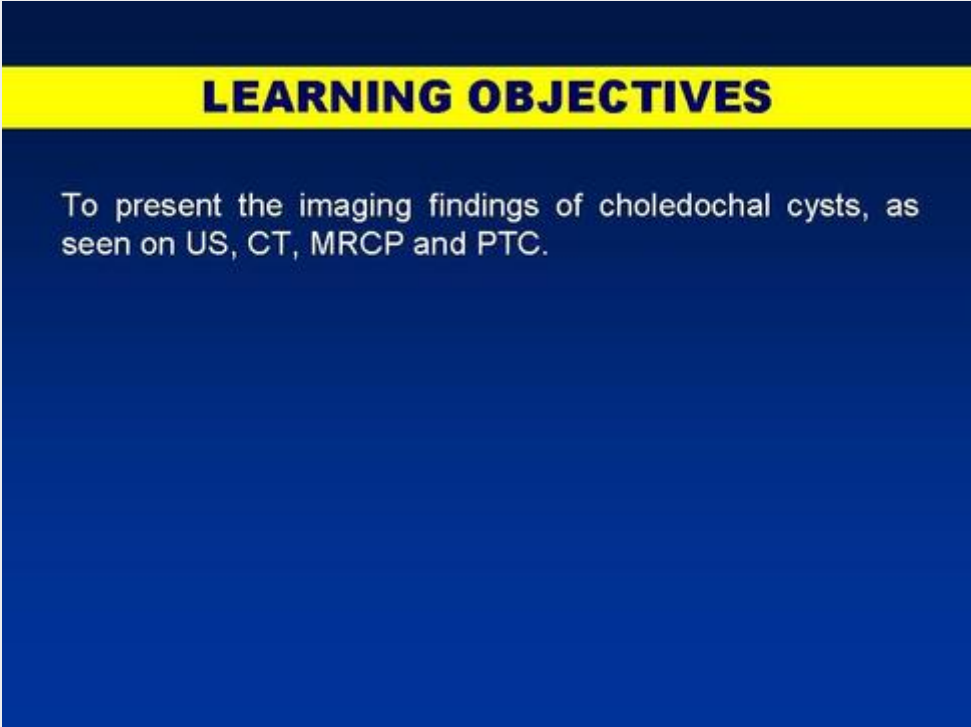
Please note: Links to movies, ppt slideshows and any other multimedia files are not available in the pdf version of presentations.

[www.esgar.org](http://www.esgar.org)

## 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

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)

### 3. Imaging findings OR Procedure details

Imaging Findings

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

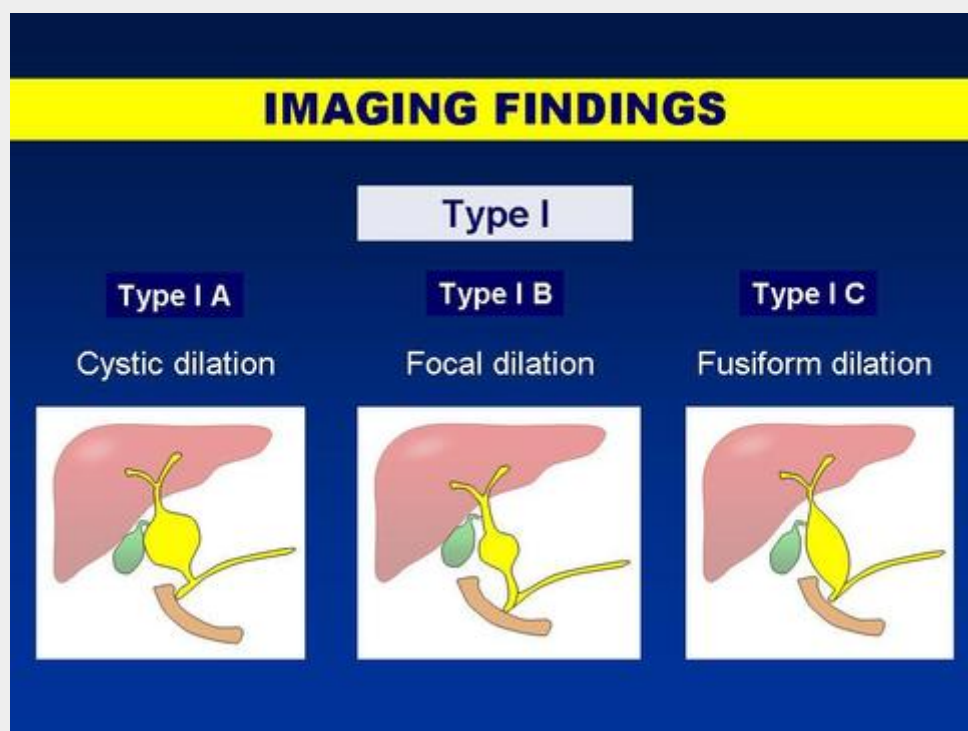
## IMAGING FINDINGS

### Classification

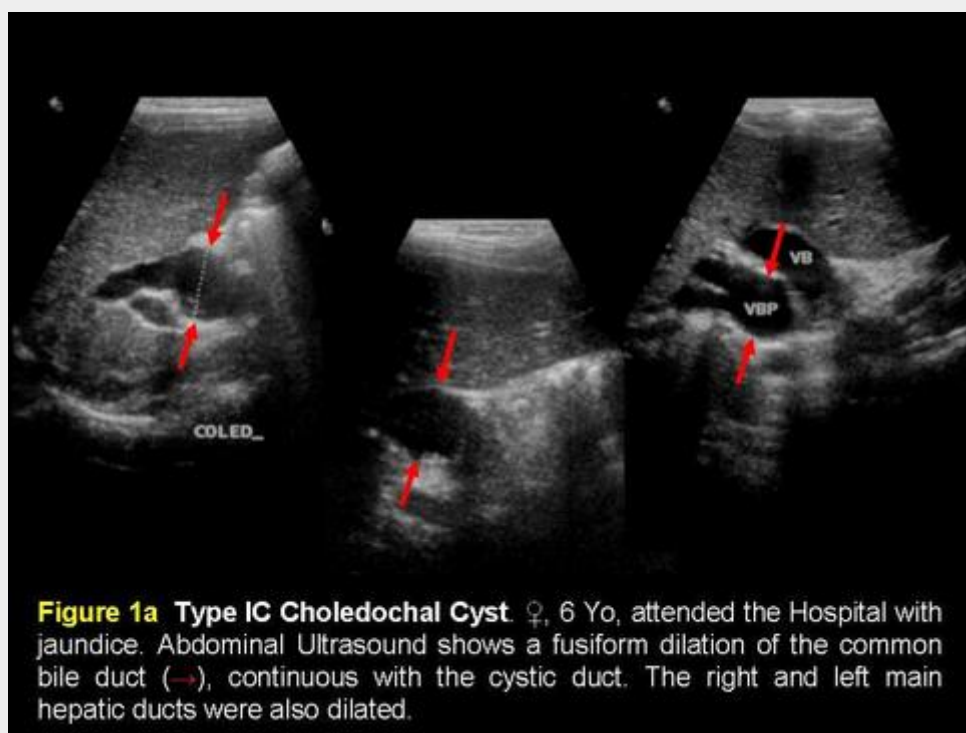
#### *Todani Modification of the Alonso – Lej Classification*

- |                 |                                     |
|-----------------|-------------------------------------|
| <b>Type I</b>   | Solitary, extrahepatic cyst         |
| <b>Type II</b>  | Extrahepatic duodenal diverticulum  |
| <b>Type III</b> | Intraduodenal cyst                  |
| <b>Type IV</b>  | Extrahepatic and intrahepatic cysts |
| <b>Type V</b>   | Multiple intrahepatic cysts         |

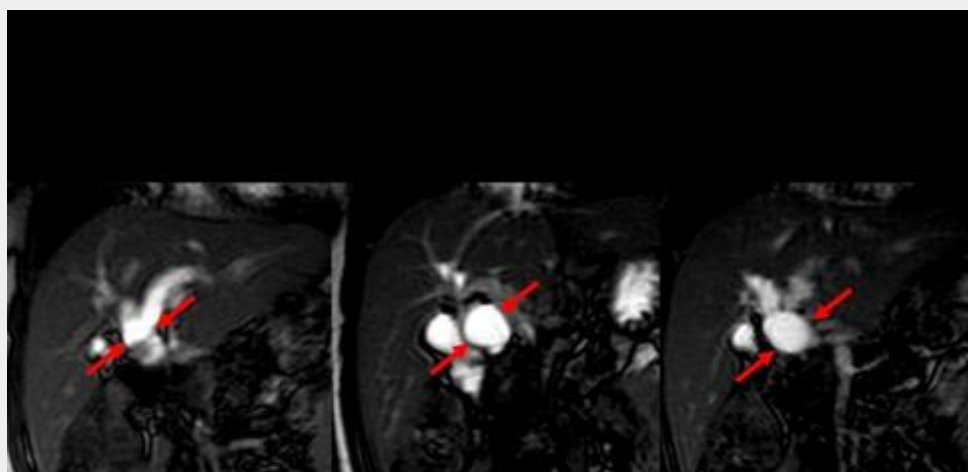
## Type I



diapositivo15.jpg

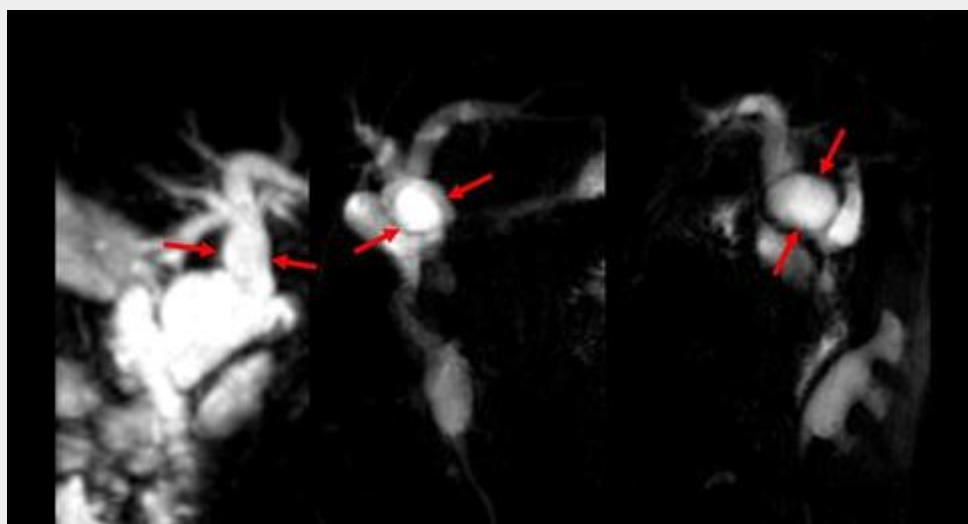


diapositivo16.jpg



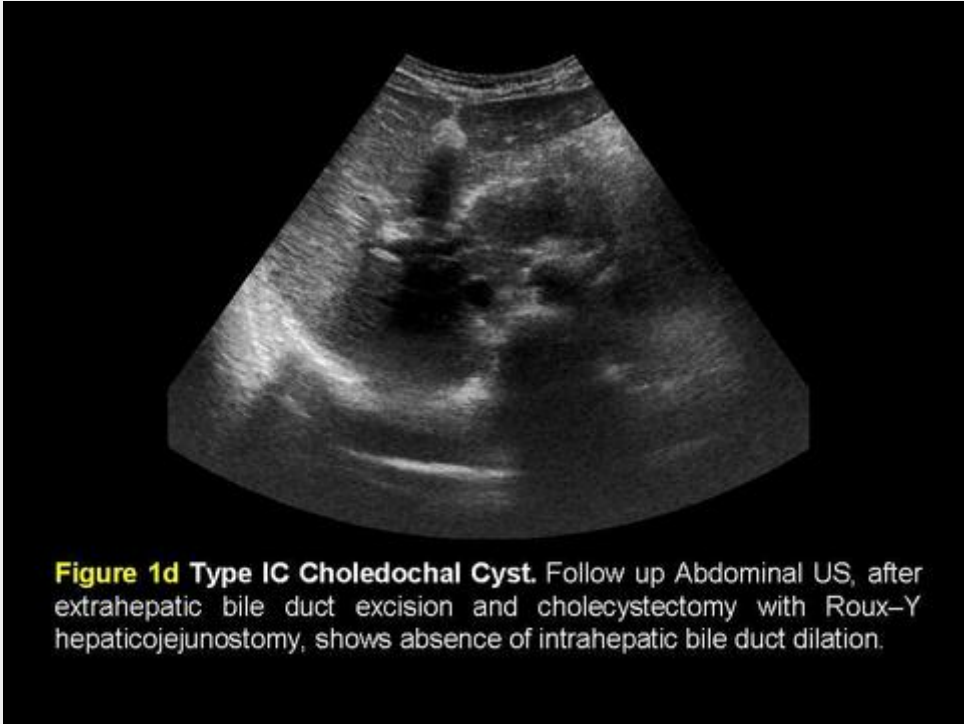
**Figure 1b** Type IC Choledochal Cyst. Magnetic Resonance SSFP sequences show a fusiform dilation of the common hepatic and common bile ducts. The cystic duct drains into the dilated extrahepatic bile duct.

diapositivo17.jpg



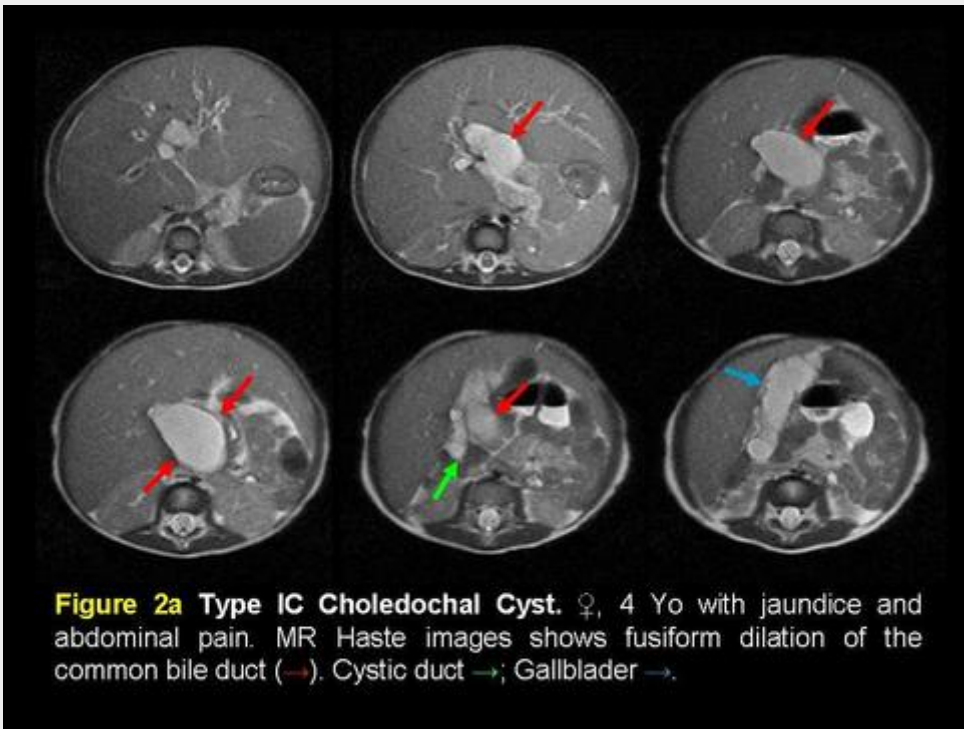
**Figure 1c** Type IC Choledochal Cyst. MR T2 reformatted images show a fusiform dilation of the common hepatic and common bile ducts. The cystic duct drains to the dilated extrahepatic bile duct. These findings correspond to a Type IC choledochal cyst, according to the Todani classification.

diapositivo18.jpg



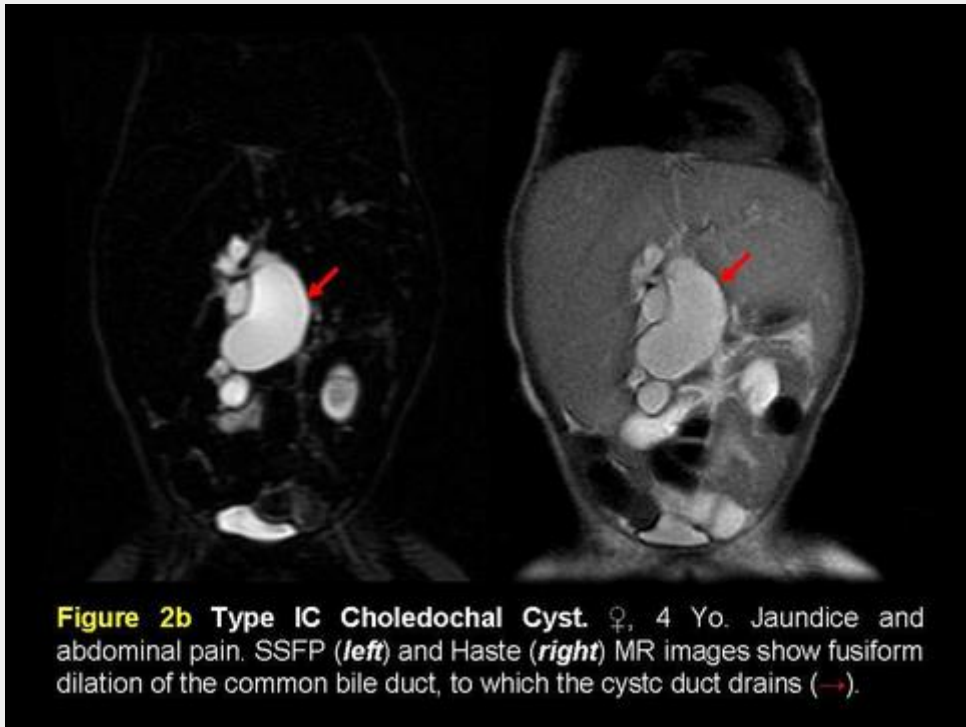
**Figure 1d** Type IC Choledochal Cyst. Follow up Abdominal US, after extrahepatic bile duct excision and cholecystectomy with Roux-Y hepaticojejunostomy, shows absence of intrahepatic bile duct dilation.

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



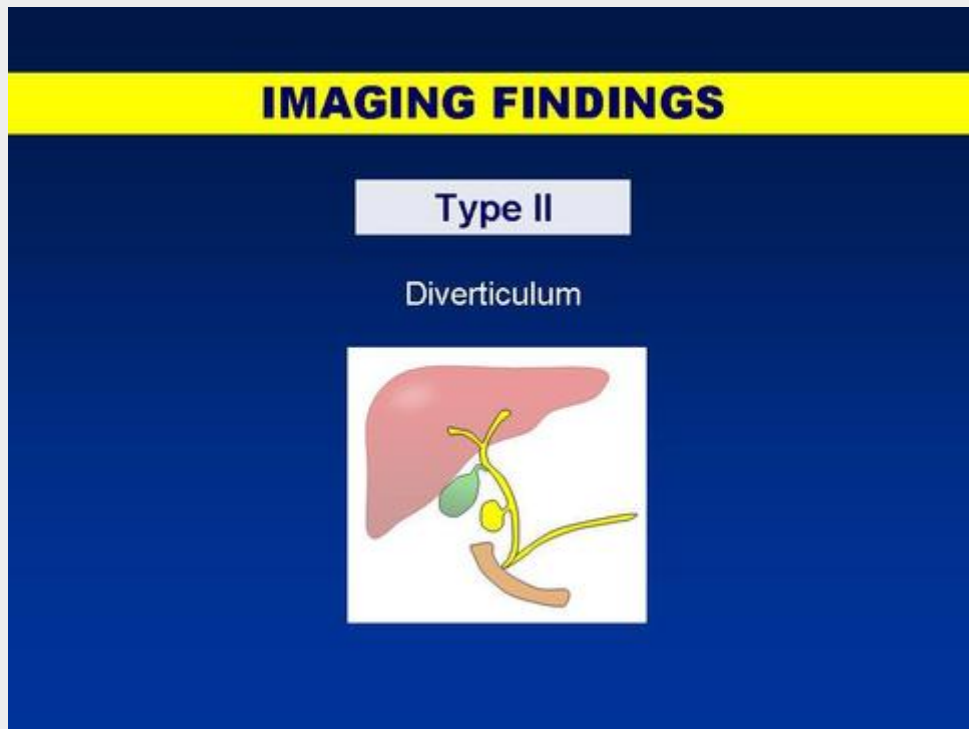
Type IC



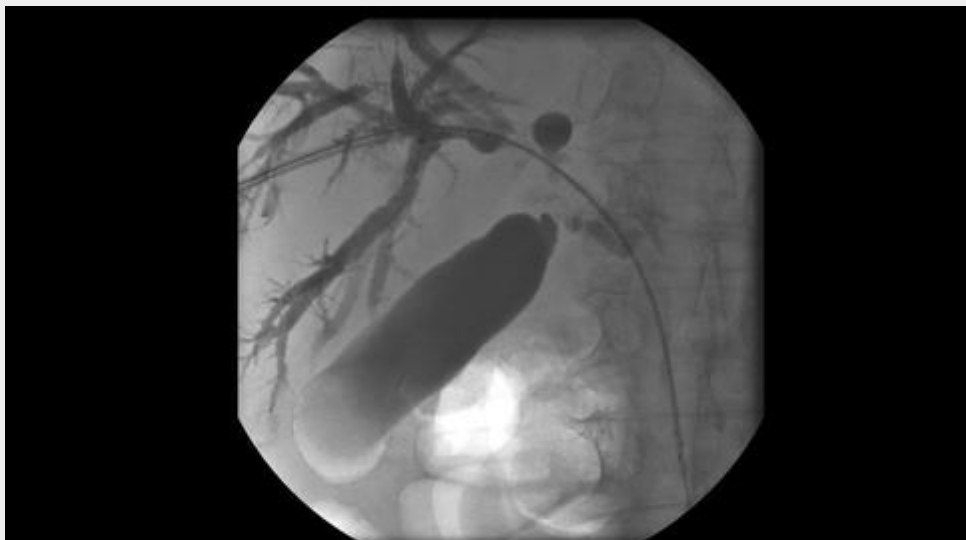
[\[Type IC Choledochal Cyst\] Video 1](#) shows a Type IC Choledochal Cyst, as seen on MR Heavily

T2-weighted reformatted images.

## Type II

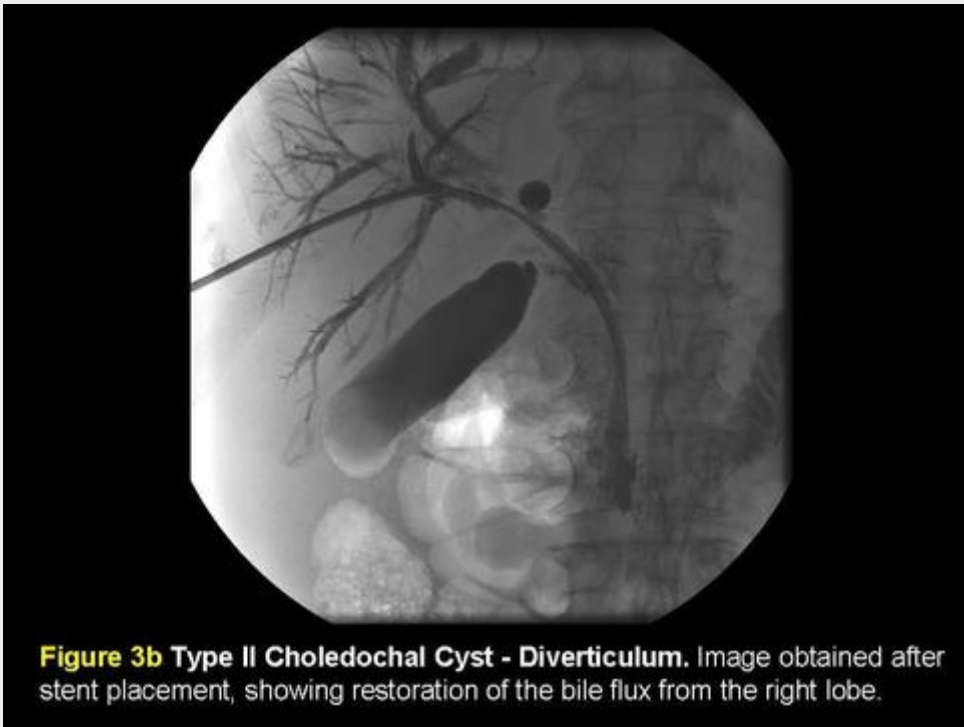


diapositivo23.jpg



**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

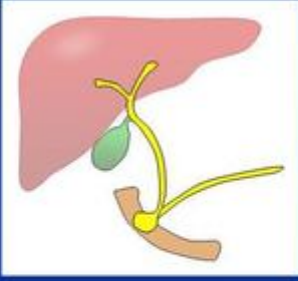


## Type III

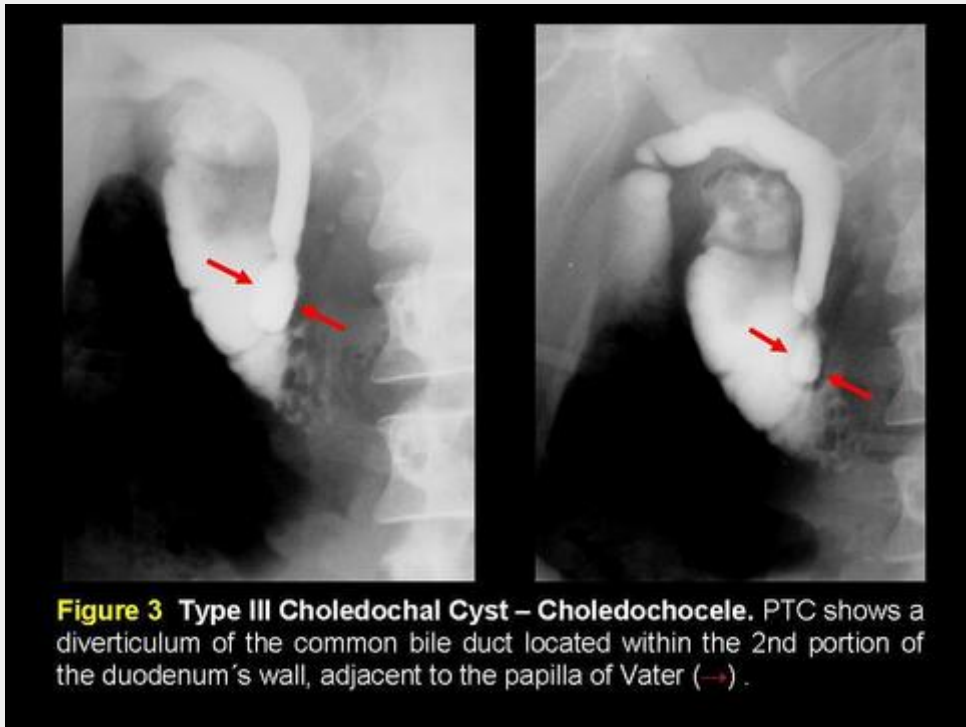
**IMAGING FINDINGS**

Type III

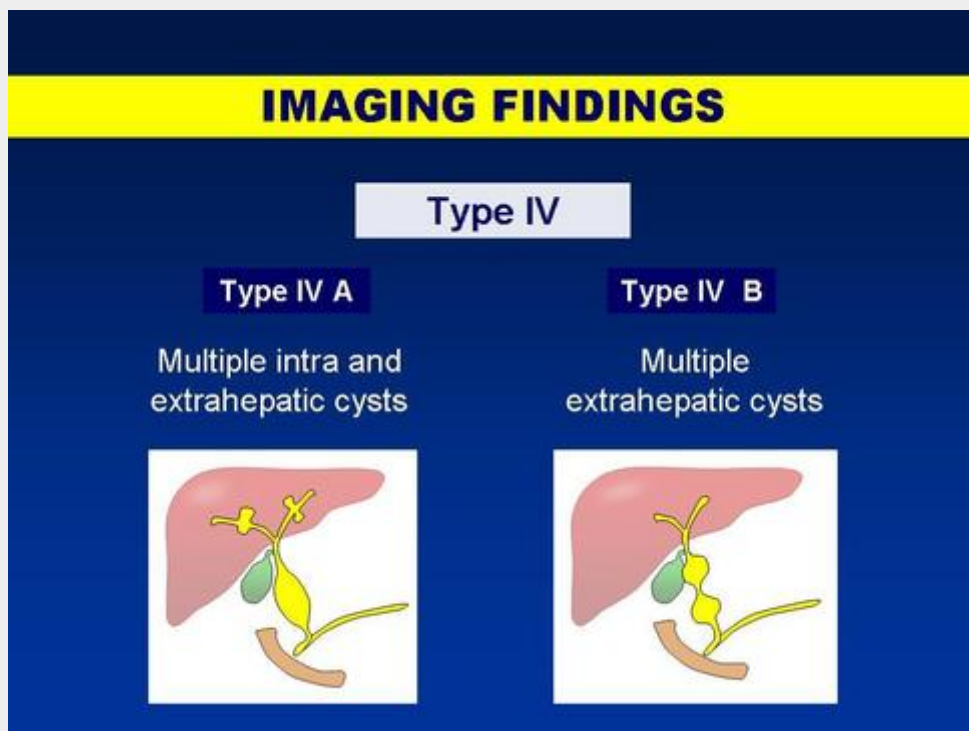
Choledochoceles



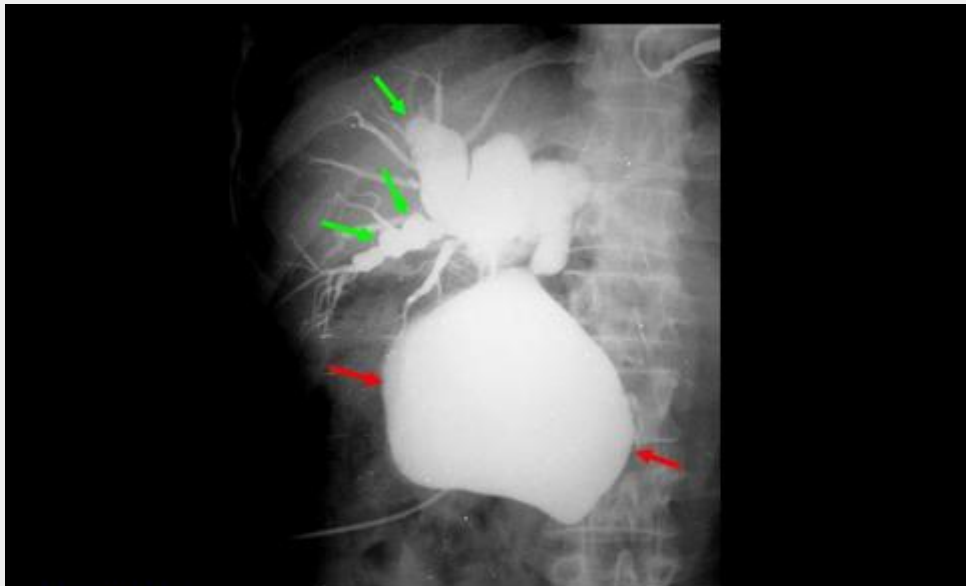
## Choledochocele



## Type IV



## 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 (→).

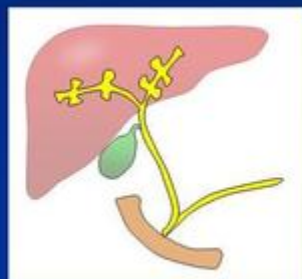
## Type V

### IMAGING FINDINGS

#### Type V

Caroli's Disease

Multiple intrahepatic cysts



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

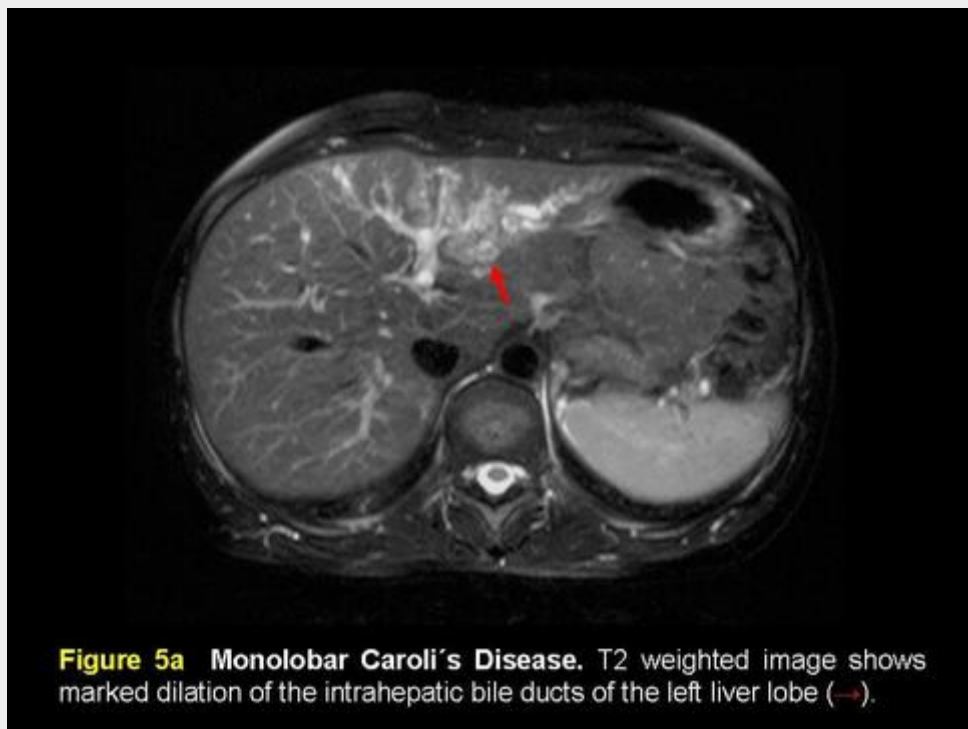
## IMAGING FINDINGS

### Caroli's Disease

#### Differential Diagnosis

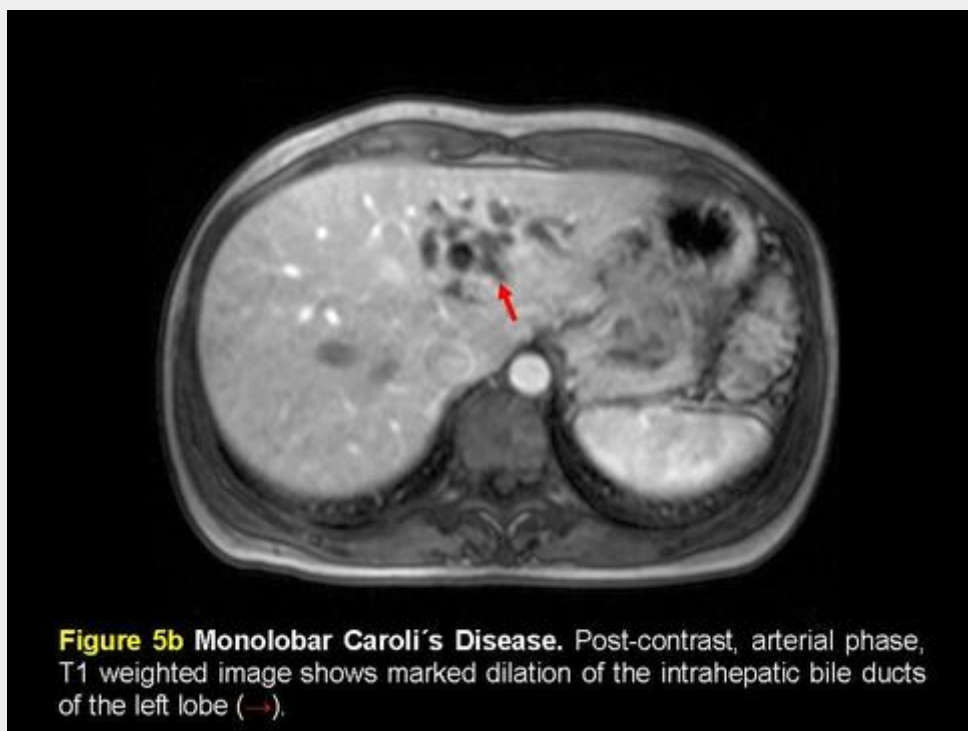
- Polycystic liver disease
- Biliary microhamartomas
- Primary sclerosing cholangitis
- Recurrent pyogenic cholangitis  
(oriental cholangiohepatitis)

diapositivo34.jpg



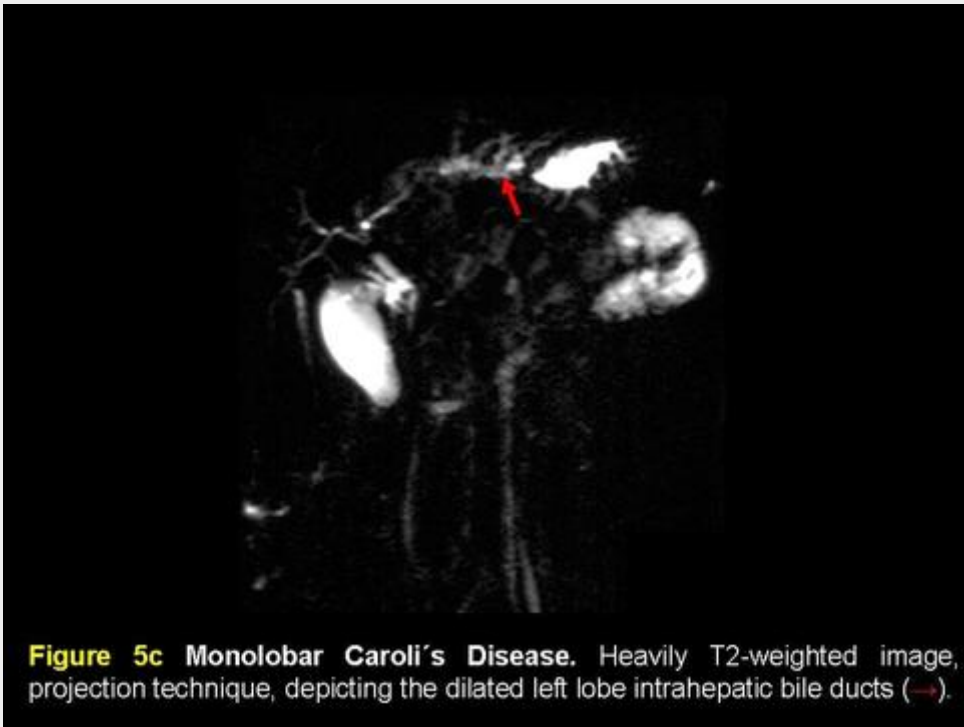
**Figure 5a Monolobar Caroli's Disease.** T2 weighted image shows marked dilation of the intrahepatic bile ducts of the left liver lobe (→).

diapositivo35.jpg



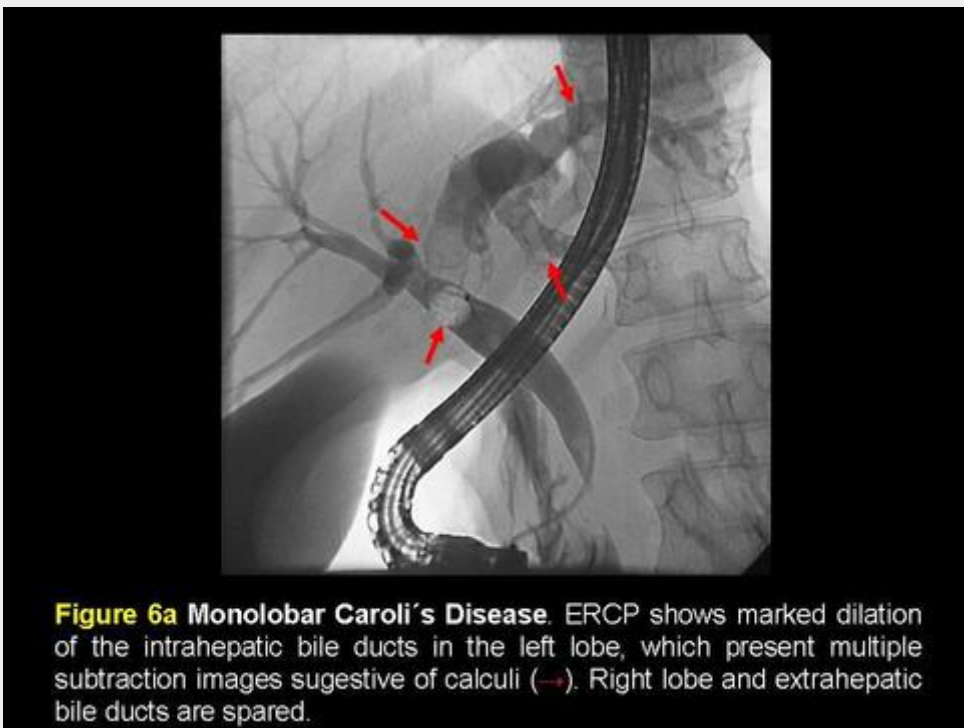
**Figure 5b Monolobar Caroli's Disease.** Post-contrast, arterial phase, T1 weighted image shows marked dilation of the intrahepatic bile ducts of the left lobe (→).

diapositivo36.jpg



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

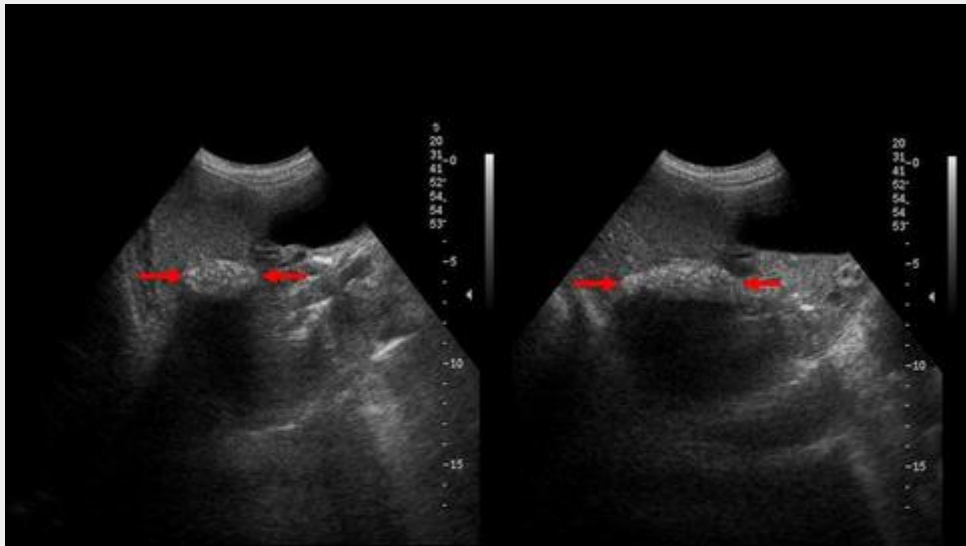
diapositivo37.jpg



**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.

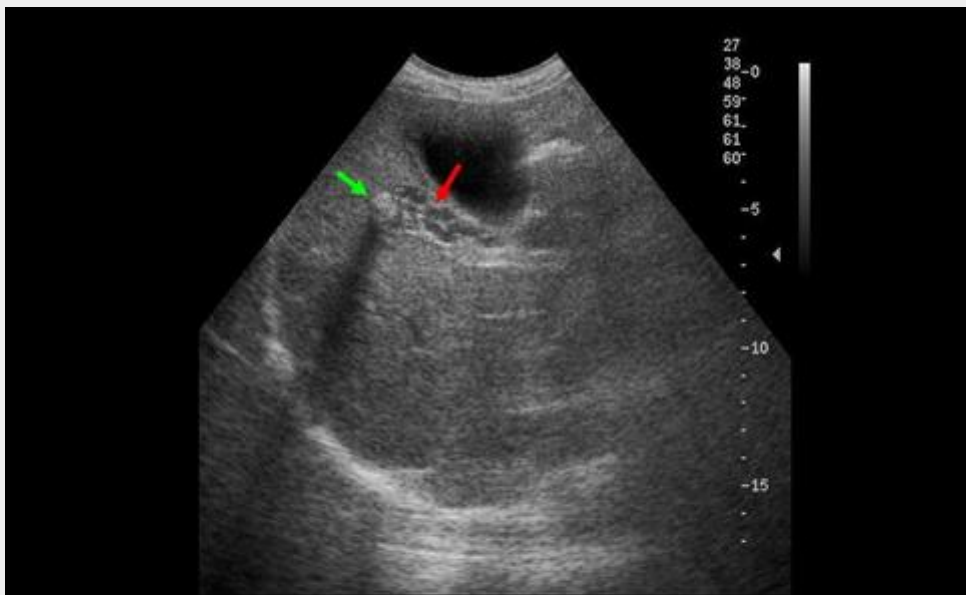


## Caroli's Syndrome



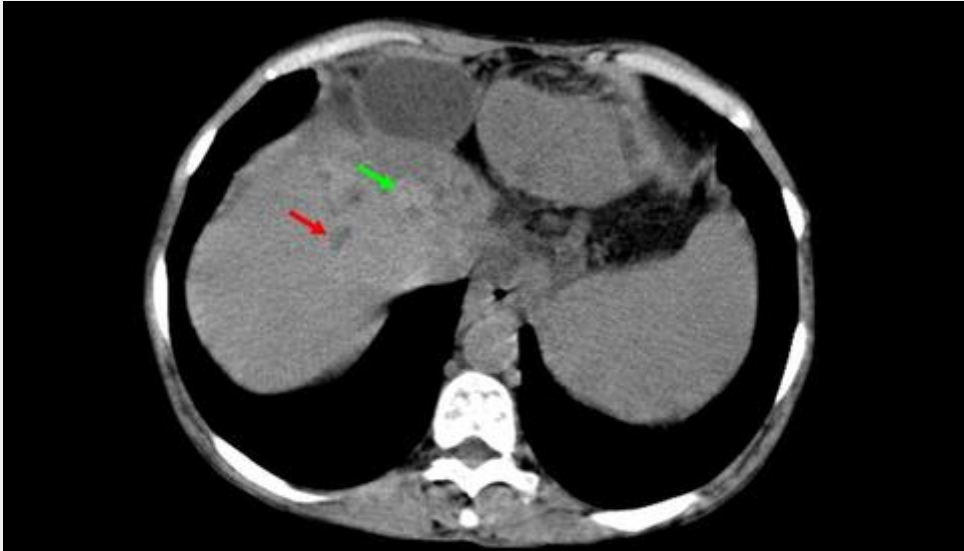
**Figure 7a Caroli's Syndrome.** Abdominal Ultrasound shows an oval hyperechogenic image with posterior acoustic shadowing in the right liver lobe (→), which corresponds to a markedly dilated intrahepatic bile duct, completely filled with calculi.

## diapositivo41.jpg



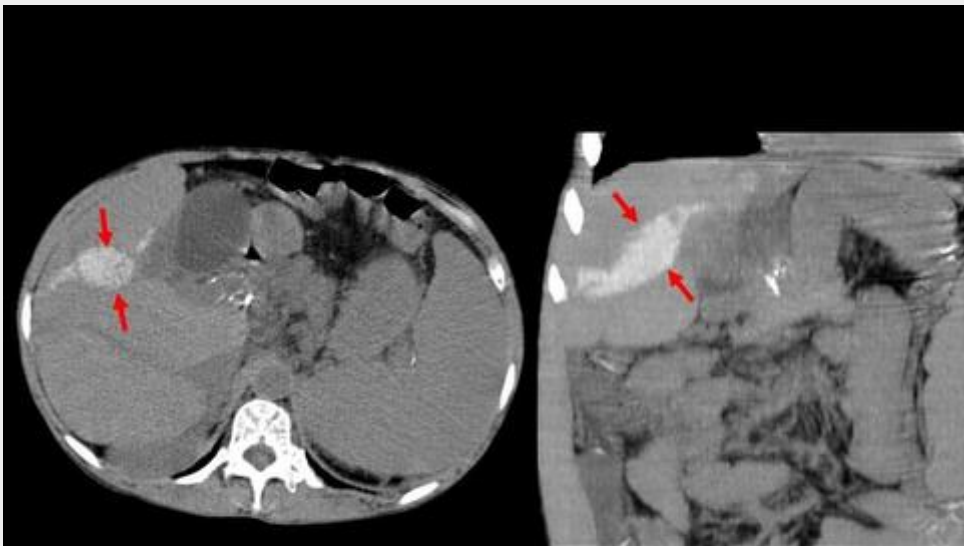
**Figure 7b Caroli's Syndrome.** Abdominal Ultrasound also shows ectatic tortuous veins in the gallbladder's bed (→) representing varicosities due to portal hypertension. → calculi within dilated intrahepatic duct.

diapositivo42.jpg



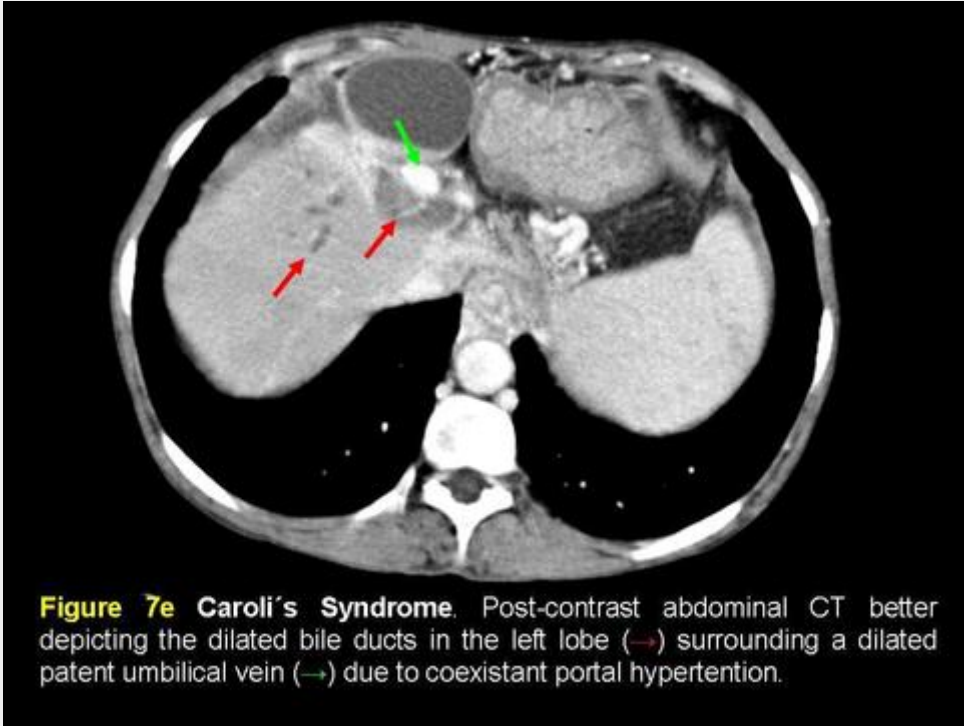
**Figure 7c Caroli's Syndrome.** Pre-contrast abdominal CT. There are dilated bile ducts in the left lobe (→), some of which filled with hyperdense material, corresponding to bile calculi (→).

diapositivo43.jpg

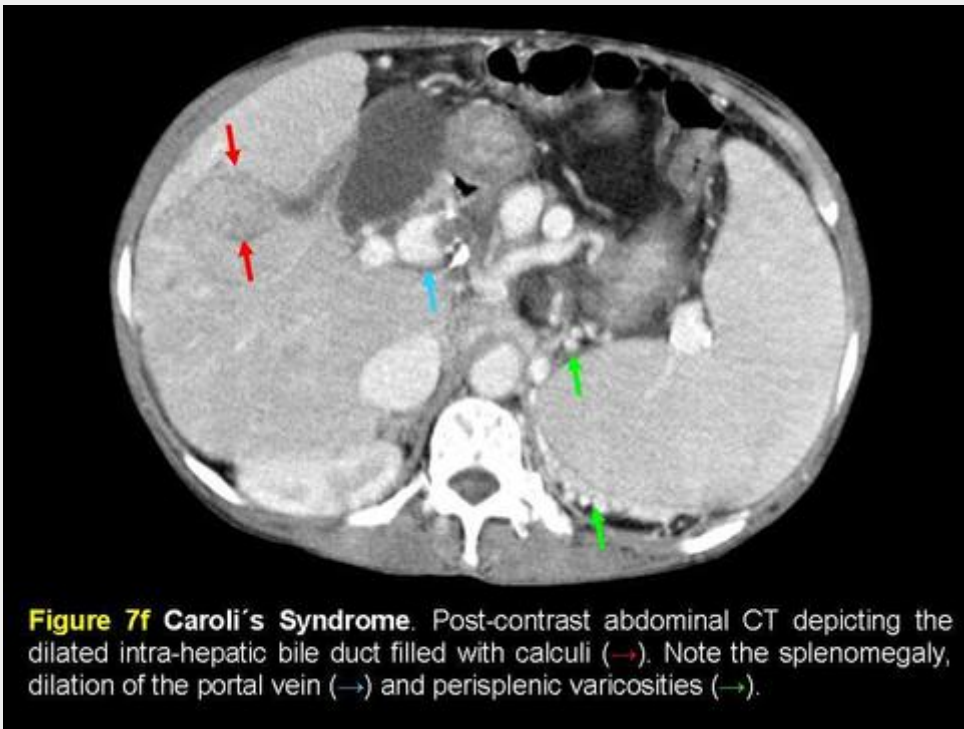


**Figure 7d Caroli's Syndrome.** Pre-contrast abdominal CT. Axial (*left*) and oblique reformed (*right*) images. There is a markedly dilated bile duct in the right lobe, completely filled with hyperdense material, corresponding to bile calculi (→).

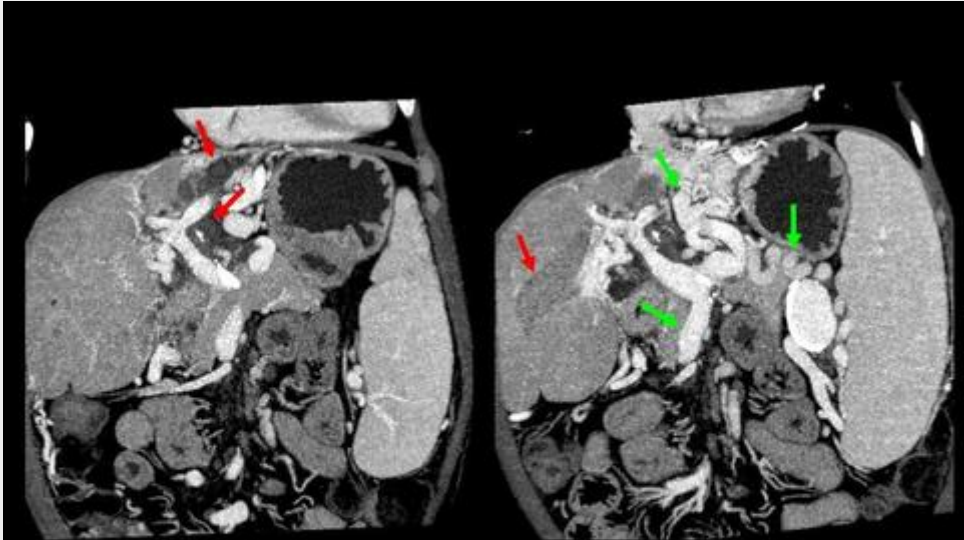
diapositivo44.jpg



diapositivo45.jpg

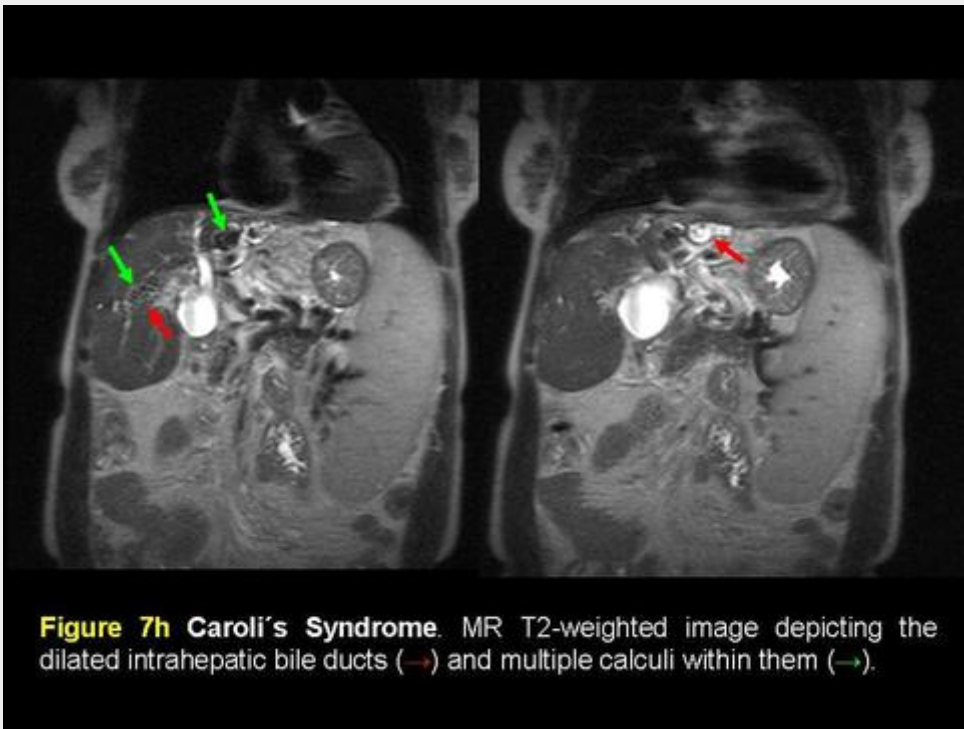


## Caroli's Syndrome



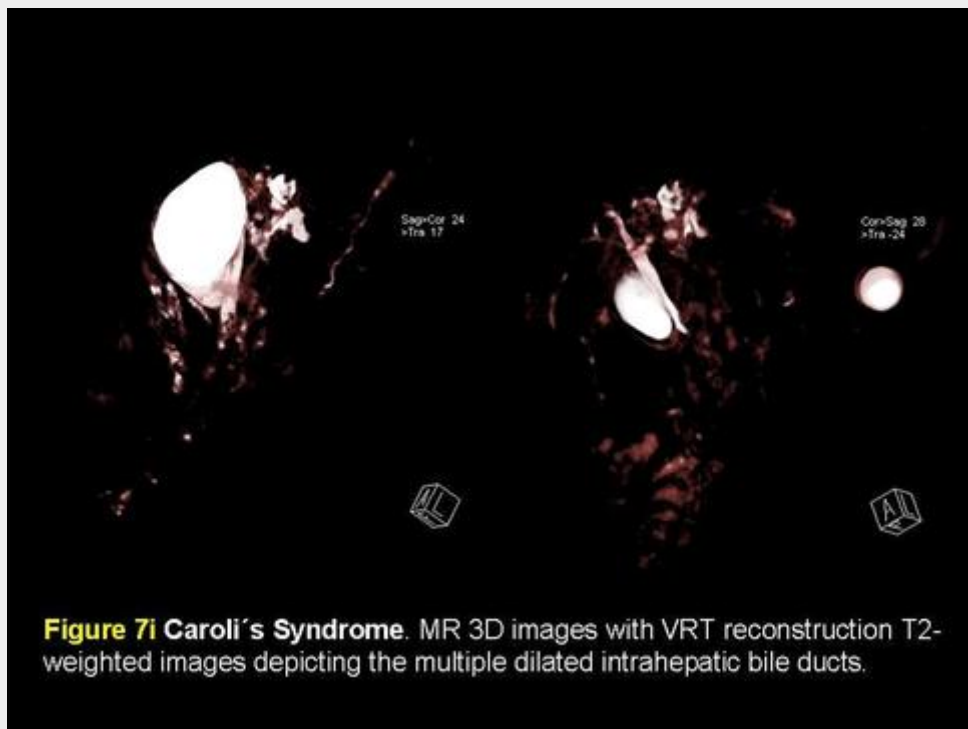
**Figure 7g Caroli's Syndrome.** Post-contrast abdominal CT. Reformed oblique MIP images depicting the dilated intra-hepatic bile ducts, some of which filled with calculi (→). Note the splenomegaly and the tortuosity and dilation of the portal, splenic and perigastric veins (→) due to portal hypertension.

## 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.

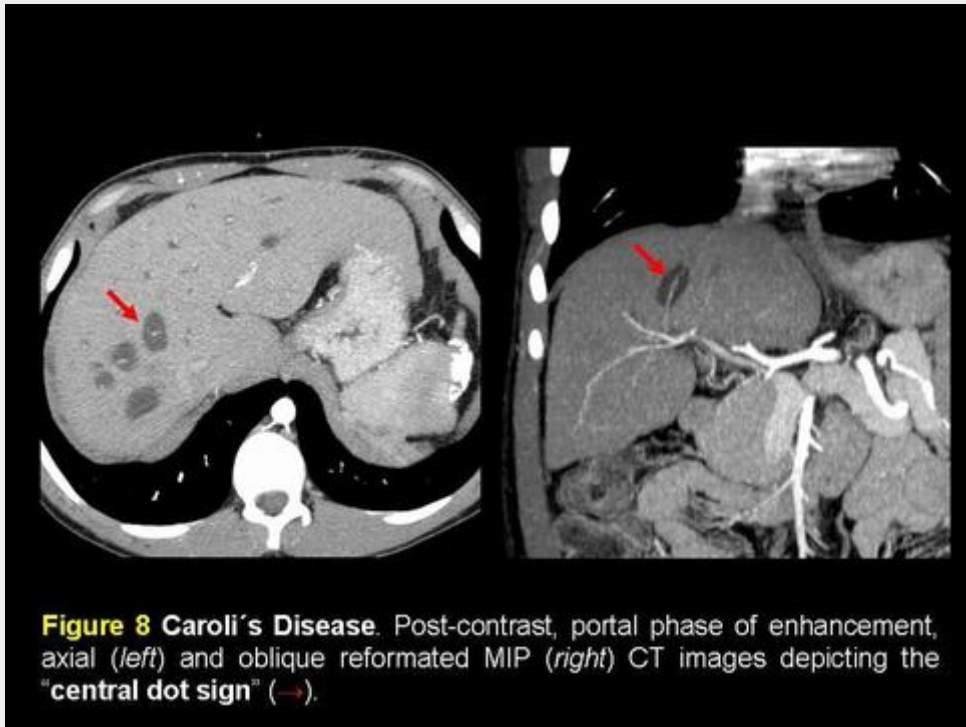
diapositivo50.jpg

## 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



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

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.

### 5. References

References

diapositivo55.jpg

## REFERENCES

Richard J. Rizzo, MD *et al*; "Congenital Abnormalities of the Pancreas and Biliary Tree in Adults"; *RadioGraphics* 1995; 15:49-68

Masahiro Kitami, MD, PhD; "Types and Frequencies of Biliary Tract Variations Associated with a Major Portal Venous Anomaly: Analysis with Multi-Detector Row CT Cholangiography" *Radiology*: Volume 238: Number 1—January 2006

David Blair Macdonald *et al*; "Relationship Between Vascular and Biliary Anatomy in Living liver Donors" *AJR*:185, July 2005

Koenraad J. Mortelé *et al*; "Anatomic Variants of the Biliary Tree: MR Cholangiographic Findings and Clinical Applications" *AJR*:177, August 2001

Yusuf BAYRAKTAR *et al*; "Agenesis of gallbladder and multiple anomalies of the biliary tree in a patient with portal thrombosis: A case report"; *Turk J Gastroenterol* 2006; 17 (3): 212-215

diapositivo56.jpg

## REFERENCES

Erik K. Insko, MD, PhD *et al*; " Benign and Malignant Lesions of the Stomach: Evaluation of CT Criteria for Differentiation", *Radiology* 2003; 228:166

Koenraad J. Mortelé, MD *et al*; "**Multimodality Imaging of Pancreatic and Biliary Congenital Anomalies**"; *RadioGraphics* 2006; 26:715–731

Jinxing Yu *et al*; "**Congenital Anomalies and Normal Variants of the pancreaticobiliary Tract and the Pancreas in Adults: Part 1, Biliary Tract**"; *AJR* 2006; 187:1536–1543

Ozlem Yonem *et al*; "**Clinical characteristics of Caroli's disease**" *World J Gastroenterol* 2007 April 7; 13(13): 1930-1933

## REFERENCES

Ahmet Mesur Halefoglul *et al*; "Magnetic resonance cholangiopancreatography: A useful tool in the evaluation of pancreatic and biliary disorders"; *World J Gastroenterol* 2007 May 14; 13(18): 2529-2534

Hyun Cheol Kim *et al*; "Multiplanar reformations and minimum intensity projections using multi-detector row CT for assessing anomalies and disorders of the pancreaticobiliary tree"; *World J Gastroenterol* 2007 August 21; 13(31): 4177-4184

Tan S S *et al*; "Management of adult choledochal cyst"; *Singapore Med J* 2007; 48 (6) : 524

Tetsuhiro Chiba *et al* "Caroli's disease: central do sign re-examined by CT arteriography and CT during arterial portography" Springer-Verlag 2001

## 6. Author Information

Author Information

## Author information

### **AUTHOR INFORMATION**

Inês Santiago

Serviço de Radiologia do Hospital Infante D. Pedro  
Aveiro, Portugal

[inês\\_agp\\_santiago@hotmail.com](mailto:inês_agp_santiago@hotmail.com)

## Special thanks

### **SPECIAL THANKS**

Nuno Neves, MD  
Luís Semedo, MD

## 7. Mediafiles

### Author information

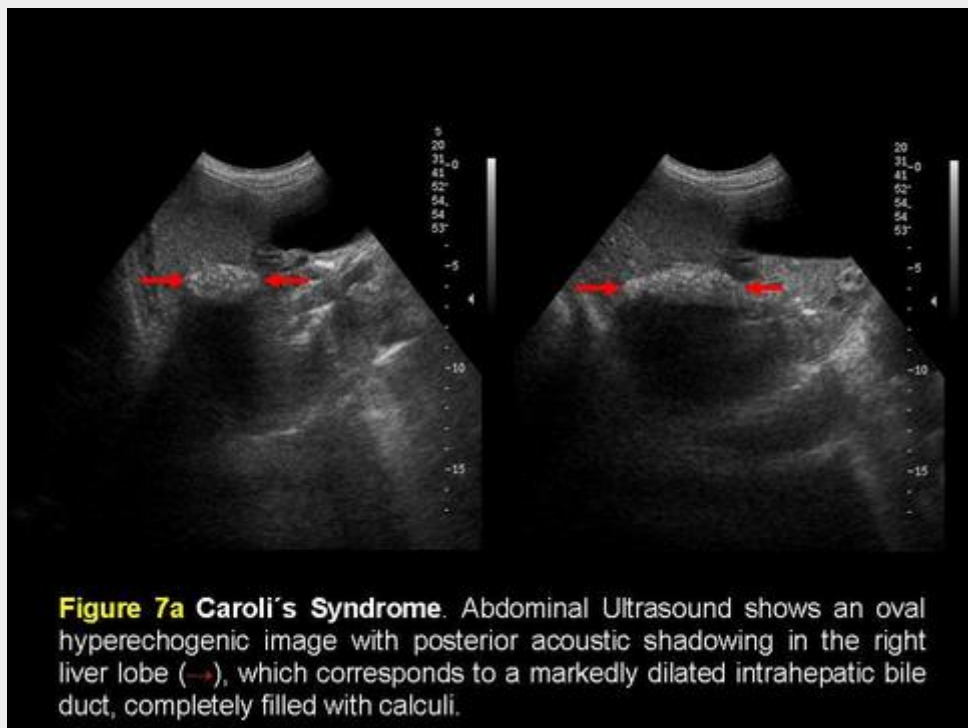
#### AUTHOR INFORMATION

Inês Santiago

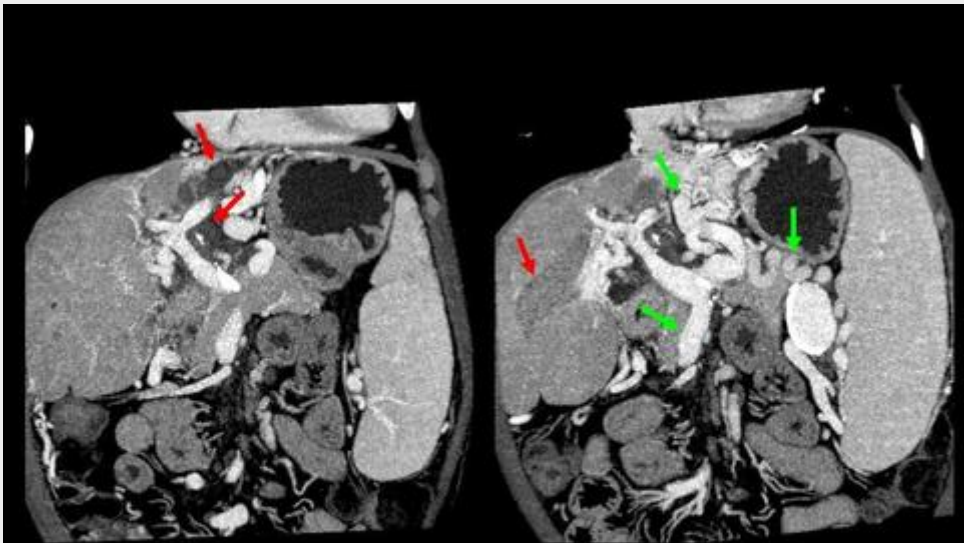
Serviço de Radiologia do Hospital Infante D. Pedro  
Aveiro, Portugal

inês\_agp\_santiago@hotmail.com

### Caroli's Syndrome

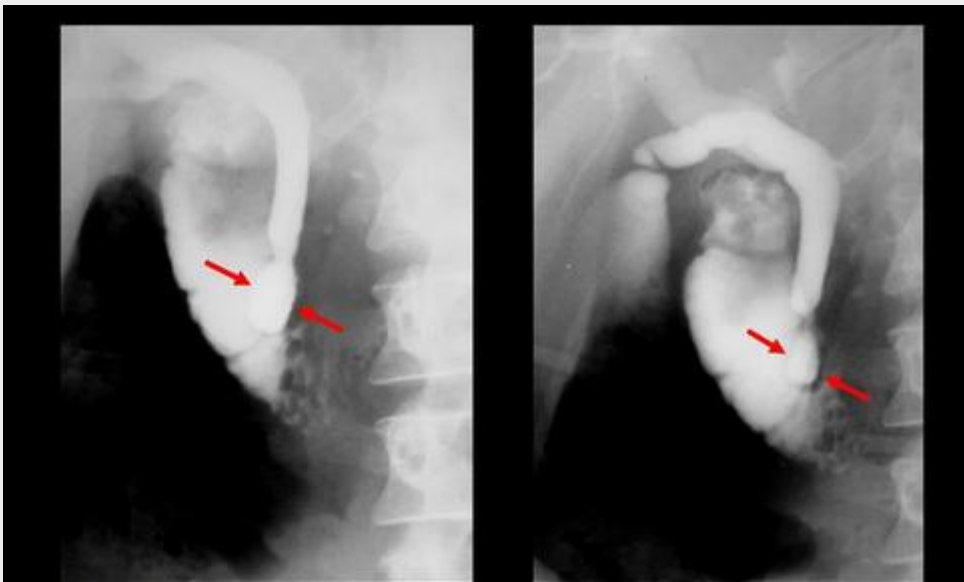


## Caroli's Syndrome



**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 (→).

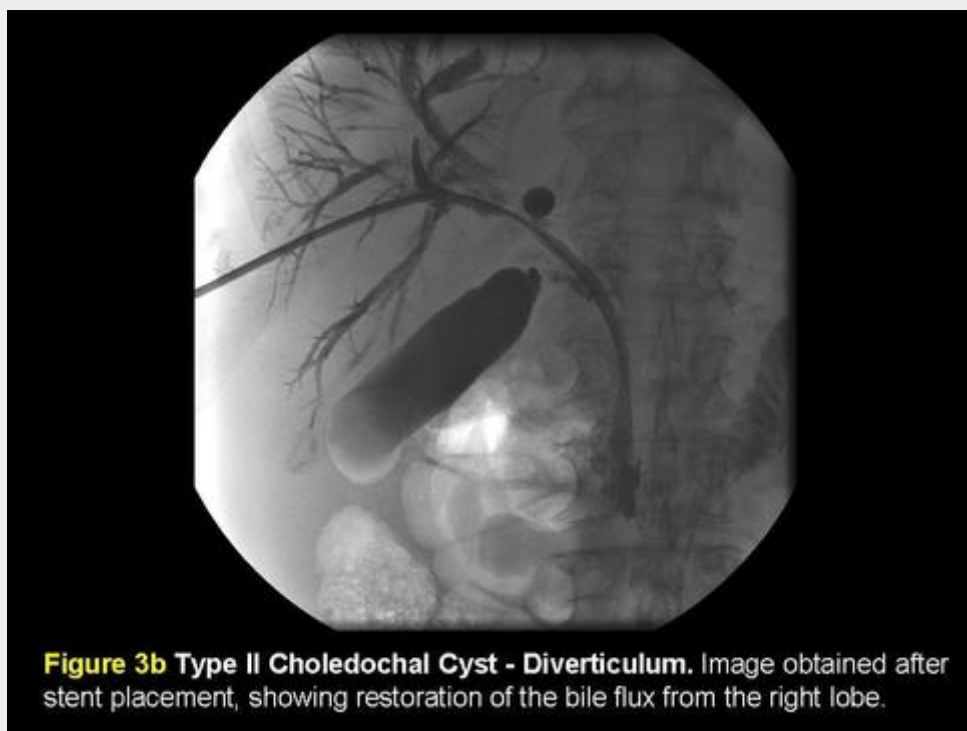
## Definition of choledochal cysts

### BACKGROUND

#### Definition

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

## Diverticulum

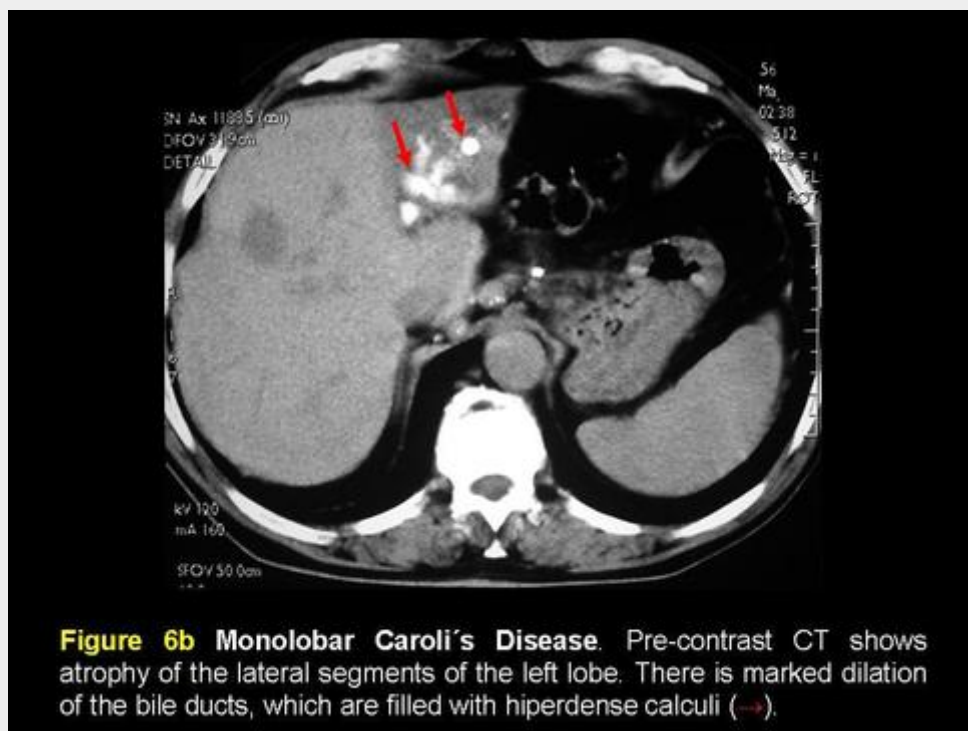


## Learning objectives

### LEARNING OBJECTIVES

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

## Monolobar Caroli's Disease



**Special thanks**

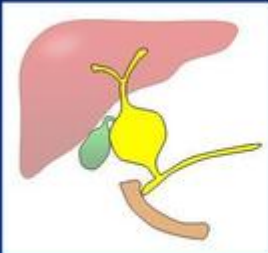


**SPECIAL THANKS**

Nuno Neves, MD  
Luís Semedo, MD

**Type I**

**IMAGING FINDINGS**

**Type I**

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

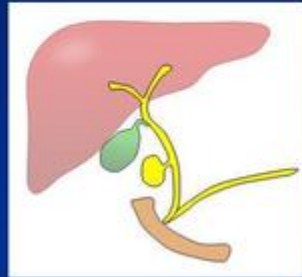


Type II

**IMAGING FINDINGS**

Type II

Diverticulum



Type III

**IMAGING FINDINGS**

Type III

Choledochocele



## Type IV

### IMAGING FINDINGS

#### Type IV

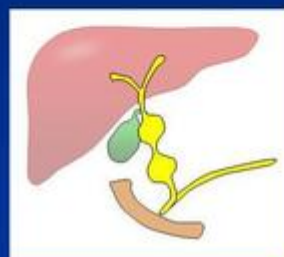
##### Type IV A

Multiple intra and extrahepatic cysts

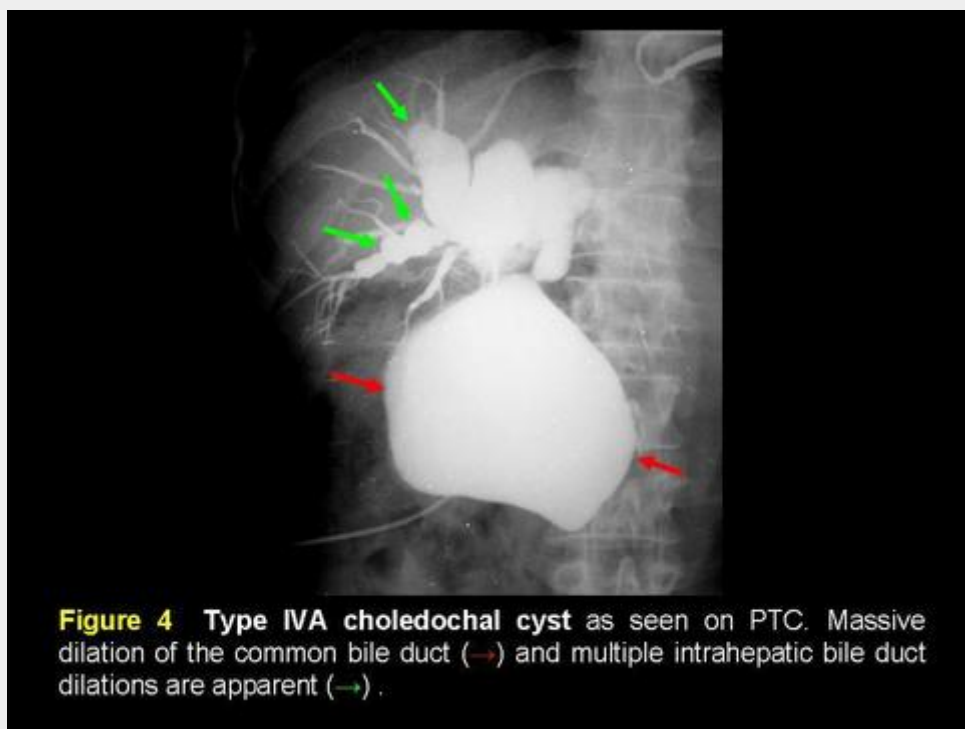


##### Type IV B

Multiple extrahepatic cysts



## Type IVA



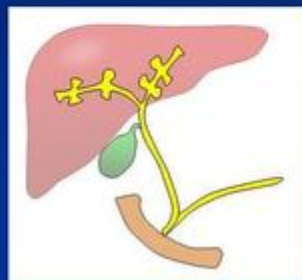
**Figure 4** Type IVA choledochal cyst as seen on PTC. Massive dilation of the common bile duct (→) and multiple intrahepatic bile duct dilations are apparent (→).

## 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

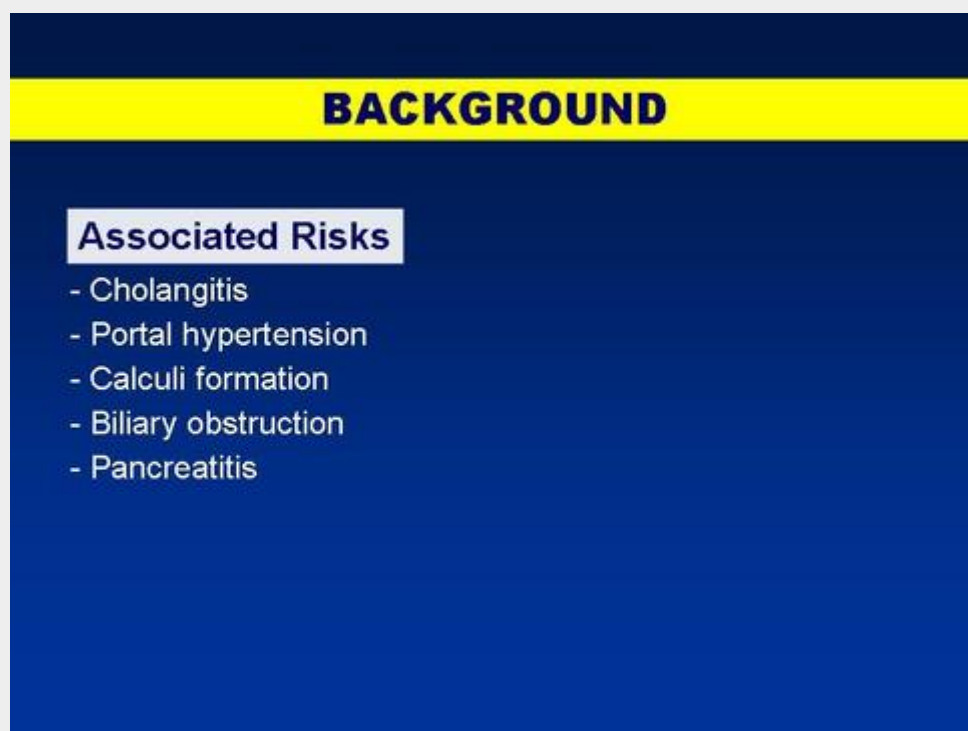
diapositivo8.jpg

**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

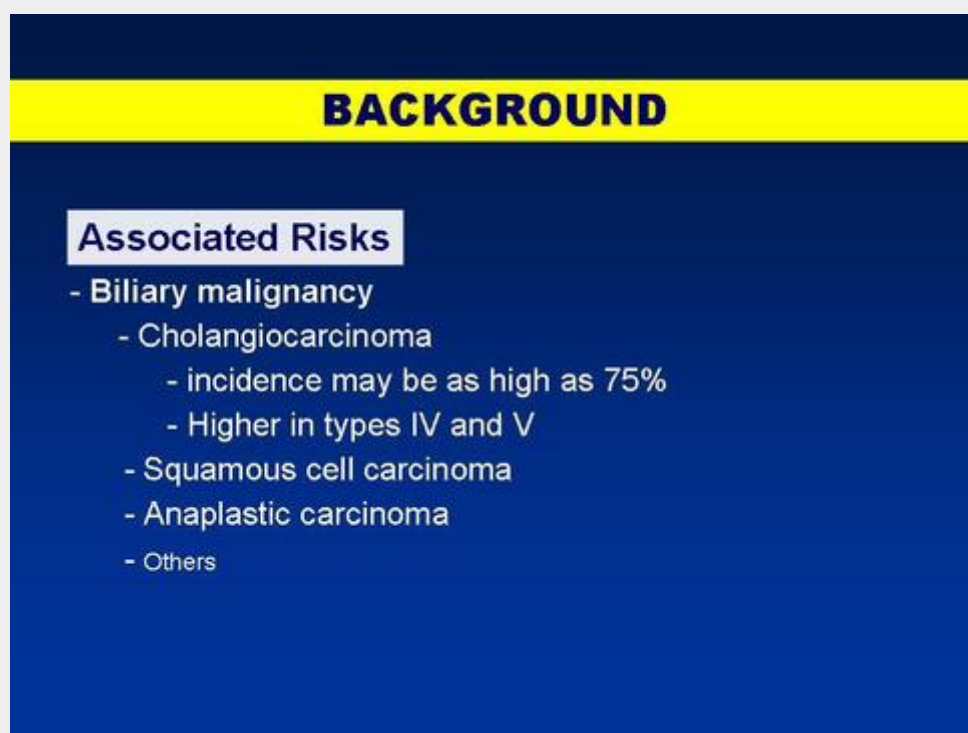


**BACKGROUND**

**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

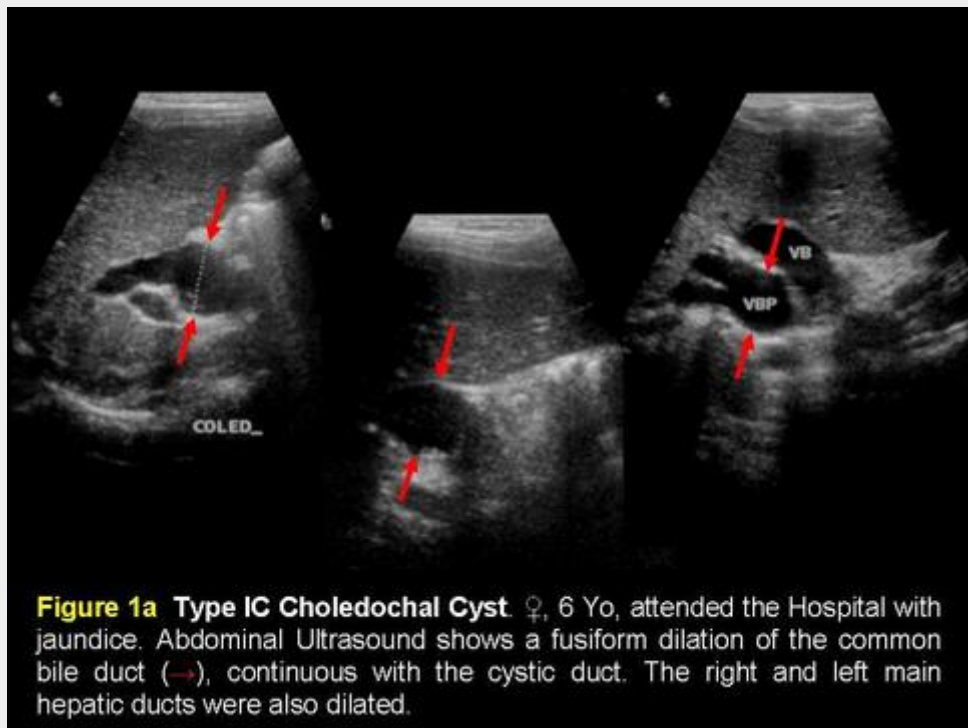
## IMAGING FINDINGS

### Classification

#### *Todani Modification of the Alonso – Lej Classification*

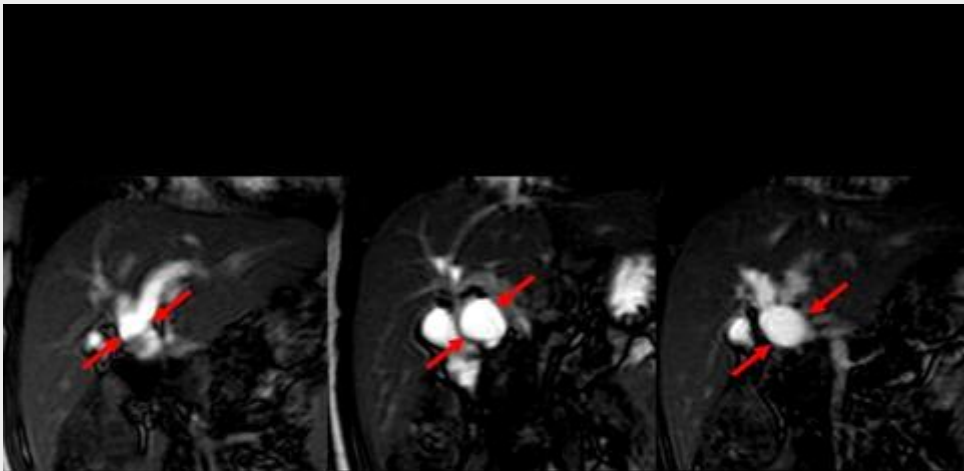
- Type I** Solitary, extrahepatic cyst
- Type II** Extrahepatic duodenal diverticulum
- Type III** Intraduodenal cyst
- Type IV** Extrahepatic and intrahepatic cysts
- Type V** Multiple intrahepatic cysts

diapositivo15.jpg



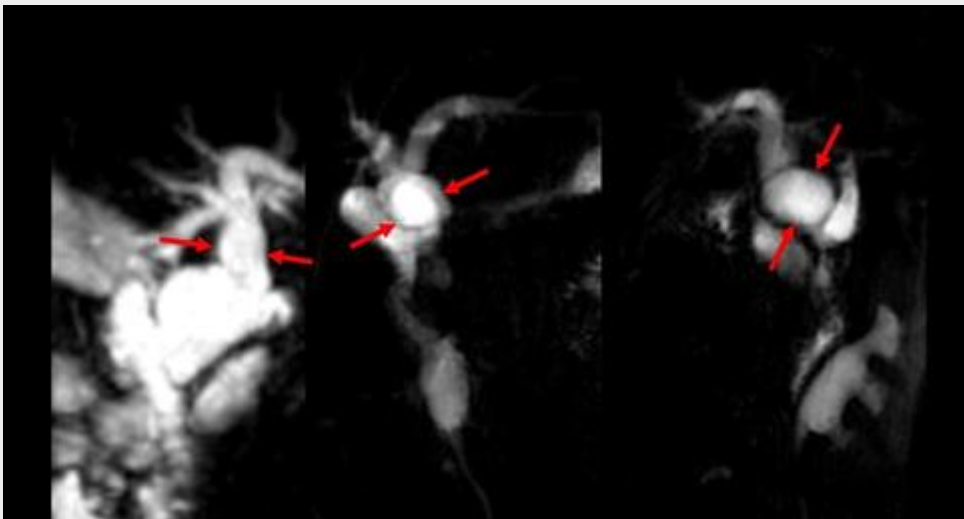
**Figure 1a Type IC Choledochal Cyst.** ♀, 6 Yo, attended the Hospital with jaundice. Abdominal Ultrasound shows a fusiform dilation of the common bile duct (→), continuous with the cystic duct. The right and left main hepatic ducts were also dilated.

diapositivo16.jpg



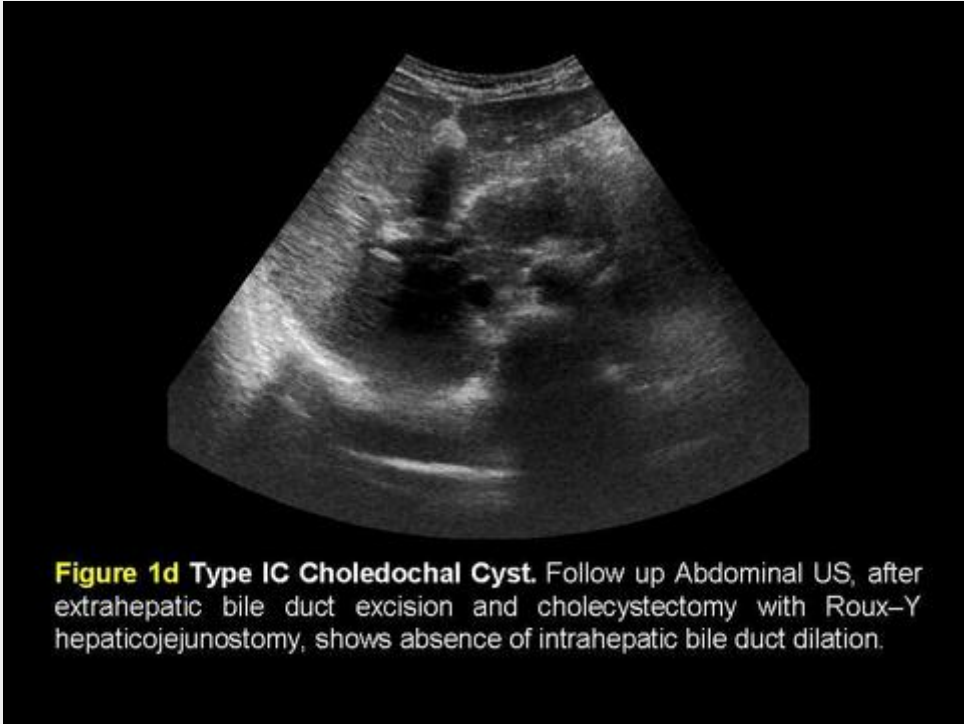
**Figure 1b** Type IC Choledochal Cyst. Magnetic Resonance SSFP sequences show a fusiform dilation of the common hepatic and common bile ducts. The cystic duct drains into the dilated extrahepatic bile duct.

diapositivo17.jpg



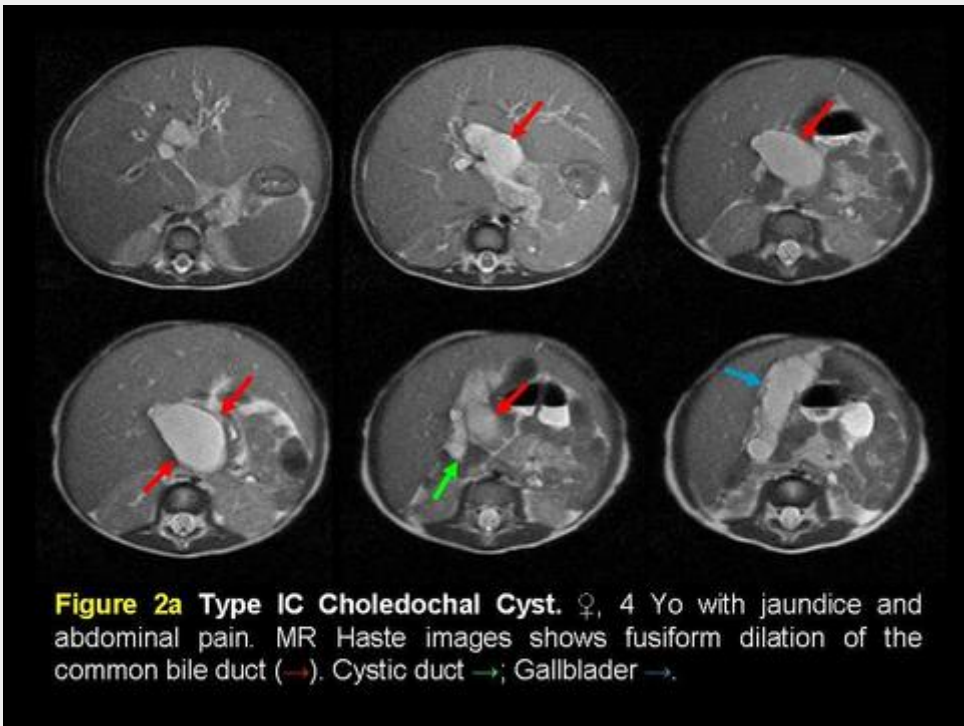
**Figure 1c** Type IC Choledochal Cyst. MR T2 reformatted images show a fusiform dilation of the common hepatic and common bile ducts. The cystic duct drains to the dilated extrahepatic bile duct. These findings correspond to a Type IC choledochal cyst, according to the Todani classification.

diapositivo18.jpg



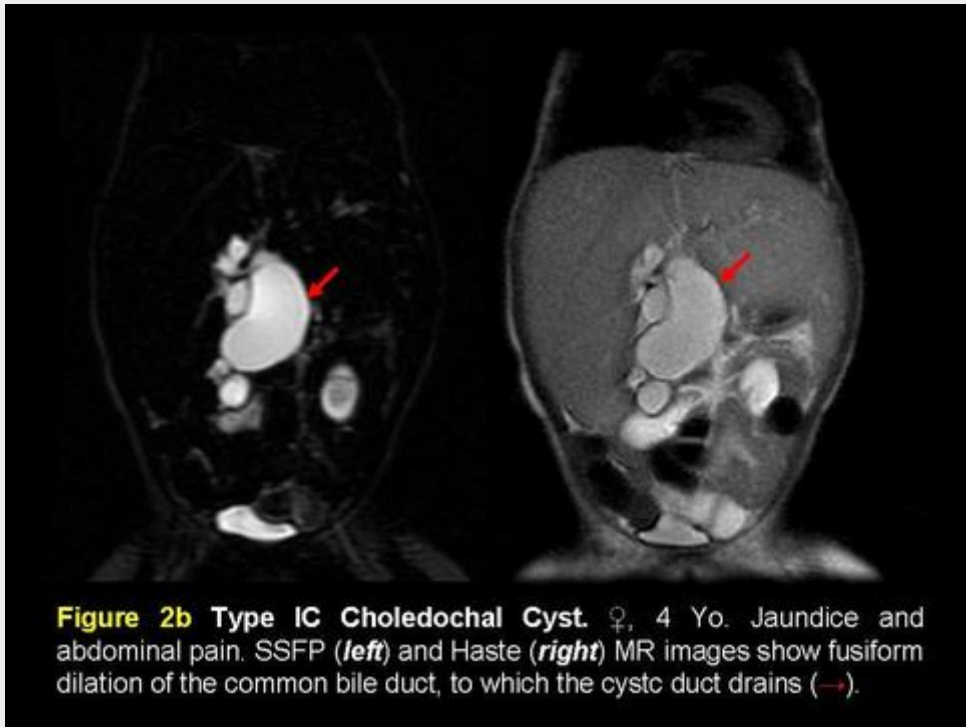
**Figure 1d** Type IC Choledochal Cyst. Follow up Abdominal US, after extrahepatic bile duct excision and cholecystectomy with Roux-Y hepaticojejunostomy, shows absence of intrahepatic bile duct dilation.

diapositivo19.jpg

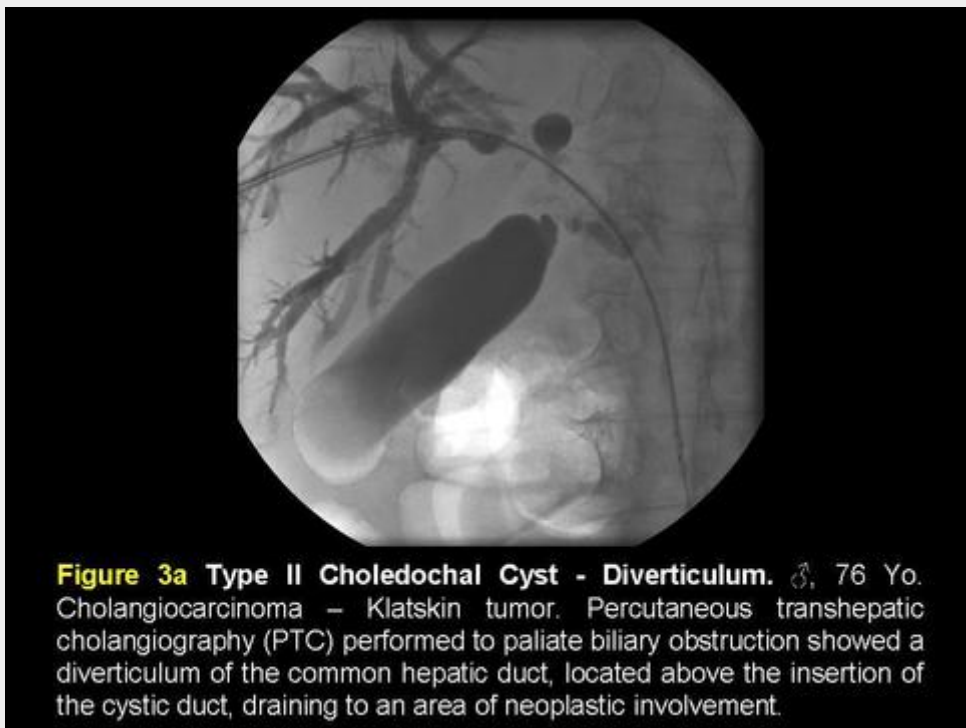


**Figure 2a** Type IC Choledochal Cyst. ♀, 4 Yo with jaundice and abdominal pain. MR Haste images shows fusiform dilation of the common bile duct (→). Cystic duct →; Gallblader →.

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

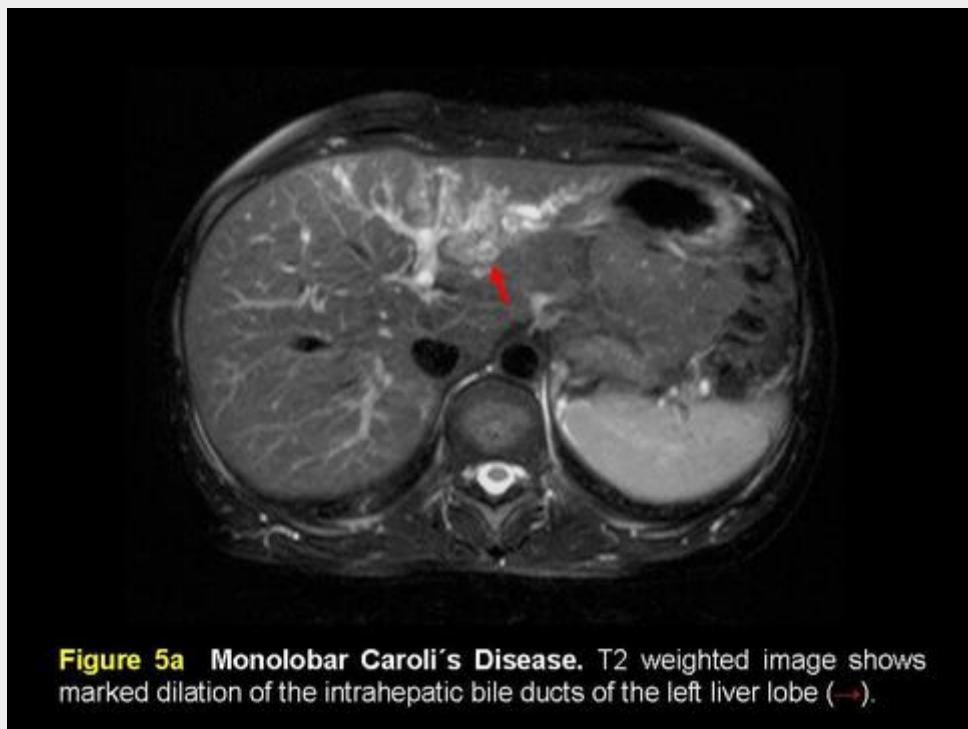
## IMAGING FINDINGS

### Caroli's Disease

#### Differential Diagnosis

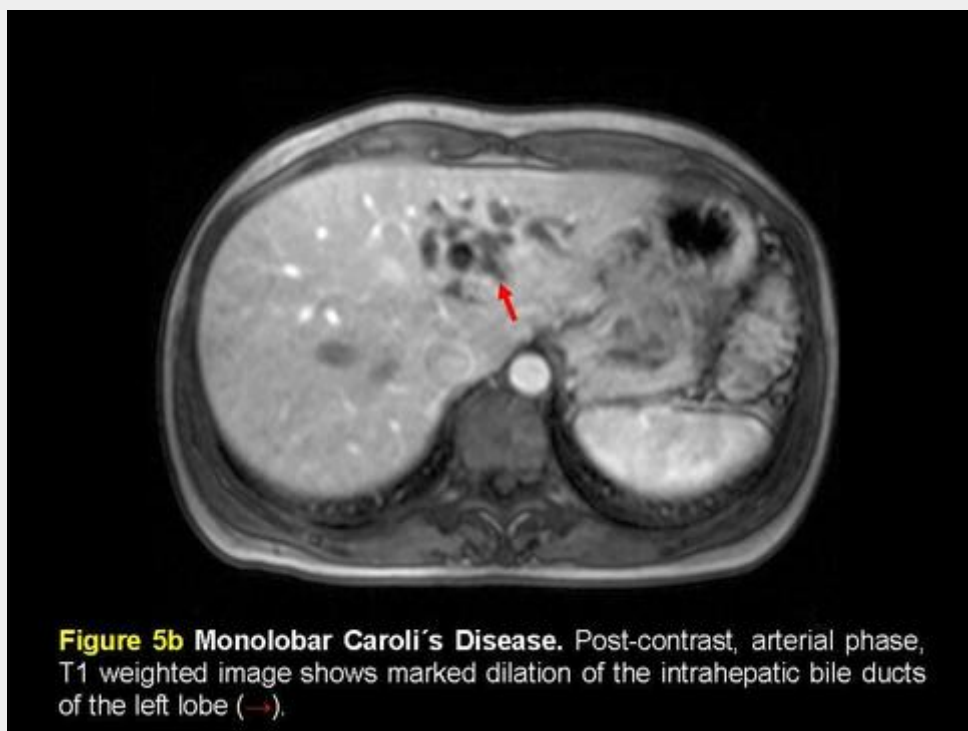
- Polycystic liver disease
- Biliary microhamartomas
- Primary sclerosing cholangitis
- Recurrent pyogenic cholangitis  
(oriental cholangiohepatitis)

diapositivo34.jpg



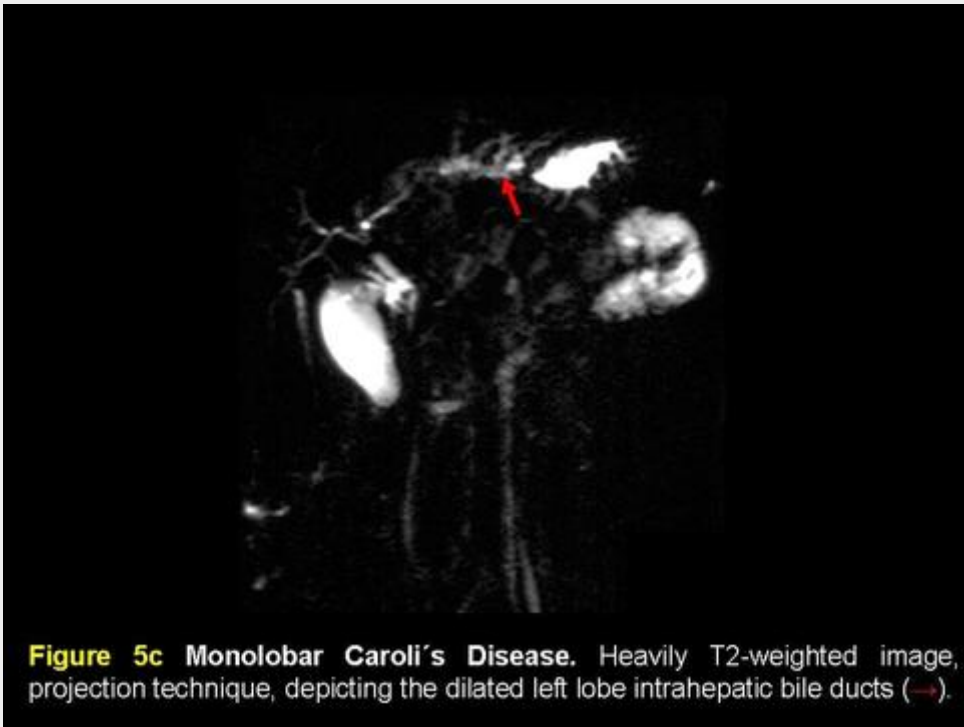
**Figure 5a Monolobar Caroli's Disease.** T2 weighted image shows marked dilation of the intrahepatic bile ducts of the left liver lobe (→).

diapositivo35.jpg

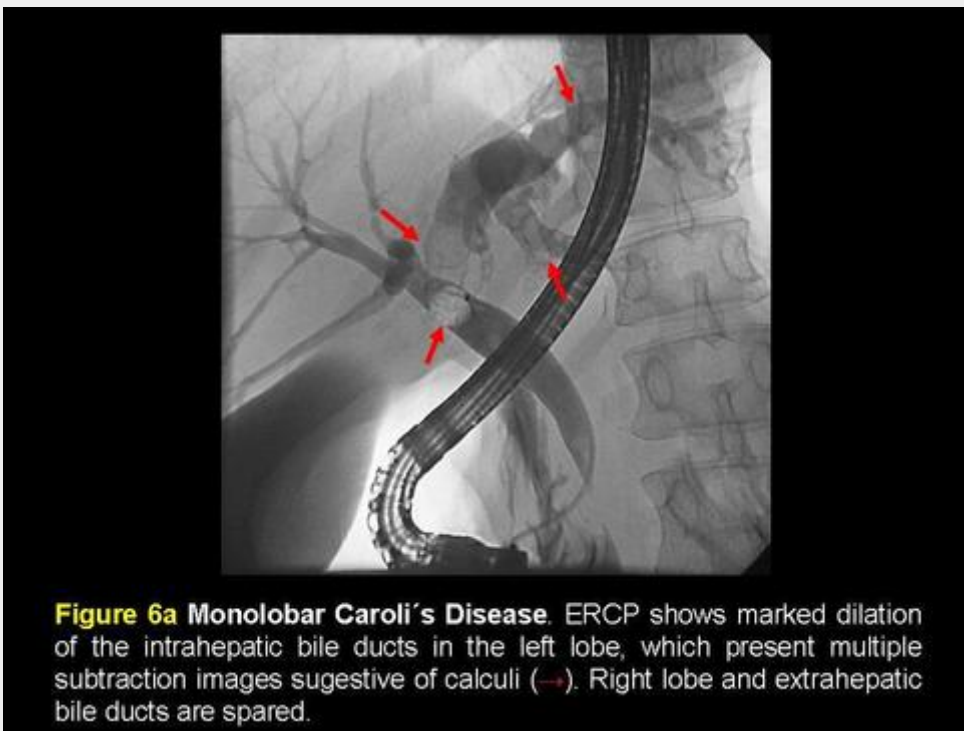


**Figure 5b Monolobar Caroli's Disease.** Post-contrast, arterial phase, T1 weighted image shows marked dilation of the intrahepatic bile ducts of the left lobe (→).

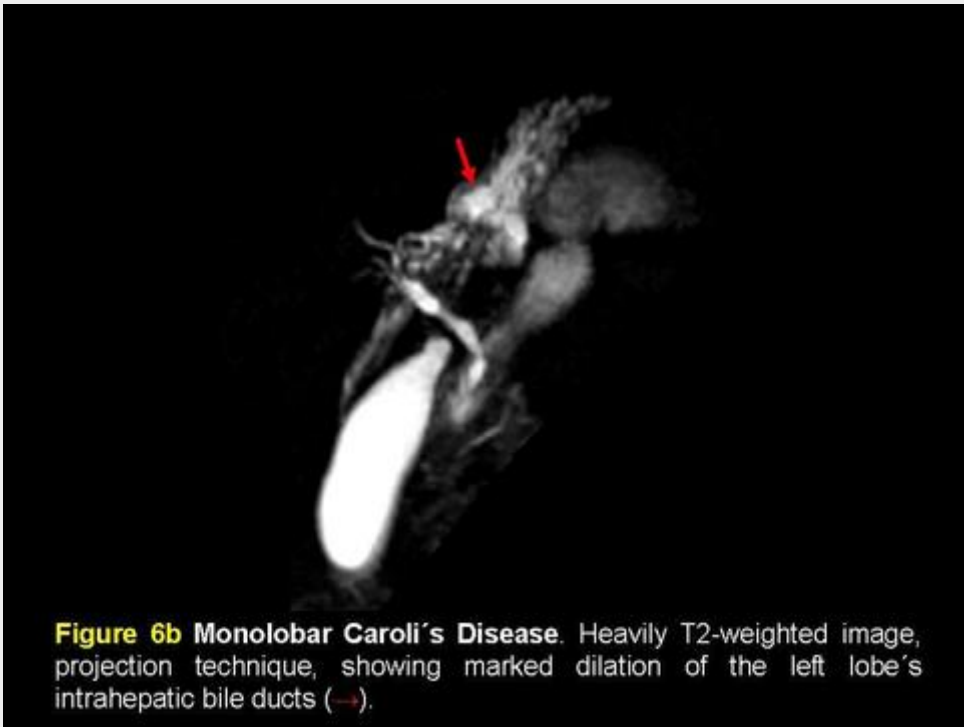
diapositivo36.jpg



diapositivo37.jpg

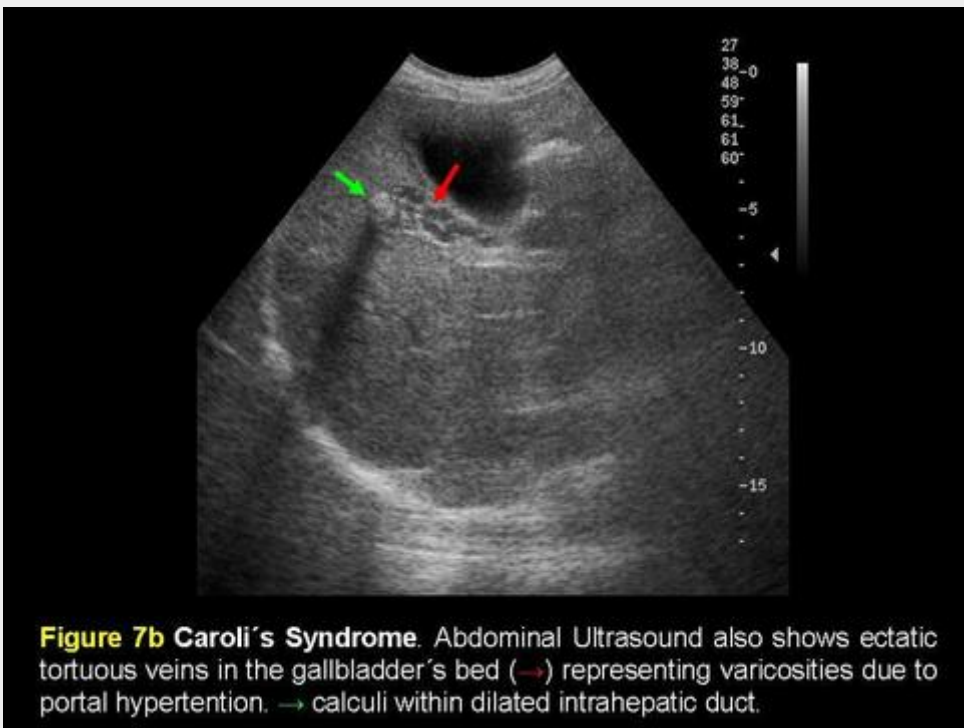


diapositivo39.jpg



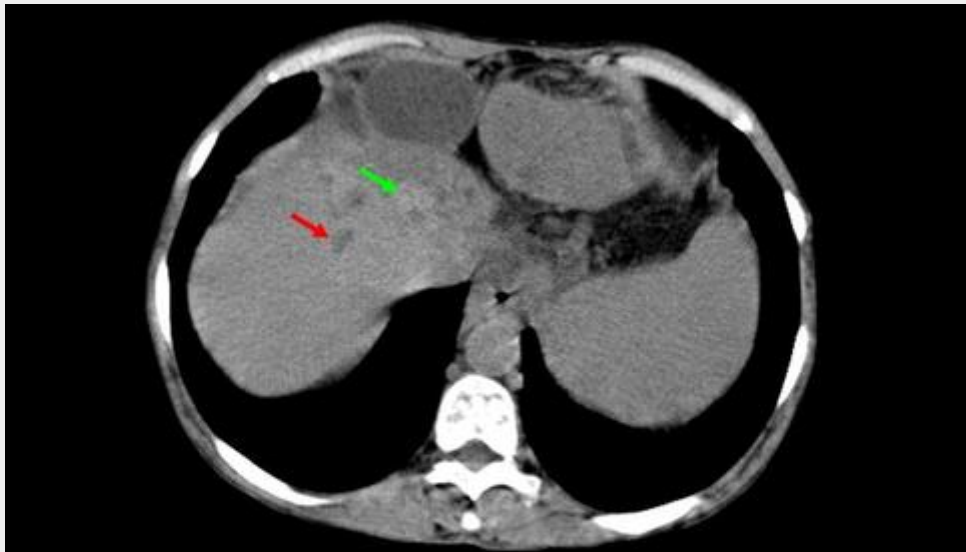
**Figure 6b Monolobar Caroli's Disease.** Heavily T2-weighted image, projection technique, showing marked dilation of the left lobe's intrahepatic bile ducts (→).

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



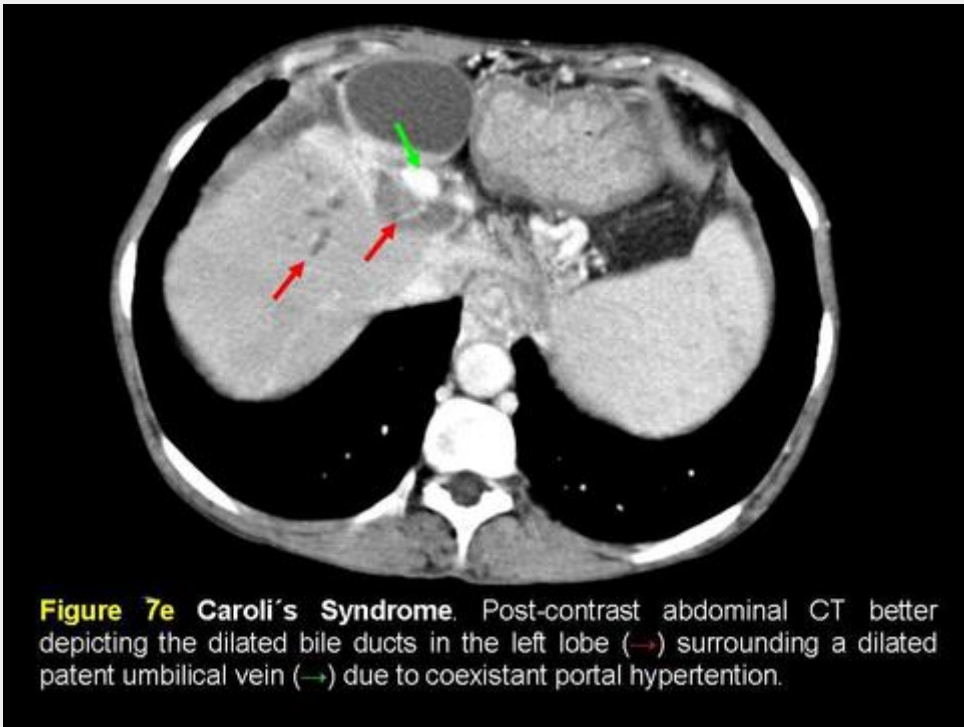
**Figure 7c Caroli's Syndrome.** Pre-contrast abdominal CT. There are dilated bile ducts in the left lobe (→), some of which filled with hyperdense material, corresponding to bile calculi (→).

diapositivo43.jpg

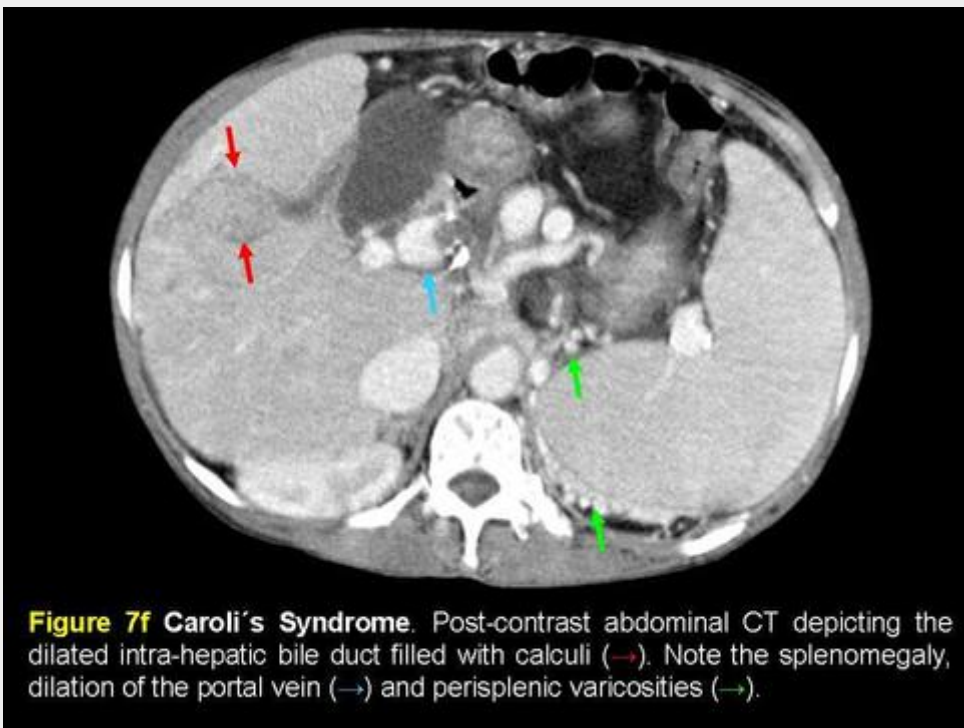


**Figure 7d Caroli's Syndrome.** Pre-contrast abdominal CT. Axial (*left*) and oblique reformatted (*right*) images. There is a markedly dilated bile duct in the right lobe, completely filled with hyperdense material, corresponding to bile calculi (→).

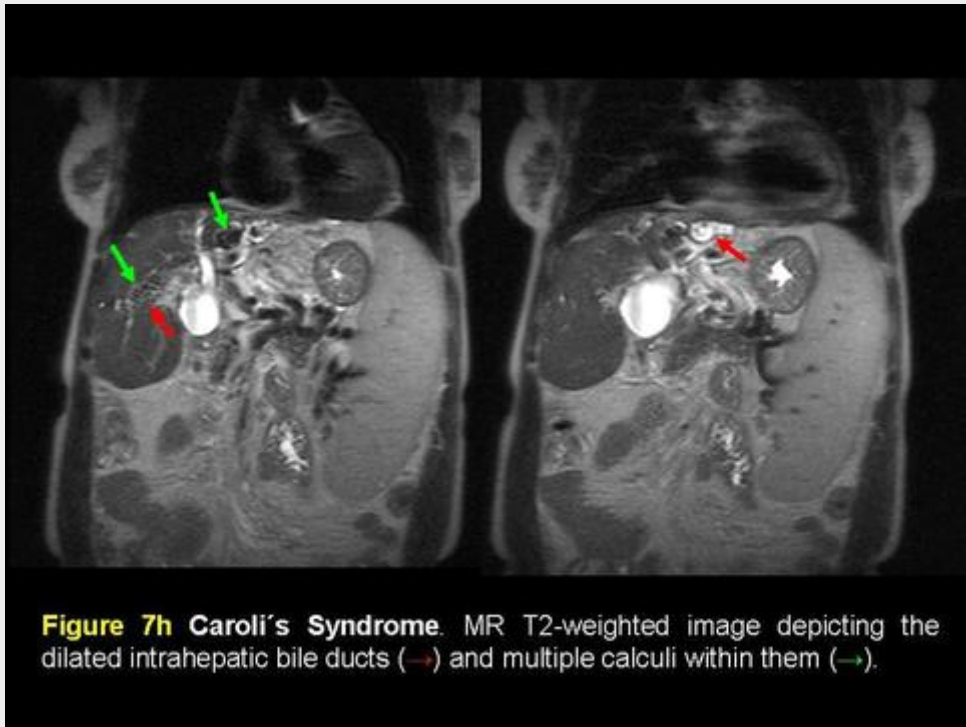
diapositivo44.jpg



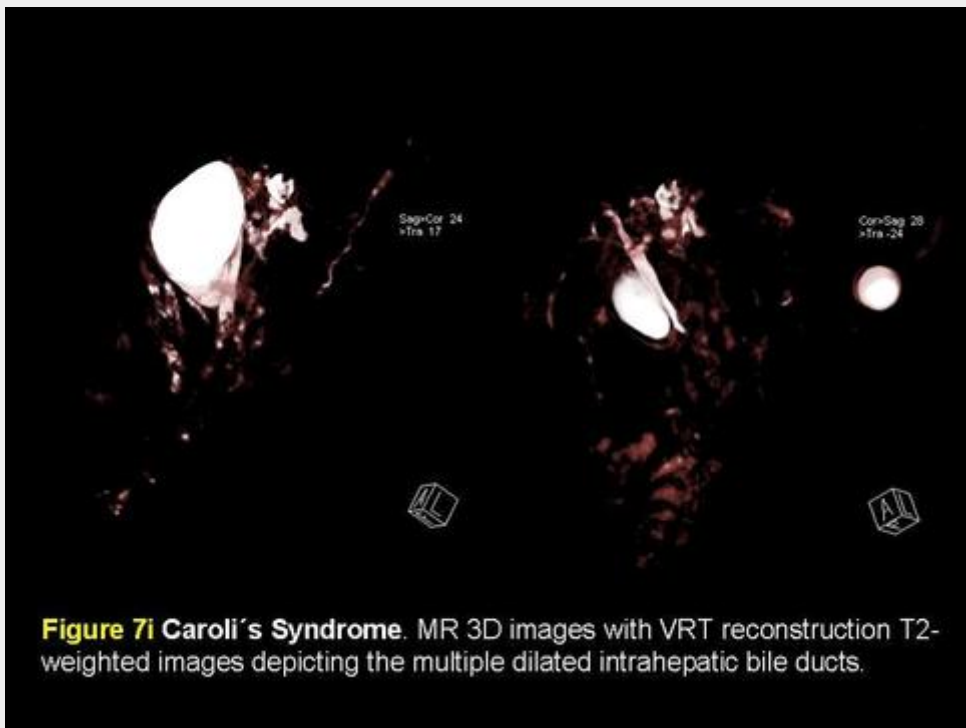
diapositivo45.jpg



diapositivo47.jpg



diapositivo48.jpg



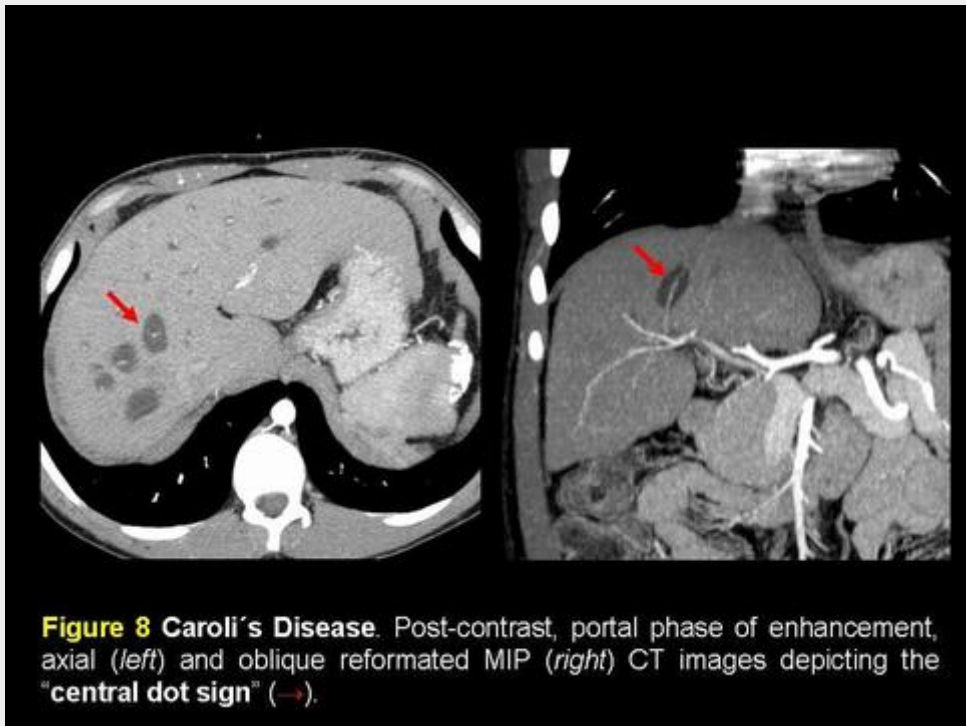
diapositivo50.jpg

## 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

## REFERENCES

Richard J. Rizzo, MD *et al*; "Congenital Abnormalities of the Pancreas and Biliary Tree in Adults"; *RadioGraphics* 1995; 15:49-68

Masahiro Kitami, MD, PhD; "Types and Frequencies of Biliary Tract Variations Associated with a Major Portal Venous Anomaly: Analysis with Multi-Detector Row CT Cholangiography" *Radiology*: Volume 238: Number 1—January 2006

David Blair Macdonald *et al*; "Relationship Between Vascular and Biliary Anatomy in Living liver Donors" *AJR*:185, July 2005

Koenraad J. Mortelé *et al*; "Anatomic Variants of the Biliary Tree: MR Cholangiographic Findings and Clinical Applications" *AJR*:177, August 2001

Yusuf BAYRAKTAR *et al*; "Agenesis of gallbladder and multiple anomalies of the biliary tree in a patient with portal thrombosis: A case report"; *Turk J Gastroenterol* 2006; 17 (3): 212-215

diapositivo56.jpg

## REFERENCES

Erik K. Insko, MD, PhD *et al*; " Benign and Malignant Lesions of the Stomach: Evaluation of CT Criteria for Differentiation", *Radiology* 2003; 228:166

Koenraad J. Mortelé, MD *et al*; "**Multimodality Imaging of Pancreatic and Biliary Congenital Anomalies**"; *RadioGraphics* 2006; 26:715–731

Jinxing Yu *et al*; "**Congenital Anomalies and Normal Variants of the pancreaticobiliary Tract and the Pancreas in Adults: Part 1, Biliary Tract**"; *AJR* 2006; 187:1536–1543

Ozlem Yonem *et al*; "**Clinical characteristics of Caroli's disease**" *World J Gastroenterol* 2007 April 7; 13(13): 1930-1933

## REFERENCES

Ahmet Mesur Halefoglul *et al*; "Magnetic resonance cholangiopancreatography: A useful tool in the evaluation of pancreatic and biliary disorders"; *World J Gastroenterol* 2007 May 14; 13(18): 2529-2534

Hyun Cheol Kim *et al*; "Multiplanar reformations and minimum intensity projections using multi-detector row CT for assessing anomalies and disorders of the pancreaticobiliary tree"; *World J Gastroenterol* 2007 August 21; 13(31): 4177-4184

Tan S S *et al*; "Management of adult choledochal cyst"; *Singapore Med J* 2007; 48 (6) : 524

Tetsuhiro Chiba *et al* "Caroli's disease: central do sign re-examined by CT arteriography and CT during arterial portography" Springer-Verlag 2001