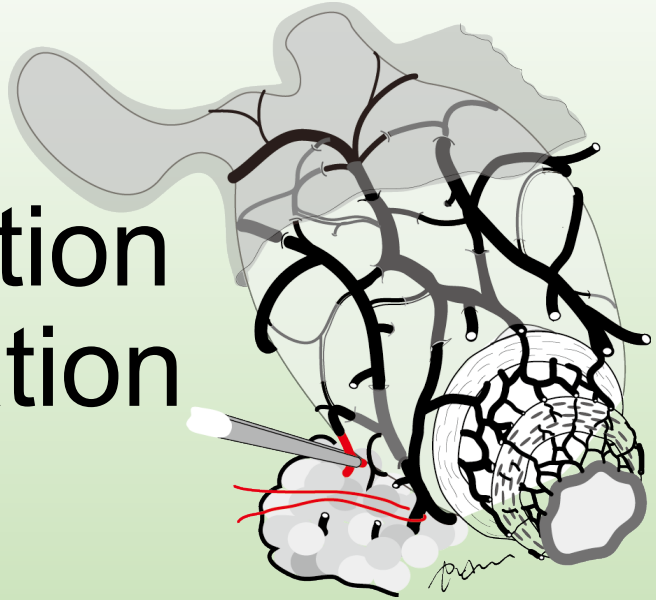
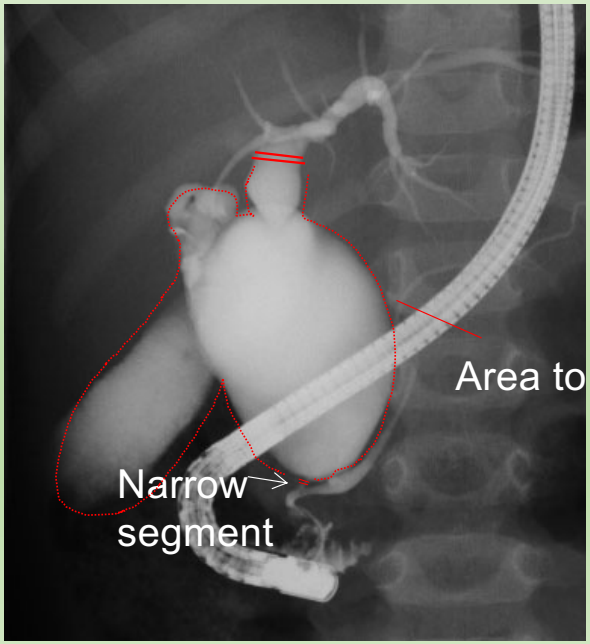
# Atlas of Pancreaticobiliary Maljunction & Congenital Biliary Dilatation






Japanese Study Group on Pancreaticobiliary Maljunction (JSGPBM)
Scientific Committee

Editor-in-Chief, Kenitiro Kaneko
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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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# Advanced

## Issues on Todani's classification

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

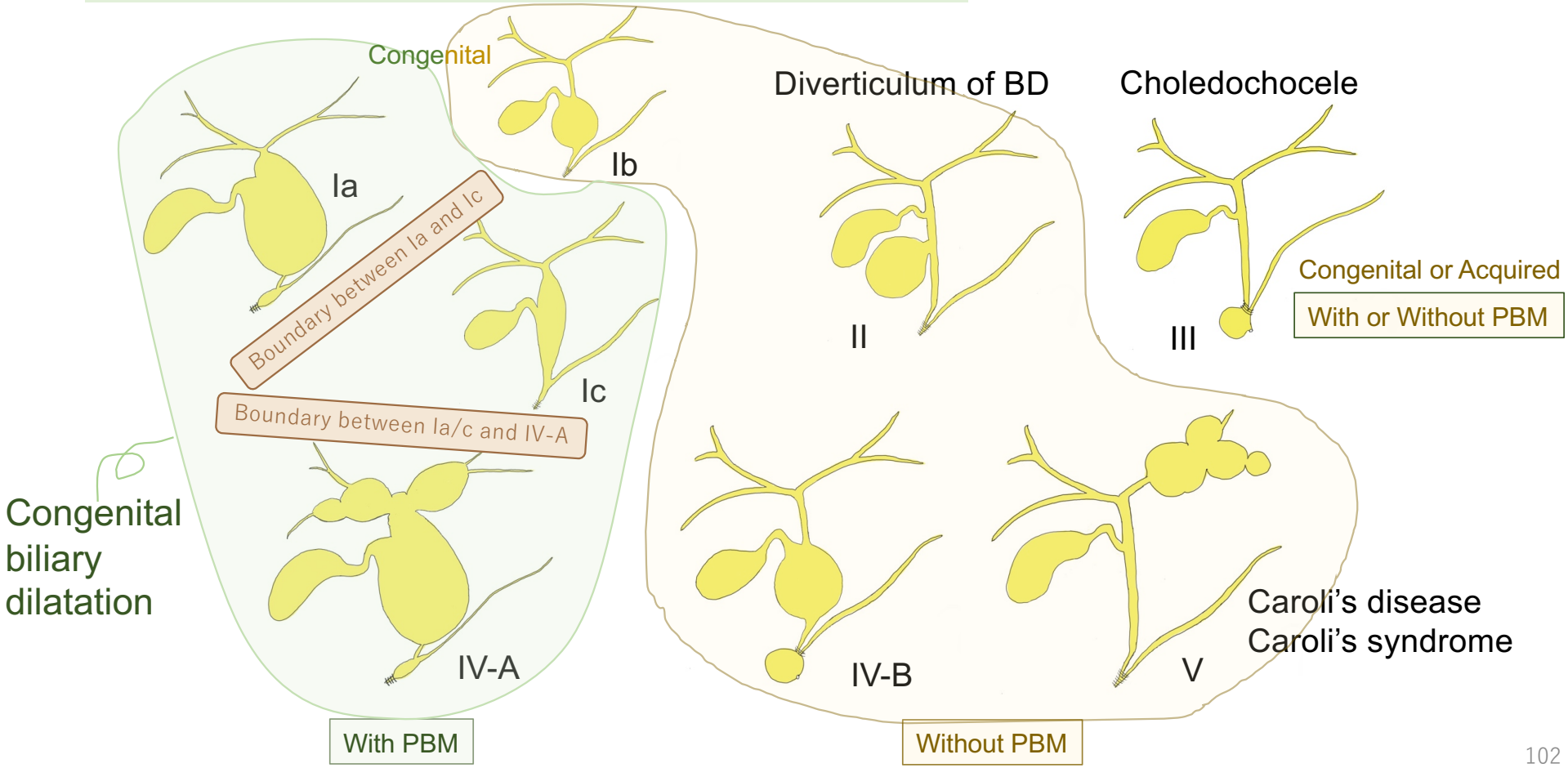
First version, September 2024

2nd version, September 2025

Revised: September 2026

# 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 type IV-A.

# Todani type IV

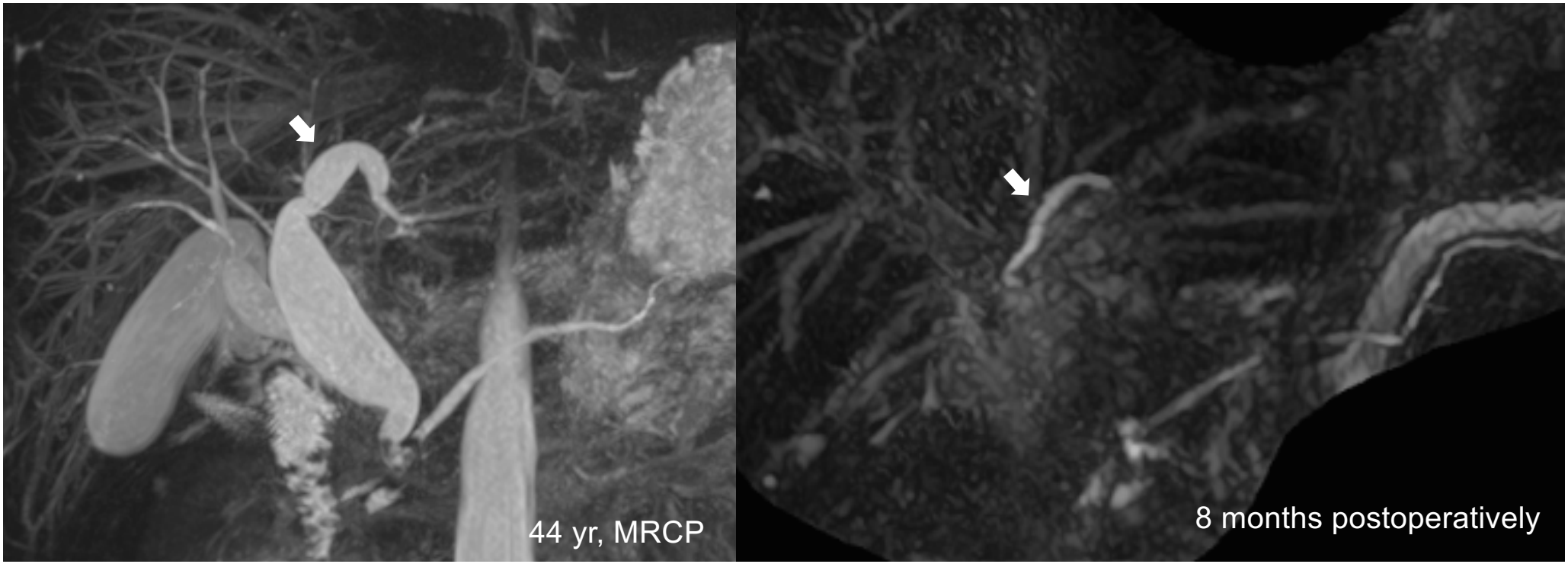
**Todani T**, Watanabe Y, Toki A, Morotomi Y.
Classification of congenital biliary cystic disease: special reference to type Ic and IVA cysts with primary ductal stricture.
*J Hepatobiliary Pancreat Surg*. 2003;10(5):340-4.
doi: 10.1007/s00534-002-0733-7. PMID: 14598133.

- **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 classify as type I or IV-A.

# Intrahepatic BD dilatation diminishes after BD resection
# Adult case, Todani type Ic




## 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.

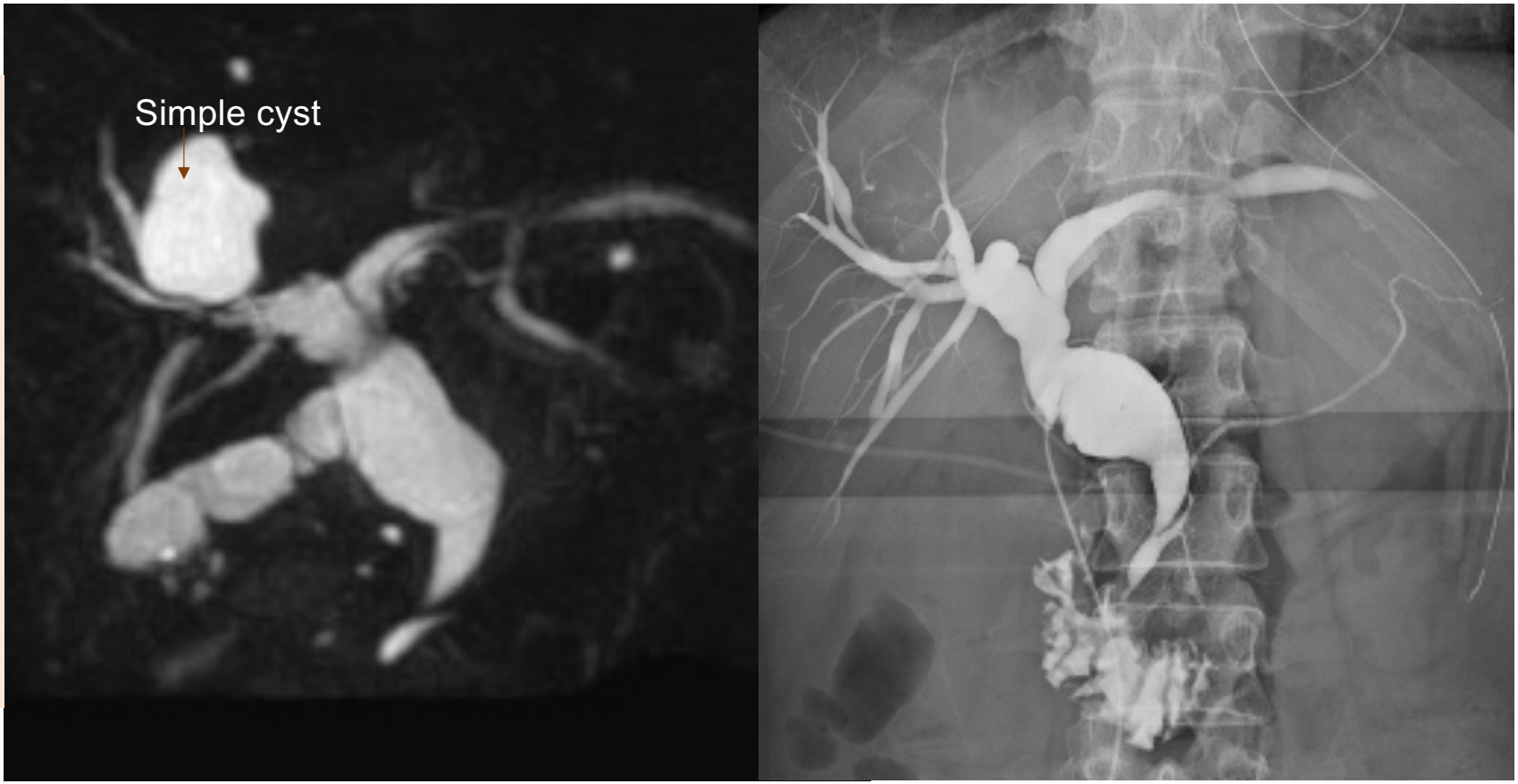
# Case 27

## Ic > Ia >> IV-A

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

A female in her 30s
Abdominal pain

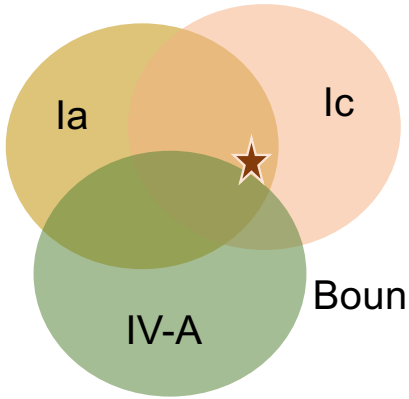
- Fusiform (Ic) rather than cystic (Ia) dilatation of the EHBD.
- Continuous dilatation extends to the right and left hepatic ducts.
  (See definition of Ic on p103)
- Narrow segment with a dilated common channel, corresponding to type C PBM
  (See JSGPBM classification on p36)



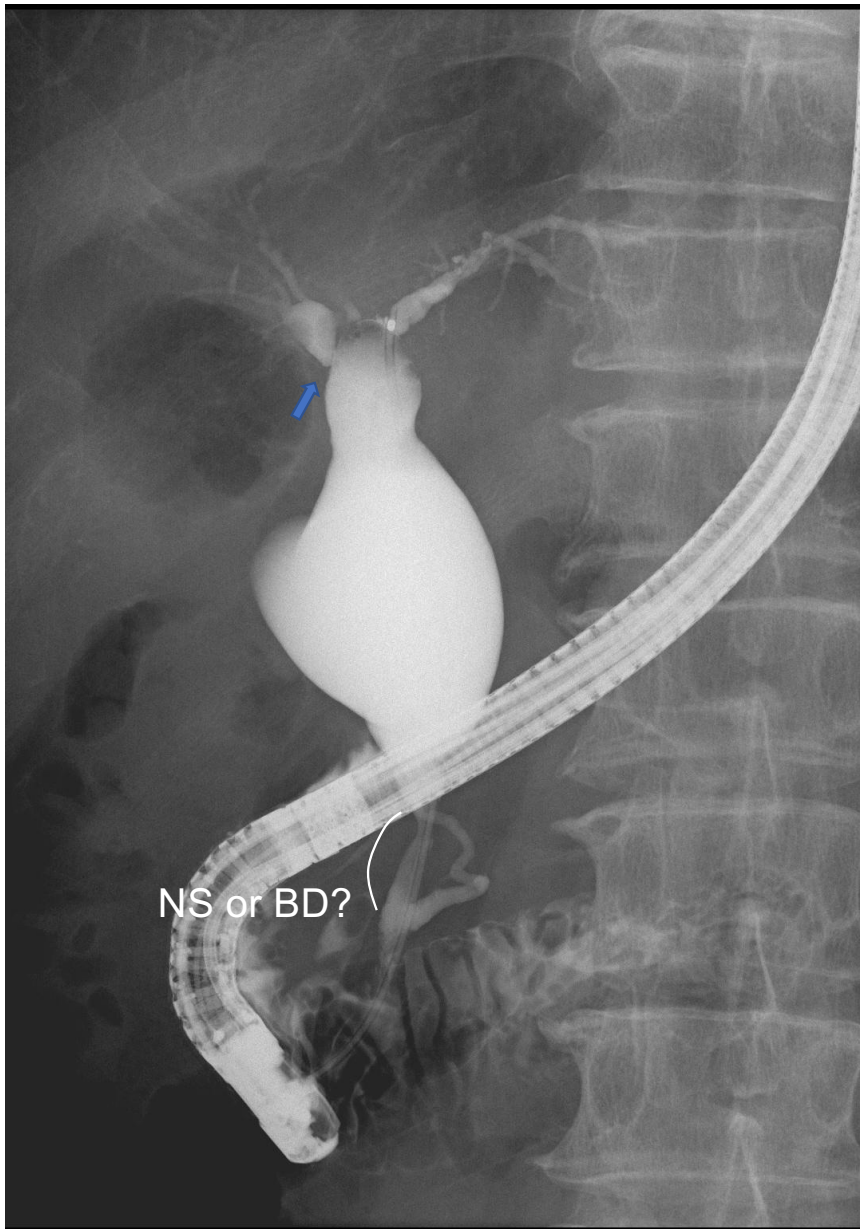
# Case 28 Ic > Ia > IV-A

ERCP in a male in his 70s
Abdominal pain and fever

- Slight dilatation of the RHD with a stenosis (arrow) that vanished 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 pp136–9)




Boundaries are arbitrary.

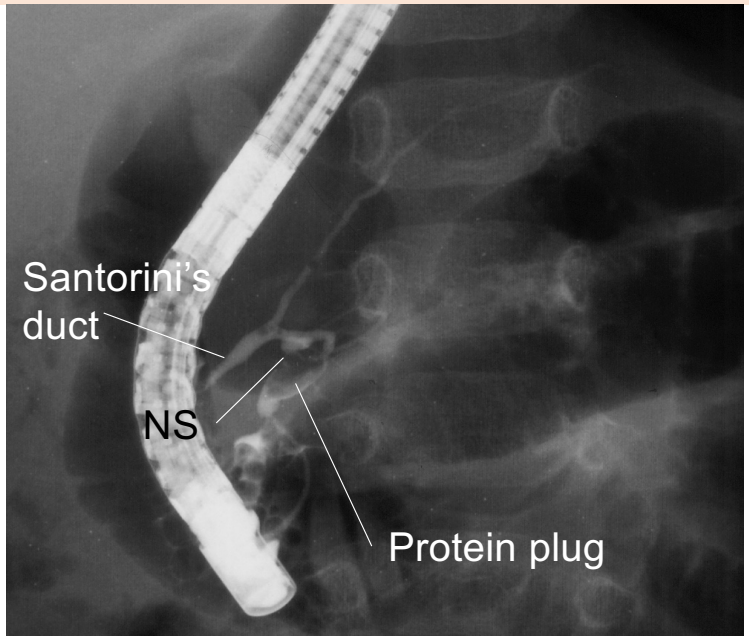


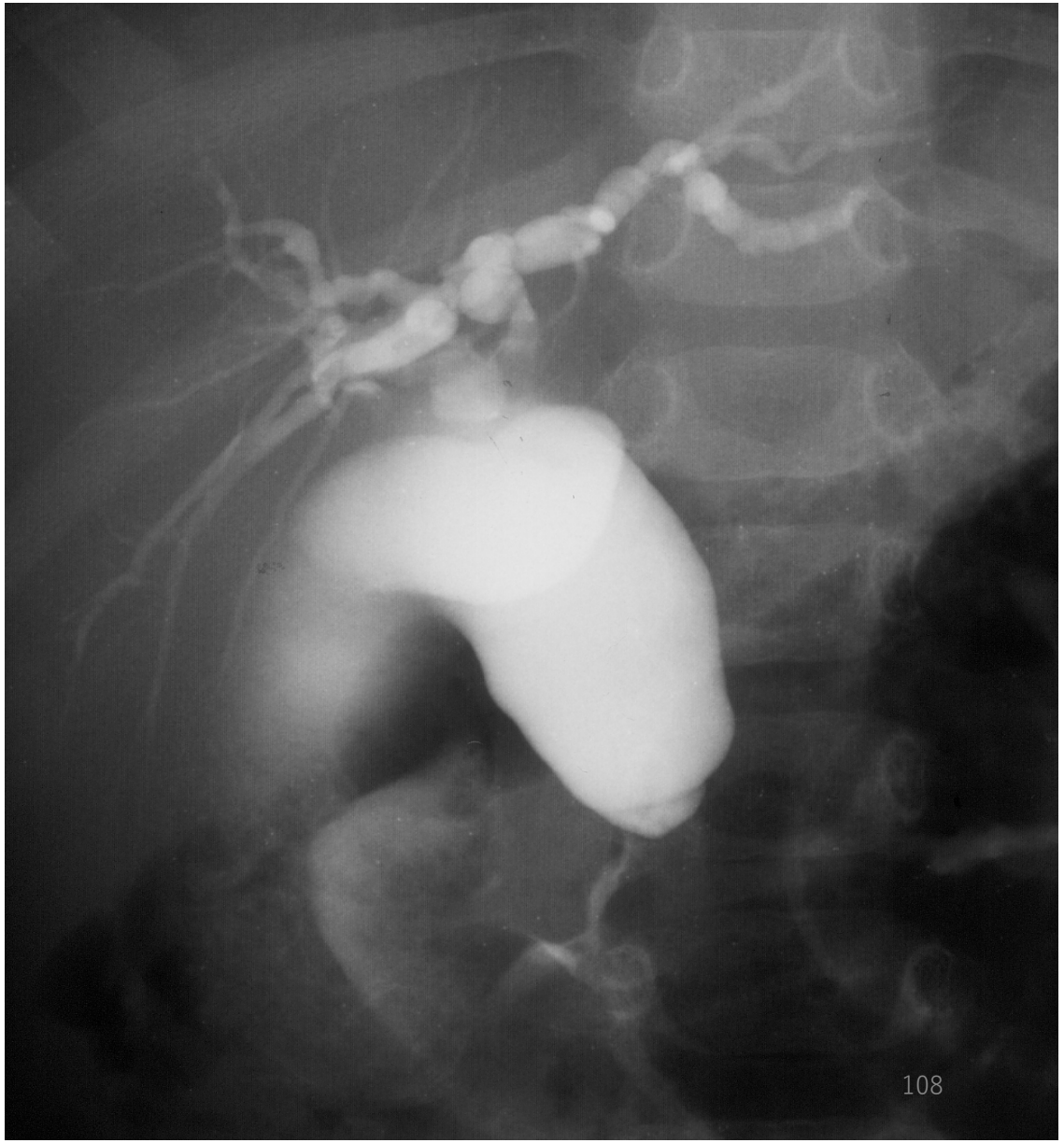
# Case 29

## Ia > IV-A

ERCP in a 9-year-old child
Abdominal pain, jaundice, acholic stool

Transient diffuse intrahepatic bile duct (IHBD) dilatation occurred due to protein plug obstruction.
The dilatation diminished after BD resection.

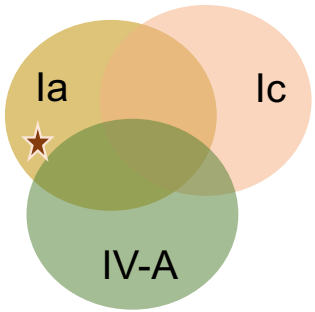


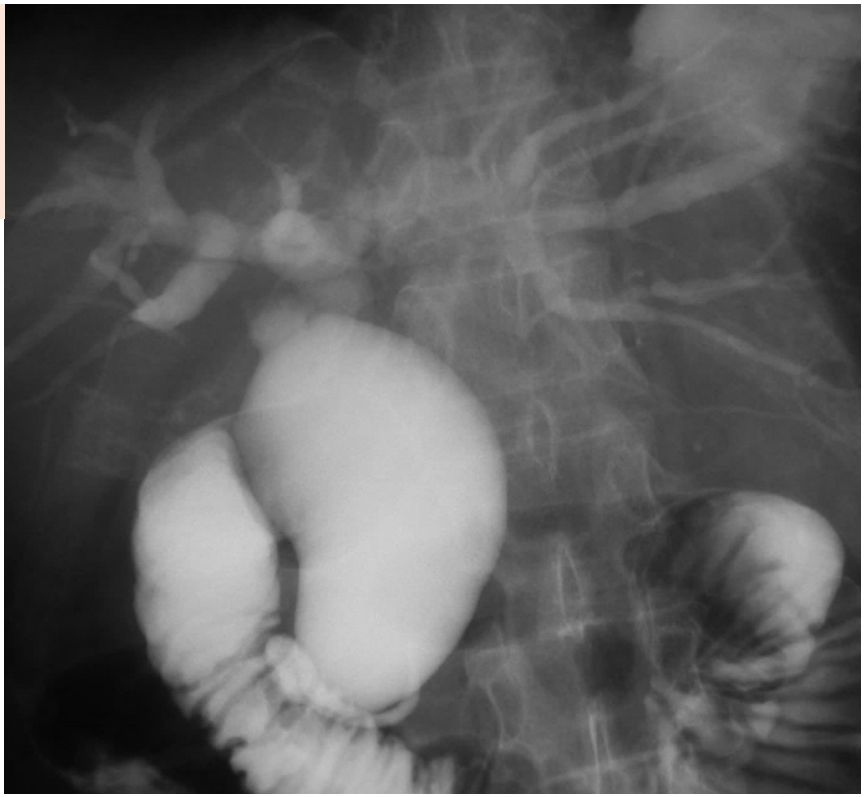



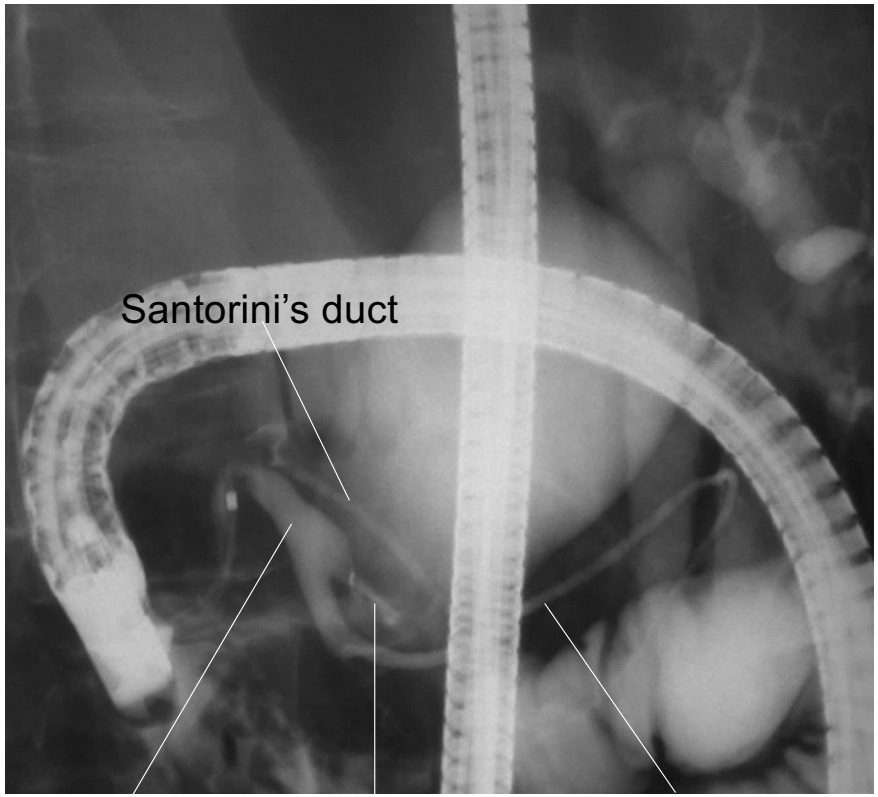
# Case 30

## Ia > IV-A

A female in her 50s. ERCP shows diffuse dilatation of the IHBD.





# Case 31

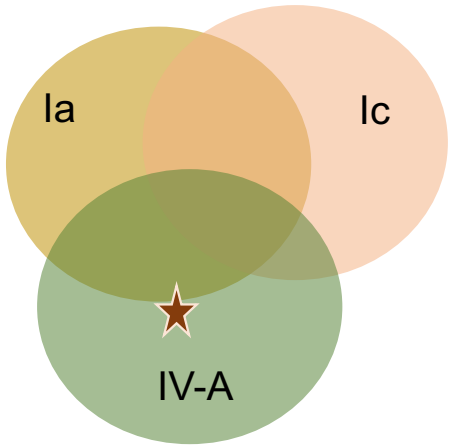
# IV-A > Ia

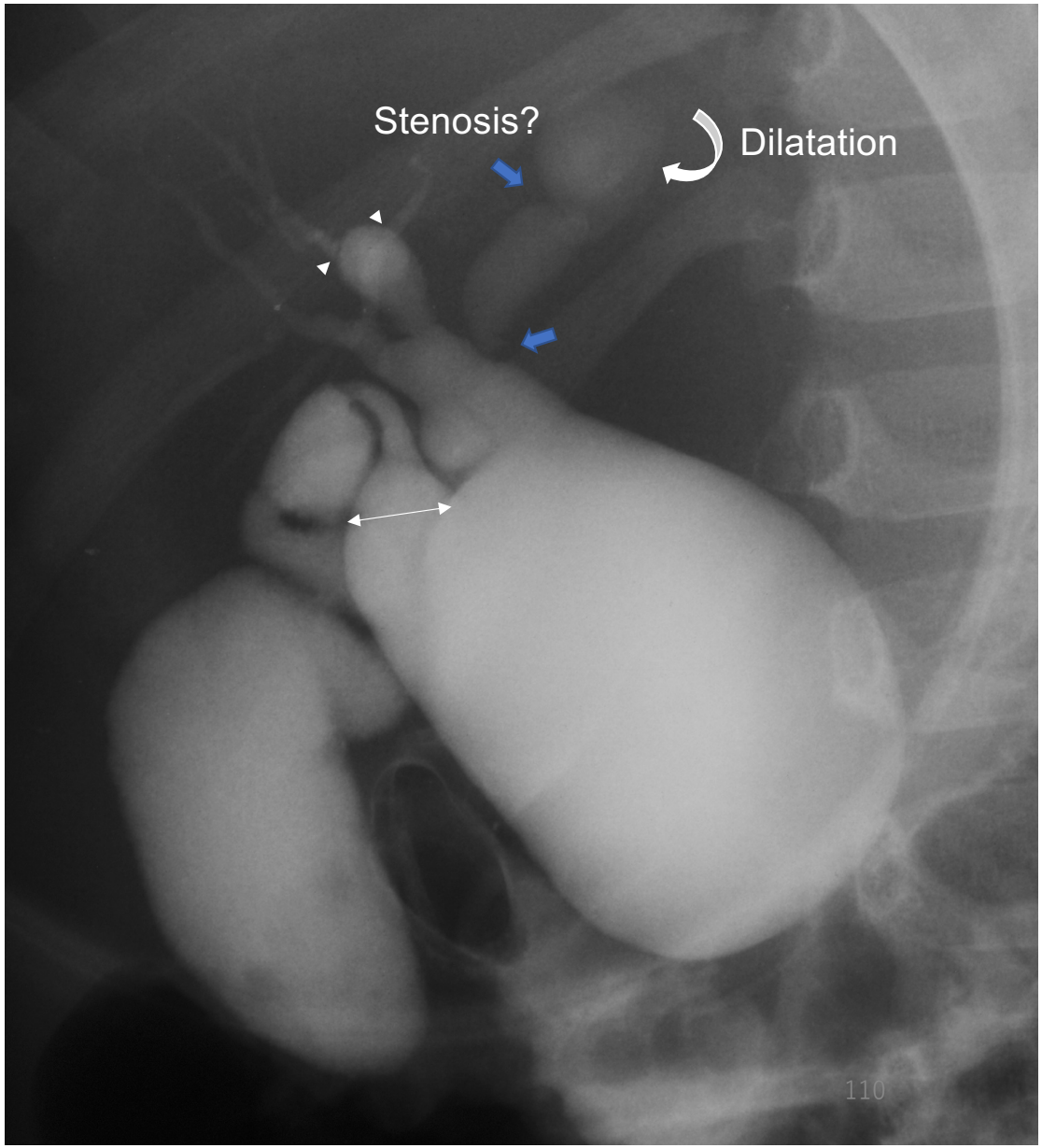
ERCP in a 2-year-old girl
Abdominal pain and vomiting

- Cystic dilatation upstream from the LHD (curved arrow) with constrictions (arrows)
- Limited dilatation (arrowheads)

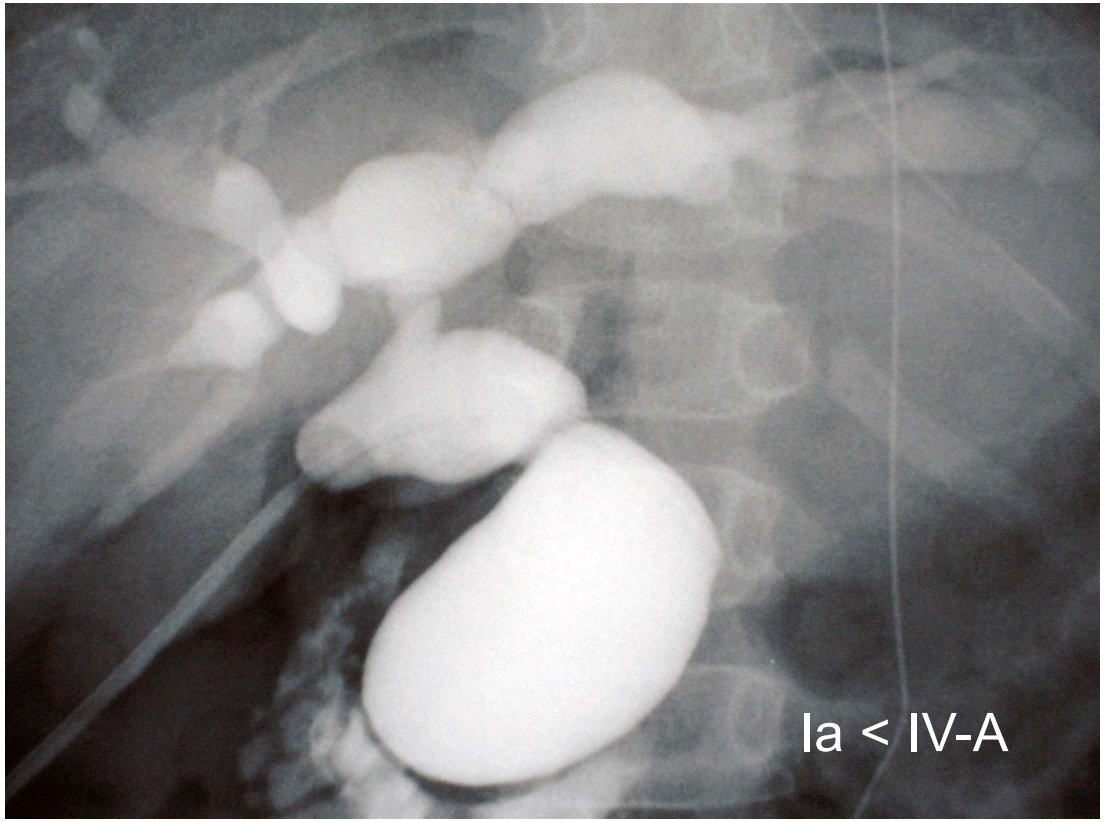
indicating type IV-A

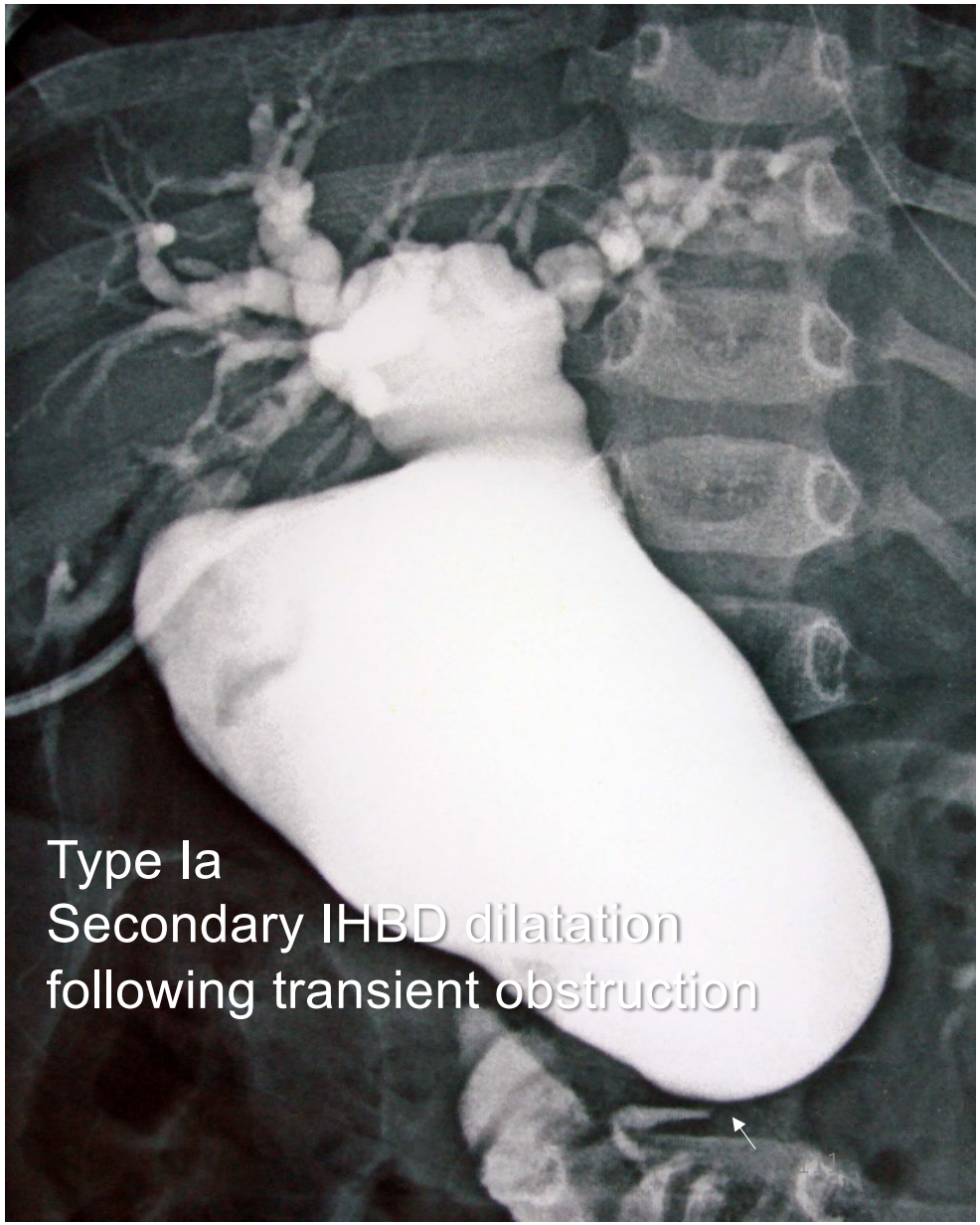
- Prominent dilatation at the base of the cystic duct (double-headed arrow)



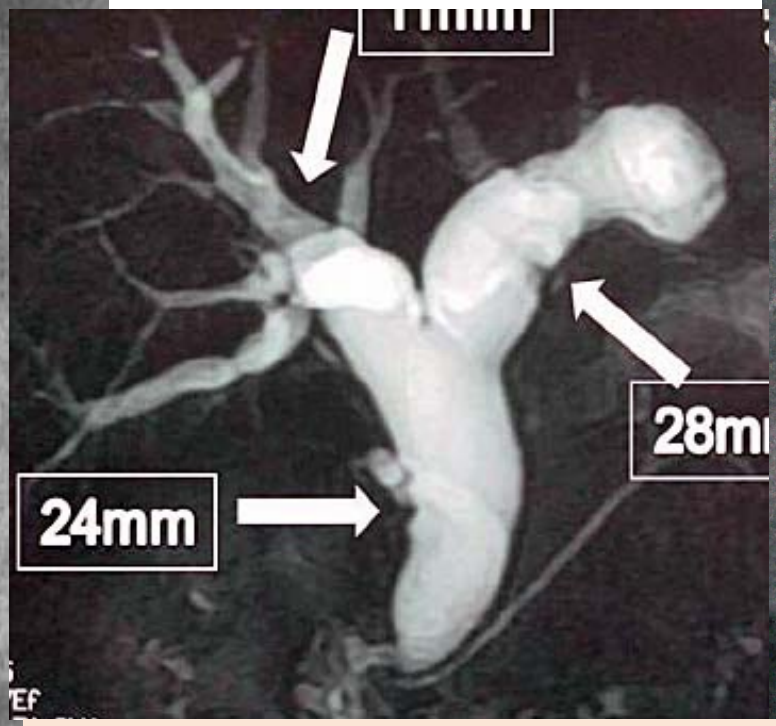
# Two cases: Ia or IV-A?



# Three cases: Ic or IV-A?




Diffuse dilatation without stenosis, but with pesisitent dilatation after BD resection

Type IV-A??

Diffuse dilatation with LHD stenosis, that diminished after BD resection



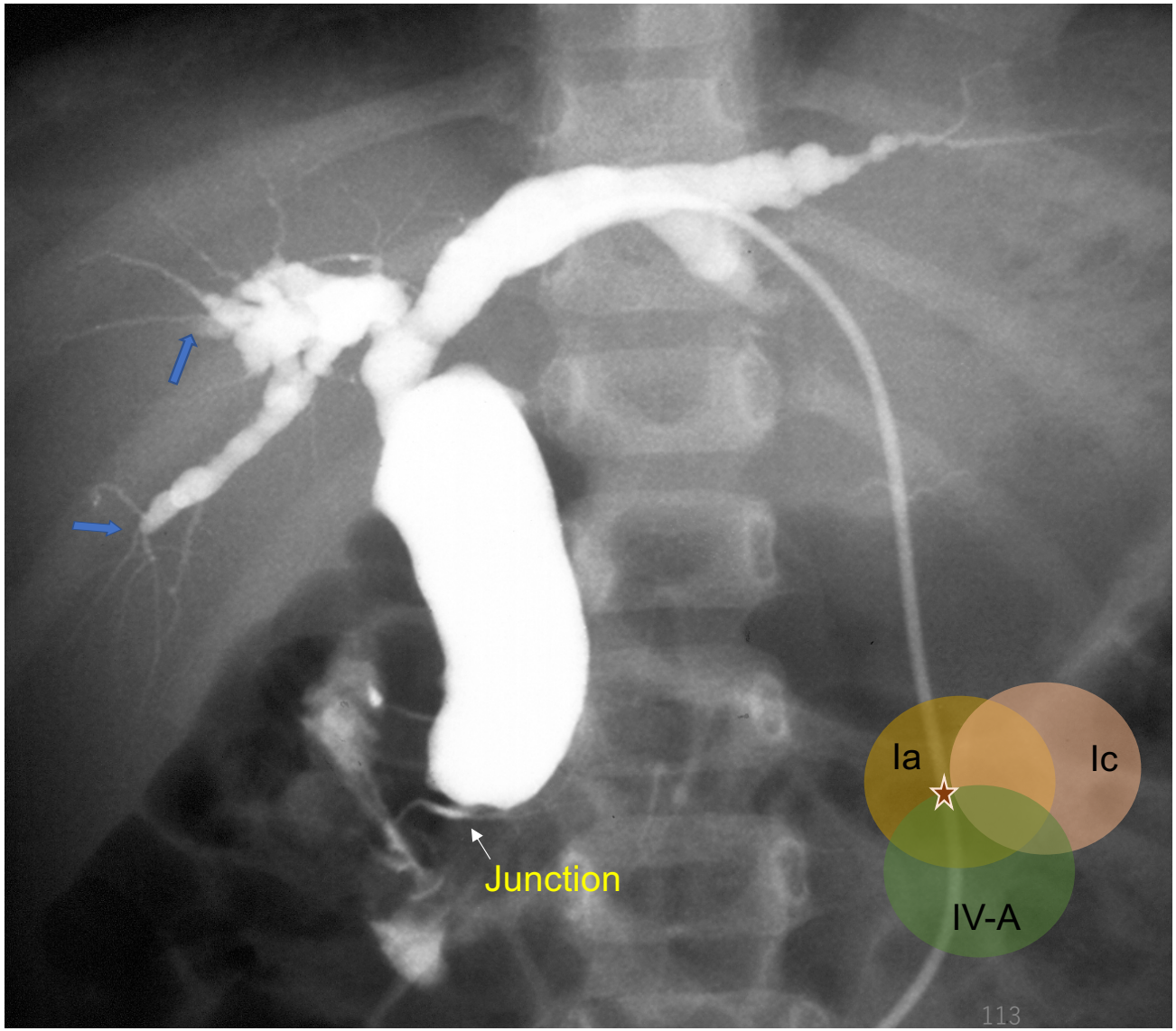
# Case 32 Ia or IV-A?

## Borderline case

Percutaneous transhepatic cholangiography in a 2-year-old boy
Abdominal pain, jaundice, elevated serum hepatobiliary enzymes

- **Cystically 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, corresponding to type A (stenotic type) PBM

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



# Case 33

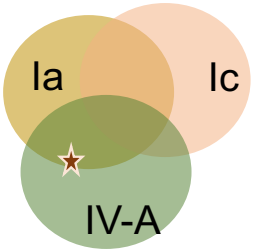
# Ia or IV-A?

## Borderline case

A female in her 20s. Percutaneous transhepatic biliary drainage was performed due to fever, jaundice, and abdominal pain.

The RHD is diffusely dilated without strictures, while the LHD is slightly dilated with a stenosis (arrow).
BD resection with stenosis treatment would relieve these dilatations.

Ia or IV-A: Questionable







Diffusely dilated RHD but without strictures

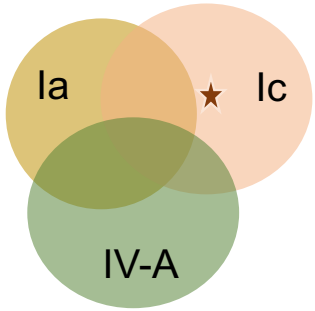
Stenosis at the orifice, but with only slightly dilated LHD

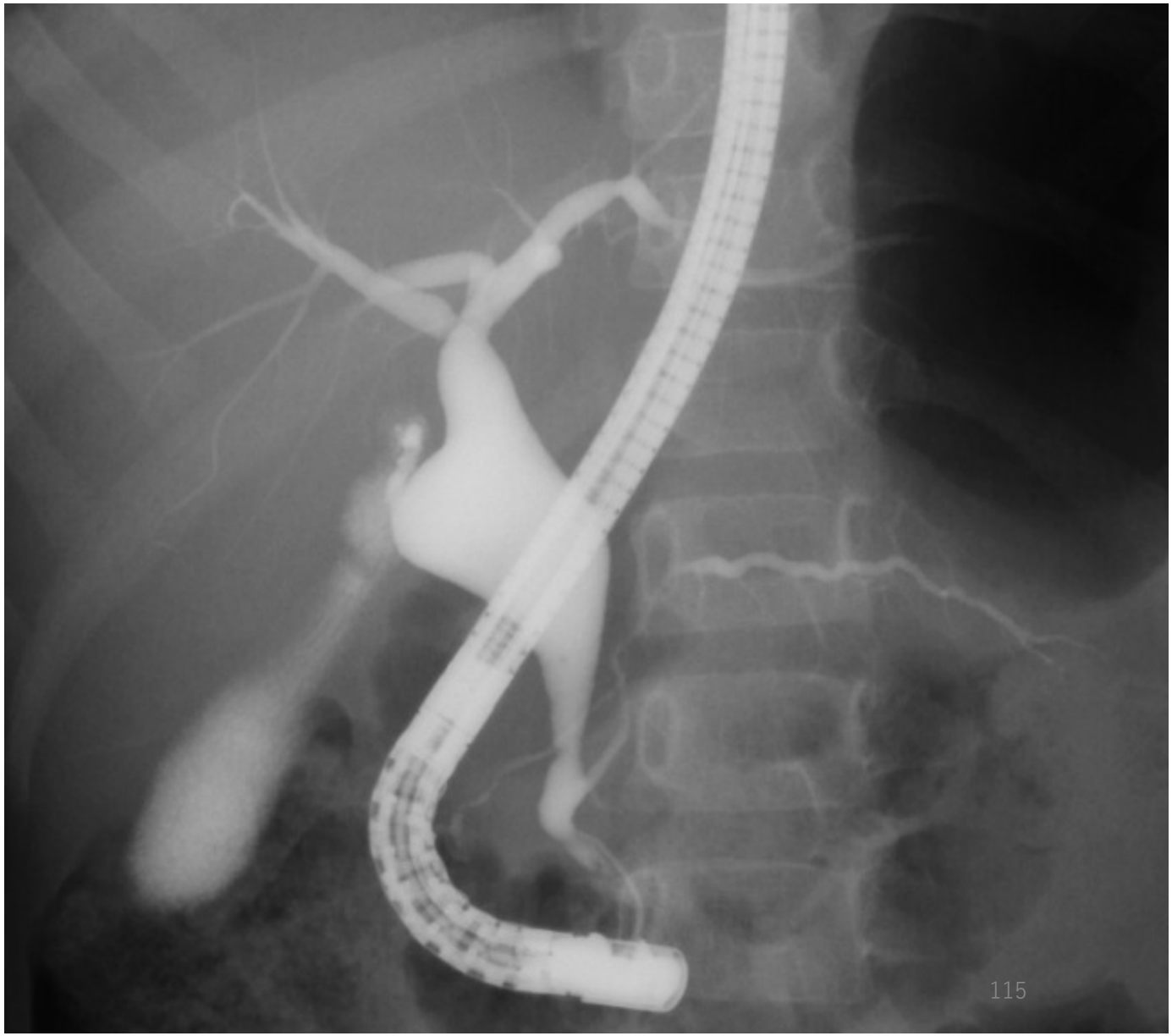


# Case 34 Ia < Ic

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

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



# Advanced

## Non-dilated type of PBM

## Definition

First version, September 2025

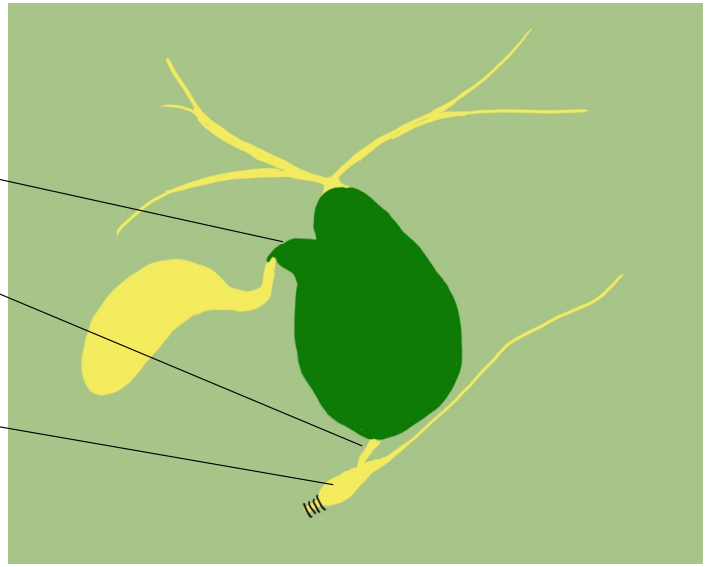
# Imaging features of congenital biliary dilatation

Features that define CBD — Introduction section

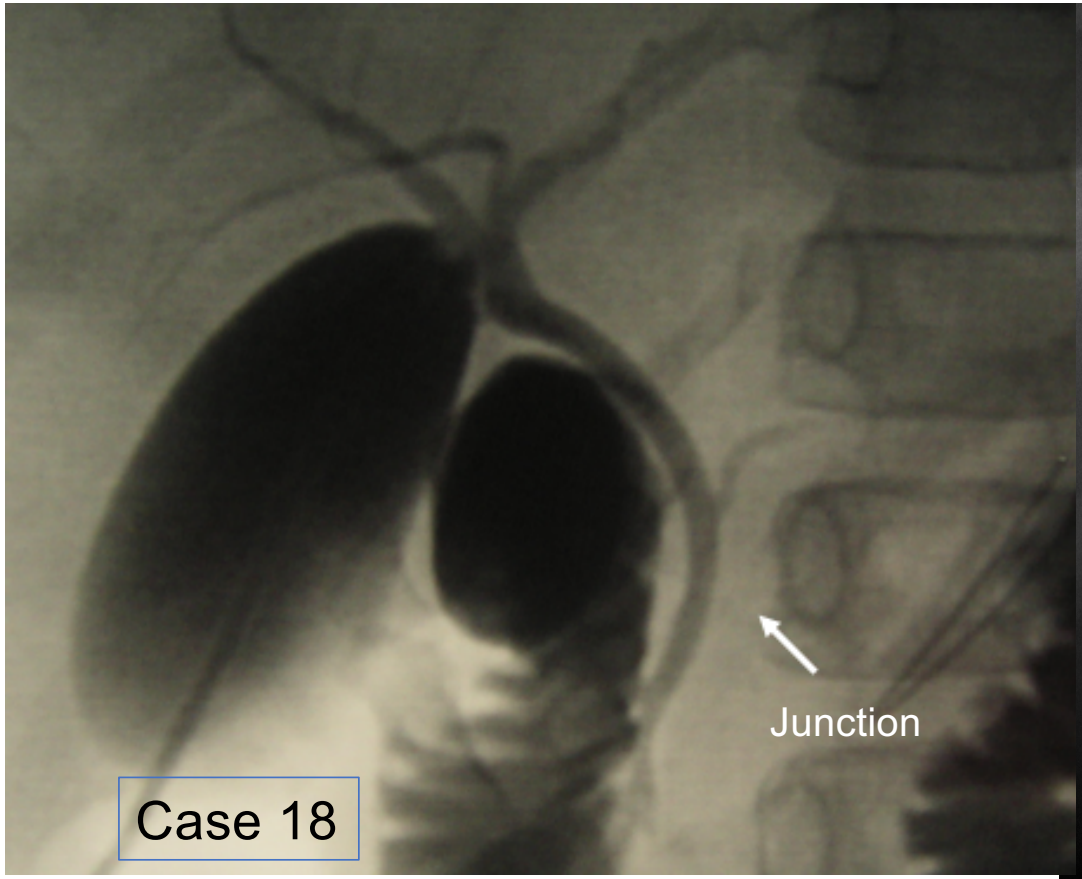
- Pancreaticobiliary maljunction
- Localized BD dilatation involving the common bile duct

Features (but not in all cases) — Basic section

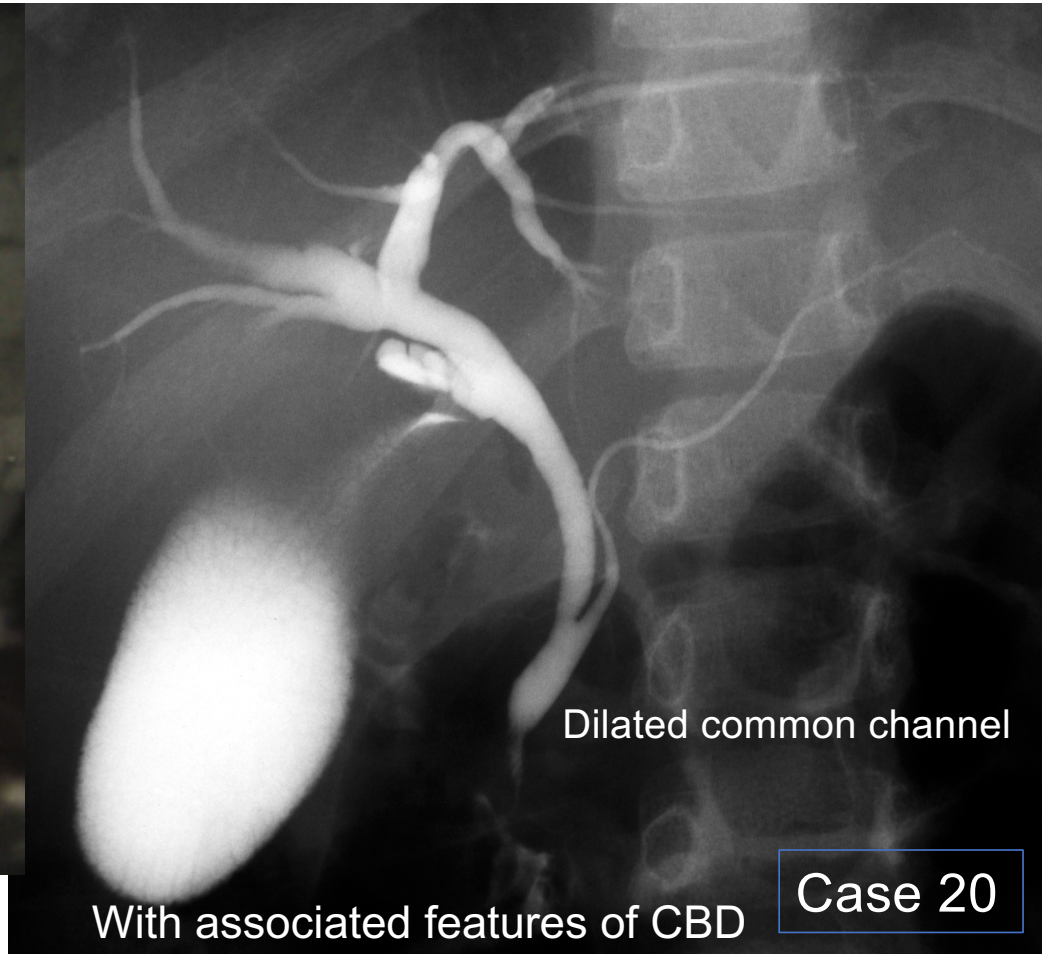
1. Dilatation at the base of the cystic duct
2. Narrow segment below the dilated BD
3. Dilated common channel



# PBM with a normal BD diameter in pediatric cases

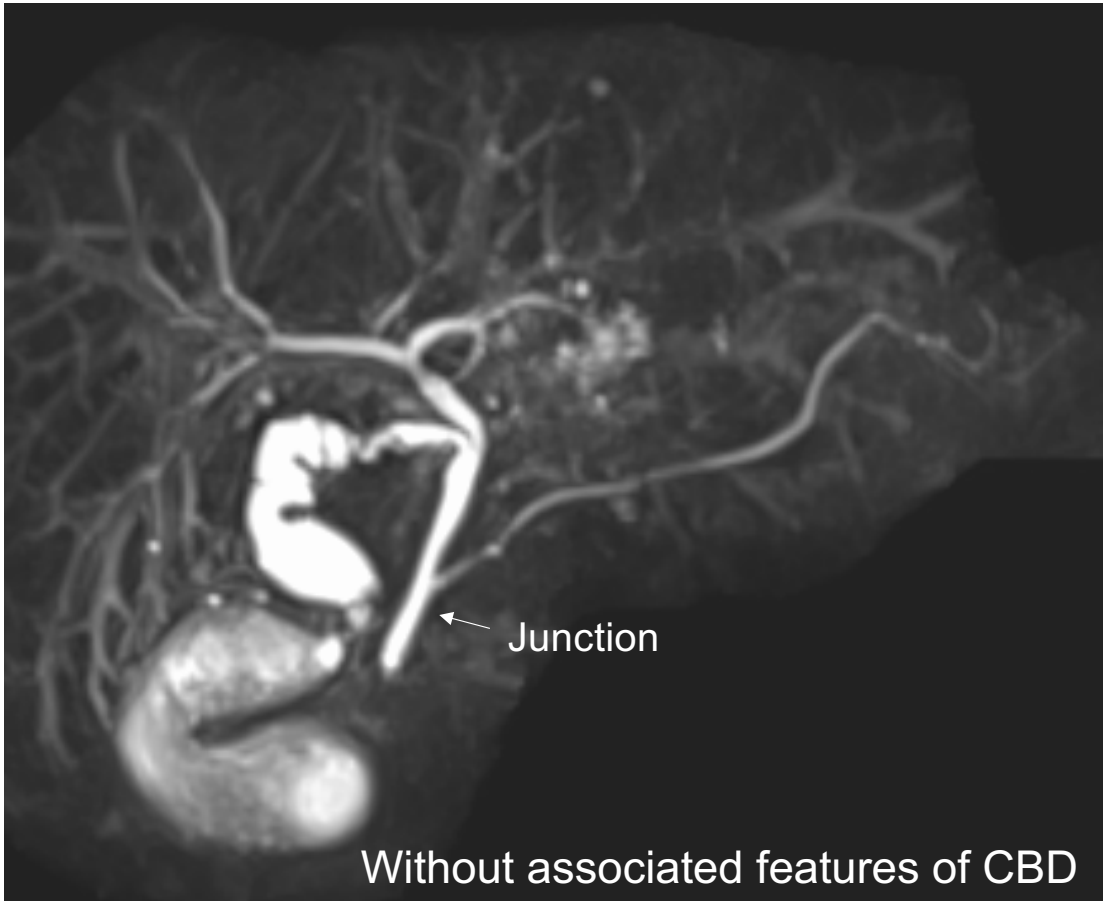



Without associated features of CBD




With associated features of CBD

# PBM with normal BD diameter in adult cases





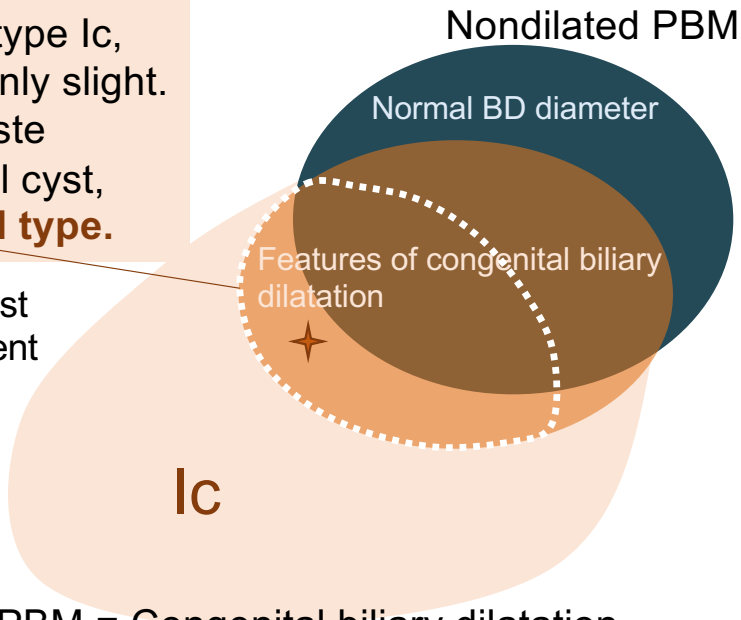

# Case 35 Forme fruste (Ic) Adult case

A female in her 80s.
Ultrasound during a medical checkup detected wall thickness at the fundus of the GB.

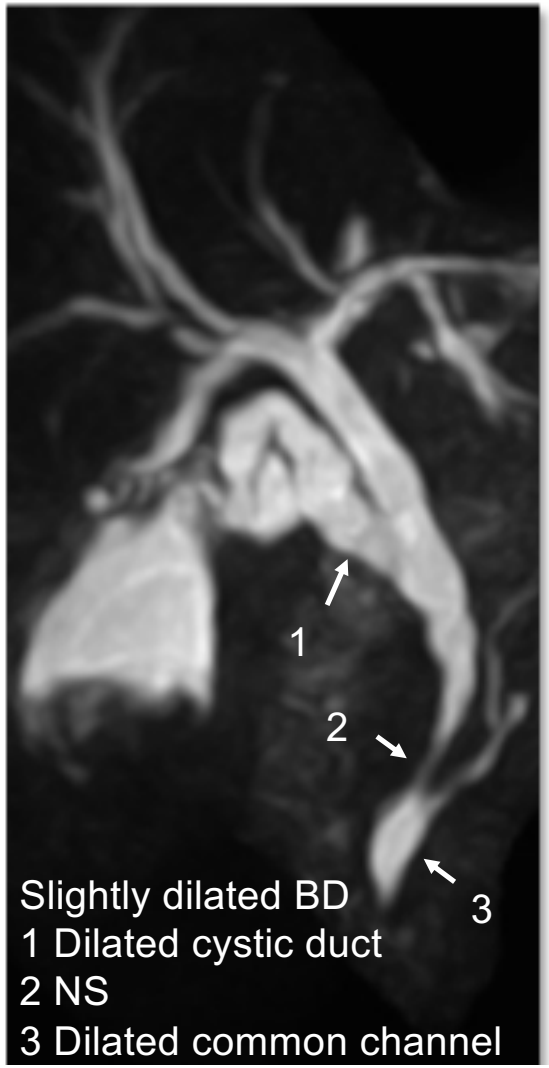
Adenosquamous carcinoma in the GB.

Strictly speaking, this is type Ic, but the BD dilatation is only slight. It represents a forme fruste (incomplete) choledochal cyst, serving as a **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.




Dilated PBM = Congenital biliary dilatation




Features of congenital biliary dilatation

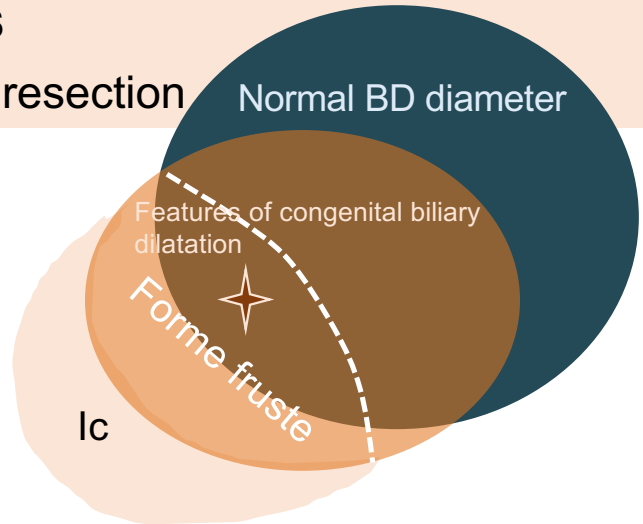
# Case 36 Pediatric case Normal BD diameter

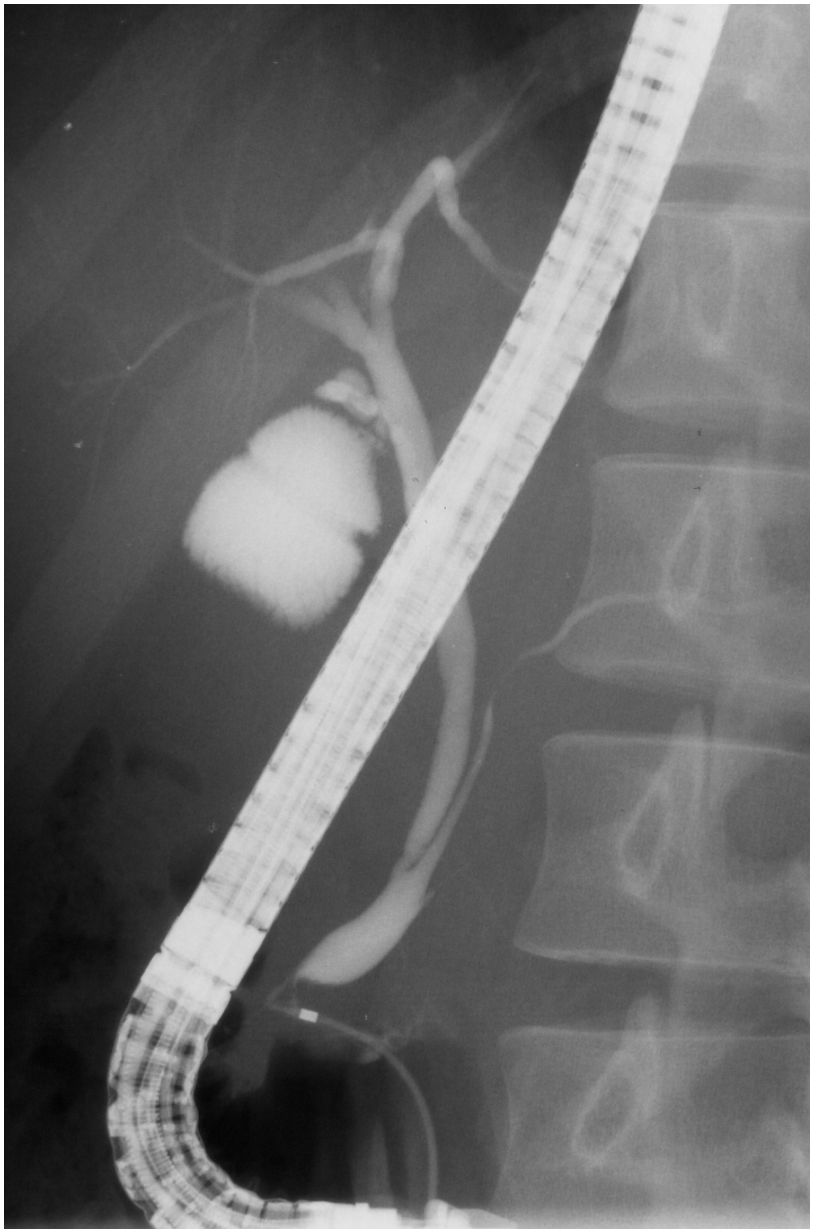
## Nondilated PBM < Forme fruste

A 15-year-old girl.
Ultrasound performed for abdominal pain detected gallbladder wall thickening.

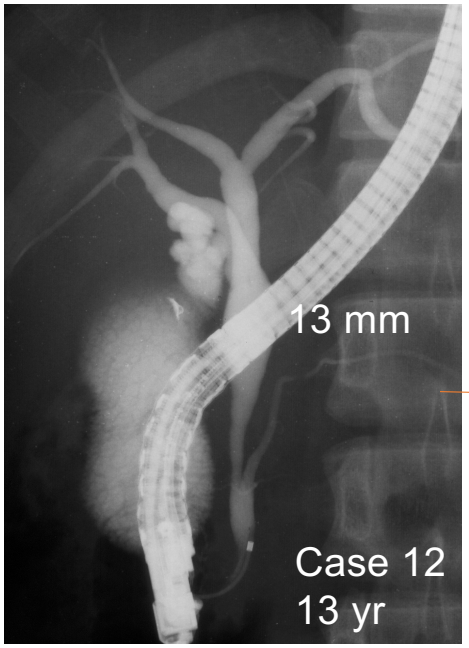
- Common bile duct diameter
  ERCP 6 mm (Ultrasound upper limit: 4.9 mm)
- **Features of congenital biliary dilatation**
  - ✓ **Dilated common channel**
  - ✓ **NS**

Bile duct resection

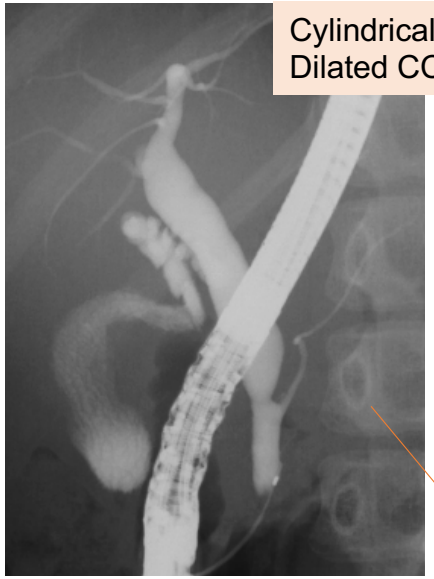


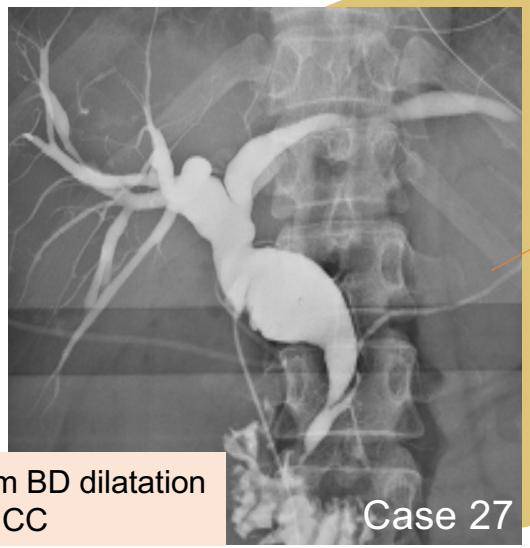
# Around forme fruste



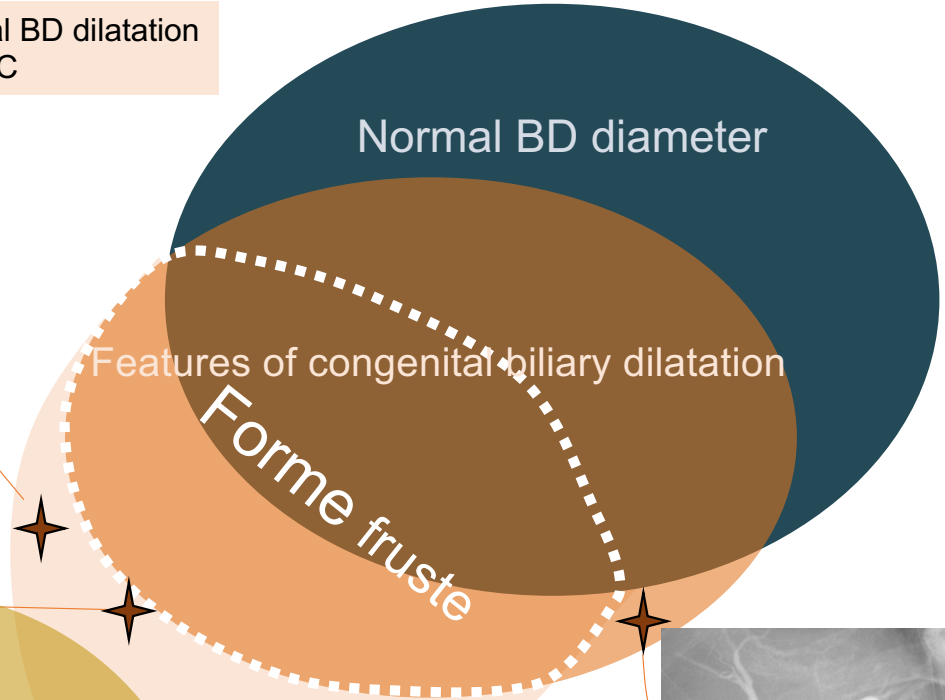

Fusiform BD dilatation
Dilated common channel (CC)
NS



Cylindrical BD dilatation
Dilated CC




Fusiform BD dilatation
Dilated CC



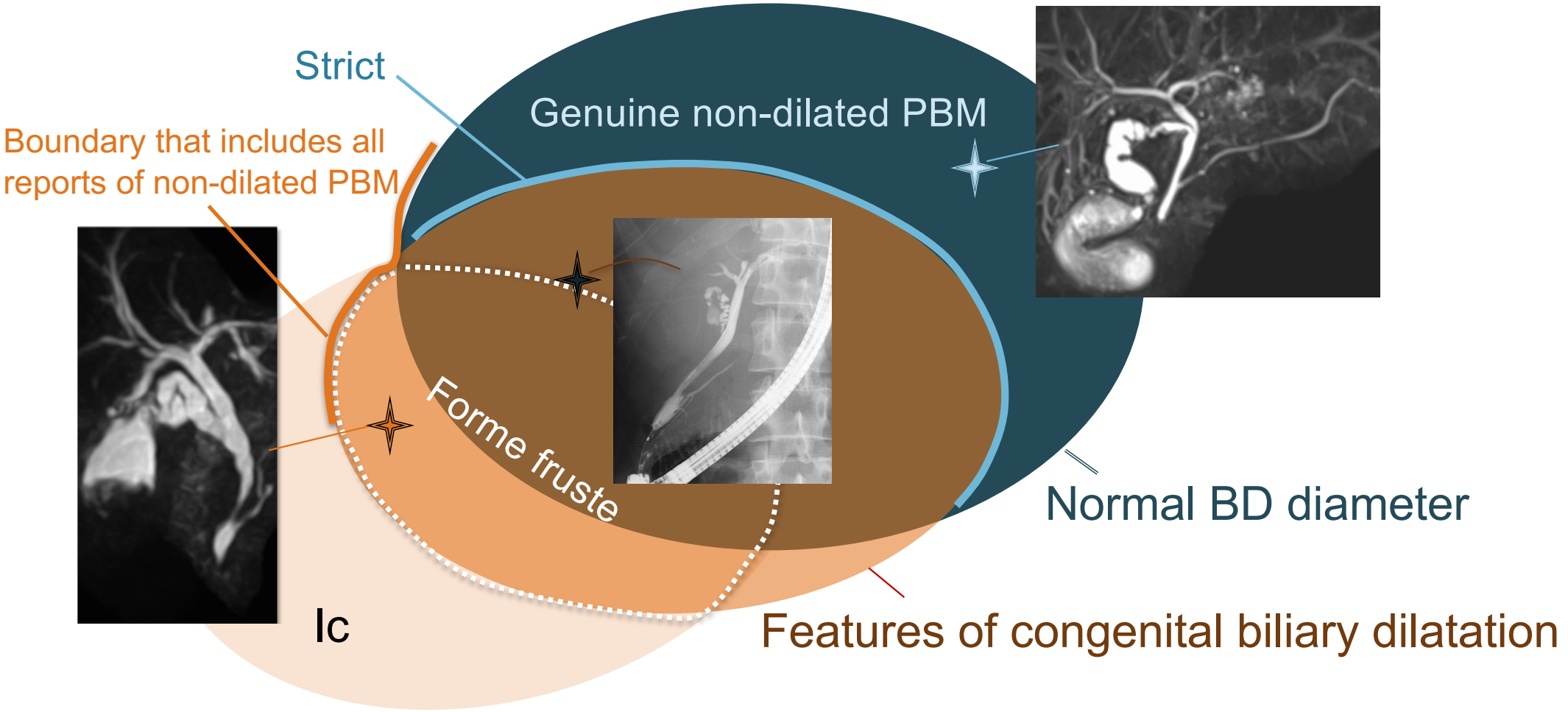

No bile stasis
except in the GB



Only slight biliary dilatation as a
feature of congenital biliary dilatation

# Definition of non-dilated PBM

Where to draw the boundary on this spectrum?

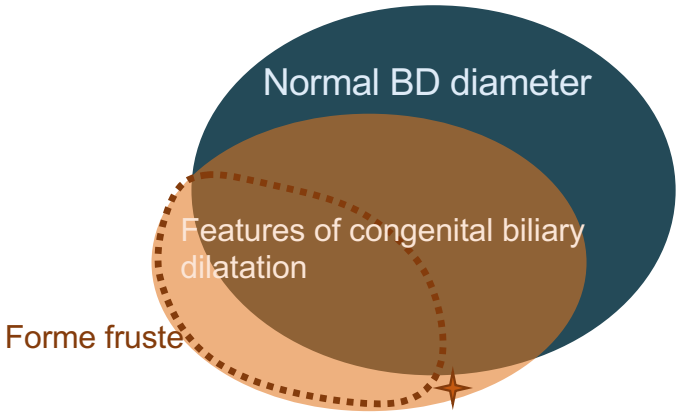


# Case 37

## Nondilated PBM > Forme fruste
## Pediatric case

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

- ERCP
  CBD diameter: 6 mm (US upper limit: 4.9 mm)
  **CHD diameter: 8 mm**
- BD was resected. Cholecystectomy alone might have been sufficient.




No bile stasis occurs except in the GB.

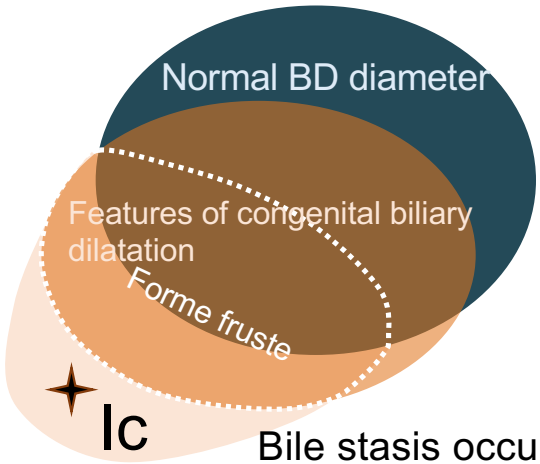
Three features of congenital biliary dilatation, but mild



From *Basic Section*

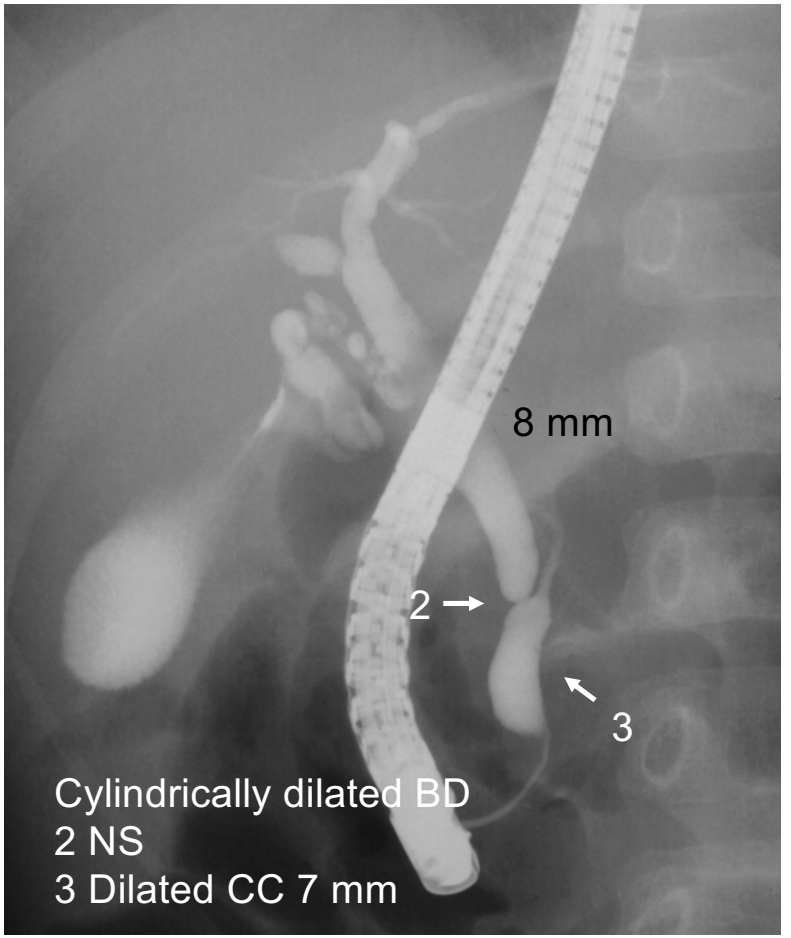
# Case 14 Type Ic: cylindrical dilatation Pediatric case

A 2-year-old boy.
Abdominal pain and vomiting.
AST: 896, ALT: 416, and amylase: 1168.
CT revealed a swollen pancreas.
BD was resected.




Bile stasis occurs in the dilated BD and CC, as well as GB

Three features of congenital biliary dilatation



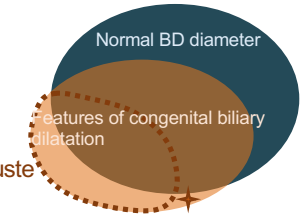

2 years old (US upper limit: 3.3 mm)

# Case 38

# Adult case

## Non-dilated PBM > Forme fruste >> Type Ic

Strictly speaking, the BD is dilated
However, no bile stasis occurs except in the GB.



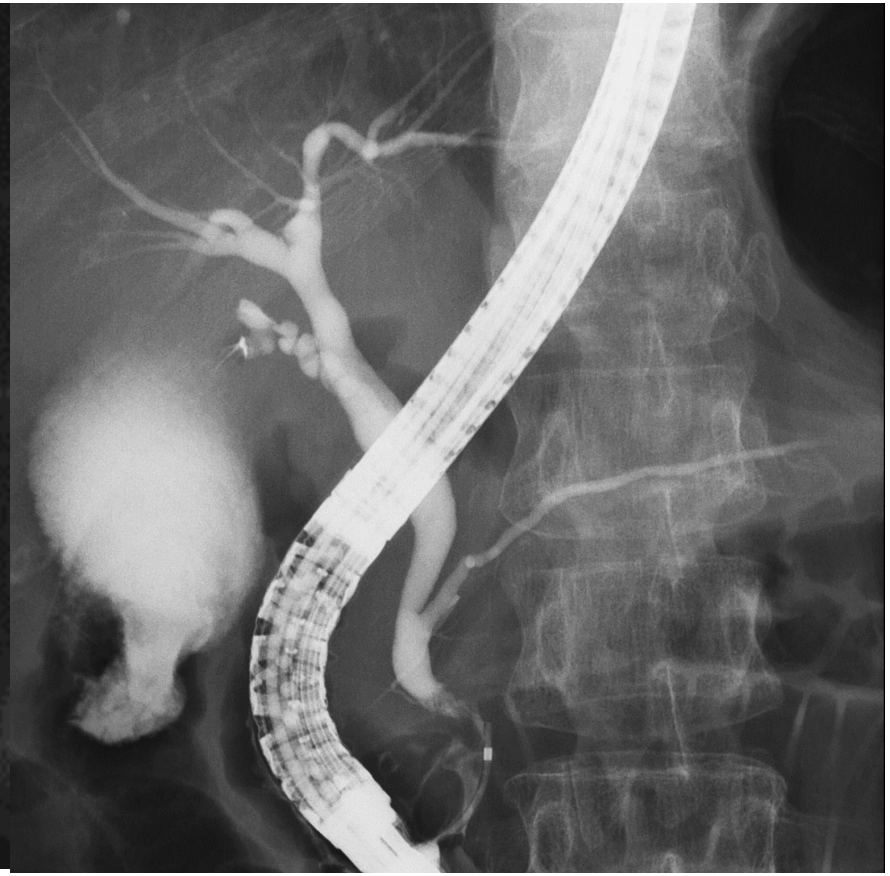

A female in her 50s.
US during medical checkup detected GB wall thickening.

BD diameter
**MRCP: 8 mm**
ERCP: 9 mm
(US upper limit: 7.2 mm)

Features of congenital biliary dilatation are all mild:
- ✓ **BD dilatation**
- ✓ **CC dilatation**
- ✓ **CD dilatation?**

**Cholecystectomy**



# Advanced

## Non-dilated type of PBM
## Pathophysiology and treatment in children

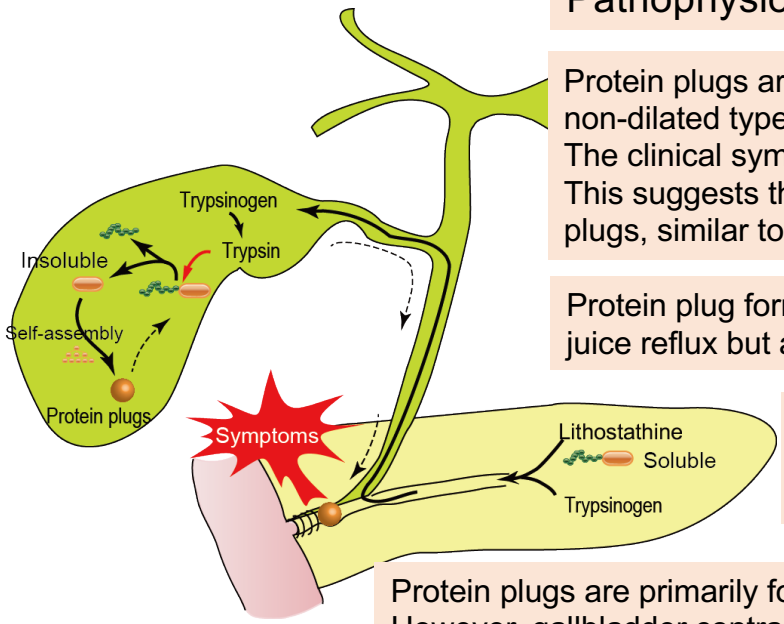
First version, September 2026

# Non-dilated PBM in children

## Current status

Patients present with the same symptoms as CBD: abdominal pain, vomiting, and jaundice.
Elevated serum liver enzymes and amylase are observed, similar to CBD.
Consequently, the same procedure (BD resection) is performed as for CBD (see *Basic Section*).

## Pathophysiological hypothesis




Protein plugs are frequently observed even in non-dilated types.
The clinical symptoms match those of CBD.
This suggests that onset is triggered by protein plugs, similar to CBD.

Protein plug formation requires not only pancreatic juice reflux but also bile stasis (see p181).

Factors contributing to stasis:
Gallbladder and
common channel dilatation

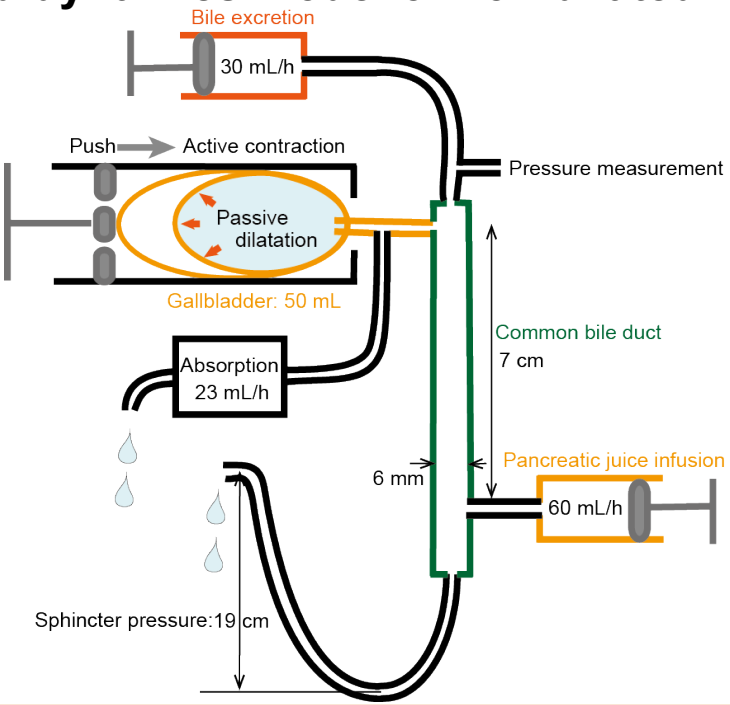
Protein plugs are primarily formed within the gallbladder. However, gallbladder contraction acts to inhibit this formation, which explains the lower overall incidence.



6-year-old, non-dilated PBM with protein plugs

# Cholecystectomy eliminates pancreatic juice reflux in the non-dilated type

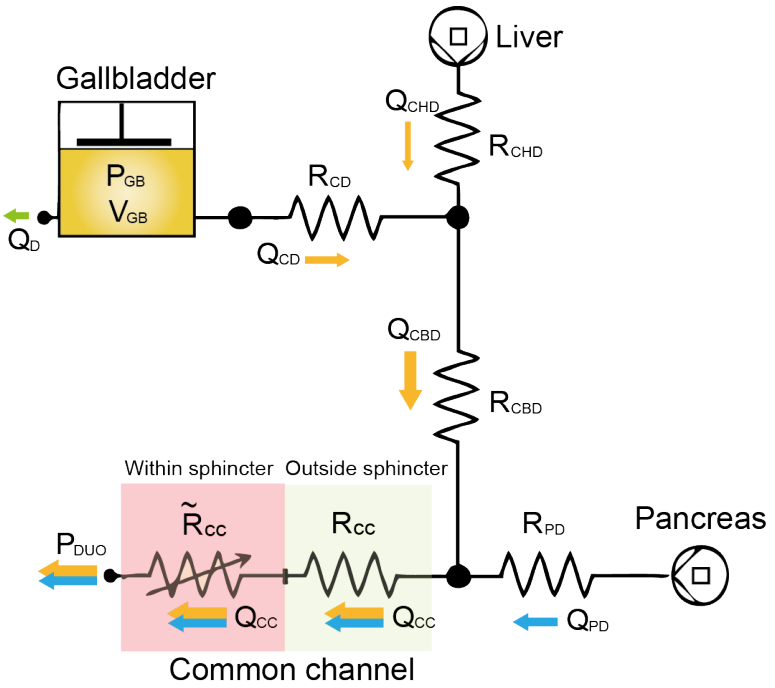
## Fluid dynamics model of non-dilated PBM




Pancreatic juice reflux occurs only during passive dilatation of the gallbladder.

Fukuzawa H, et al. *J Hepatobiliary Pancreat Sci.* 2020;27:265–272.

## Mathematical model of non-dilated PBM




Condition for pancreatic juice reflux: $\tilde{R}_{cc}Q_{PD} > P_{GB} - P_{DUO}$

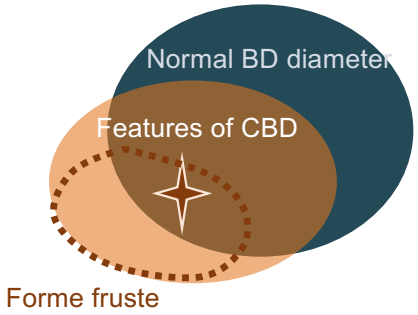
Tajikawa T, et al. *J Biorheol* 2023;37(2):44–55
*Biomed J Sci & Tech Res* 2024;55:47447–56

# Case 39

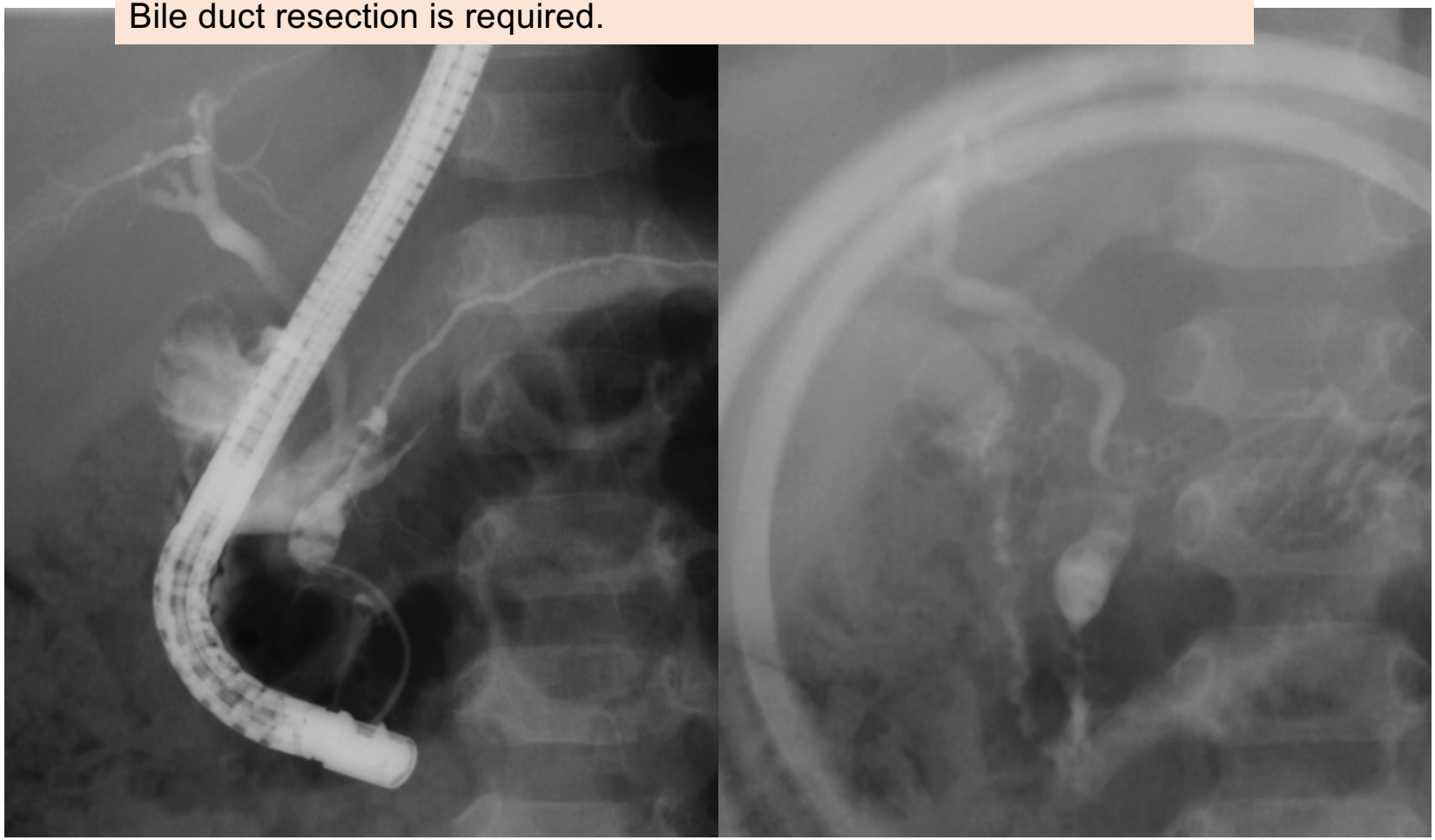
## PBM with a normal BD diameter and prominent common channel dilatation, Forme fruste > Non-dilated

A 3-year-old boy.
Abdominal pain and vomiting.
AST: 834, ALT: 443, and amylase: 682 U/L.

ERCP
CBD diameter: 3.8 mm
(US upper limit: 3.5 mm)
Common channel diameter: 22 mm




Bile stasis occurs not only in the GB but also in the common channel.
Bile duct resection is required.



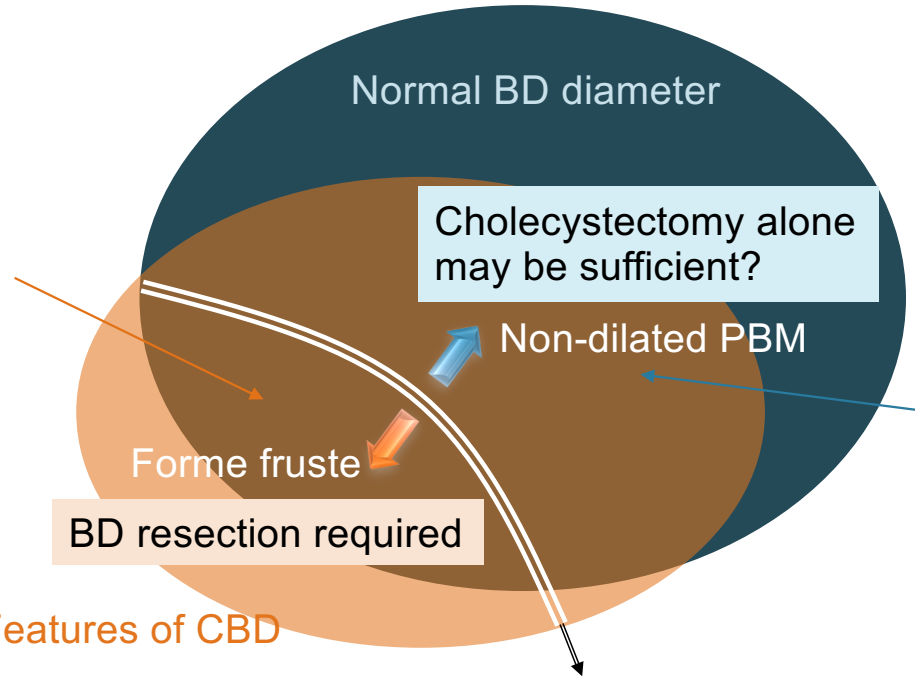
# Therapeutic considerations in children

## Varying features of common channel dilatation




Normal BD diameter but prominent common channel dilatation

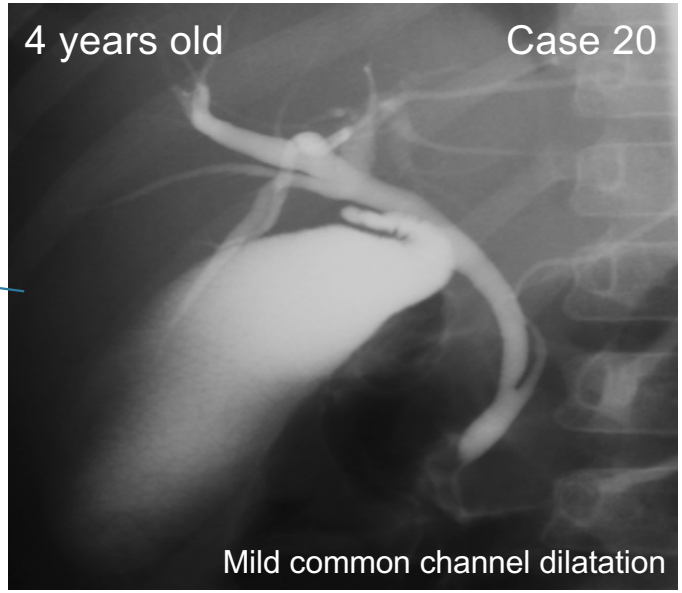
Bile stasis in both the GB and common channel




Reflecting differences in pathophysiology and treatment

**Proposed definition line for non-dilated PBM**

Bile stasis limited to the GB




Although BD resection was performed in this case, cholecystectomy alone might have been sufficient.

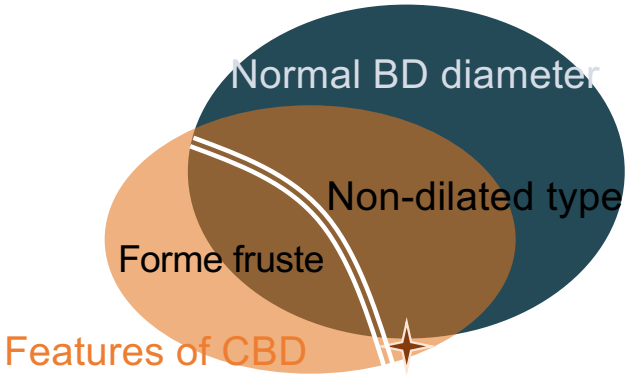
# Case 40

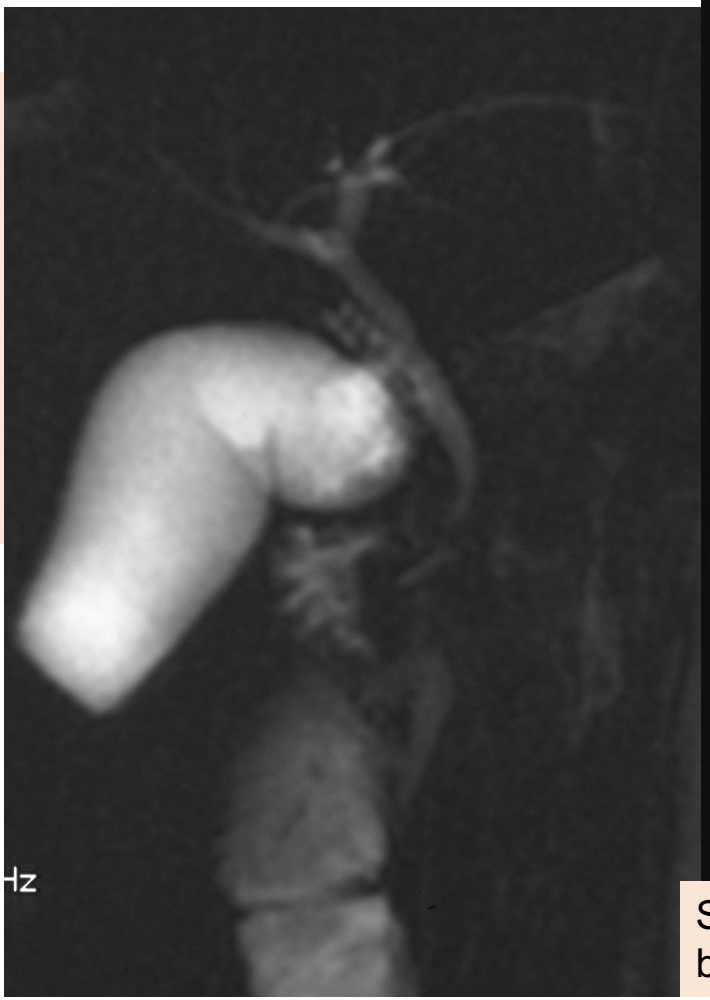
## Non-dilated type > Forme fruste
## Pediatric case

A 13-year-old girl
Abdominal pain, diarrhea, and elevated serum amylase

- Common bile duct diameter
  **MRCP: 5.5 mm**
  ERCP: 5.9:mm (US upper limit 4.9 mm)
- **Features of CBD**
  - ✓ **NS?**

Although the bile duct was resected, cholecystectomy alone might have been sufficient in this case.

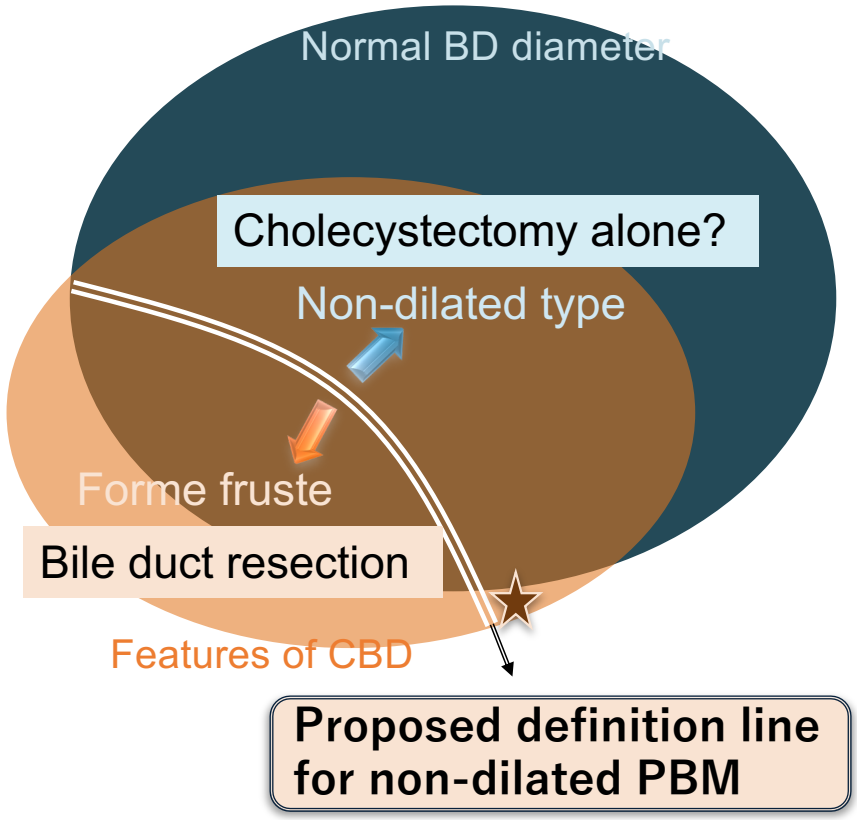




Strictly speaking, the BD is dilated,
but no bile stasis occurs except in the GB.

# Slight BD dilatation and mild features of CBD



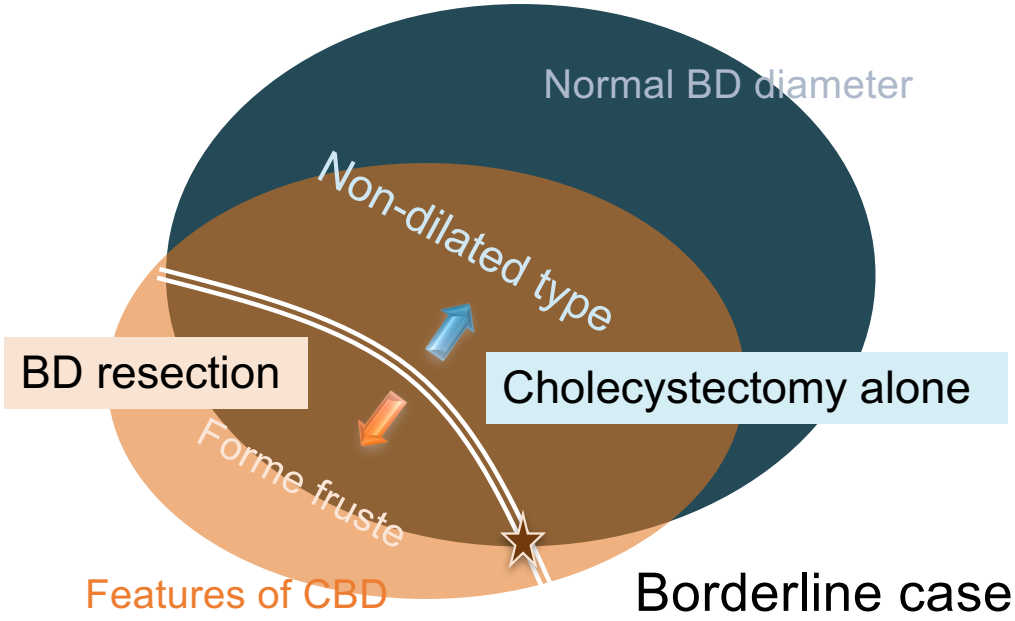

The definition and treatment may depend on the existence of bile stasis except in the GB.






# case 41 Normal common bile duct diameter in a child

An 11-year-old girl.
Ultrasound showed a common bile duct diameter of 4.5 mm (Upper limit: 4.6 mm).
Intraoperative cholangiography showed a slightly dilated common hepatic duct and common channel.




Borderline case



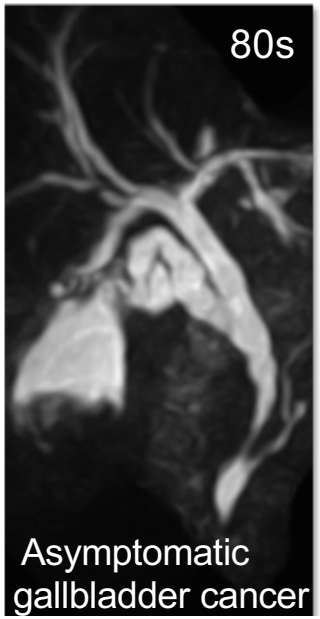
# Advanced

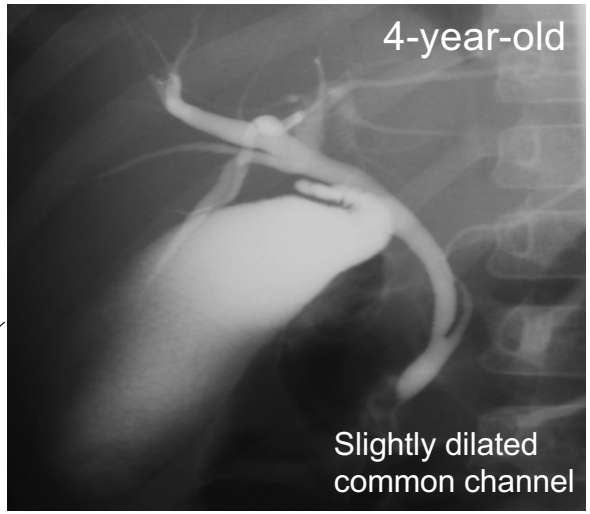
## Non-dilated type of PBM

## Pathophysiology and treatment in adults

First version, September 2026

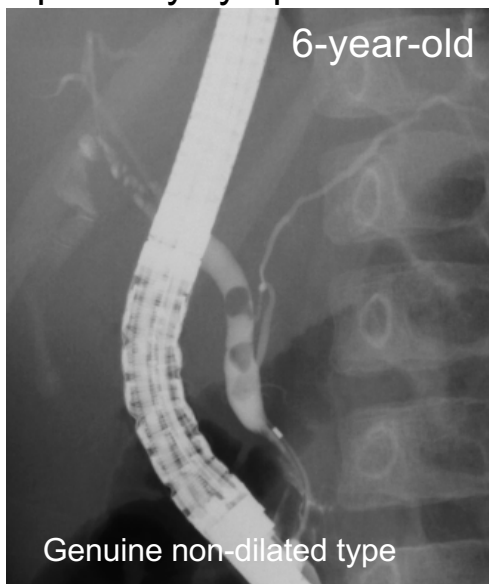
# Non-dilated type
# Why are so many cases asymptomatic?

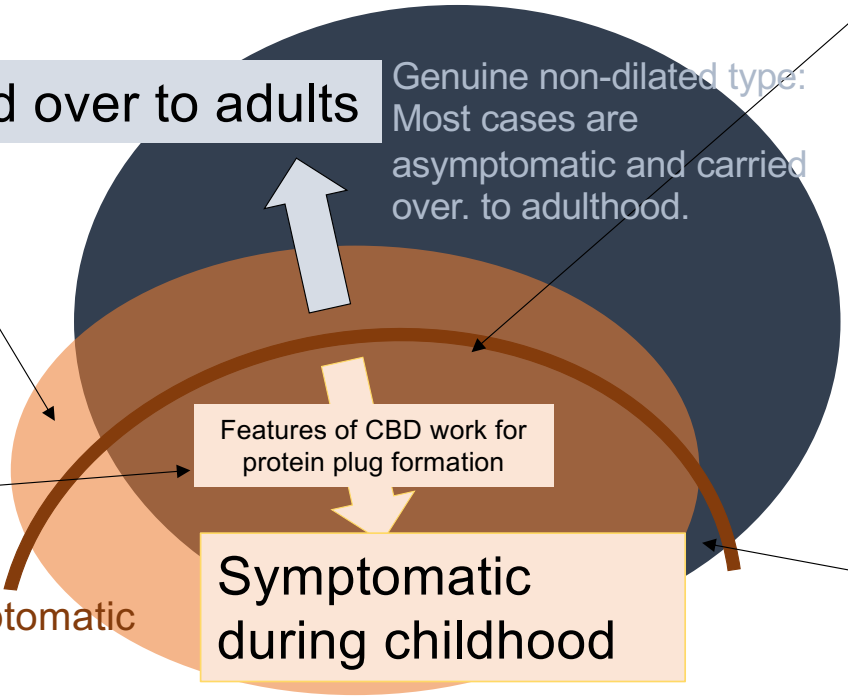



Exceptionally symptomatic cases



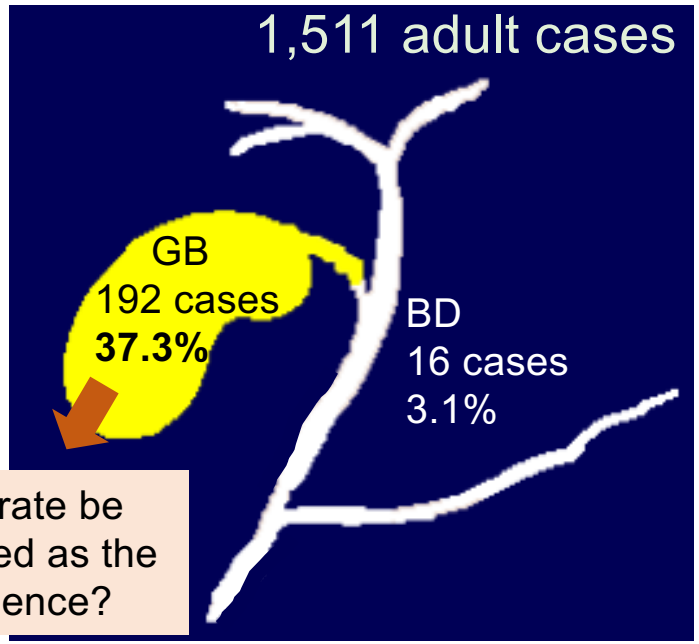



# Is the incidence of gallbladder cancer in non-dilated type really high?

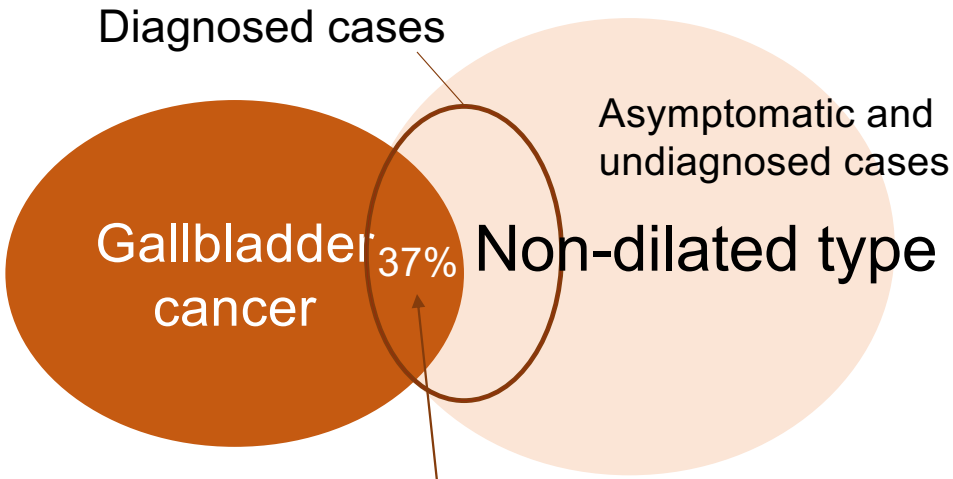
From *Introduction Section*

1990–2007 Nationwide survey




Can this rate be considered as the true incidence?

Biliary cancer rates in the non-dilated type:
42.4% 218/514 cases




This figure must be the rate of gallbladder cancer only among cases diagnosed with non-dilated PBM.

# Estimation of the prevalence of non-dilated PBM from the rate of PBM among the patients undergoing cholecystectomy for gallstone using preoperative MRCP

| Cholecystectomy for benign diseases | | PBM |
|---|---|---|
| Gallstone/cholecystitis | 2,002 cases | 7 cases (0.35%) |
| Polyp/adenomyomatosis | 44 cases | 2cases (4.5%) |

Shirai S, Kaneko K, Kato S, Kondo R, Osawa T, Fukami Y, Sano T. Prevalence of Pancreaticobiliary Maljunction and High Confluence of the Pancreaticobiliary Ducts: True Incidence of Gallbladder Cancer. *J Hepatobiliary Pancreat Sci*. 2025 Sep;32(9):649-655. doi: 10.1002/jhbp.12187. Epub 2025 Jul 7. PMID: 40624738; PMCID: PMC12466845.

This significantly high rate supports its known relation with PBM ($P < 0.01$).

Little evidence supports that pancreatobiliary reflux induces gallstone formation.

Gallstone/cholecystitis and PBM can be assumed to be unrelated.

The figure of **0.35%** is estimated as the **prevalence of non-dilated PBM** in the Japanese population.

# cf. Prevalence of dilated PBM in adults

**Ultrasound in health checkups**

- GB wall thickening ≧ 3 mm
- BD dilatation ≧ 10 mm

➡ EUS ➡ ERCP

27,076 individuals ➡ PBM 9 cases

- Dilated type 4 (0.015%)
- Non-dilated type 5 (0.018%)

The dilated type (congenital biliary dilatation) is assumed not to be overlooked by US screening.

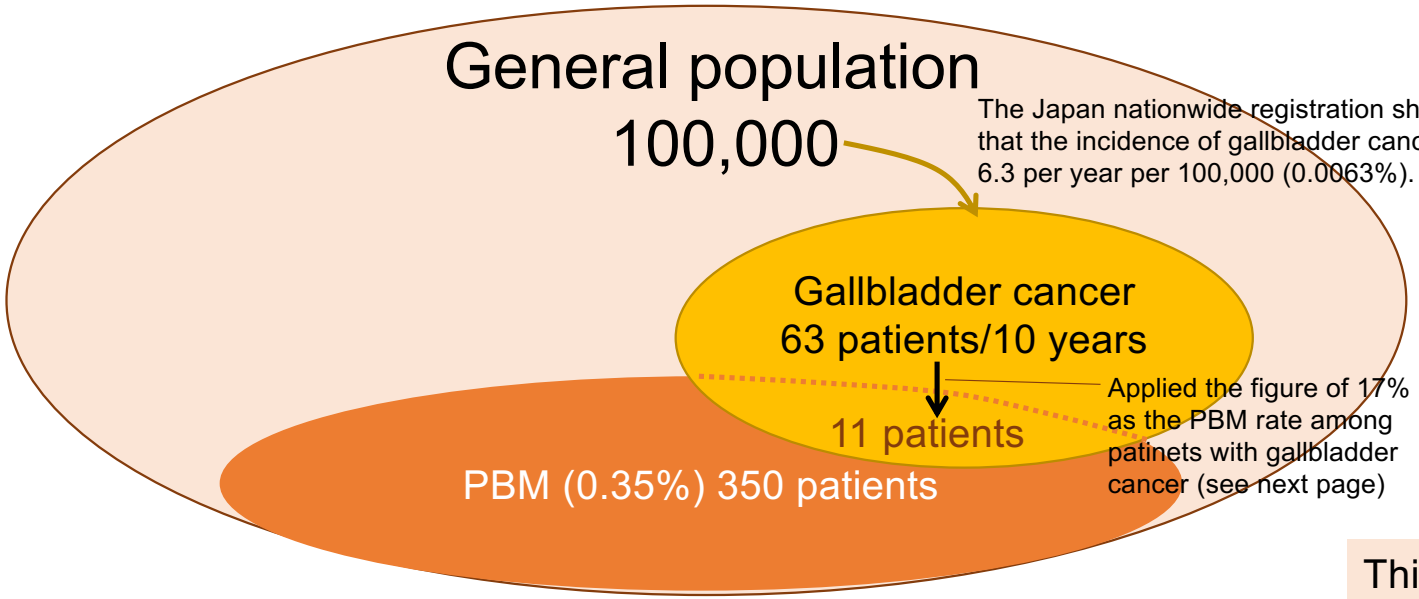
$$\frac{1}{6{,}700}$$

Yamao K, et al.
Prospective study of the detection of anomalous connections of pancreatobiliary ducts during routine medical examinations.
*Hepatogastroenterology 1996; 43: 1238–1245.*

# Estimation of the incidence of gallbladder cancer

The figure of 0.35% is assumed to be the prevalence of non-dilated PBM.




The Japan nationwide registration shows that the incidence of gallbladder cancer is 6.3 per year per 100,000 (0.0063%).

Applied the figure of 17% as the PBM rate among patinets with gallbladder cancer (see next page)

Incidence of gallbladder cancer among patients with non-dilated PBM

$$\frac{11}{350} = 3.1\% \text{ / } 10 \text{ years}$$

This is 1/10 of the gallbladder cancer rate reported in the nationwide survey

This is 52 times higher than that in the general population (0.063% / 10years)

# Literature survey
# Rate of gallbladder cancer among patients with PBM

Diagnostic criteria：
common channel length ≥15mm

Diagnostic criteria：
outside the sphincter

Similar data in the Western

| Author | Country, city | Year | PBM (n) | Gallbladder cancer (n) | Rate (%) | Source |
|---|---|---|---|---|---|---|
| Kato O | Japan, Nagoya | 1983 | 4 | 8 | 50.0 | Gastrointest Endosc 29:94–8 |
| Kinoshita H | Japan, Osaka | 1984 | 10 | 48 | 20.8 | Cancer 54:762–9 |
| Kimura K | Japan, Chiba | 1985 | 16 | 96 | 16.7 | Gastroenterology 89:1258–65 |
| Misra SP | India, New Delhi | 1989 | 2 | 21 | 9.5 | Gastroenterology 96:907–12 |
| 内村 | Japan, Hamamatsu | 1991 | 19 | 106 | 17.9 | 胆と膵12:397–404 |
| 須田 | Japan, Yamanashi | 1991 | 18 | 41 | 43.9 | 胆道 5:513–6 |
| Chijiiwa K | Japan, Fukuoka | 1993 | 4 | 37 | 10.8 | Am Surg 59:430–4 |
| Chao TC | **Taiwan**, Taipei | 1995 | 4 | 25 | 16.0 | J Clin Gastroenterol 21:306–8 |
| Sandoh N | Japan, Niigata | 1997 | 10 | 58 | 17.2 | Hepatogastroenterology 44:1580–3 |
| Egami K | Japan, Tama | 1998 | 14 | 43 | 32.6 | Nihon Ika Daigaku Zasshi 65:7–13 |
| Yoshida T | Japan, Ooita | 1999 | 9 | 59 | 15.3 | J Am Coll Surg 189:57–62 |
| Hasumi A | Japan 133 institutes | 2000 | 80 | 769 | 10.4 | J Hepatobiliary Pancreat Surg 7:551–5 |
| Elnemr A | Japan, Kanazawa | 2001 | 23 | 126 | 18.3 | Hepatogastroenterology 48:382–6 |
| Hu B | **China**, Shanghai | 2003 | 7 | 54 | 13.0 | Gastrointest Endosc 57:541–5 |
| Kamisawa T | Japan, Tokyo | 2006 | 36 | 268 | 13.4 | Hepatogastroenterology 53:816–8 |
| Kang CM | **Korea**, Seoul, Yonsei Univ. | 2007 | 10 | 218 | 4.6 | Can J Gastroenterol 21:383–7 |
| Kim Y | **Korea**, Ansan | 2014 | 10 | 197 | 5.1 | Langenbecks Arch Surg 399:1071–6 |
| Park JS | **Korea**, Seoul, Asan Medical Center | 2016 | 76 | 1,111 | 6.8 | Medicine 95:e3526 |
| Chang J | **Korea**, Seoul, Seoul National University | 2016 | 69 | 401 | 17.2 | World J Surg 40:1211–7 |
| Fujimoto T | Japan, Fukuoka | 2017 | 7 | 22 | 31.8 | J Hepatobiliary Pancreat Sci 24:103–8 |
| Hyvärinen I | **Finland**, Helsinki | 2019 | 3 | 18 | 16.7 | Scand J Surg 108:285–90 |
| Kawakami S | Japan, Yamanashi | 2021 | 8 | 34 | 23.5 | BMC Cancer 21:1245 |
| Muraki T | **USA** | 2022 | 24 | 300 | 8 | Ann Surg 2022;276:e32-e39 |
| Muraki T | Japan, Nagano | 2022 | 15 | 171 | 8.8 | Ann Surg 2022;276:e32-e39 |
| Shirai S | Japan, Nagakute | 2025 | 8 | 49 | 16.3 | J Hepatobiliary Pancreat Sci 32:649-55 |
| | | | | **Median rate** | 16.3 | |
| | | | | **Mean rate** | 17.8 | |
| | | Total | 486 | 4280 | 11.4 | |

# Assuming a 3% carcinogenesis rate per 10 years:

- Lifetime risk of **gallbladder** cancer (from age 20s to 80s)

$$1 - (0.97)^6 = 0.167$$ 17%

Simple calculation (3% × 6 = 18%) suggests nearly 20% will develop gallbladder cancer.

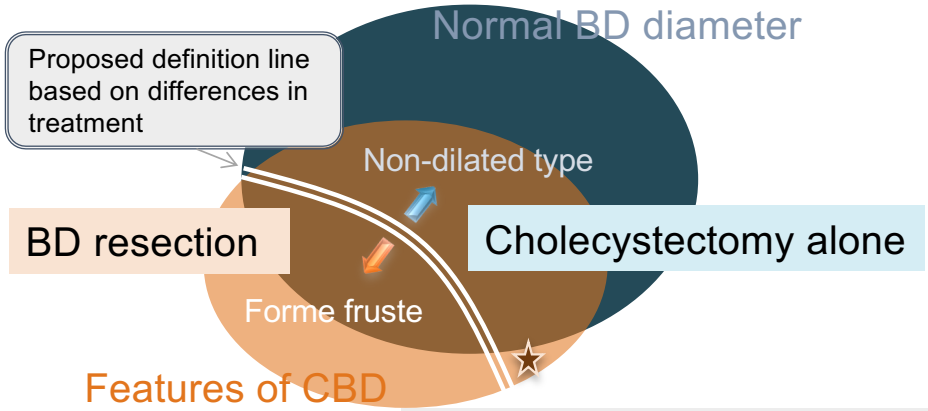
- Non-dilated **bile duct cancer** incidence is only ~1/10 of gallbladder cancer. ~ 2%

- Considering postoperative complications:
  Cholecystectomy: Strongly recommended.
  Bile duct resection: Highly debatable.

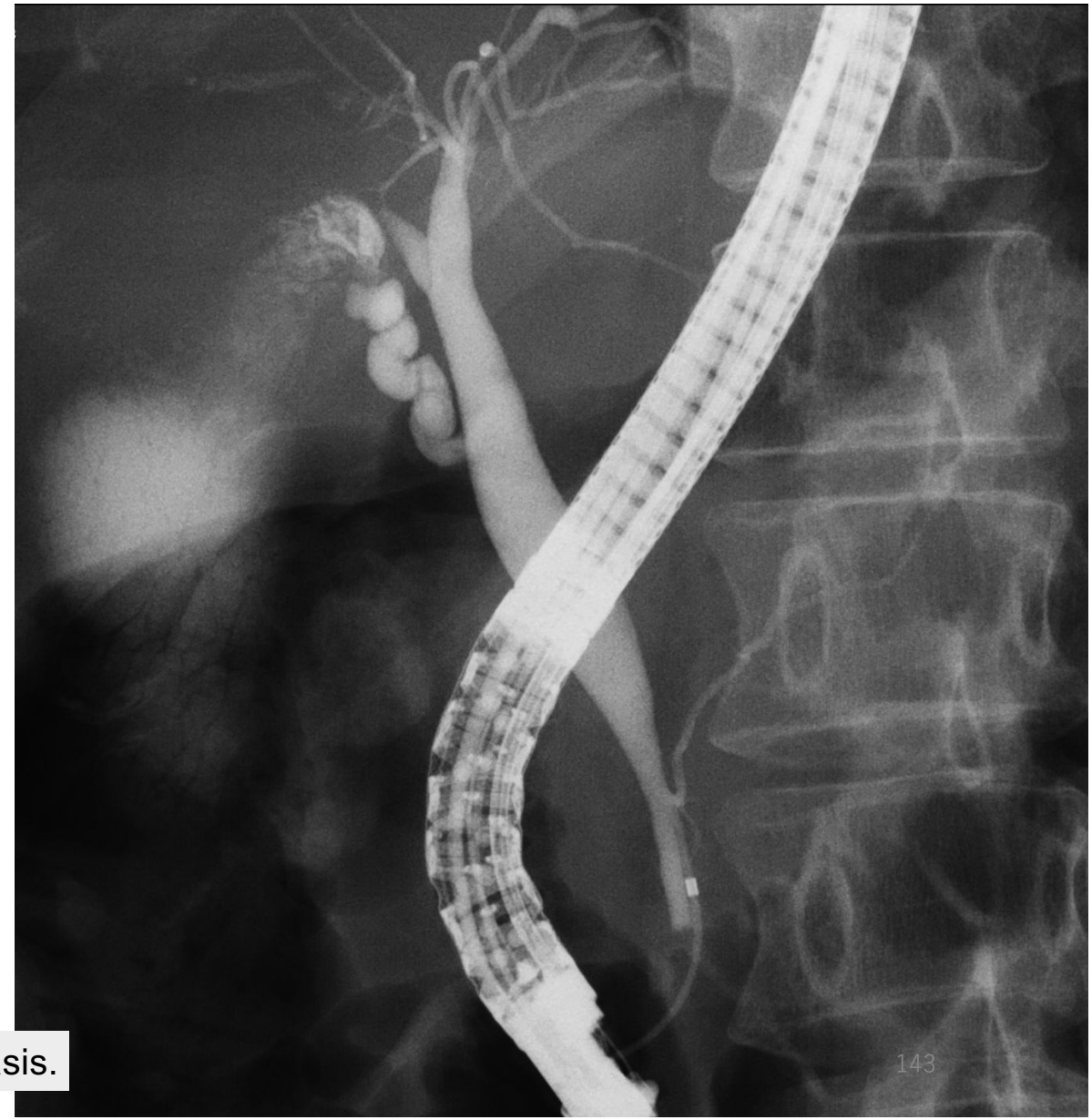
# Case 42 Non-dilated type > Forme fruste (Ic)

ERCP in a female in her 40s
She had no symptoms.
Ultrasonography during a medical checkup revealed gallbladder wall thickening.

- PBM classification: type B (non-stenotic type)
- The BD dilatation is only slight, causing no bile stasis.
  No other features of CBD.
  Bile stasis occurs only in the gallbladder.
- Cholecystectomy alone is sufficient for treatment.



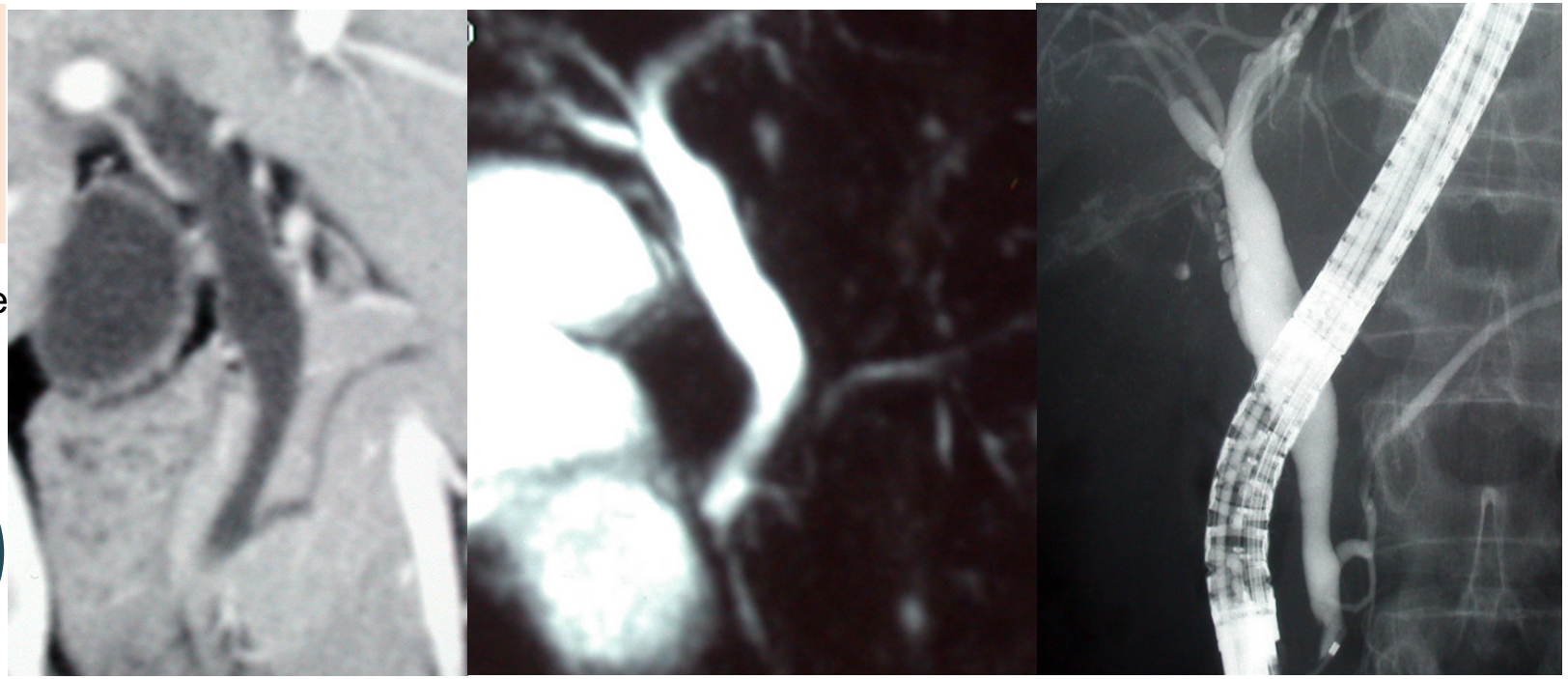

Carcinogenesis needs bile stasis.



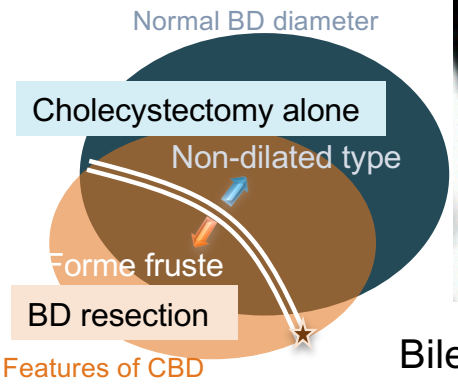
# Case 43

## Borderline case
## Fusiform dilatation (Ic) ≤ forme fruste ≤ non-dilated type

A female in her 40s
- Slight BD dilatation.
- Slight common channel dilatation.
- PBM classification: type B (non-stenotic type).



This case is closer to the boundary than Case 42.




Bile stasis does not occur except in the gallbladder.
By conventional criteria, this case corresponds to type Ic dilated PBM,
but BD resection would be oversurgery when weighing the risk of postoperative complications.