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Advanced

Issues on Todani's classification

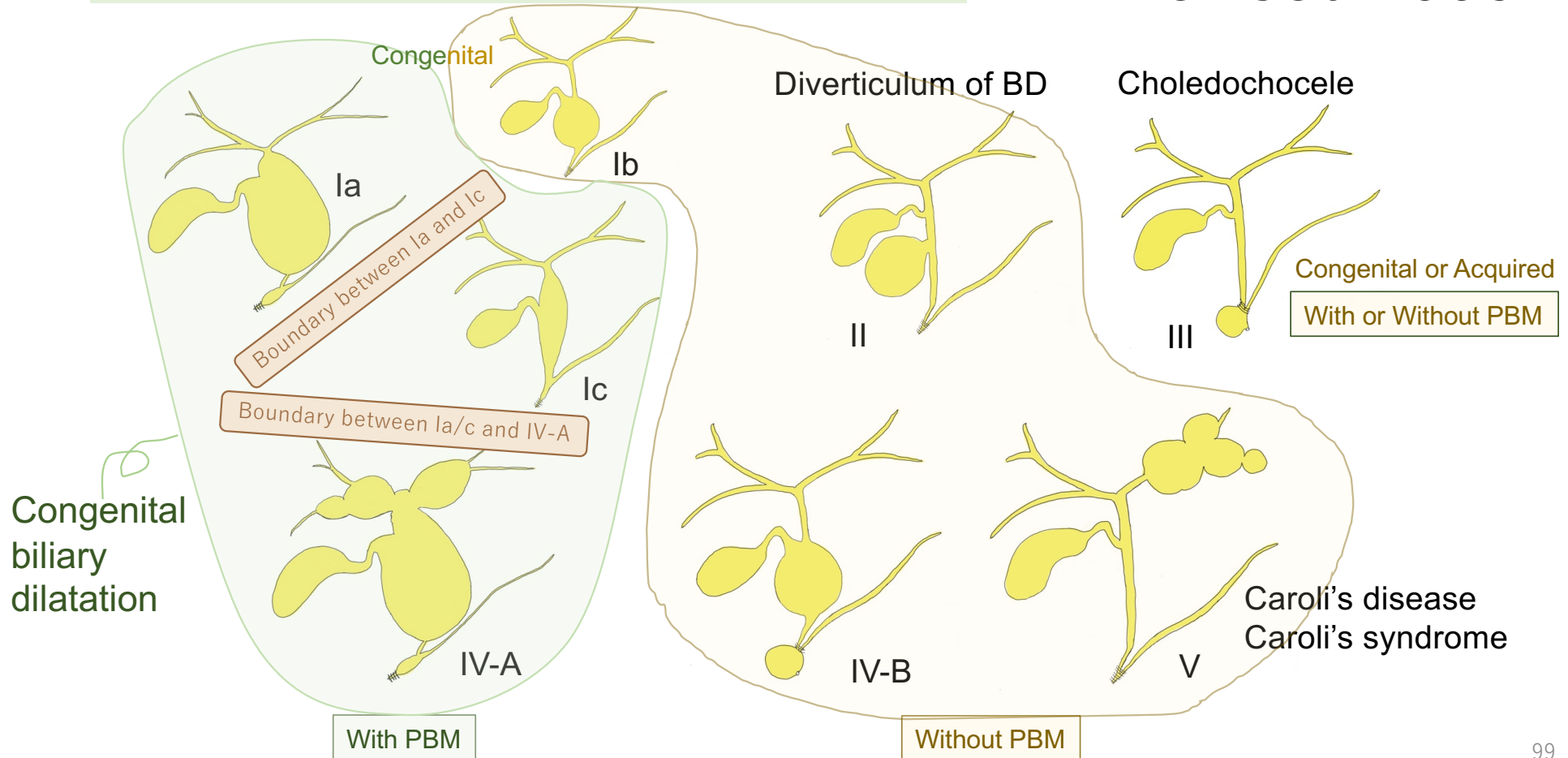
Arbitrary boundaries between Ia/c and IV-A, and between Ia and Ic

First version, September 2024

2nd version, September 2025

Todani's classification of choledochal cysts 1977, revised 1995

The term choledochal cyst involves different entities



Todani type I

Todani T, Watanabe Y, Toki A, Morotomi Y.

Classification of congenital biliary cystic disease: special reference to type Ic and IVA cysts with primary ductal stricture.

J Hepatobiliary Pancreat Surg. 2003;10(5):340-4.

doi: 10.1007/s00534-002-0733-7. PMID: 14598133.

- **Type I** cysts have no intrahepatic component and the common hepatic duct proximal to the cyst is usually normal. This type is morphologically divided into three subtypes:
 - **Type Ia** is a cystic dilatation of the choledochus
 - **Type Ib** is a segmental dilatation with no pancreatobiliary malunion present, and hepaticocholedochostomy after cyst excision may be feasible See Expert Section
 - **Type Ic** refers to a fusiform, diffuse, or cylindrical dilatation of the choledochus with pancreatobiliary malunion, and the dilatation often extends continuously to the intrahepatic duct

The form of these dilatations is not cystic but fusiform, and these dilatations diminish after bile duct excision. This type should not be included in IV-A.

Todani type IV

Todani T, Watanabe Y, Toki A, Morotomi Y.

Classification of congenital biliary cystic disease: special reference to type Ic and IVA cysts with primary ductal stricture.

J Hepatobiliary Pancreat Surg. 2003;10(5):340-4.

doi: 10.1007/s00534-002-0733-7. PMID: 14598133.

- **Type IV** is a multiple cyst

- **Type IVA** involves multiple cysts in both the intrahepatic and extrahepatic bile ducts. Intrahepatic dilatation affects both lobes, although the left lobe is predominantly involved.

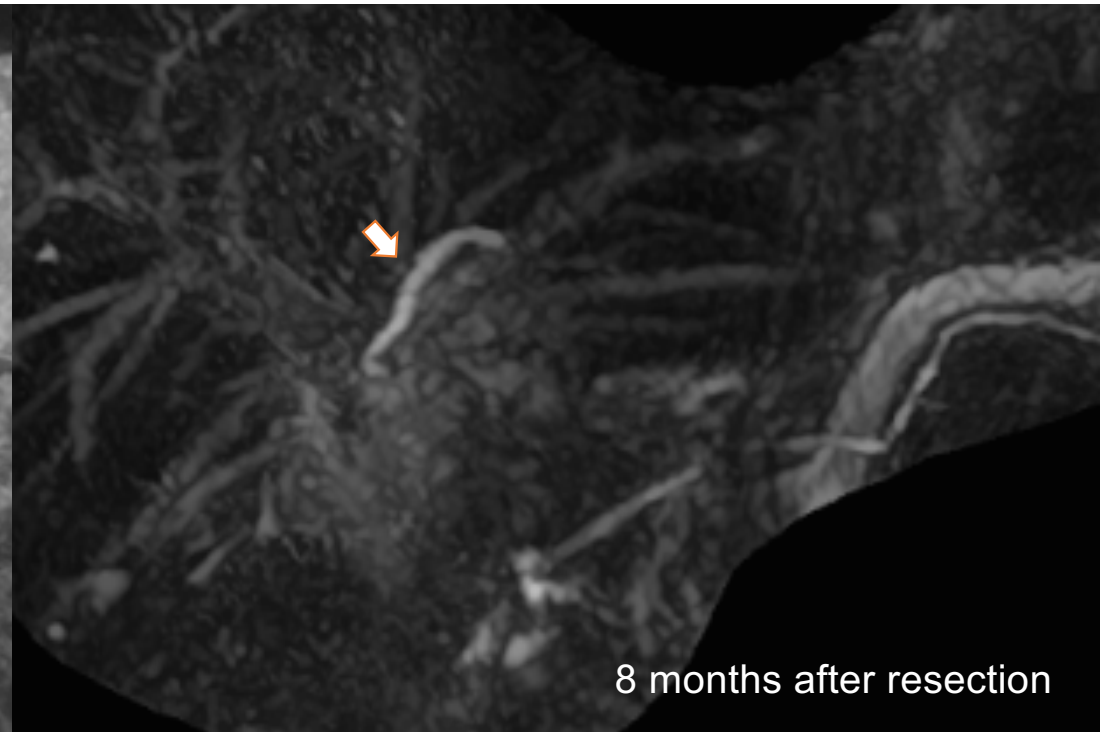
Based on the shape of the extrahepatic and intrahepatic ducts, type IV is subclassified into three forms, cystic–cystic, cystic–fusiform, and fusiform–fusiform.

Necessary condition: BD dilatation upstream from CHD

Sufficient conditions: **Limited** BD dilatation upstream from CHD accompanying **strictures** and the **dilatation remaining** after BD resection

Cases falling short of sufficient conditions are difficult to judge I or IV-A.

Intrahepatic BD dilatation diminishes after BD resection Adult case, Todani type 1c



Literature

Koshinaga T, Inoue M, Ohashi K, Sugito K, Ikeda T, Hagiwara N, Tomita R. Persistent biliary dilatation and stenosis in postoperative congenital choledochal cyst. J Hepatobiliary Pancreat Sci. 2011 Jan;18(1):47-52

Hill R, Parsons C, Farrant P, Sellars M, **Davenport M**. Intrahepatic duct dilatation in type 4 choledochal malformation: pressure-related, postoperative resolution. J Pediatr Surg. 2011 Feb;46(2):299-303.

Case 27

Ic>Ia>>IV-A

Dilatation of the bile ducts upstream from CHD, but diffuse dilatation without strictures

Female in her thirties
Abdominal pain

- Fusiform (Ic) rather than cystic (Ia) dilatation of EHBD
- Continuous dilatation extends to right and left hepatic ducts
See the definition of Ic on P96.
- Narrow segment with a dilated common channel
= Type C PBM
according to the JSGPBM classification on P35

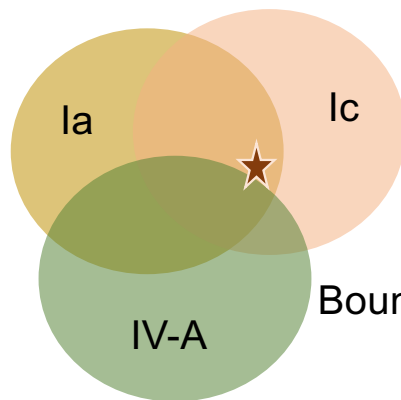


Case 28

Ic>Ia>IV-A

ERCP in a male in his seventies
Abdominal pain and fever

- Slight dilatation of RHD with a stenosis (arrow) that would vanish after BD resection
- Fusiform (Ic) rather than cystic (Ia) dilatation
- Narrow segment (NS) or normal thin BD? Operative findings showed no constriction, indicating a normal BD rather than NS. Type A < Type B (non-stenotic type) PBM See P133–6.



Boundaries are arbitrary.



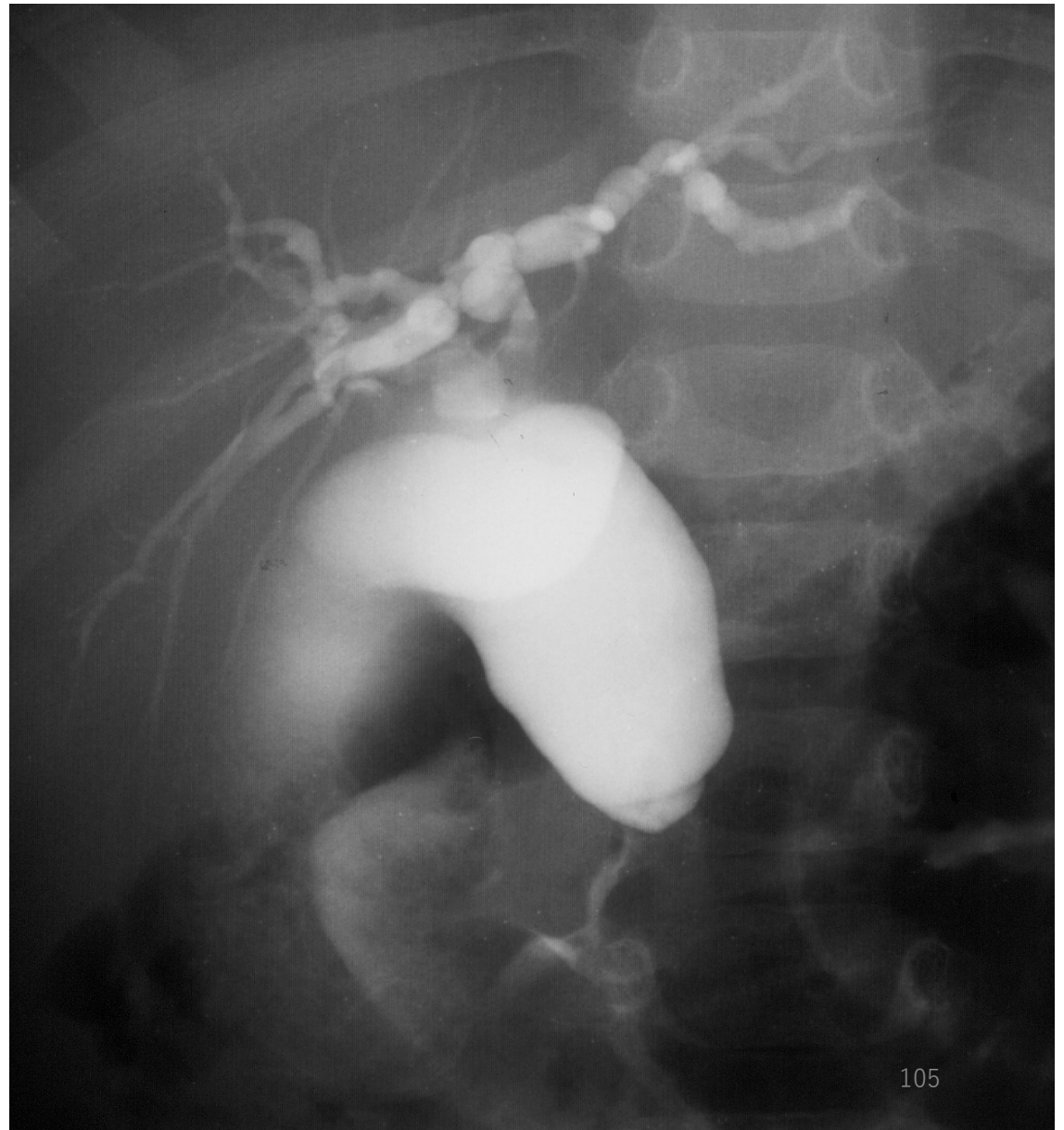
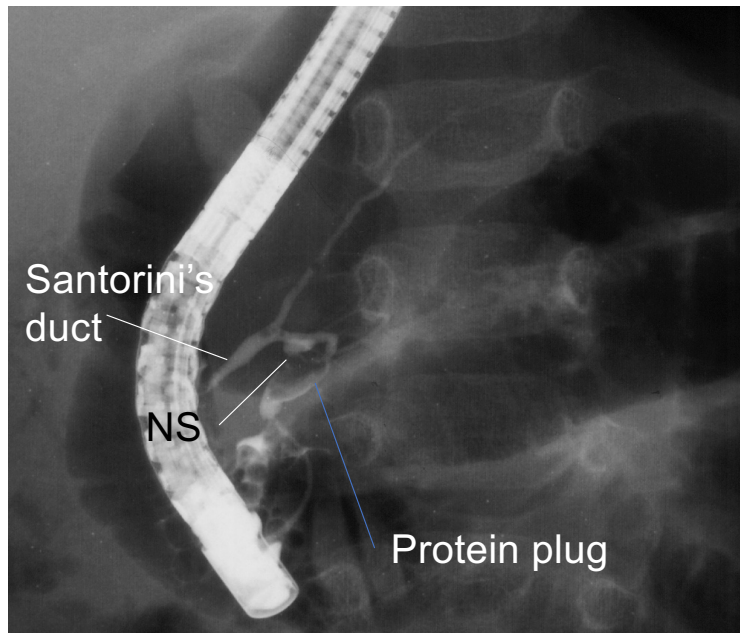
Case 29

Ia>IV-A

ERCP in a 9-year-old child

Abdominal pain, jaundice, acholic stool

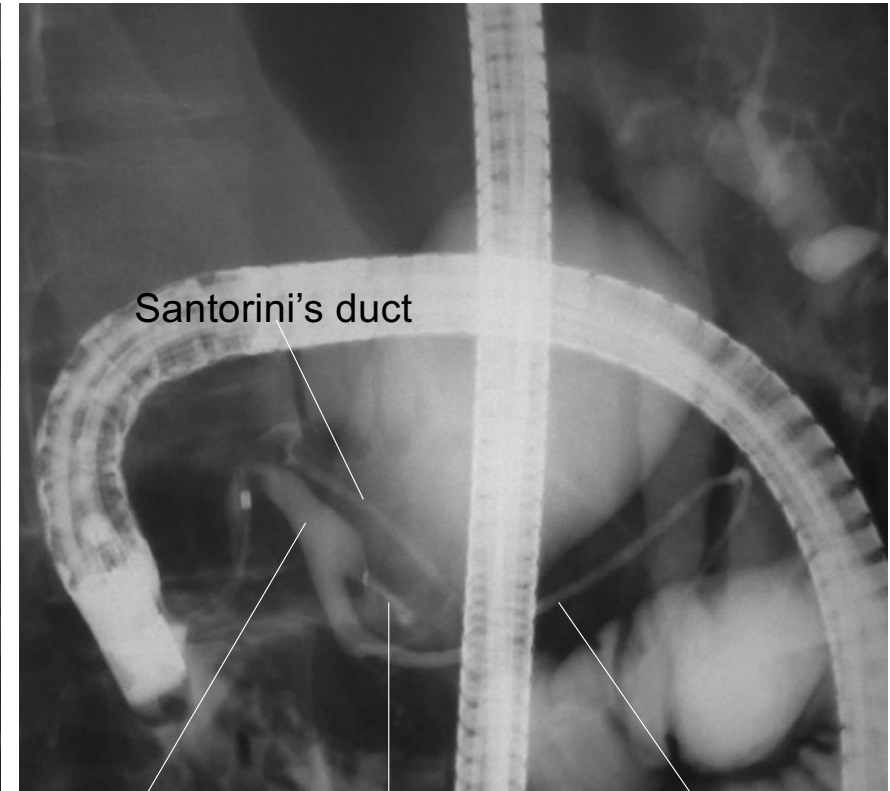
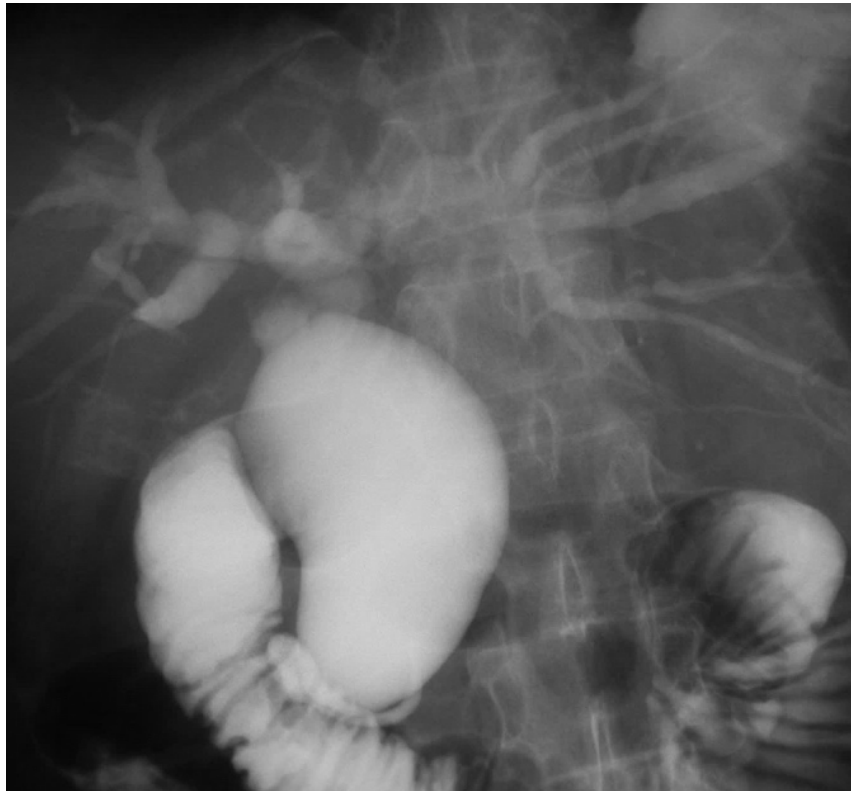
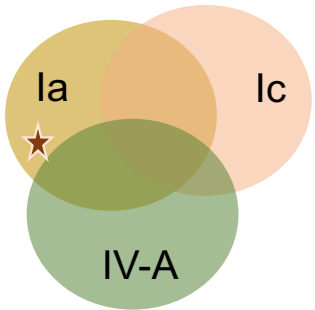
Transient diffuse intrahepatic BD (IHBD) dilatation due to protein plug obstruction
Dilatation diminished after BD resection.



Case 30

la > IV-A

Female in her
fifties
ERCP shows
diffuse dilatation
of IHBD



Common channel

NS

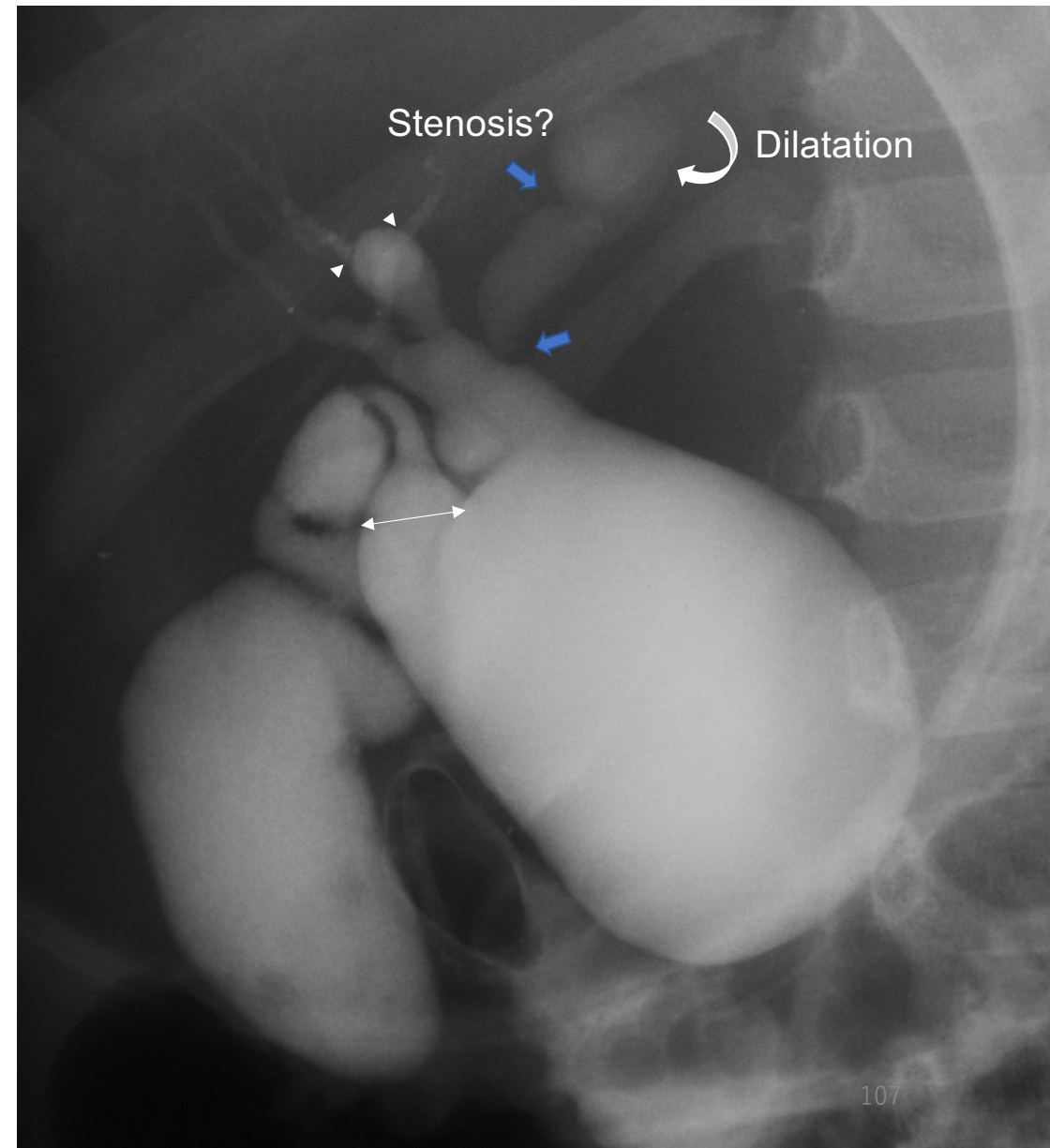
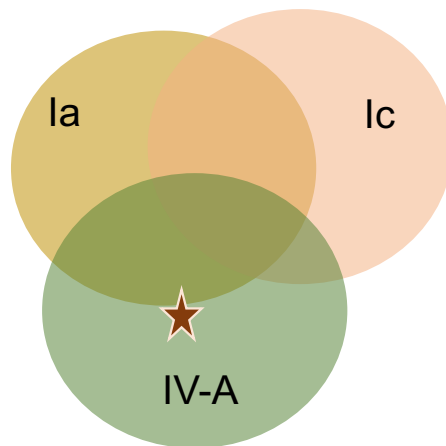
Main PD

Case 31

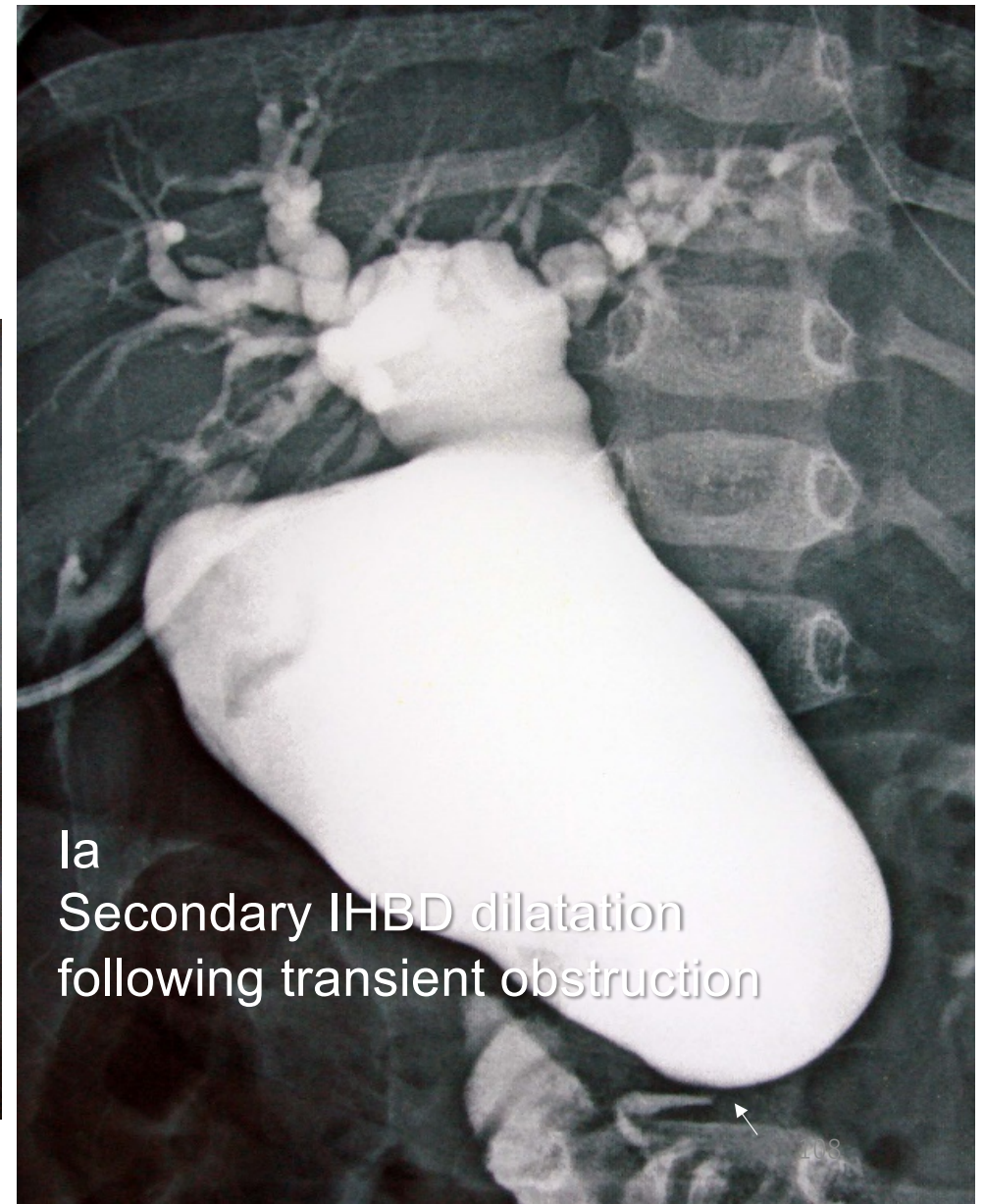
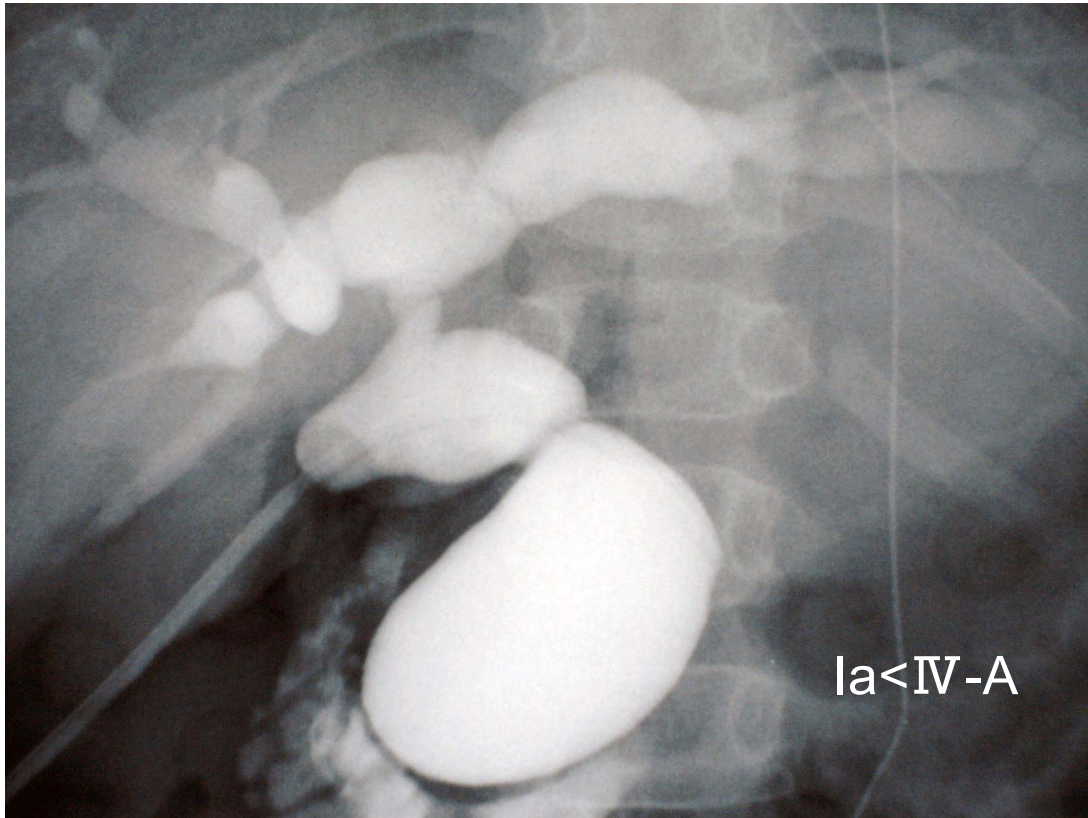
IV-A > Ia

ERCP in a 2-year-old girl
Abdominal pain and vomiting

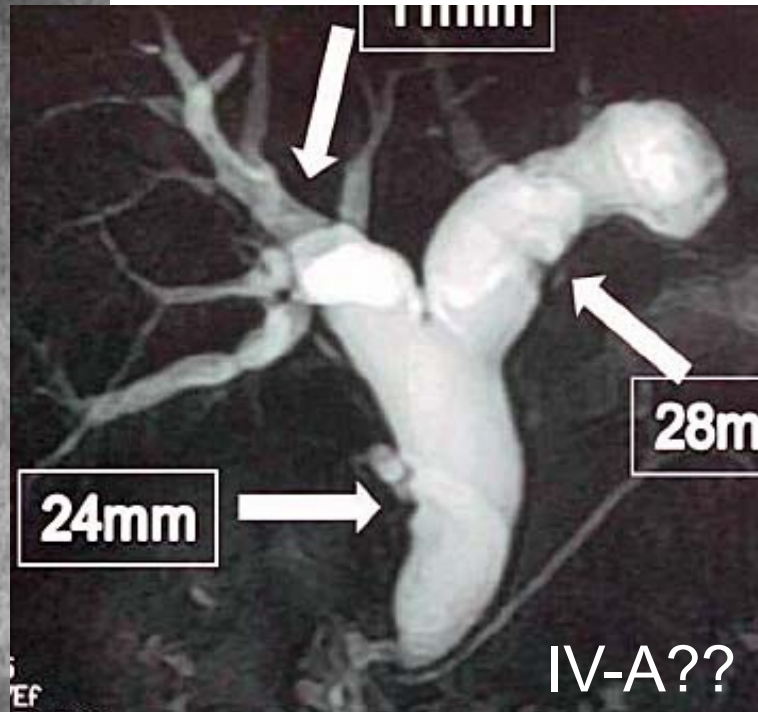
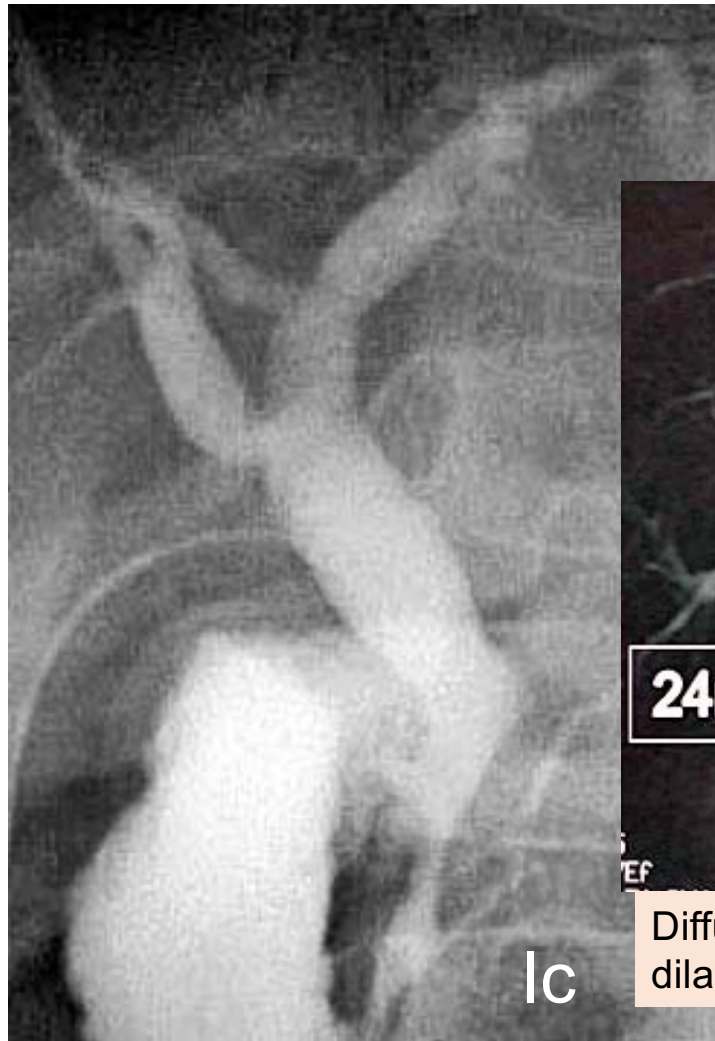
- Cystic dilatation upstream from LHD (curved arrow) with constrictions (arrows)
- Limited dilatation (arrow heads) indicating type IV-A
- Eminent dilatation at the base of the cystic duct (double-headed arrow)



Two cases: Ia or IV-A?

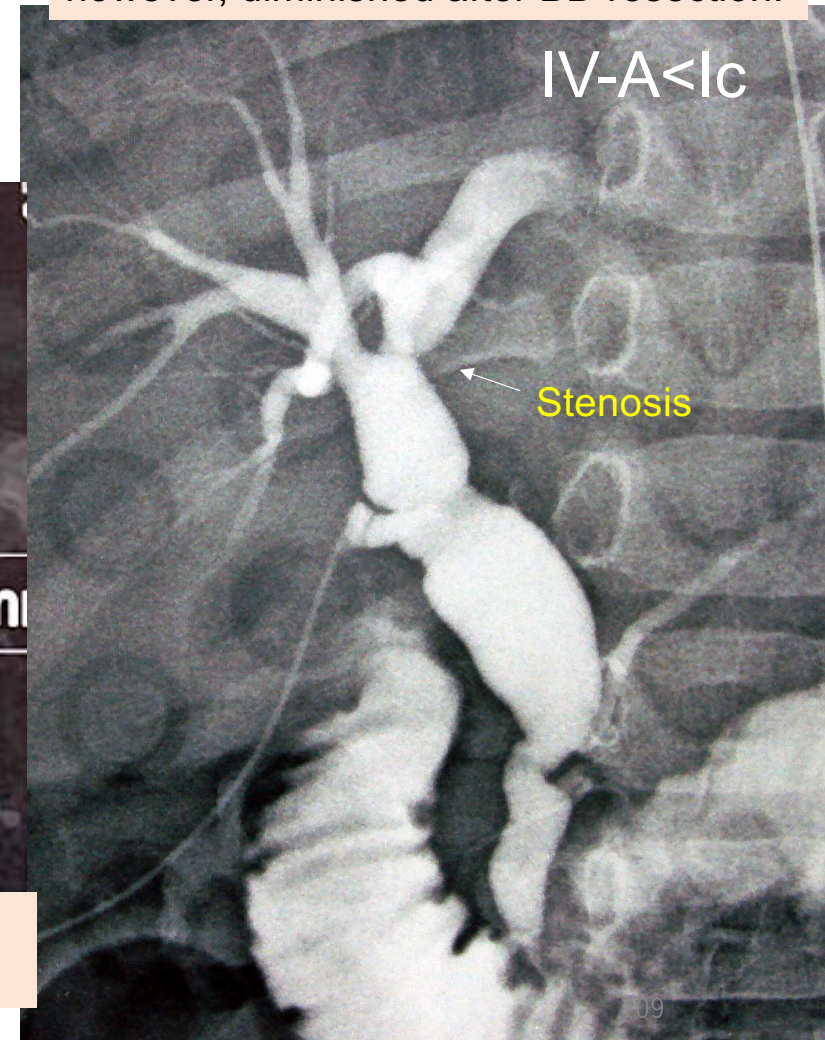


Three cases: Ic or IV-A?



Diffuse dilatation without stenosis but dilatation remaining after BD resection

Diffuse dilatation with LHD stenosis, however, diminished after BD resection.

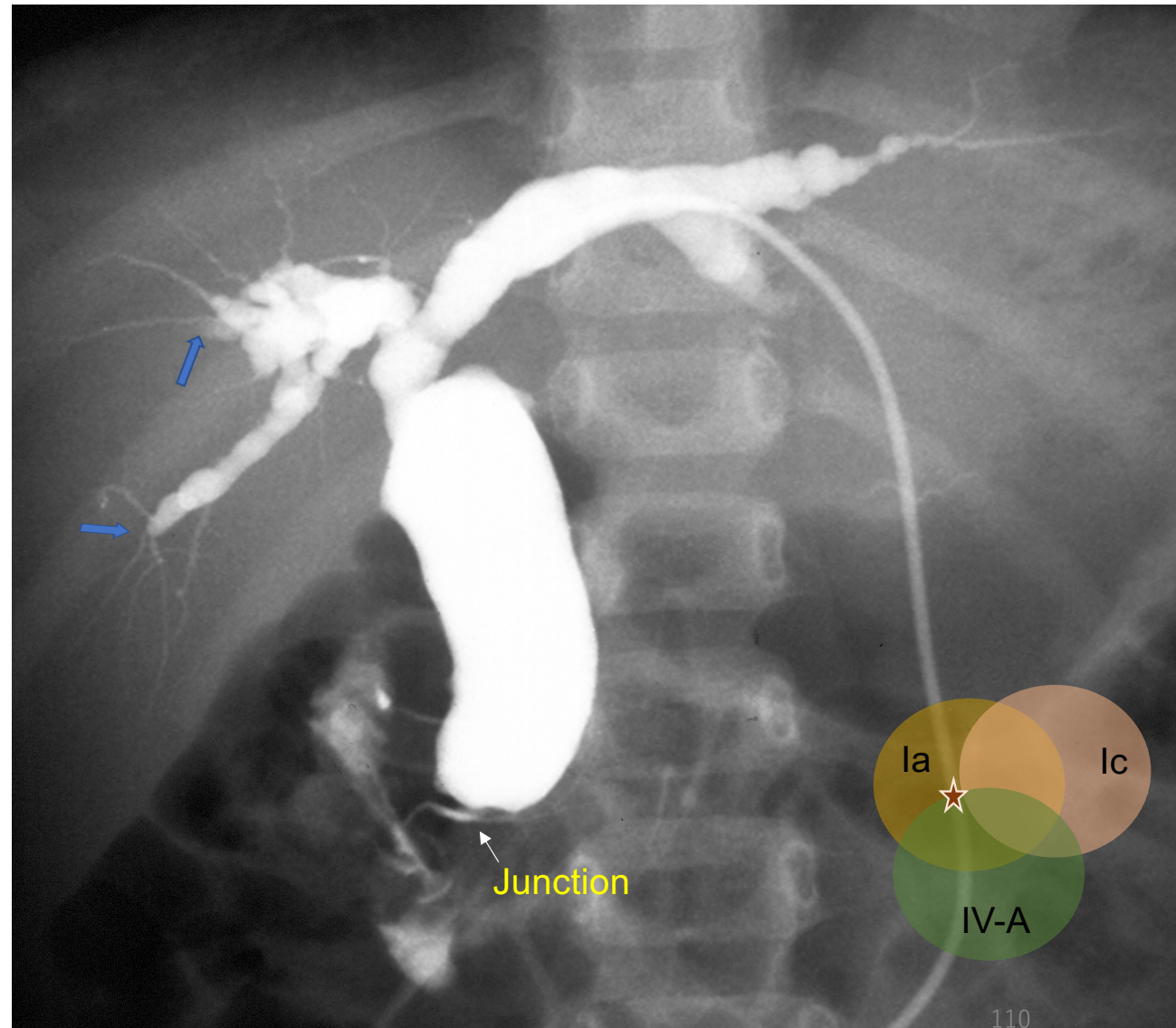


Case 32 Ia or IV-A?

Percutaneous transhepatic cholangiography in a 2-year-old boy
Abdominal pain, jaundice, elevated serum levels of hepato-biliary enzymes

- **Cystic-dilated EHBD** 17 mm
- **Fusiform-dilated IHBDs** (LHD 8mm) abruptly change to those of normal caliber (→), indicating **localized dilatation**.
However, this IHBD dilatation would diminish after BD resection, which contradicts IV-A criteria.
- Very short narrow segment
Type A (stenotic type) PBM

EHBD, extrahepatic bile duct (BD); IHBD, intrahepatic BD;
LHD, left hepatic duct



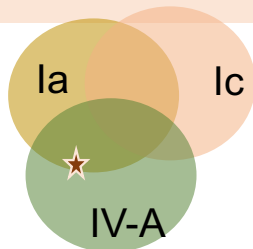
Case 33

Ia or IV-A?

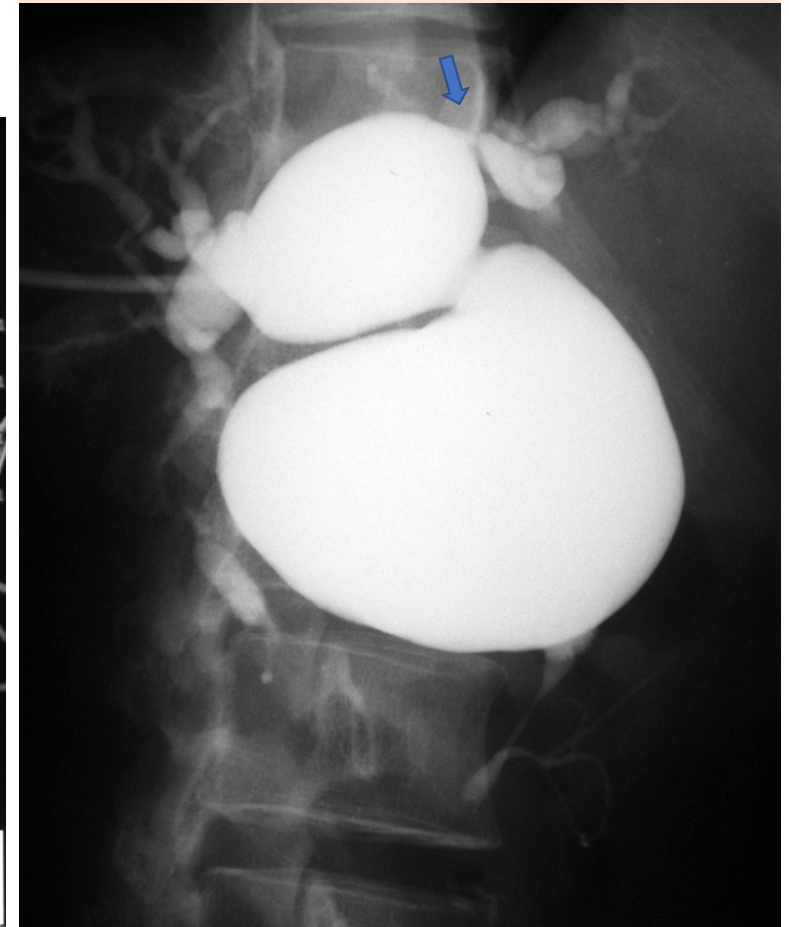
Female in her twenties
Percutaneous transhepatic
biliary drainage due to fever,
jaundice, and abdominal
pain

Diffusely dilated RHD
without stenoses
Slightly dilated LHD with a
stenosis (arrow)
BD resection with stenosis
treatment would relieve
these dilatations

Ia or IV-A, questionable



Stenosis at the orifice but only slightly
dilatated LHD



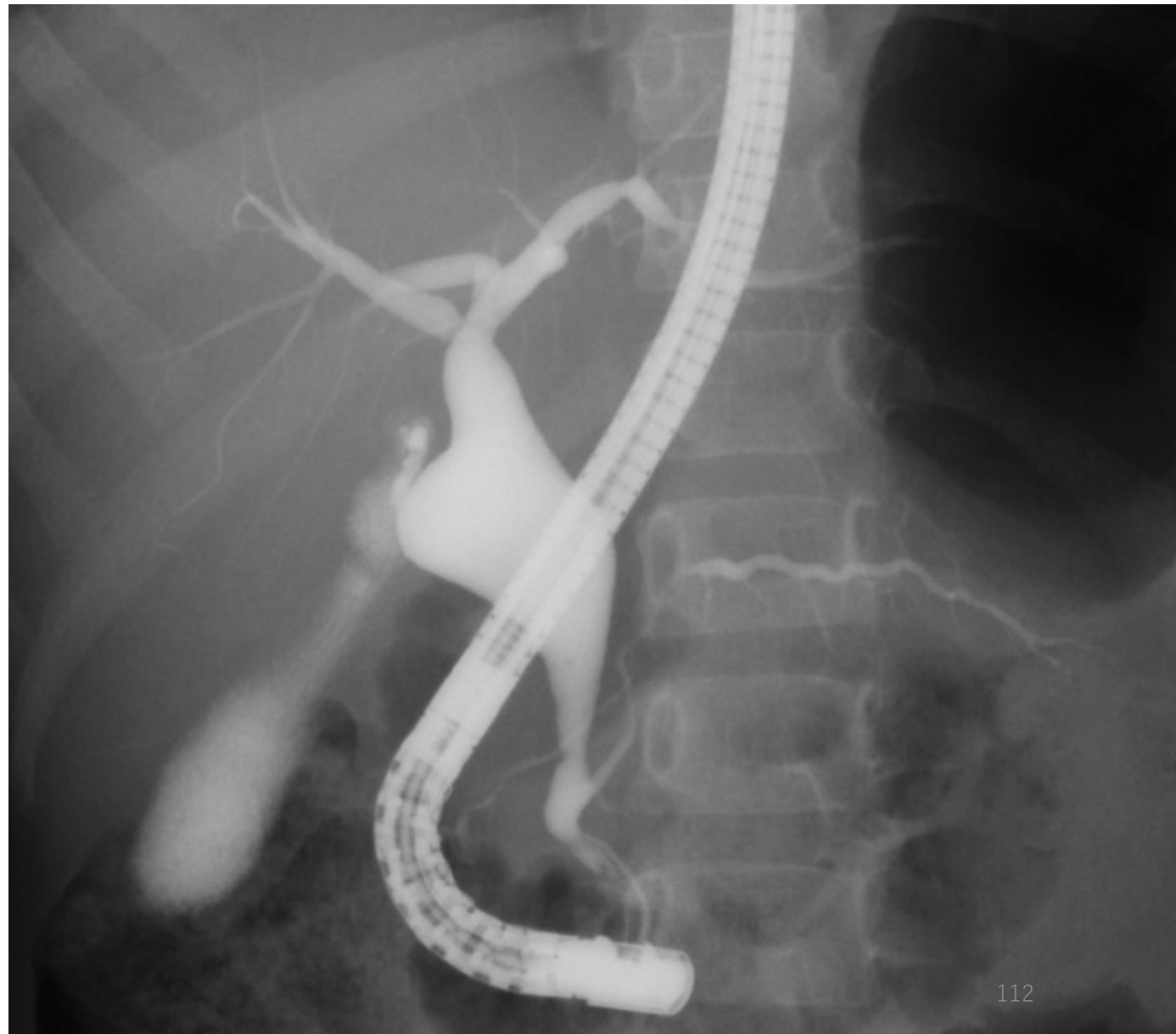
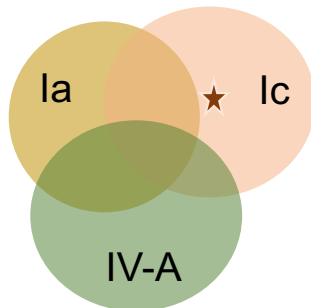
Diffusely dilated RHD but without strictures

Case 34

Ia<Ic

ERCP in a 4-year-old child
Vomiting, jaundice, elevated serum
levels of liver enzymes and amylase

- Dilatation up to CHD
- Dilated cystic duct composing the
common bile duct
Cystic or fusiform, either is fine
- No clear narrow segment indicating
type B (non-stenotic type) PBM
often seen in Ic choledochal cyst



Advanced

Nondilated type of PBM

Definition, pathophysiology, and diagnosis

First version, September 2025

Imaging features of congenital biliary dilatation

Features that define CBD

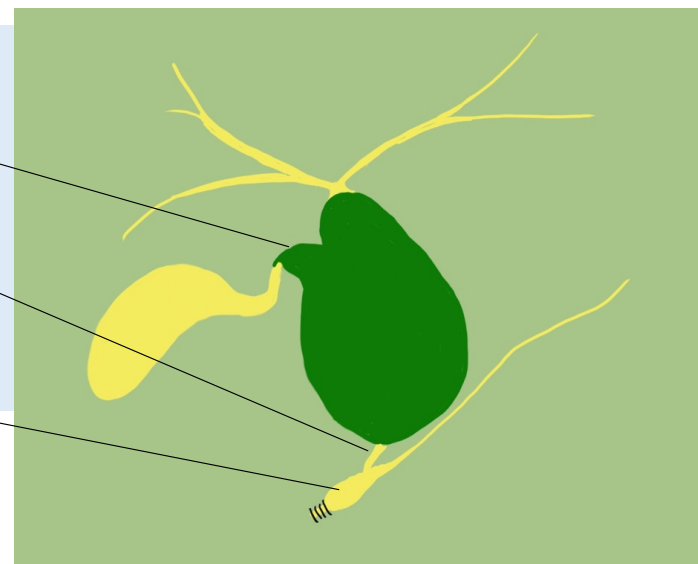
Introduction section

- Pancreaticobiliary maljunction
- Localized BD dilatation involving the common bile duct

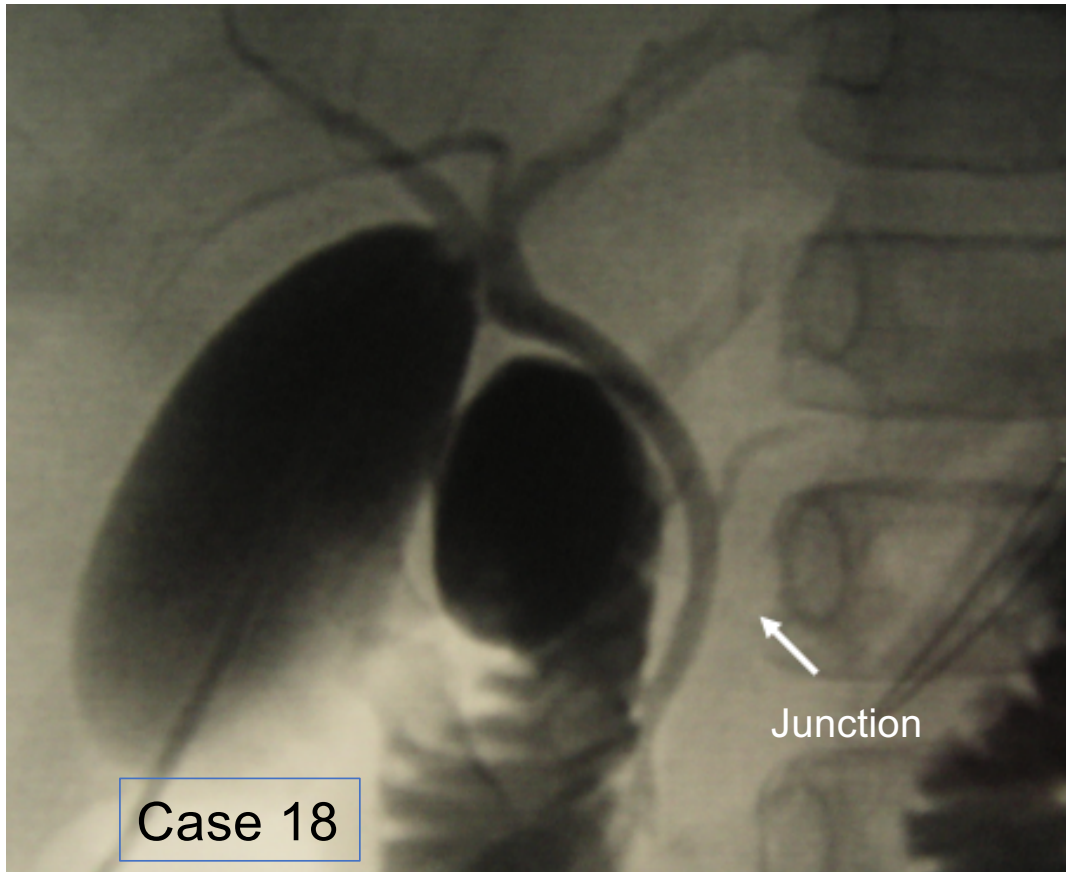
Features (but not in all cases)

Basic section

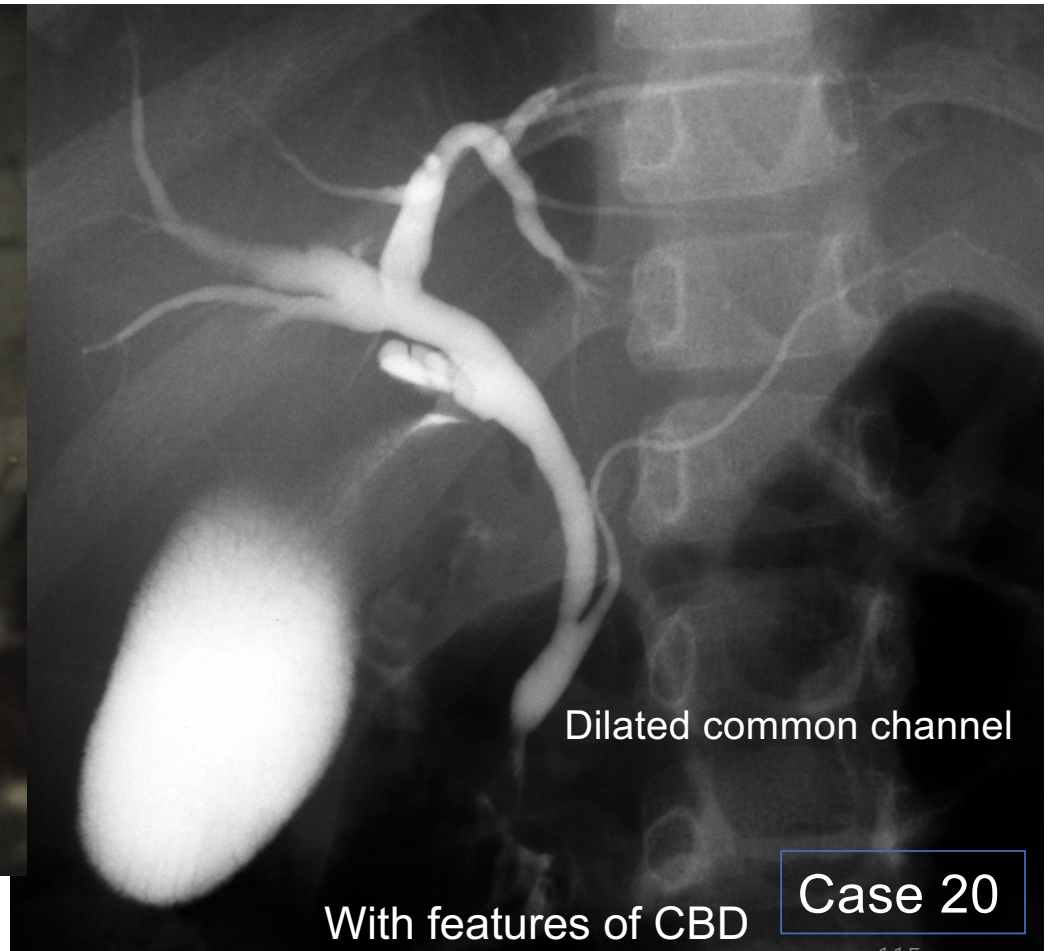
- Dilatation at the base of the cystic duct
- Narrow segment below the bile duct
- Dilated common channel



PBM with normal BD diameter Pediatric cases

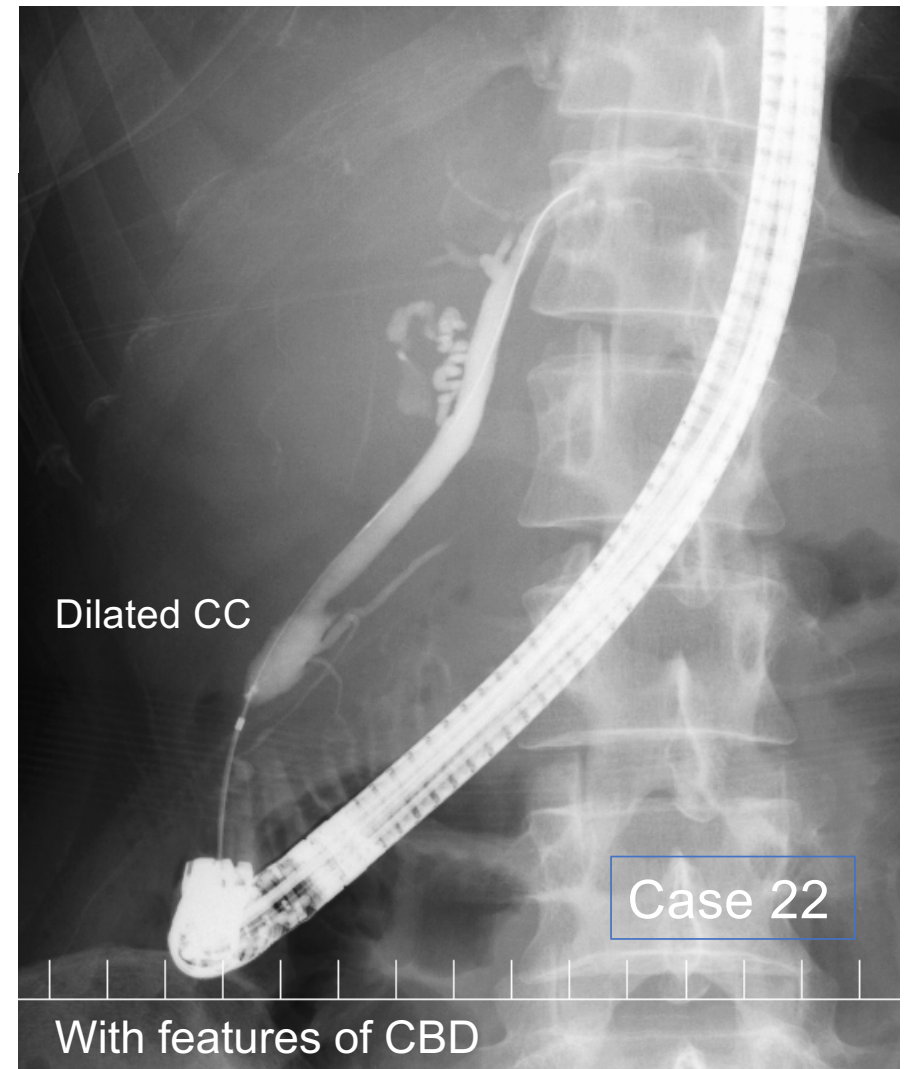
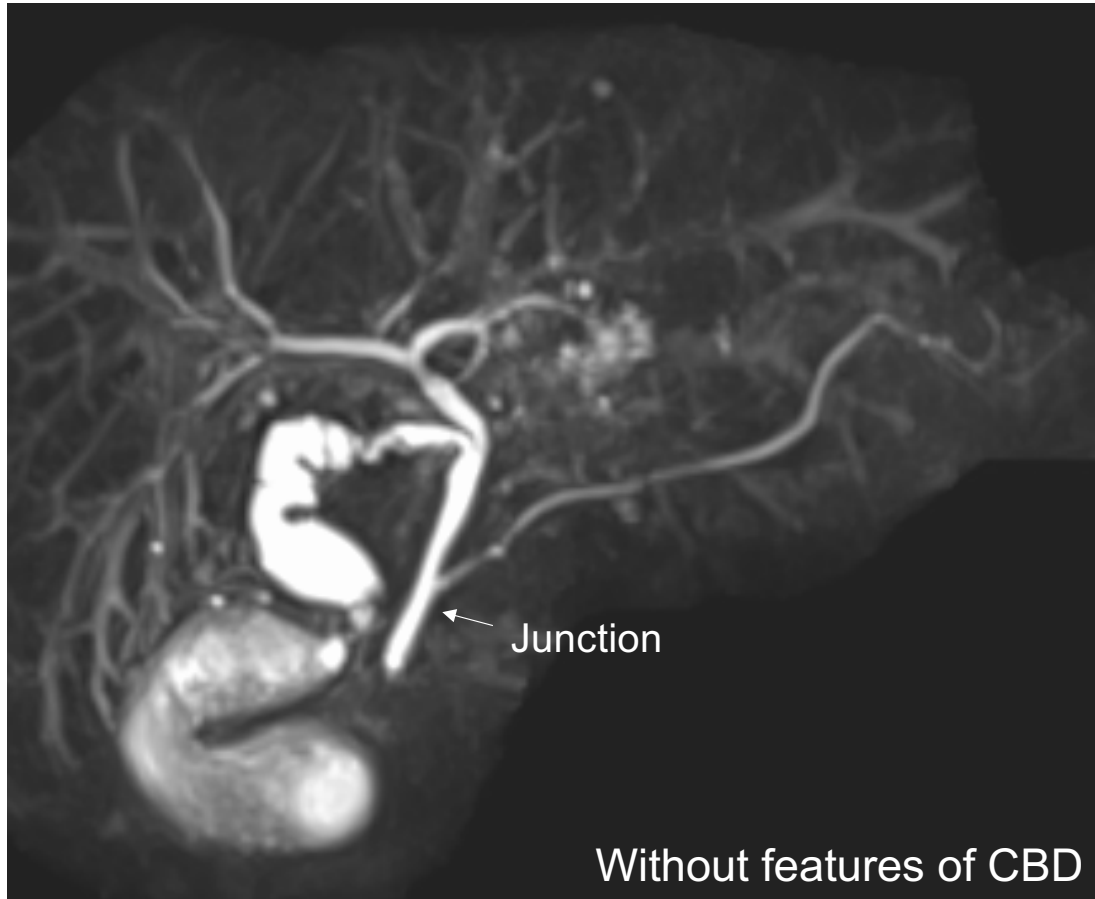


Without features of CBD



With features of CBD

PBM with normal BD diameter Adult cases



Case 35

Forme fruste (Ic) Adult case

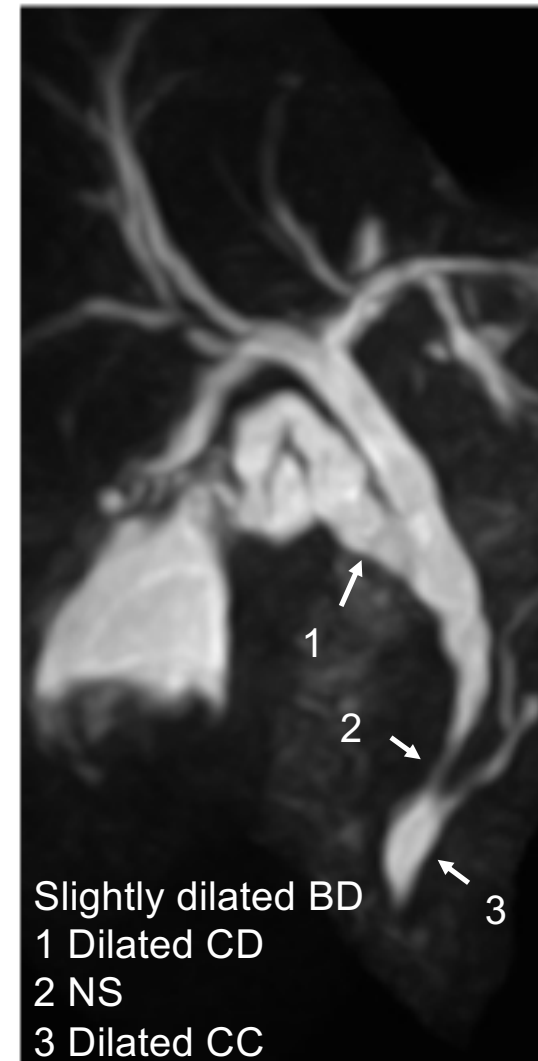
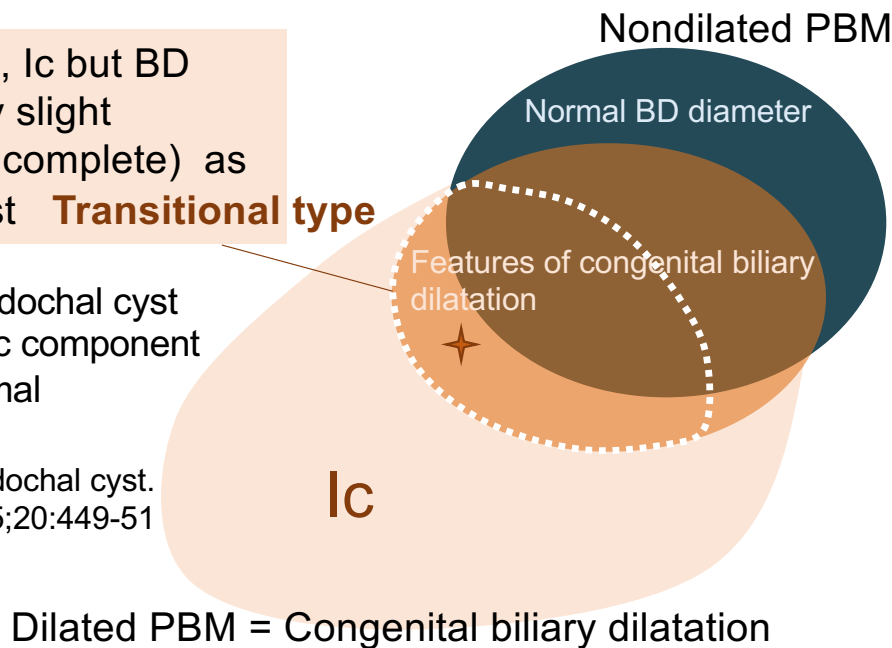
Female in her eighties
Ultrasound during medical check up detected
wall thickness at the fundus of the GBr
adenosquamous carcinoma in the GB

Strictly speaking, Ic but BD
dilatation is only slight
Forme fruste (incomplete) as
choledochal cyst **Transitional type**

A variant of choledochal cyst
in which the cystic component
is absent or minimal

Lilly JR et al.

Forme fruste choledochal cyst.
J Pediatr Surg 1985;20:449-51



Features of congenital biliary dilatation

Case 36

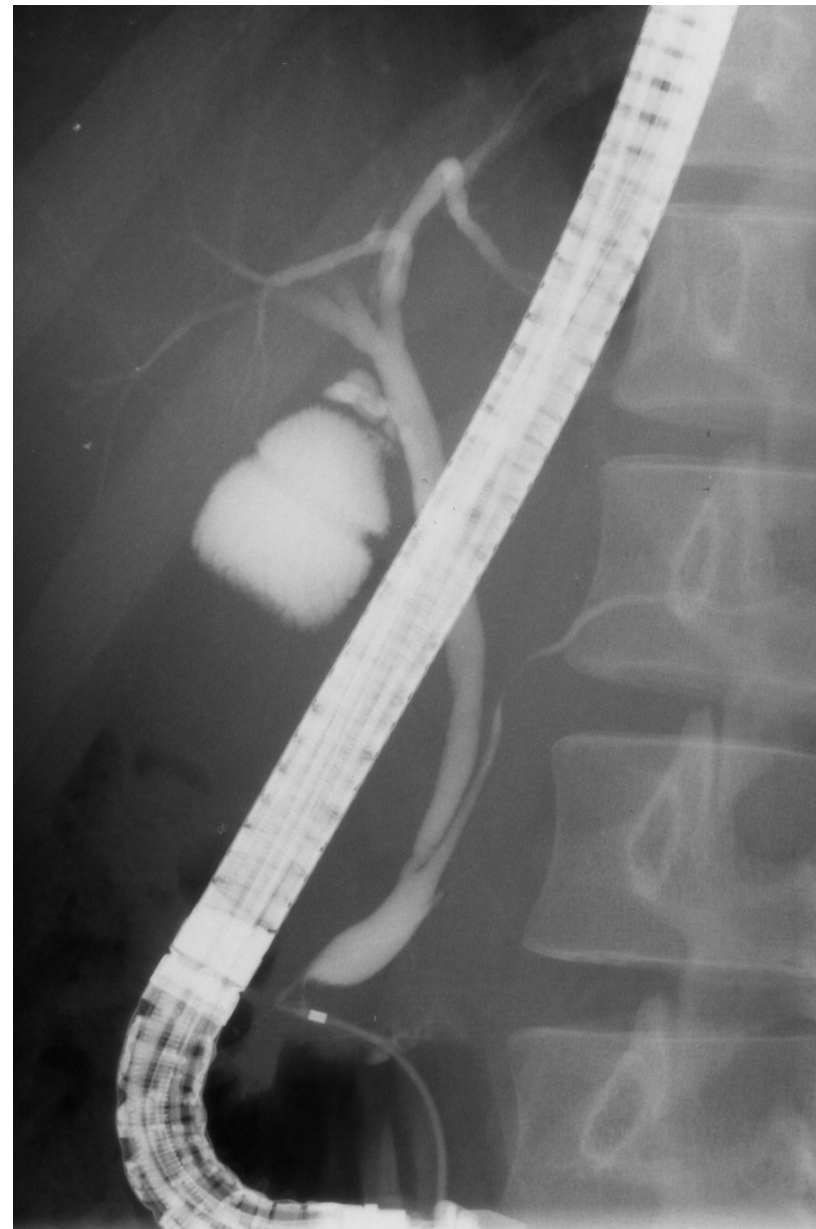
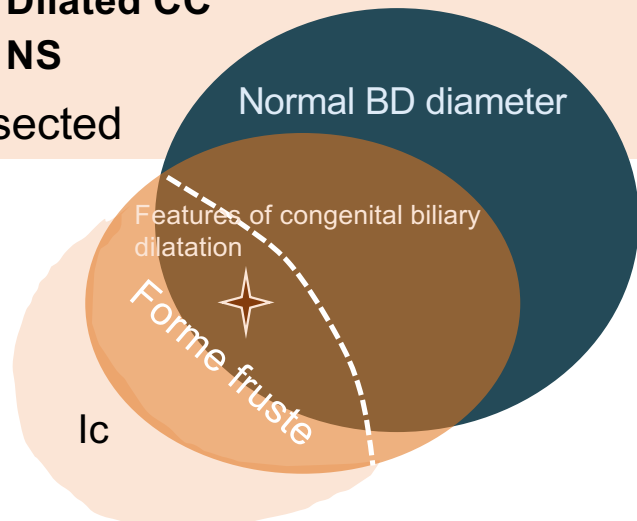
Pediatric case Normal BD diameter

Nondilated PBM < Forme fruste

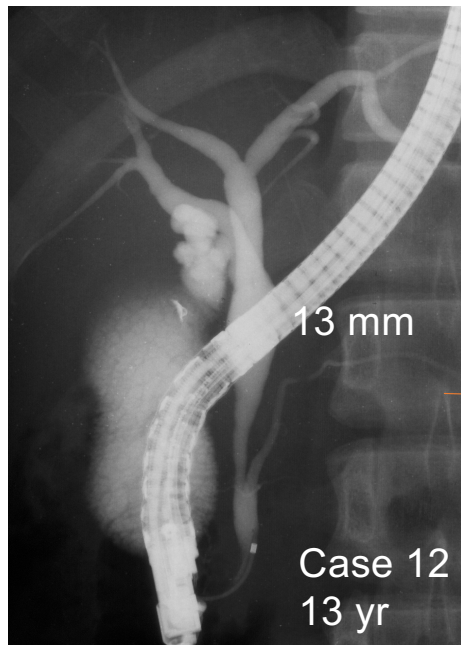
15-year-old girl
Ultrasound due to abdominal pain detected
gallbladder wall thickness

- CBD diameter
ERCP 6mm (Ultrasound Upper Limit 4.9 mm)
- **Features of congenital biliary dilatation**
 - ✓ Dilated CC
 - ✓ NS

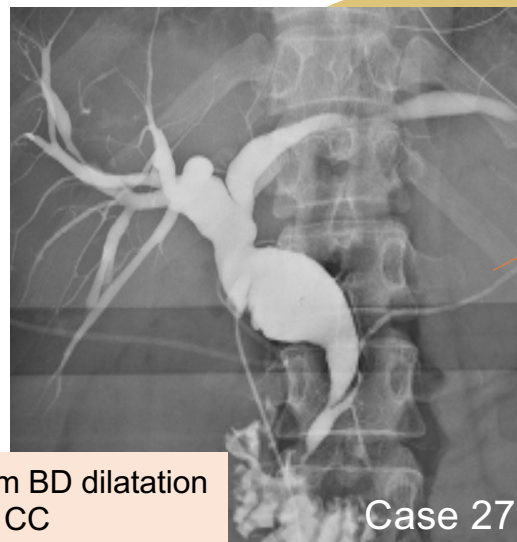
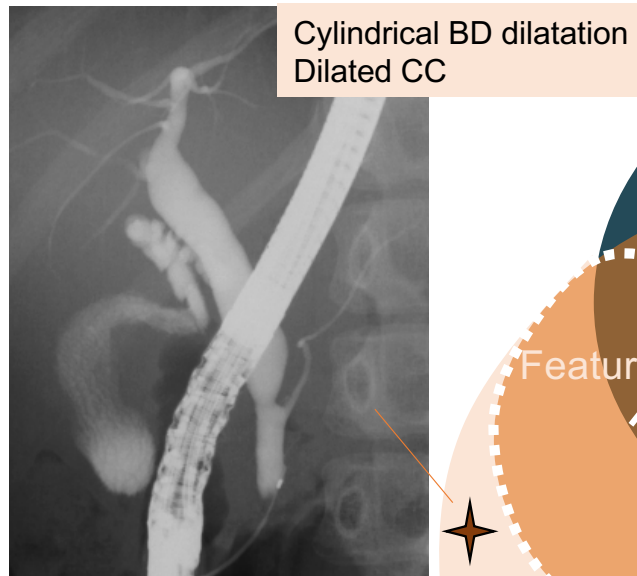
BD resected



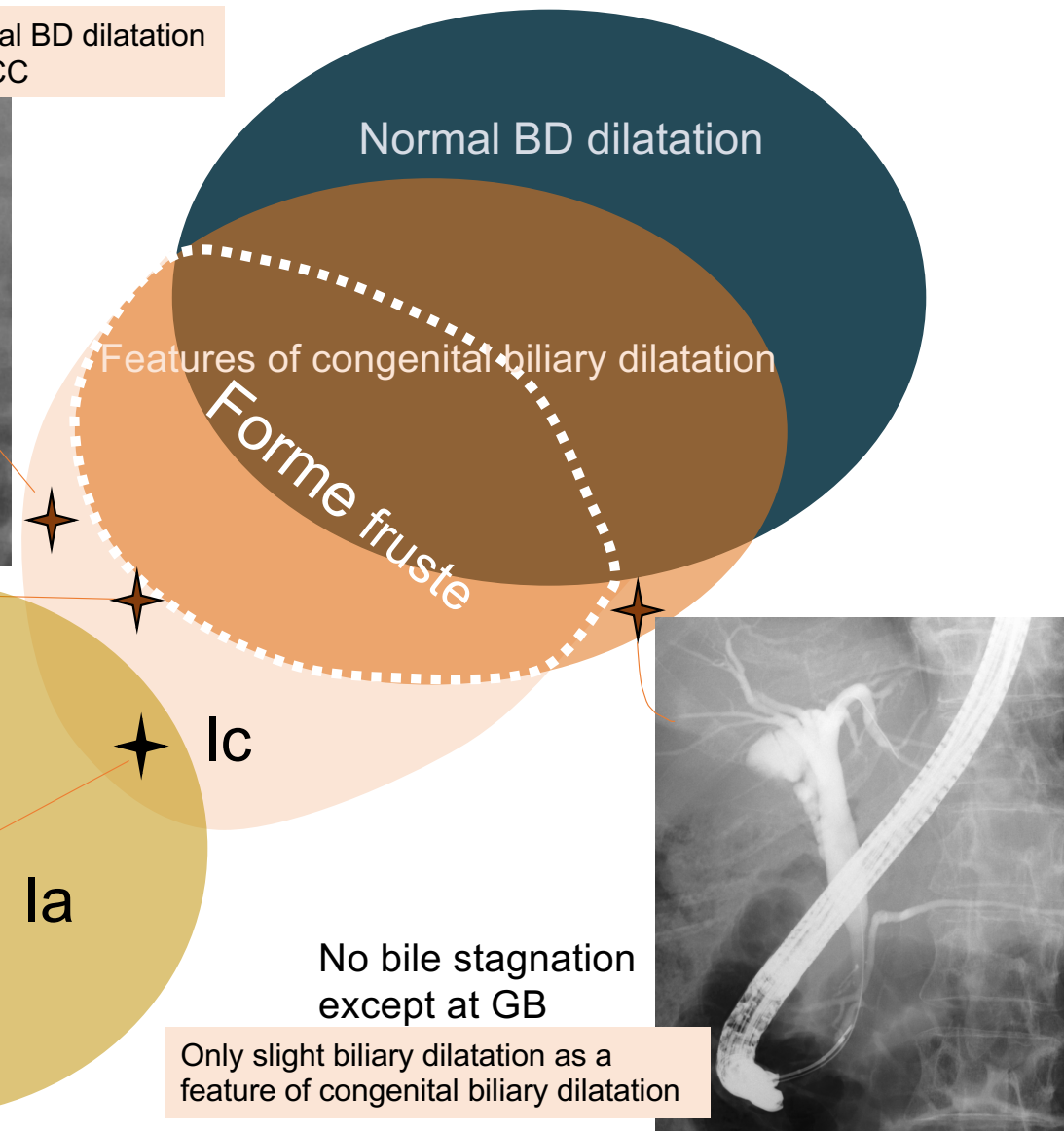
Around Forme fruste



Fusiform BD dilatation
Dilated CC
NS

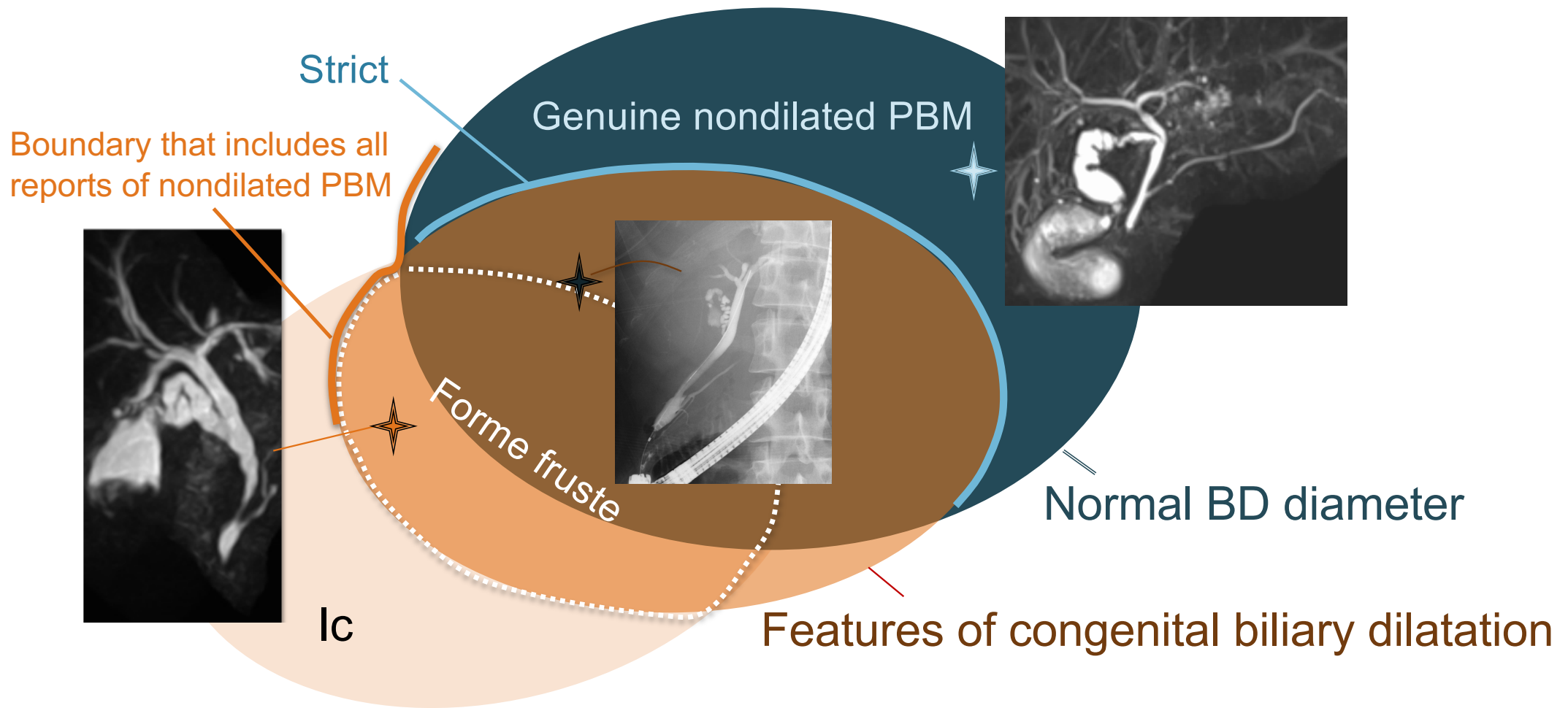


Fusiform BD dilatation
Dilated CC



Definition of Nondilated PBM

Where to draw the boundary on this spectrum?

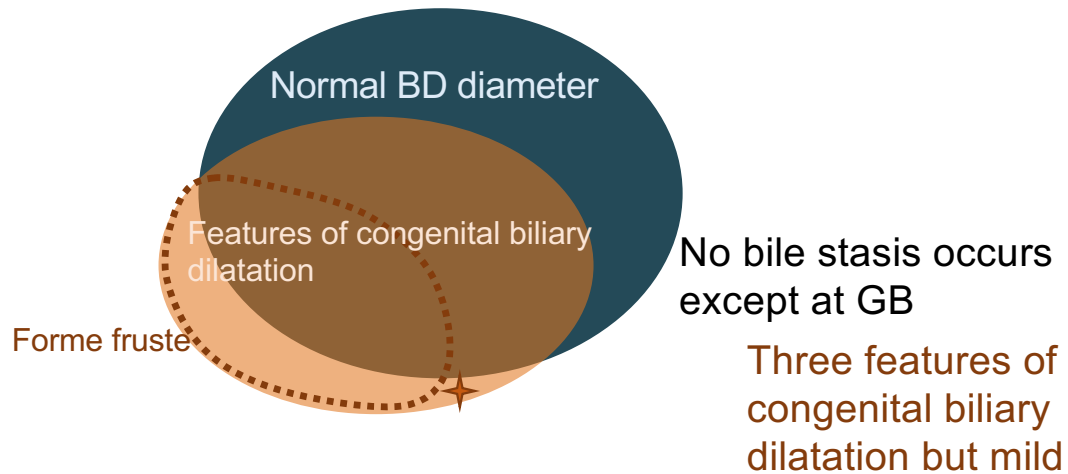


Case 37

Nondilated PBM >Forme fruste Pediatric case

14-year-old girl
Abdominal pain, fever, hyperamylasemia

- ERCP
CBD diameter 6 mm
(US upper limit 4.9 mm)
CHD diameter 8 mm
- BD resected

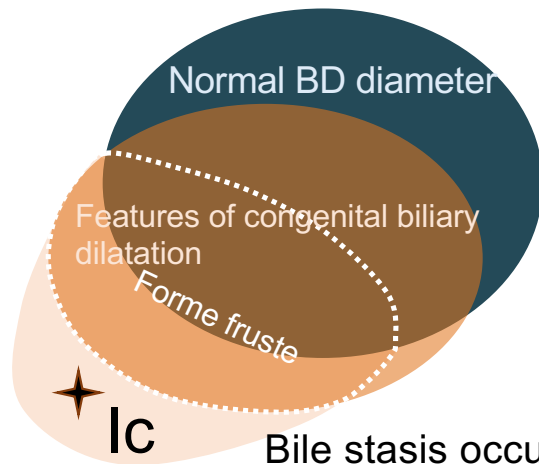


From basic section

Case 14

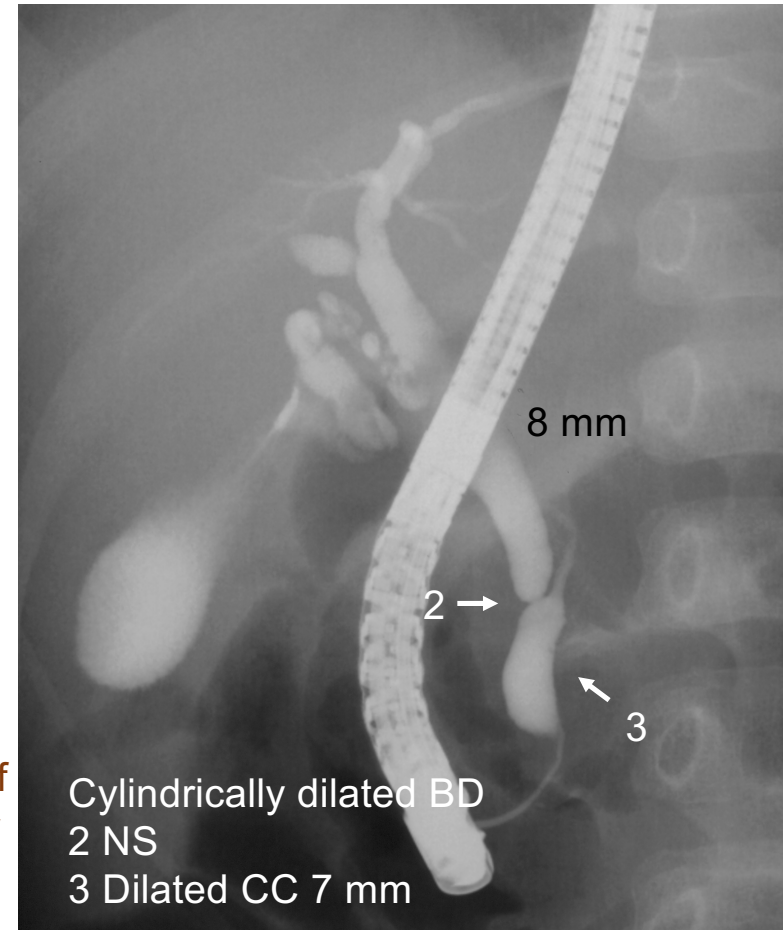
Ic Cylindrical dilatation Pediatric case

2-year-old boy
abdominal pain, vomiting
AST 896, ALT 416, Amy 1168
CT swollen pancreas
BD resection



Bile stasis occurs
at dilated BD and CC
as well as GB

Three features of
congenital biliary
dilatation



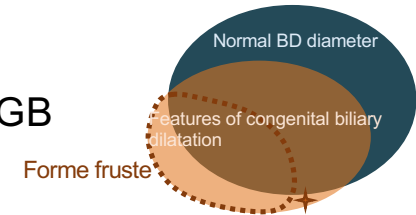
2 yr old, US upper limit 3.3 mm

Case 38

Adult case

Nondilated PBM > forme fruste >> Ic

Strically speaking, BD is dilated
However, no bile stagnate except at GB



Femal in her fifties
US during medical
check up detected GB
wall thickness

BD diameter
MRCP 8 mm

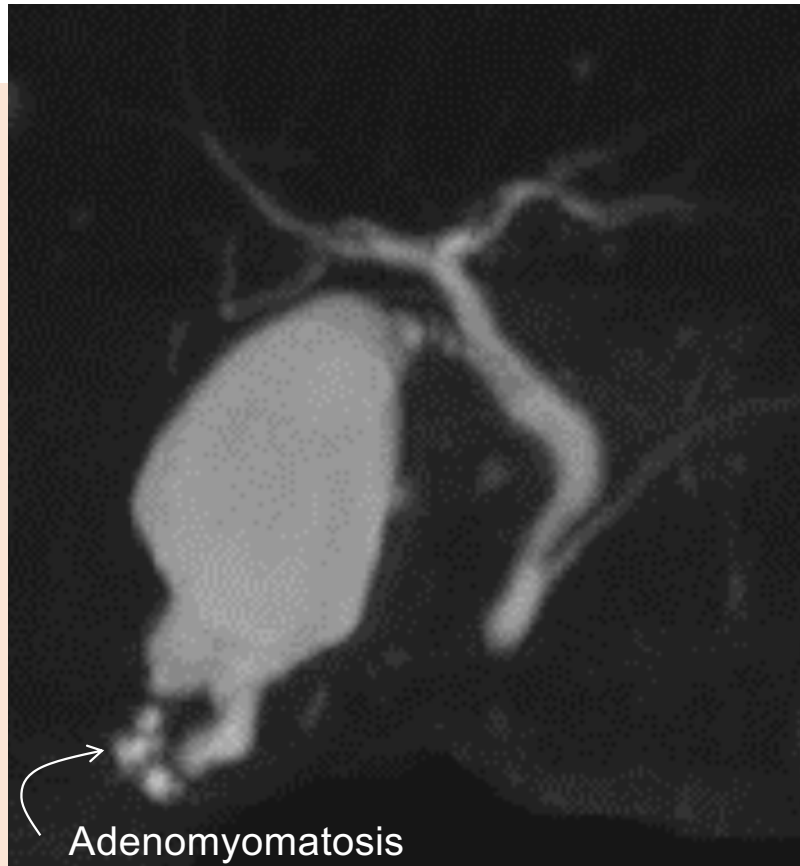
ERCP 9 mm

US upper limit 7.2 mm

Features of congenital
biliary dilatation are
mild

- ✓ **BD dilatation**
- ✓ **CC dilatation**
- ✓ **CD dilatation?**

Cholecystectomy



Advanced

PBM seemingly complicated

Rare type of PBM

First version, September 2024

Case 39

Protein plugs complicate PBM

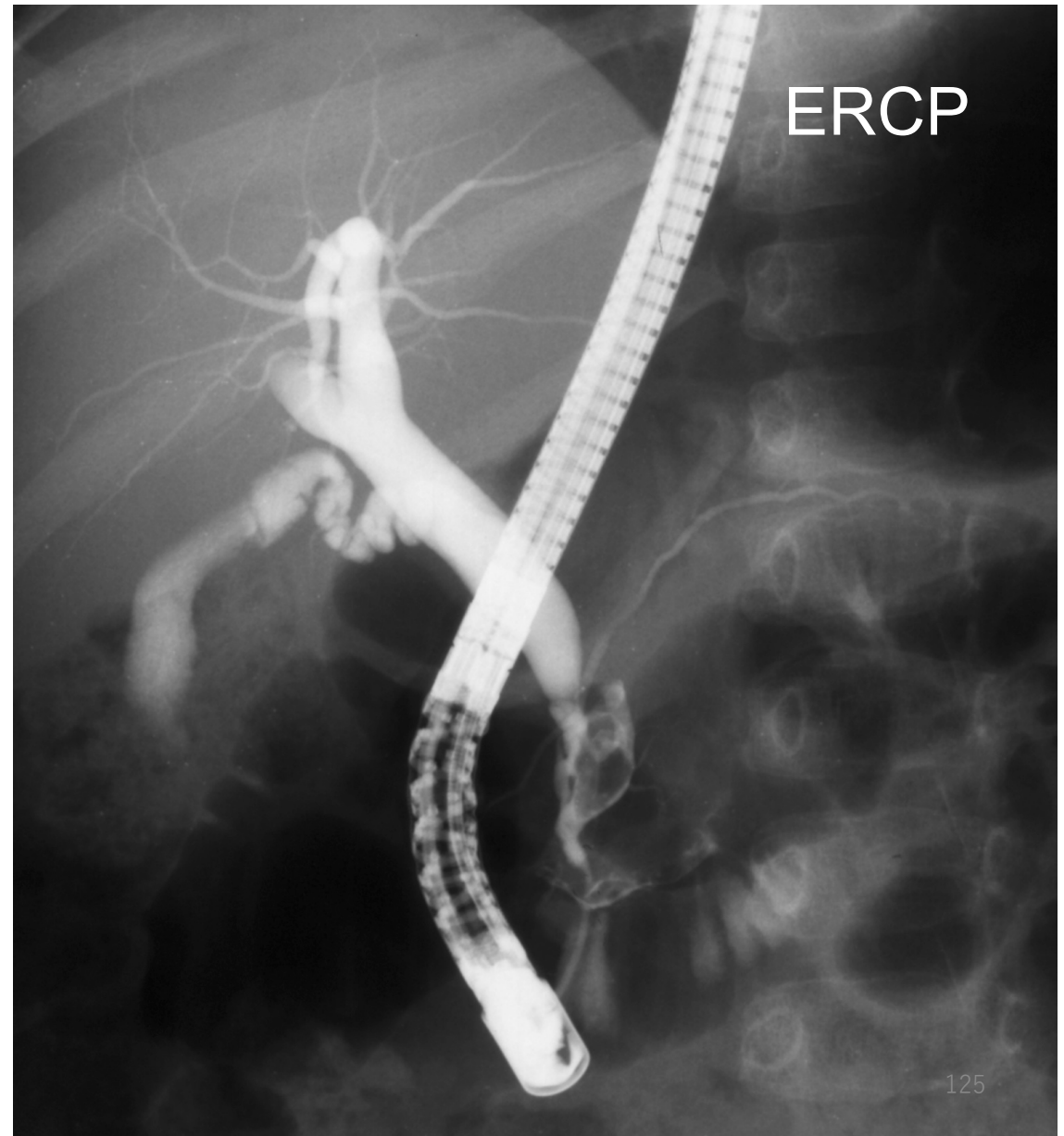
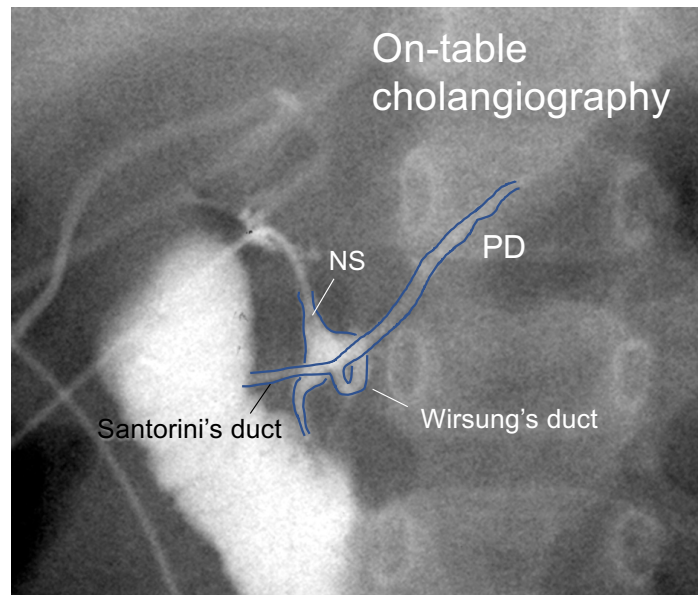
A 3-year old girl developed abdominal pain and vomiting, and jaundice after five days

Protein plugs compacted in the common channel caused dilatation, and complicated appearance of PBM.

She underwent BD resection after symptoms subsided.

On-table cholangiography showed disappearance of protein plugs, which disclosed the structure of PBM.

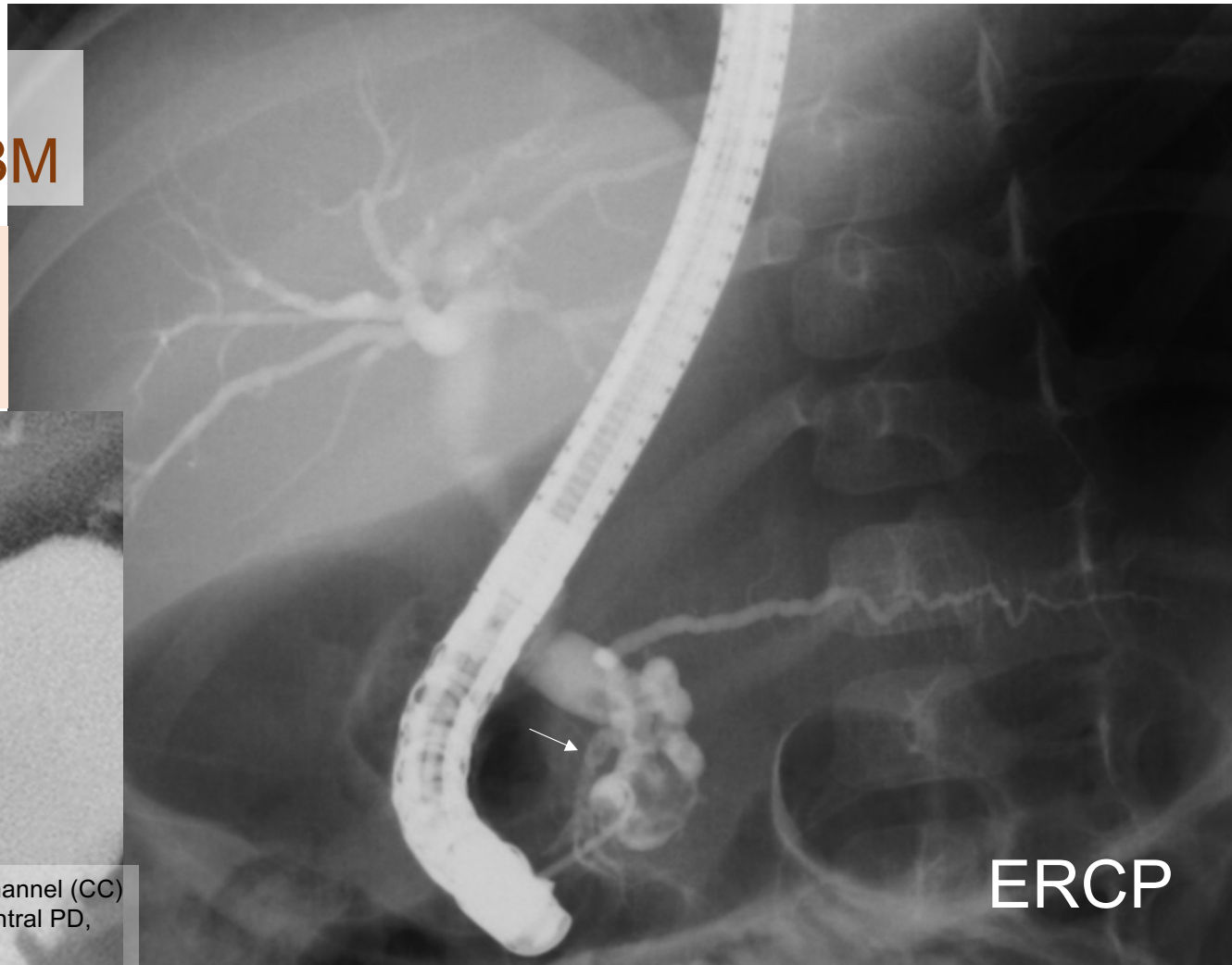
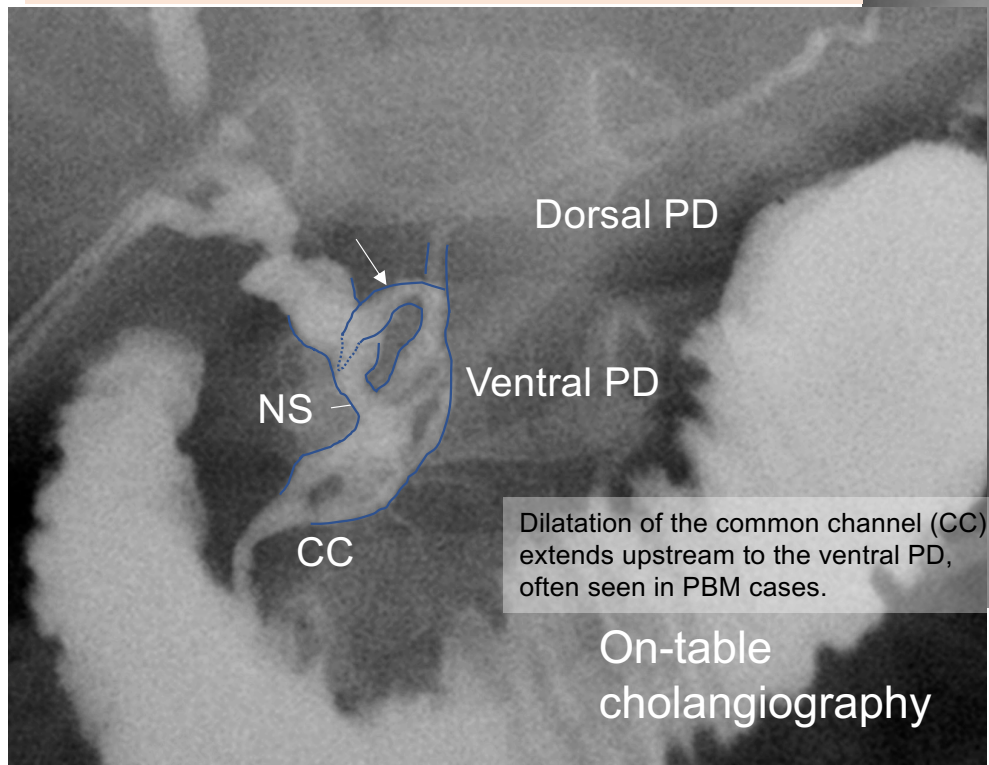
Dilatation of the common channel had disappeared.



Case 40

Protein plugs complicate PBM

A one-year-old girl developed acholic stool, abdominal pain, and jaundice with elevated serum levels of hepatobiliary enzymes. Protein plugs compacted in the common channel complicated appearance of PBM. Most of plugs had disappeared until surgery



Arrows indicate the same PD branch.

Case 41

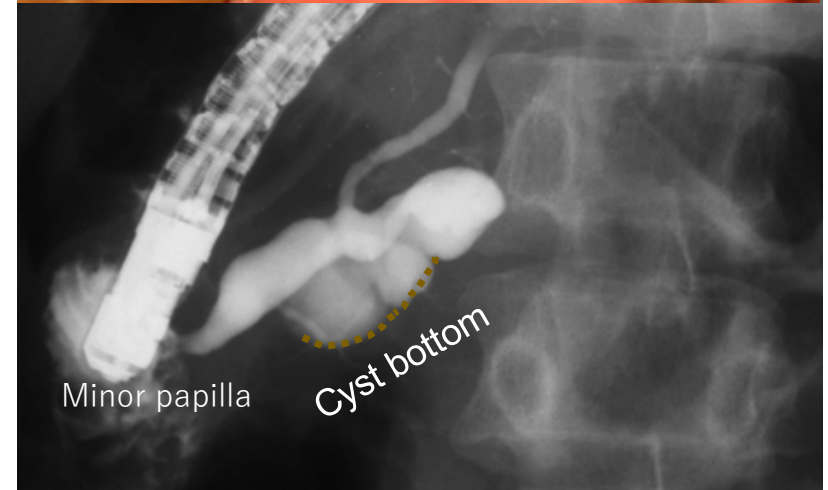
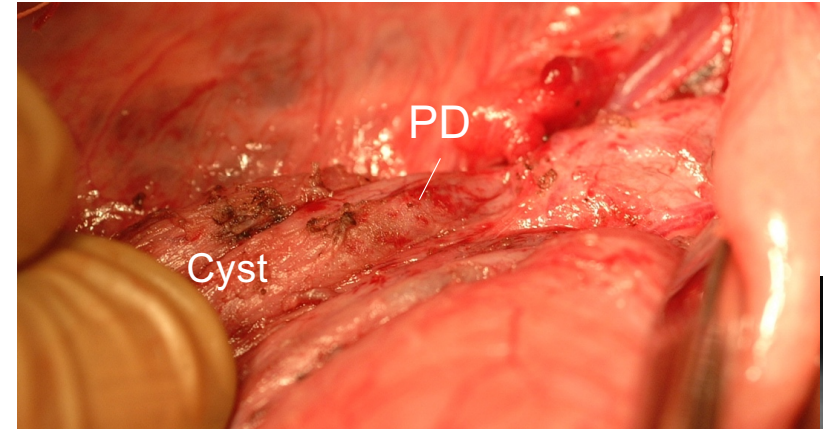
A female in her twenties developed abdominal pain with hyperamylasemia caused by protein plugs.

The narrow segment, that the cyst must have, was not found during surgery, ERCP was performed to disclose the relationship with PD. The guidewire through the main pailla was observed running at the bottom of the cyst.

Main (Ventral) pancreatic duct forms cyst bottom



Dotted line of the cyst was composed of PD



Pancreatography through the minor papilla after cyst excision

Case 42

Rare type Bile duct joins the inferior branch of PD

Male in his twenties
Ultrasonography found a dilated BD
during his medical check-up.

Type Ia congenital biliary dilatation

- **BD joins the inferior branch of PD**

This case supports Matsumoto's theory:
PBM is the malformation where the
choledochus misconnects with the ventral PD or
its branches outside the duodenal wall.
See P120.

