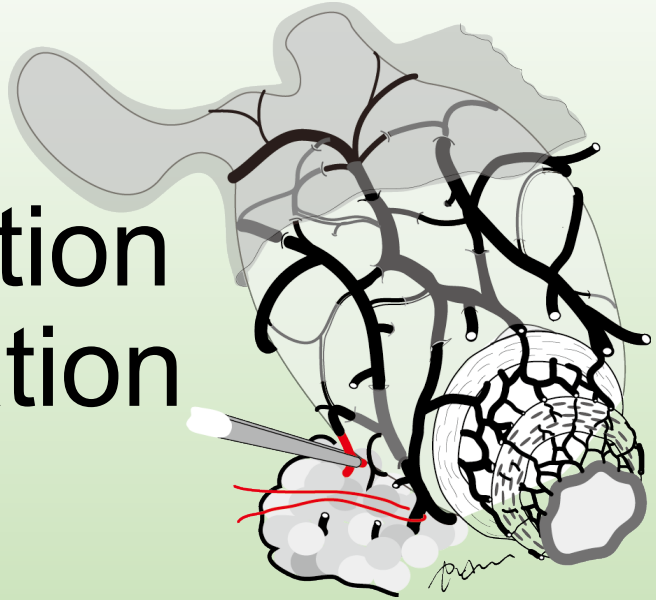
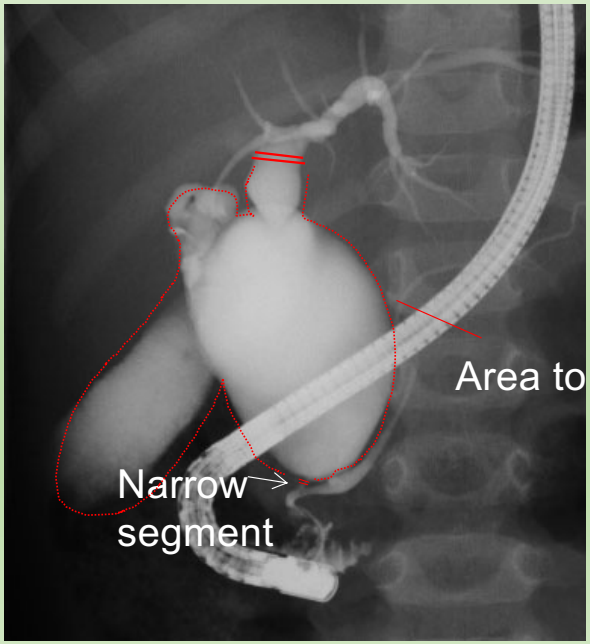
Atlas of Pancreaticobiliary Maljunction & Congenital Biliary Dilatation





Japanese Study Group on Pancreaticobiliary Maljunction (JSGPBM)
Scientific Committee

Editor-in-Chief, Kenitiro Kaneko
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Yoshiki Morotomi

In collaboration with Naoto Urushihara

Preface

Pancreaticobiliary maljunction (PBM) was discovered as a malformation accompanying choledochal cysts. In Japan, where choledochal cysts are common, the concept of PBM is widespread, but it seems unfamiliar in other countries where choledochal cysts are rare. However, the term “choledochal cyst” includes various entities of congenitally dilated bile ducts with or without PBM. Almost all choledochal cysts seen in Asia have localized dilatation of the bile duct and PBM, where PBM plays a major role in pathophysiology. In countries outside Asia, the frequency of choledochal cysts without PBM is relatively high, leading to confusion when discussing “ choledochal cysts”. We felt it necessary to differentiate this type with PBM from other types without PBM and defined the common type with PBM in Asia as “congenital biliary dilatation”.

PBM is also observed in patients without bile duct dilatation. Gallbladder cancers arise at an extraordinarily high frequency in these non-dilated PBM patients. However, the definition of non-dilatation has been unclear. We have developed ultrasonographic criteria for bile ducts of normal diameter, which we present here.

In this Atlas, we attempt to demonstrate clear and precise explanations of PBM and congenital biliary dilatation and hope this Atlas will help people unfamiliar to this field. The contents of the Atlas are free to use providing the source (JSGPBM Atlas of PBM & CBD) is cited.

September 2022
Editor-In-Chief, Kenitiro Kaneko

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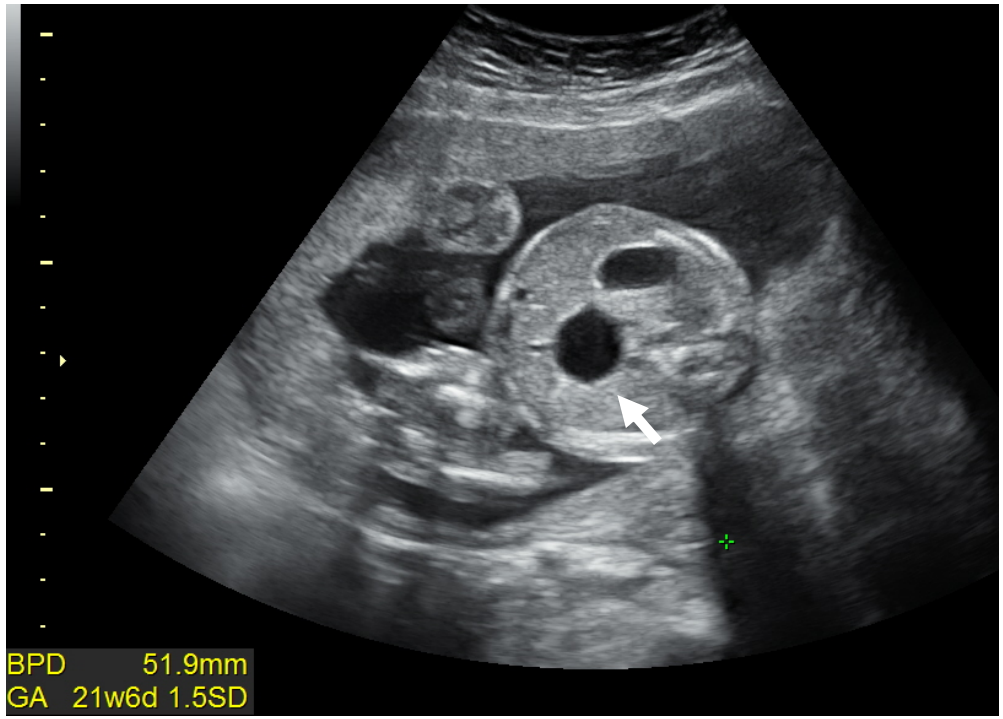
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Expert

Prenatal diagnosis
Transition to biliary atresia

First version, September 2024

Both congenital biliary dilatation (CBD) and biliary atresia (BA) are found prenatally as a biliary cyst



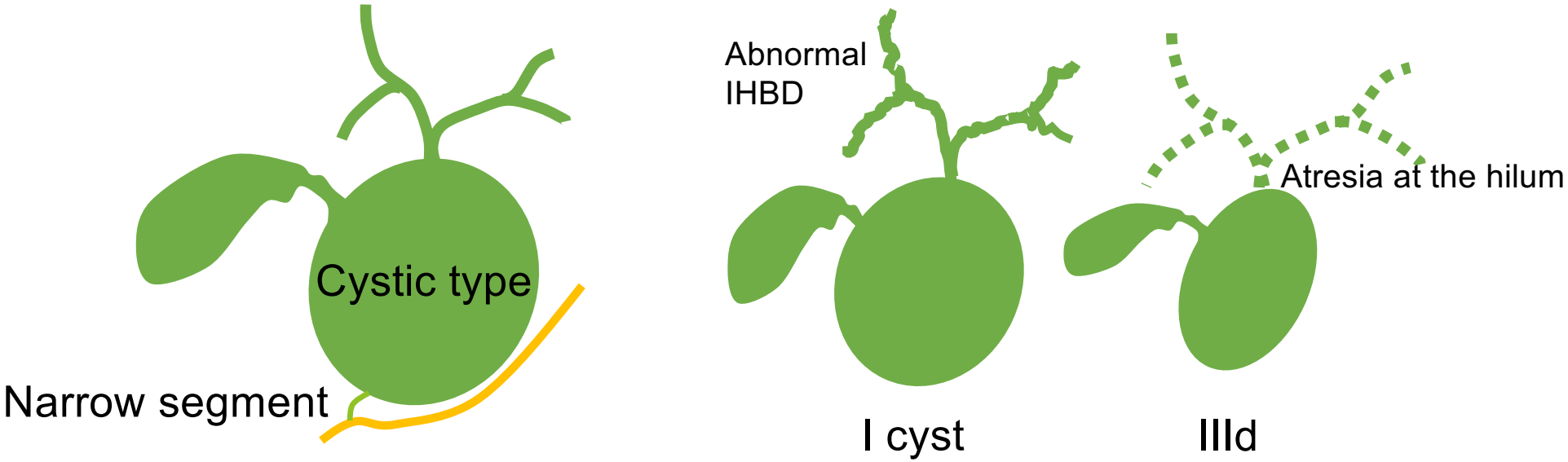
Differential diagnosis is difficult even after birth

Congenital biliary dilatation?
or
Biliary atresia?

Why?

This fetal ultrasonogram showed a connection between the extrahepatic cyst and the intrahepatic bile ducts. However, type IIId biliary atresia was diagnosed after birth.

Fetal biliary cysts present a similar form regardless of final diagnoses



Congenital biliary dilatation

Hepatic fibrosis is prone to occur

Cystic biliary atresia

Hepatic fibrosis occurs inevitably

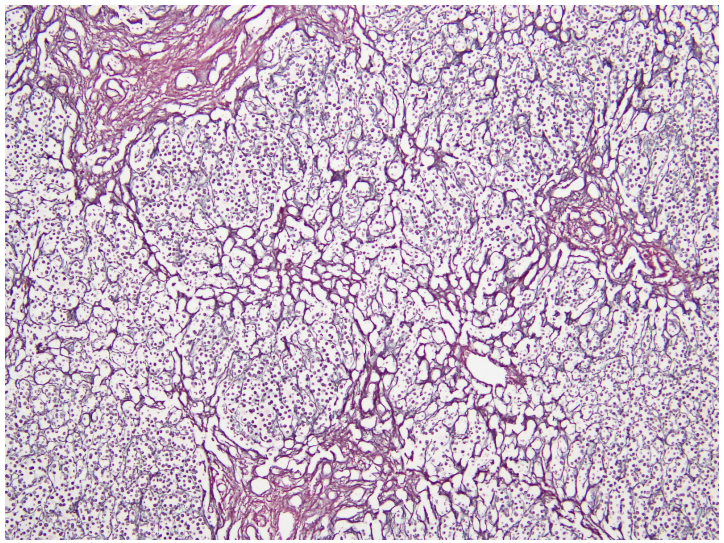
Biliary obstruction causes cholangiopathy specific to neonates and early infants.

Prenatally-diagnosed CBD and hepatic fibrosis

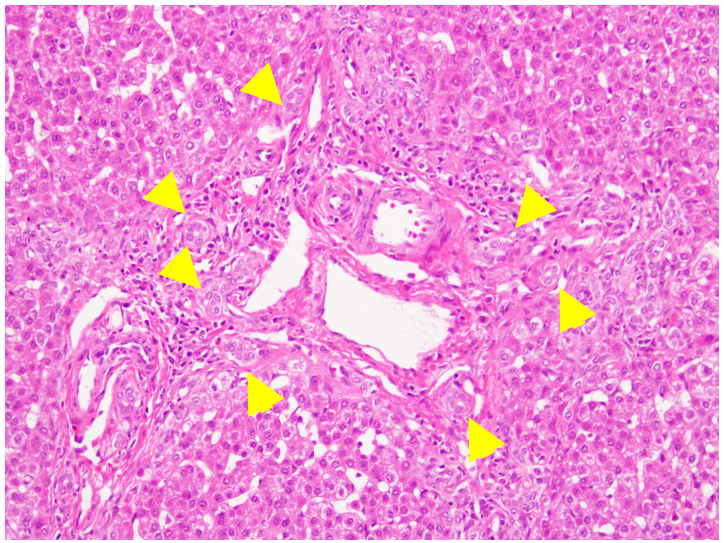
- These cases are prone to hepatic fibrosis.
- Fibrosis can occur even in asymptomatic cases.
- The fibrosis improves after bile duct resection, suggesting that the fibrosis is reversible.

Diagnosis at 29 weeks‘ gestation. The BD was resected upon the onset of acholic stool at 6 months.

Liver pathology shows the same cholangiopathy as seen in biliary atresia.



Bridging F1-2

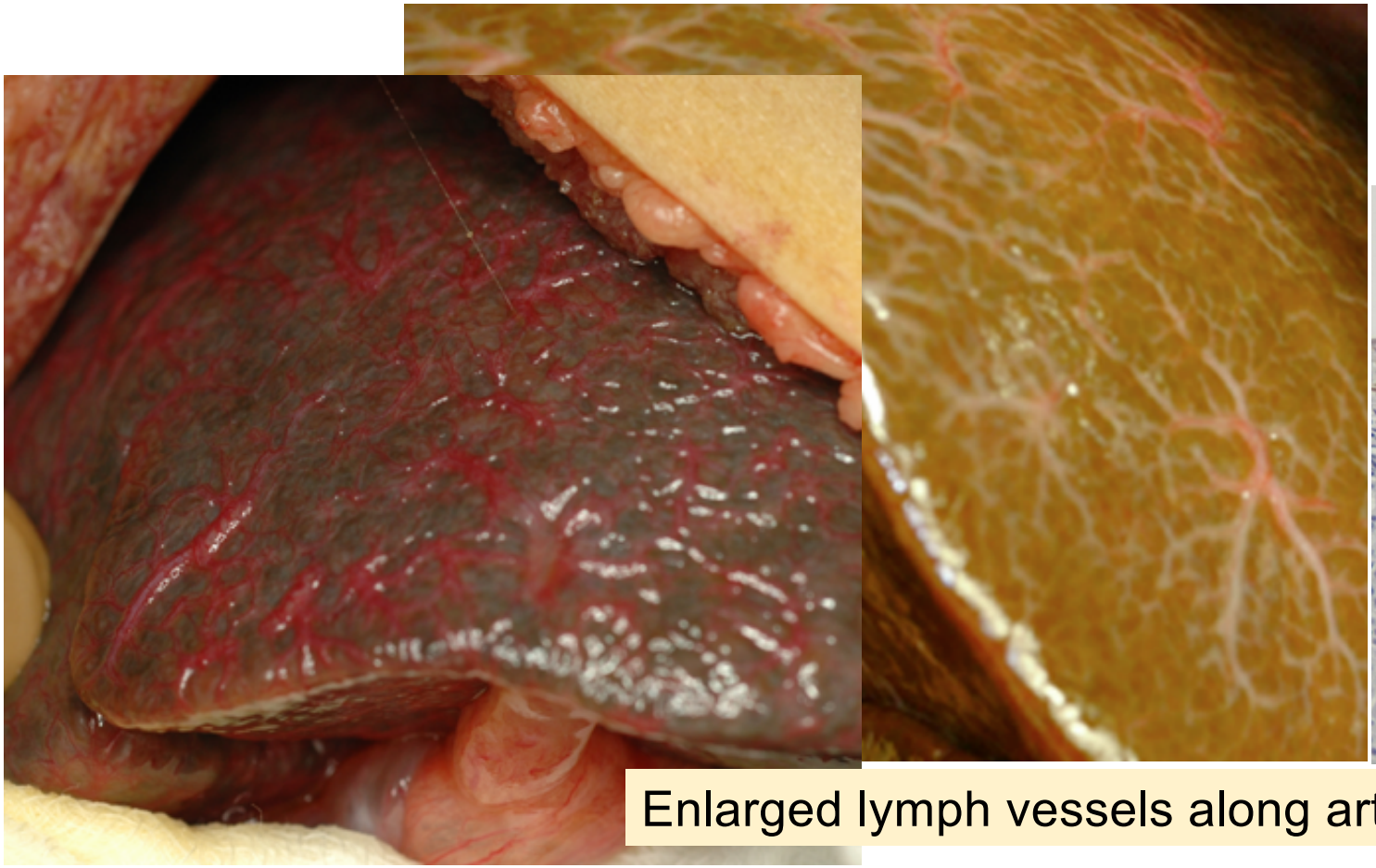


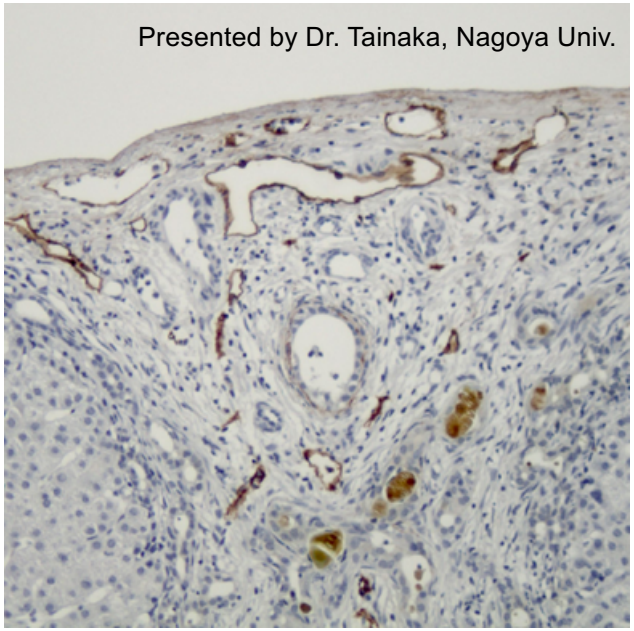
Cholangiocyte proliferation

Makibishi sign specific to biliary atresia



Makibishi, ninjas' tool





Presented by Dr. Tainaka, Nagoya Univ.

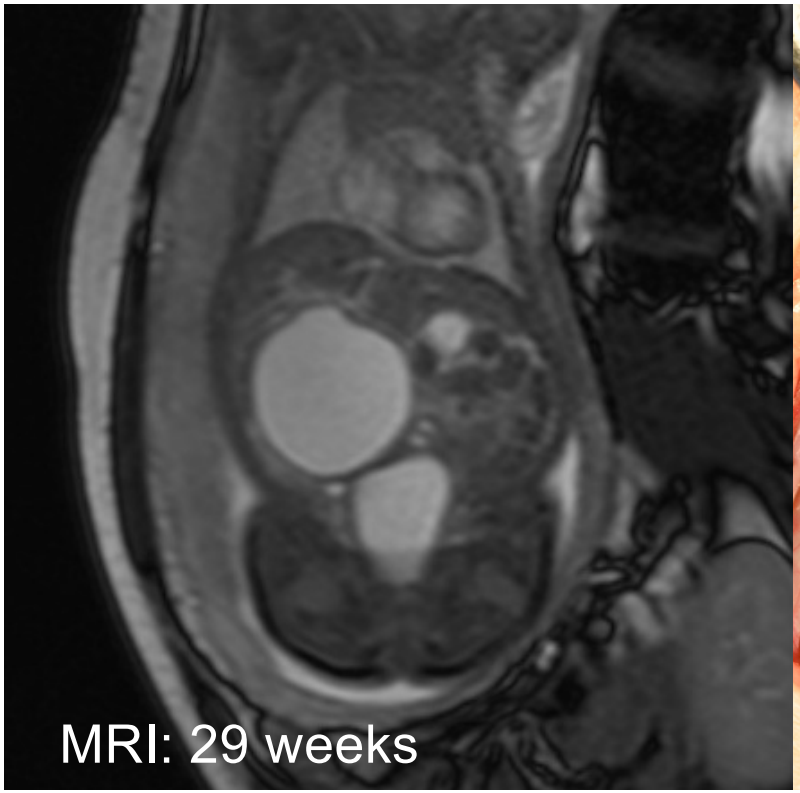
Enlarged lymph vessels along arteries on the liver surface

Case 48

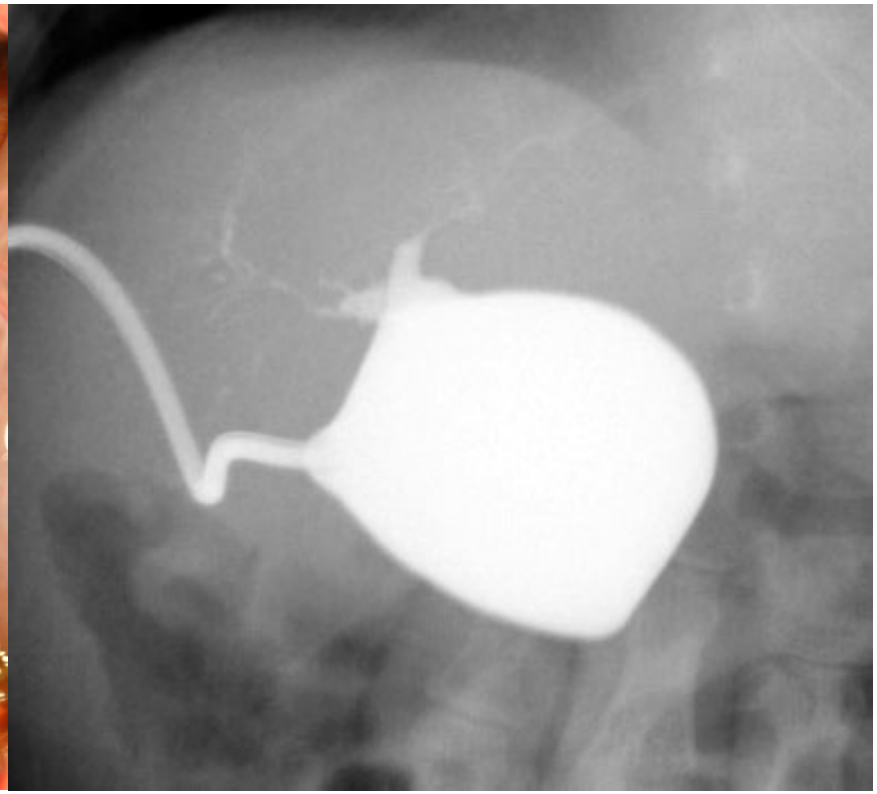
Type I cyst biliary atresia

Makibishi sign

Treelike intrahepatic BDs





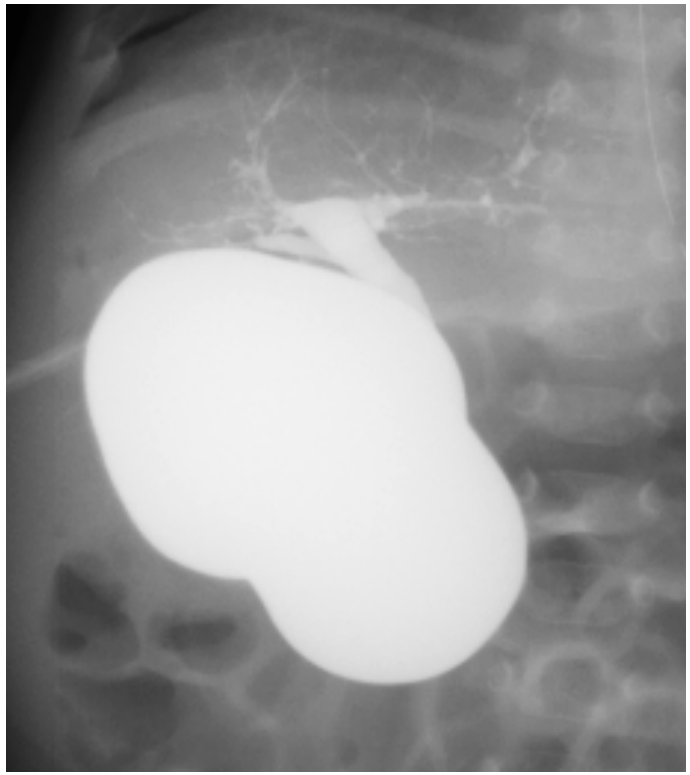


Contrast material did not go out into the duodenum.

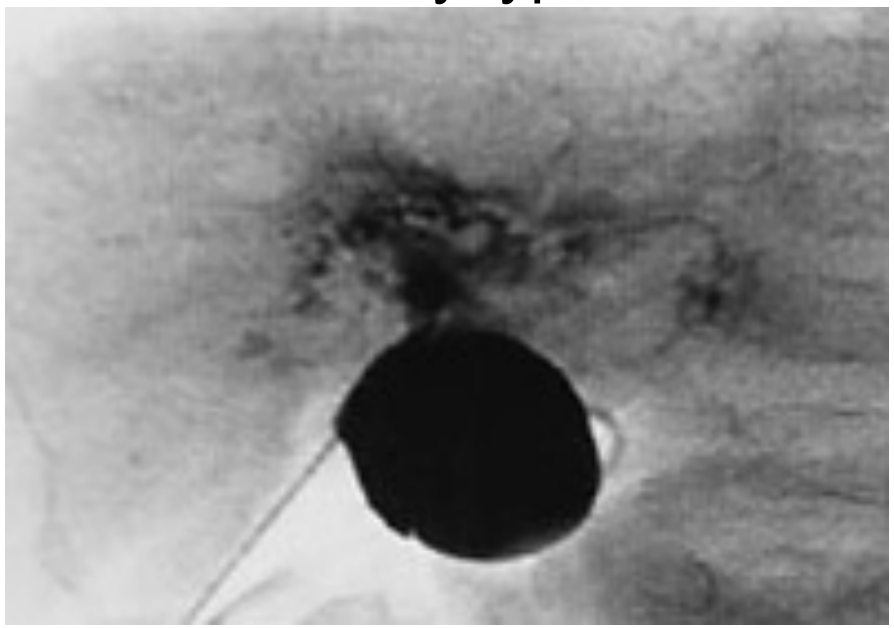
Type I cyst biliary atresia

Cholangiography findings:
- No flow into the duodenum
- Abnormal IHBD shapes

Treelike type



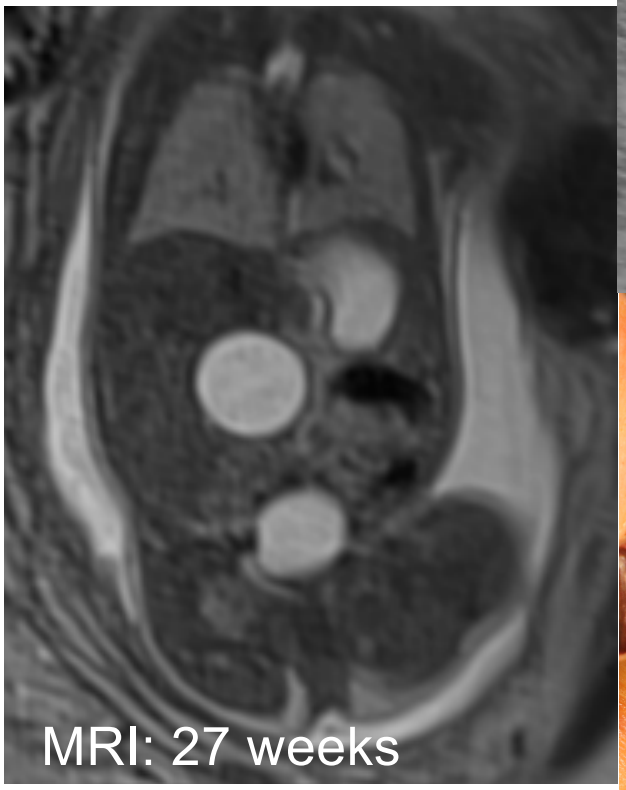
Cloudy type

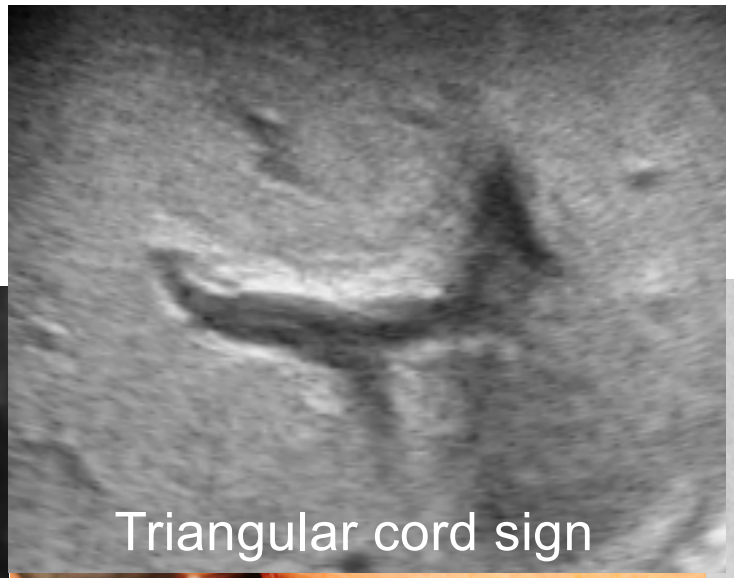


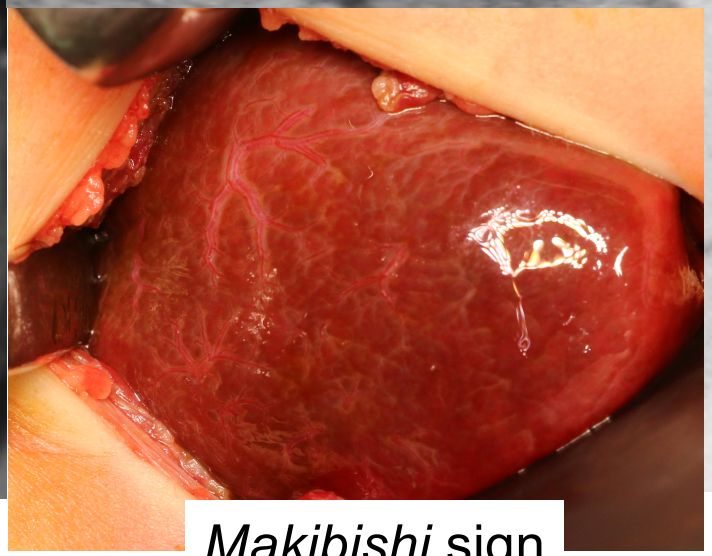
Nio M, et al. Long-term outcome in type I biliary atresia. *J Pediatr Surg.* 2006;41:1973–5.

Case 49

Type IIId BA



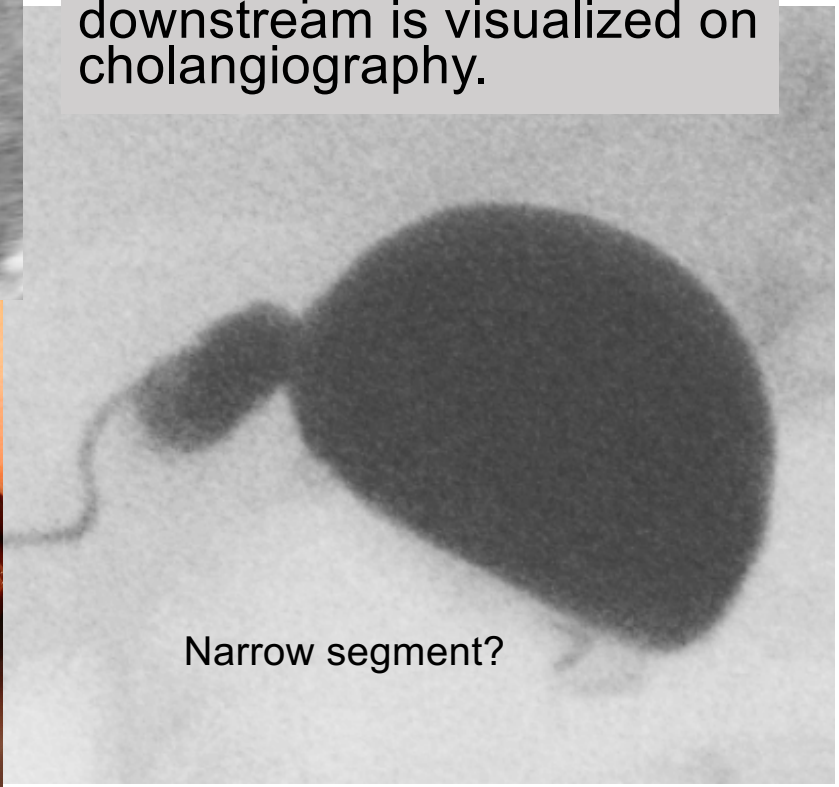




Makibishi sign

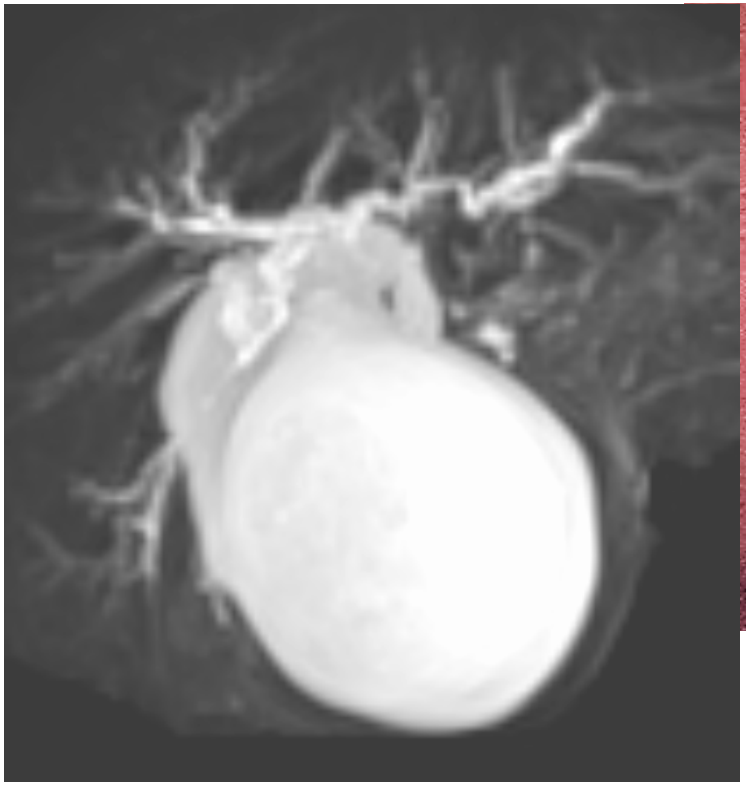
Atresia at the hepatic hilum requires the Kasai operation.

Neither upstream nor downstream is visualized on cholangiography.



Case 50

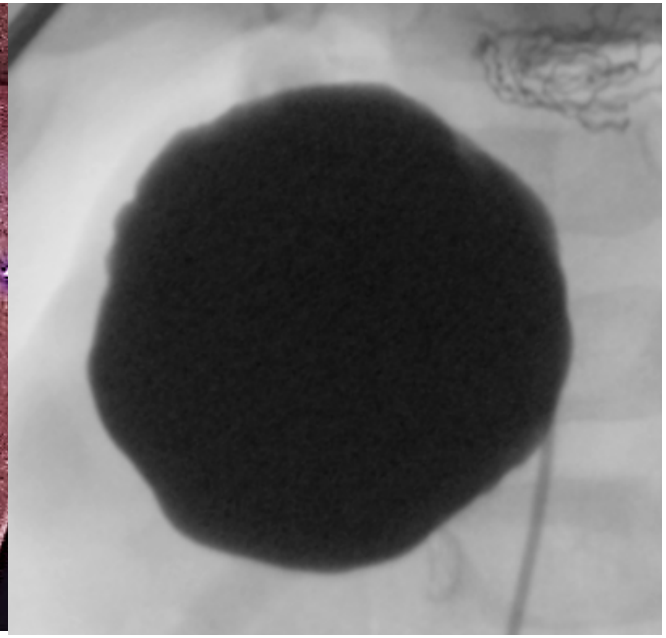
Type Ia CBD > Type I cyst BA



Almost normal IHBD



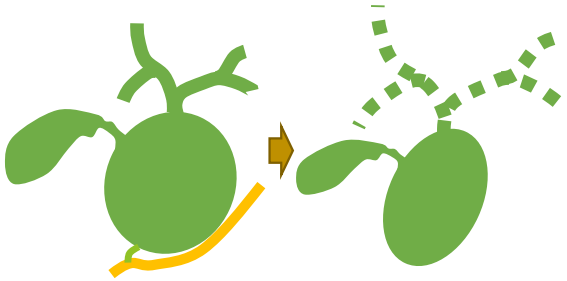
Makibishi sign



Contrast material did not enter the duodenum

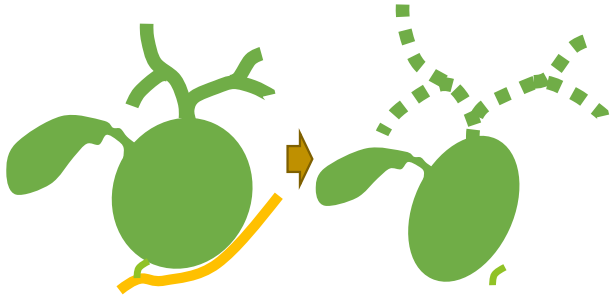
CBD featuring elements of BA
Intermediate/Transitional type
Bile duct resection yields good result.

There exists a transitional type from CBD to BA in the literature



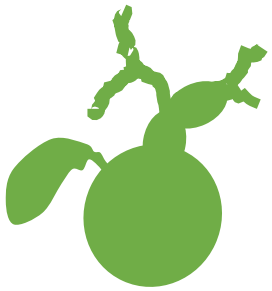
CBD → IIId BA

Caponcelli E, et al.
J Pediatr Surg. 2008;43:1619



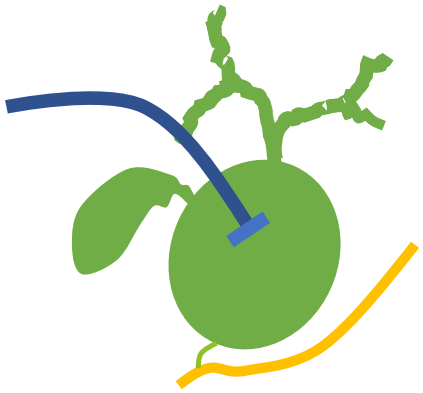
CBD → IIId BA (+NS)

Koshinaga T, et al.
BMC Pediatrics. 2018;18:158



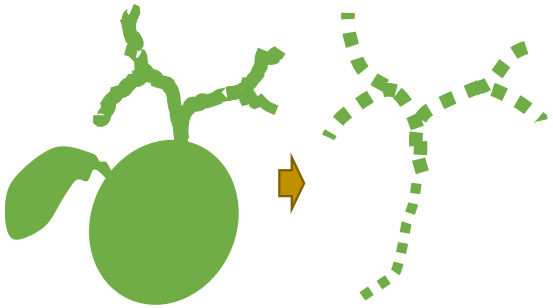
I cyst BA with a dilated LHD
Rapid progression of liver cirrhosis resulting in liver transplantation.
(The authors described a type IV-A CBD).

Fumino S, et al.
Pediatr Surg Int. 2013;29:1097



I cyst BA
After biliary drainage, contrast material began to enter the duodenum, revealing an NS and PBM.

Suzuki T, et al.
J Hepatobiliary Pancreat Sci. 2013;20:465

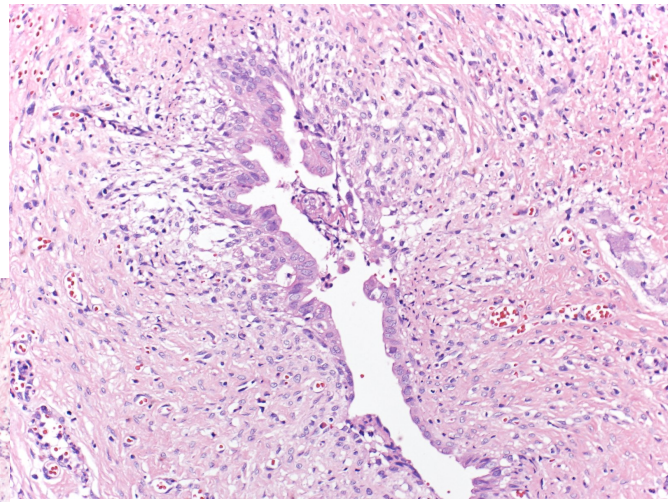


I cyst → III or IIId

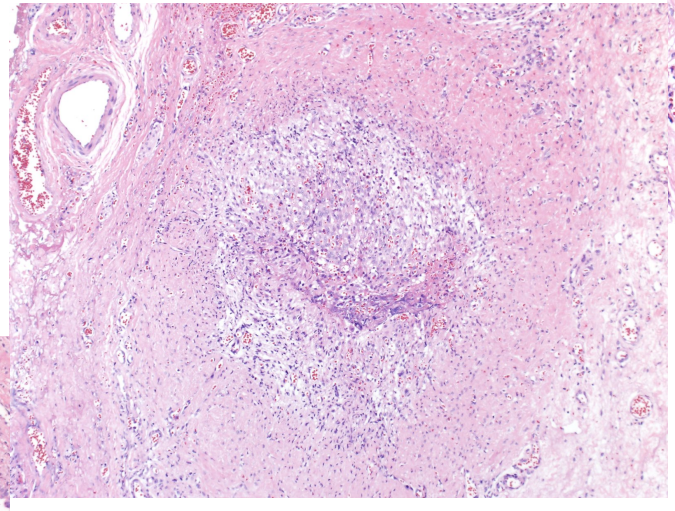
Nakata, et al.
J Jpn Pediatr Surg. 2000;36:332
Fujishiro J, et al.
Pediatr Sur Int. 2005;21:116
Kawasaki, et al.
J Jpn Pediatr Surg. 2009;45:729
Masumoto K, et al.
Pediatr Sur Int. 2011;27:99
Odashima, et al.
J Jpn Pediatr Surg. 2016;52:1042

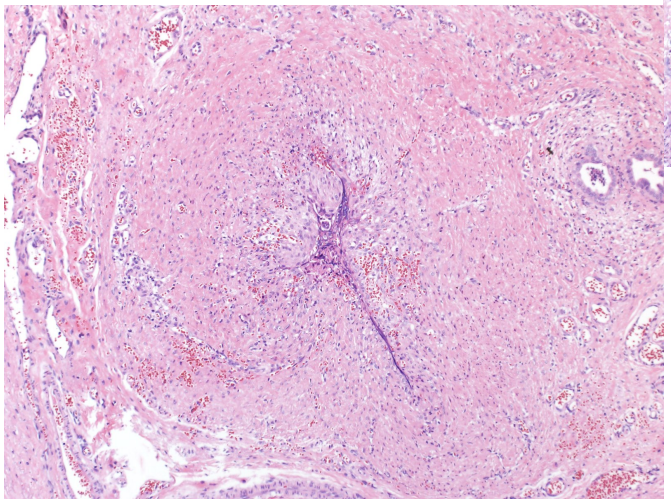
Pathology of hilar remnant in type IIId BA

Obstruction progresses from downstream to upstream?

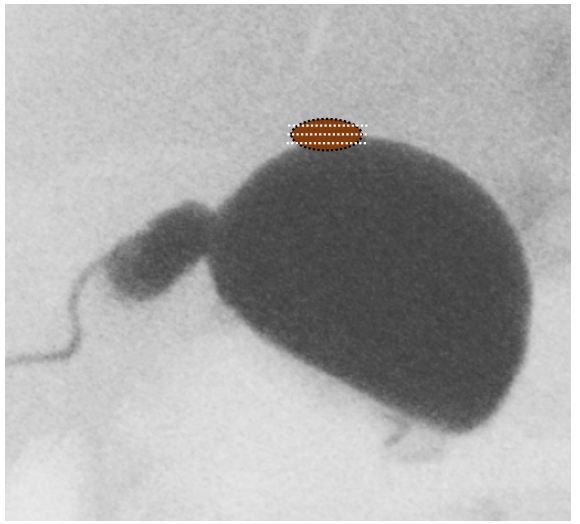


Upstream





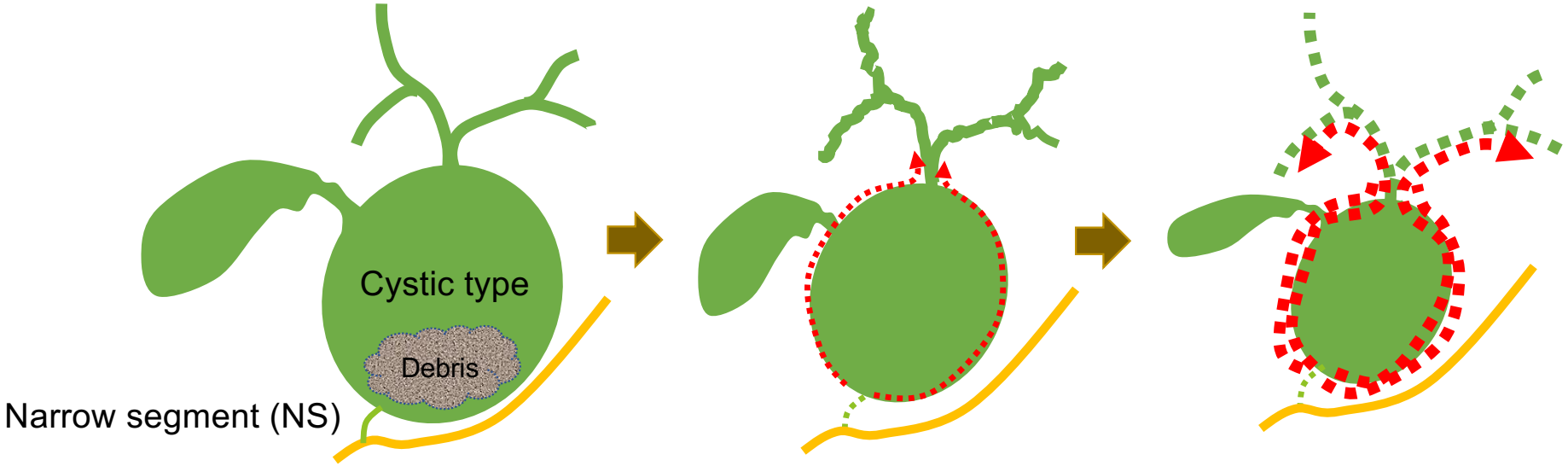
Downstream



Hypothesis

Cystic BA originates from CBD with an NS

Infantile obstructive cholangiopathy progresses according to the degree of obstruction at the NS.



Ia CBD

Pure congenital malformation

I cyst BA

IIId BA

Kato S, et al. *J Hepatobiliary Pancreat Sci*. 2022;29(9):961–963.

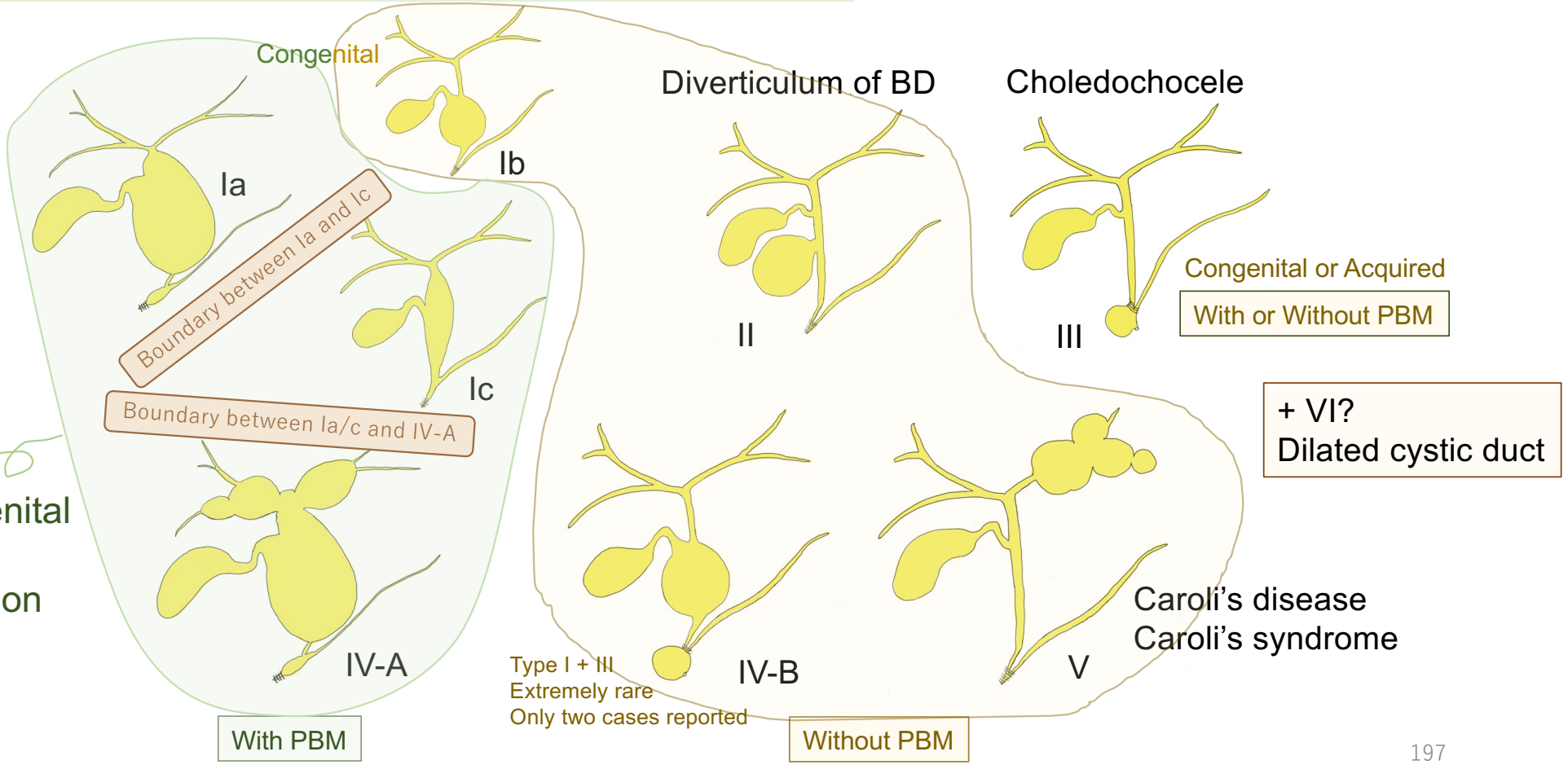
Expert

Rare congenital dilatation of the biliary tract

First version, September 2026

Todani's classification of choledochal cysts 1977, revised 1995

The term choledochal cyst involves different entities



Case 51

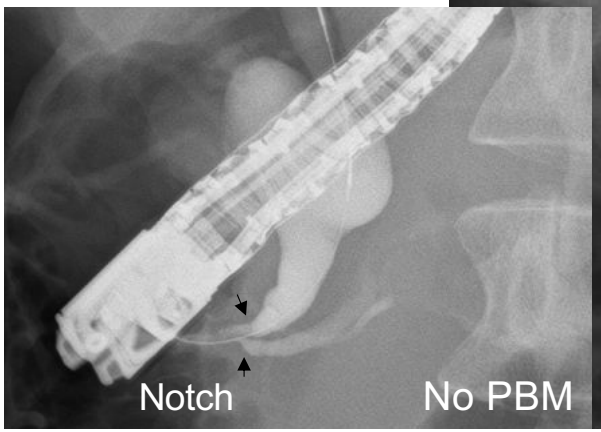
Type Ib:
segmentally dilated common bile duct without PBM

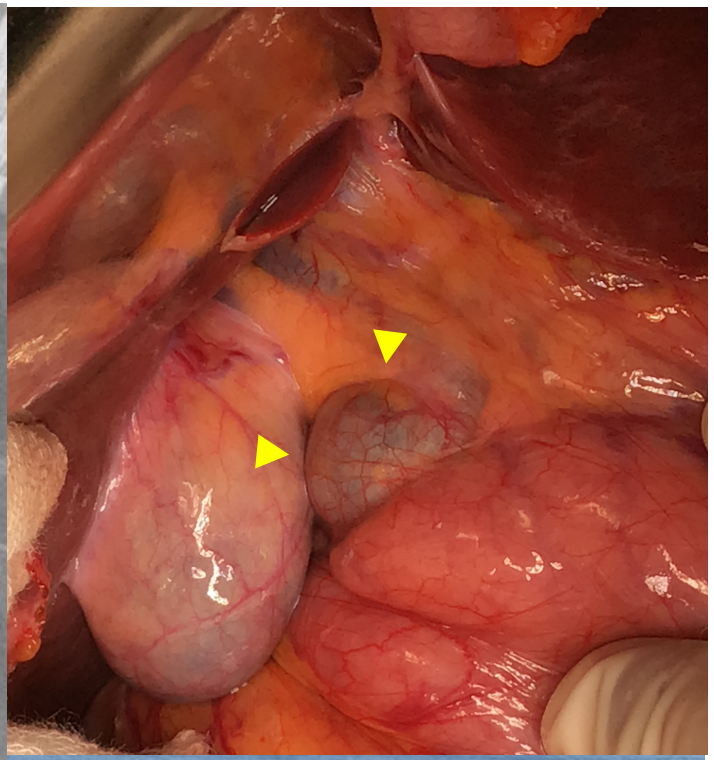
A female in her 30s.

She experienced repeated episodes of abdominal pain.

The BD was resected.

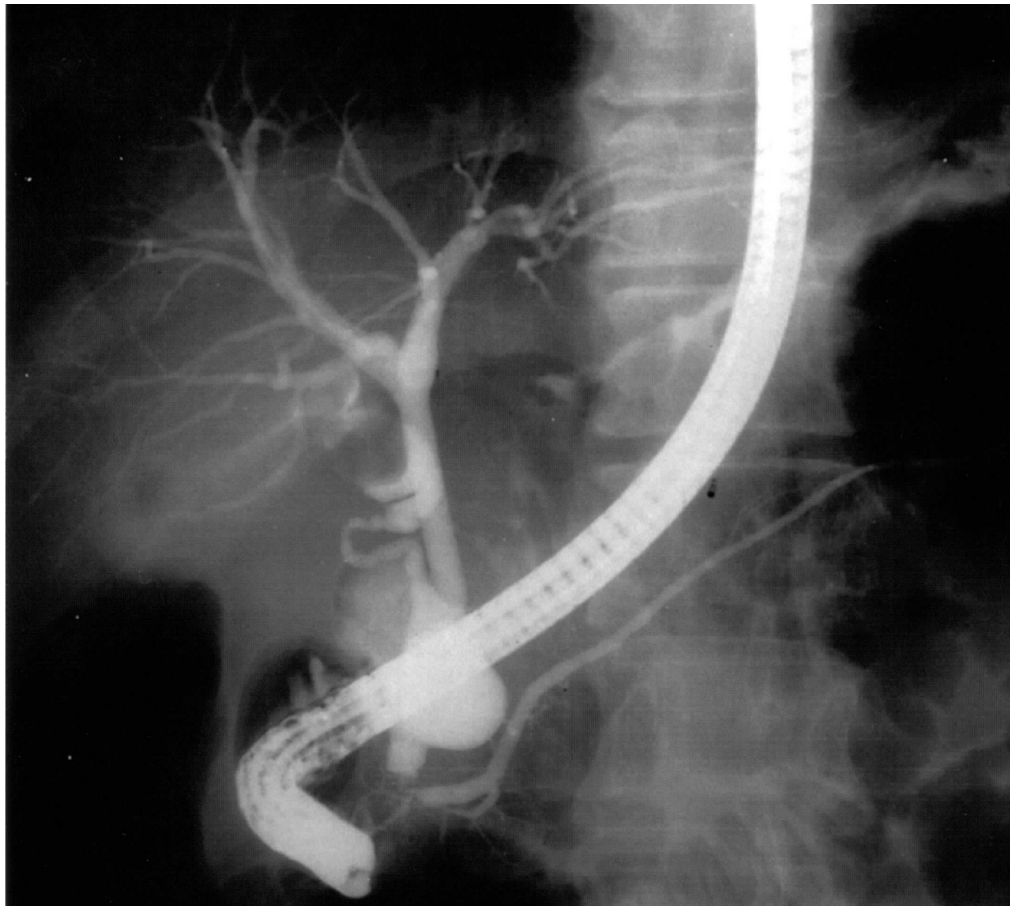








Type Ib



The sphincter action affects both the BD and the PD, proving no PBM.

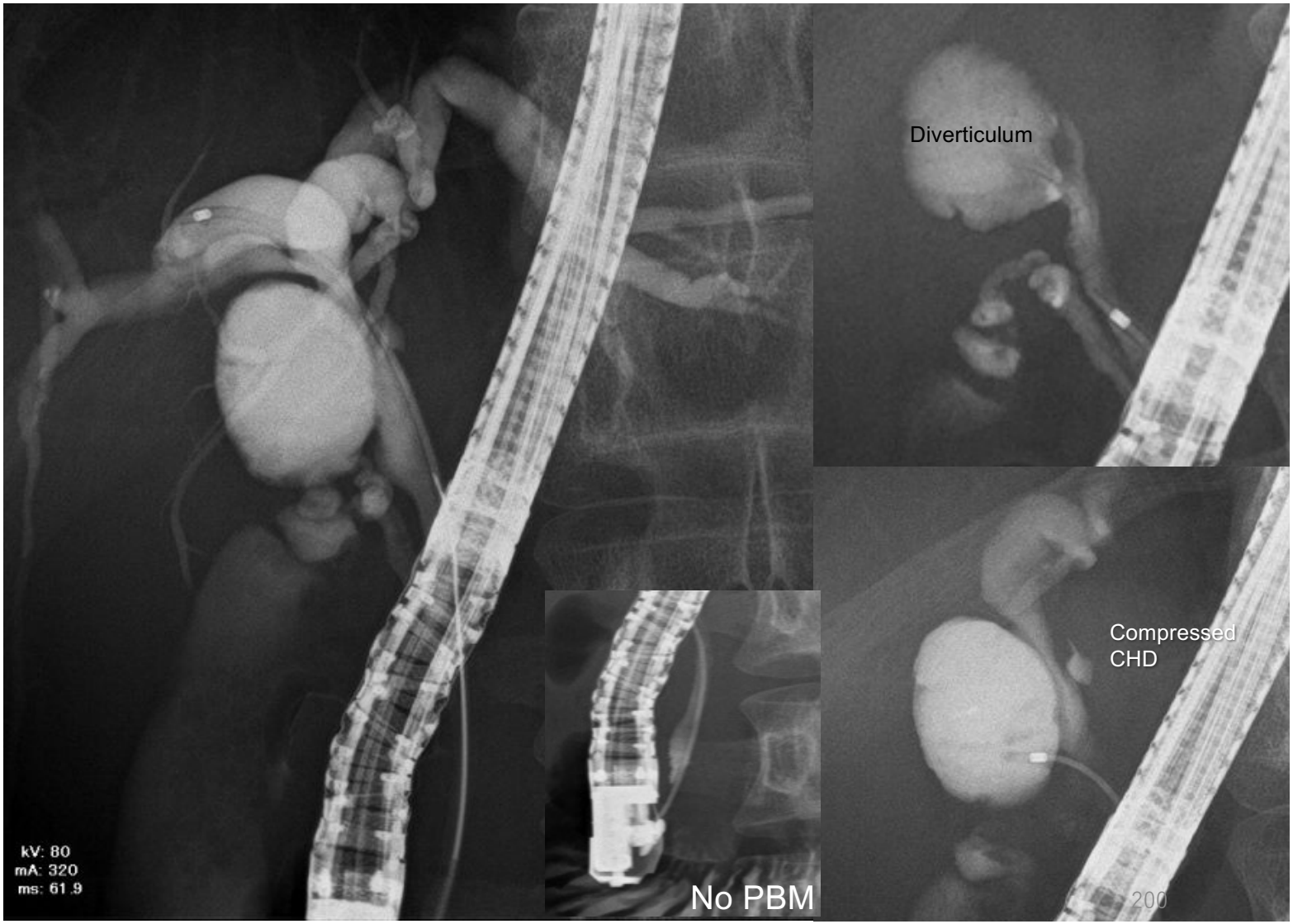
Case 52

Type II: diverticulum

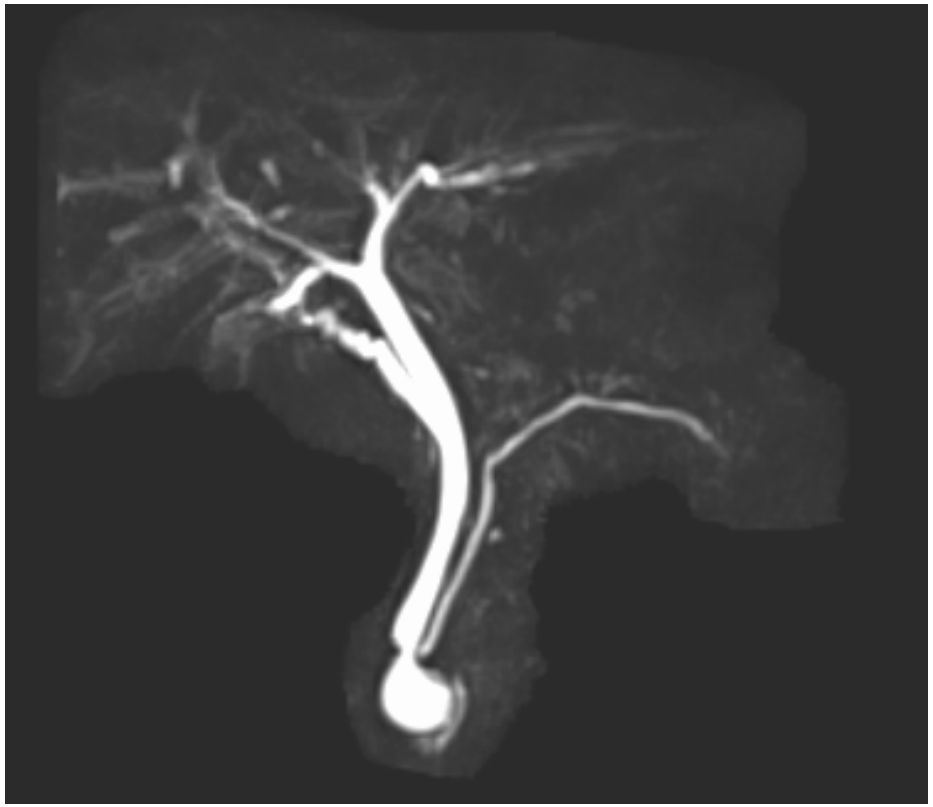
A male in his 20s.
He presented with epigastric pain and obstructive jaundice (T-bil 5.8 mg/dL, D-bil 4.2 mg/d, AST 195 U/L, ALT 595 U/L, γ-GT 733 U/L).

The diverticulum compressed the common hepatic duct, causing obstructive jaundice and IHBD dilatation.

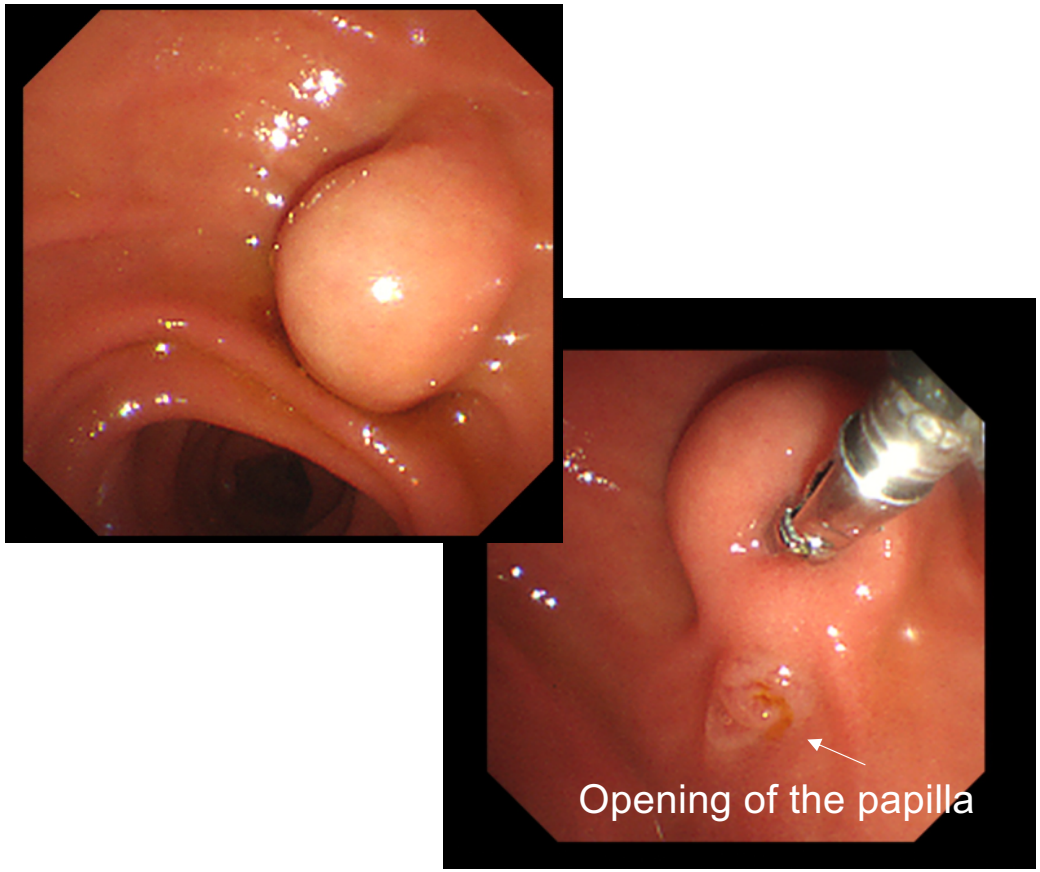
The BD was resected.



Case 53 Type III: Choledochocele



Kamisawa's classification: cystic dilatation of the intramural BD

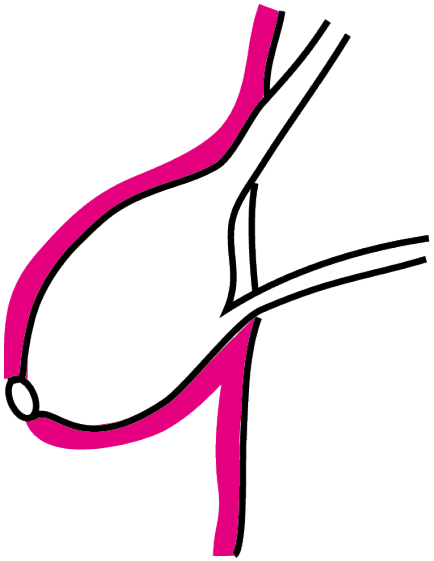


A male in his 60s. A choledochocele was detected during endoscopic examination for reflux esophagitis, and was managed by observation alone due to the absence of symptoms.

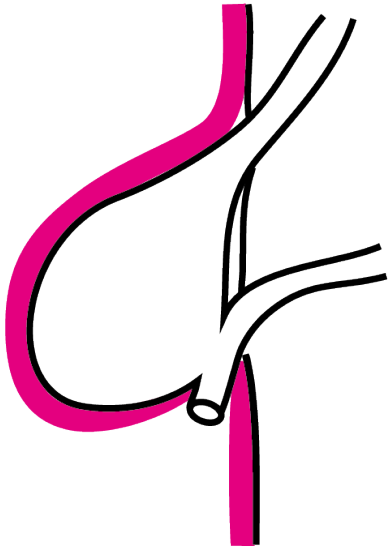
Classification of choledochocele

Kamisawa T, et al. *Tando (Biliary Tract).* 2002;16:26–32 (in Japanese)
Kamisawa T, et al. *Hepatogastroenterology.* 2005;52:29–32

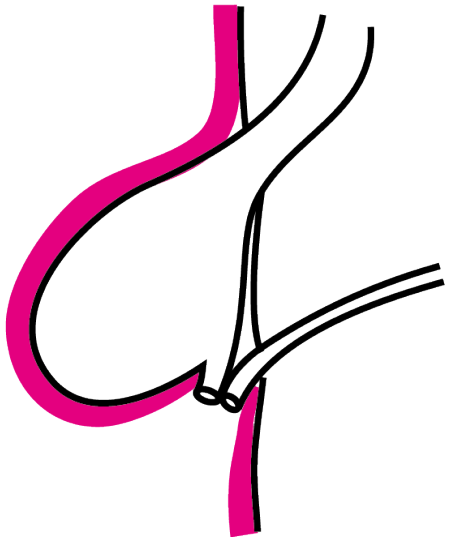
This type contains acquired cases or cases without PBM.



Cystic dilatation of the common channel



Cystic dilatation of the intramural BD forming a common channel



Cystic dilatation of the intramural BD without forming a common channel

These types can cause acute pancreatitis.