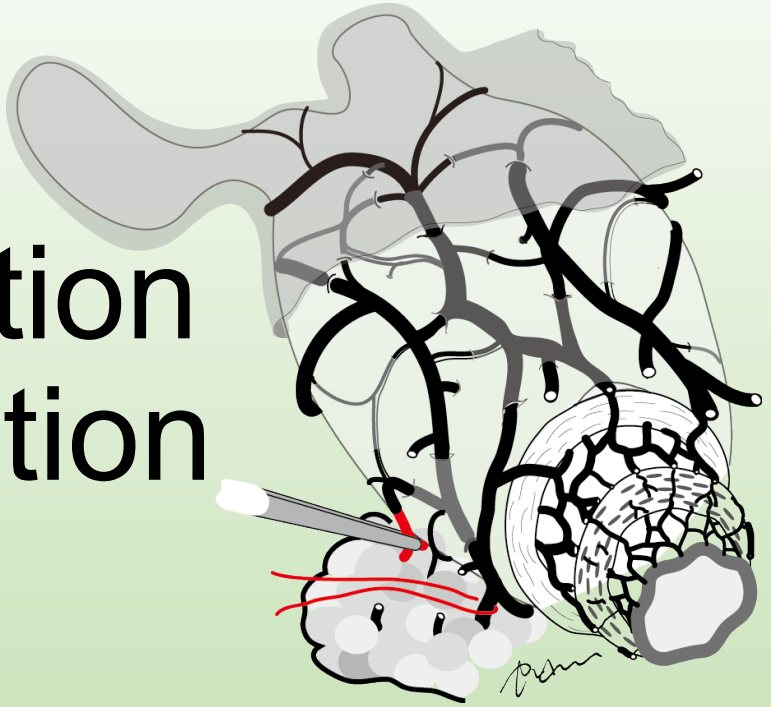
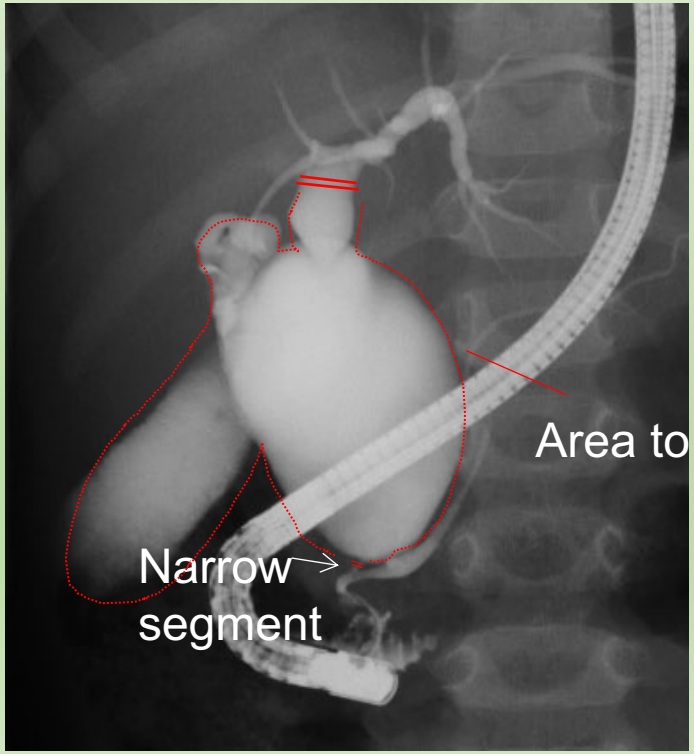
Atlas of Pancreaticobiliary Maljunction & Congenital Biliary Dilatation





Japanese Study Group on Pancreaticobiliary Maljunction (JSGPBM)
Scientific Committee

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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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PBM, pancreaticobiliary maljunction
CBD, congenital biliary dilatation
BD, bile duct

Advanced

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Introduction

First version, September 2022; Revised, September 2026

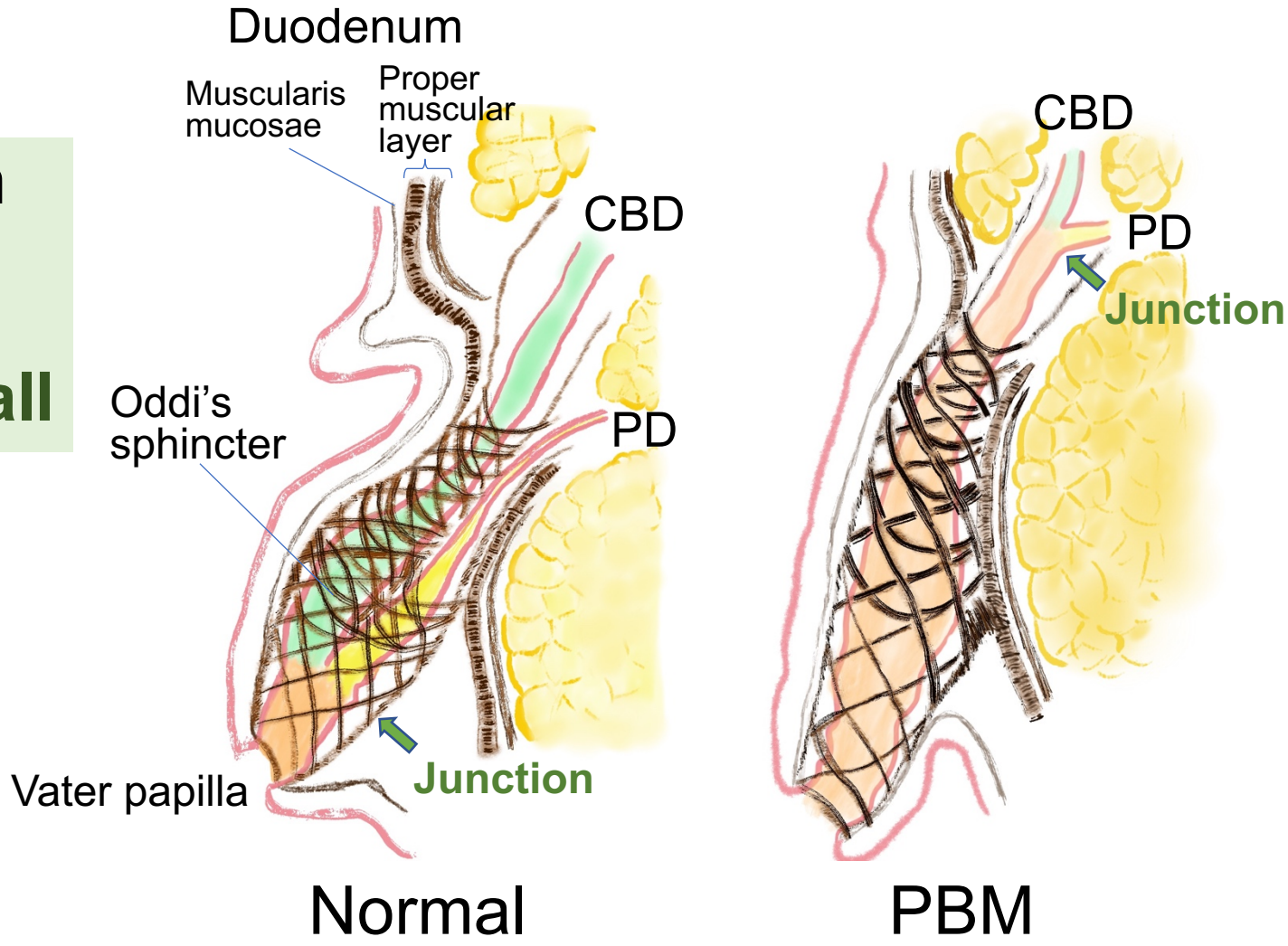
What is Pancreaticobiliary Maljunction (PBM)?

Definition

A congenital malformation in which the **pancreatic duct** and **bile duct join outside the duodenal wall**

The junction outside of the sphincter action is a functional phenomenon of PBM but is not the definition of it. However, when the confluence outside the duodenal wall cannot be shown, identification of the junction outside the sphincter action is enough for a diagnosis of PBM.

See *Basic Section*.

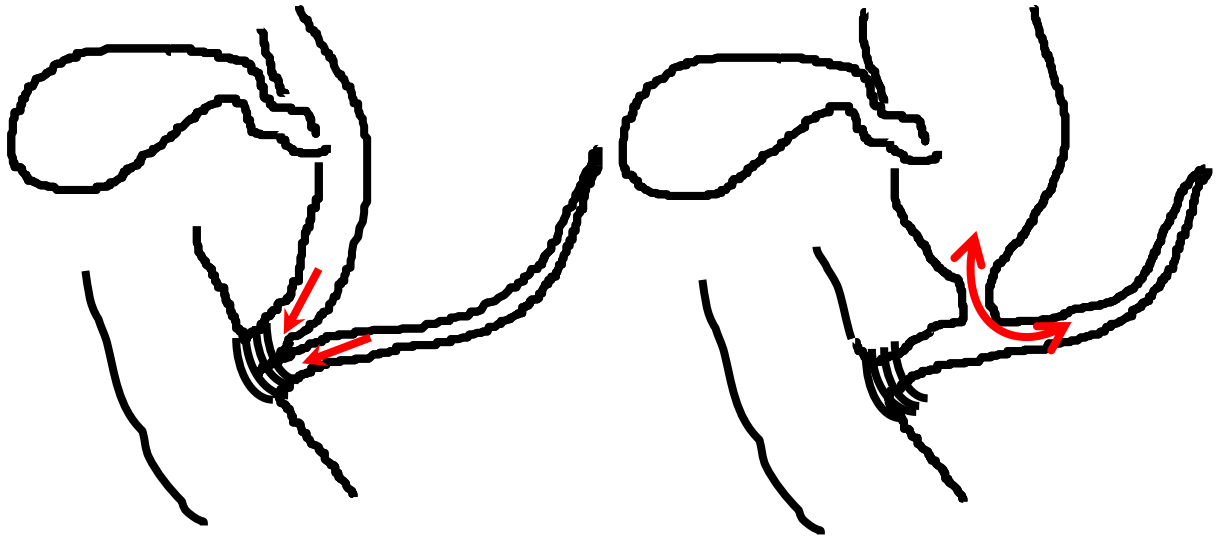


What is Pancreaticobiliary Maljunction (PBM)?

Pathophysiology

In PBM, the sphincter action does not regulate the pancreaticobiliary junction and **two-way regurgitation** of pancreatic juice and bile occurs, resulting in various pathological conditions of the biliary tract and pancreas.

During **childhood**, intermittent **symptoms** including abdominal pain, vomiting, and jaundice occur. During **adulthood**, **biliary cancers** develop.

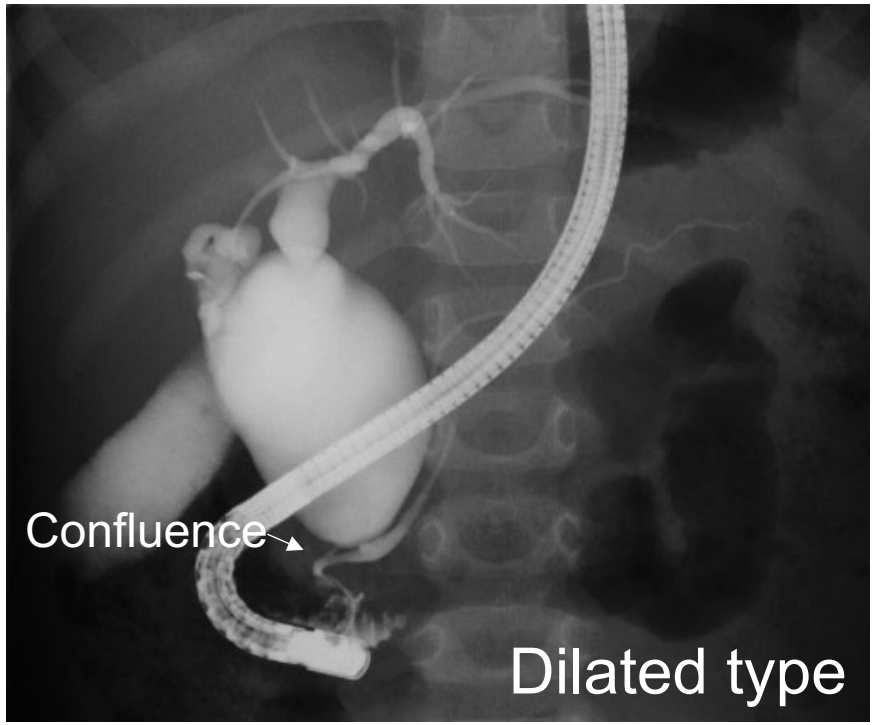


Normal PBM

What is Pancreaticobiliary Maljunction?

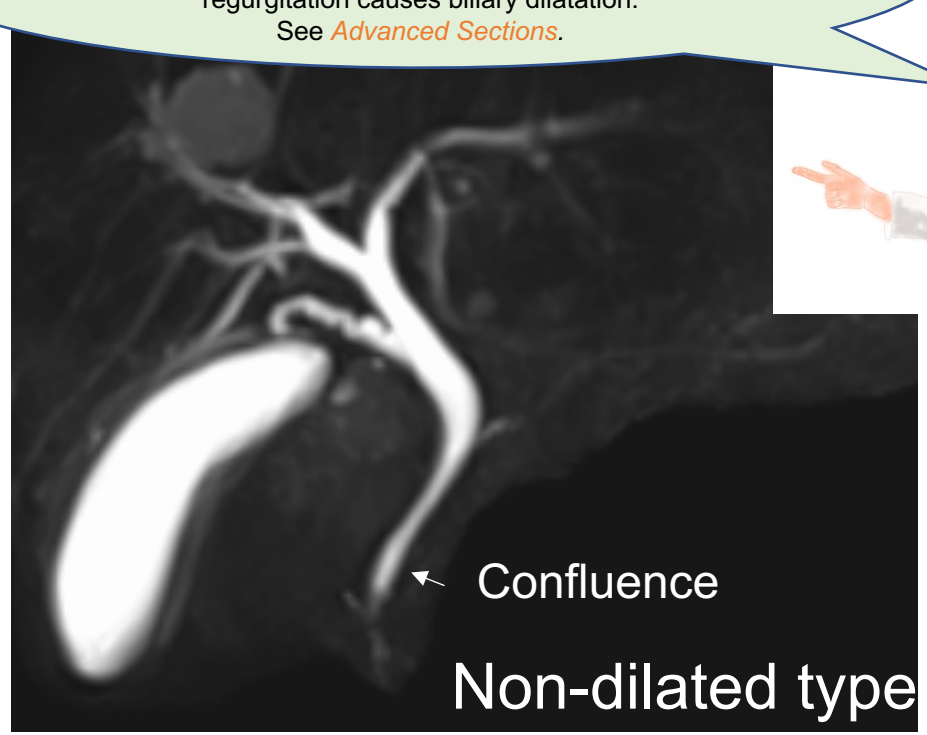
Two major categories according to whether the bile duct is dilated or not:

- Dilated type = congenital biliary dilatation (CBD)
- Non-dilated type



The definition of non-dilated type remains problematic.
See *Basic* and *Advanced Sections*.
Existence of non-dilated type denies that pancreatobiliary regurgitation causes biliary dilatation.
See *Advanced Sections*.



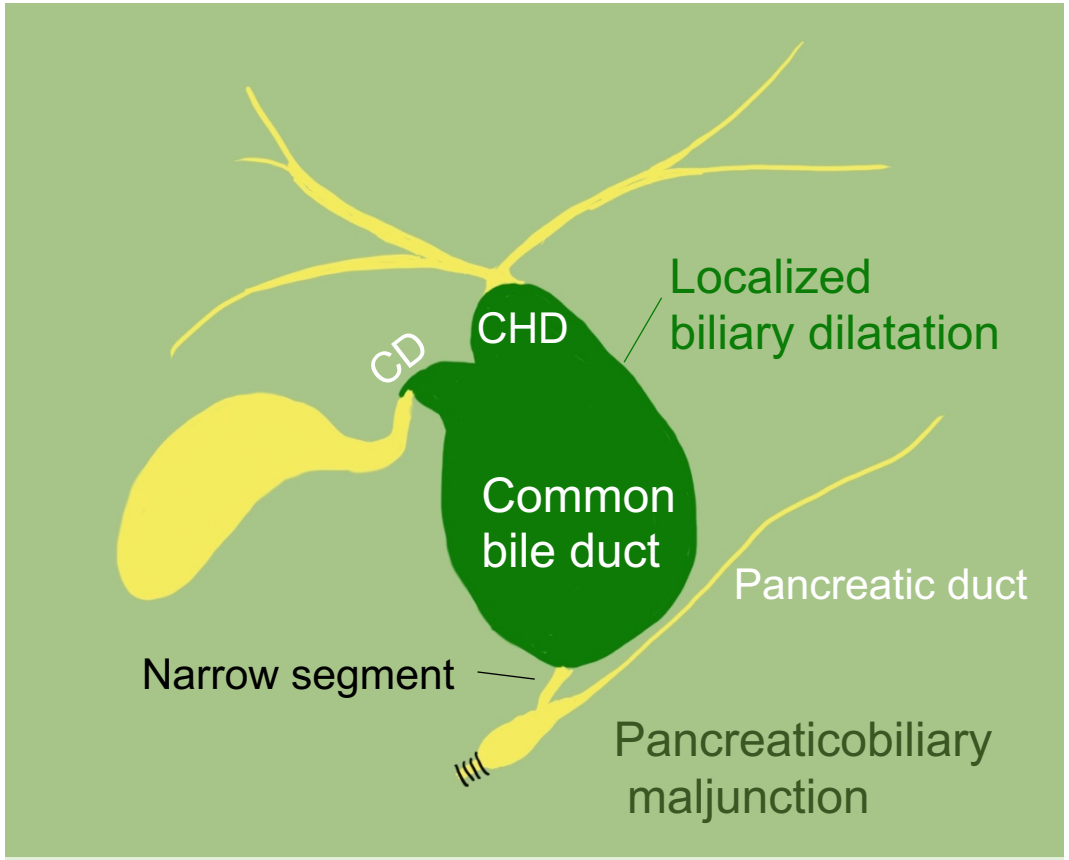


What is Congenital Biliary Dilatation (CBD)?

Definition

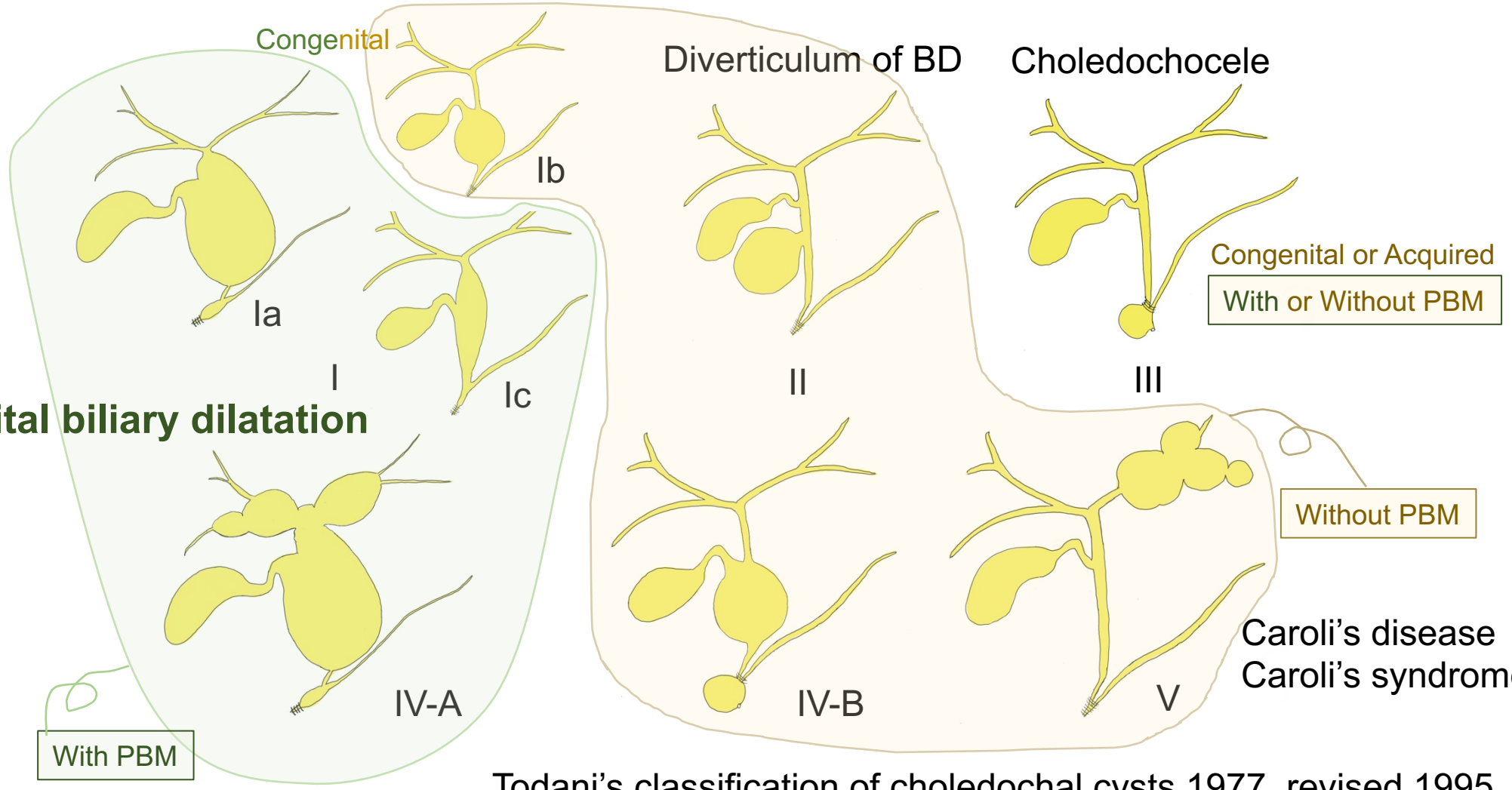
Congenital malformation involving **PBM** and **localized dilatation of the bile duct (BD)** including the common bile duct

Definition of CBD proposed by the Japanese Study Group on Pancreaticobiliary Maljunction (JSGPBM) in 2015



In this case, the common hepatic duct (CHD) and the base of the cystic duct (CD) are dilated, but bile ducts upstream to the CHD and the gallbladder are not dilated (localized dilatation).

The term choledochal cyst involves different entities

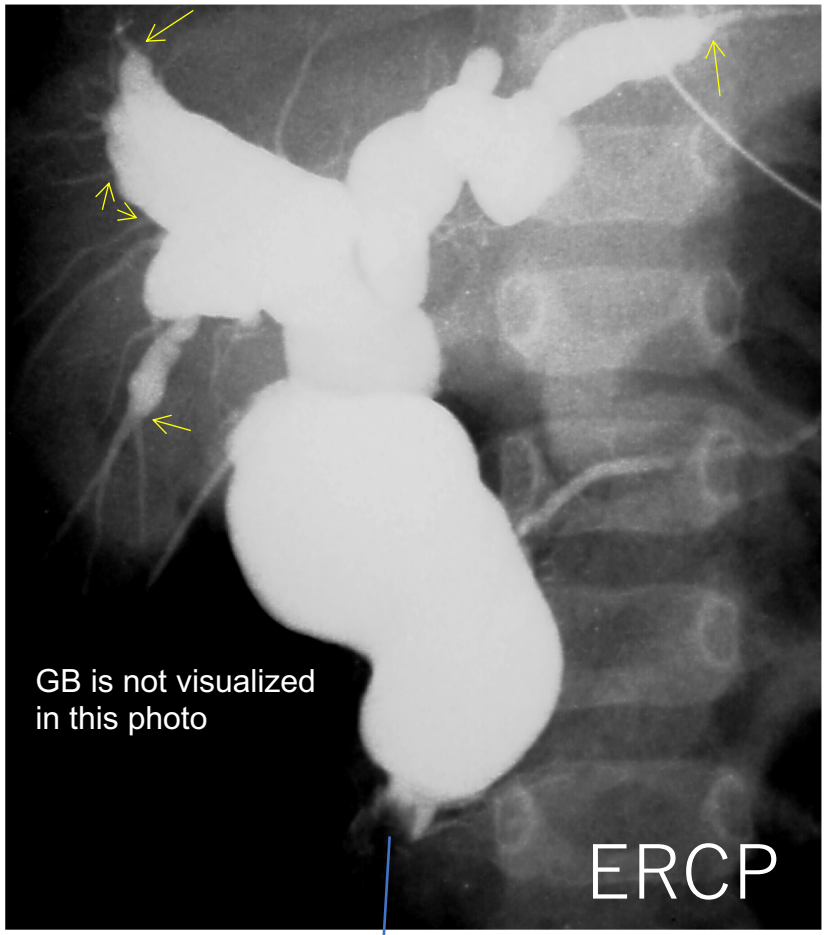


Todani's classification of choledochal cysts 1977, revised 1995

“Localized dilatation of BD” indicates “congenital” disease

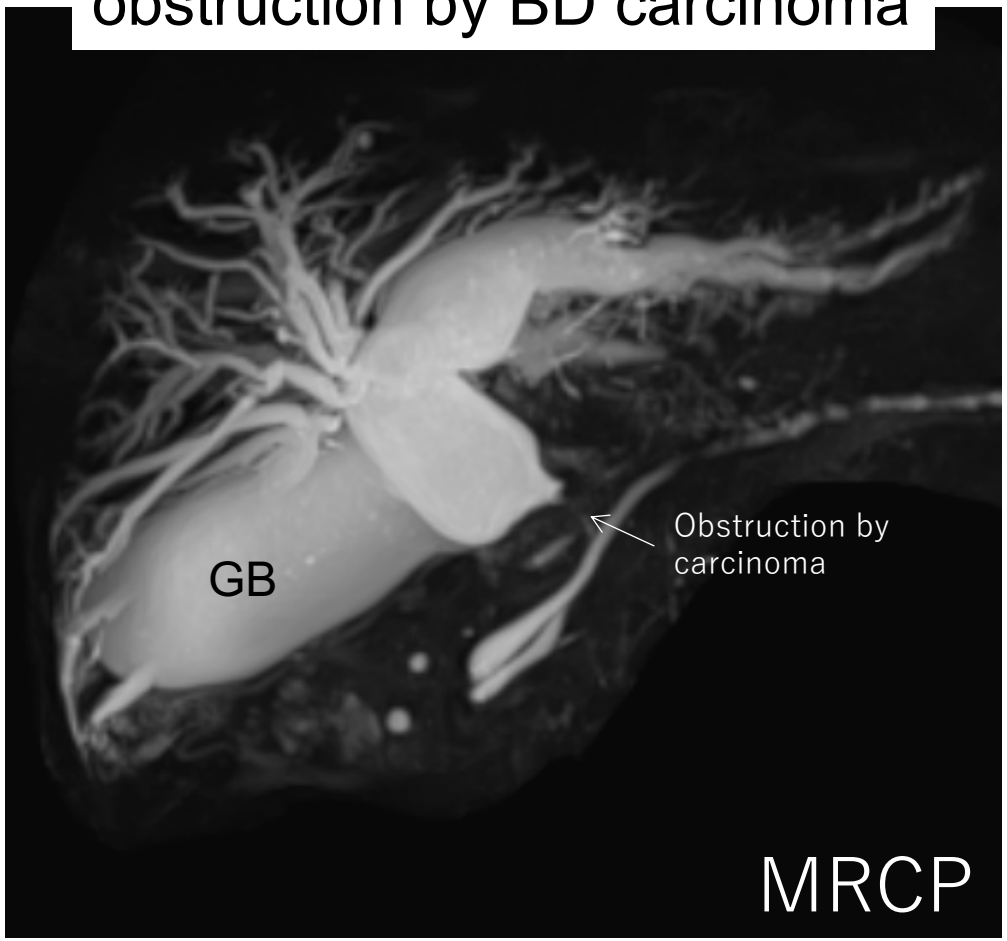
Even when dilatation extends to intrahepatic BDs, dilated areas are localized. **Dilated BDs abruptly change to those of normal caliber** (→).
The GB is never enlarged, although the cystic duct is dilated at its base.
However, in secondary dilatation, dilatation extends smoothly upstream to the peripheral intrahepatic BD.
GB is also enlarged (Courvoisier sign).

Congenital biliary dilatation



Protein plug
compacted in the common channel

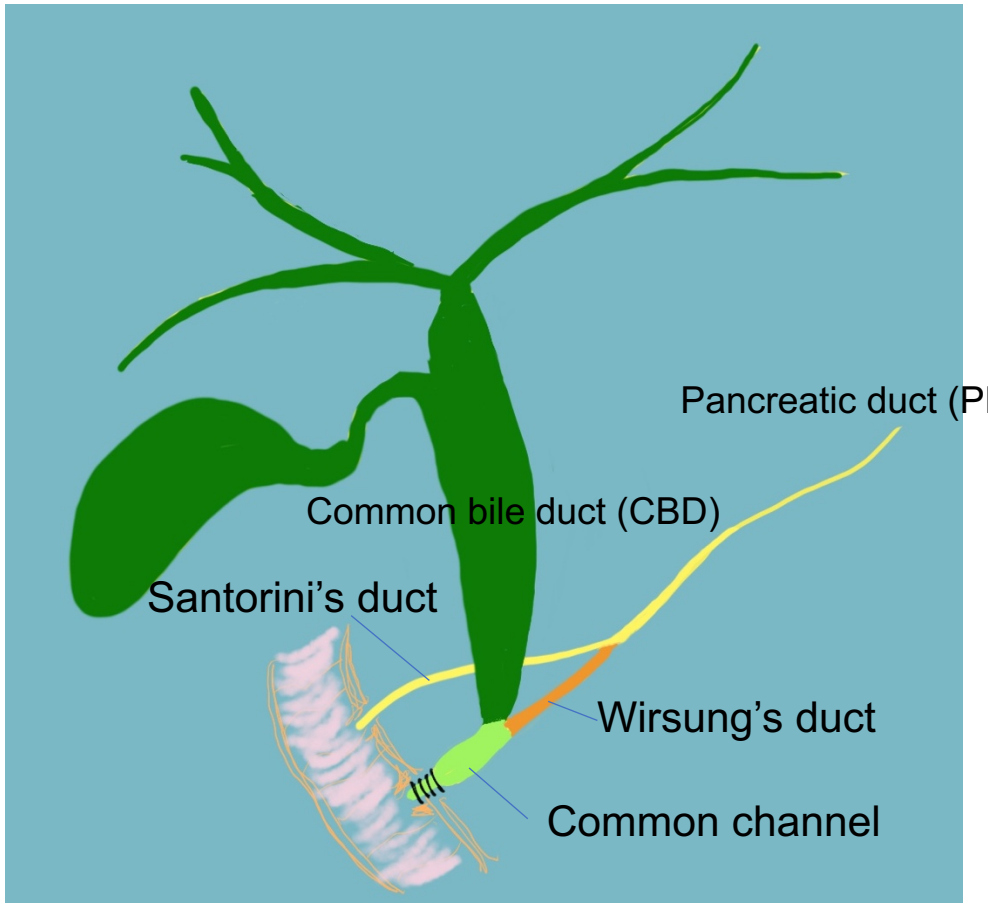
Secondary dilatation due to obstruction by BD carcinoma



Smoothly dilated until peripheral intrahepatic BD.
GB is also enlarged.

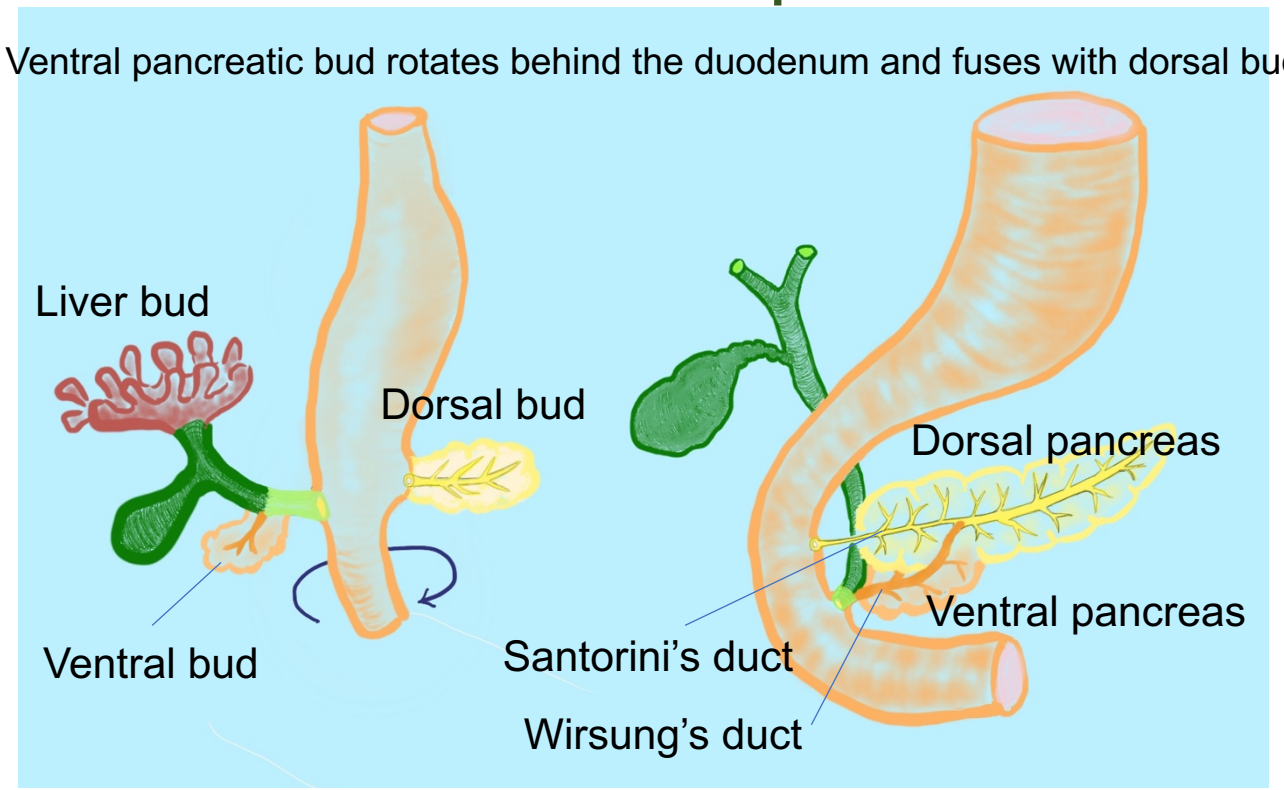
PBM arises from anomalous genesis of ventral pancreatic duct and bile duct

What is wrong in this figure?



Answer: Santorini's duct (dorsal PD) is depicted behind the CBD. It must be ventral to the CBD.

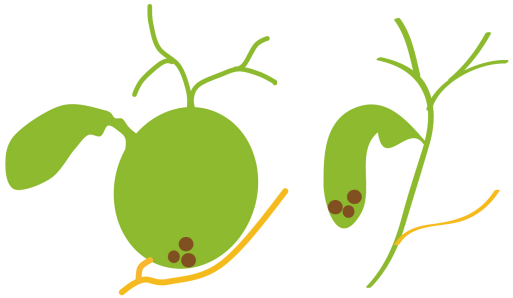
Pancreas development



Connection anomaly never happens between the BD and dorsal pancreatic duct in PBM.

Fusion failure between the ventral and dorsal pancreatic ducts (i.e., pancreas divisum) can occur together with PBM. See *Advanced Section*.

PBM causes symptoms only with secondary factors

Age	Neonates	Infants	Children	Adults	Elderly
Cause of symptoms	Debris	Protein Plugs		Gallstones	Biliary cancers
	See *Advanced Section*, pp160–163	See *Basic Section*, pp21–25 *Advanced Section*, pp148–158			See **pp15,16,** *Basic Section* pp26–28
					
Type of PBM	Dilated Type A	Dilated Prone to Type C	Dilated Type A	Incidental gallstones Non-dilated	Dilated Non-dilated
					Cancer remains asymptomatic Until advanced

See **p8**, Basic Section, pp35–40

Pathogenesis varies by life stage

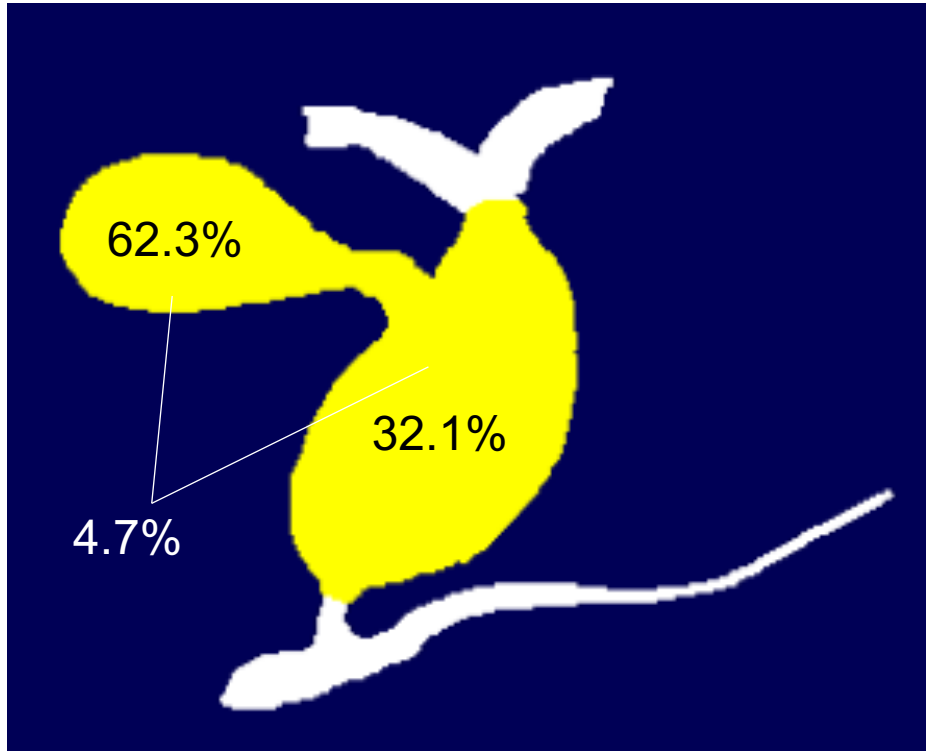
Treatment for pancreaticobiliary maljunction

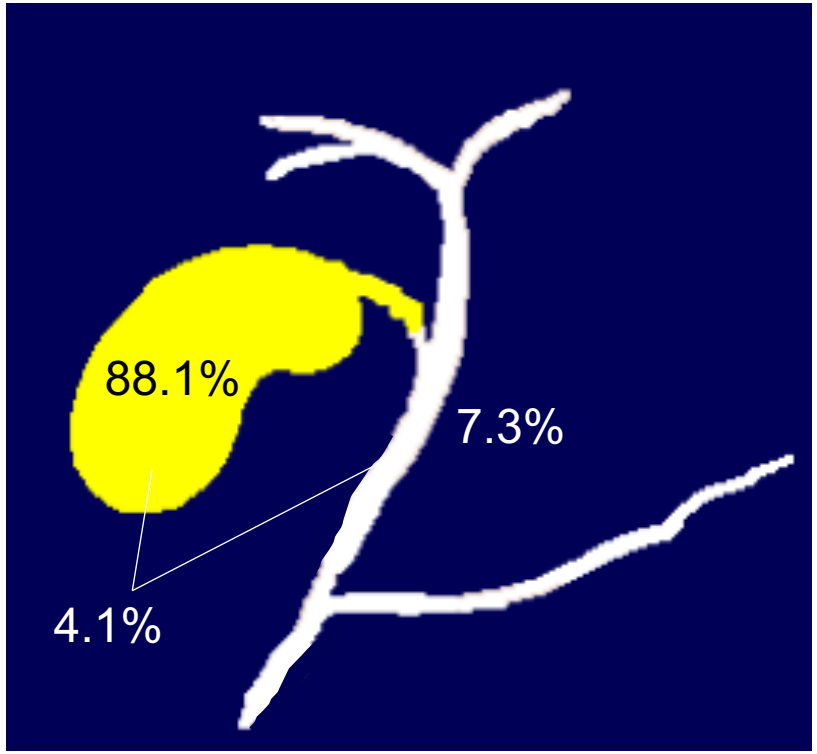
- Rationale for treatment:
  - ✓Not merely to correct malformation!
  - ✓Treatment is indicated to prevent complications.
- Complications of PBM:
  - ✓**Pediatric cases**: Recurrent **symptoms** including abdominal pain, vomiting, and jaundice.
  - ✓**Adult cases**: High incidence of **biliary tract cancer** (see p15).
- Surgical Management:
  - **Congenital biliary dilatation**: Requires **bile duct resection.**
  - **Non-dilated PBM**: Requires **at least cholecystectomy.***

*There is currently no consensus on the necessity of bile duct resection for non-dilated PBM due to the low incidence of carcinogenesis within the bile duct.

Biliary carcinogenesis

1,511 adult cases of PBM
Japan nationwide survey 1990–2007





Cancer rate	CBD 21.6%	Non-dilated PBM 42.4%
	215/997 cases	218/514 cases

Biliary cancers arise 10–20 years earlier in PBM

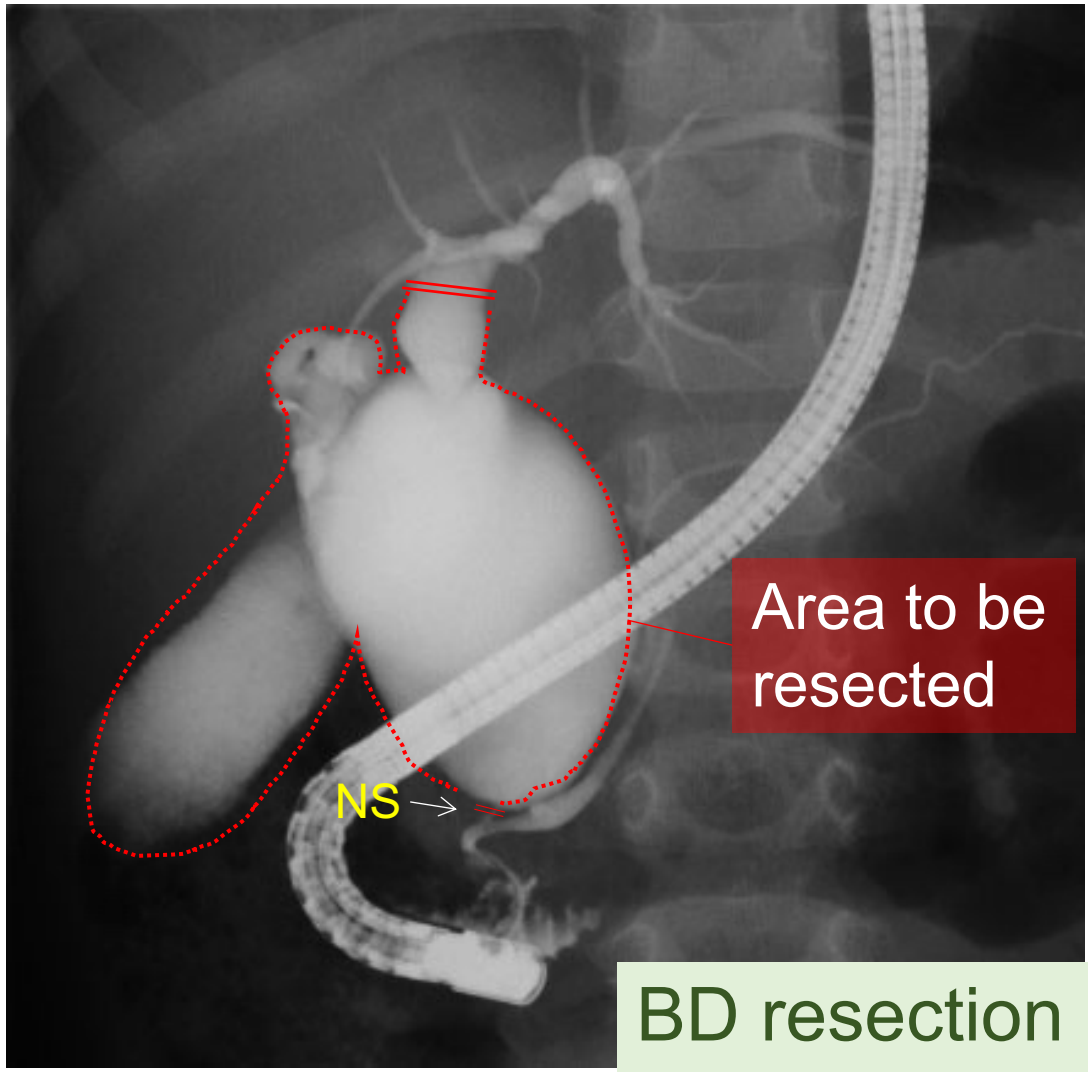
	Gallbladder cancer		*P*
PBM	Yes	No	
	36 cases	232 cases	
Age (years)	**58.9** ± 8.8	**69.4** ± 8.7	< 0.01
M:F	1:2.1	1:2.9	NS

	Bile duct cancer		P
PBM	Yes	No	
	6 cases	159 cases	
Age (years)	**54.8** ± 14.3	**68.8** ± 9.1	< 0.01
M:F	1:2	1:0.7	NS

Kamisawa T, Tu Y, Kuwata G, Egawa N, Nakajima H, Tsuruta K, Okamoto A, Matsukawa M.
Biliary carcinoma risk in patients with pancreaticobiliary maljunction and the degree of extrahepatic bile duct dilatation.
Hepatogastroenterology. 2006;53(72):816-8.

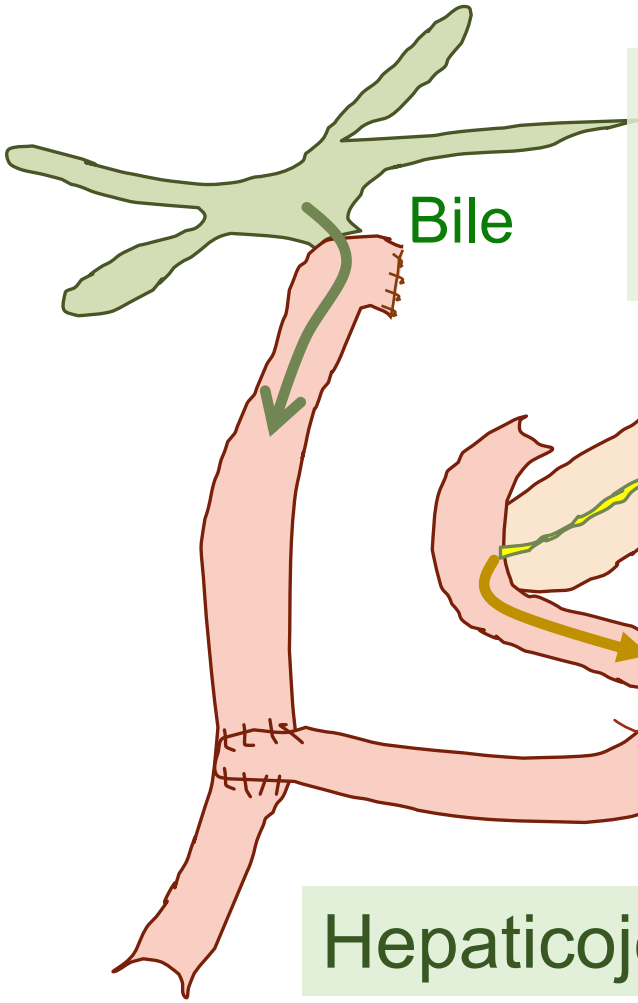
Surgery for CBD

Must resect where cancers would arise and stop the reflux of pancreatic juice to prevent cancers and symptoms



BD resection

NS, narrow segment



Bile and pancreatic juice flows are separated

Pancreatic juice

Hepaticojejunostomy

Recommended terms

- **Pancreaticobiliary maljunction (PBM)** (Official MeSH term since 2020)
- **Congenital biliary dilatation (CBD)**
- **Bile duct resection** as a procedure for CBD

Non-recommended terms

- Pancreaticobiliary malunion or pancreatobiliary malunion
- Choledochal cyst (when indicating CBD [Todani types Ia, c, and IV-A])

The term "choledochal cyst" encompasses various entities, such as CBD, choledochocele, and Caroli's disease.

Basics

Pathophysiology, diagnosis, and classification

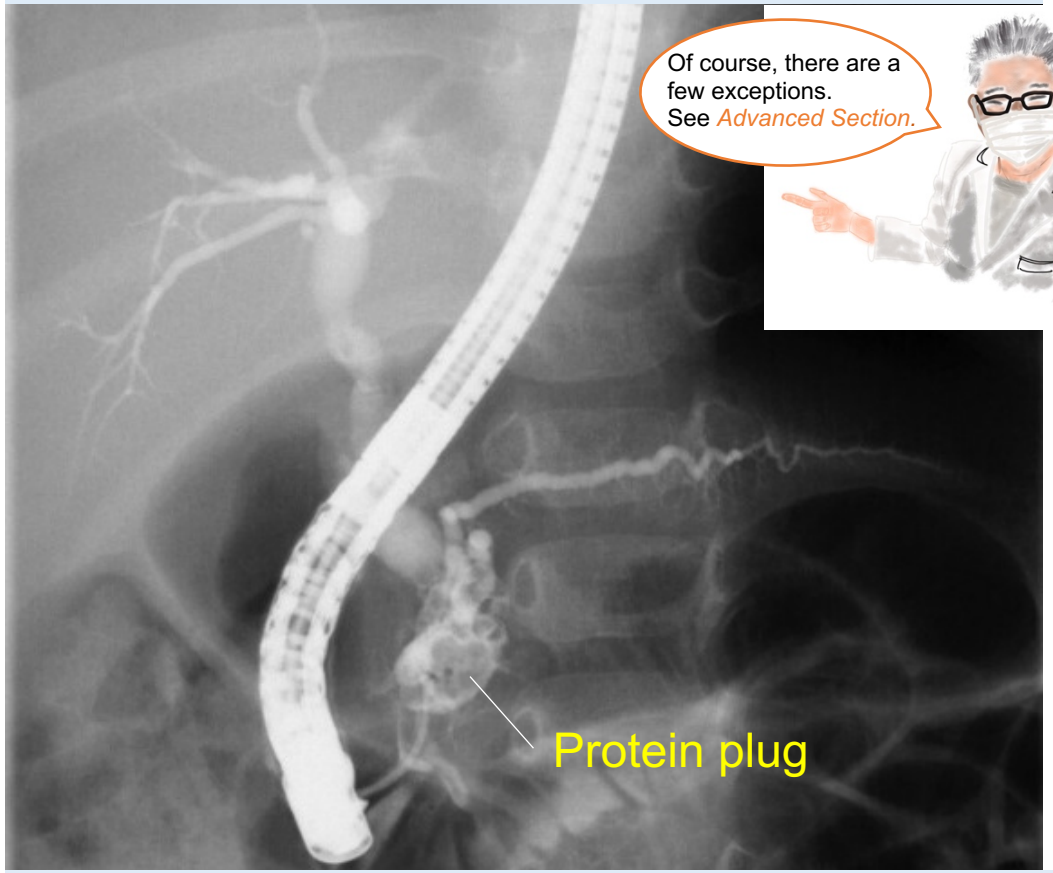
First version, September 2022

Symptoms in 128 pediatric patients

Symptoms & Laboratory data	Number of patients	%
Abdominal pain	108	85.7
Nausea/vomiting	81	64.3
Jaundice	58	46.0
Abdominal mass	29	23.0
Fever	33	26.2
Acholic stool	35	27.8
T.bil > 1.0 mg/dL	61	48.4
ALT > 50 IU/L	73	57.9
Amylase > 200 IU/L	53	42.1

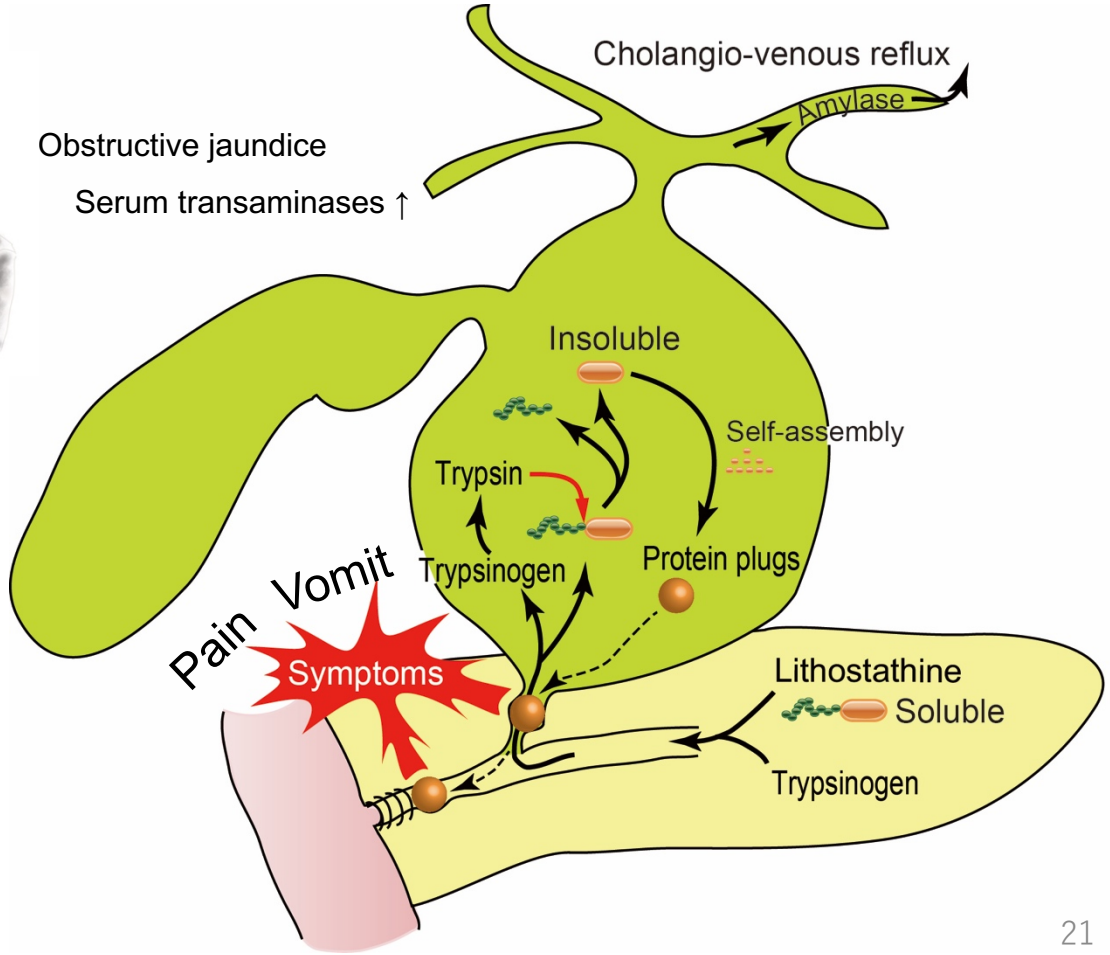
Symptomatology in childhood

Protein plugs cause symptoms in children.
Pancreatobiliary reflux produces protein plugs, which obstruct the common channel or narrow segment and increase intraductal pressure.



Children are sensitive to increased intra-bile duct pressure and are liable to vomit and feel pain.

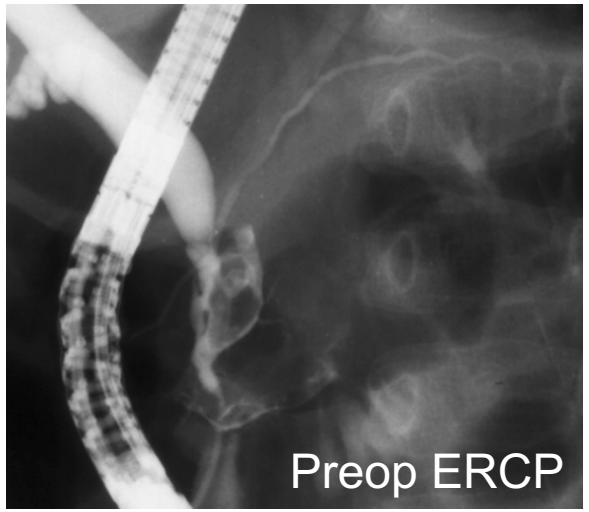
Hyperamylasemia is observed in 60% of pediatric cases but is caused by **cholangiovenous reflux** in most cases. True pancreatitis is rare (7%). See two slides ahead.

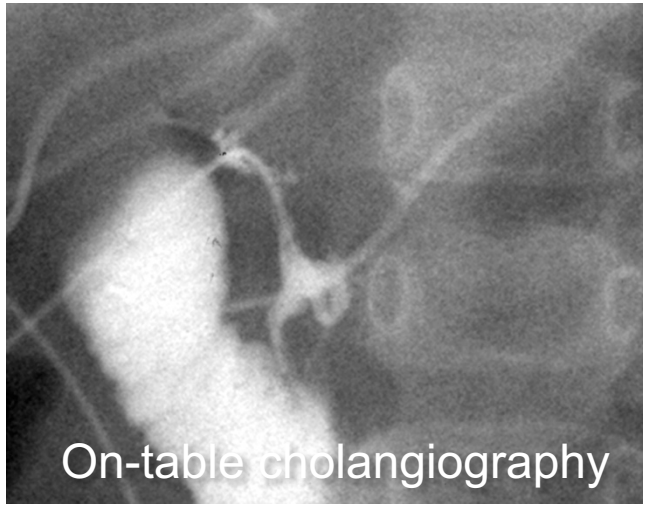


Kaneko K, et al. *Dig Dis Sci* 2007;52:1979-86

Protein plug theory explains intermittent symptoms

Characteristics of protein plugs		Nature of symptoms
Fragile	→ Spontaneous dissolution →	Self-limited
Repeating production	→	Recurrence





Spontaneous dissolution

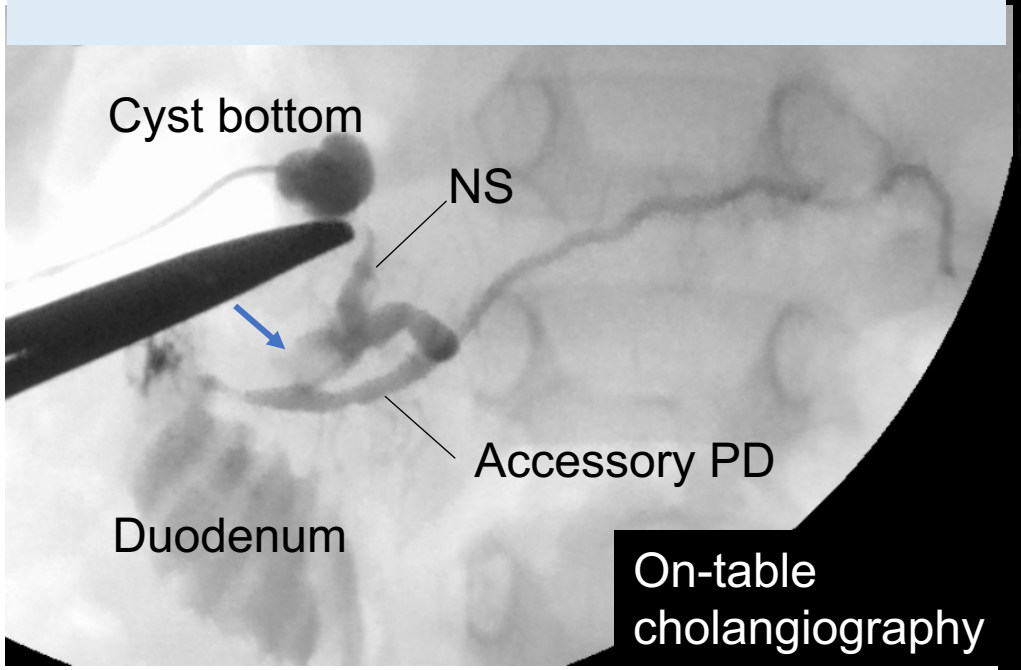
This is the reason why people often say they have never seen such protein plugs!



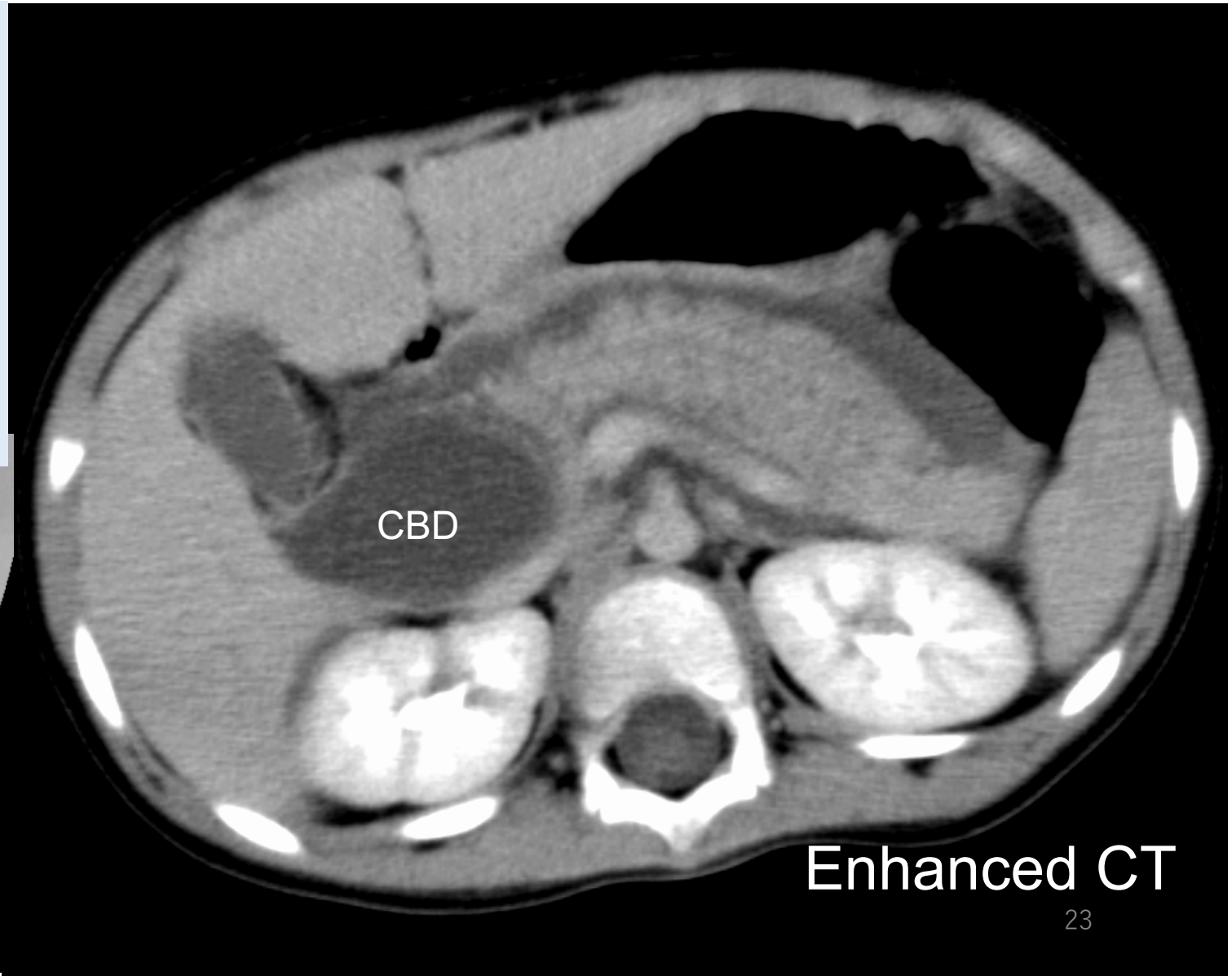
Case 1 True pancreatitis is observed only in 7% of pediatric patients

A 3-year-old boy
Abrupt onset of severe abdominal pain
T.bil 0.8, D.bil 0.1 mg/dL, AST 499, ALT 156 IU/L, amylase 1146, lipase 1816 U/L

- **Swollen pancreas surrounded by effusion**
- After pancreatitis had subsided, surgical cholangiography revealed a protein plug compacted in the common channel (arrow) and drainage through the accessory PD, suggesting protein plugs were hard in this case.



On-table cholangiography



Enhanced CT

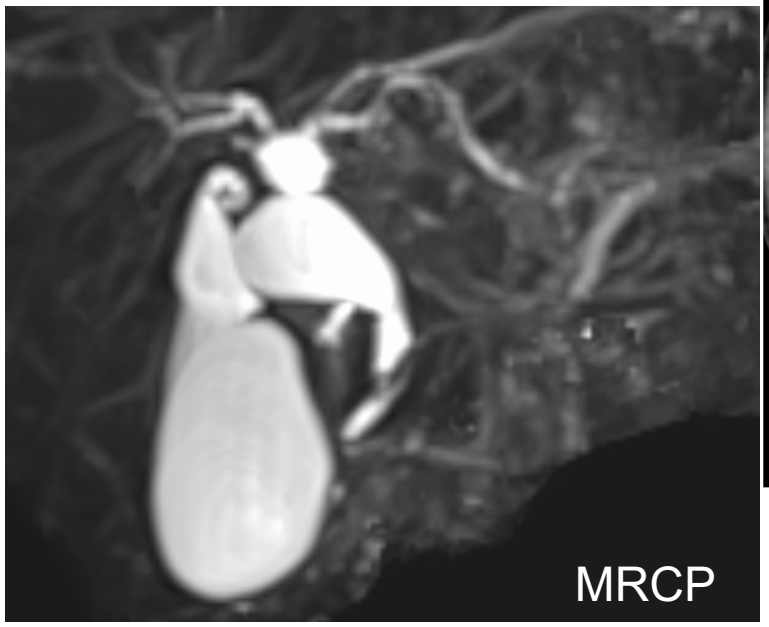
Case 2

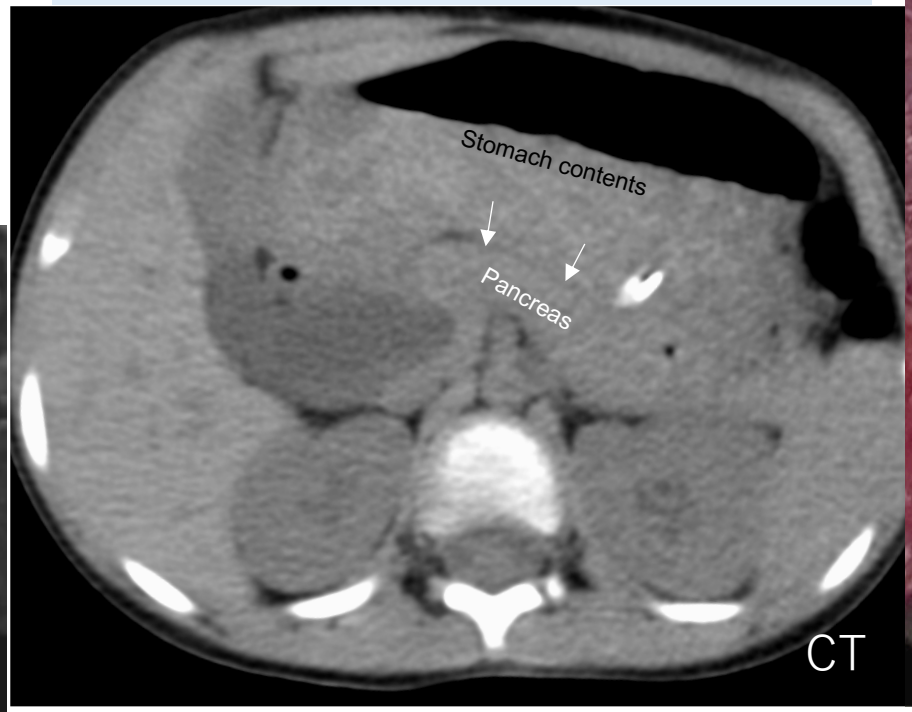
Pseudopancreatitis Hyperamylasemia without evidence of pancreatitis

A 3-year-old girl
Repeating **abdominal pain** and vomiting

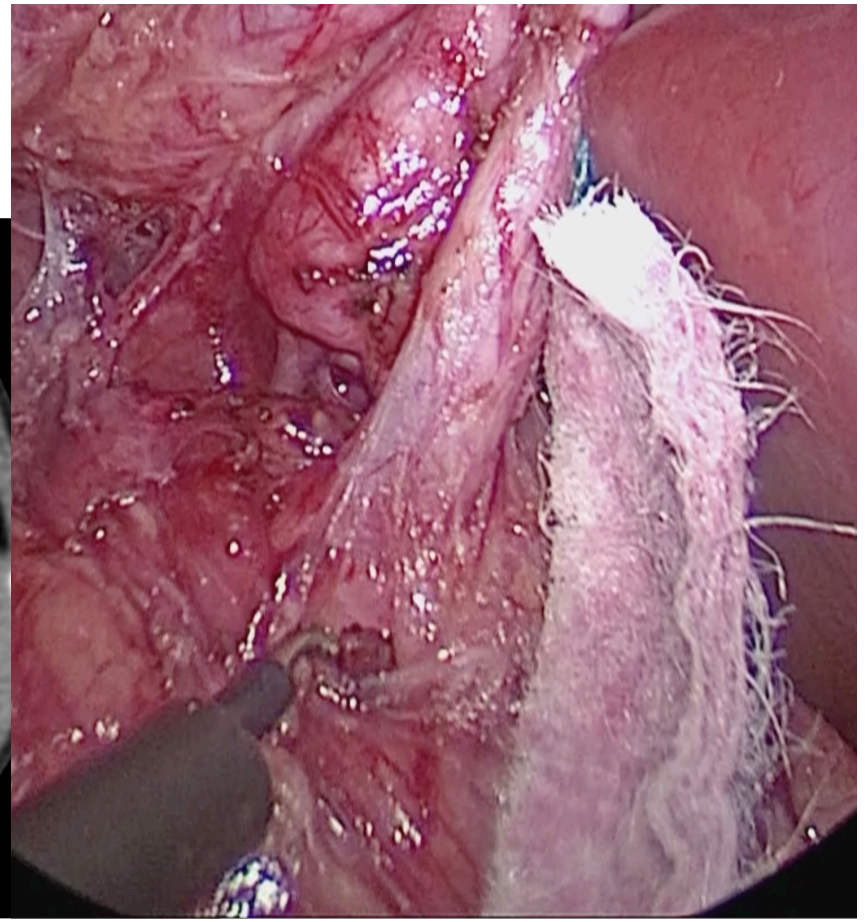
T.bil 0.7 D.bil 0.1 mg/dL
AST 176 ALT 235 IU/L
Amylase 1385 U/L

Pancreas is normal on imaging studies and operative findings despite abdominal pain and elevated serum amylase level.
Hyperamylasemia in most cases are due to cholangio-venous reflux.





No swollen pancreas



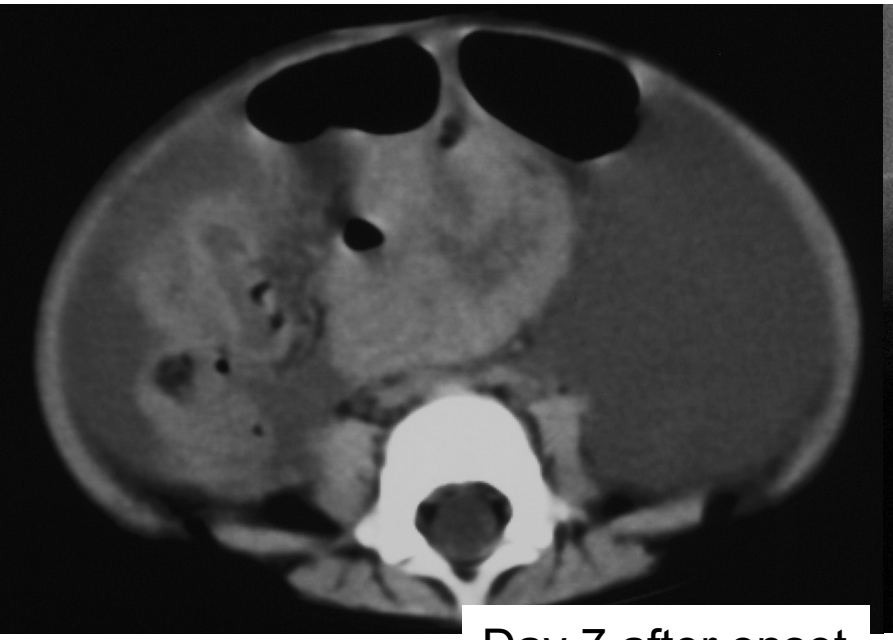
No adhesion
around the intrapancreatic BD

Case 3 Biliary perforation is rare: 3%–6% of children

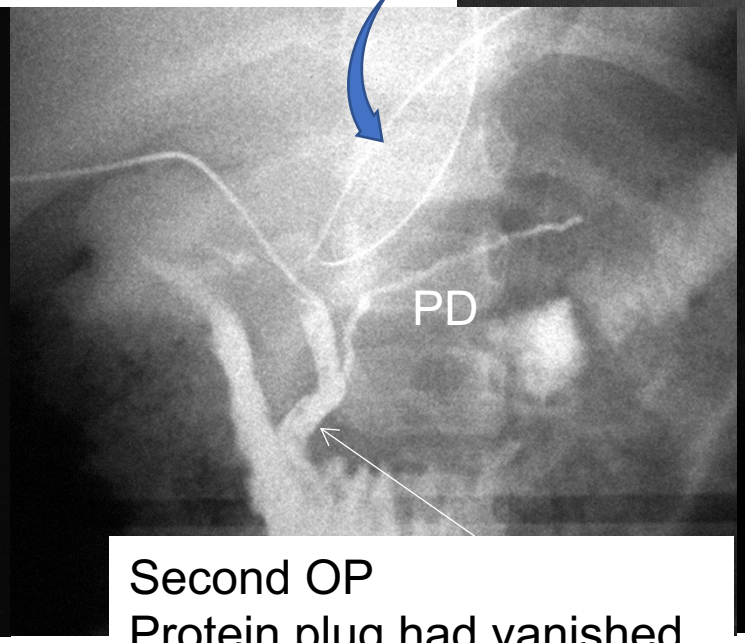
Most protein plugs are fragile and dissolve spontaneously (transient symptoms). However, exceptionally hard protein plugs produce abrupt increases in intraductal pressure, causing a rupture.

A 1-year-old girl
Sudden onset of vomiting and abdominal pain on day 4,
Diarrhea and abdominal distention on day 5,
Fever on day 6, and pleural effusion on chest Xp on day 7

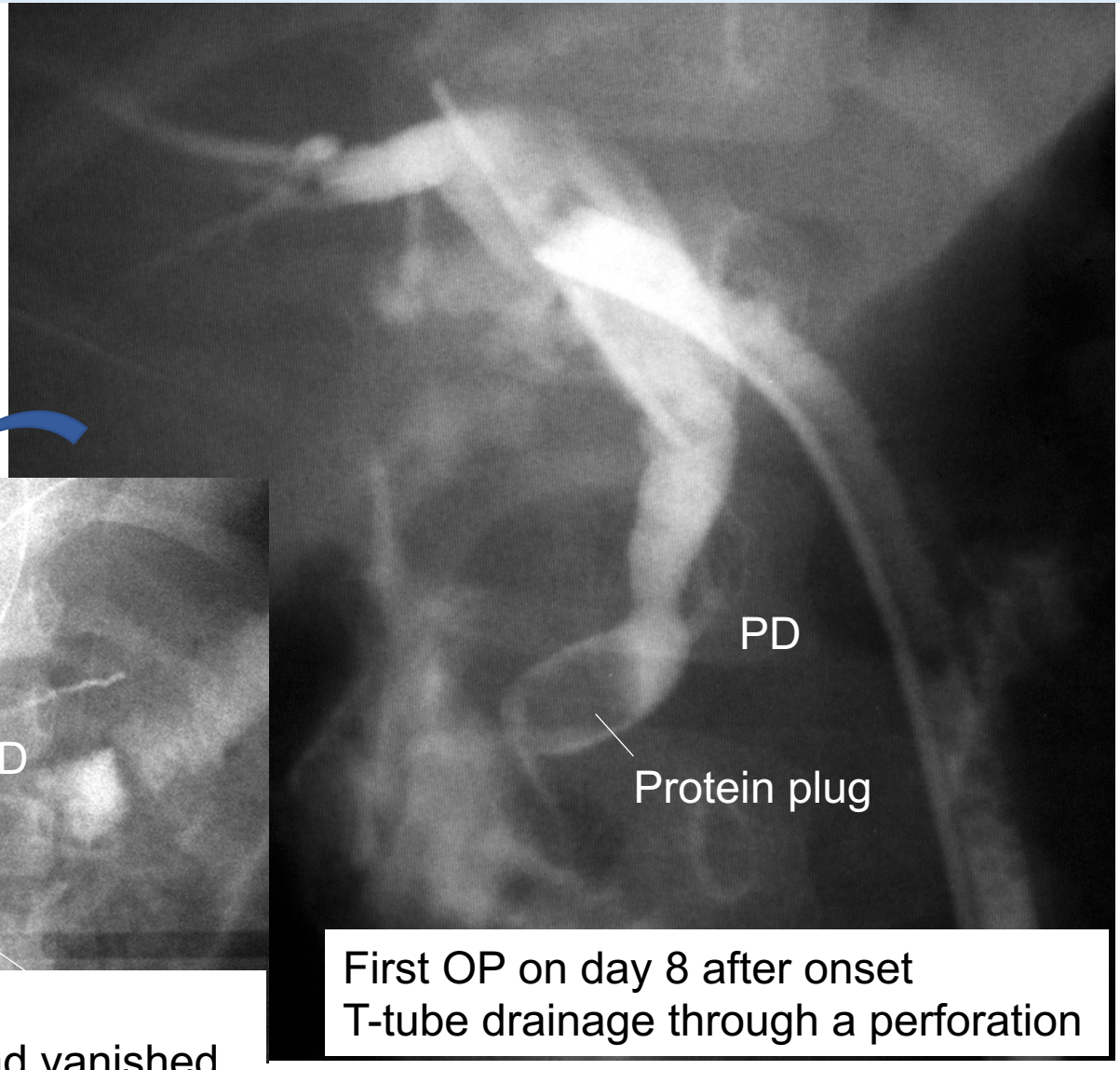
Bile peritonitis causes relatively mild irritation and is often overlooked.



Day 7 after onset



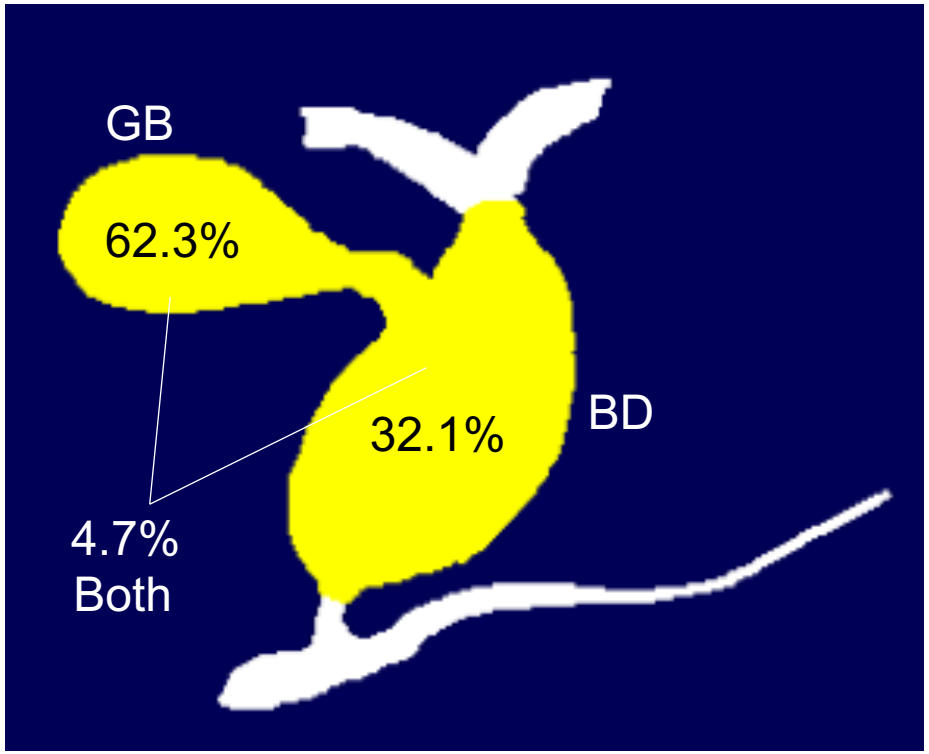
Second OP
Protein plug had vanished

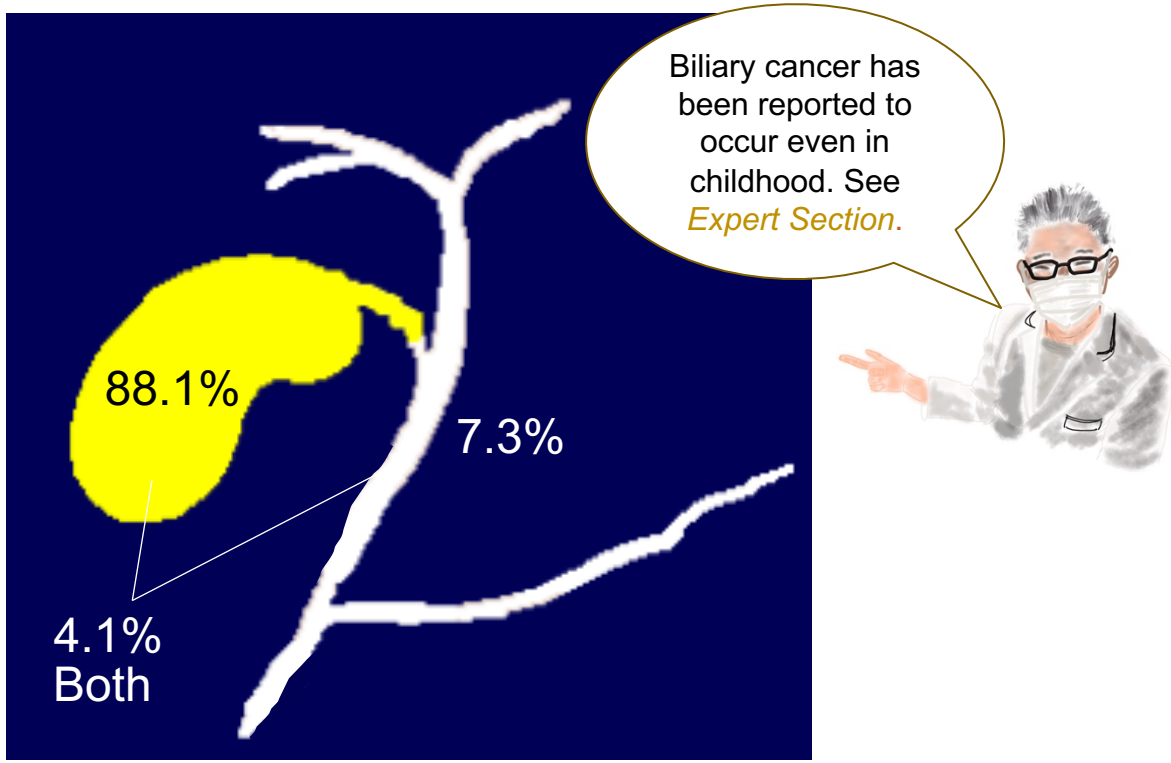


First OP on day 8 after onset
T-tube drainage through a perforation

Biliary carcinogenesis

1,511 adult cases of PBM
Japan nationwide survey 1990–2007



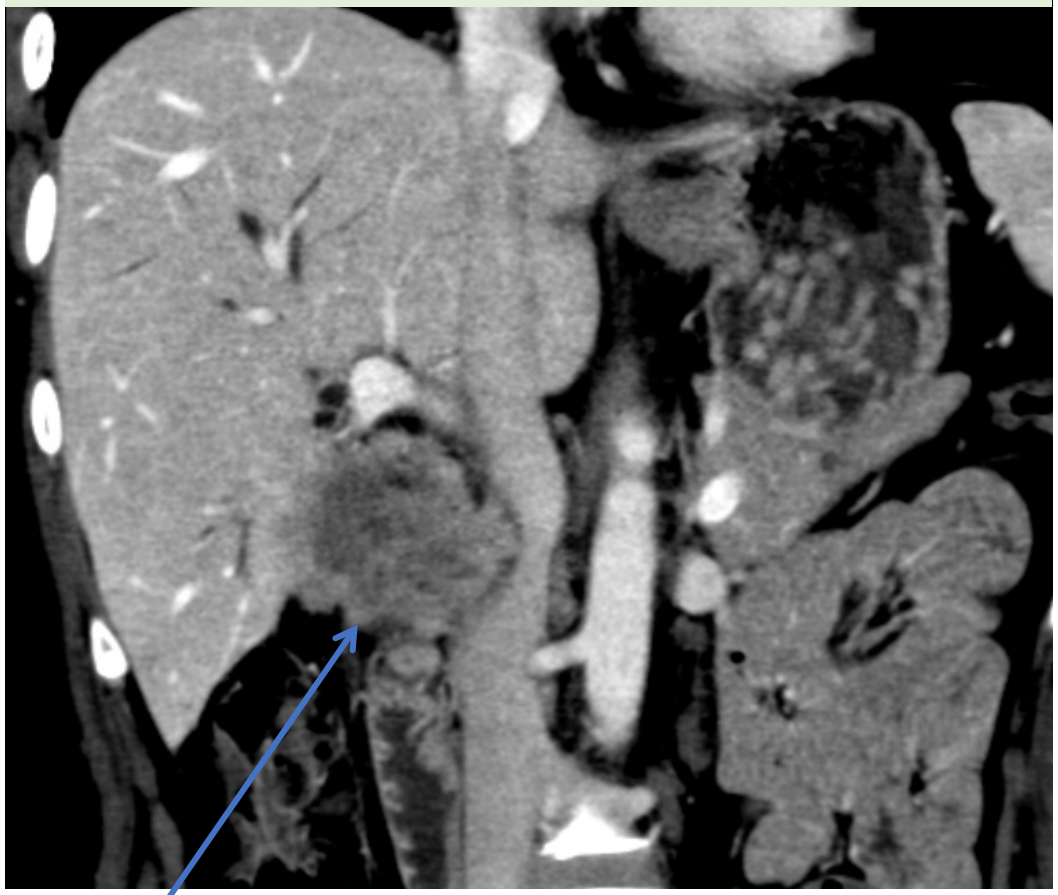


Cancer rate	CBD 21.6%	Non-dilated PBM 42.4%

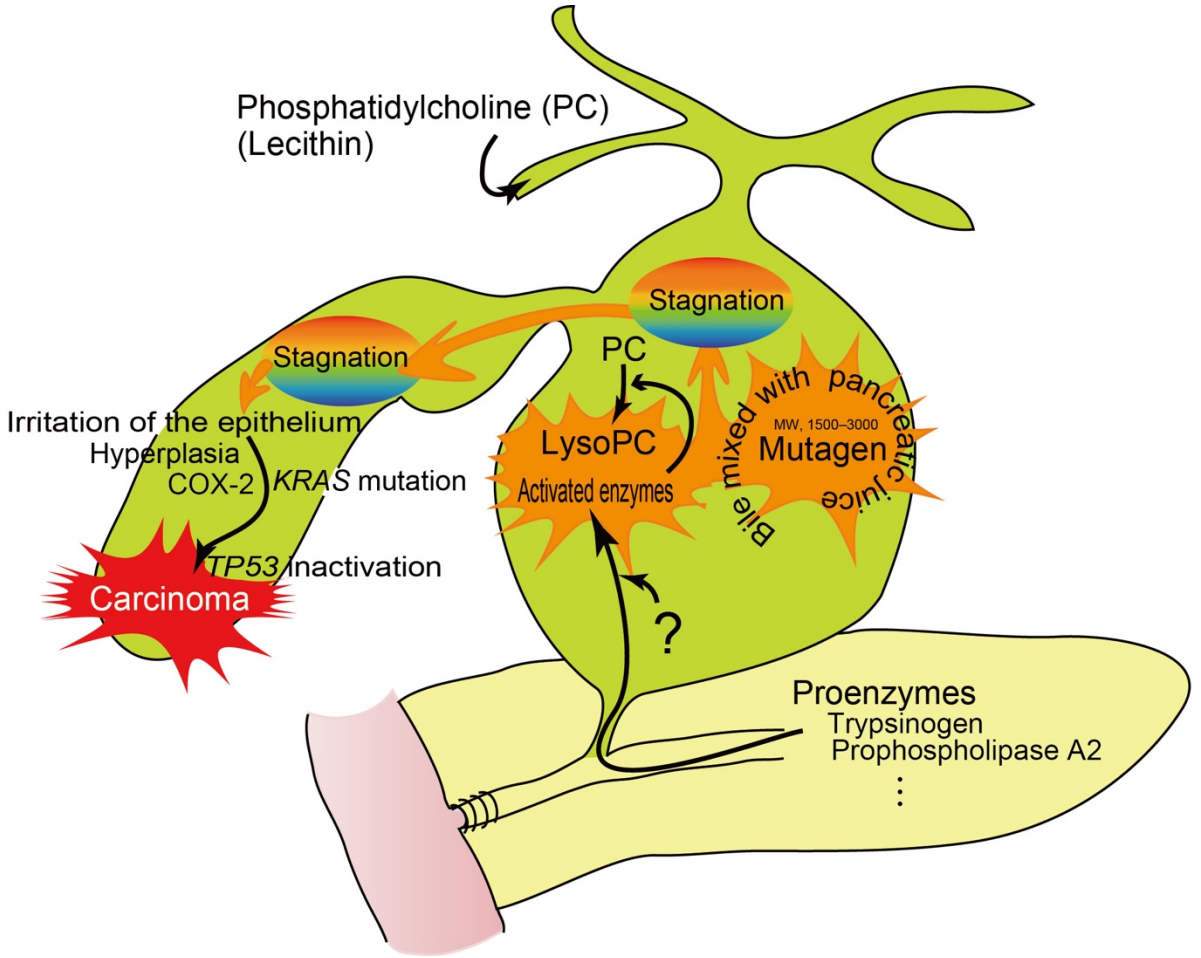
Carcinogenesis in region where bile stagnates → Indicates → Hazardous substances, produced by a mixture of bile and pancreatic juice, stagnate and cause chronic inflammation, which induces carcinogenesis

Cancer develops during adulthood

Occurs in patients 10 to 20 years younger than in patients without PBM



BD cancer in a CBD patient in her 40s



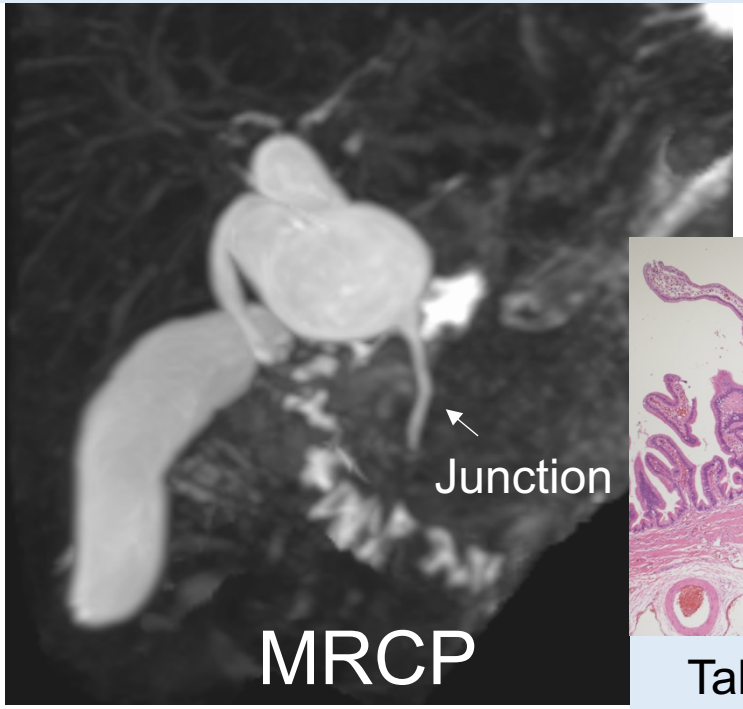
Hyperplasia–dysplasia–carcinoma sequence

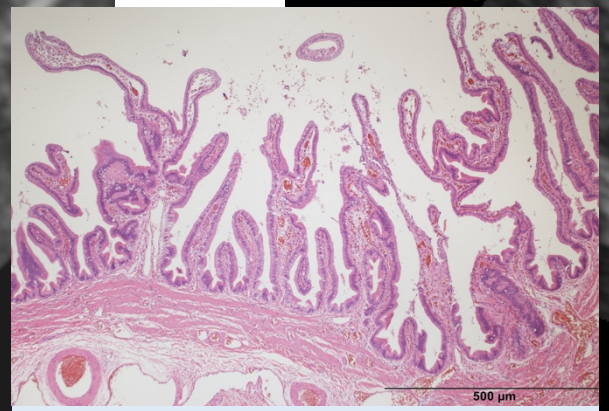
Case 4 Diffuse Hyperplasia of GB mucosa observed in a child

Epithelial hyperplasia in GB is detected as a thick wall on US and is observed from childhood. US during a medical check provides an opportunity to identify PBM without bile duct dilatation in adults.

A 3-year-old girl
Recurrent vomiting from 1 year of age
Abdominal pain, vomiting, and elevated serum transaminase levels led to US, which identified a Todani Type Ia CBD.

Thick wall of GB on ultrasonography (US)

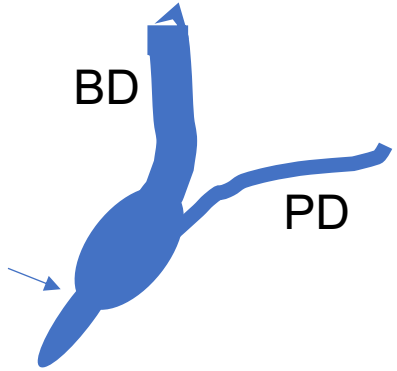




Tall GB epithelial folds

Diagnosis of PBM by imaging studies

- EUS or MD-CT demonstrates the junction of BD and PD outside the duodenal wall (definition)
- Demonstration of the junction of BD and PD above the sphincter action (functional diagnosis) = **Notch within the common channel**



Notch, indentation made by sphincter action

Criteria 1 requires judging whether the common channel is abnormally long, or the junction has an abnormal shape.
However, it is sometimes difficult because the judgement is subjective.
Rather, it is easier to find a notch, which the sphincter creates.

Imaging diagnosis

1. An abnormally long common channel and/or an abnormal union between the pancreatic and bile ducts must be evident on direct cholangiography, such as endoscopic retrograde cholangiopancreatography (ERCP), percutaneous transhepatic cholangiography (PTC), intraoperative cholangiography, magnetic resonance resonance cholangiopancreatography (MRCP), or three-dimensional drip infusion cholangiography computed tomography (3D-DIC-CT). However, in cases with a relatively short common channel, it is necessary to confirm that the effect of the papillary sphincter does not extend to the junction by direct cholangiography.
2. Pancreaticobiliary maljunction can be diagnosed if the pancreaticobiliary junction outside the wall can be depicted by endoscopic ultrasonography (EUS) or multi-planar reconstruction (MPR) images provided by multi-detector row computed tomography (MD-CT).

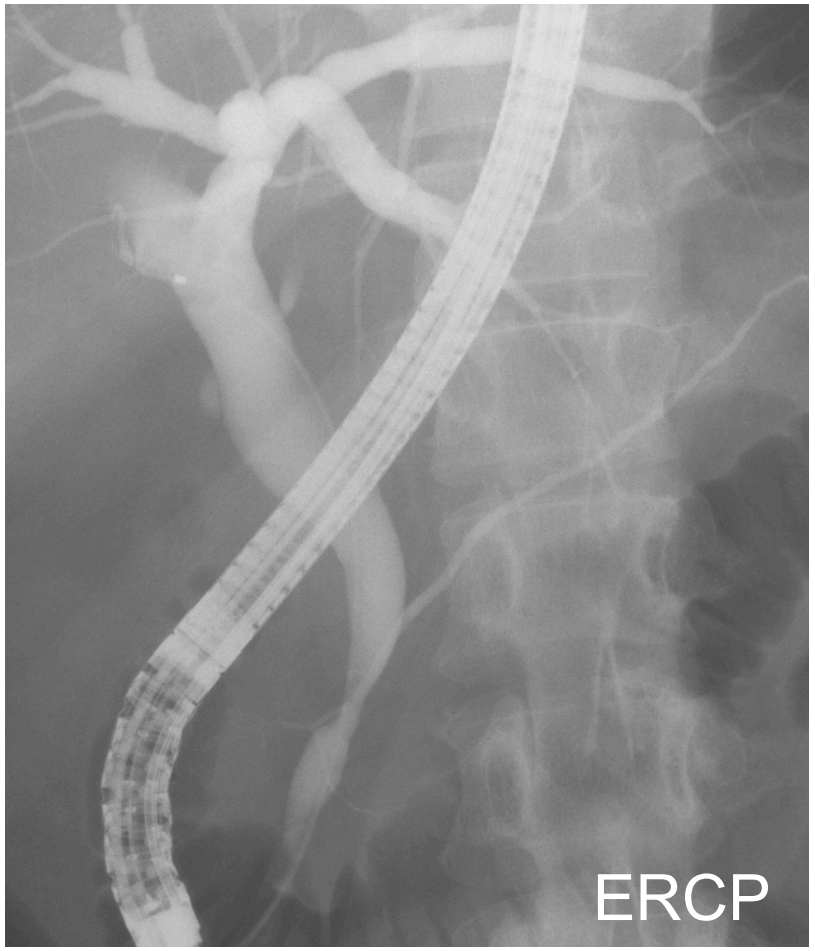
Diagnostic criteria for pancreaticobiliary maljunction 2013
J Hepatobiliary Pancreat Sci 2014;21:159–161

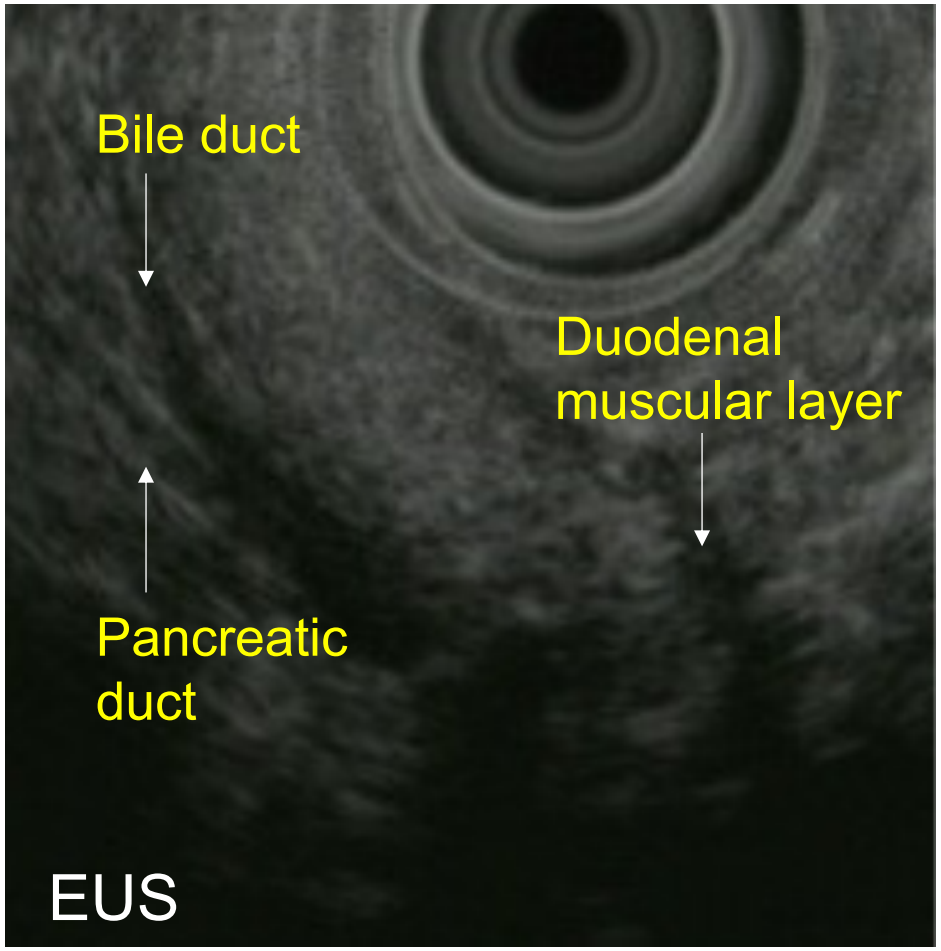
Case 5

EUS directly demonstrates the junction outside the duodenal wall

- A female in her 40s Abdominal pain
- EUS shows **union outside the duodenal wall = PBM**

EUS, endoscopic ultrasonography



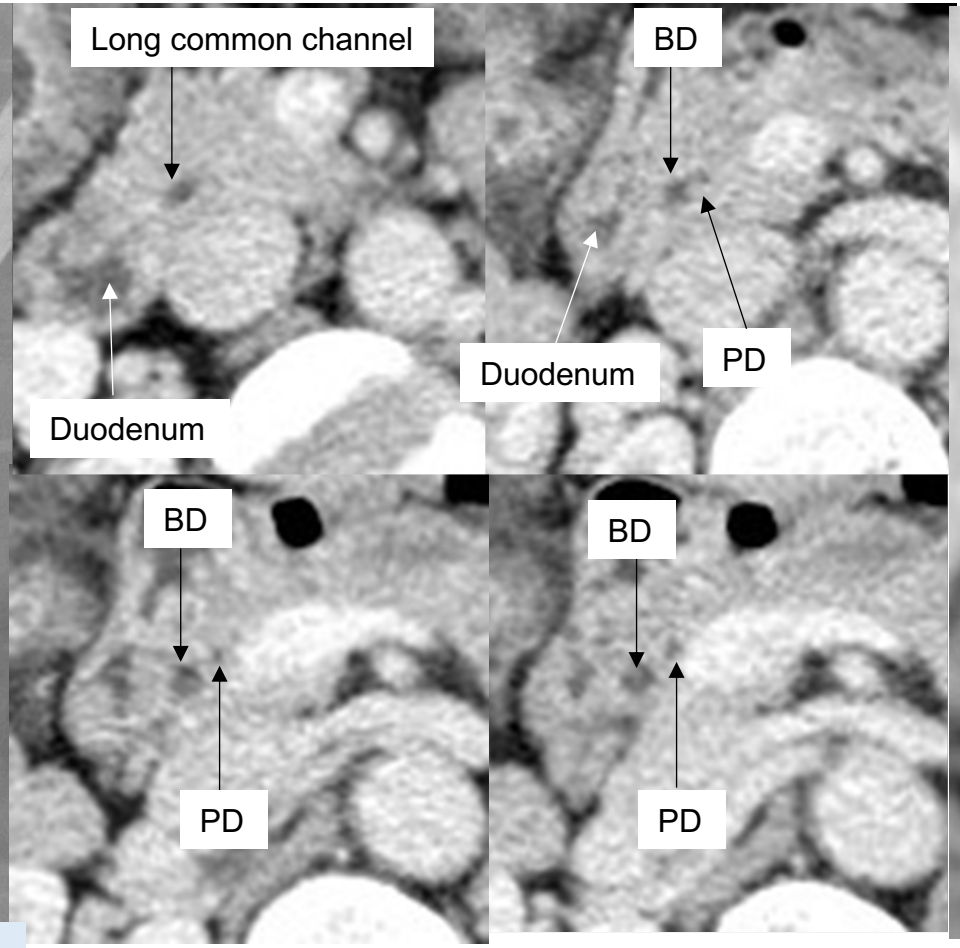


Case 6

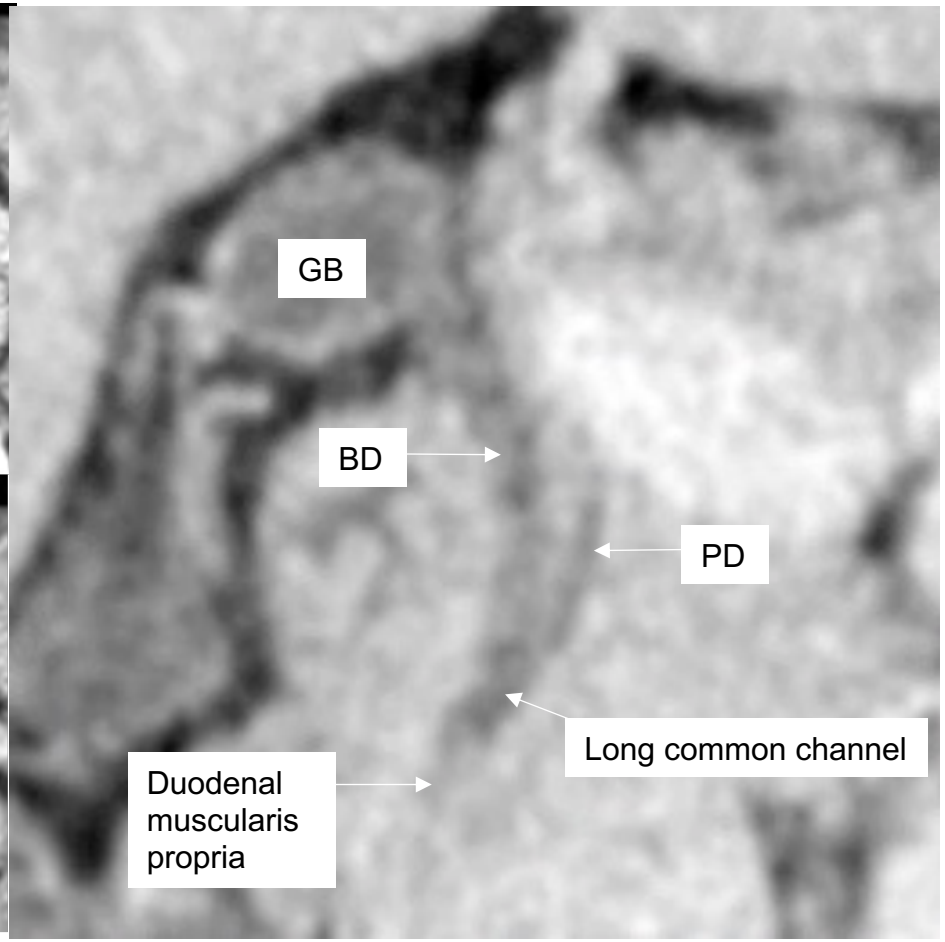
CT directly demonstrates the junction outside the duodenal wall



A female in her 40s
Right hypochondrial pain

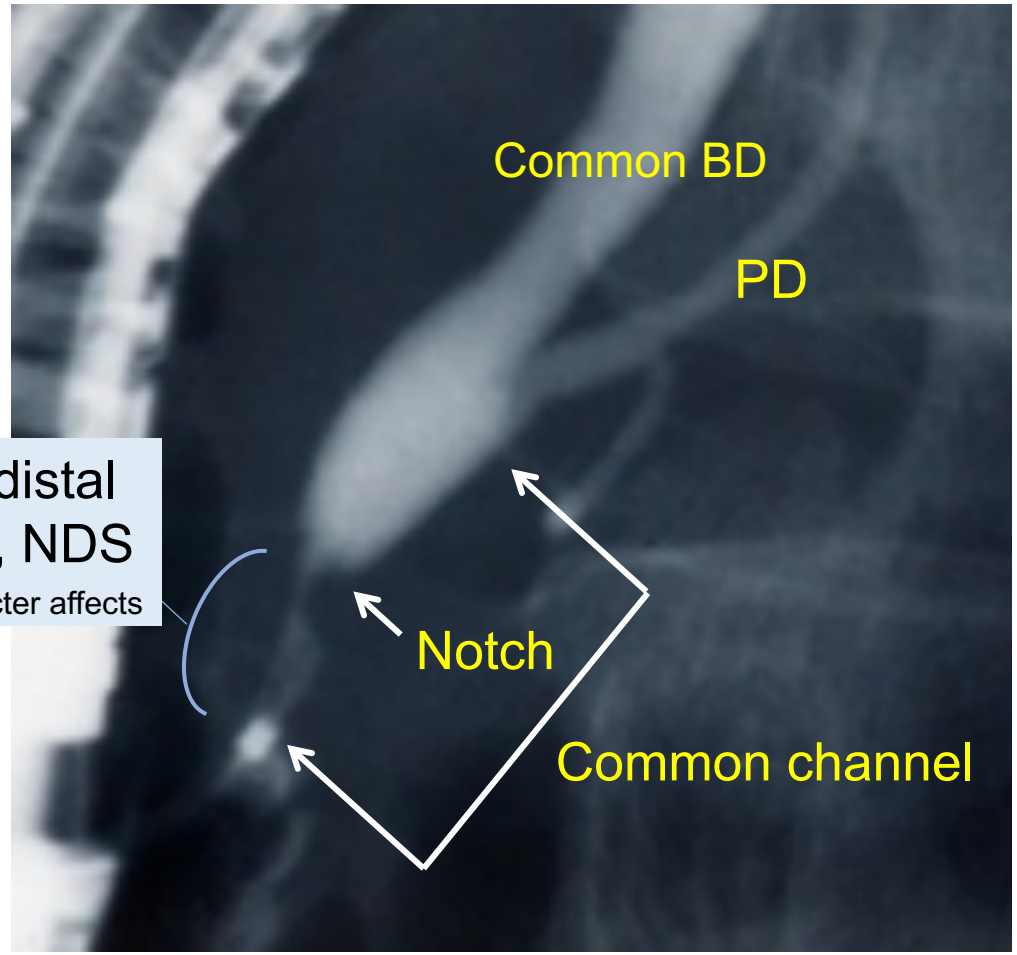


Axial section



Coronal section

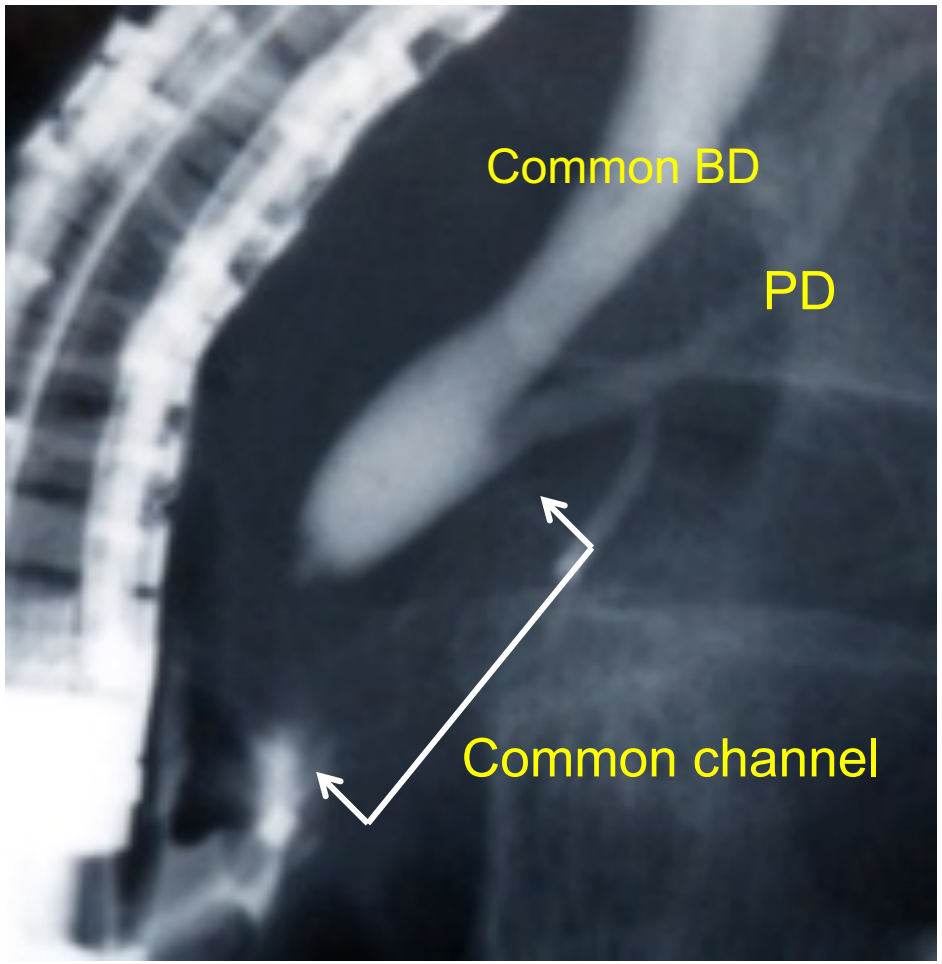
Notch on the common channel = PBM



Narrow distal segment, NDS

Area the sphincter affects

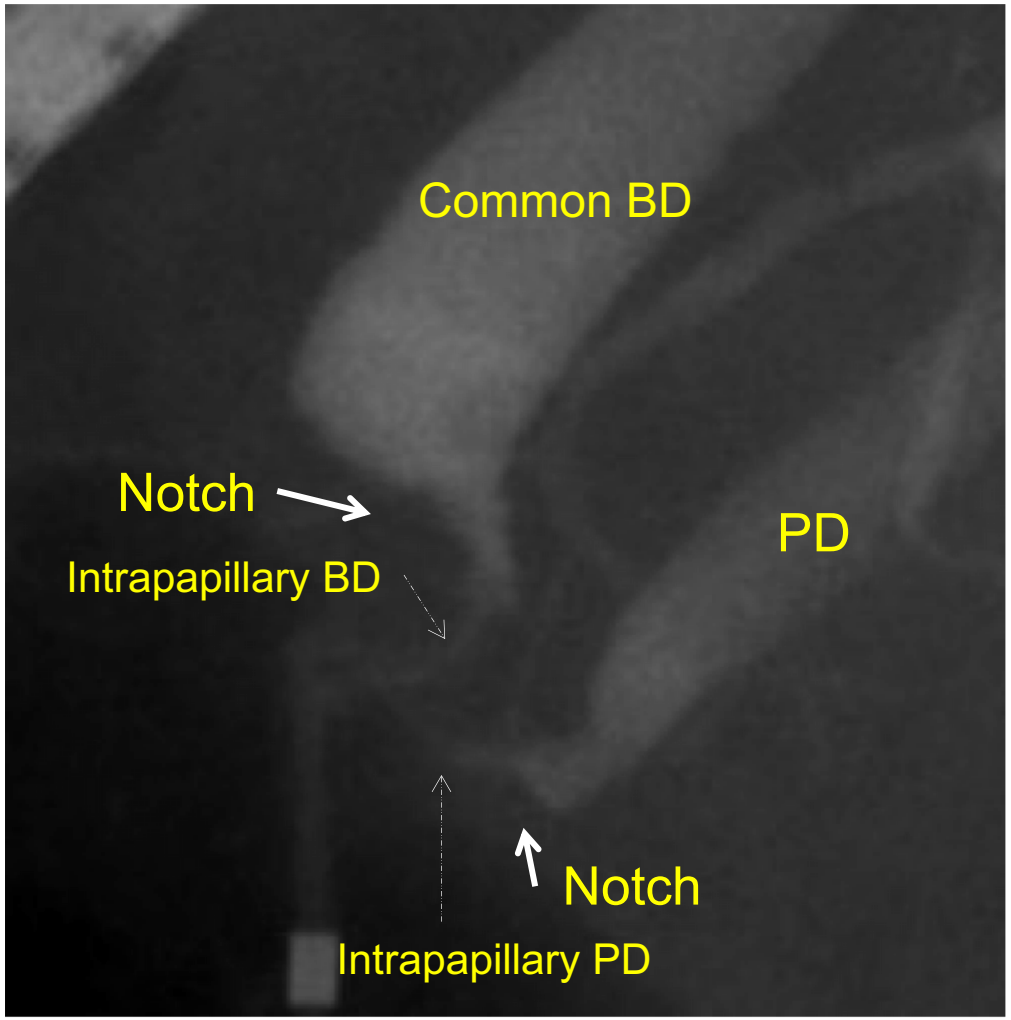
Relaxation phase of Oddi's sphincter



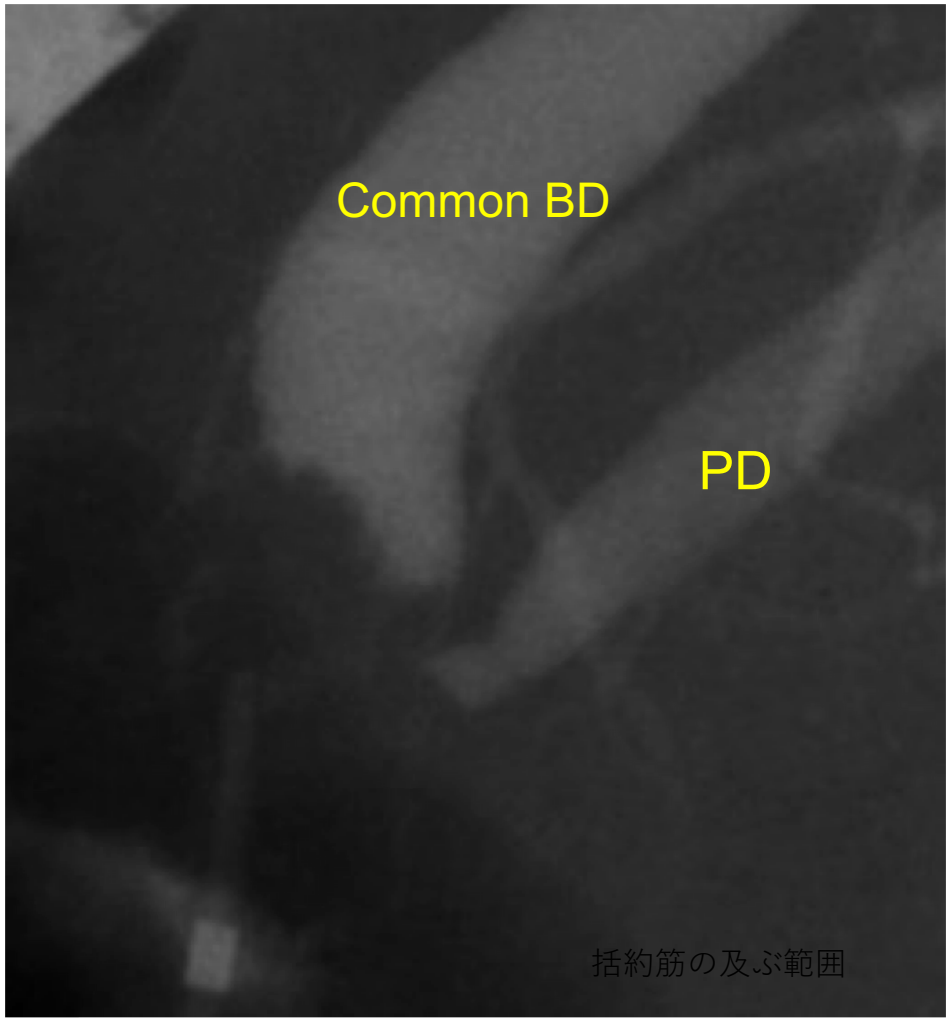
Contraction phase of Oddi's sphincter

Normal control

Because sphincter action affects both BD and PD, the notch is seen at each side of BD and PD.



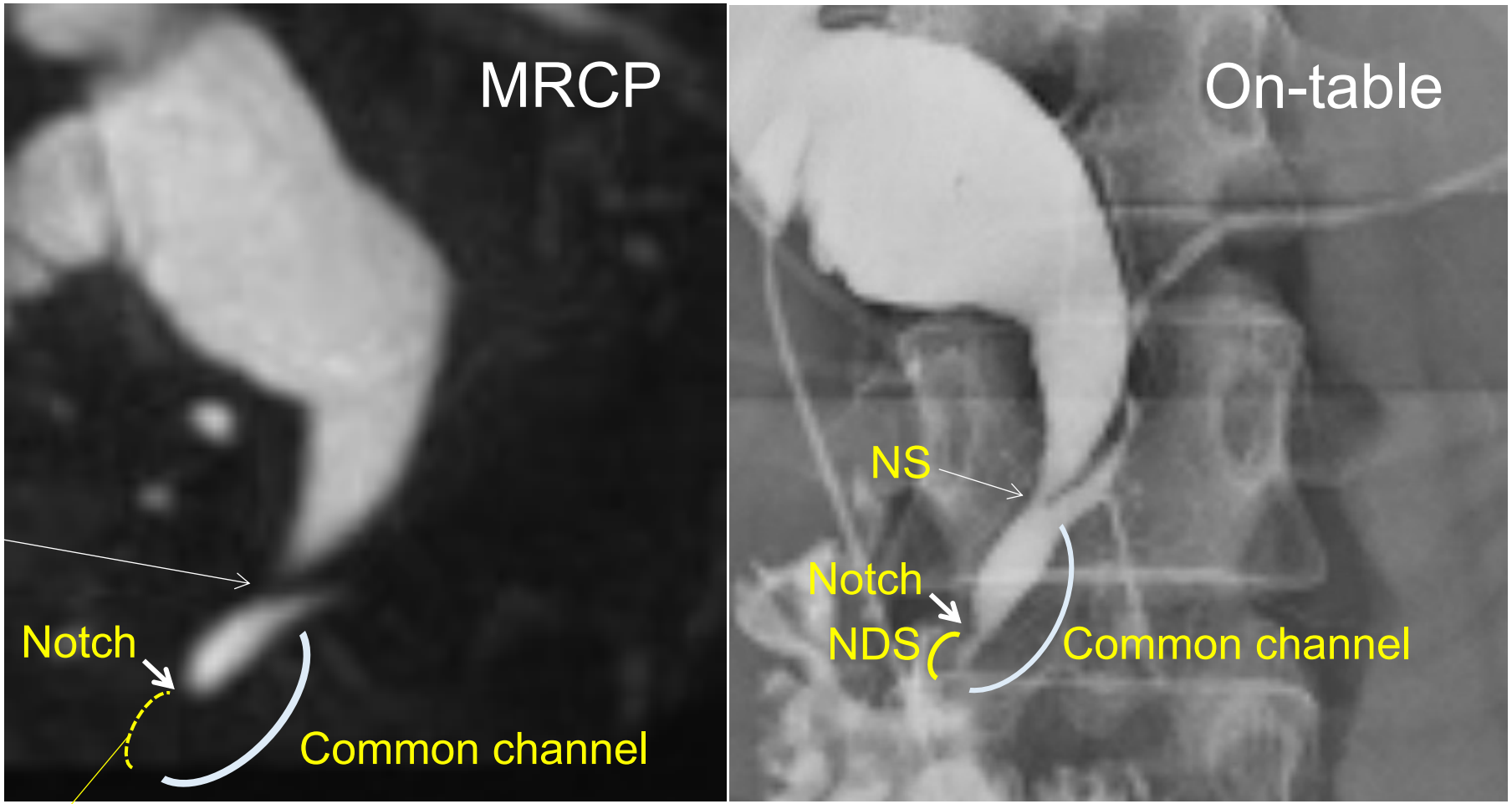
Relaxation phase
of Oddi's sphincter



Contraction phase
of Oddi's sphincter

MRCP often does not depict NDS

Narrow distal segment (NDS) is the area the sphincter affects



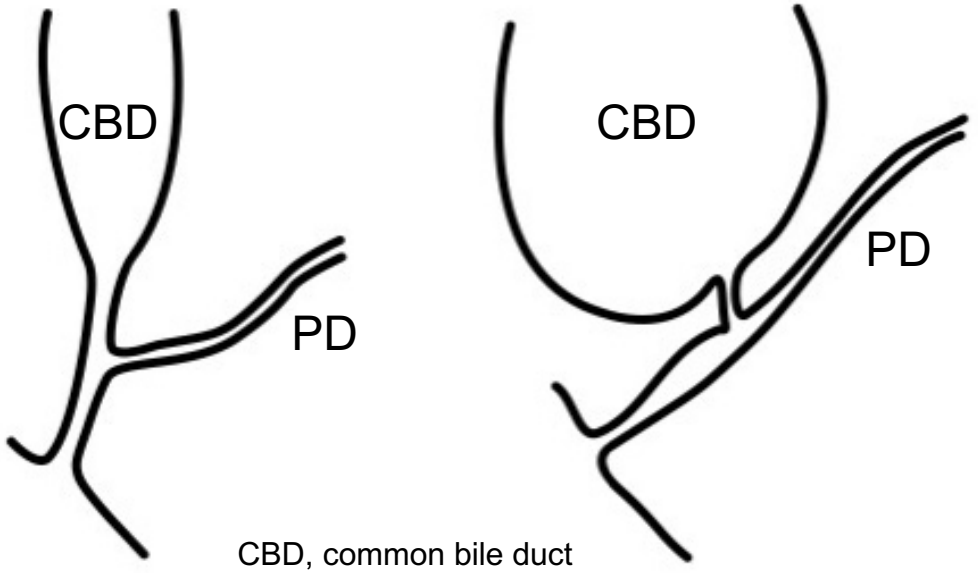
Narrow segment (NS)
Stenotic portion distal to the bile duct

NDS not depicted →MRCP may not elucidate precise length of the common channel

Old Classification of PBM

Adding
dilated common channel and patent Ap (III)
to Kimura's classification (I, II)

Kimura's classification 1976



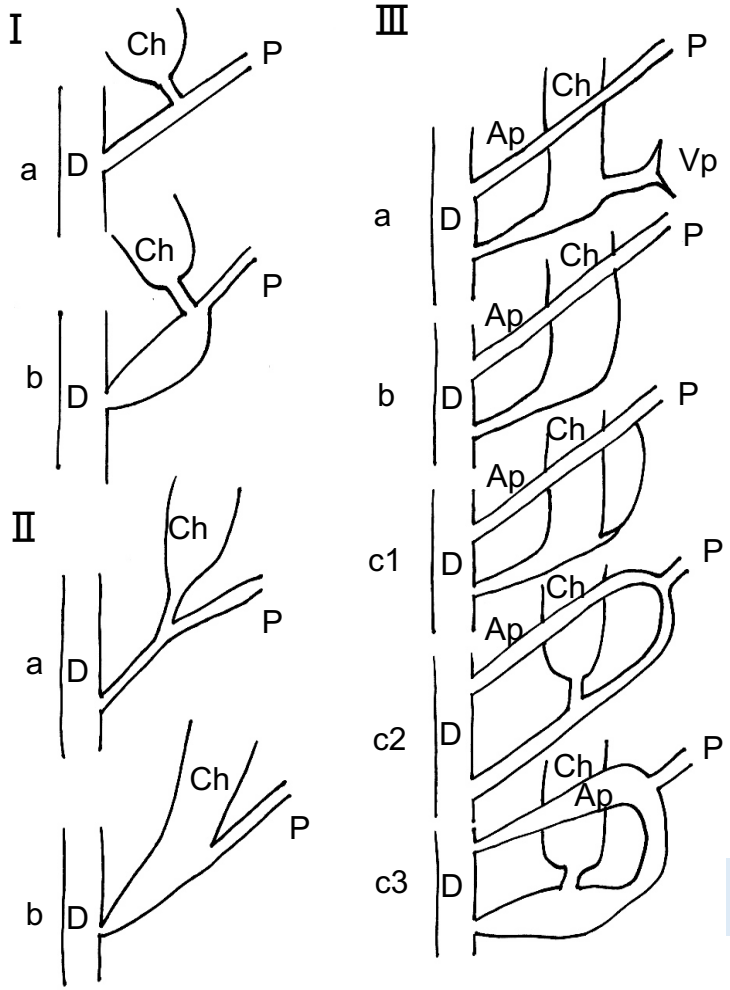
CBD, common bile duct

PC type
PD enters CBD

CP type
CBD enters PD

Remains basic even today

Komi's revised classification 1991



→ Divisum

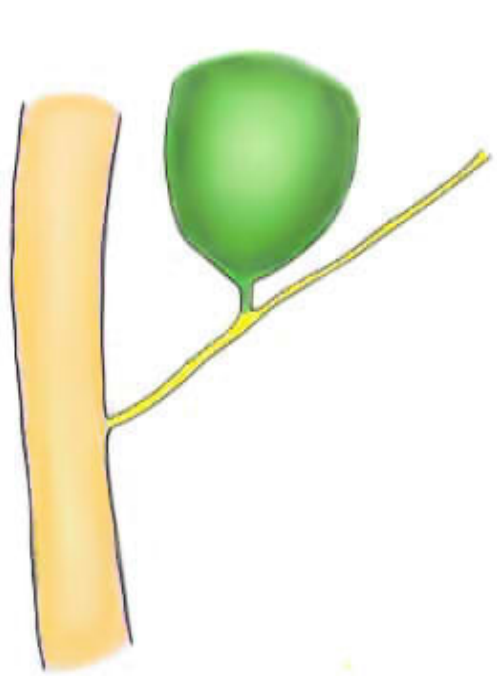
Complicated

Ch, choledochus; Ap, accessory PD

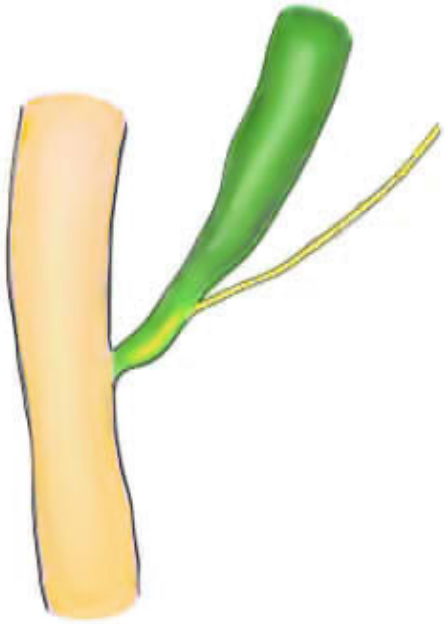
Classification by JSGPBM

J Hepatobiliary Pancreat Sci. 2017 Aug;24(8):449–455

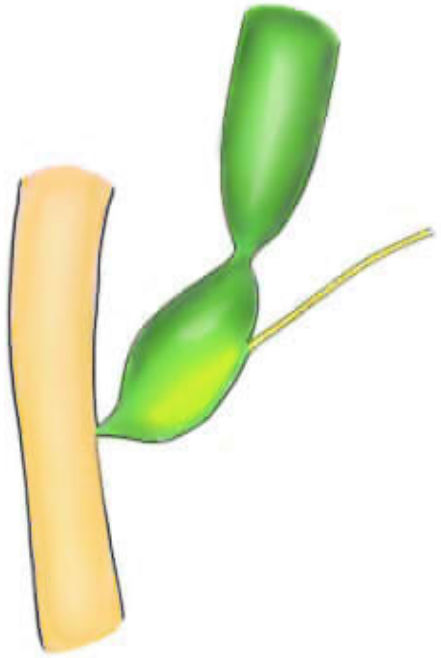
Based on the existence of narrow segment and dilatation of common channel



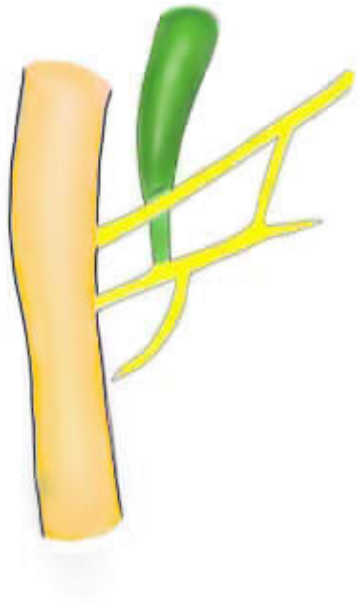
Type A
Stenotic type



Type B
Non-stenotic type



Type C
Dilated channel type

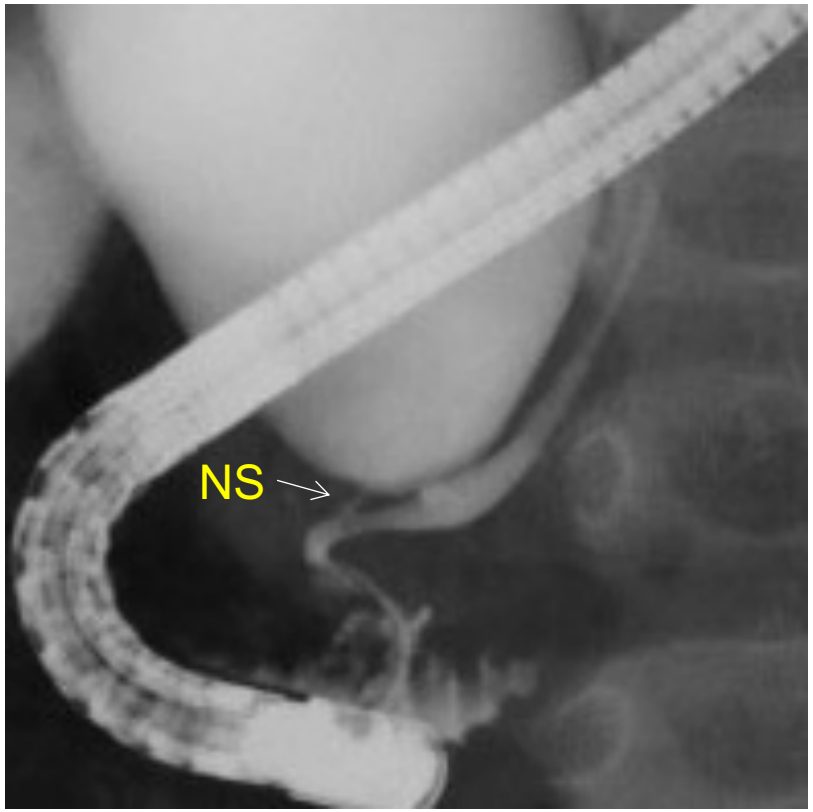


Type D
Complex type

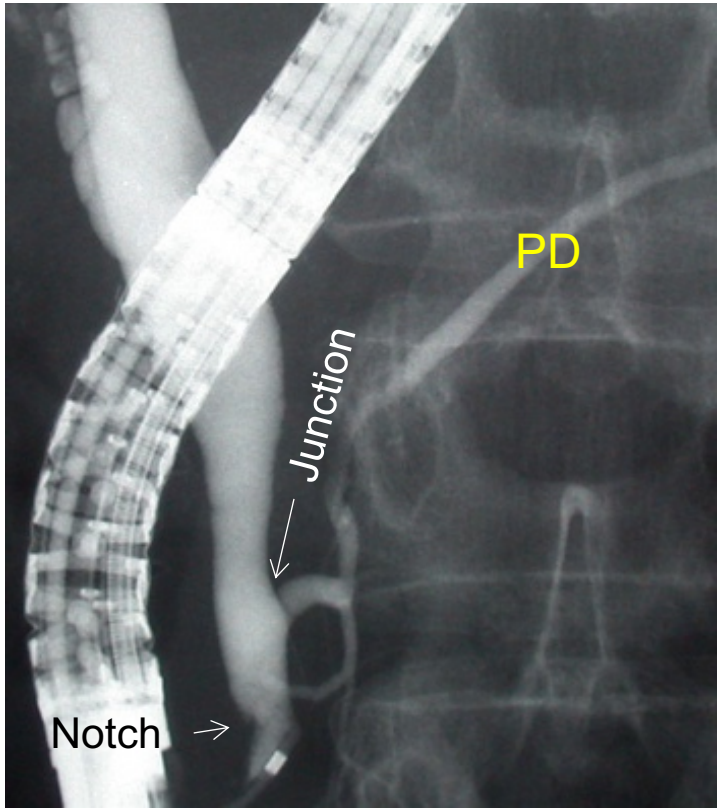
This classification is
indifferent to existence of accessory PD (Santorini's duct)

These are the only exceptions that are not applied for the left three types.
These are often associated with annular pancreas and pancreas divisum

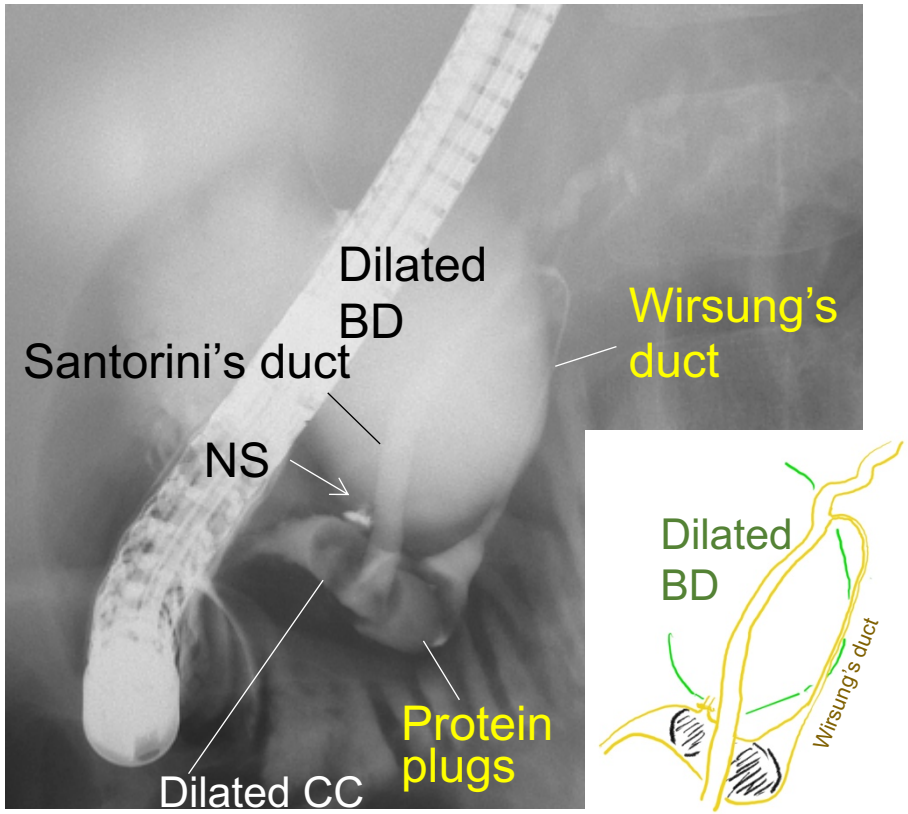
Classification by JSGPBM



Type A
Stenotic type



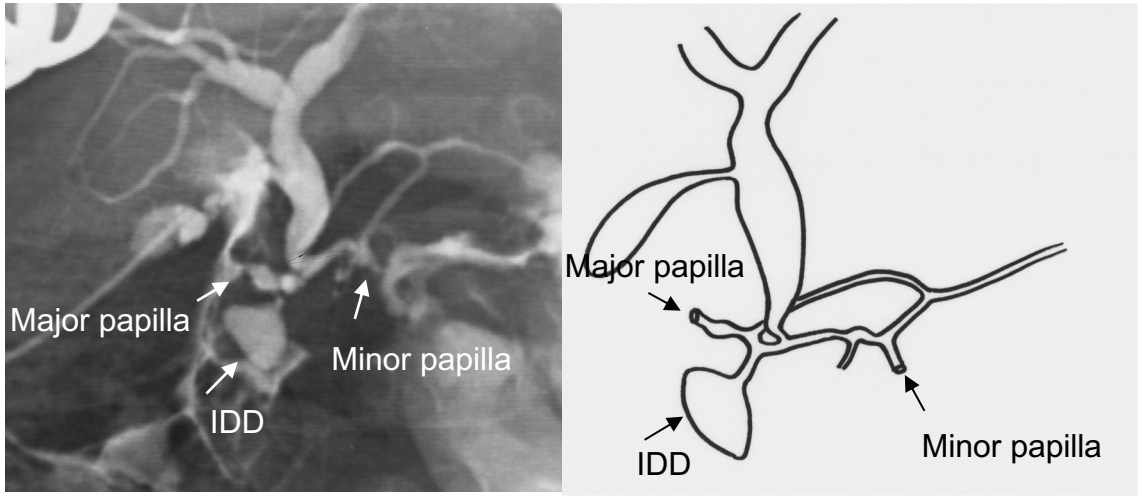
Type B
Non-stenotic type

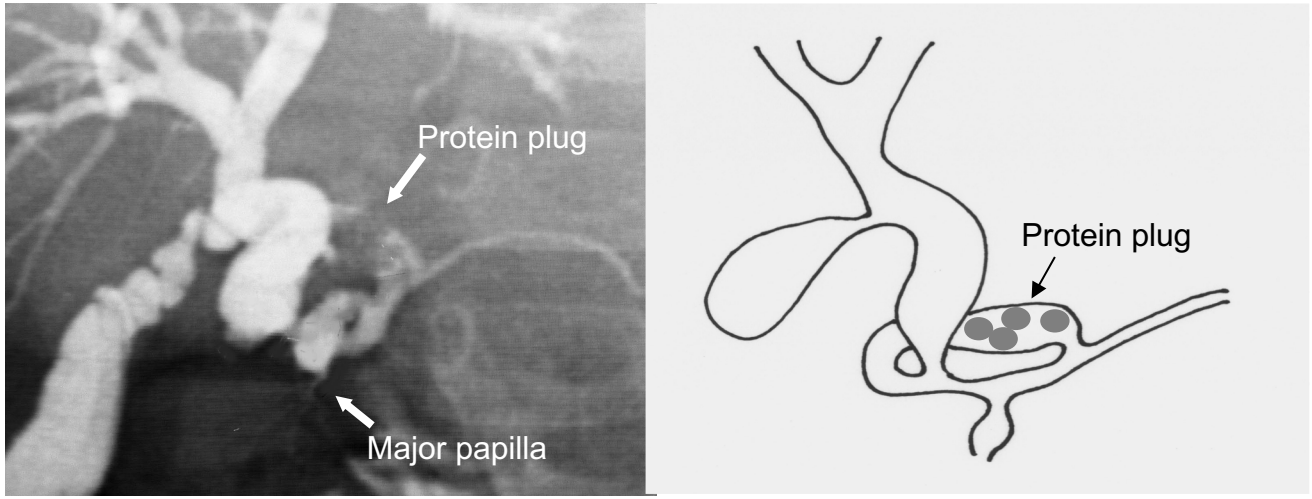


Type C
Dilated channel type

Type D: Complex type

This type was created only for exceptions, and not cases that have an accessory PD





IDD, intraluminal duodenal diverticulum

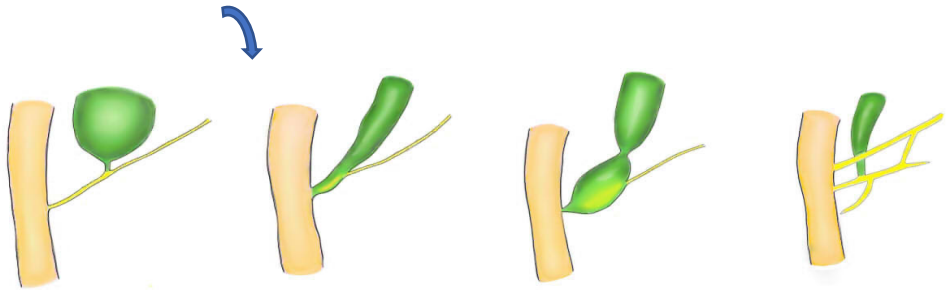
A 1-year-old girl:
Annular pancreas and intraluminal duodenal diverticulum

A 4-year-old boy:
Annular pancreas

JSGPBM classification reflects clinical characteristics in children

Urushihara N, Hamada Y, Kamisawa T, Fujii H, Koshinaga T, Morotomi Y, Saito T, Itoi T, Kaneko K, Fukuzawa H, Ando H.
Classification of pancreaticobiliary maljunction and clinical features in children.
J Hepatobiliary Pancreat Sci. 2017 Aug;24(8):449-455. doi: 10.1002/jhbp.485. Epub 2017 Jul 31. PMID: 28639336.

317 pediatric cases



Type A Stenotic type	Type B Non-stenotic type	Type C Dilated channel type	Type D Complex type
131	76	98	12

Clinical characteristics	Type	The other types	P
Younger（avg. age）	A 1.8 y	3.2 - 4.6 y	＜0.01
Cystic dilated BD	A 89%	24% - 30%	＜0.01
Non-dilated type	B 22％	0% - 17%	＜0.01
Hyperamylasemia	B 86% C 73%	A 38% D 67%	＜0.01
Acute pancreatitis (7% in all cases)	A 5%, B 7%, **C 11%,** D 8%		NS
Biliary perforation	**C 14%**	0% - 3%	＜0.01
Protein plugs	**C 56%**	22% - 40%	＜0.01

This classification is not suitable for adults in whom patency of Santorini's duct is associated with biliary cancers

Yoshimoto K, Kamisawa T, Kikuyama M, Kuruma S, Chiba K, Igarashi Y. Classification of pancreaticobiliary maljunction and its clinical features in adults. *J Hepatobiliary Pancreat Sci.* 2019 Dec;26(12):541-547. doi: 10.1002/jhbp.691. Epub 2019 Nov 16. PMID: 31633314.

Basics

What is congenital biliary dilatation (CBD)?

First version, September 2022
2nd version, September 2024

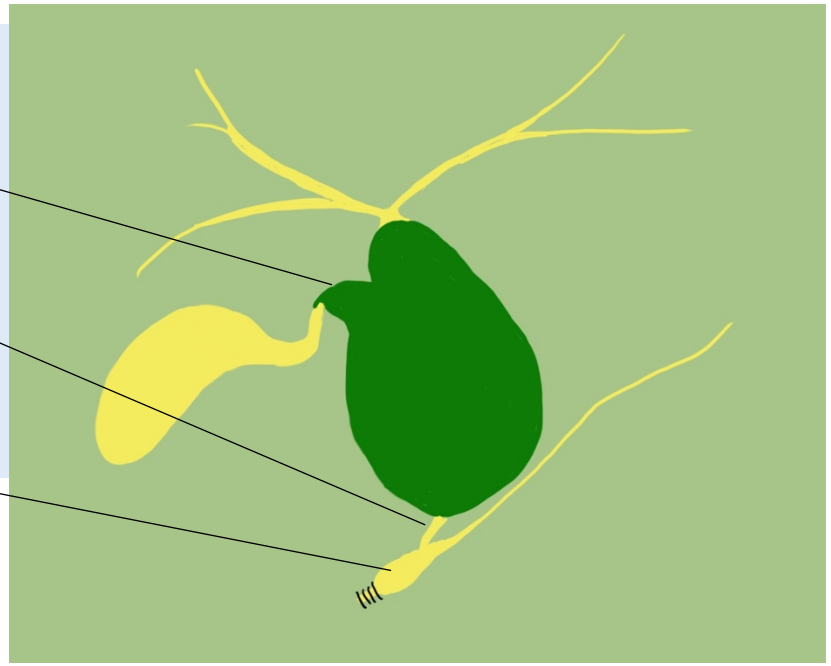
Imaging features of congenital biliary dilatation

Features that define CBD

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

Features (but not in all cases)

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



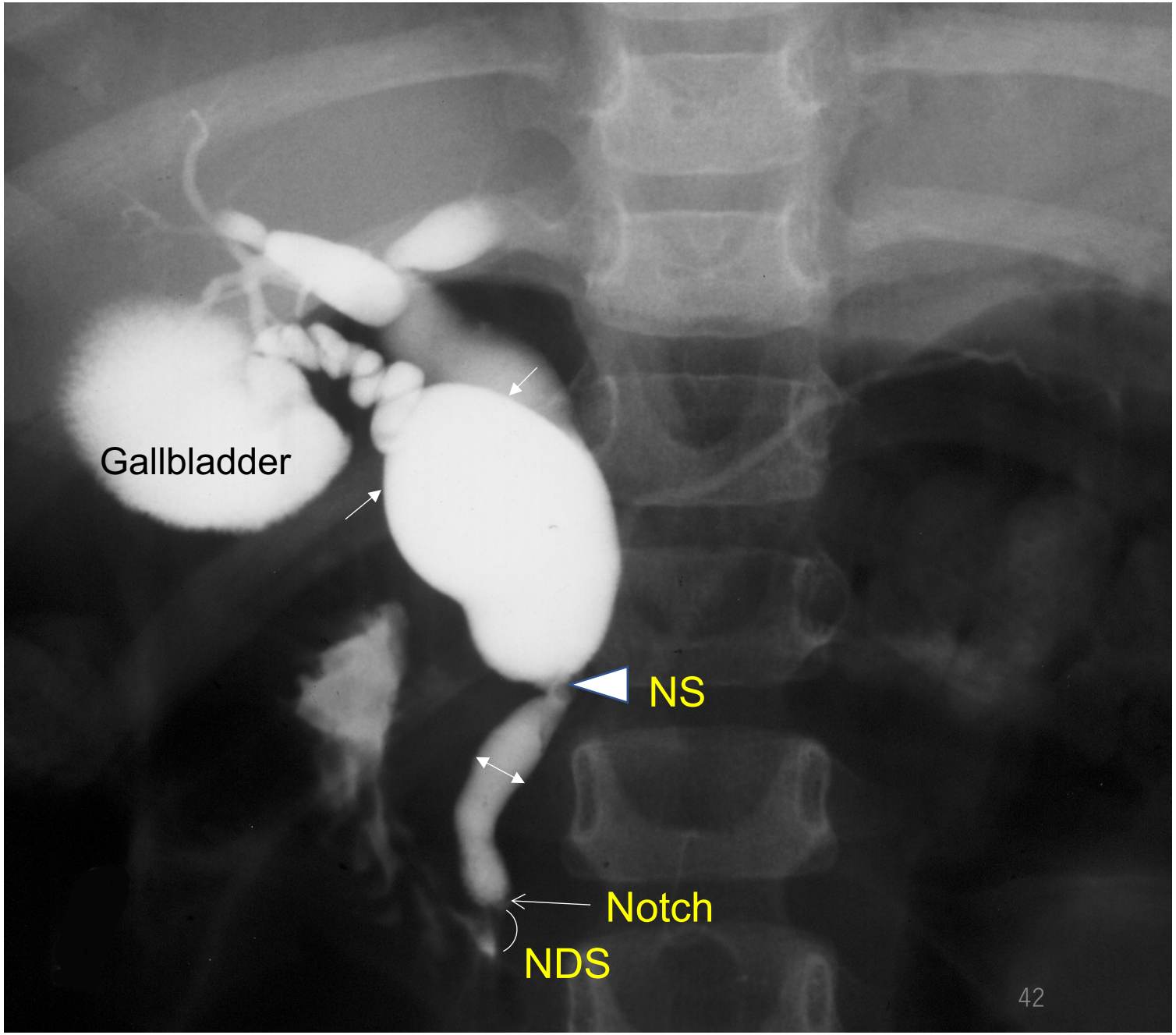
Criteria of
PBM classification by JSGPBM
See pp36–39

Case 7

Dilated cystic duct
Narrow segment
Dilated common channel

ERCP in a 4-year-old girl
Onset with abdominal pain and vomiting

- The common bile duct seems to be formed by a **dilated cystic duct** at its base (arrow). However, the gallbladder and the cystic duct at its gallbladder side are not dilated (i.e., localized dilatation).
- The lower part of the cystic-dilated BD abruptly becomes thin (arrowhead) and joins the PD. The thin portion is called the **narrow segment (NS).**
  c.f. The narrow distal segment (NDS) denotes a thin portion affected by the sphincter function.
- **Dilated common channel (two-way arrow)** is often observed.
- Type C (dilated channel type) PBM

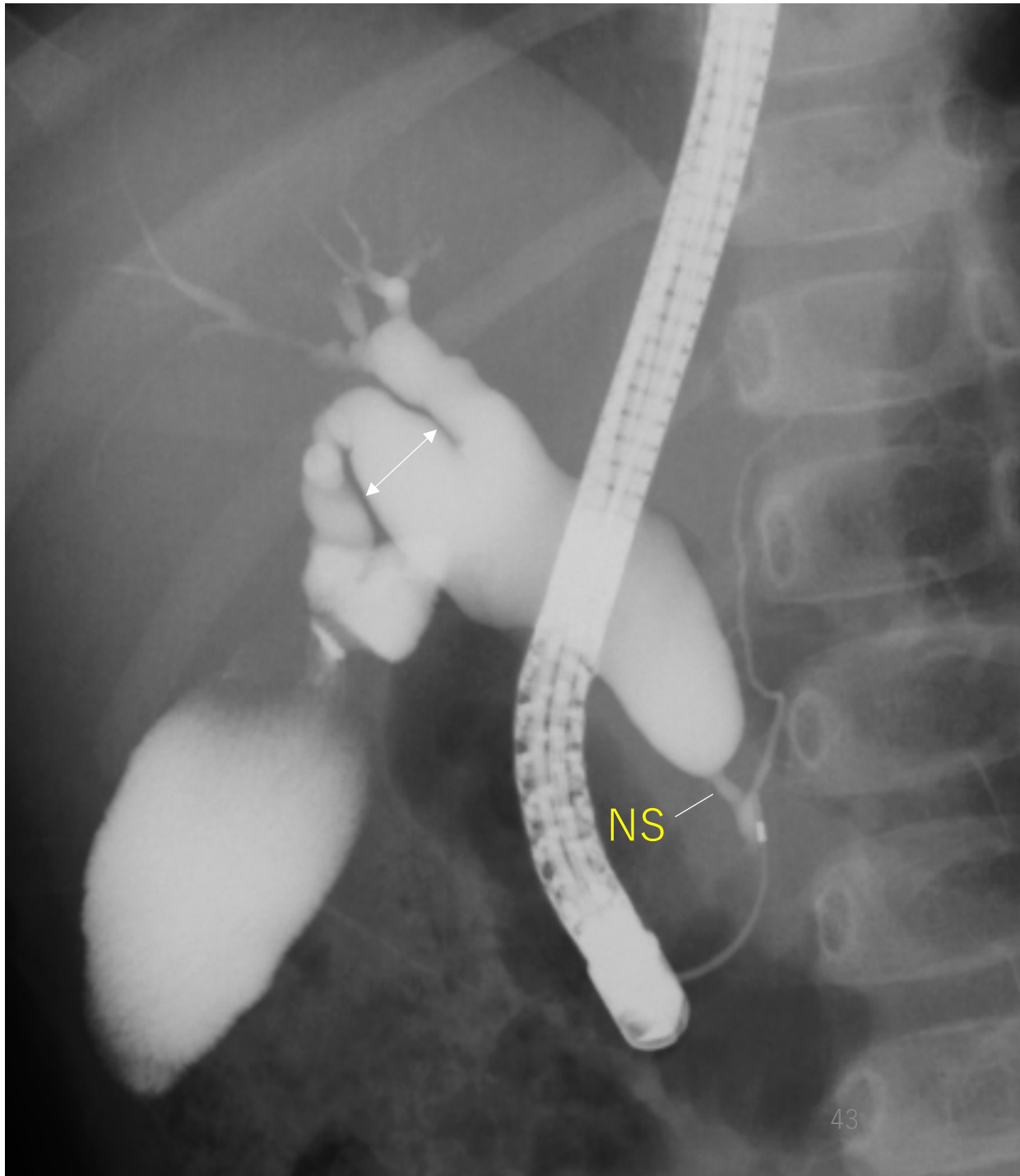


Case 8

Dilated cystic duct & long narrow segment

ERCP in a 3-year-old girl
Onset with abdominal pain, vomiting, and elevated levels of serum hepatobiliary enzymes

- **The base of the cystic duct (arrow) is more dilated** than the common hepatic duct.
- **The NS** is long.
- The common channel is not seen on this ERCP.
- Type A (stenotic type) PBM



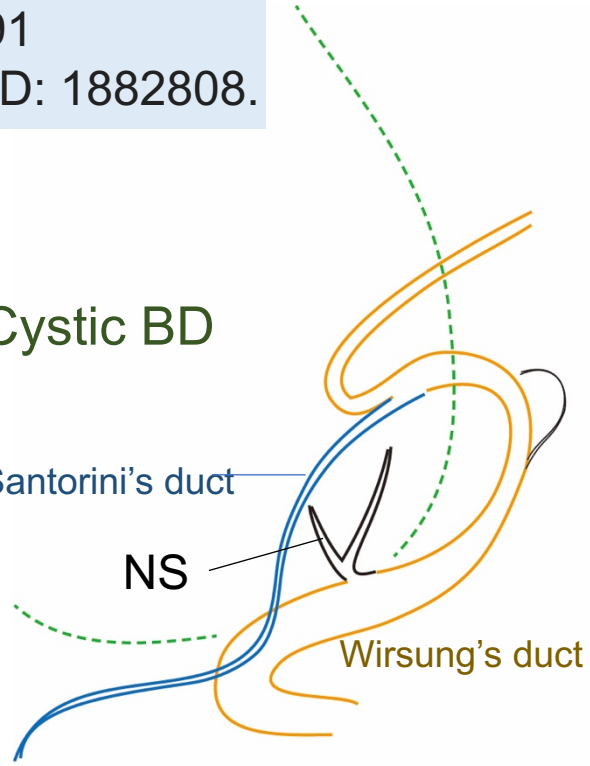
NS is a branch of the PD

Histopathological demonstration

Suda K, Matsumoto Y, Miyano T. Narrow duct segment distal to choledochal cyst. *Am J Gastroenterol*. 1991 Sep;86(9):1259-63. PMID: 1882808.

Case 9

ERCP in a 1-year-old girl
Vomiting and elevated pancreatic enzymes



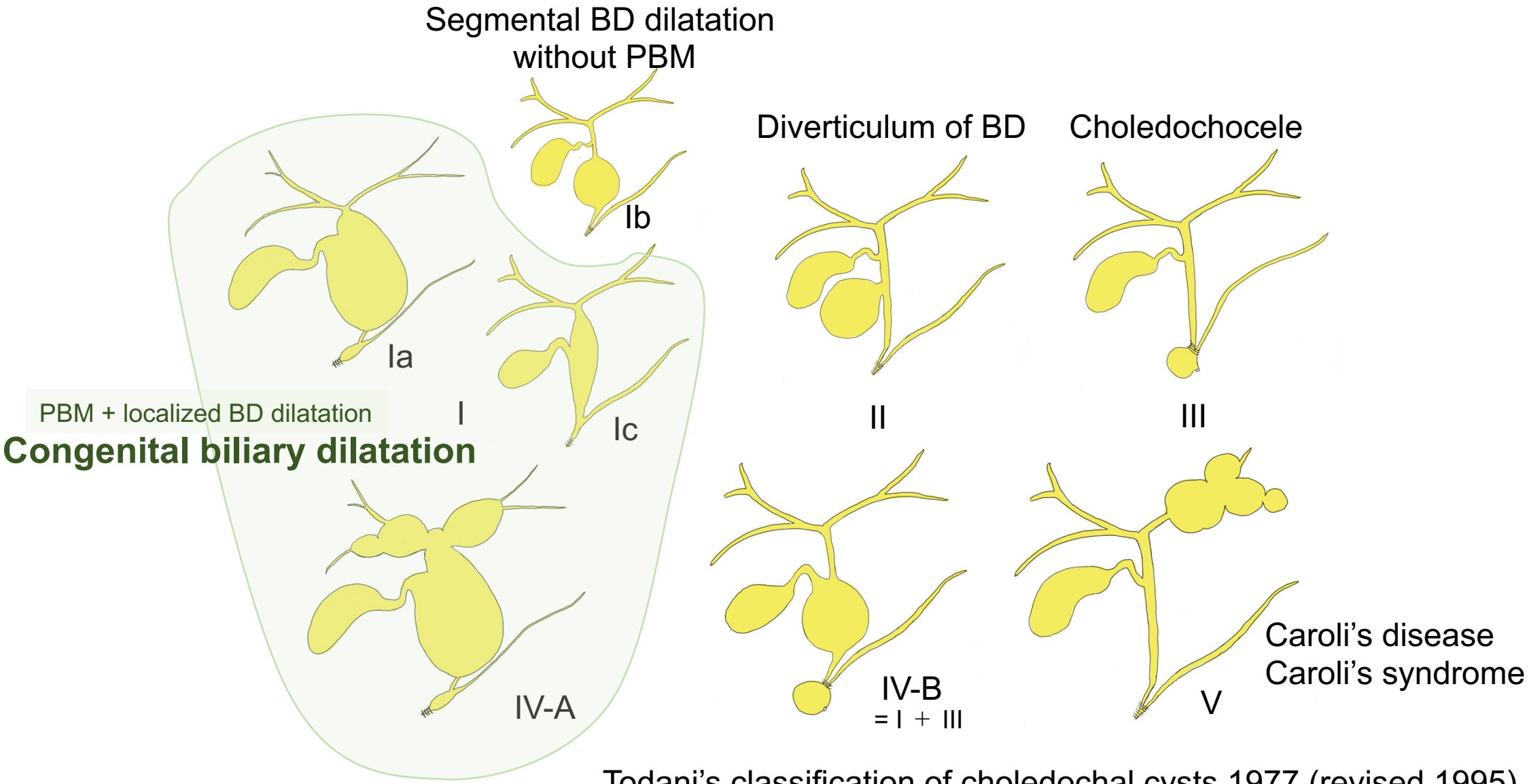
Radiological demonstration

NS is one branch of the PD



Matsumoto's theory of PBM embryogenesis:
PBM results from the BD misconnecting with the ventral PD or its branches (see *Advanced Section*).

The term choledochal cyst involves different entities



Todani's classification of choledochal cysts 1977 (revised 1995)

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
  - **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

Extremely rare
See
Expert Section

This statement may cause confusion regarding distinction from IV-A.
No constriction or stenosis is important.

See Advanced Section, pp99–113

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**.
  - **Type IVB** refers to multiple dilatations confined only to the extrahepatic bile duct.

Extremely rare

Basics

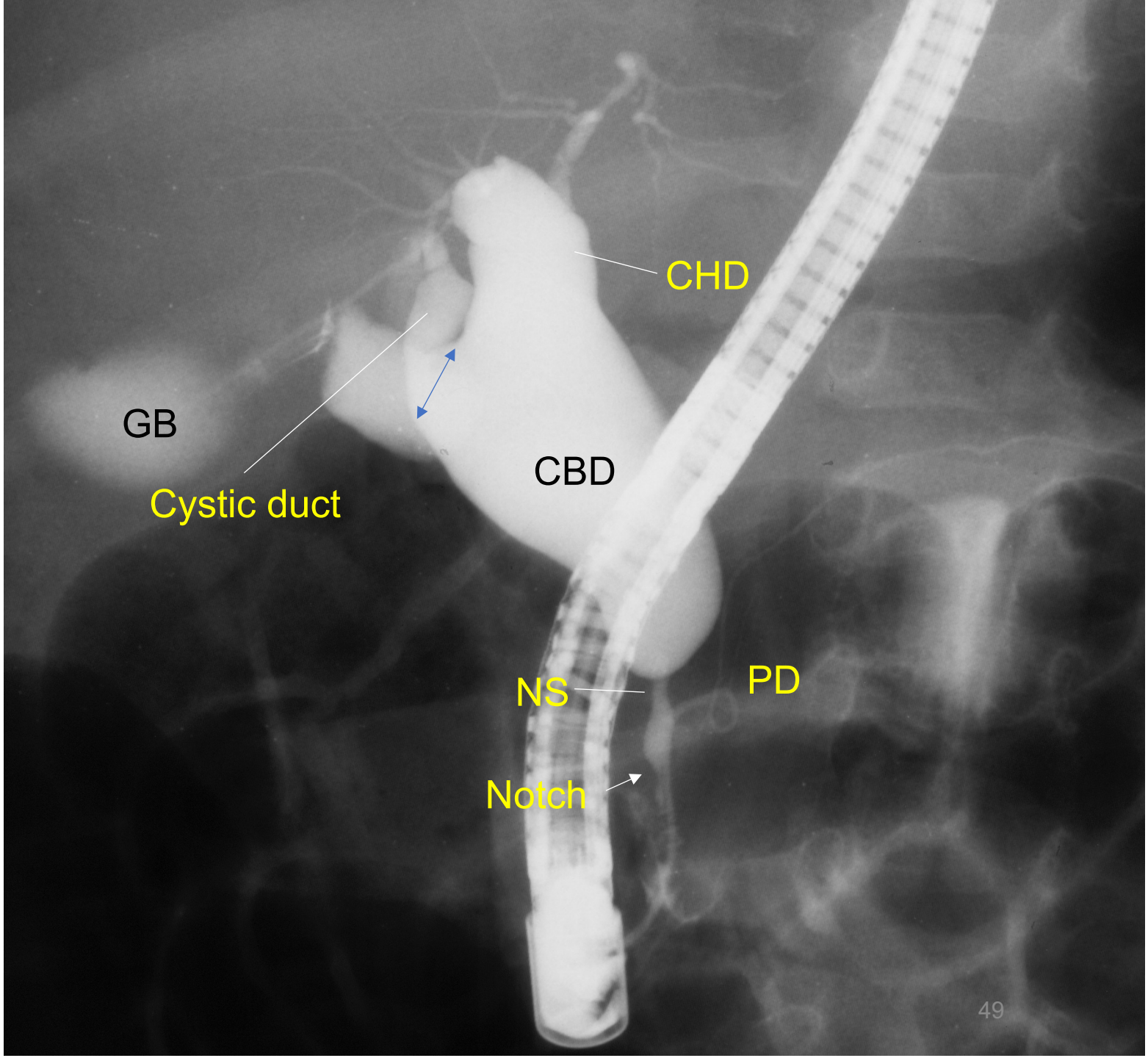
Typical congenital biliary dilatation

First version, September 2022

Case 10 Typical Ia: Pediatric case

ERCP in a 1-year-oid girl
Elevated serum transaminase levels during the course of a viral infection

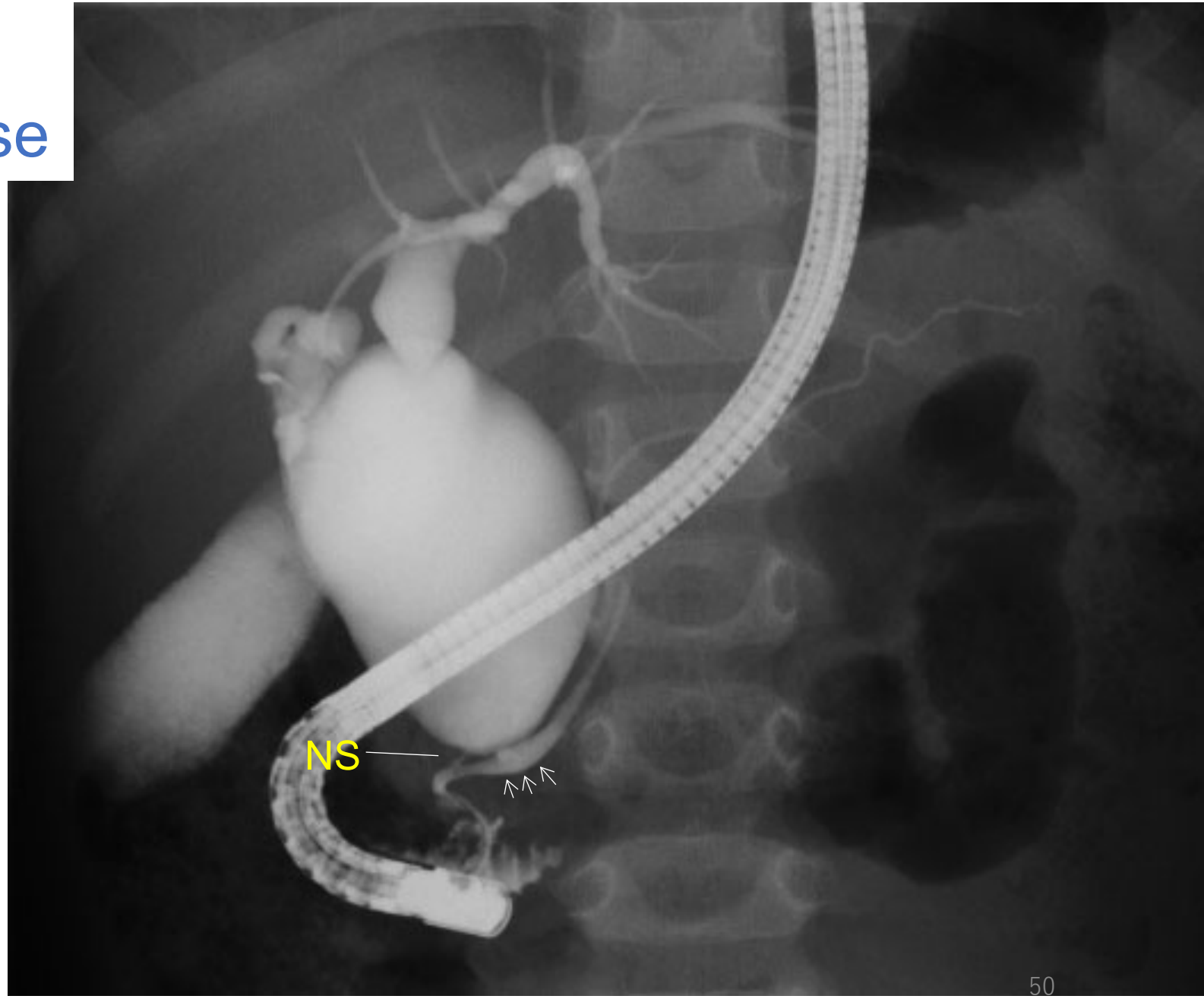
- **Cystically dilated CBD** of 21 mm in diameter
- **Dilatation involves CHD but not its upstream**
- **Cystic duct is dilated at its base** (↔) but GB is not dilated
- **Narrow segment (NS)** exists
- The common channel is not dilated
- JSGPBM classification:
  Type A, stenotic type



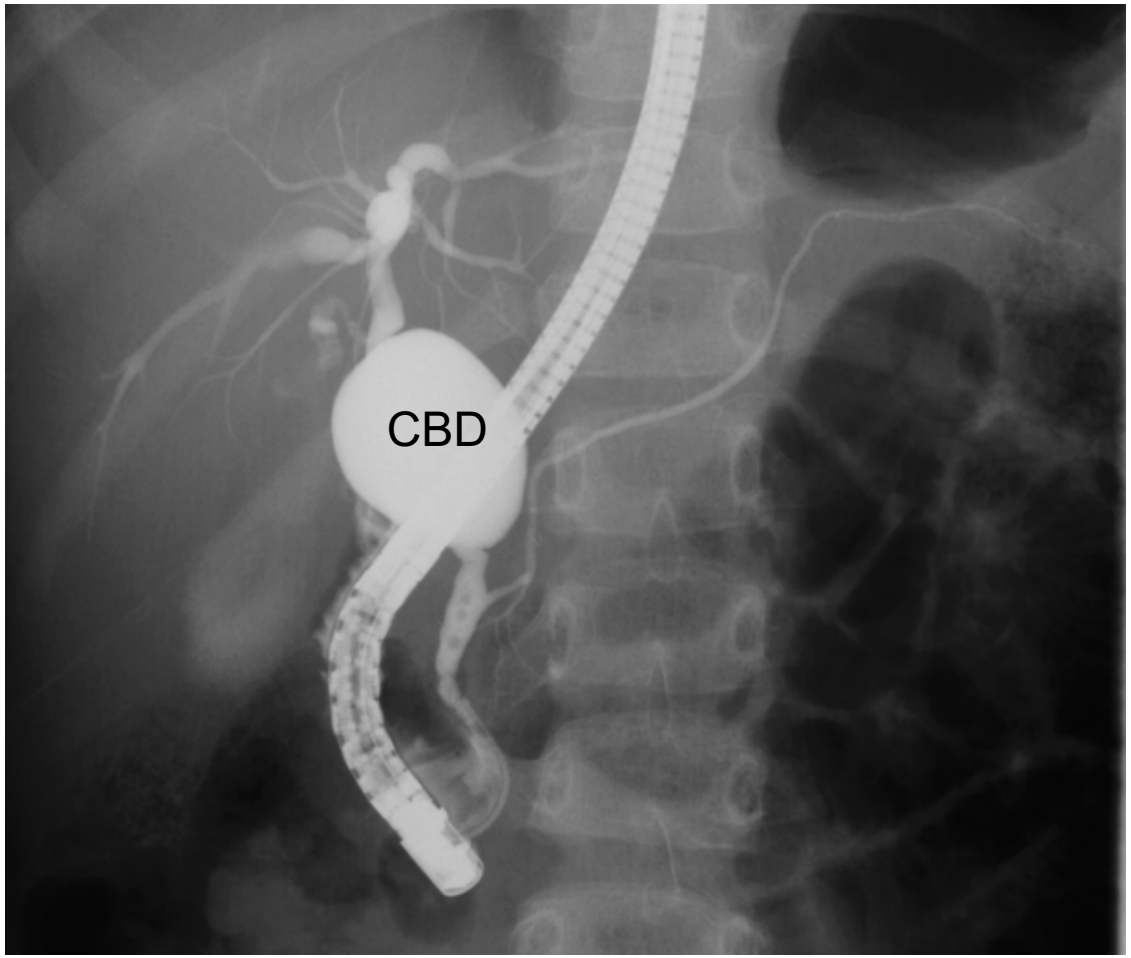
Case 11 Typical Ia: Pediatric case

ERCP in a 3-year-old girl
Abdominal pain, vomiting, and elevated serum transaminase levels

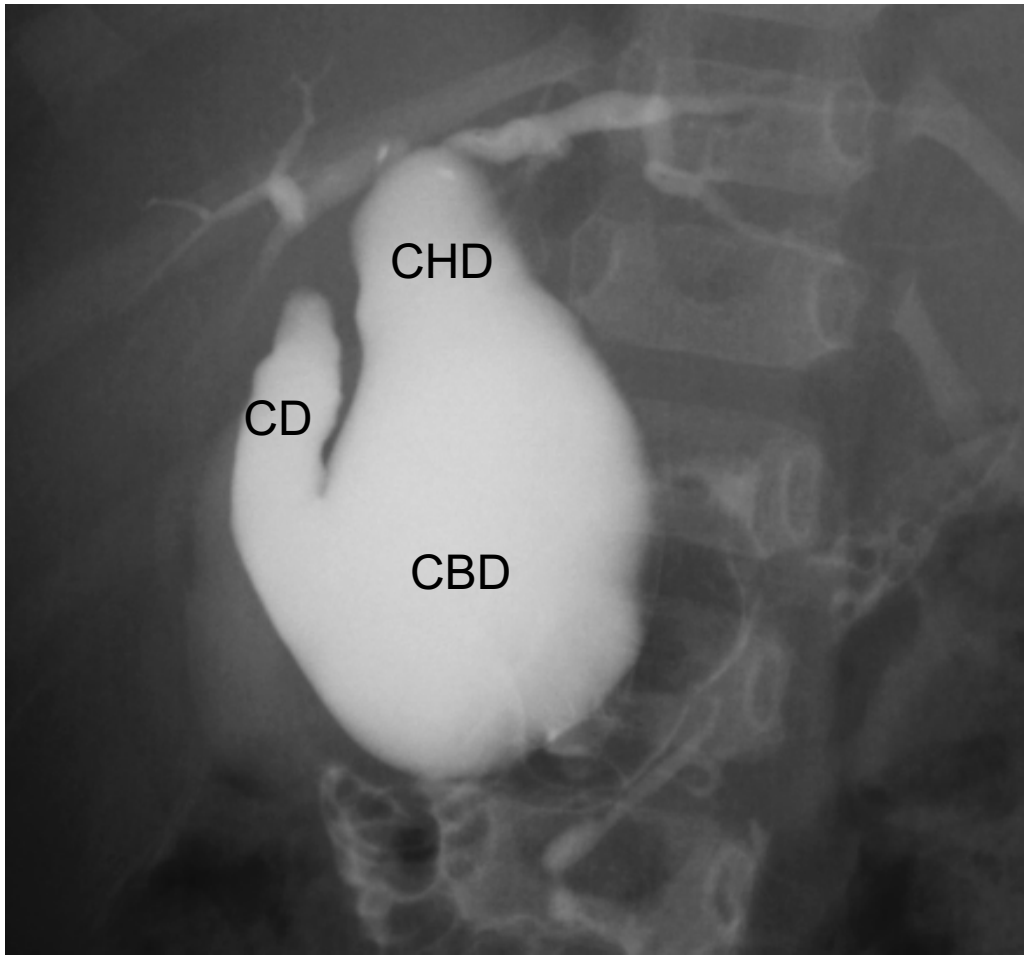
- **Cystically dilated CBD** with a diameter of 40 mm
- **Dilatation involves CHD but not its upstream processes**
- **Cystic duct is dilated at its base**, but GB is not dilated
- **NS** exists
- The common channel is not dilated
- **NS locates not at the cyst bottom but right-ventrally**
  Ventral PD attaches to the cyst wall (arrows). Care not to injure PD during dissection
- Type A, stenotic type



Localized dilatation to various extents even within type Ia pediatric cases



Dilatation limited to the common bile duct (CBD)

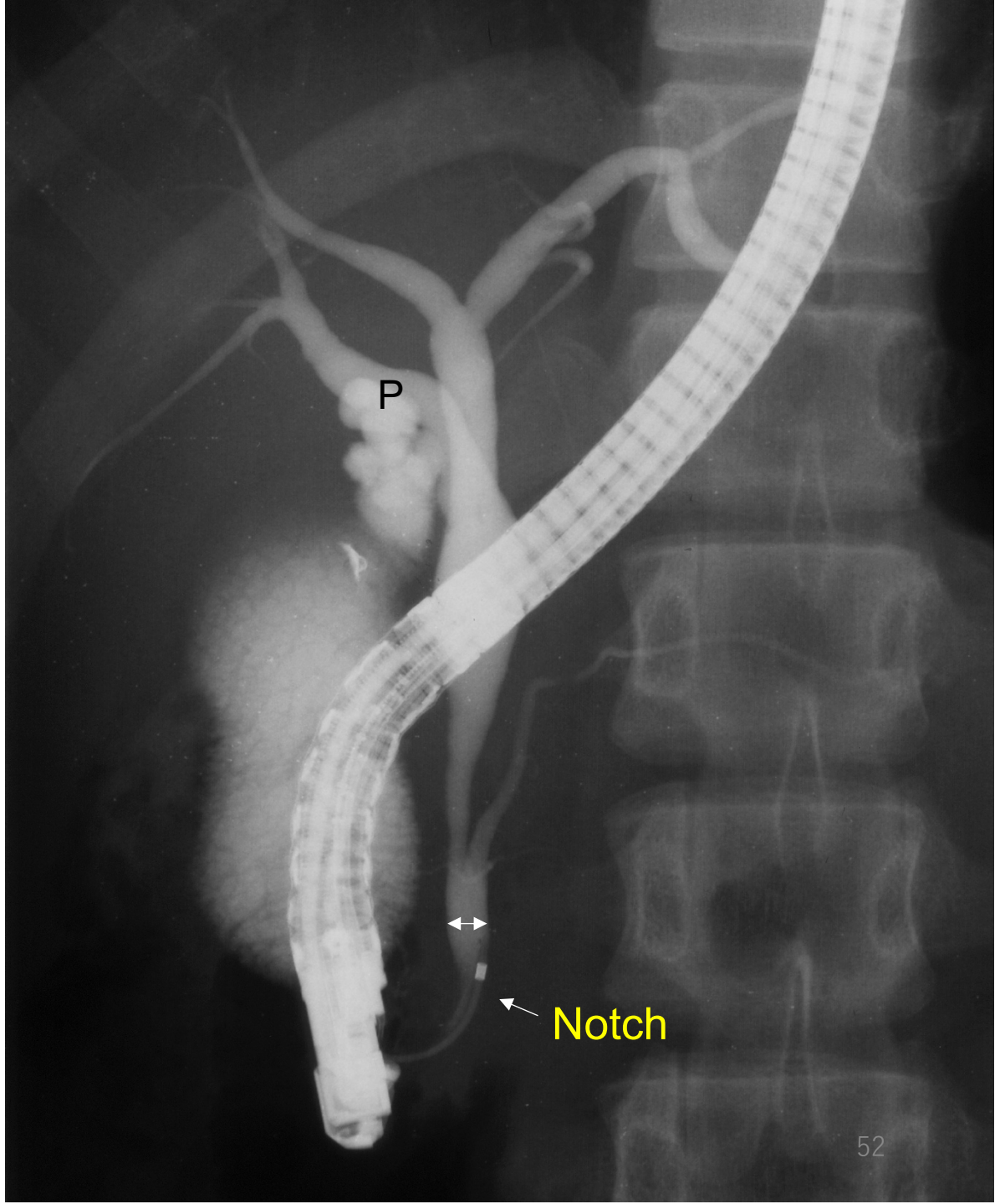


Dilatation involving CBD, CD, and CHD

Case 12 Typical Ic: Fusiform dilatation pediatric case

ERCP in a 13-year-old girl
Abdominal pain, vomiting, hyperamylasemia

- **Fusiform-dilated** CBD with a 13-mm diameter
- **Posterior branch (P) is slightly dilated, but the dilatation is smooth and continuous from CHD**, indicating not IV-A but Ic
- **Dilated common channel (↔)**
- Existence of NS is equivocal
- Type C, dilated channel type

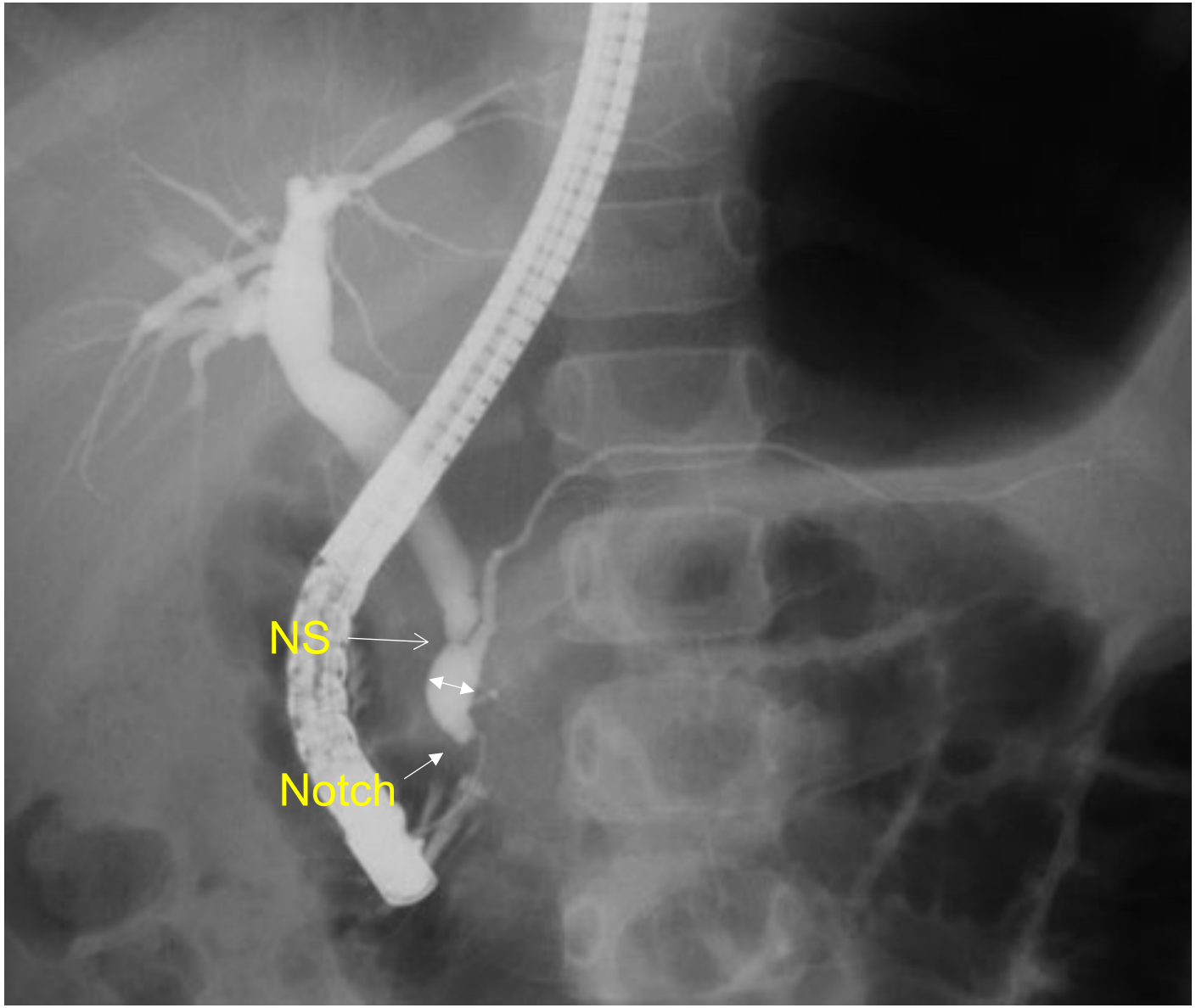


Case 13

Typical Ic: Cylindrical dilatation pediatric case

ERCP in a 5-year-old boy
Cholecystectomy at the age of 1

- **Cylindrically dilated CBD** with an 8.4-mm diameter
- **No dilatation upstream from the CHD**
- **NS** exists but is short
- **Dilated common channel (↔)**
- Type C, dilated channel type

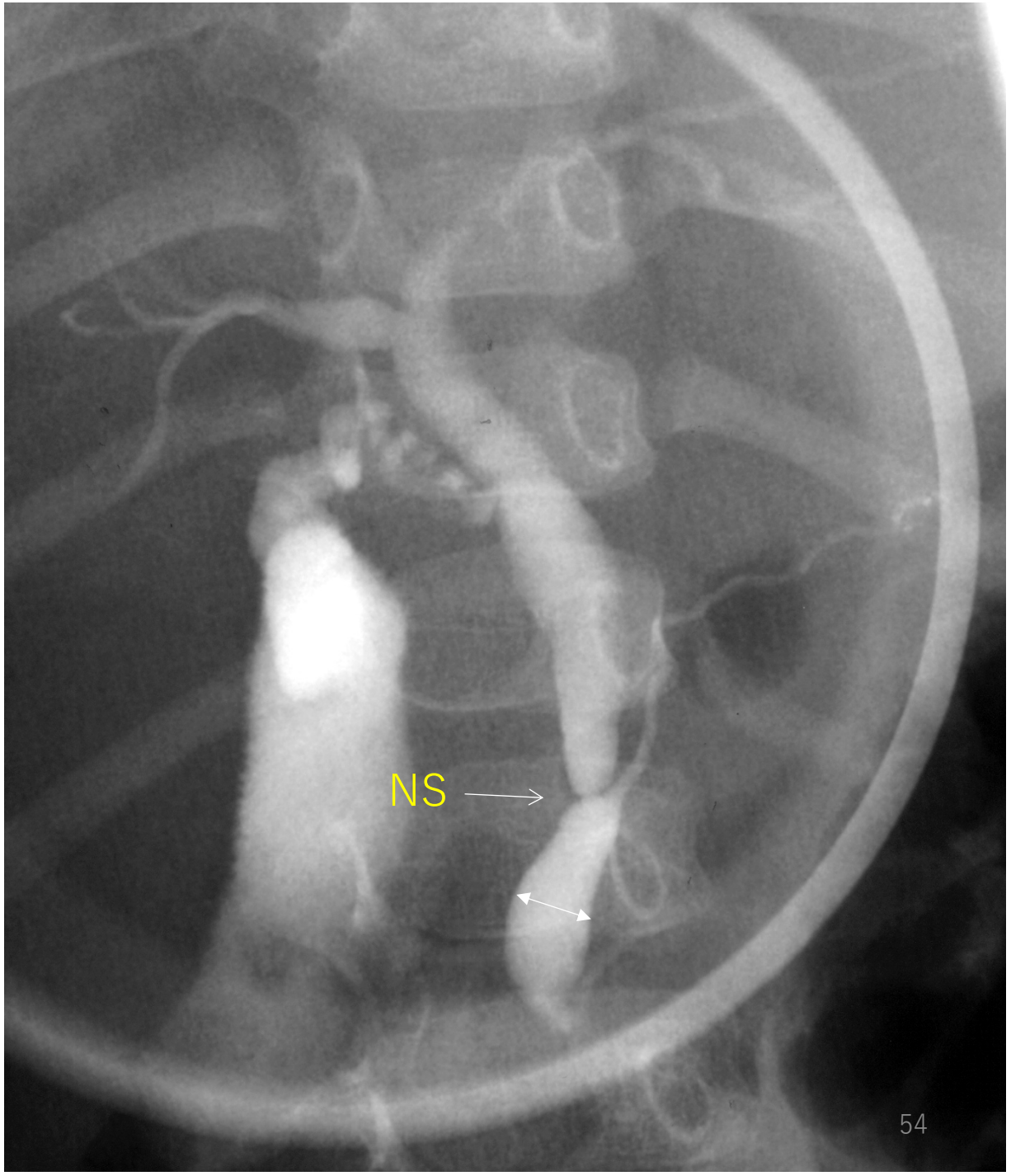


Case 14

Typical Ic: Cylindrical dilatation pediatric case

ERCP in a 2-year-old
Abdominal pain, vomiting, elevated serum hepatic and biliary enzymes
Swollen pancreas on CT (true pancreatitis)

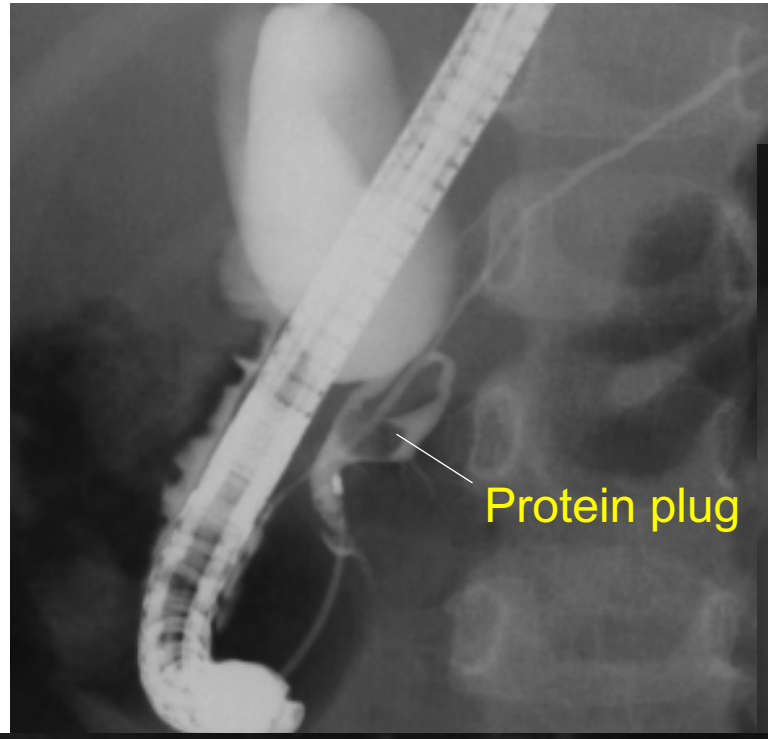
- **Cylindrically dilated CHD** with an 8.0-mm diameter
- **No dilatation upstream from the CHD**
- **NS** exists but is short
- **Dilated common channel (↔)**
- Type C, dilated channel type

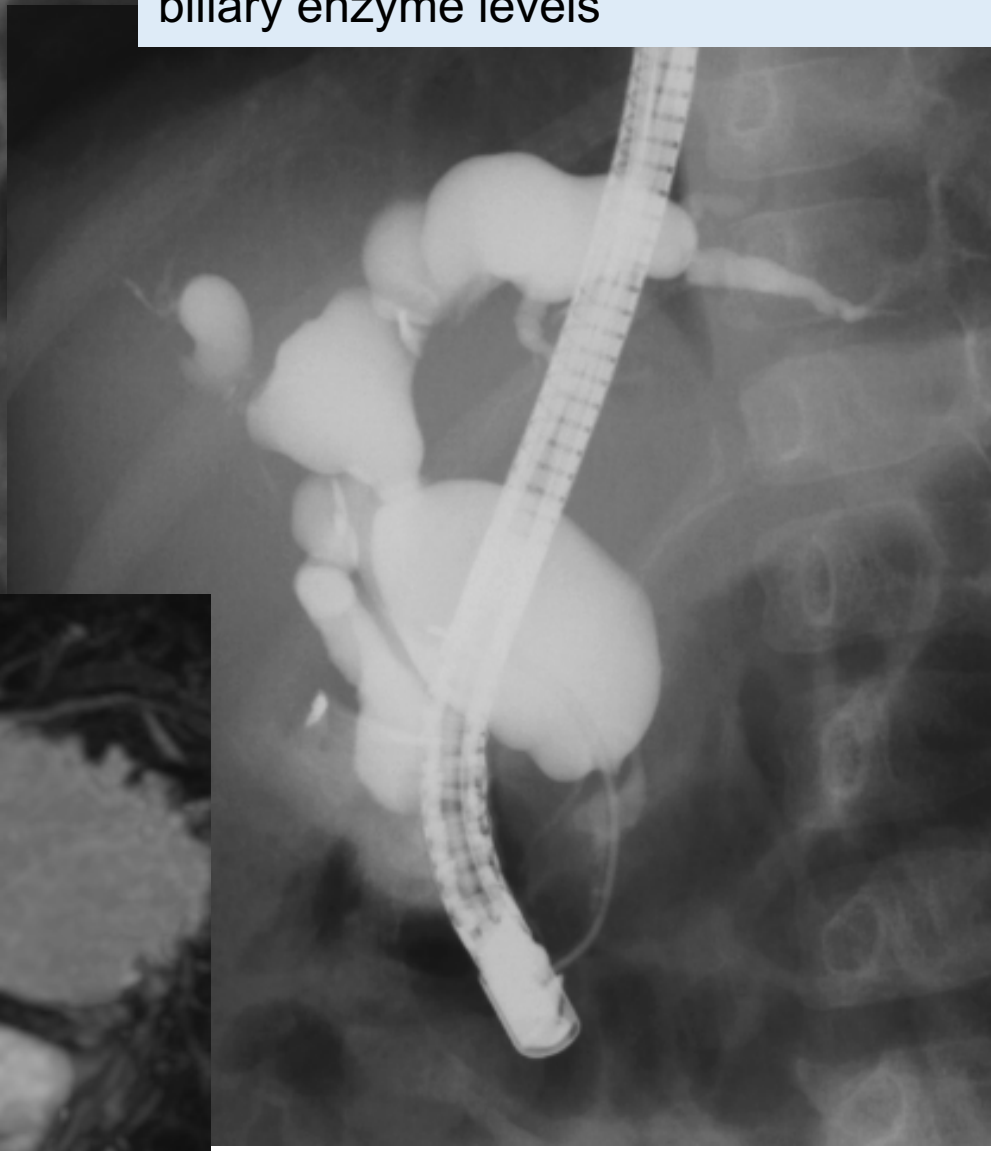


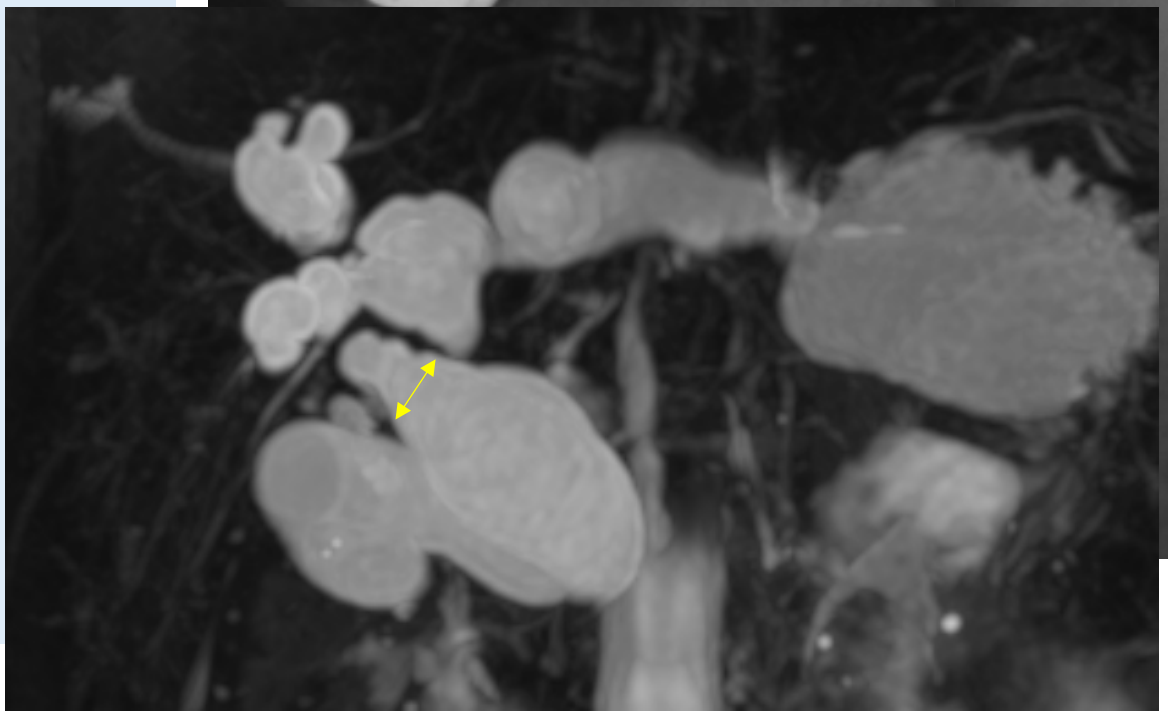
Case 15

Typical VI-A: Cystic–cystic type pediatric case

A 4-year-old girl
Abdominal pain, elevated hepatic and biliary enzyme levels







- **Cystic dilatation at both extra- and intrahepatic BDs**
- **Dilated cystic duct (CD) at the base** (↔) **Dilated common channel** contains protein plugs, **NS** exists
- Type C, dilated channel
- Intrahepatic gallstones occurred after BD resection in this case

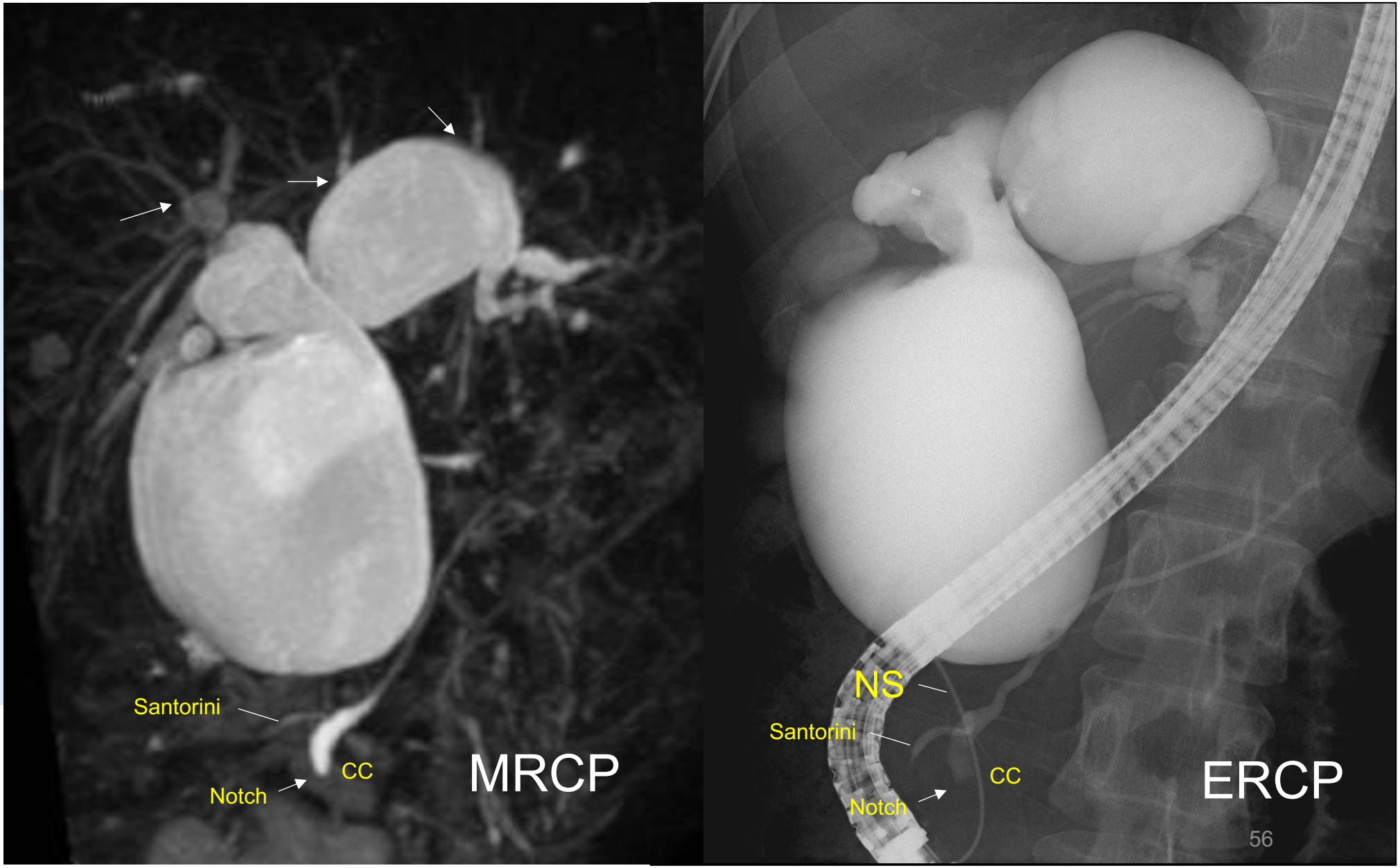
See p85.

Case 16

Typical VI-A: Cystic–cystic type, adult case

A female in her 30s. Ultrasonography detected BD dilatation on her medical check-up.

- **Cystically dilated CBD**
- **Cystically dilated BD upstream from CHD**
- Dilated BDs abruptly change to those of normal caliber (→).
- NS exists
- Type A, stenotic type



CC, common channel

Case 17

Typical IV-A: Fusiform–fusiform type pediatric case

ERCP in a girl

- **Fusiform dilatation at both extra- and intrahepatic BDs**
- **Cystic dilatation at the base of the cystic duct**
- **NS** exists
- **Dilated common channel**
- Type C, dilated channel type
- Localized dilatation: Dilated BDs abruptly change to those of normal caliber (→).



Basics

Non-dilated type of PBM

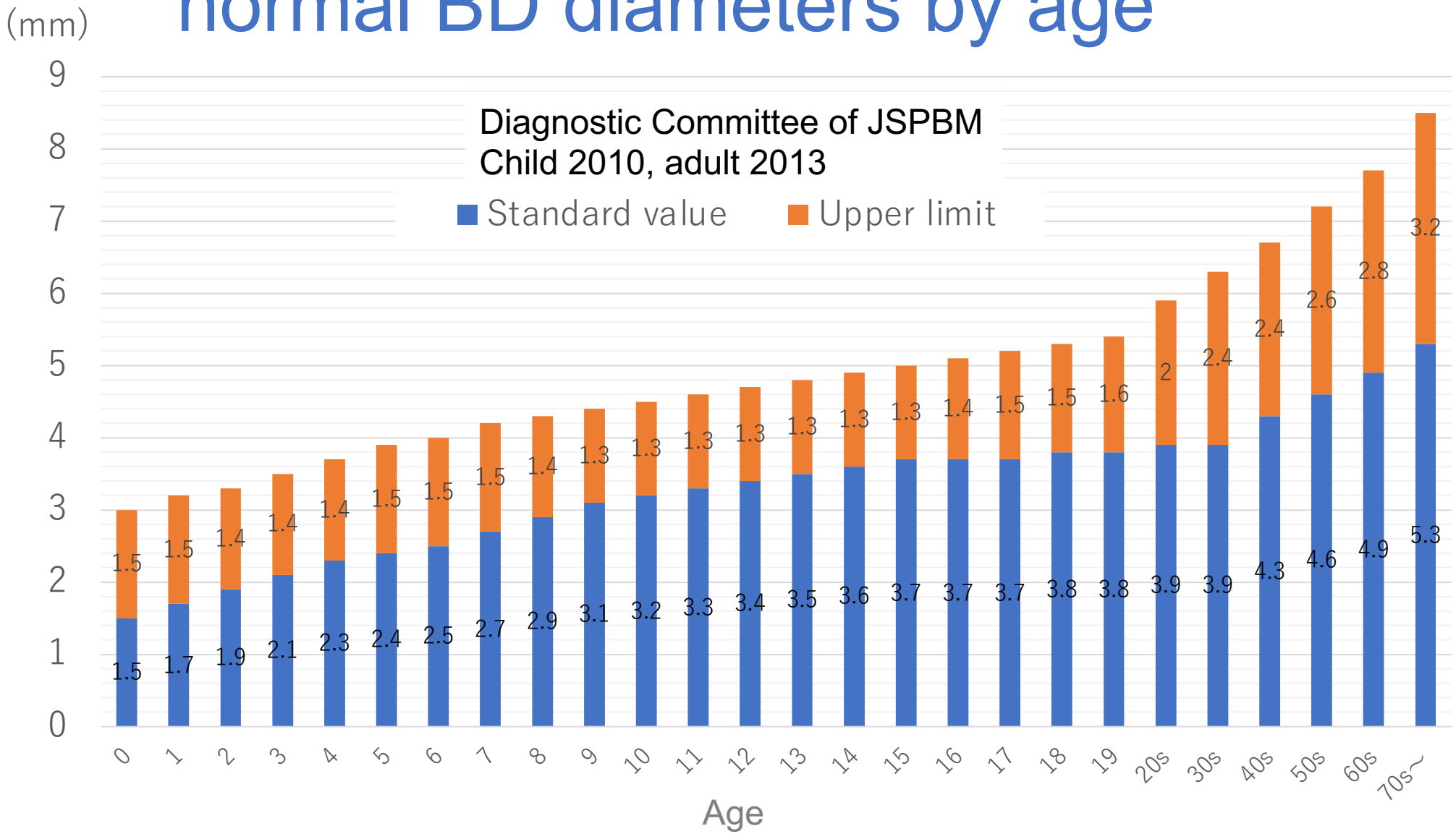
First version, September 2022
2nd version, September 2024

What is non-dilated type of PBM?

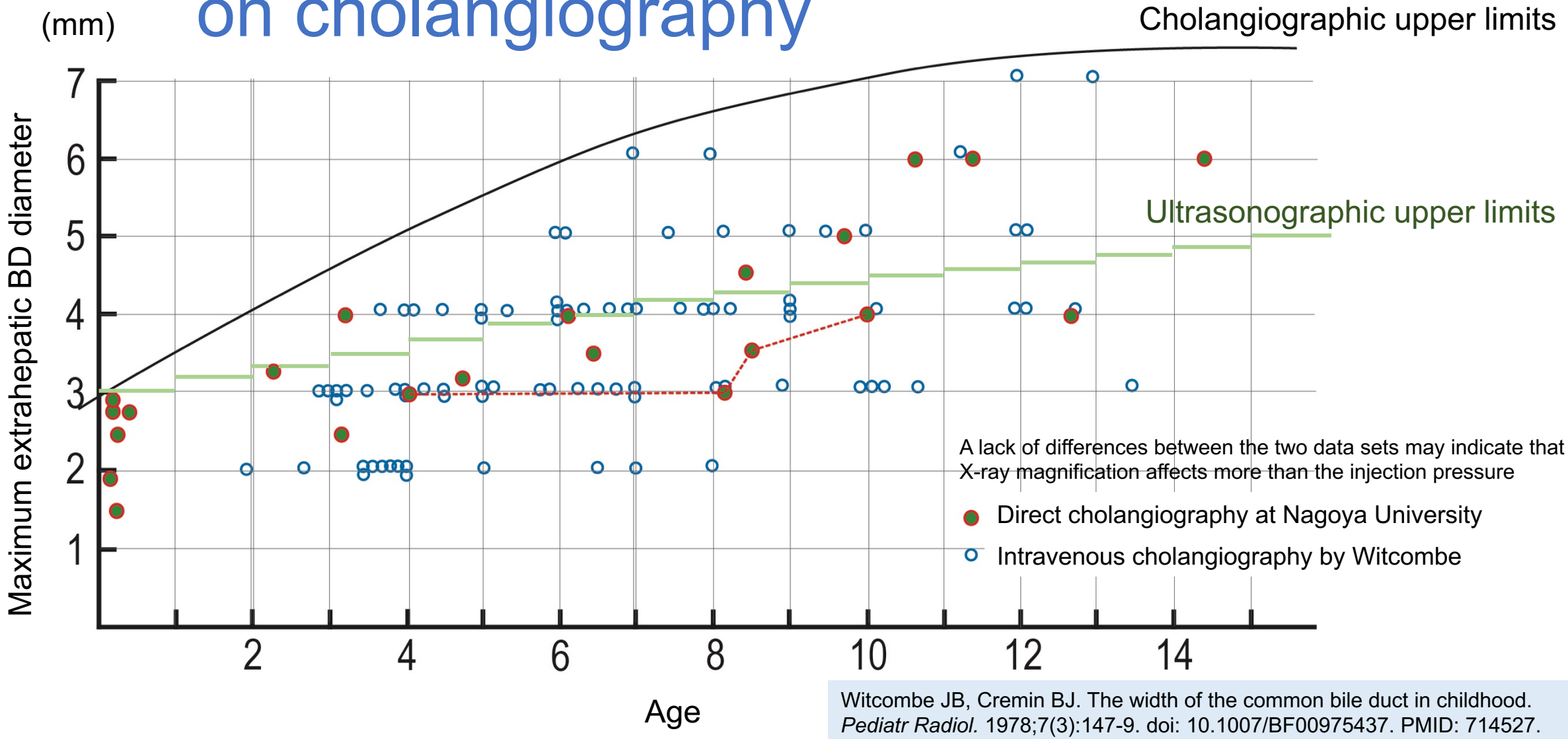
- Definition has not yet been determined.
- BD diameter **increases with age** in both children and adults. Ultrasonographic upper limits of normal BDs by age are determined.
- There is little data of normal BD diameters on direct cholangiography* or MRCP** at present.
- Non-dilated type should have a BD diameter within normal upper limits.
- Despite diameters within limits, cases with features of congenital biliary dilatation including dilated CC, NS, and dilated CD are in dispute for inclusion (see *Advanced Section*).

* larger than US due to Xray magnification and injection pressure; ** presumed similar to US

Ultrasonographic upper limits of normal BD diameters by age



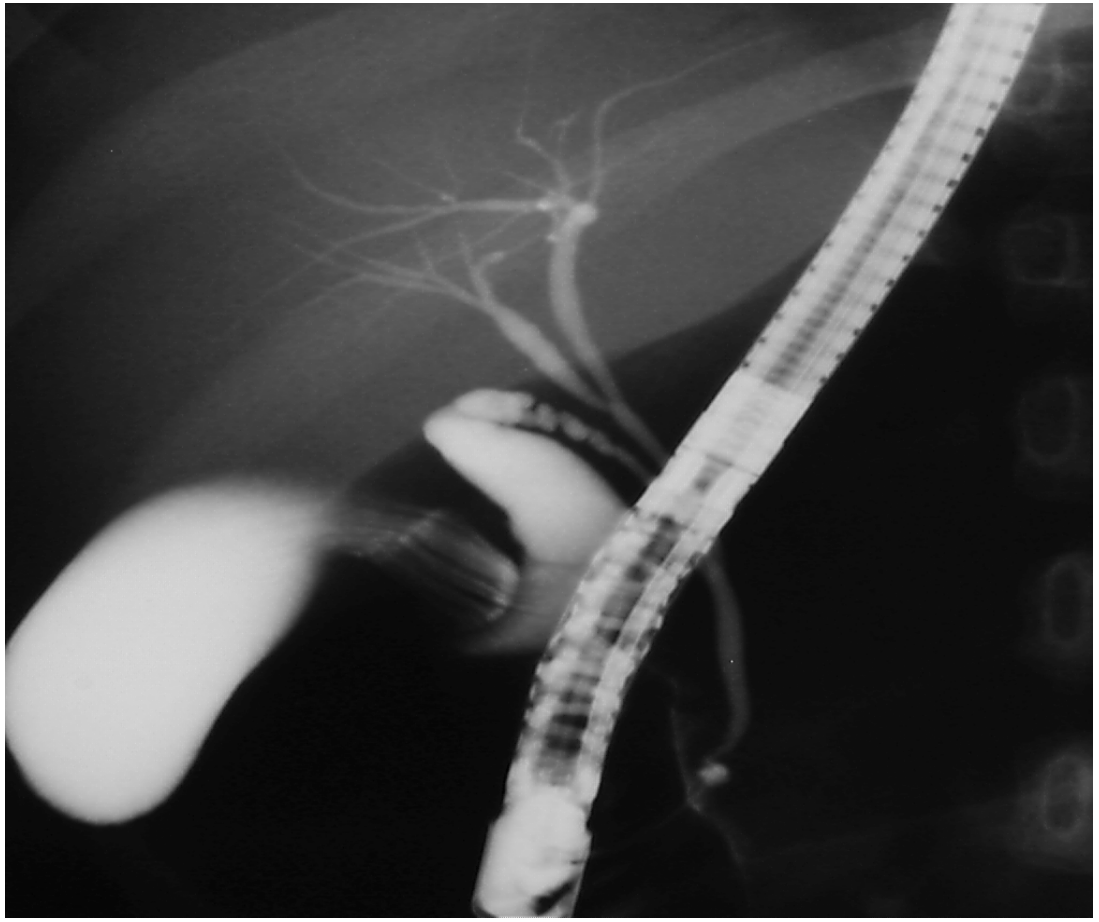
Normal pediatric BD diameters on cholangiography



Witcombe JB, Cremin BJ. The width of the common bile duct in childhood. *Pediatr Radiol.* 1978;7(3):147-9. doi: 10.1007/BF00975437. PMID: 714527.

Normal pediatric BD

Appearance varies from thin to thick.



3-year-old girl



6-year-old girl

Case 18

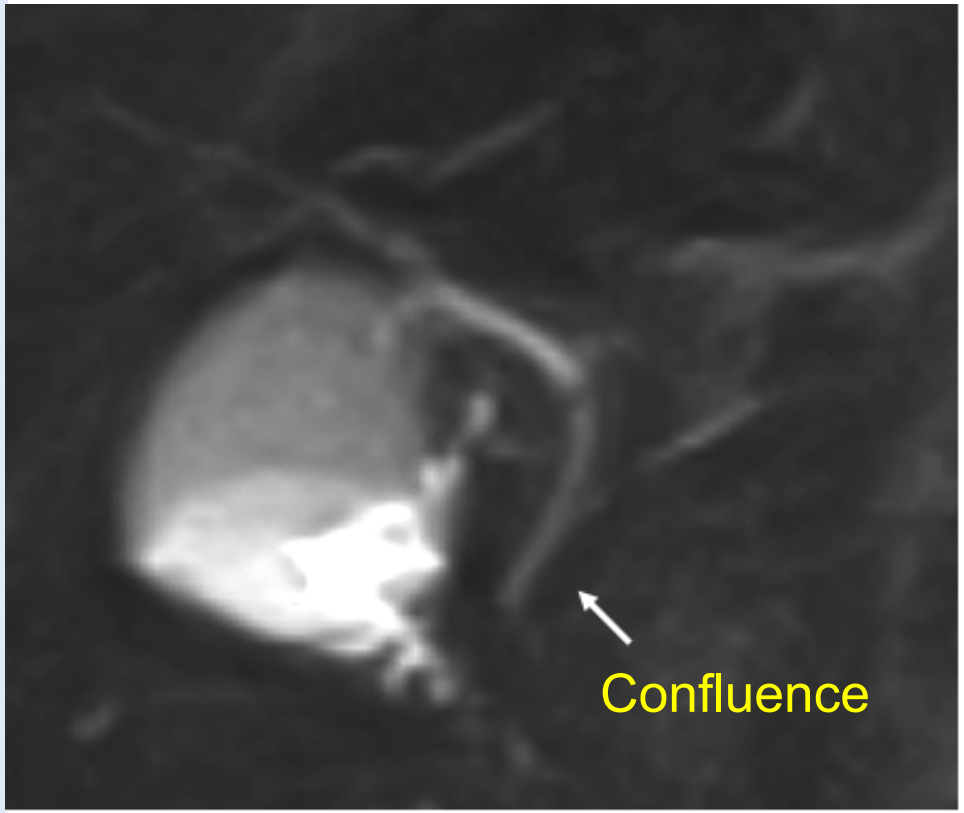
Typical non-dilated type of PBM: Pediatric case

This case had no features of congenital biliary dilatation such as a dilated common channel. True non-dilated type, such as this case, is extremely rare in children.

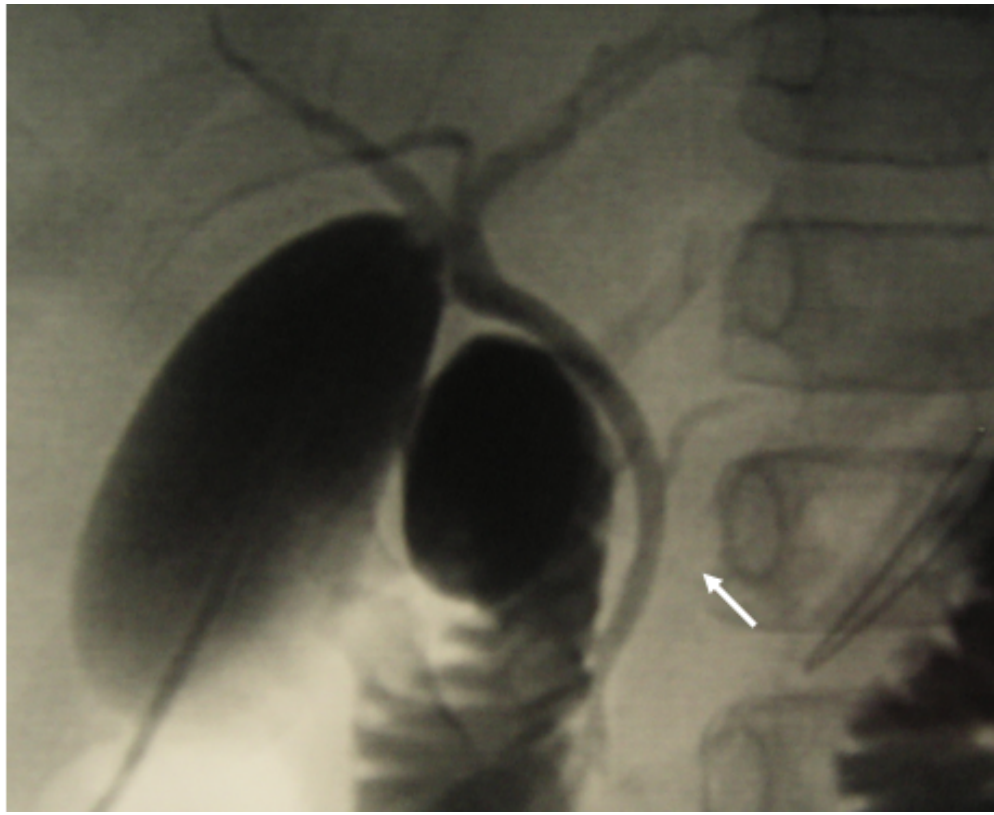
A 6-year-old girl
Abdominal pain, vomiting, hyperamylasemia

- **Abnormally long common channel = PBM**
  Type B, non-stenotic type
- **Diameter of CBD**
  4 mm on MRCP and cholangiography **within US limits**
- No features of congenital biliary dilatation except PBM

NS, dilated CC, dilated CD



MRCP



On-table cholangiography

See p41.

Case 19

Typical non-dilated type of PBM: Pediatric case

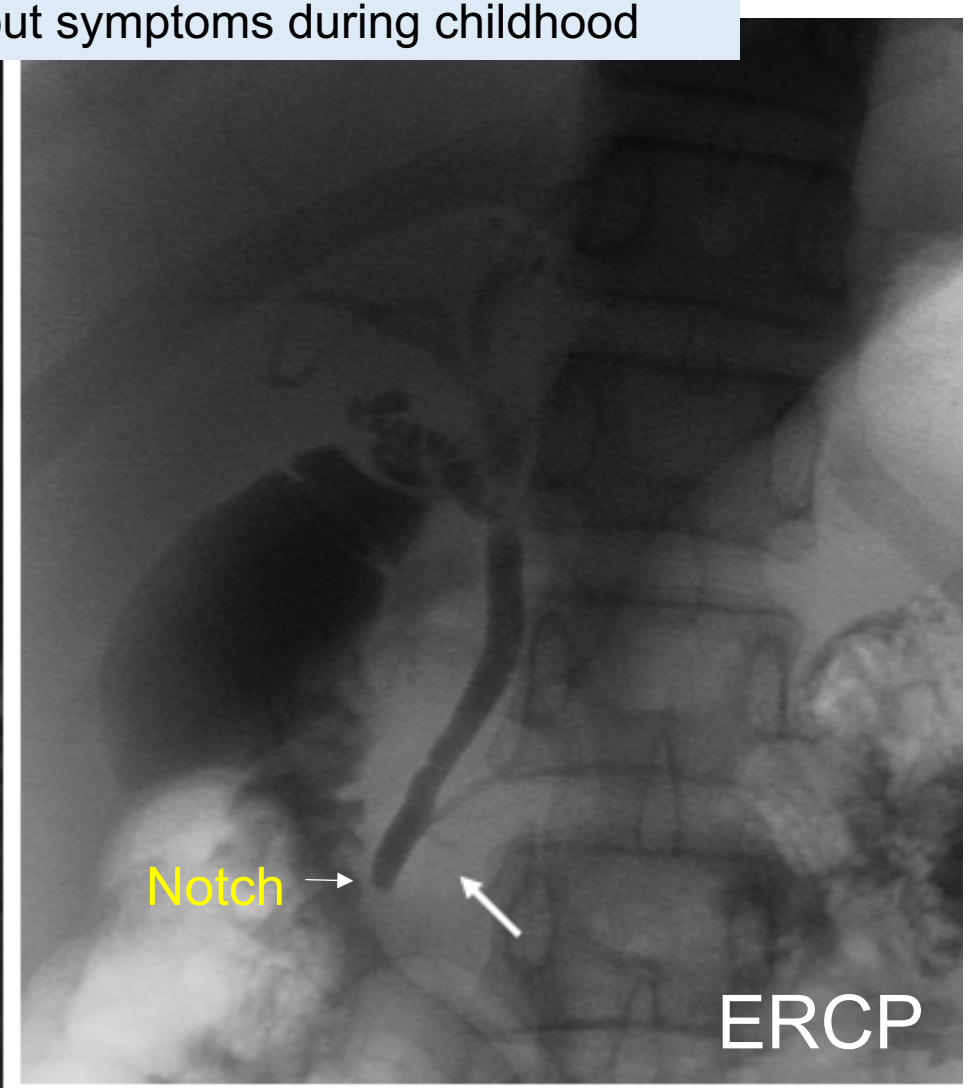
Non-dilated type has not been found without symptoms during childhood

Symptoms are the same as those of congenital biliary dilatation

A 12-year-old girl
Abdominal pain, vomiting, hyperamylasemia

- **Notch on the common channel = PBM**
  Type B, non-stenotic type
- **Diameter of CBD**
  MRCP 4 mm
  ERCP 4 mm
  within US limits
- No features of congenital biliary dilatation except PBM





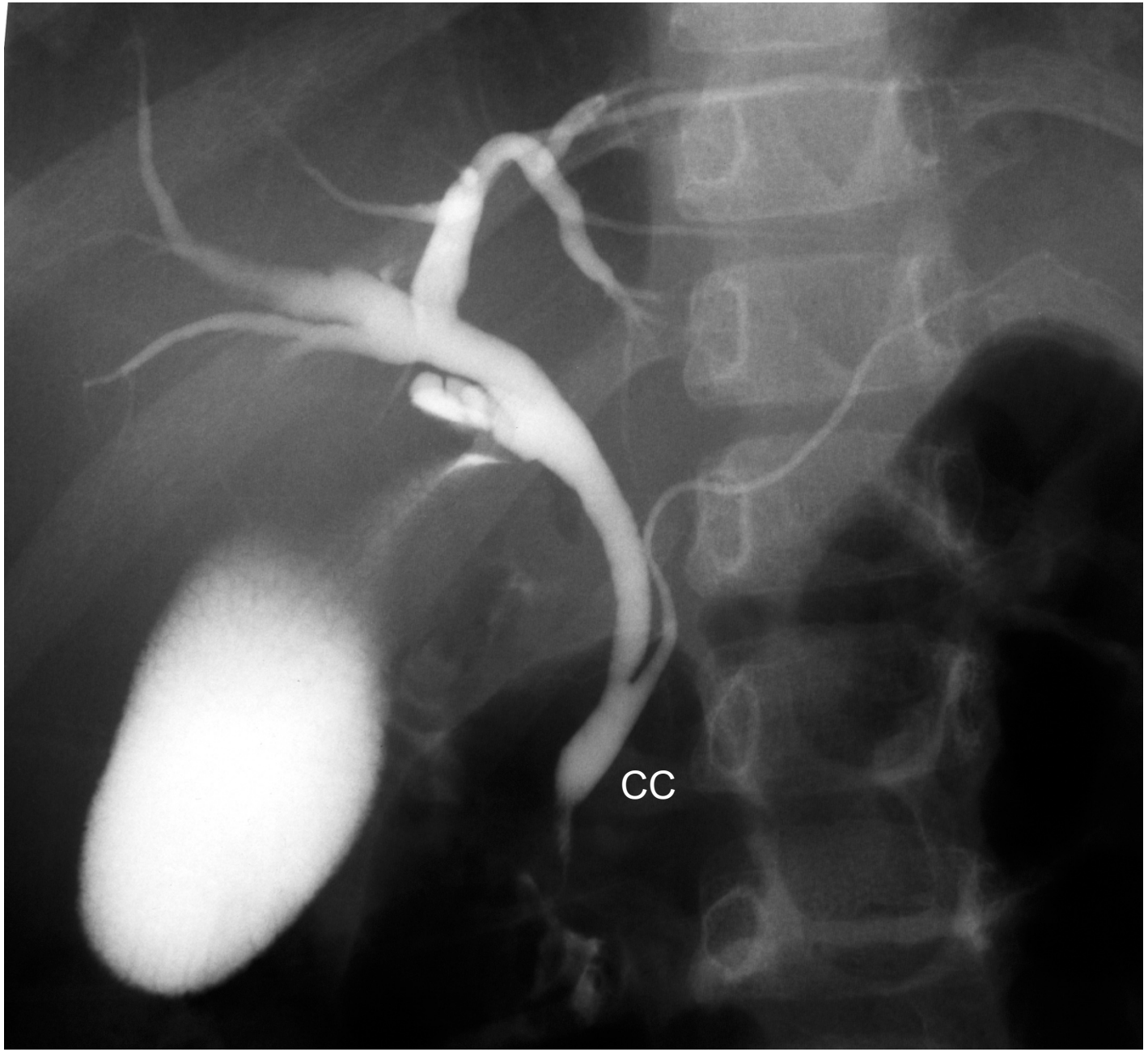
Case 20

Non-dilated type?

Despite normal BD calibers, many pediatric cases have other features of congenital biliary dilatation than PBM, such as in this case.
See *Advanced Section.*

ERCP in a 4-year-old girl
Abdominal pain, vomiting

- BD 5 mm in diameter on cholangiography
  cf. US upper limit, 4.2 mm at 4 years of age
- **Dilated common channel (CC)**
- CD seems dilated at its base



Case 21 Typical non-dilated type of PBM: Adult case

Non-dilated type is often found without symptoms in adults

Gallbladder cancer often arises in adult non-dilated type

MRCP in a female in her 50s.
GB cancer found incidentally on CT for another disease.

- **Abnormally long CC = PBM**
  Type B, non-stenotic type
- Diameter of the CBD
  6.2 mm on MRCP
  Within US limits
- No features of congenital biliary dilatation except PBM



From *Introduction Section*, see p31.

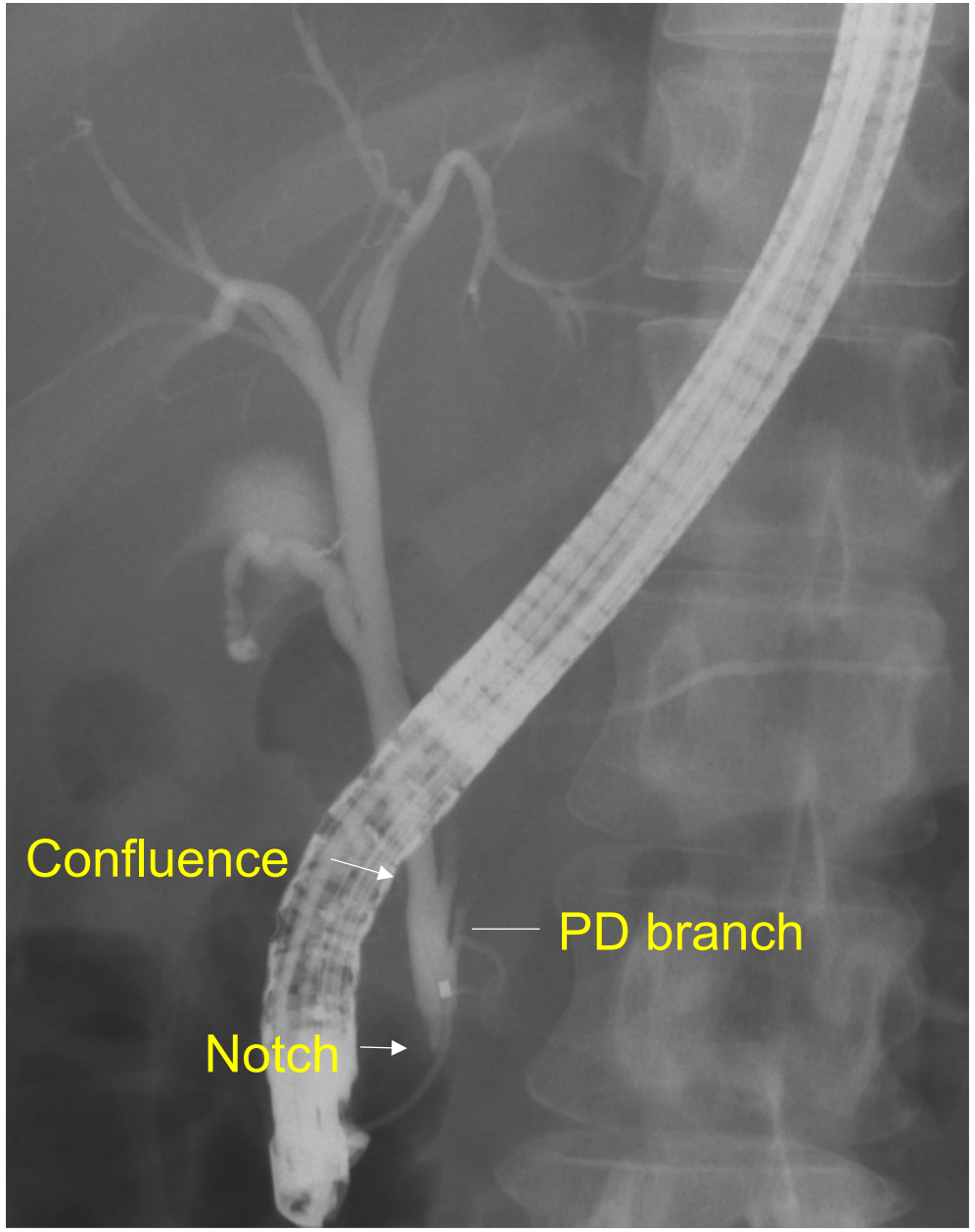
Case 6 Typical non-dilated PBM: Adult case

ERCP in a female in her 40s
Right hypochondrial pain

- **Abnormally long common channel**
  = Notch on the common channel
  = PBM
- **Diameter of the extrahepatic BD**

US	4 mm
MRCP	4 mm
ERCP	4 mm

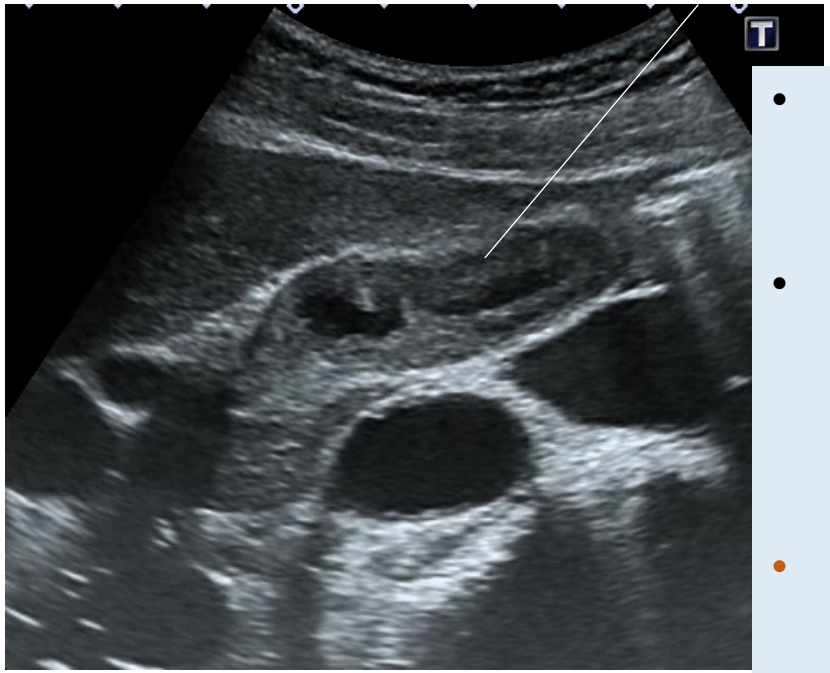
- **No features of congenital biliary dilatation** but for PBM
- A PD branch from the common channel
  The common channel is composed of PD?
  See *Advanced Section*.



Case 22 Non-dilated type of PBM: Typical opportunity for discovery in adults

A female in her 50s

Screening US found GB wall thickness

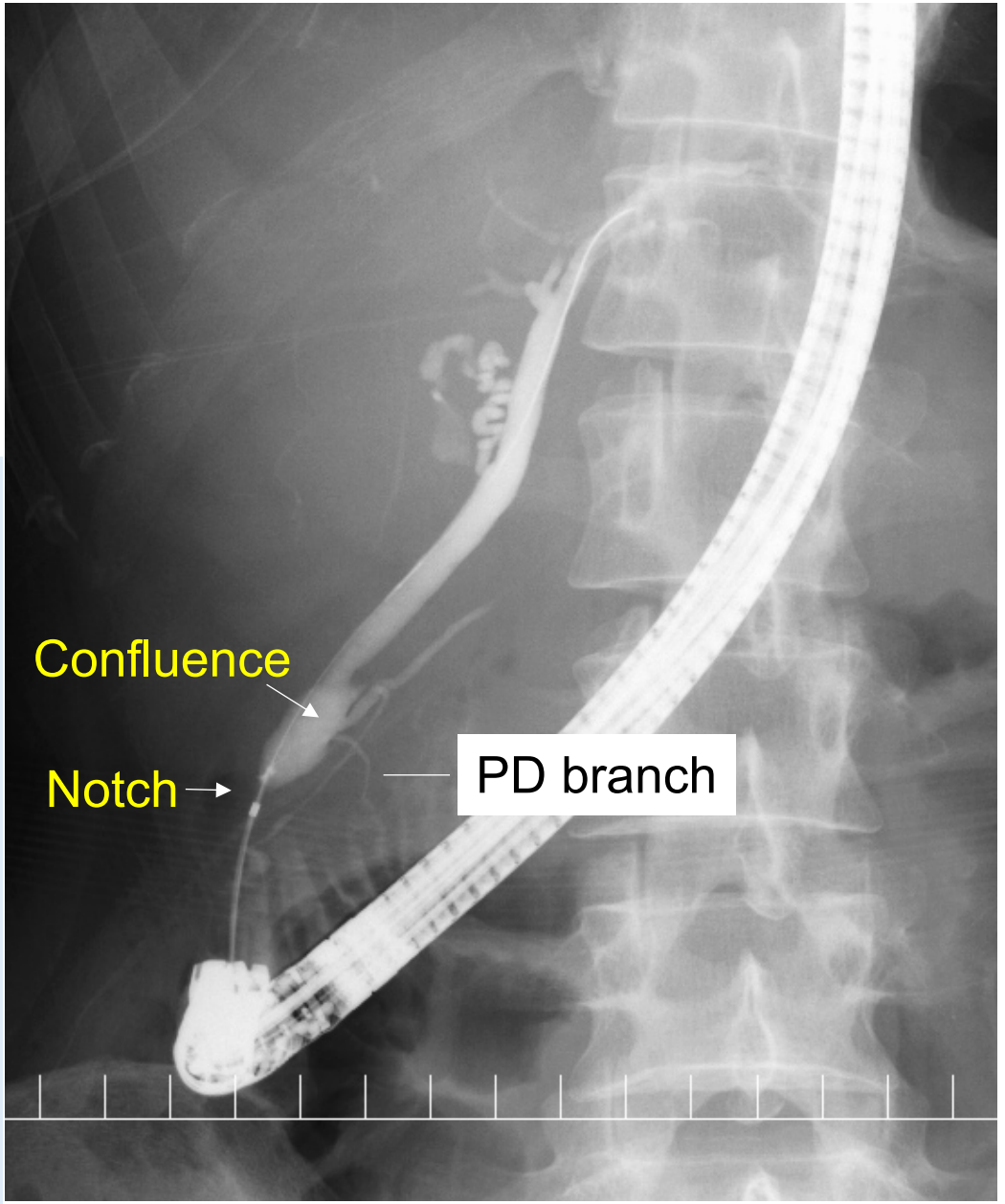


See *Advanced Section*. ←

- **Notch on the common channel = PBM**
- **Diameter of the extrahepatic BD**

US	4.2 mm
MRCP	6.2 mm
ERCP	5.8 mm

- **Feature of CBD**
  Dilated common channel
- A PD branch from the common channel



Basics

Treatment for PBM

First version, September 2022
2nd version, September 2023

Treatment for pancreaticobiliary maljunction

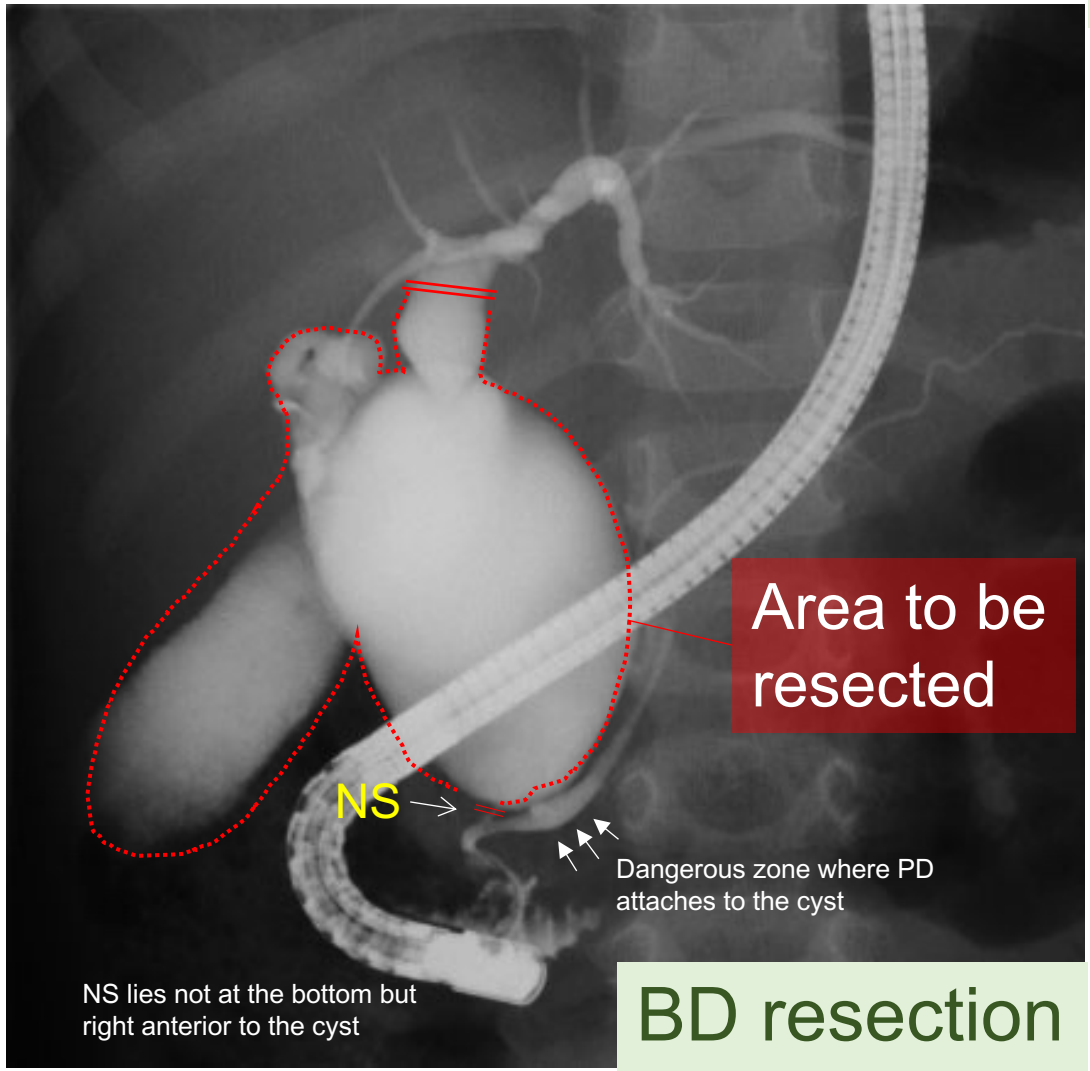
- Rationale for treatment:
  - ✓Not merely to correct malformation!
  - ✓Treatment is indicated to prevent complications.
- Complications of PBM:
  - ✓**Pediatric cases**: Recurrent **symptoms** including abdominal pain, vomiting, and jaundice.
  - ✓**Adult cases**: High incidence of **biliary tract cancer** (see p15).
- Surgical Management:
  - **Congenital biliary dilatation**: Requires **bile duct resection.**
  - **Non-dilated PBM**: Requires **at least cholecystectomy.***

*There is currently no consensus on the necessity of bile duct resection for non-dilated PBM due to the low incidence of carcinogenesis within the bile duct.

In pediatric cases, however, bile duct resection is commonly performed because protein plugs can induce symptoms via the same pathophysiology observed in congenital biliary dilatation.

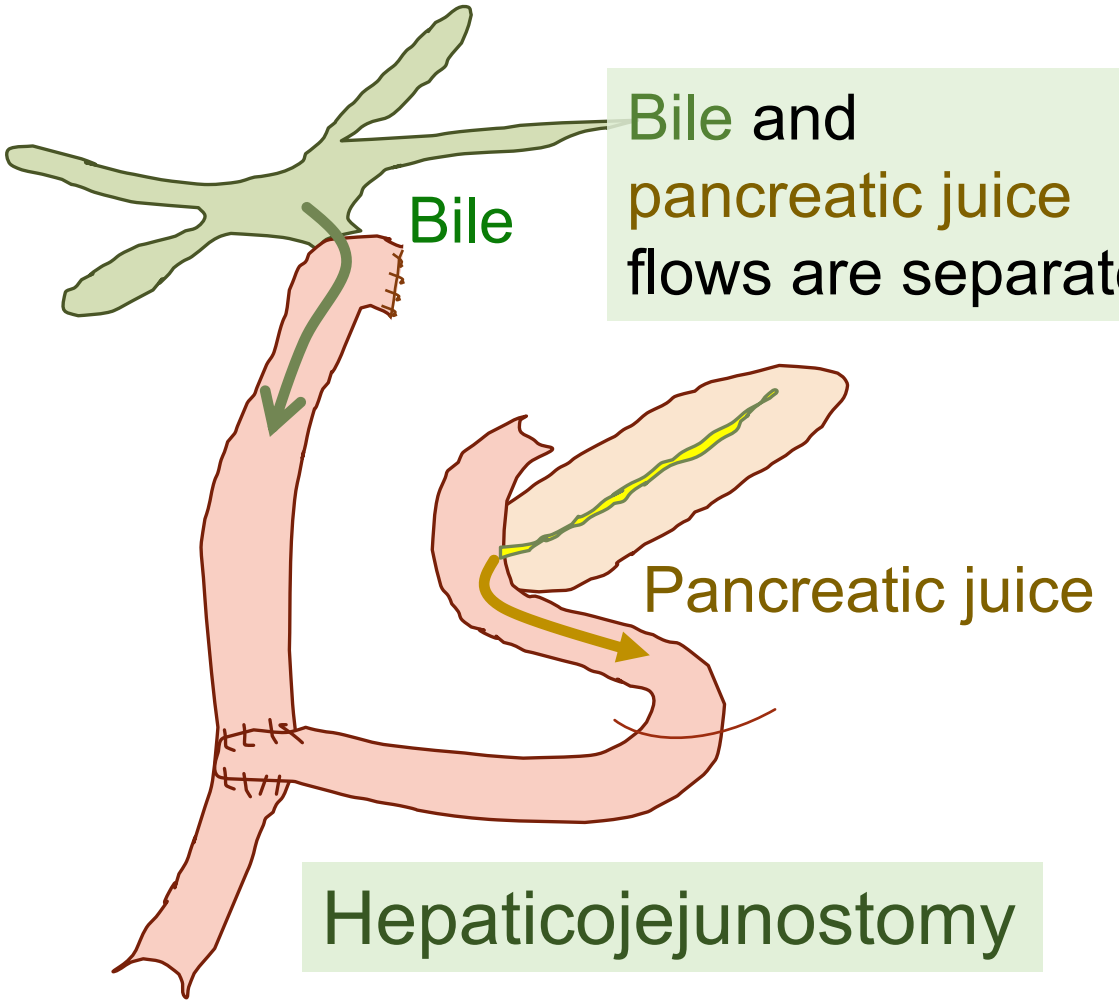
Surgery for CBD

Must resect where cancers would arise and stop the reflux of pancreatic juice to prevent cancers and symptoms



BD resection

NS, narrow segment



Bile and pancreatic juice flows are separated

Hepaticojejunostomy

Cf. Hepaticoduodenostomy allows reflux of pancreatic juice into the biliary system

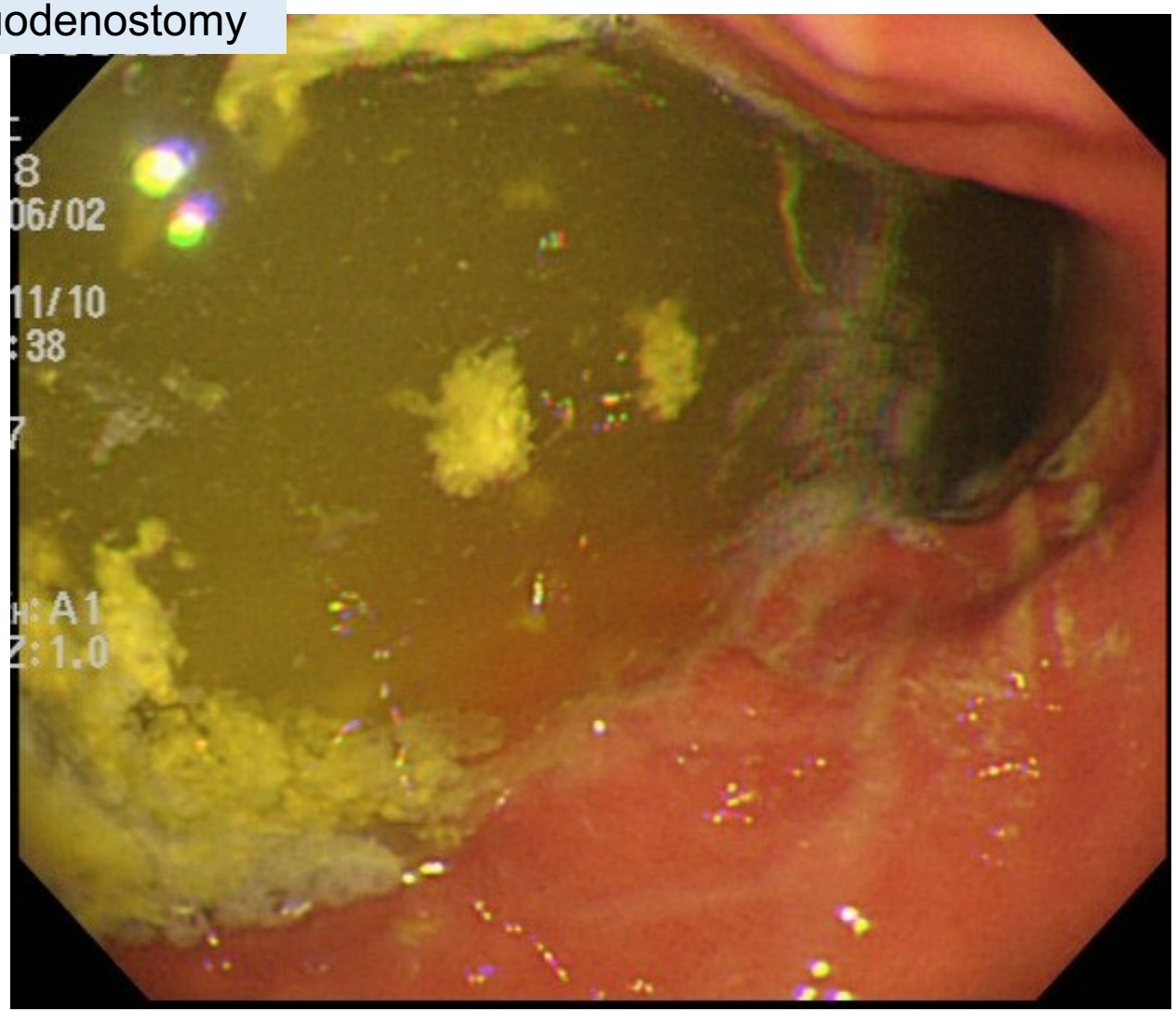
Bile gastritis after hepaticoduodenostomy

One of the reasons not to recommend hepaticoduodenostomy

A female in her twenties underwent bile duct resection with hepaticoduodenostomy for congenital biliary dilatation (CBD).

Seven years later, she developed epigastric pain. Endoscopy revealed bile regurgitation into the stomach, along with redness, edema, and erosion in some areas of gastric mucosa.

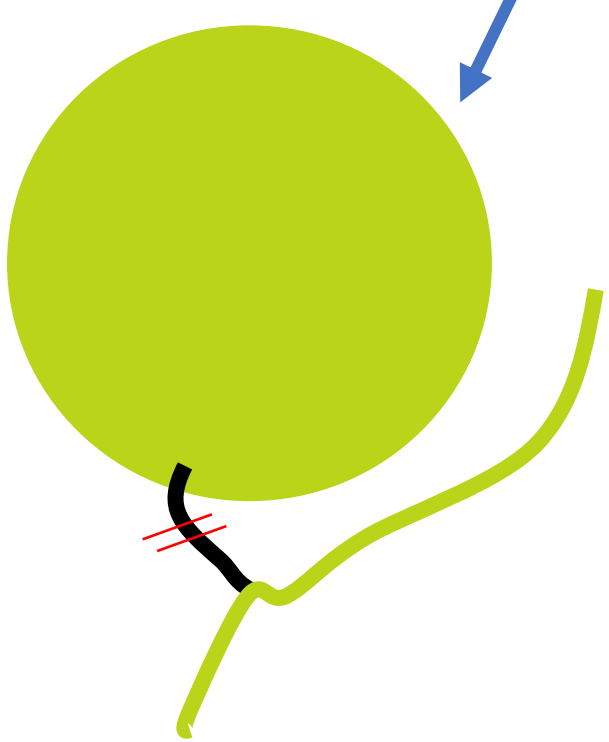
Three months after the examination, she underwent conversion to hepaticojejunostomy, which completely relieved her epigastric pain and endoscopic abnormality.



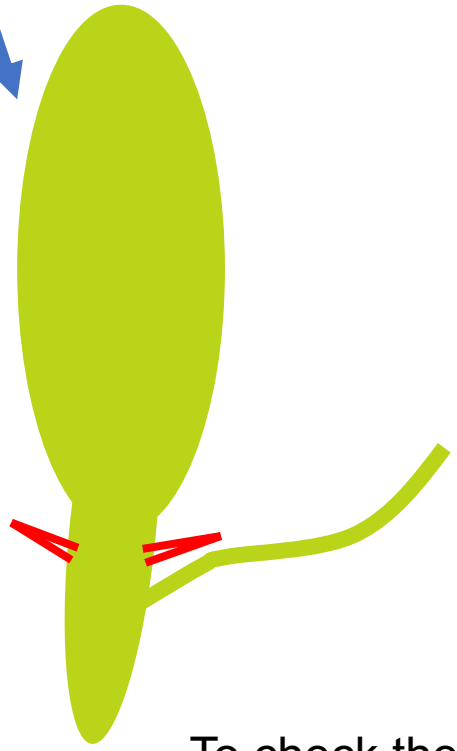
Complete resection of intrapancreatic BD

Residual intrapancreatic BD would produce protein plug reformation and carcinogenesis. See *Complications Section*.

Different procedures **with** and **without** a narrow segment



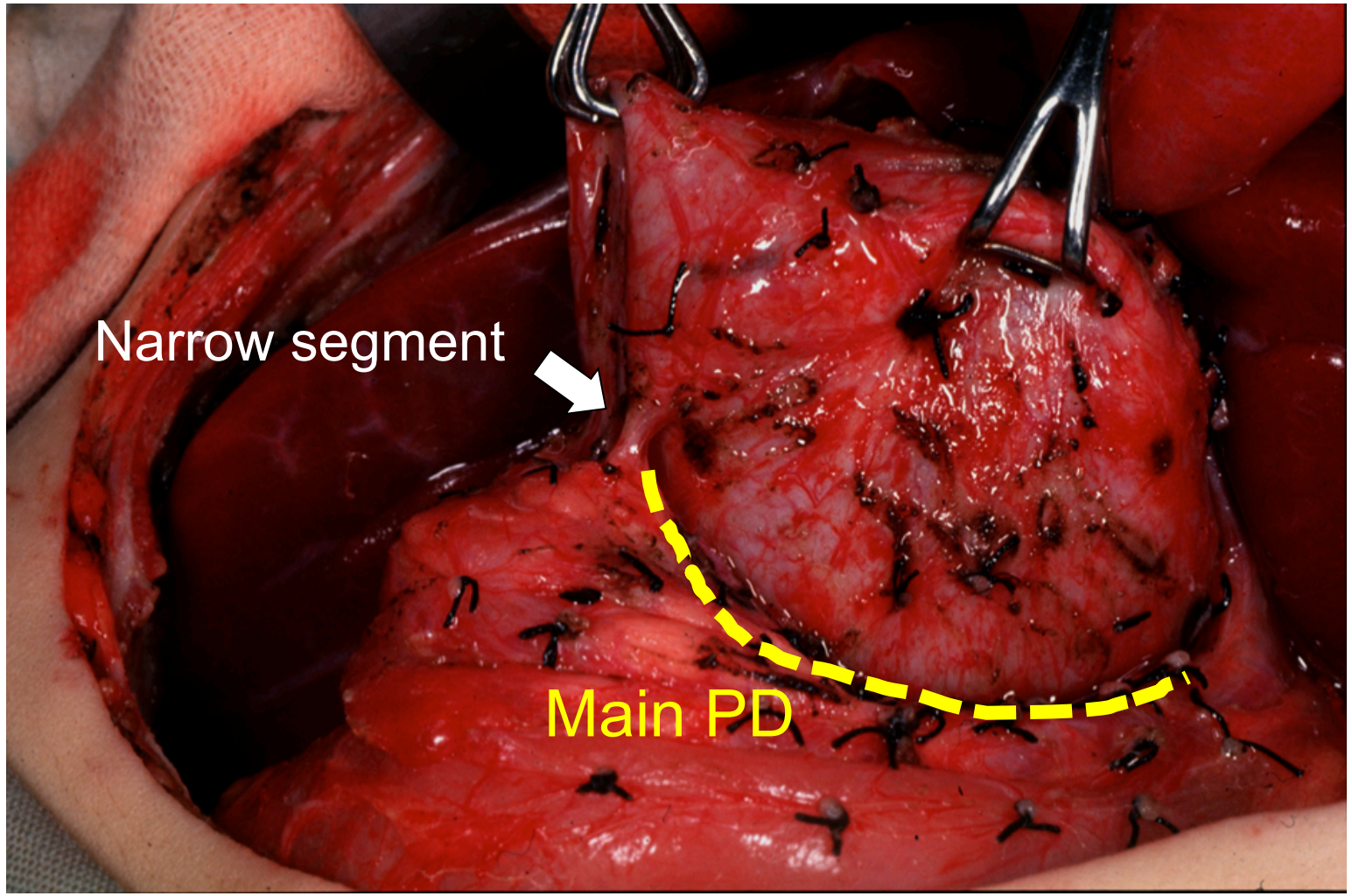
Visual confirmation of narrow segment



To check the distance to PD

Fluoroscopy using clips

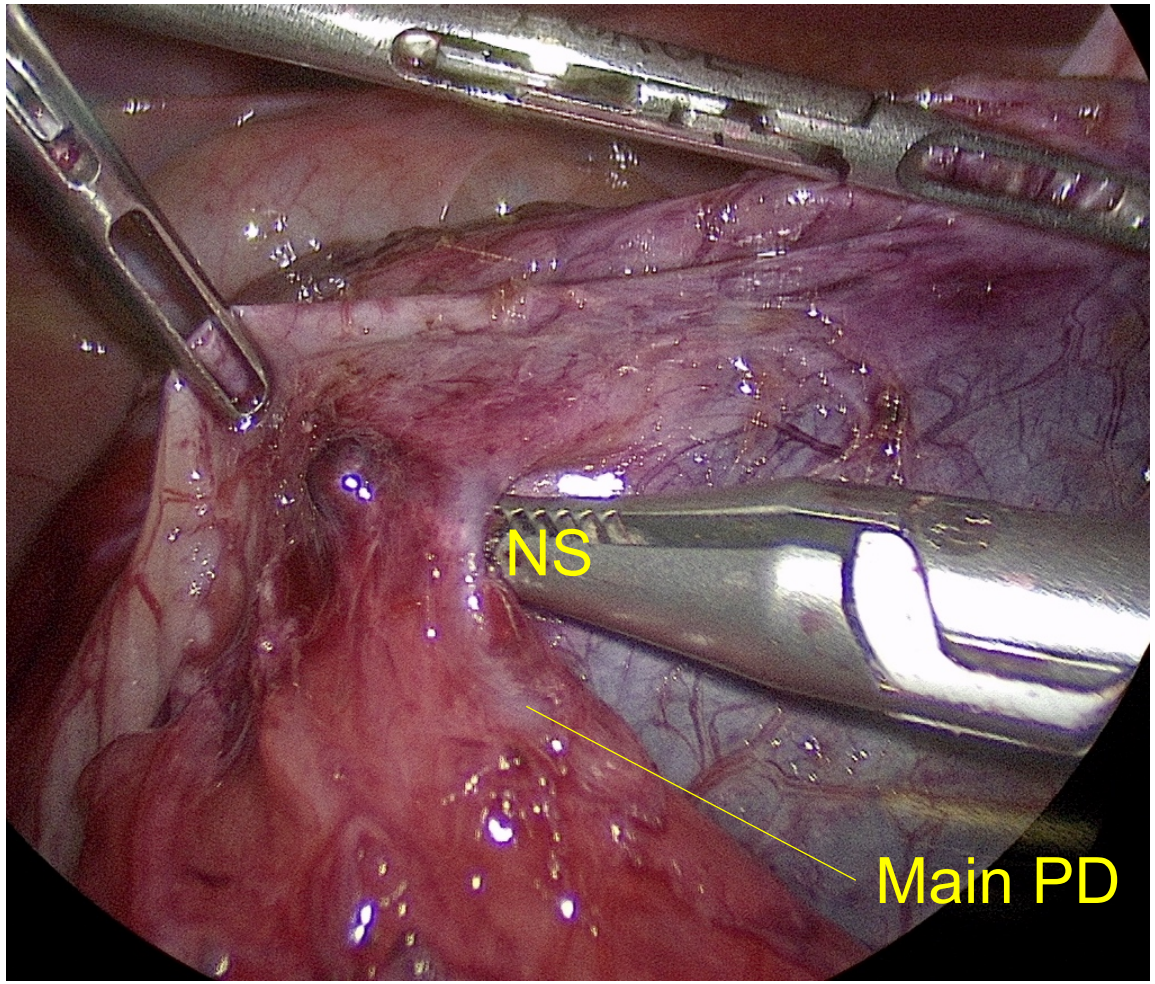
Visual confirmation of narrow segment

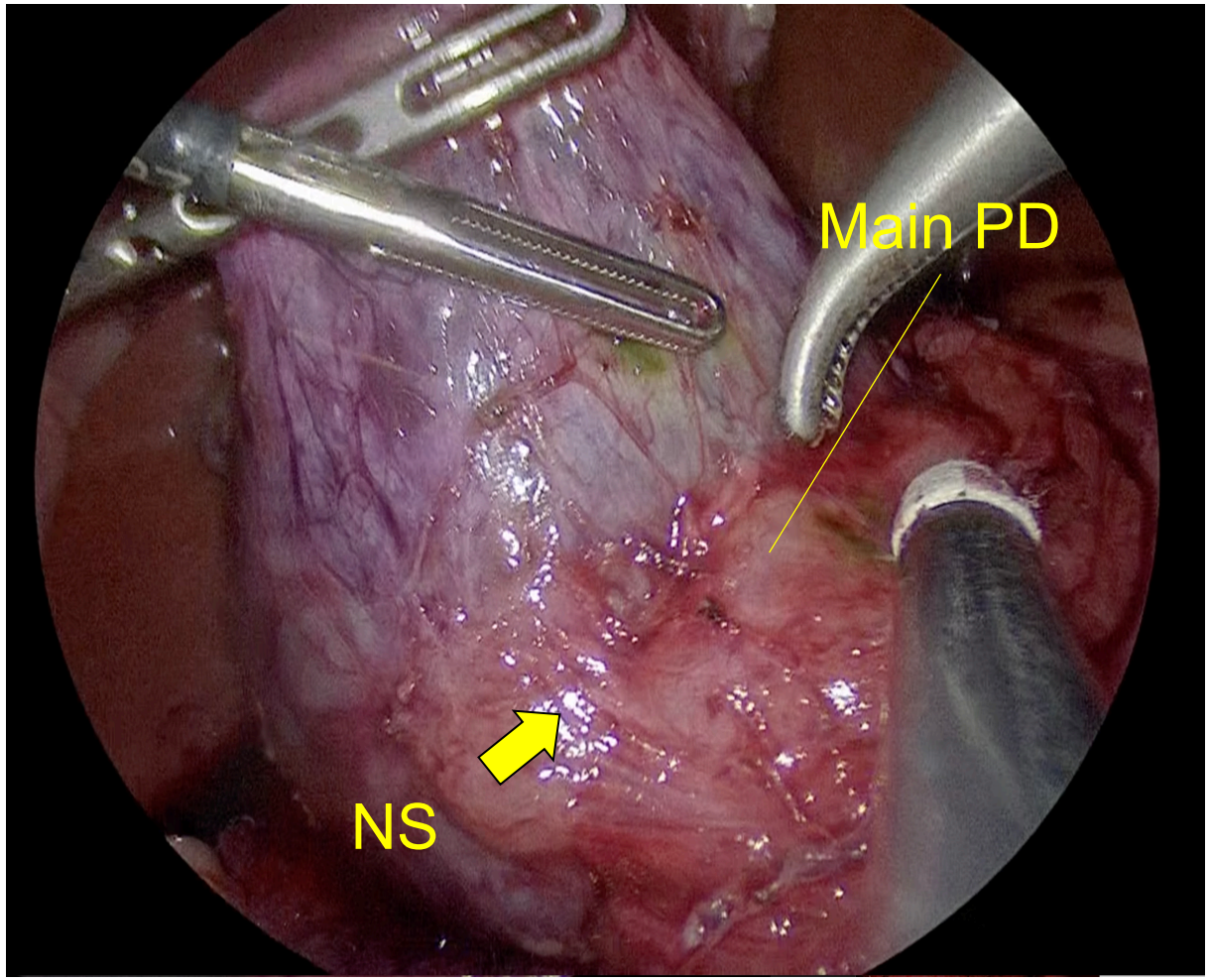


The narrow segment is often located not at the bottom but on the right-ventral side of the cyst.
In this case, care must be taken not to injure the PD because it runs over the cyst surface.

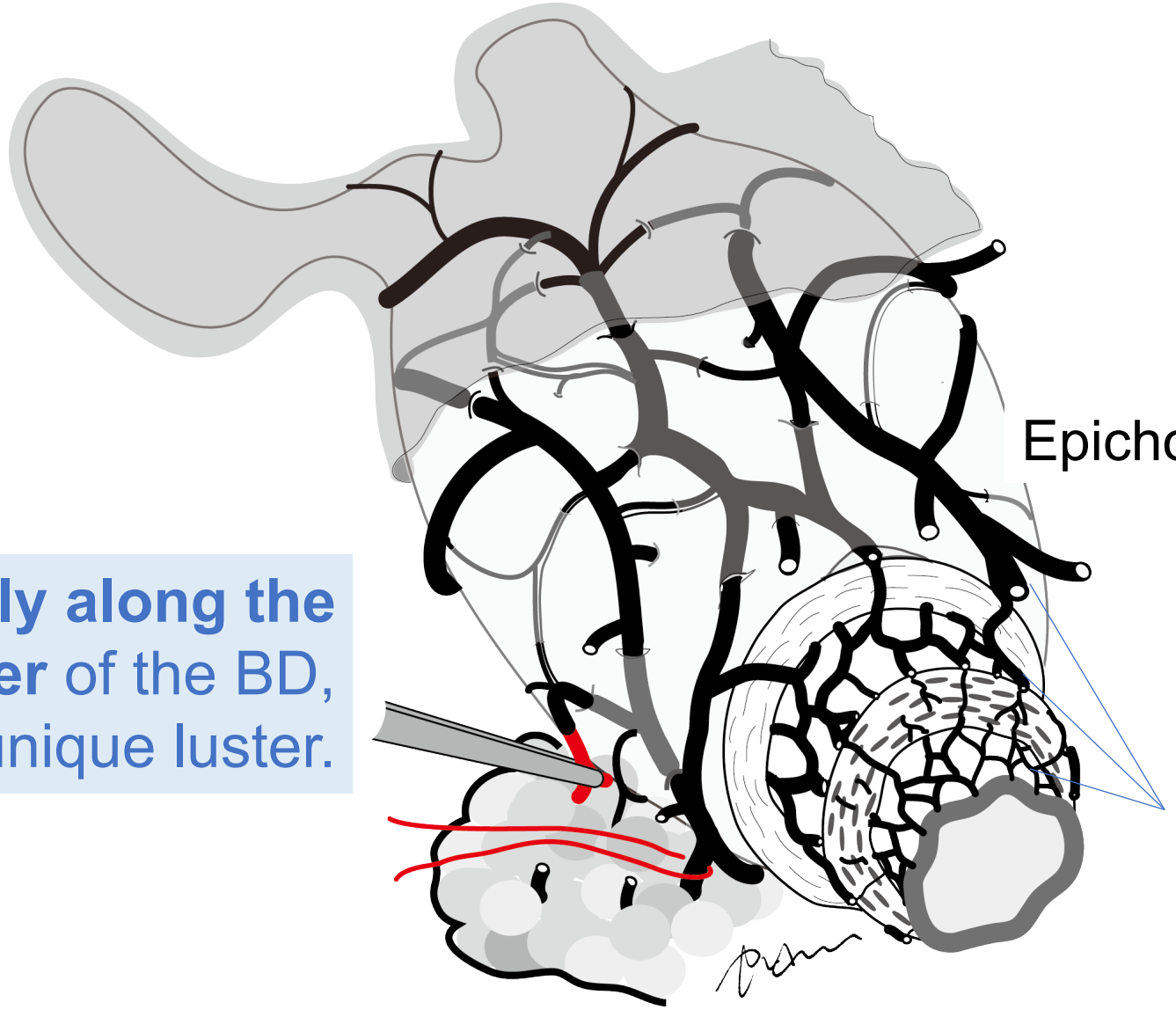
Main PD adheres to the cystically dilated BD

Infant case





How to avoid pancreatic duct injury



Epicholedochal plexus

Multilayer vascular plexuses

→ Cutting into the BD wall causes bleeding that obscures the layer.

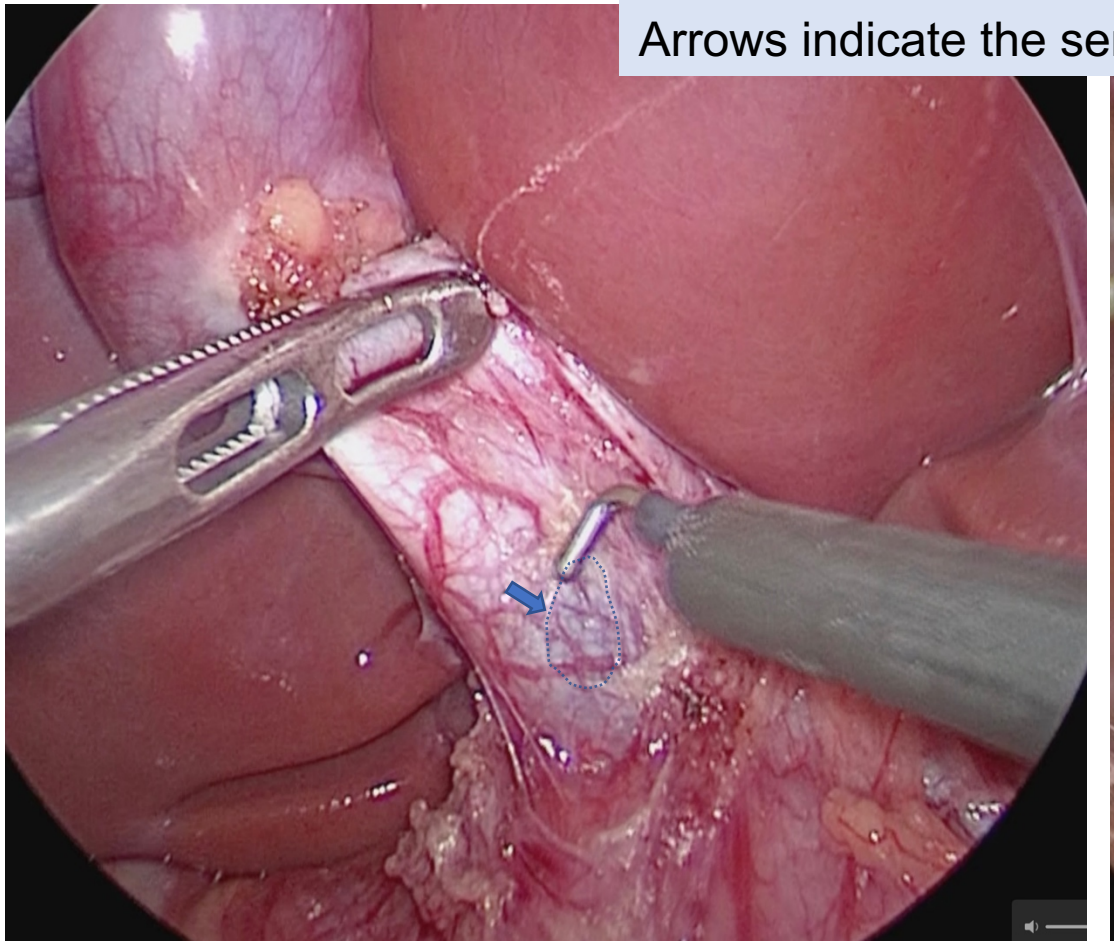
Dissect strictly along the serosal layer of the BD, which has a unique luster.

BD serosa has a unique luster

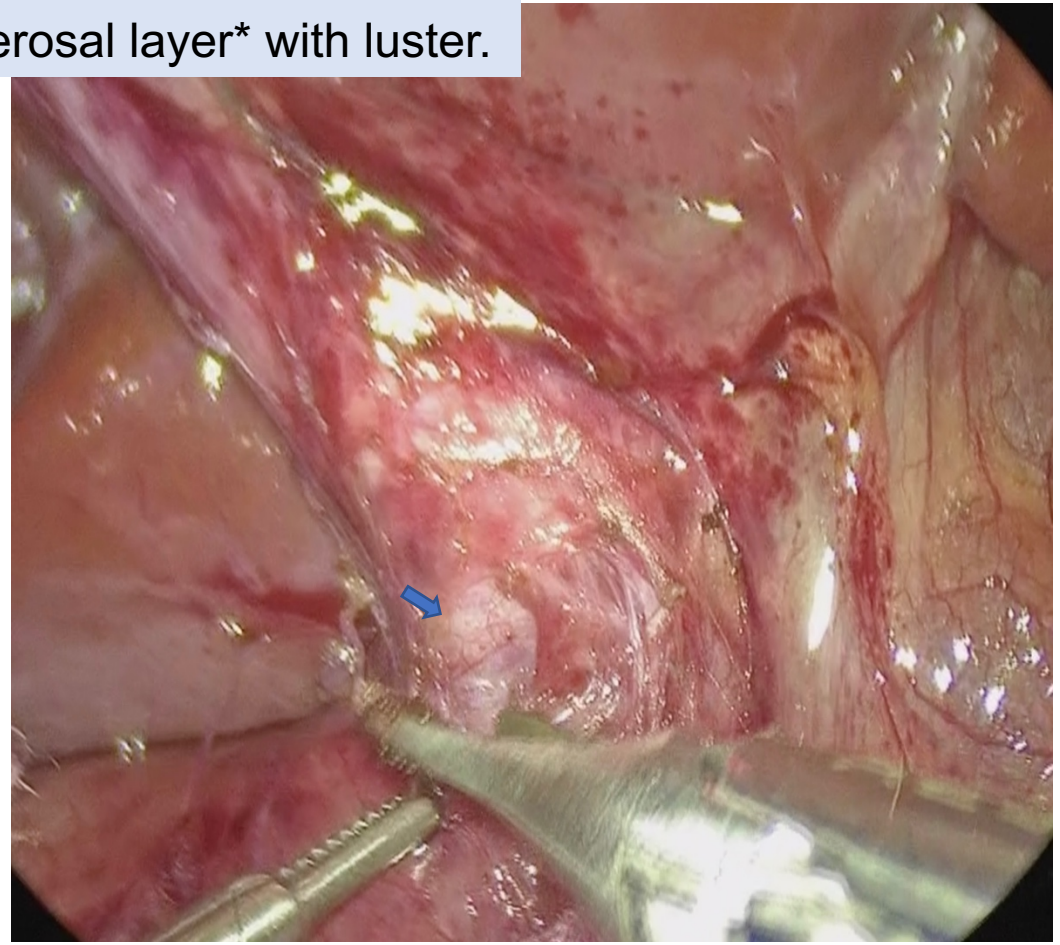
*To be precise, this layer is not the serosa but corresponds to the inner subserosal (ss inner) layer of the gallbladder.

Honda G, Iwanaga T, Kurata M. Dissection of the gallbladder from the liver bed during laparoscopic cholecystectomy for acute or subacute cholecystitis. *J Hepatobiliary Pancreat Surg*. 2008;15(3):293-6. doi: 10.1007/s00534-007-1260-3. Epub 2008 Jun 6. PMID: 18535767.

Arrows indicate the serosal layer* with luster.



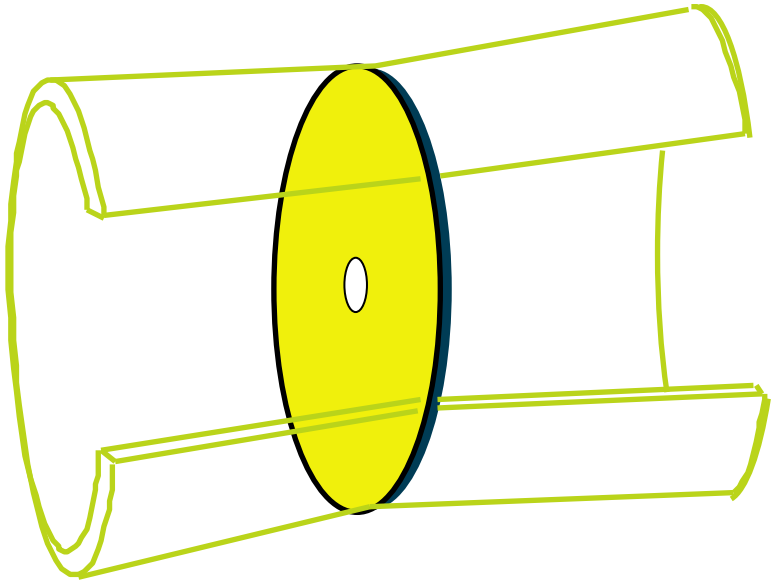
Case without inflammatory adhesion



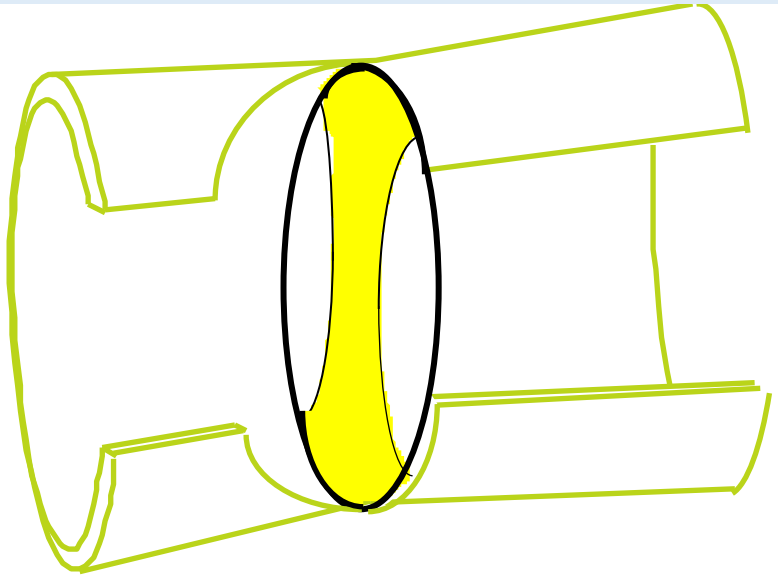
Case with inflammatory adhesion

Two types of congenital BD stenosis around the hepatic hilum

Overlooking these stenoses during BD resection can cause postoperative intrahepatic gallstone formation.
Stenoses is present in 85% of IV-A congenital biliary dilatation.



Membranous stenosis

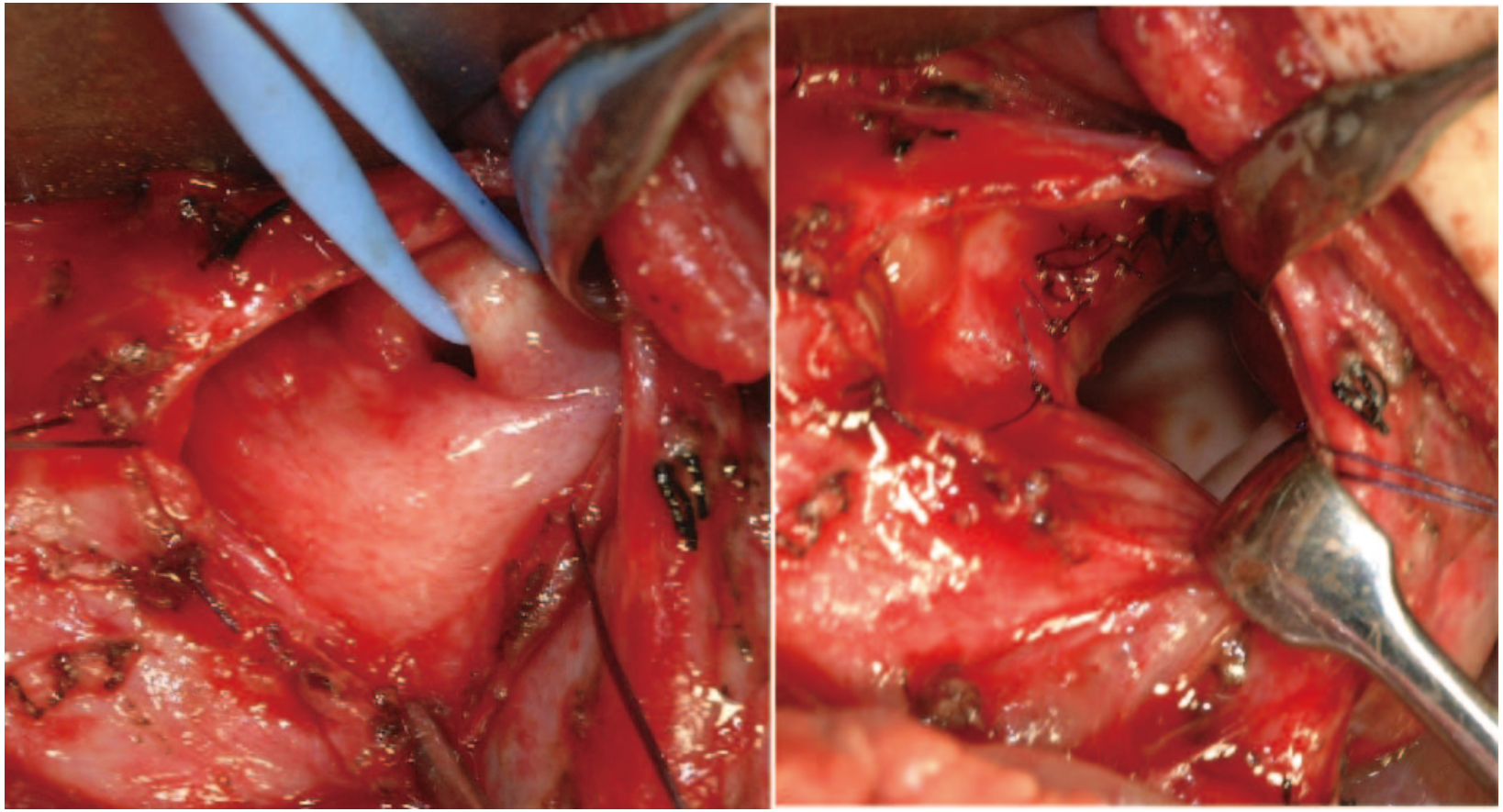


Septal stenosis

Balloon dilatation is ineffective for these stenoses.

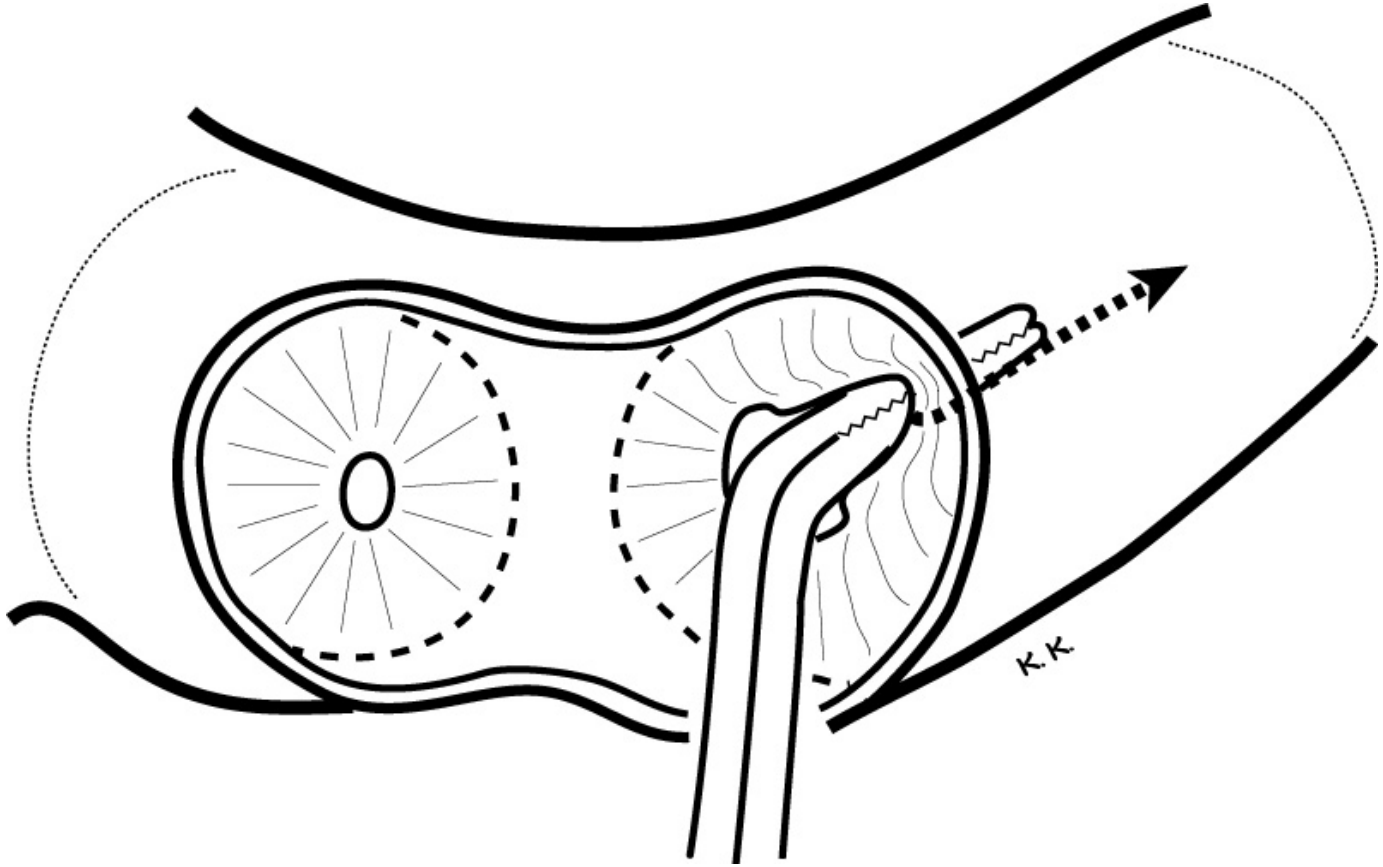
Ando H, Ito T, Kaneko K, Seo T.
Congenital stenosis of the intrahepatic bile duct associated with choledochal cysts. J Am Coll Surg. 1995 Nov;181(5):426-30. PMID: 7582210.

Resection of septal stenosis



During BD resection

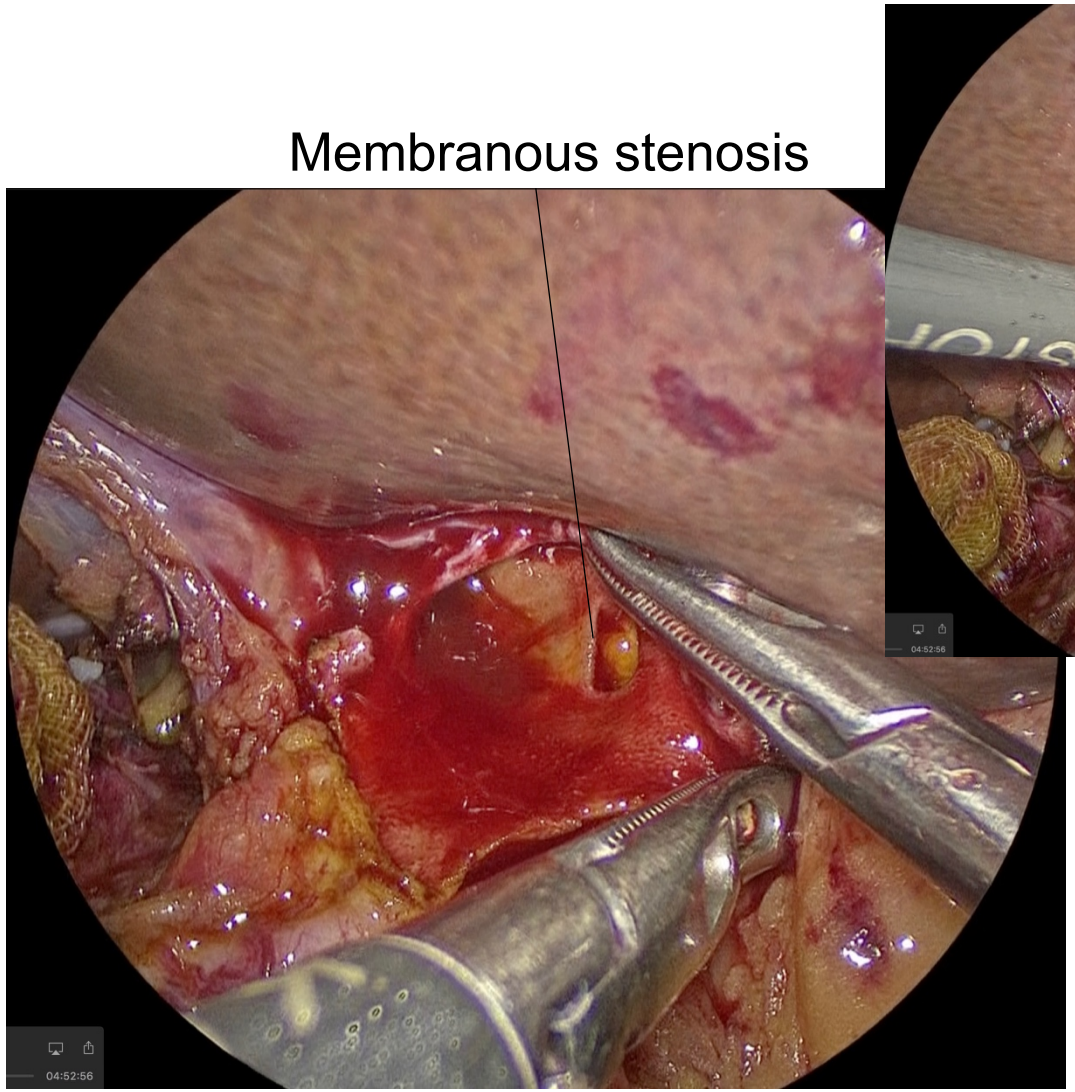
Todani and Lilly's BD plasty for membranous stenosis



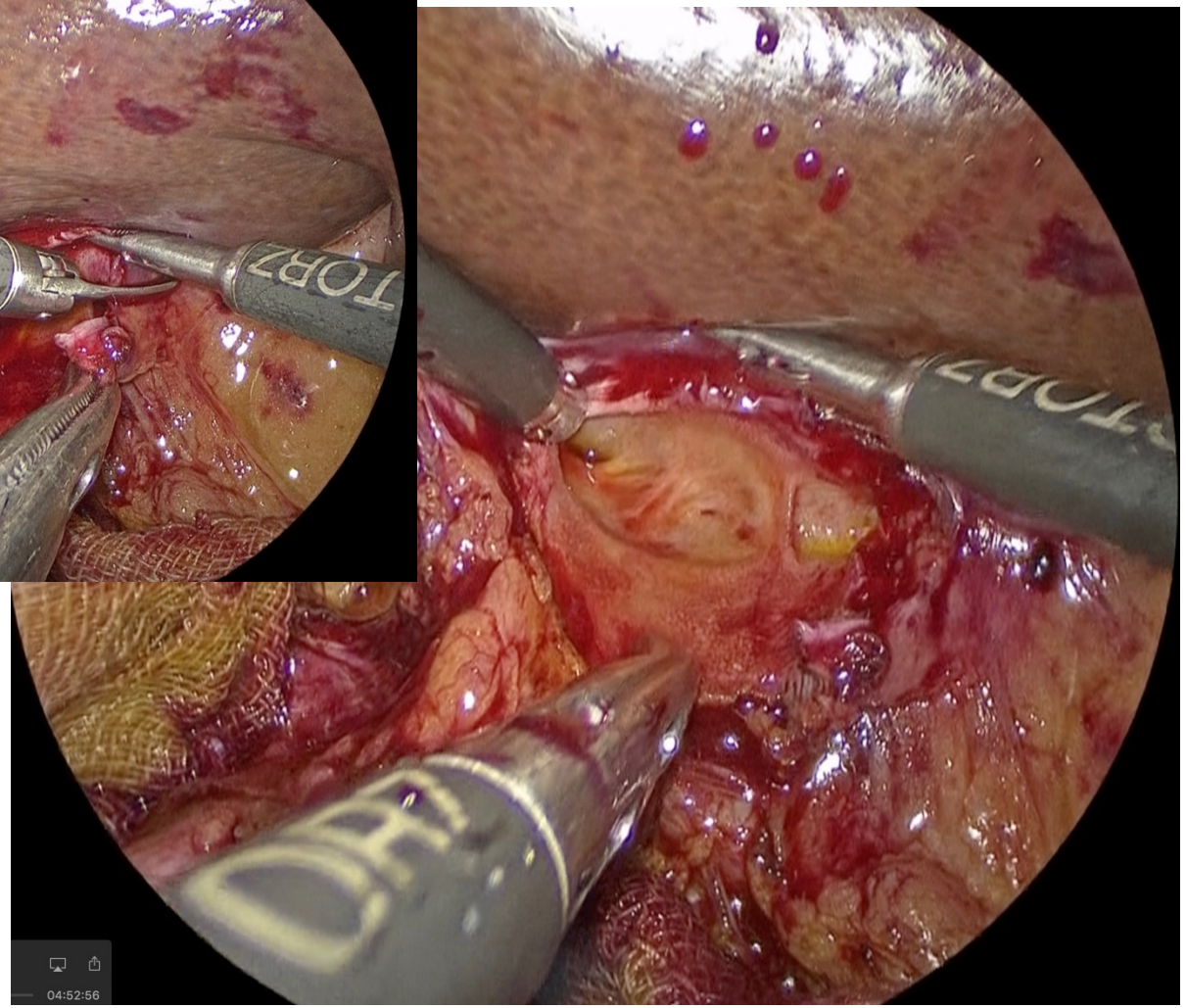
This procedure is useful for cases with small-caliber CHD when membranous stenosis is difficult to resect.

Todani’s BD plasty

Membranous stenosis



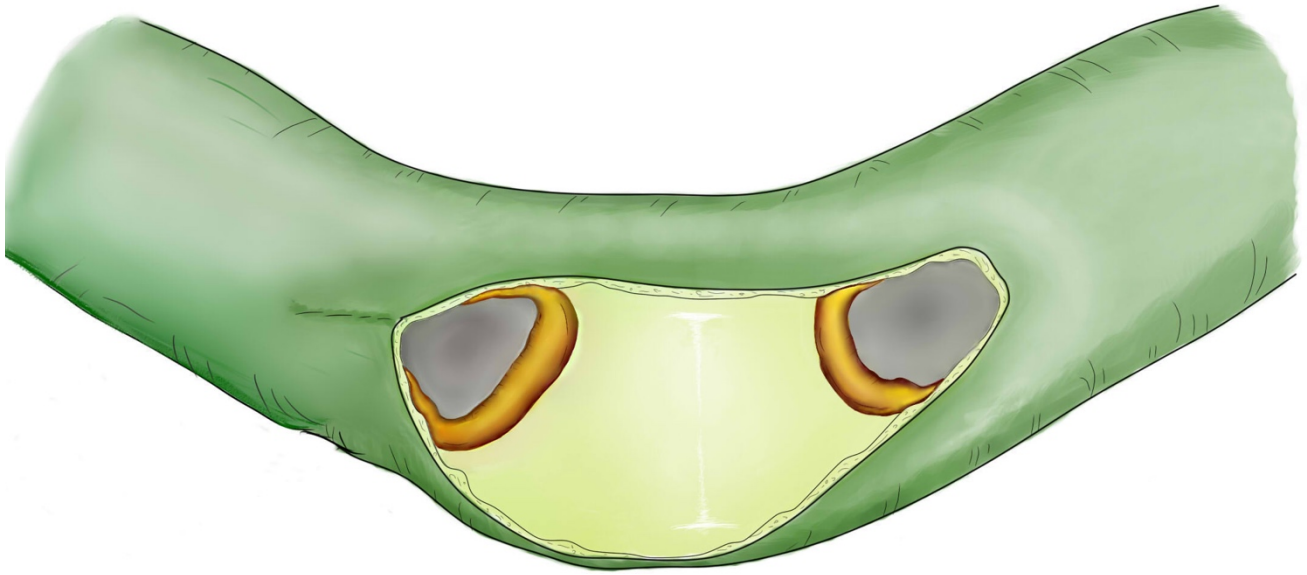




Hepaticojejunostomy with a wide stoma

Especially important for type IV-A cases





Hepaticojejunostomy in a case with membranous stenoses

This procedure prevents anastomotic stenosis and also relieves congenital stenosis.

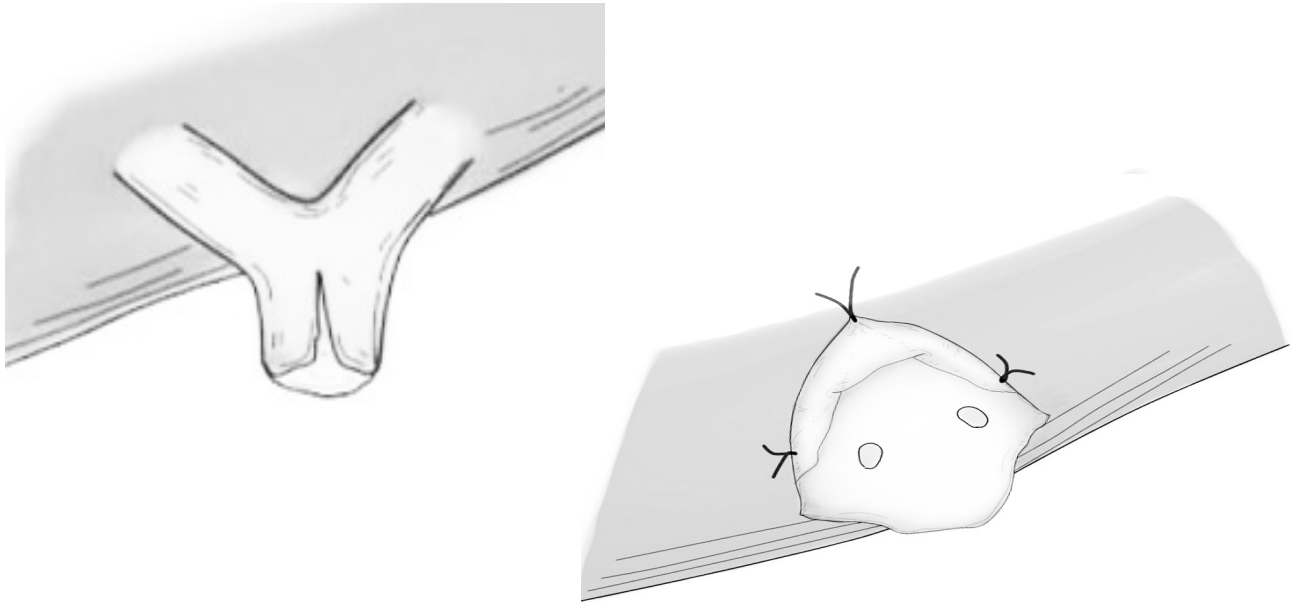
Procedures to prevent anastomotic stenosis in cases with small-caliber hepatic ducts

Open surgery



The Todani and Lilly procedure is applicable in open surgery, but difficult in laparoscopic surgery.

Laparoscopic surgery



An inverted bile duct fixed to the hilar plate provides a good operative field and secure anastomosis.

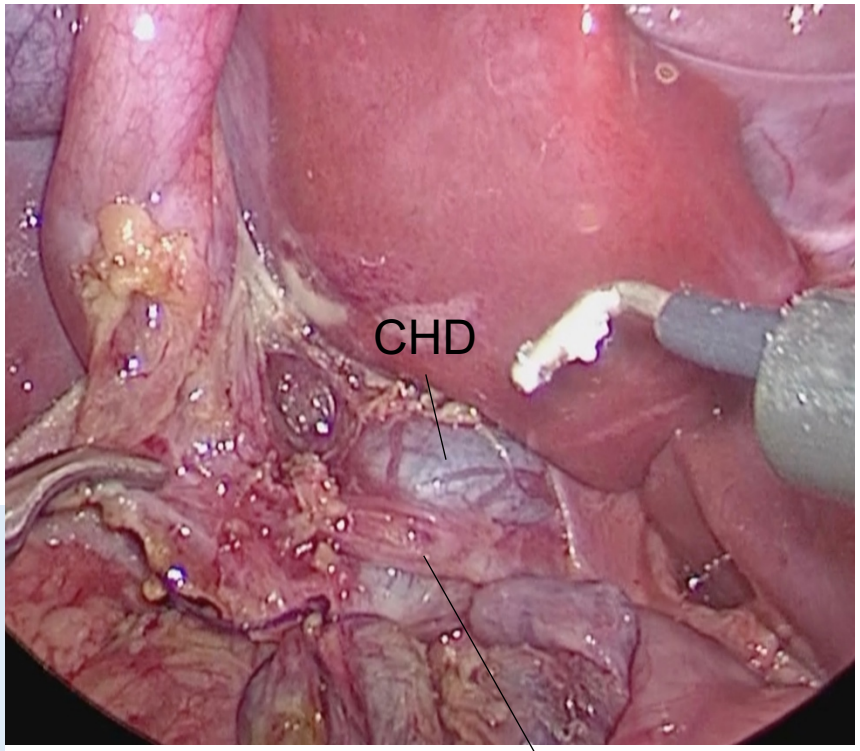
Urushihara et al. 2015

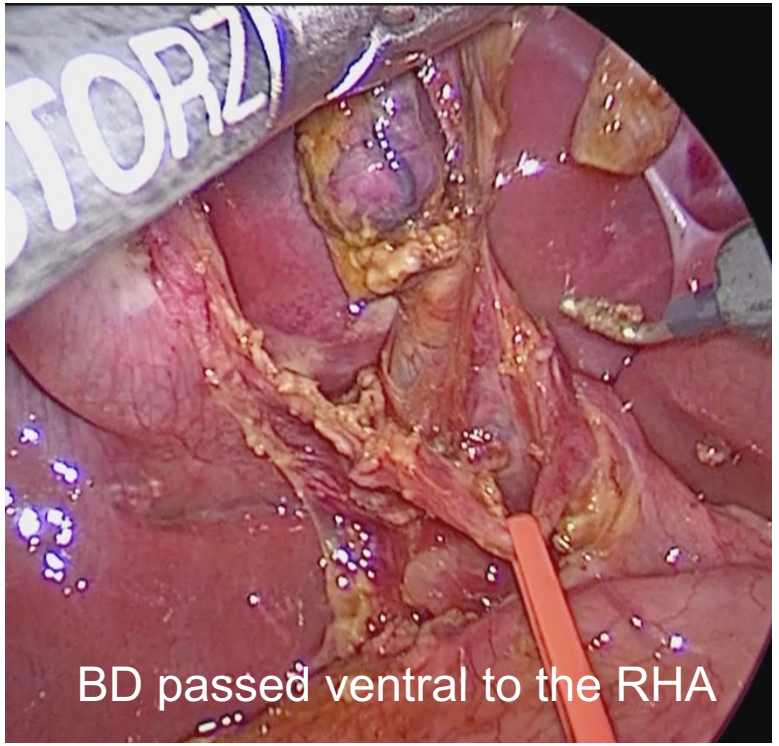
Excessive dissection impairs blood flow, resulting in anastomotic stricture.

Right hepatic artery runs ventral to BD

Case 23

A 4-year-old boy
Type Ia CBD





RHA ventral to CHD
Japanese (normal controls): 10% [1)]
Japanese (adult PBM): 21% [1)]
Japanese (pediatric PBM): 26% [2)]
Chinese (pediatric PBM): 17% [3)]

Hepaticojejunostomy must be performed ventral to the RHA.
Otherwise, bile outflow can be impaired because the anastomosed jejunum pulls the anastomosis ventrally.

1) Shinozaki K, Ajiki T, Matsumoto T, Yoshida Y, Murakami S, Okazaki T, Toyama H, Matsumoto I, Fukumoto T, Ku Y. Anatomical variations of liver blood supply in patients with pancreaticobiliary maljunction. *Surg Today.* 2016 Feb;46(2):169-75. doi: 10.1007/s00595-015-1118-2. Epub 2015 Feb 5. PMID: 25649536.

2) Takada S, Uchida H, Hinoki A, Shirota C, Sumida W, Tainaka T, Makita S, Takimoto A, Nakagawa Y, Maeda T. Variations of the hepatic artery and bile duct in patients with pancreaticobiliary maljunction: Impact on postoperative outcomes. *J Hepatobiliary Pancreat Sci.* 2023 Nov;30(11):1241-1248. doi: 10.1002/jhbp.1381. Epub 2023 Oct 25. PMID: 37876298.

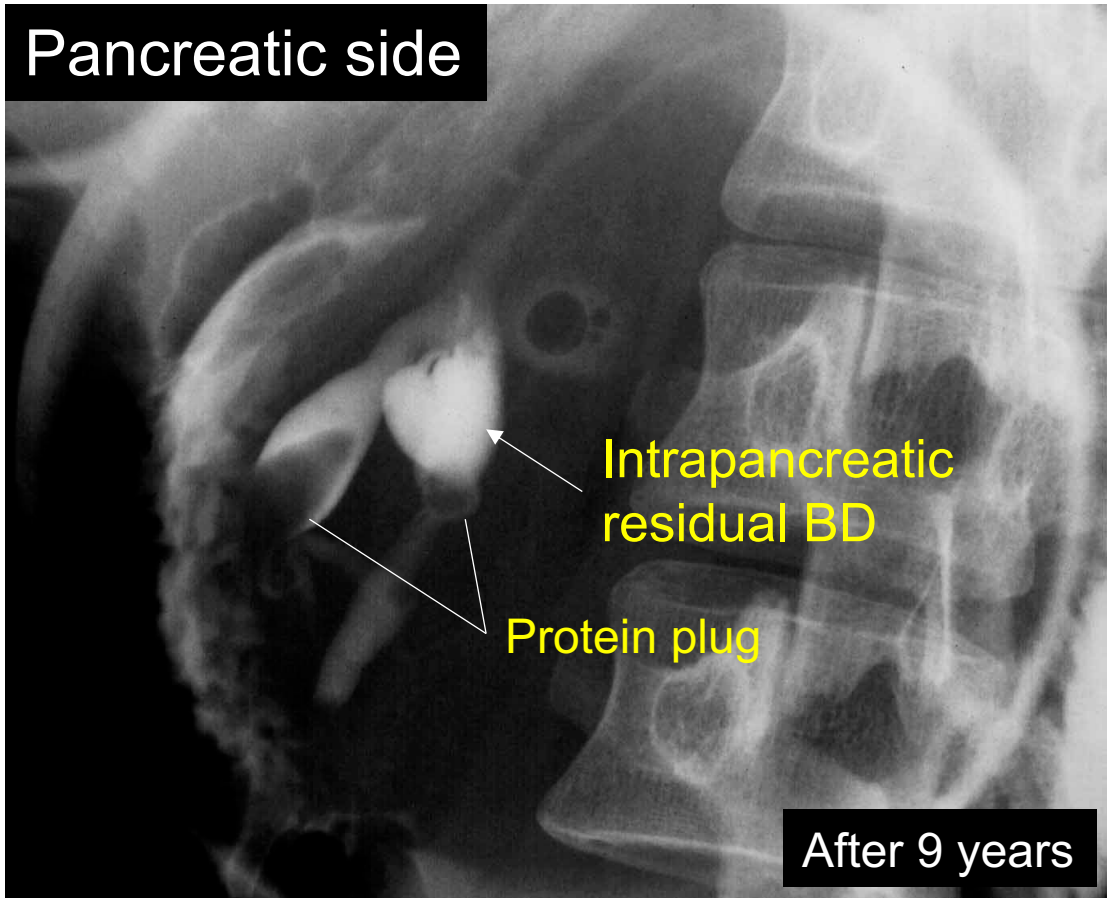
3) Guo WL, Wang J. Preoperative one-stop magnetic resonance imaging evaluation of the pancreaticobiliary junction and hepatic arteries in children with pancreaticobiliary maljunction: a prospective cohort study. *Surg Today*. 2021 Jan;51(1):79-85. doi: 10.1007/s00595-020-02077-5. Epub 2020 Jul 12. PMID: 32656698.

Basics

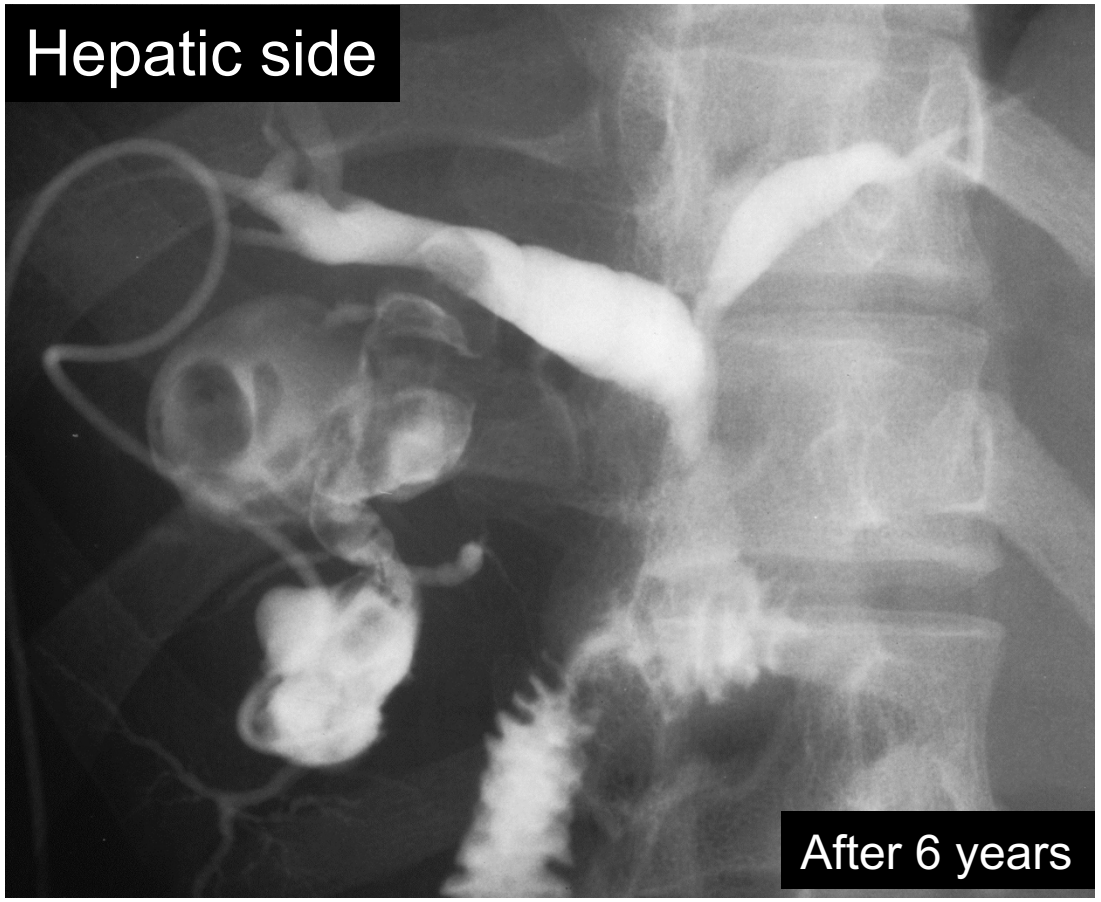
Postoperative complications

First version, September 2022; Revised, September 2026

Long-term complications after BD resection



A residual intrapancreatic BD can cause protein plugs, stones, or carcinogenesis. Complete excision of the intrapancreatic BD is necessary.



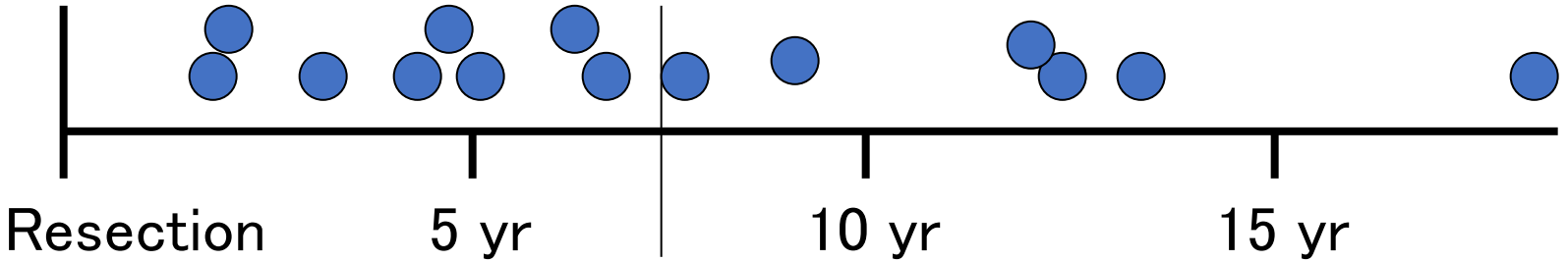
Intrahepatic gallstones and/or cholangitis occur in 7% to 8% of cases several years after BD resection.

Outcomes of residual intrapancreatic bile duct

Author	Residual intrapancreatic BD Age at resection	Pancreatitis / Pancreatic stones Postoperative years	Cancer Postoperative years	Journal / Year
Xia (Beijing)	41 cases 42 (25–57)	18 (44%)	6 (15%) 12 (0.5–22)	*Surgery* **2016**
Fan (Shanghai)	78 cases Adults	51 (65%) 7 (5–9)	11 (14%) 4–11	*J Int Med Res* **2018**

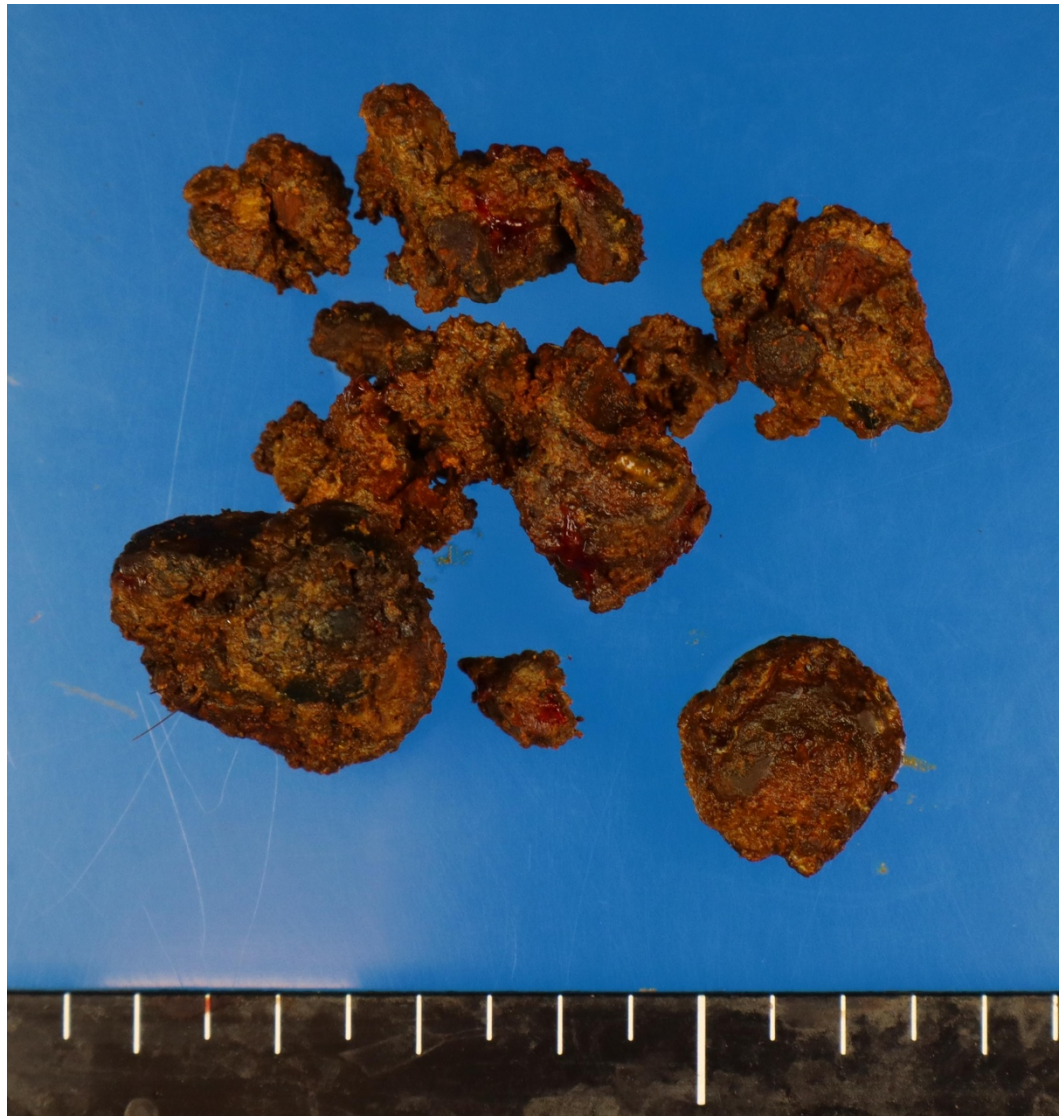
Time from resection to intrahepatic gallstone formation

Pediatric cases (n=14)



Mean: 7.3 yr

Postoperative gallstones are brown pigment stones

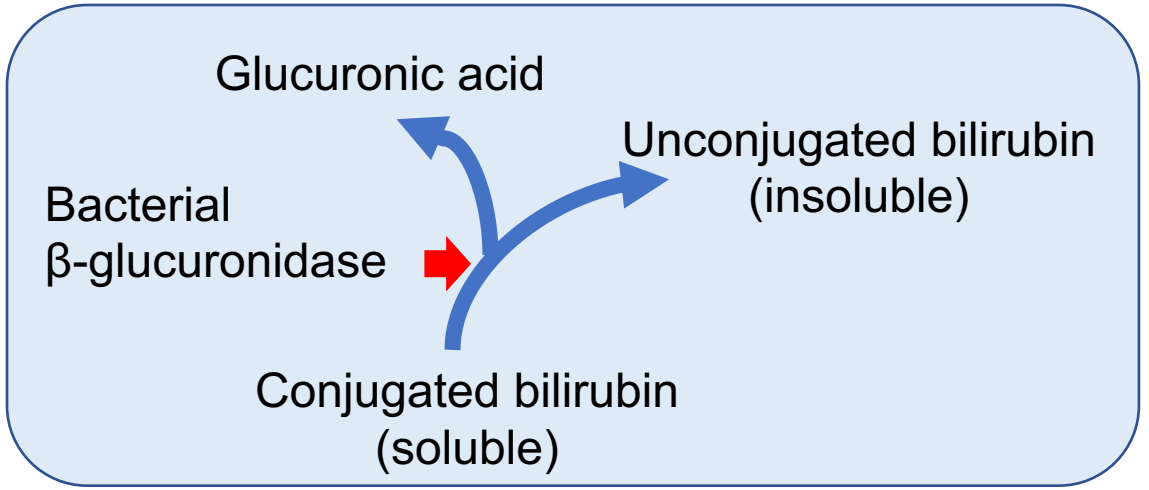


Composition:

Bilirubin calcium + calcium fatty acid

Bacterial β-glucuronidase and phospholipase A1

Bile stasis and bacterial infection



Risk factors for postoperative hepatolithiasis

- Todani type IV-A
- Hepaticojejunal anastomotic stricture
- Congenital stenoses overlooked during BD resection

Todani type IV-A



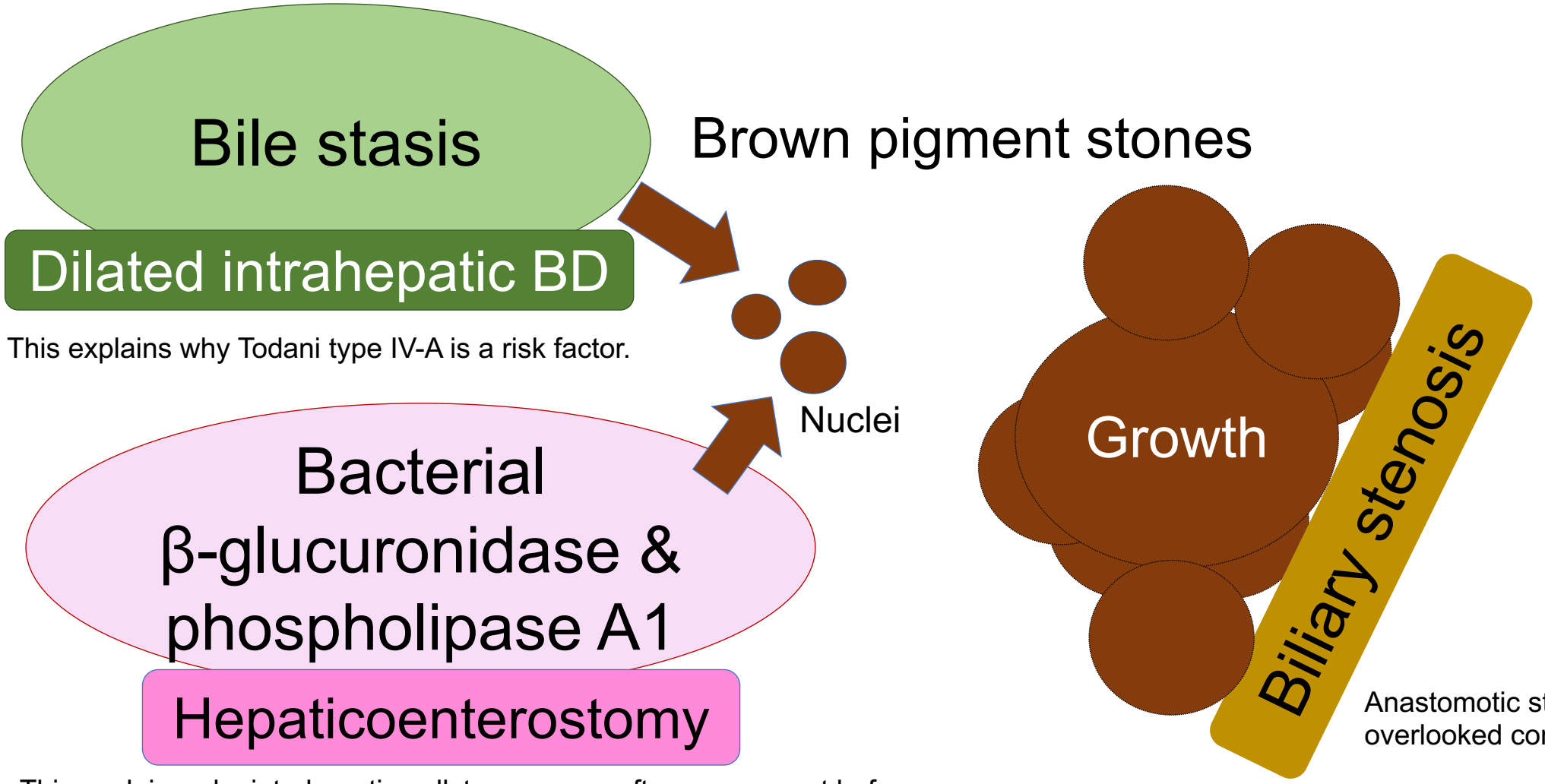
Anastomotic stricture
in non-dilated PBM



Congenital septal stenosis



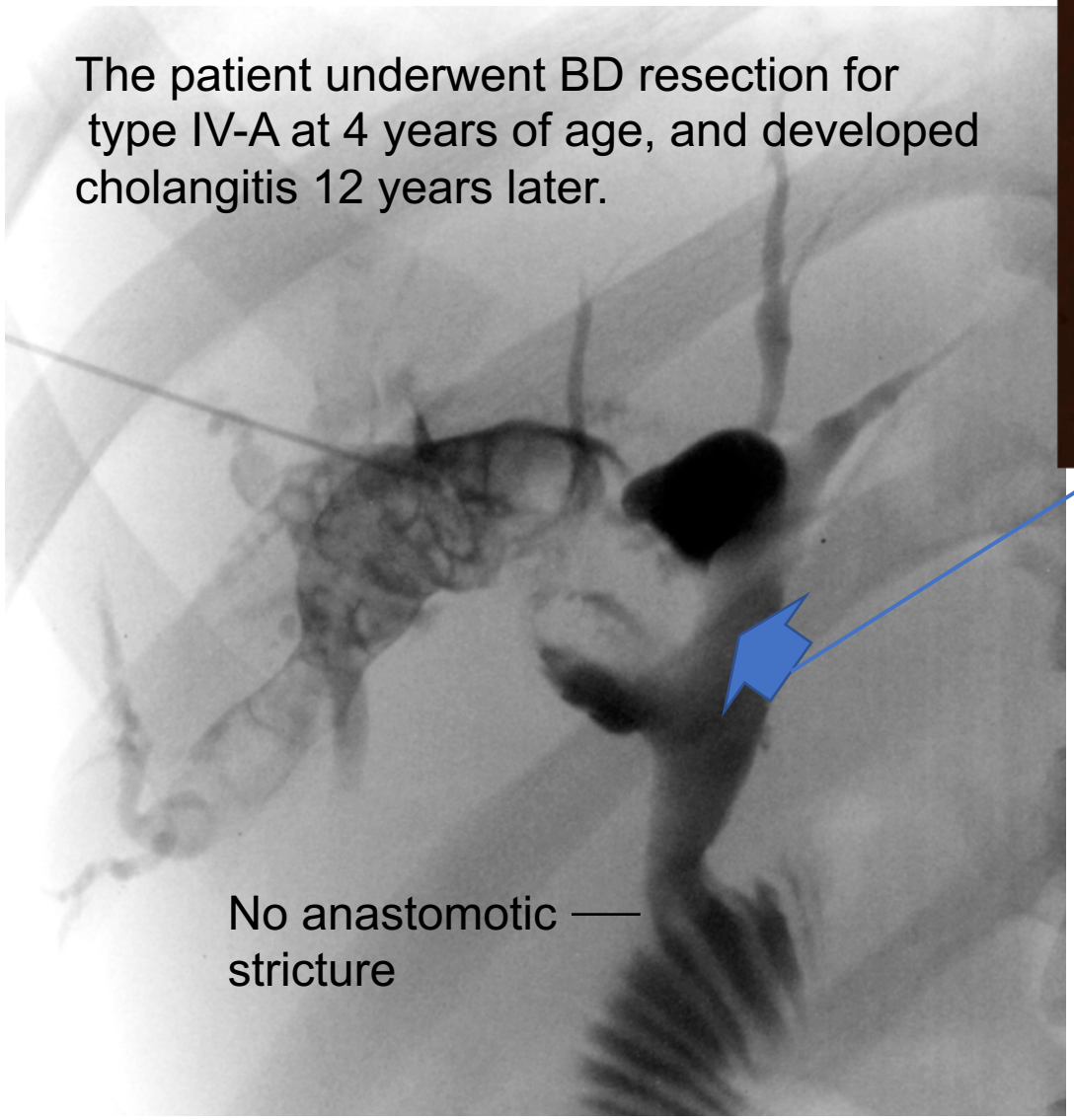
Mechanism of postoperative gallstone formation

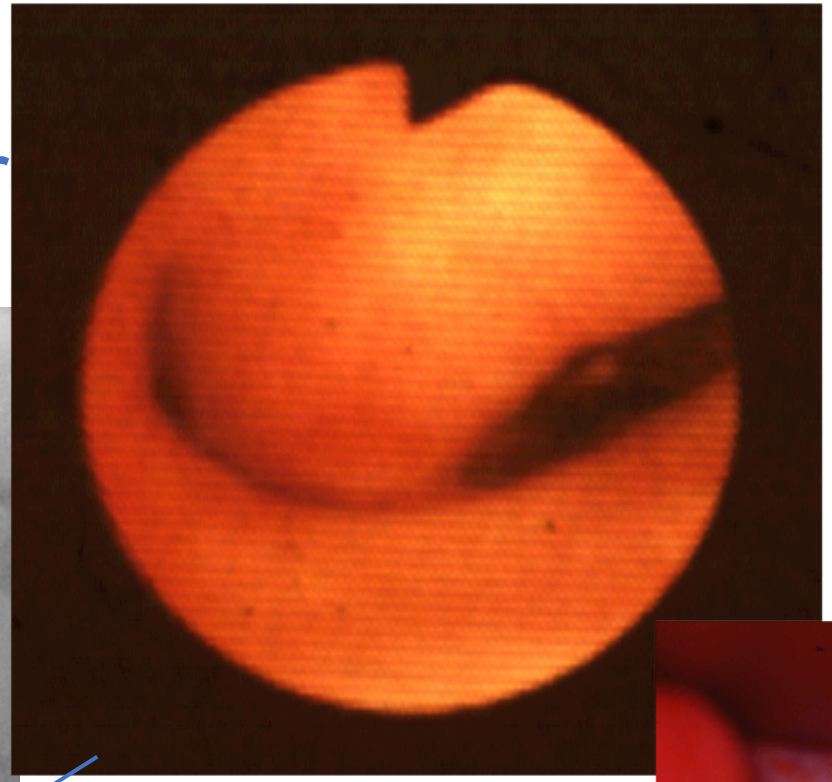


Stone removal alone is insufficient for a permanent cure;
relieving stenosis or resecting the dilated BD (hepatectomy) is required.

Case 24 Intrahepatic gallstones after BD resection

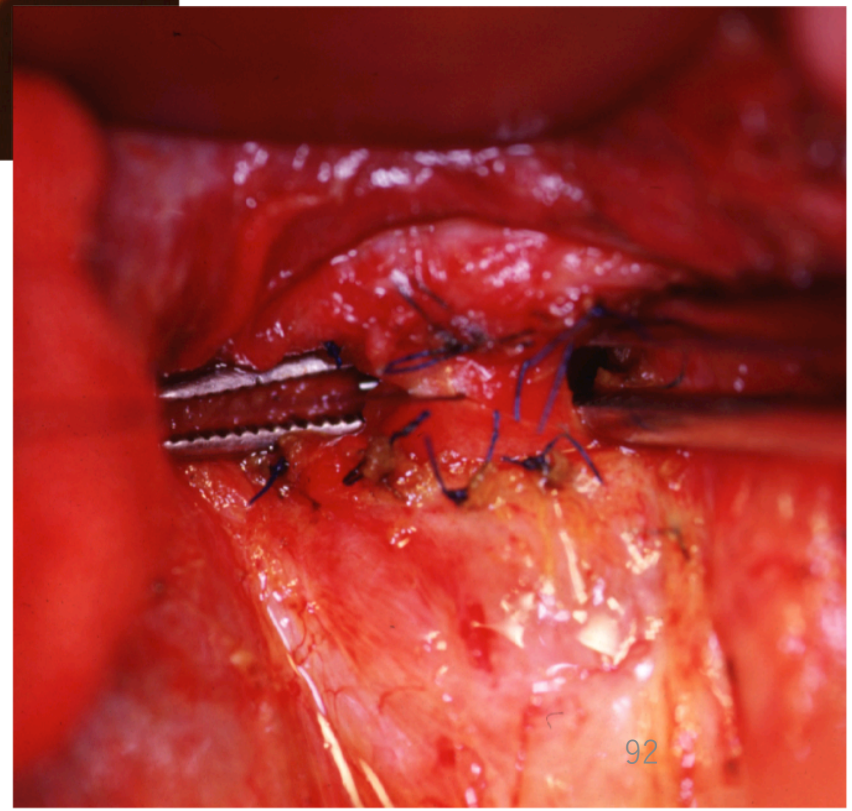
The patient underwent BD resection for type IV-A at 4 years of age, and developed cholangitis 12 years later.





Apparent separate orifices on endoscopy actually merged into a single duct proximal to the stenosis.

Septal stenosis trapping gallstones

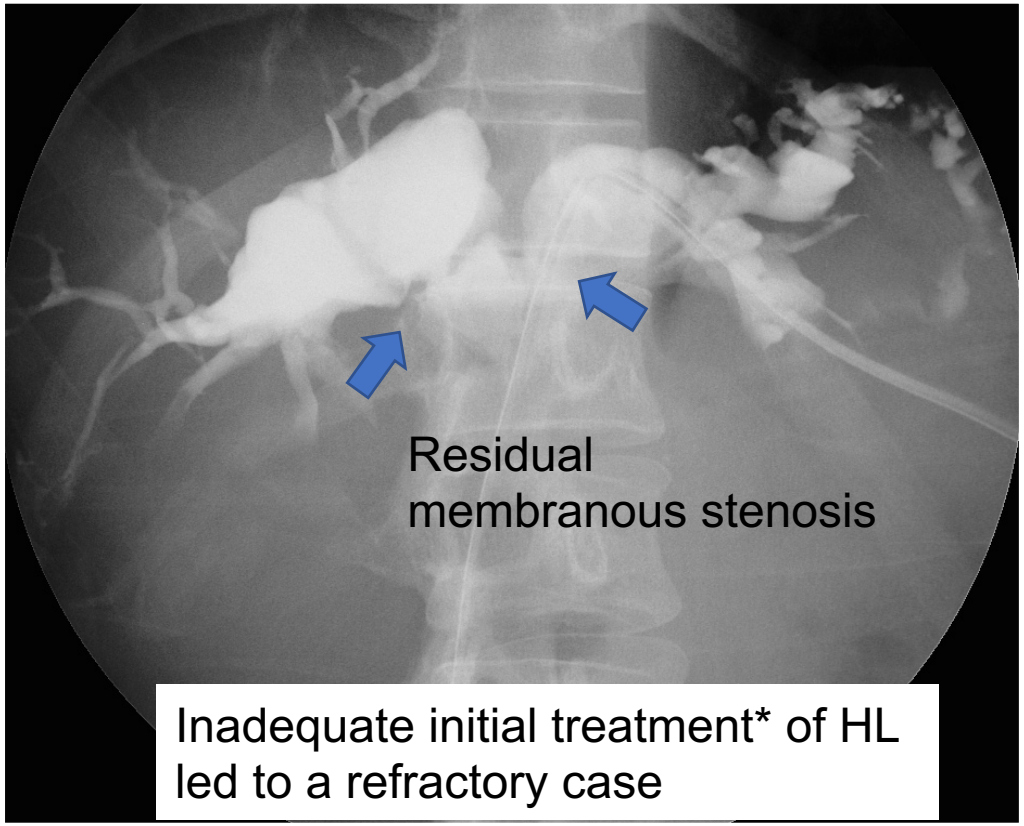


Case 25

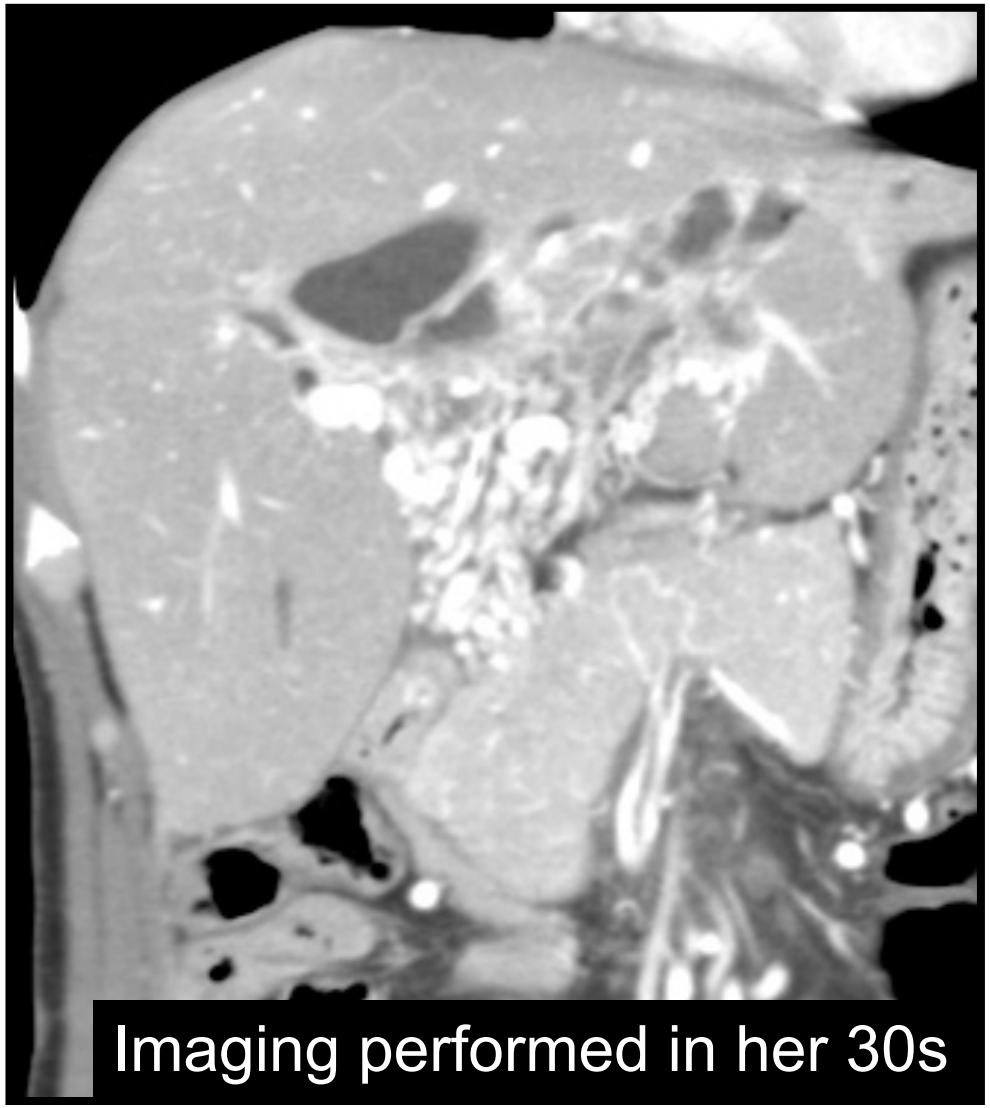
Refractory cholangitis will occur

BD resection of type IV-A CBD at 1 year of age.
↓
In her 20s, hepatolithiasis (HL) developed, and PTCS* was performed.
↓
Mild cholangitis recurred repeatedly, but she did not seek medical advice.
↓
In her 30s, she had severe cholangitis with recurrent HL.

Cavernous transformation of the portal vein



Inadequate initial treatment* of HL led to a refractory case



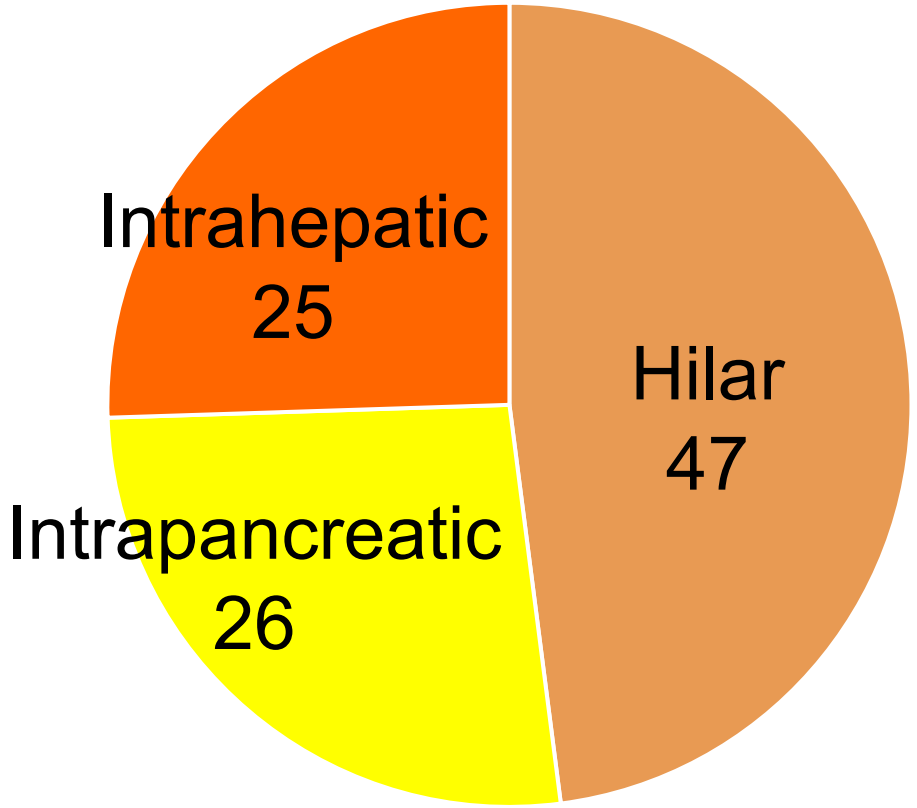
*PTCS, percutaneous transhepatic cholangioscopic lithotomy

Long-term postoperative complications: Carcinogenesis in the residual BD

Higher incidence even after BD resection

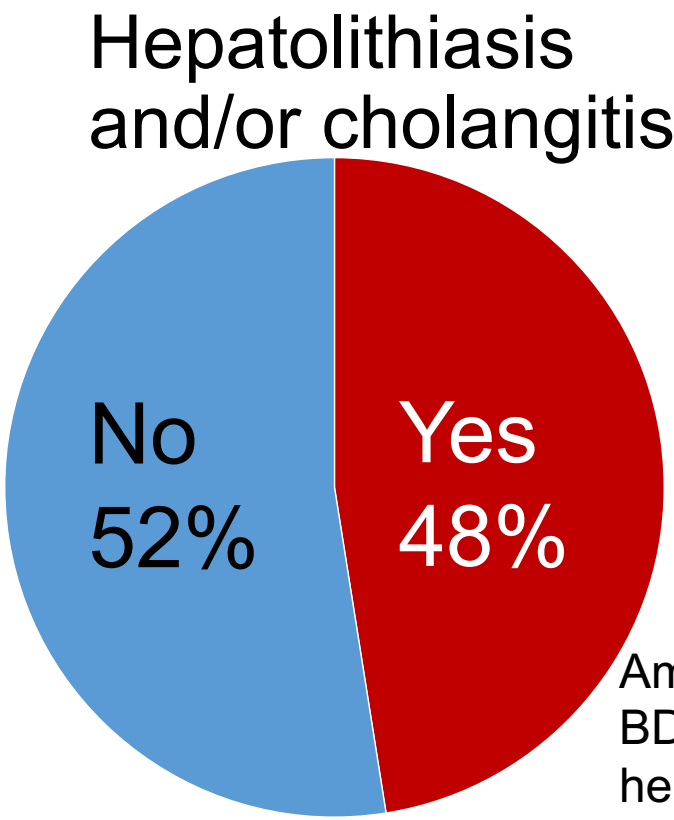
- 107 cases collected from the literature
- Carcinogenesis rate: 2.0%
- Mean interval to carcinogenesis:
  12 postoperative years (mean age: 47 years)

Kamisawa T, Kaneko K, Itoi T, Ando H. Pancreaticobiliary maljunction and congenital biliary dilatation. *Lancet Gastroenterol Hepatol.* 2017 Aug;2(8):610-618. doi: 10.1016/S2468-1253(17)30002-X. PMID: 28691687.



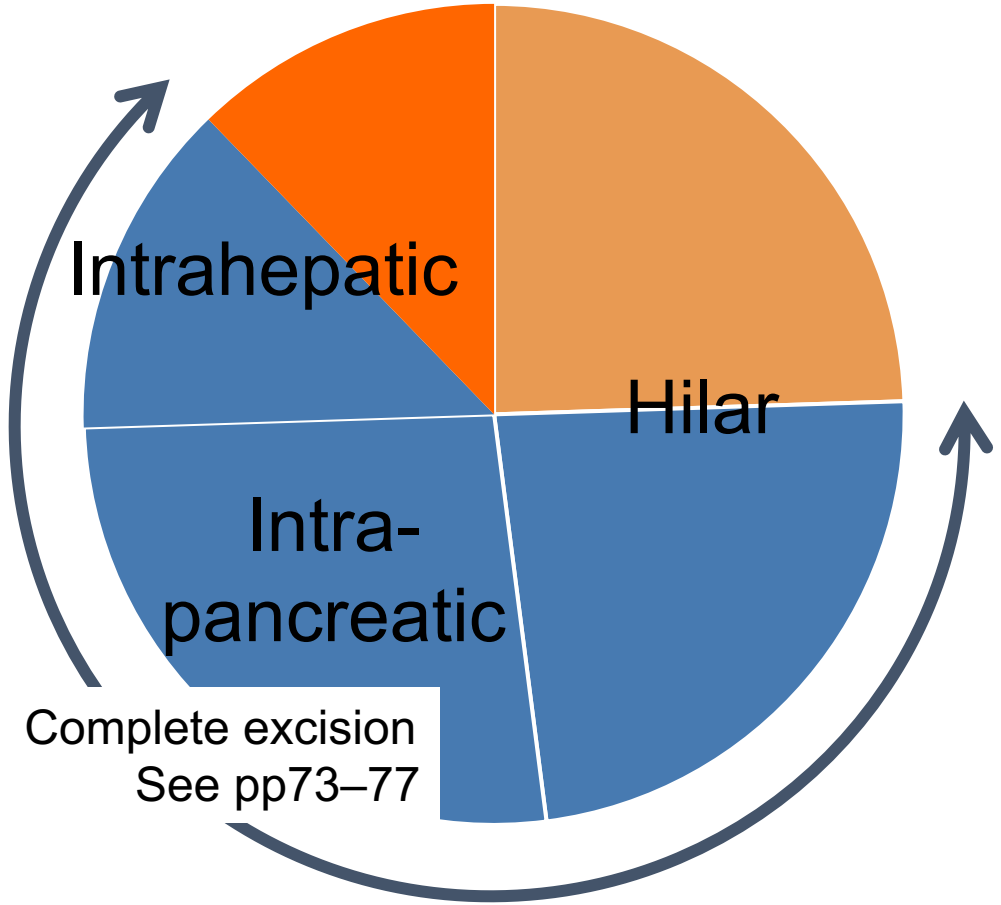
cf. US Survey
Biliary cancer after BD resection: 13/394 (3.3%)
Pediatric cases 4/135 (3.0%) / Adult cases 18/259 (6.9%)
Soares et al. *JAMA Surg* 2015

Many cases of residual BD cancer could have been prevented



Among 80 residual BD cancers on the hepatic side

Approximately half of patinets with postoperative residual BD cancer present with hepatolithiasis and/or cholangitis. Anastomotic stricture or overlooked congenital stenosis leads to hepatolithiasis and/or cholangitis, ultimately resulting in carcinogenesis.

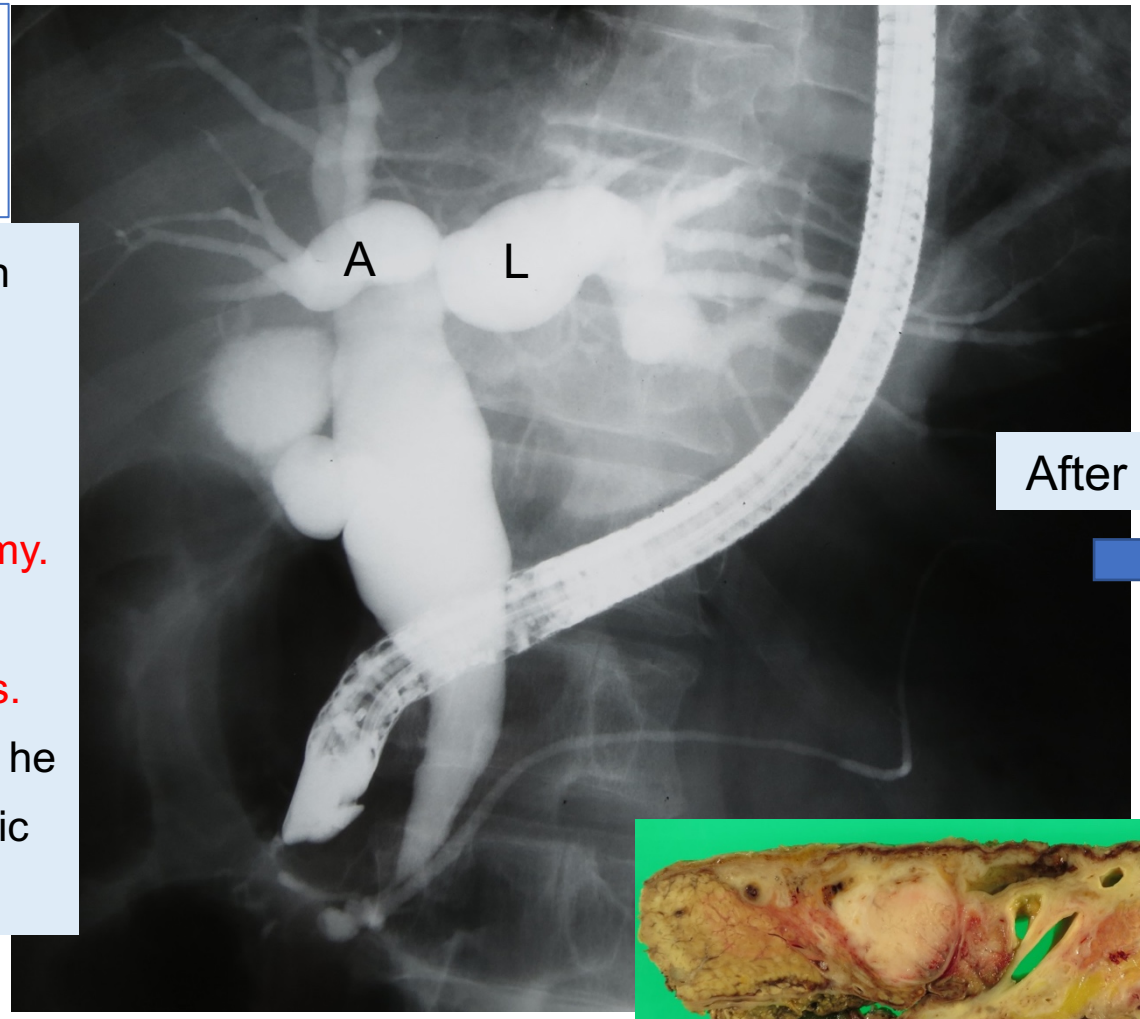


Appropriate procedures could have prevented these cancers.

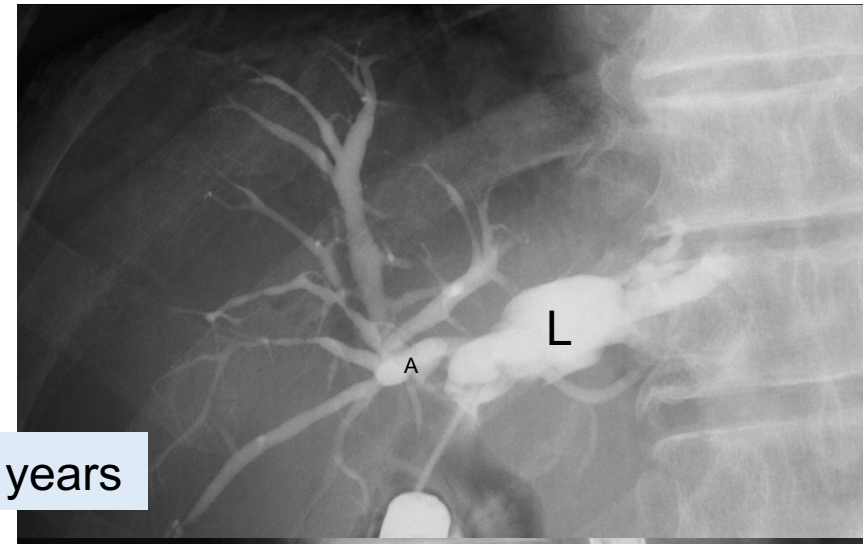
Intrahepatic BD cancer after BD resection

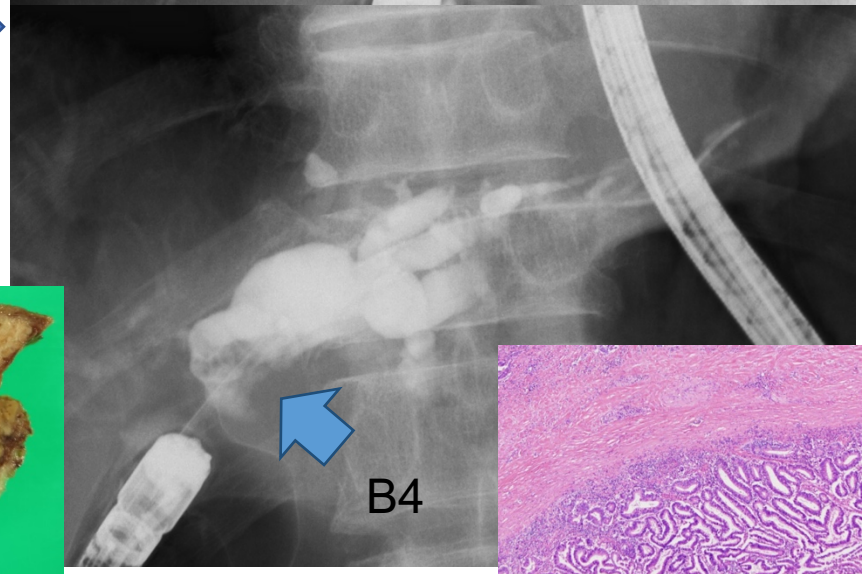
Case 26

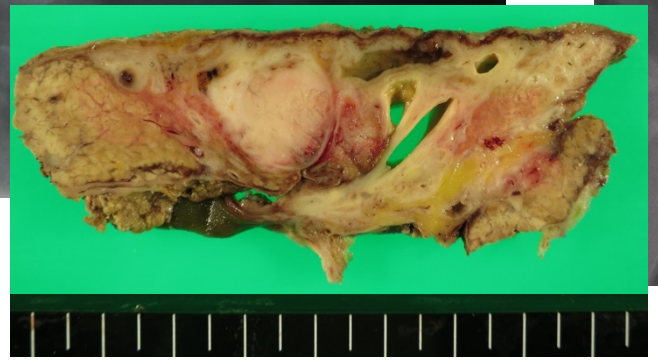
A male in his 60s with type IV-A CBD underwent BD resection with hepaticoduodenostomy. Four years late, he developed cholangitis. Nine years after that, he developed intrahepatic BD cancer in B4.



After 13 years







A, anterior section duct; L, left hepatic duct. Dilatation of the anterior segment diminished postoperatively.

Basics

High confluence of the pancreaticobiliary ducts

First version, September 2024; Revised, September 2026

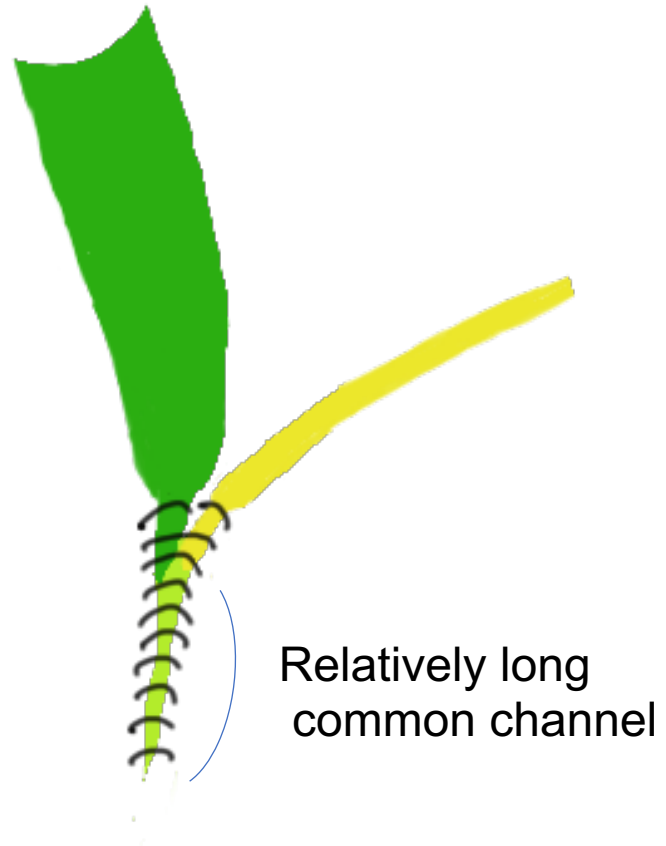
What is High confluence of pancreaticobiliary ducts?

Definition

The length of the common channel is **6 mm or longer**, although the **sphincter effect reaches the pancreaticobiliary junction**.

Kamisawa T, et al. *Pancreatology*. 2002;2:122-8

Normal — **High confluence** — PBM



High confluence is a **transitional type between normal and PBM**, and frequently causes **pancreatobiliary reflux**. Patients with high confluence often are known to have a high incidence of gallbladder cancer, similar to those with PBM. Sai et al. proposed a similar concept of **"occult pancreatobiliary reflux"**, defined as a normal confluence that still yields **pancreatobiliary reflux.**

Sai JK et al. *Gastrointest Endosc.* 2003:57:364-8

Spectrum from normal to PBM

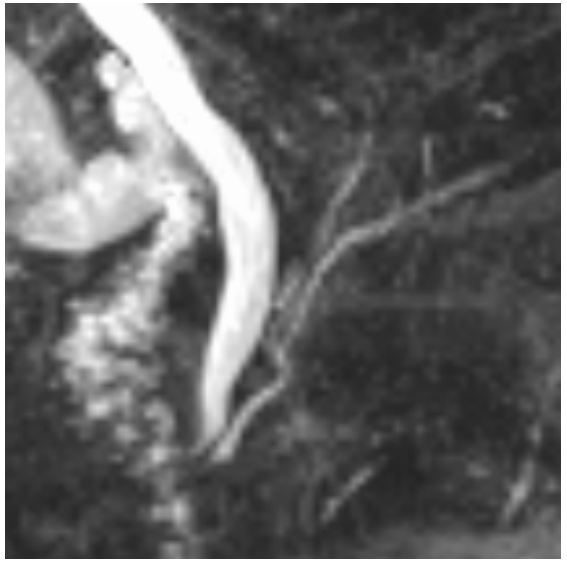
Normal

Occult pancreatobiliary reflux

High confluence

PBM

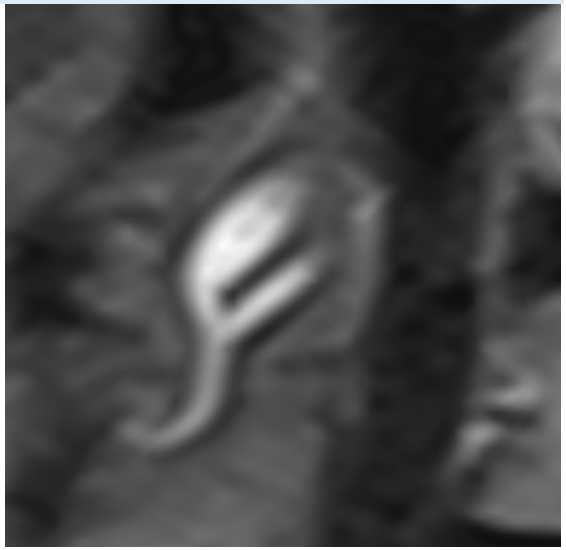
The sphincter function affecting the confluence



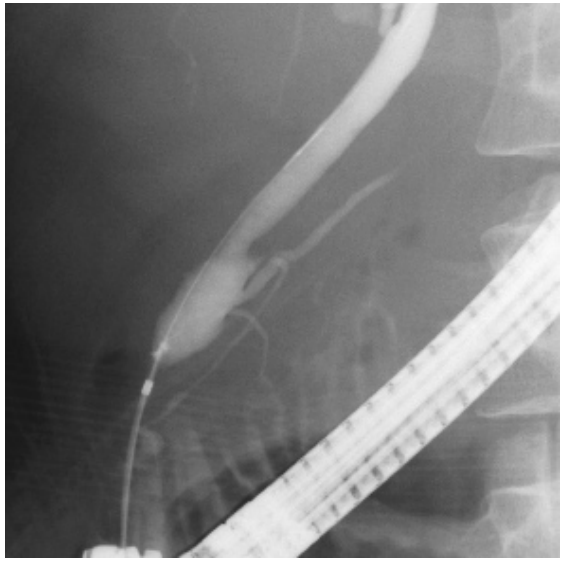
Separate openings



Common channel formation
Cholangiography occasionally visualizes the PD.



Relatively long common channel



Abnormally long common channel

High confluence is a transitional type

High confluence and PBM among patients undergoing ERCP

	High confluence	Dilated-type PBM	Non-dilated-type PBM	Controls
n	50	30	33	2,867
Sex: Male	27	7	9	1,480
Female	23	23	24	1,387
Mean age, years	61.4 (32–75)	49.8 (9–78)	55.8 (30–77)	57.6 (16–92)
Gallbladder cancer	6 (12%)	4 (13%)	22 (67%)	72 (3%)
Bile duct cancer	1 (2%)	6 (20%)	0	48 (2%)

Kamisawa T, et al. *Pancreatology*. 2002;2:122-8

Advanced

Issues on Todani's classification

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

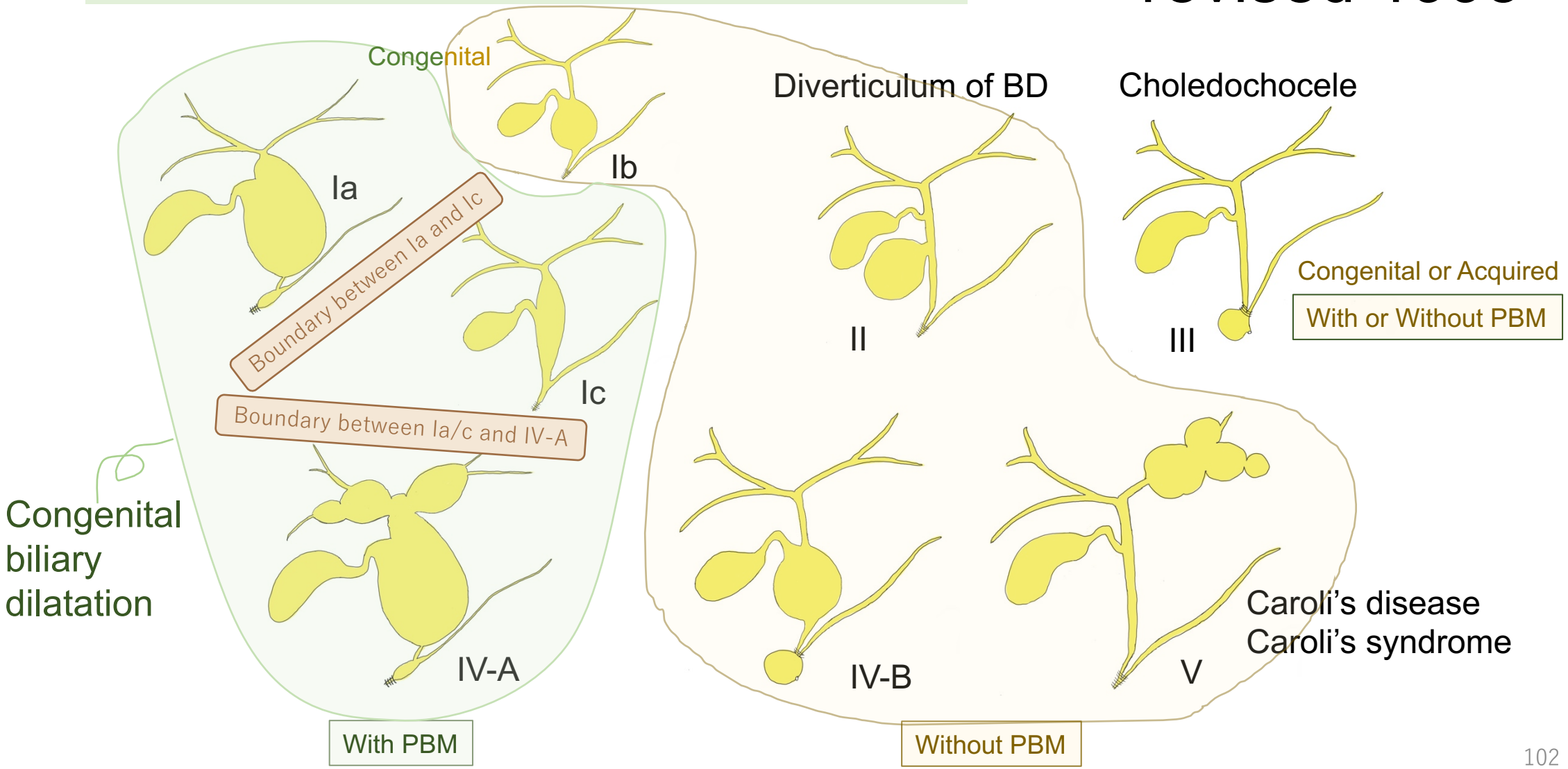
First version, September 2024

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

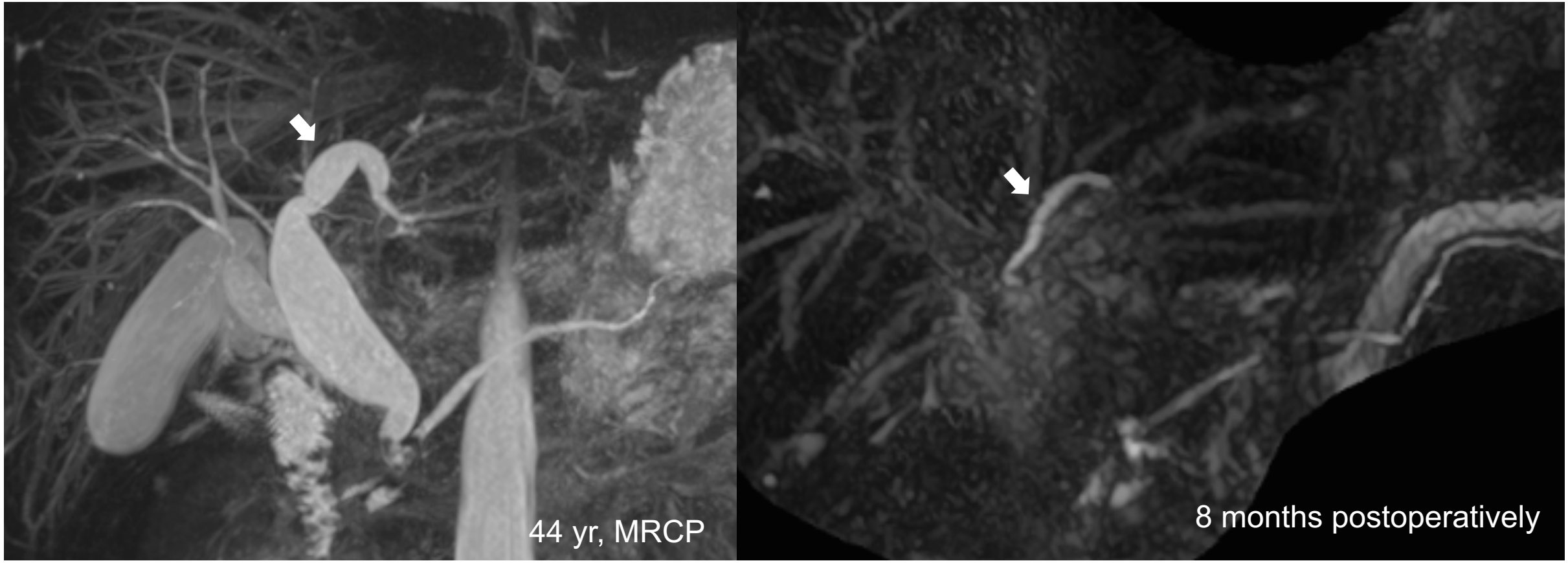
Todani T, Watanabe Y, Toki A, Morotomi Y.
Classification of congenital biliary cystic disease: special reference to type Ic and IVA cysts with primary ductal stricture.
J Hepatobiliary Pancreat Surg. 2003;10(5):340-4.
doi: 10.1007/s00534-002-0733-7. PMID: 14598133.

- **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.

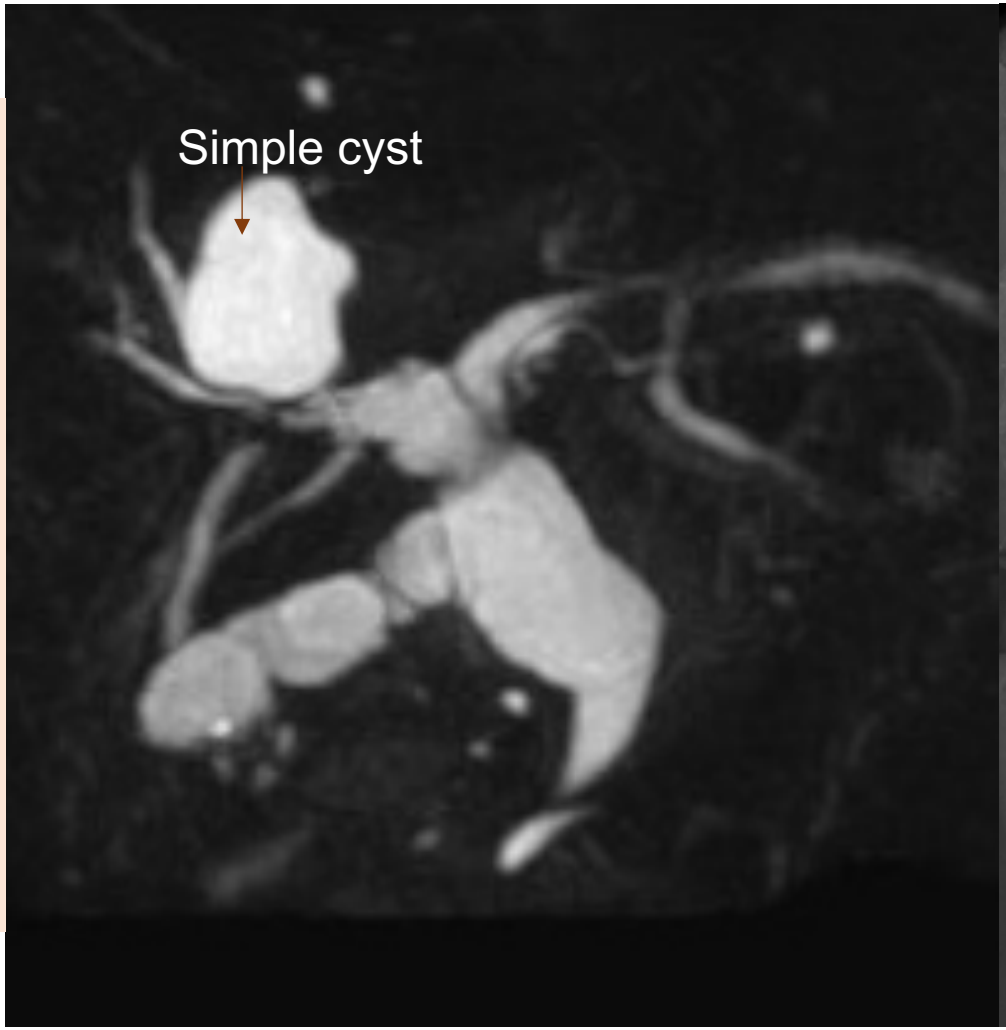
Case 27

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)

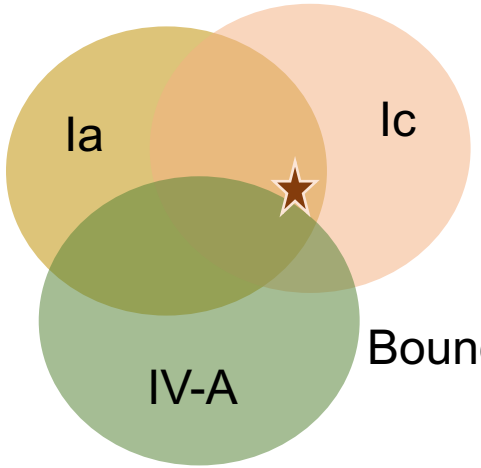




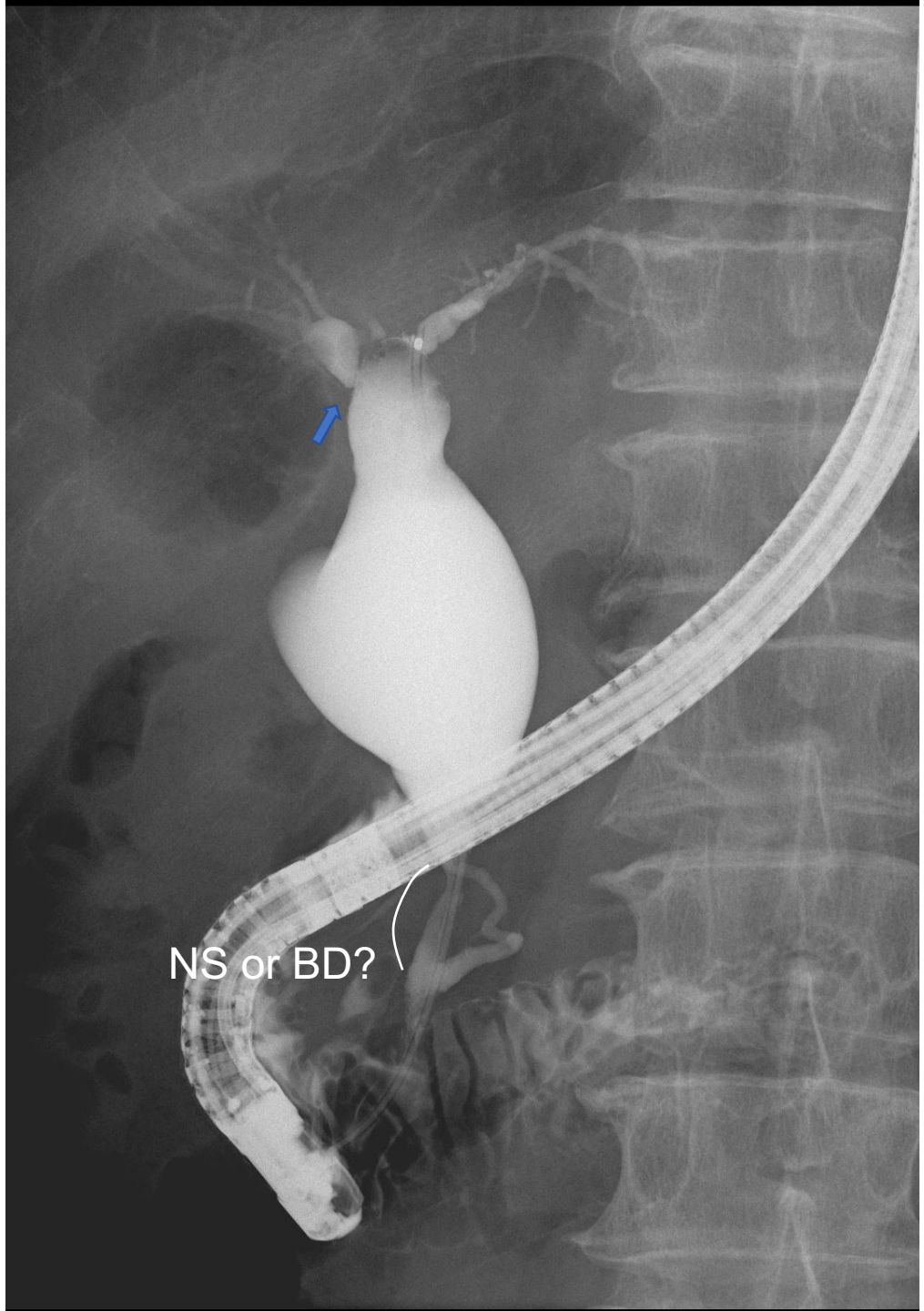
Case 28 Ic > Ia > IV-A

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.

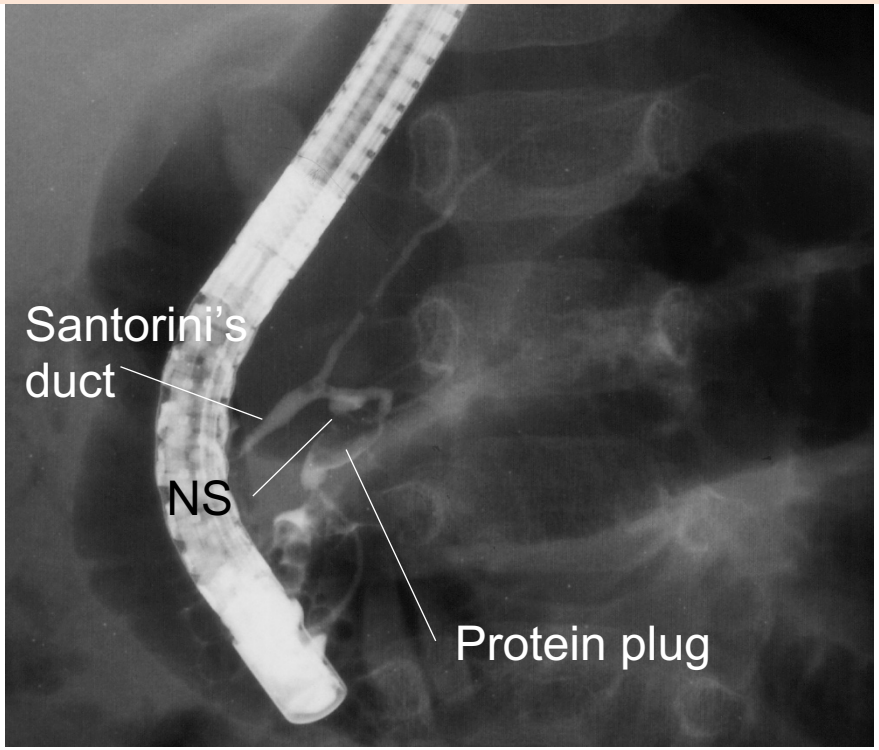


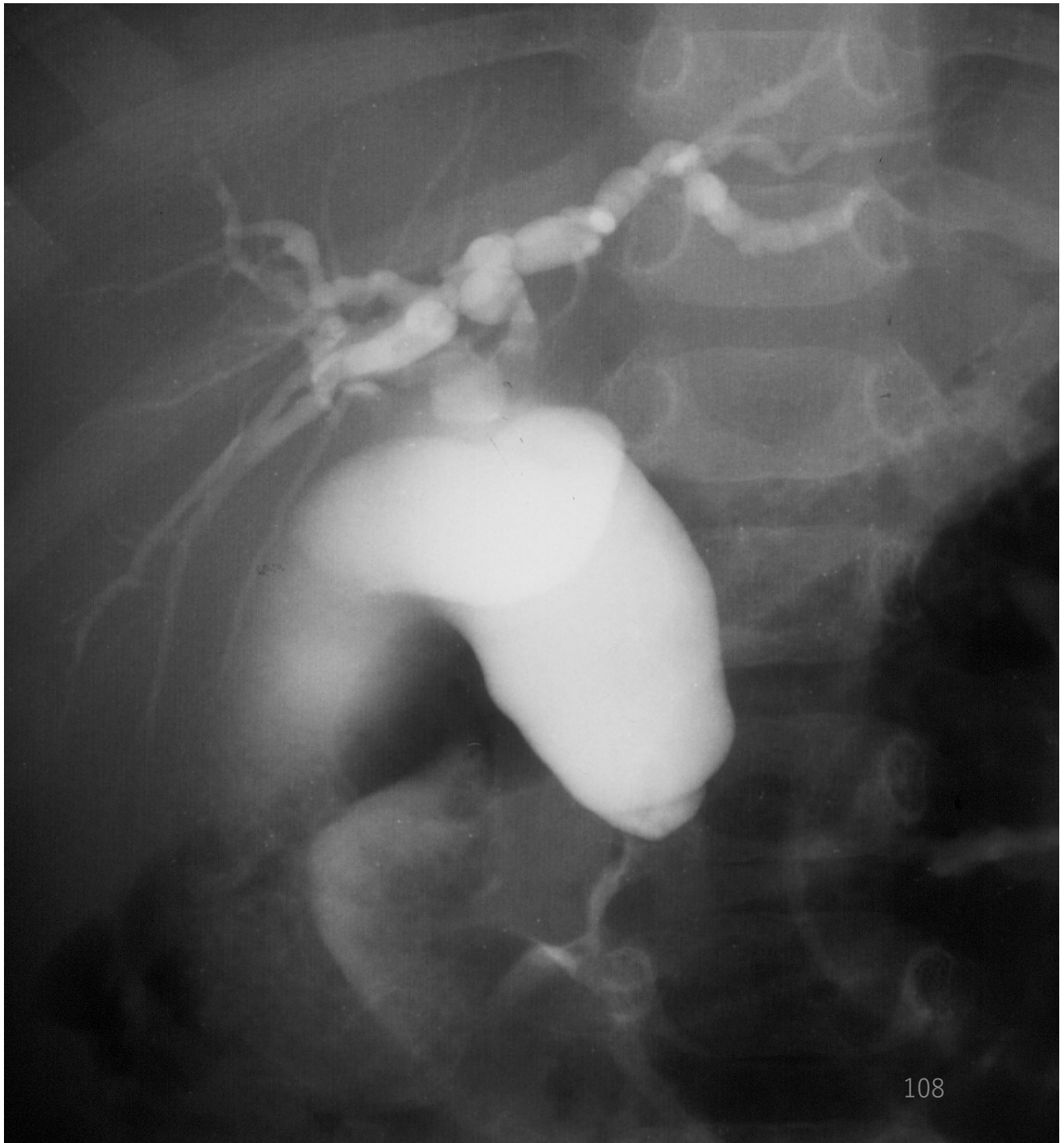
Case 29

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.

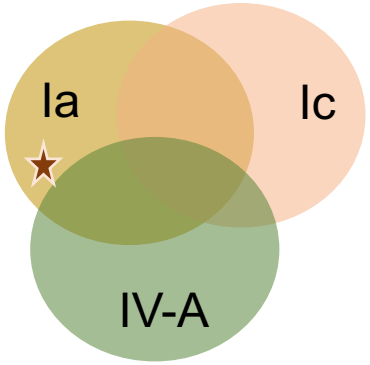


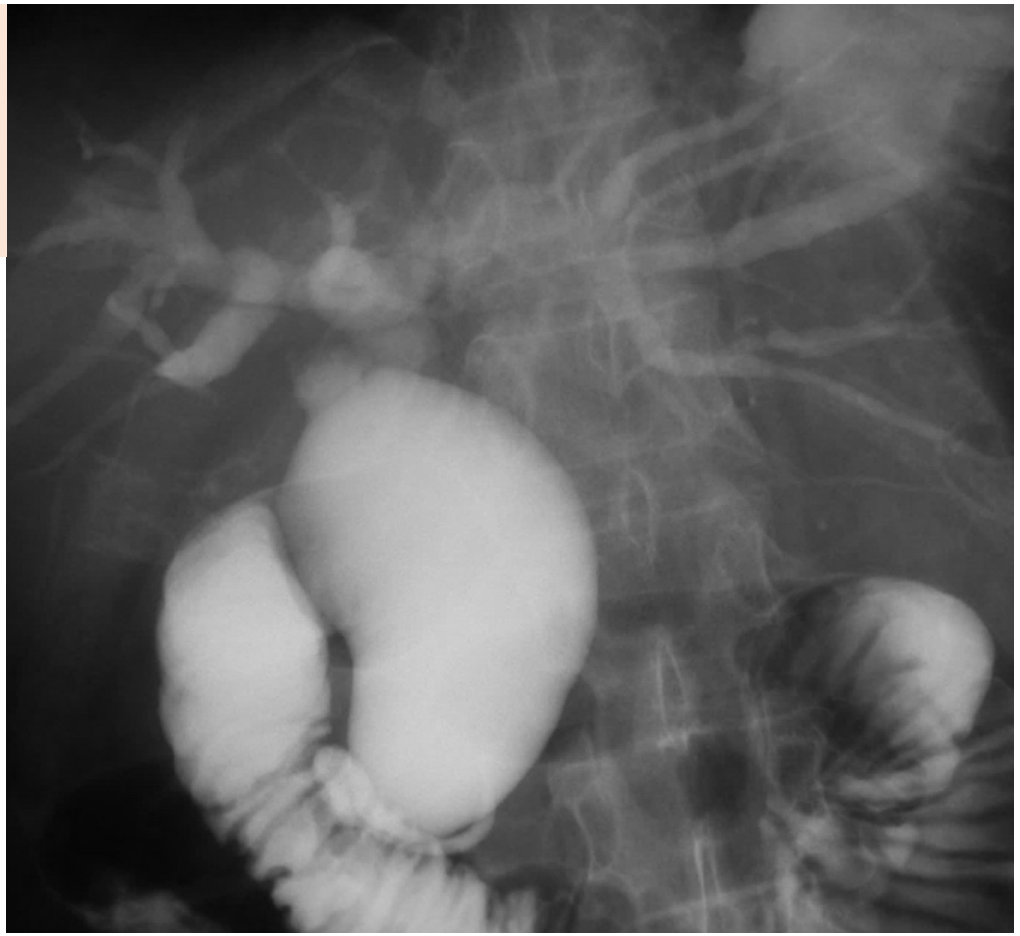


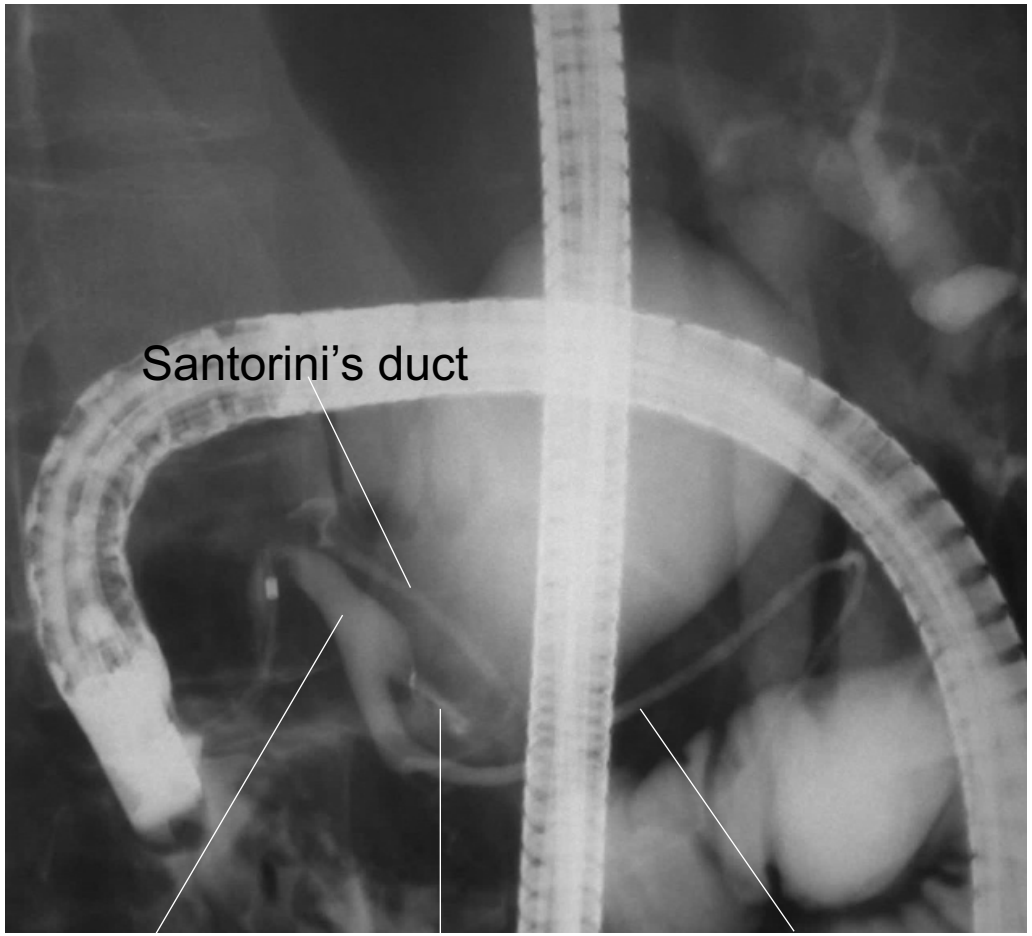
Case 30

Ia > IV-A

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







Case 31

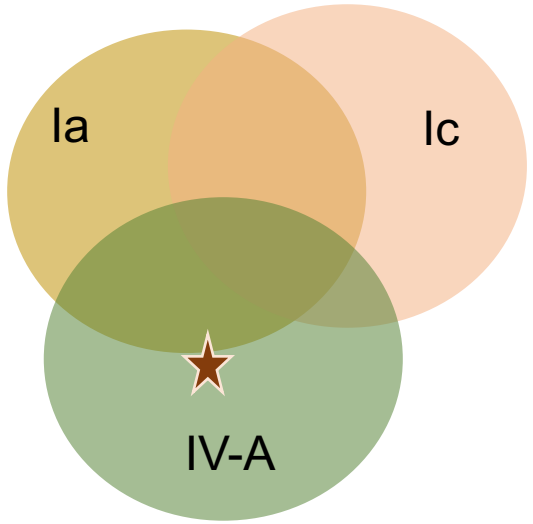
IV-A > Ia

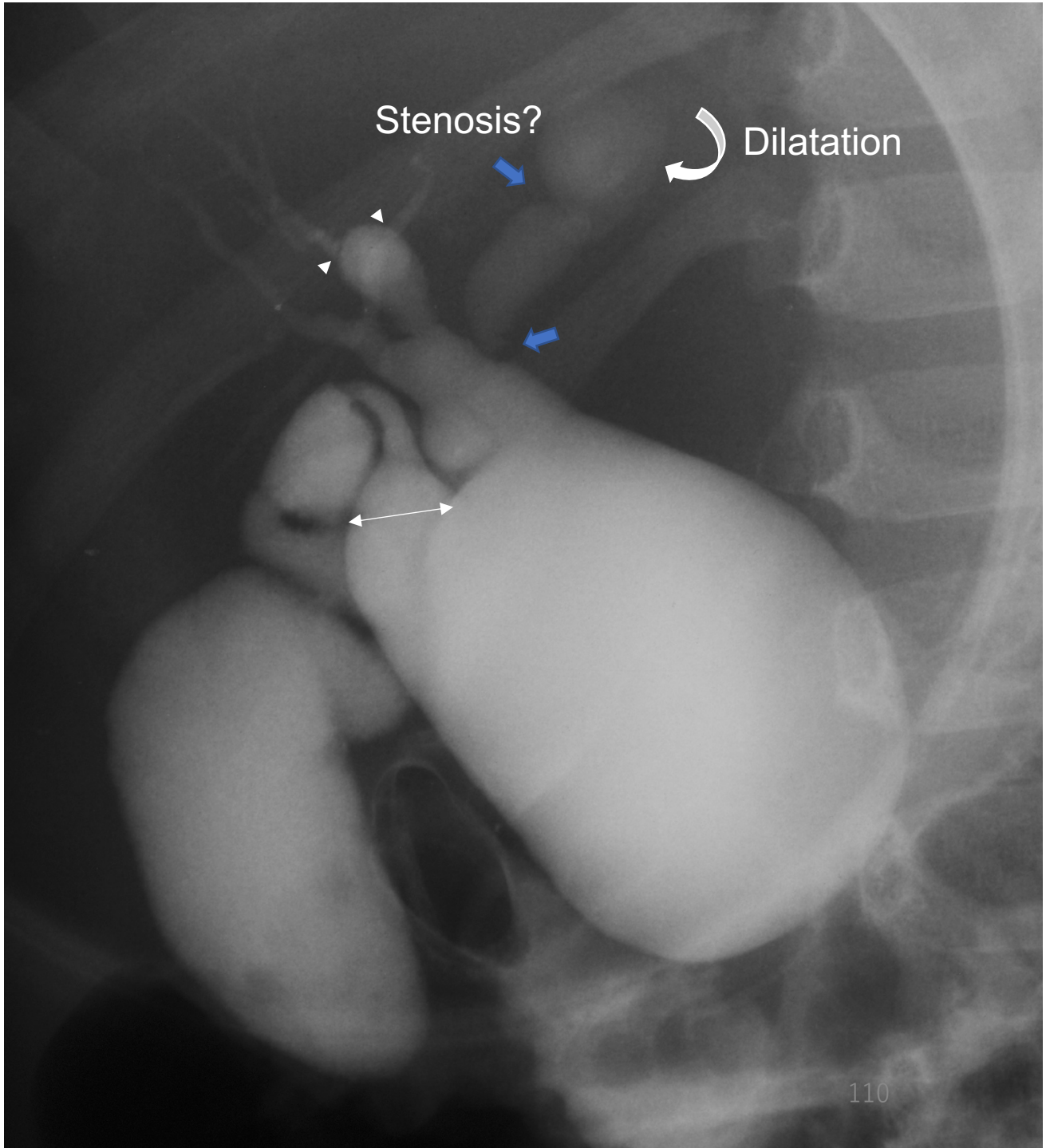
ERCP in a 2-year-old girl
Abdominal pain and vomiting

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

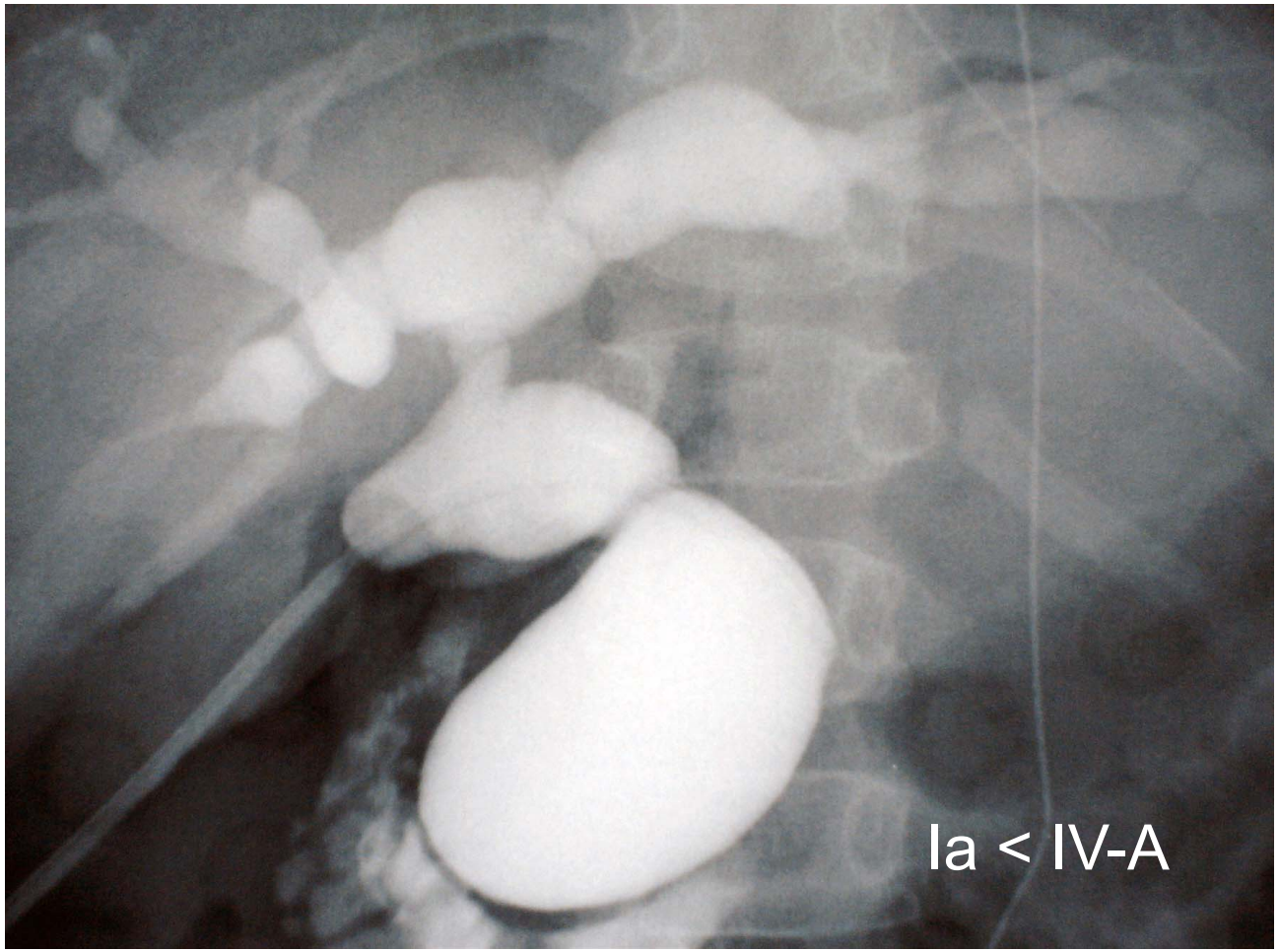
indicating type IV-A

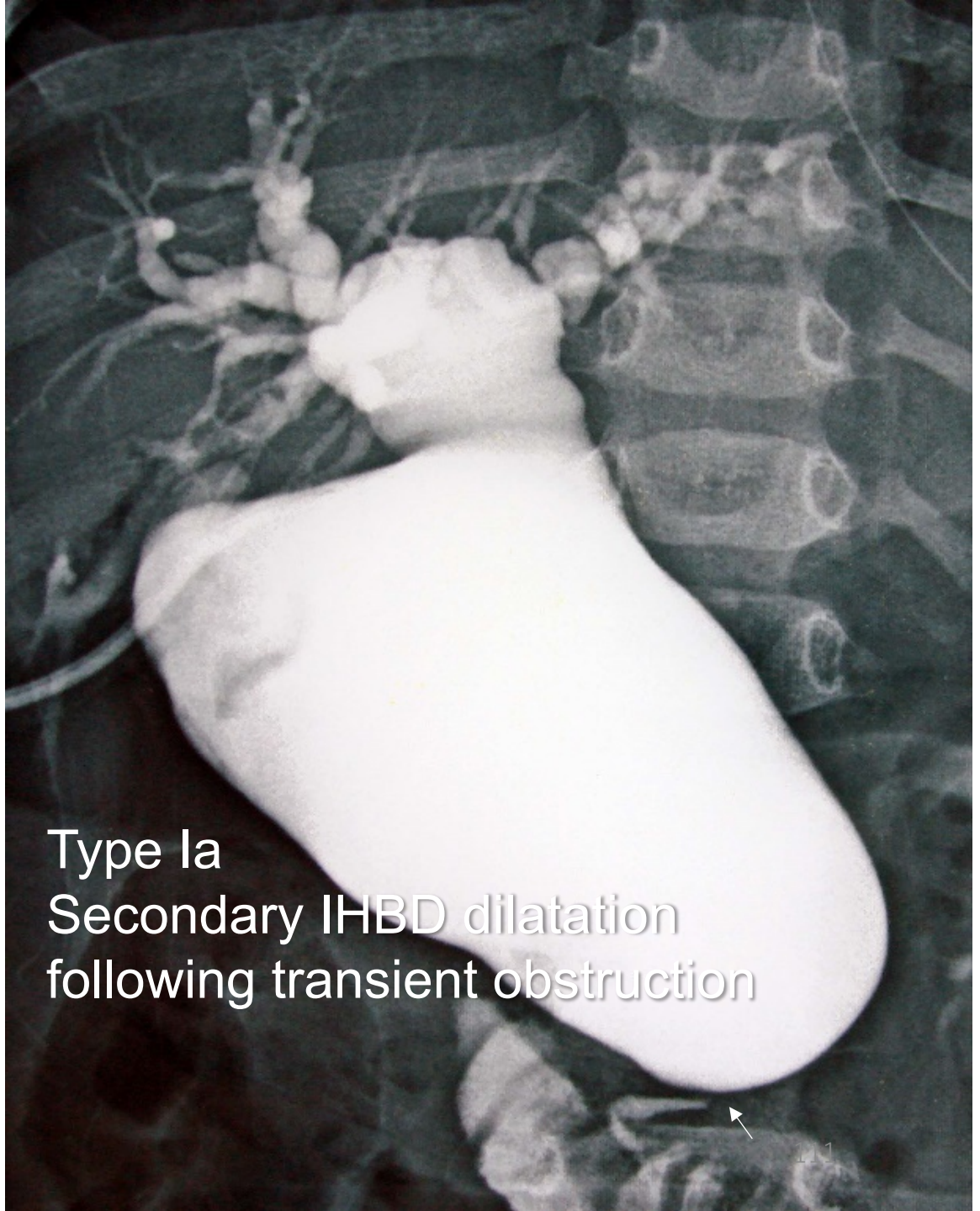
- Prominent dilatation at the base of the cystic duct (double-headed arrow)





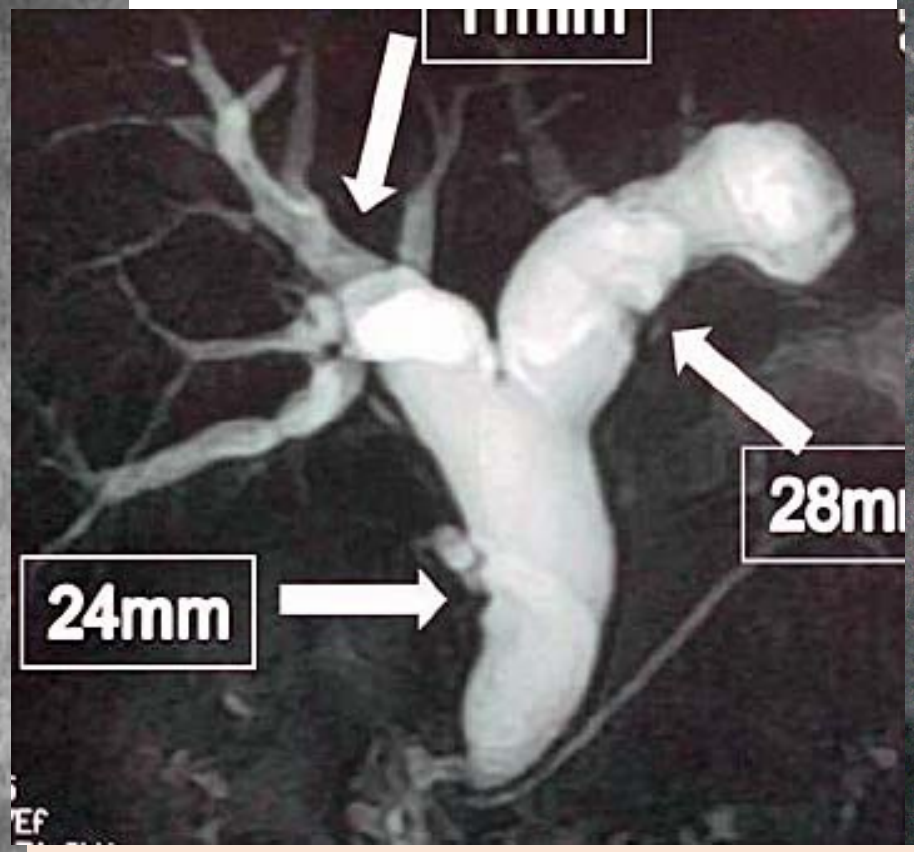
Two cases: Ia or IV-A?





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



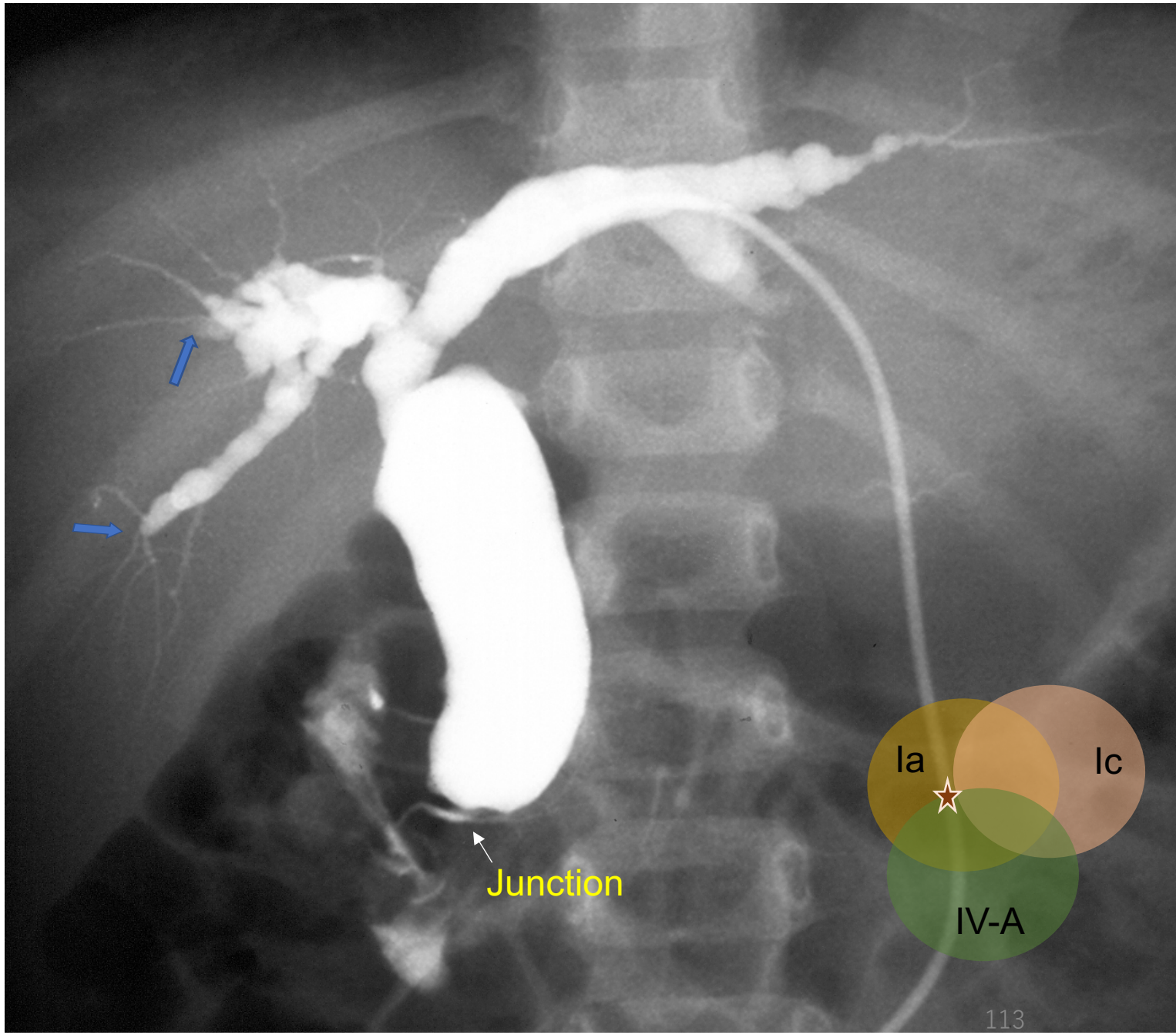
Case 32 Ia or IV-A?

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



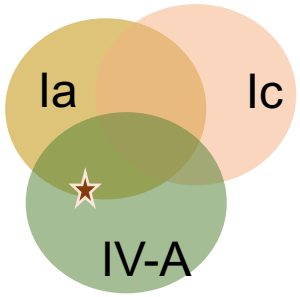
Case 33

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

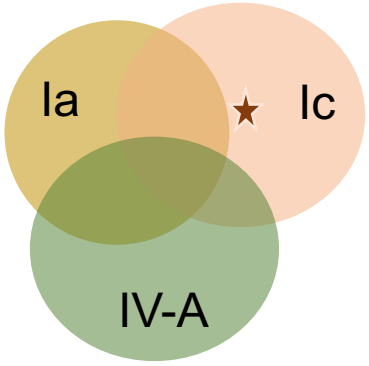
Stenosis at the orifice, but with only slightly dilated LHD

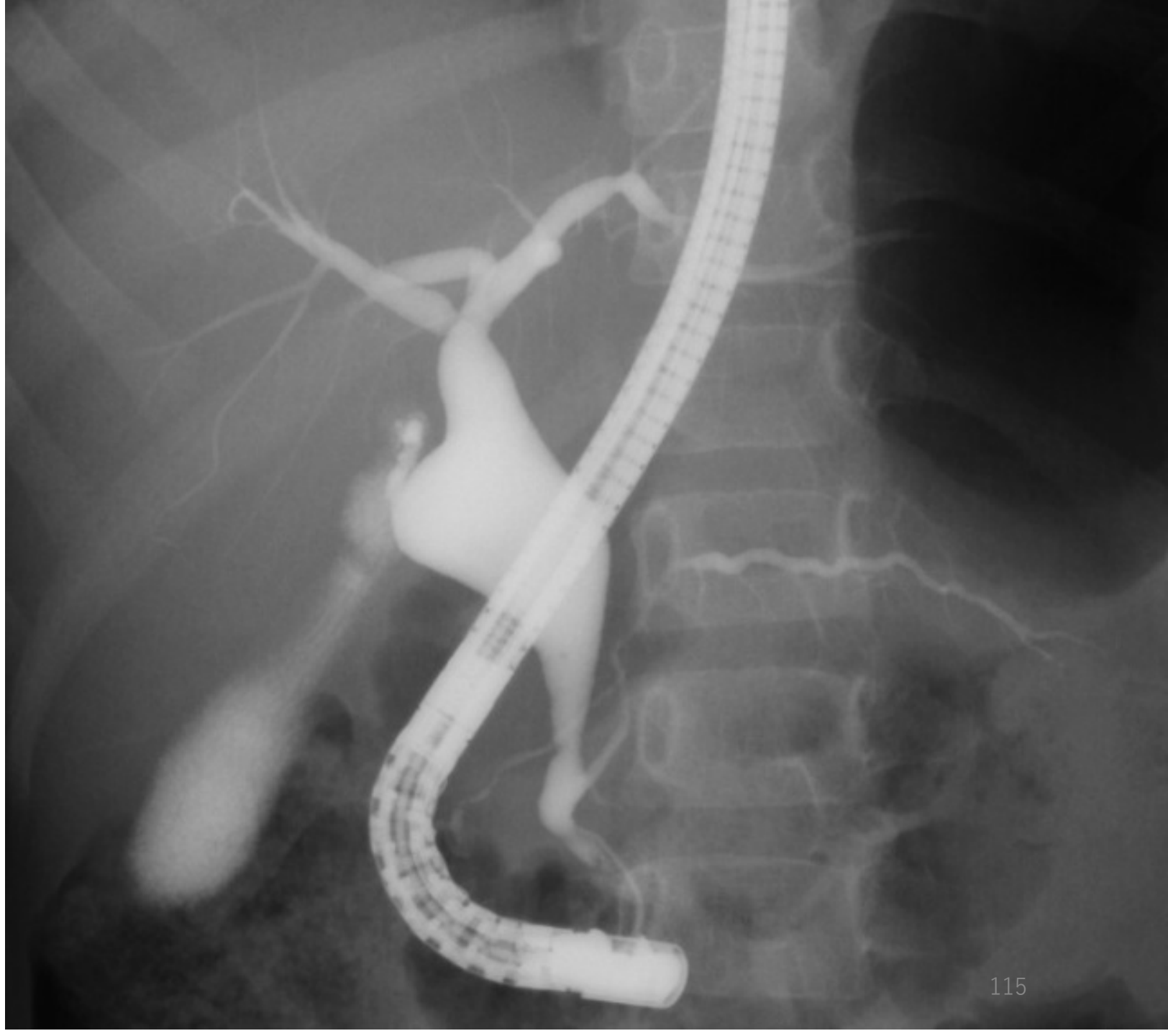


Case 34 Ia < Ic

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

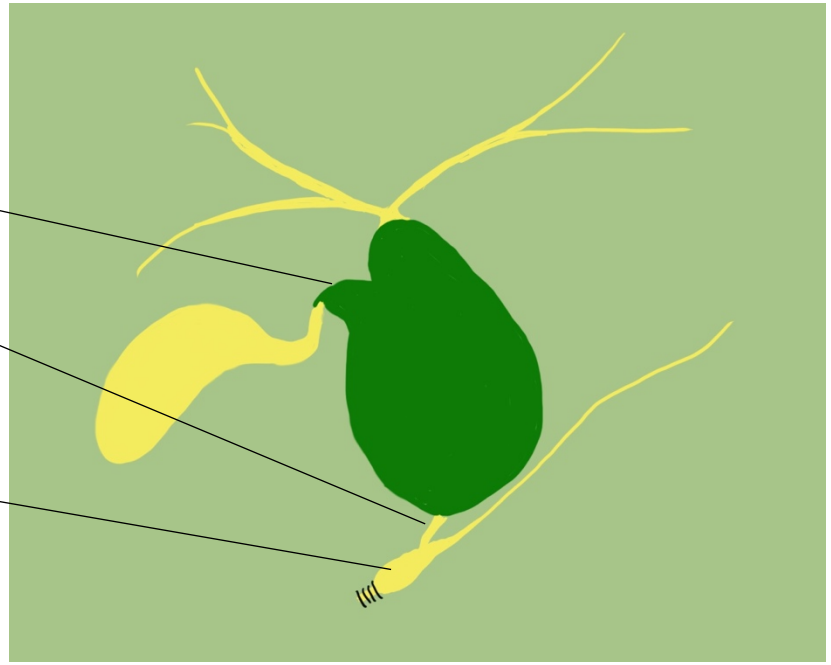
Imaging features of congenital biliary dilatation

Features that define CBD — Introduction section

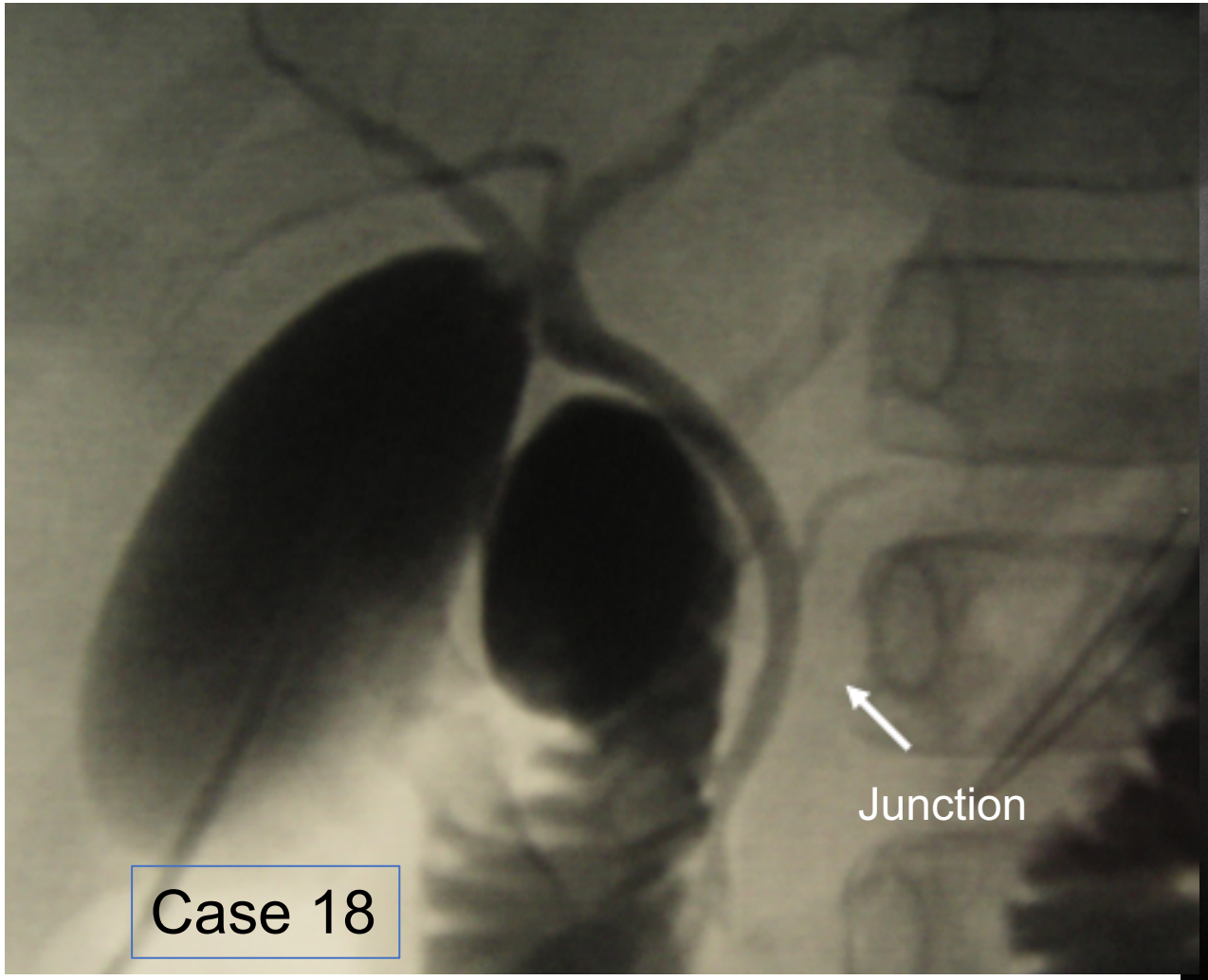
- Pancreaticobiliary maljunction
- Localized BD dilatation involving the common bile duct

Features (but not in all cases) — Basic section

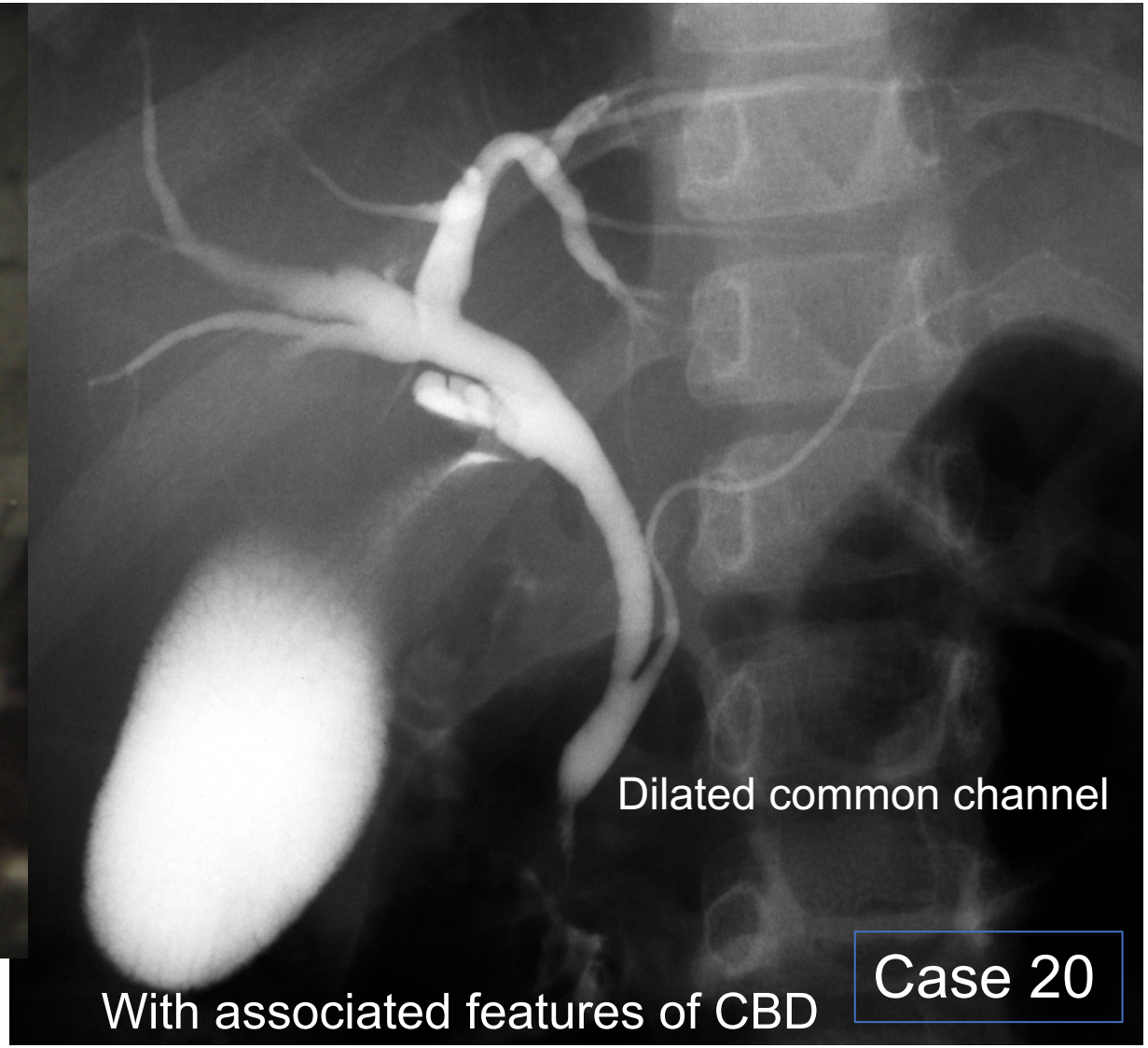
1. Dilatation at the base of the cystic duct
2. Narrow segment below the dilated BD
3. Dilated common channel



PBM with a normal BD diameter in pediatric cases

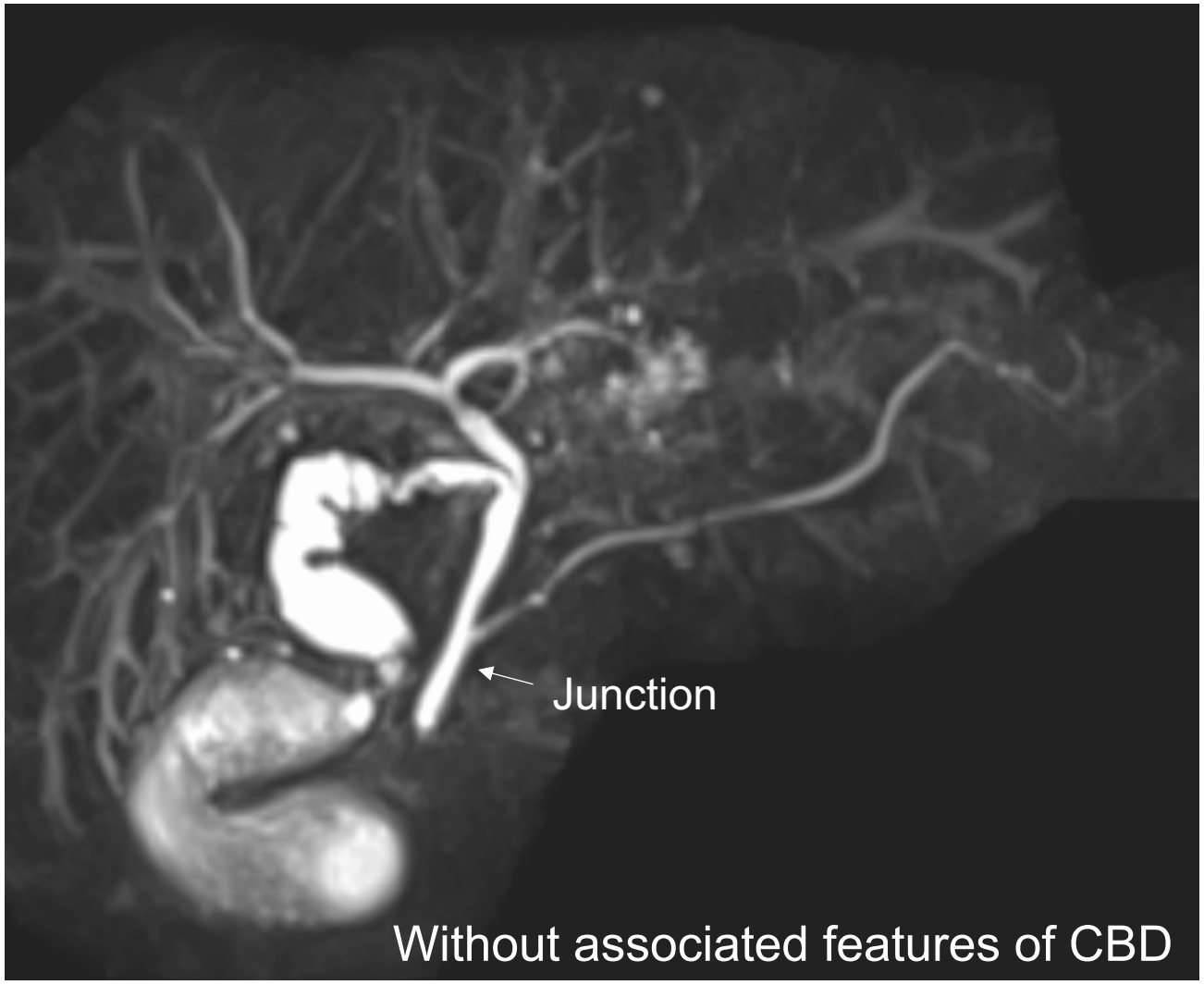


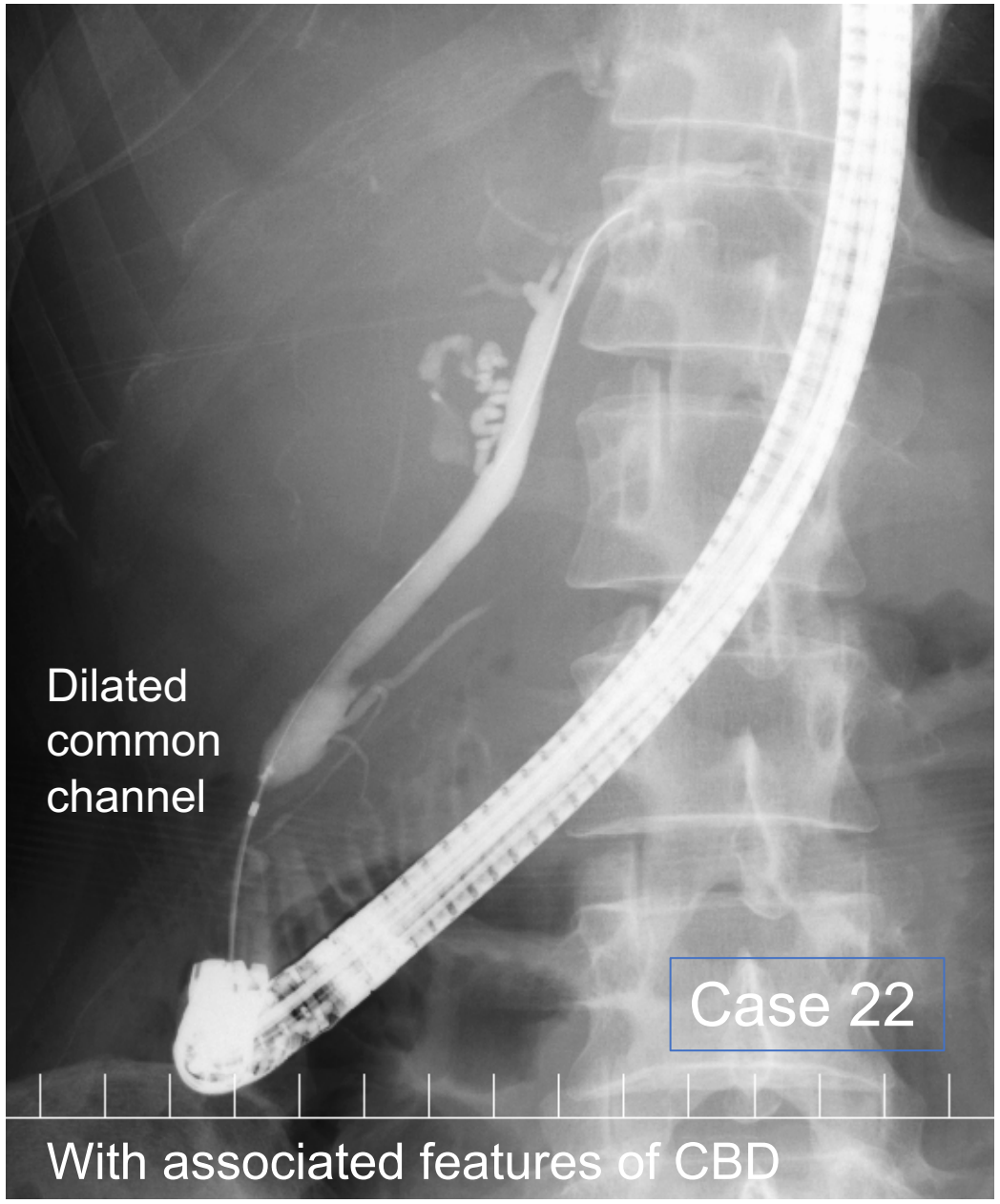
Without associated features of CBD



With associated features of CBD

PBM with normal BD diameter in adult cases





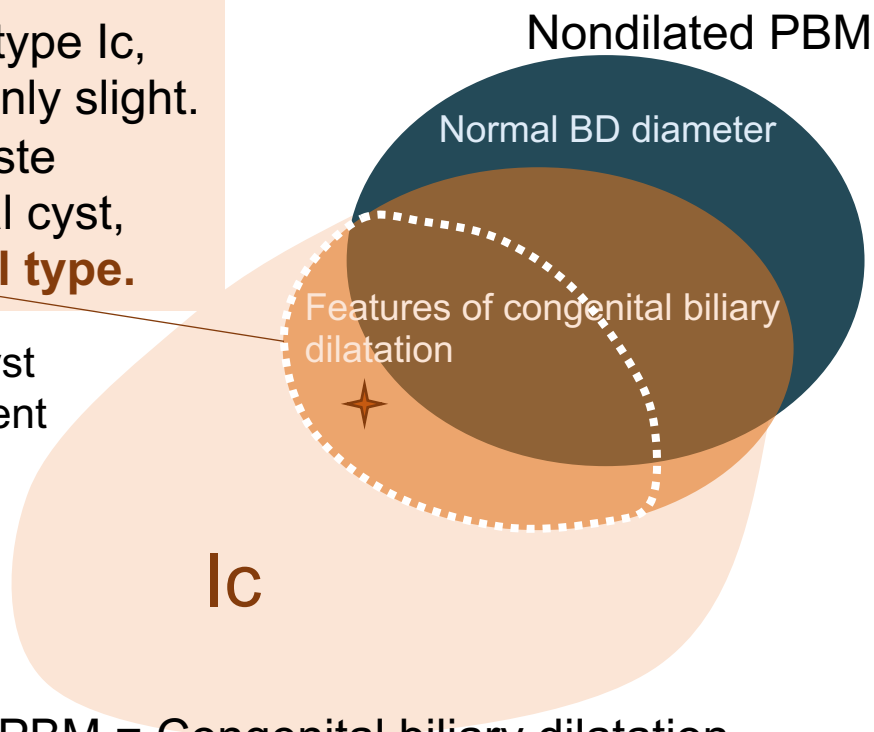
Case 35 Forme fruste (Ic) Adult case

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

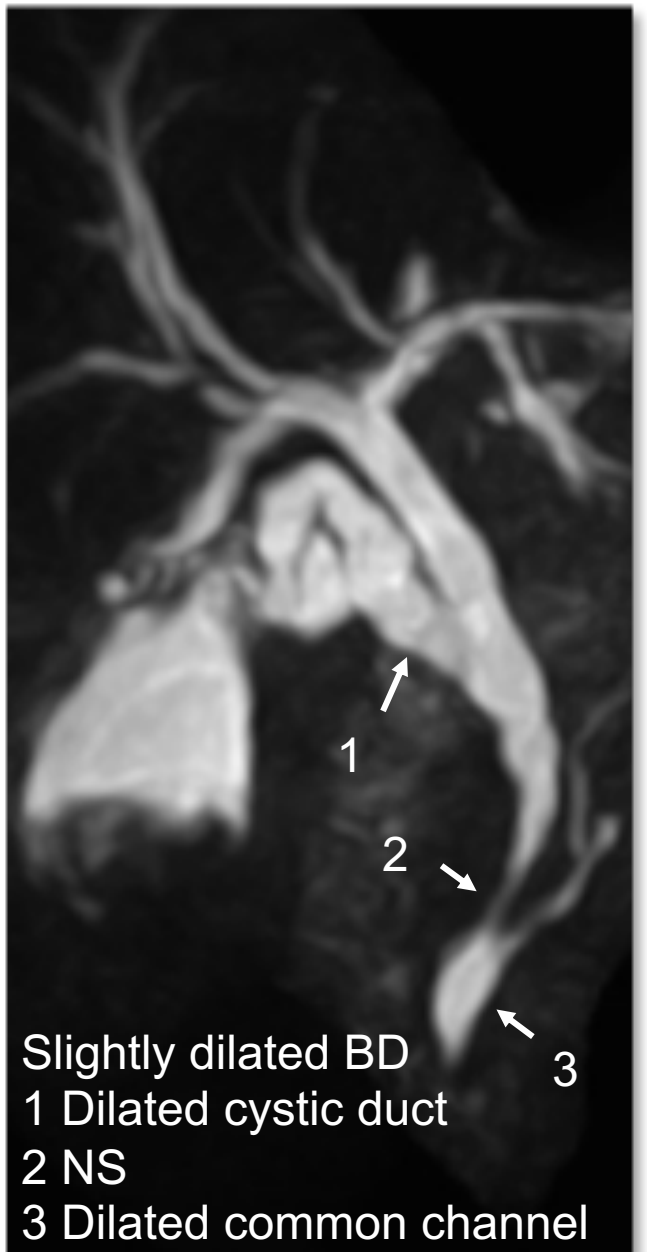
Adenosquamous carcinoma in the GB.

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

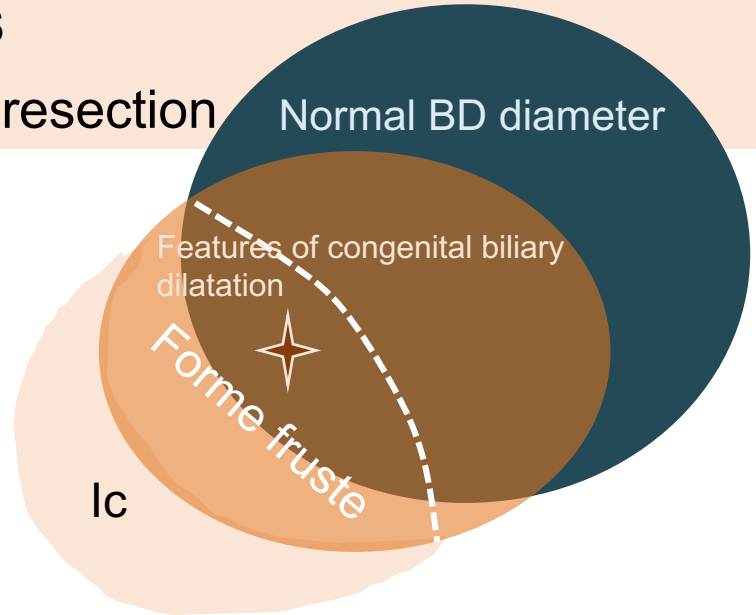
Case 36 Pediatric case Normal BD diameter

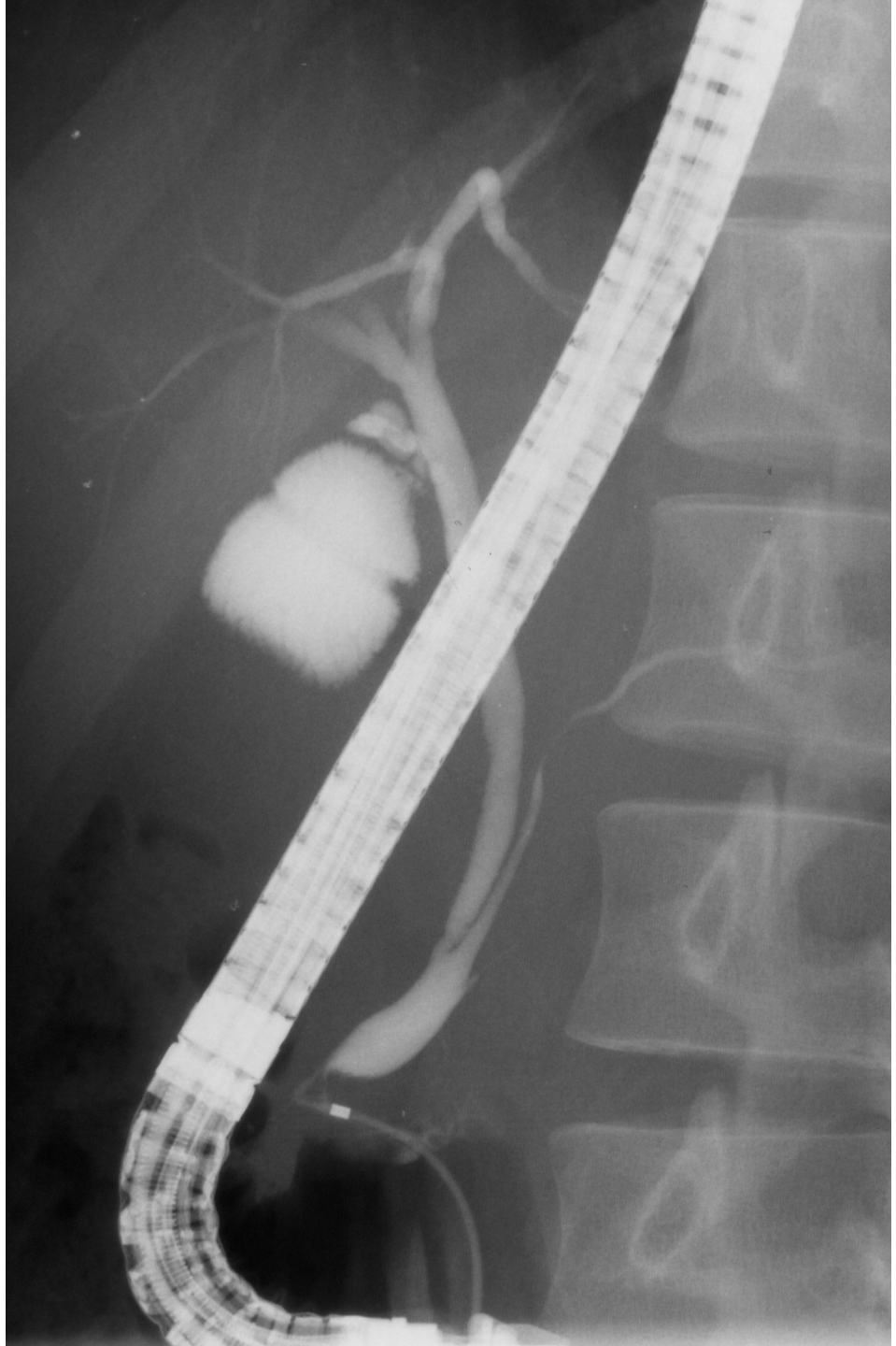
Nondilated PBM < Forme fruste

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

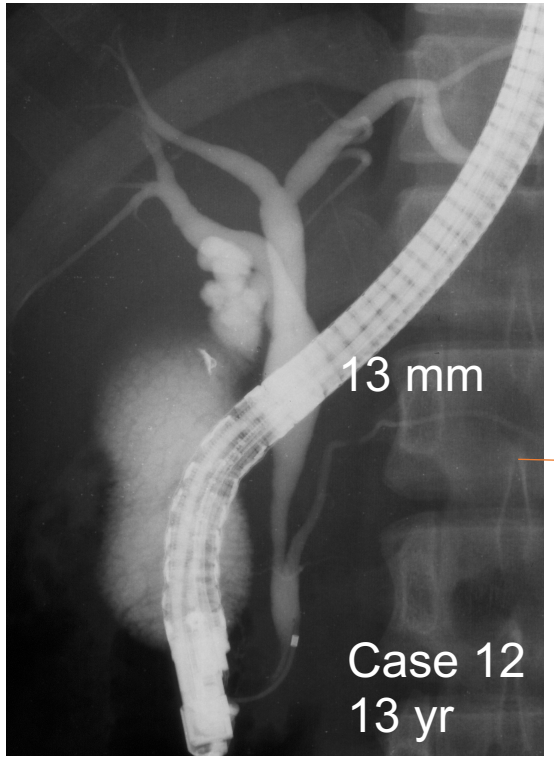
- Common bile duct diameter
  ERCP 6 mm (Ultrasound upper limit: 4.9 mm)
- **Features of congenital biliary dilatation**
  - ✓ **Dilated common channel**
  - ✓ **NS**

Bile duct resection





Around forme fruste



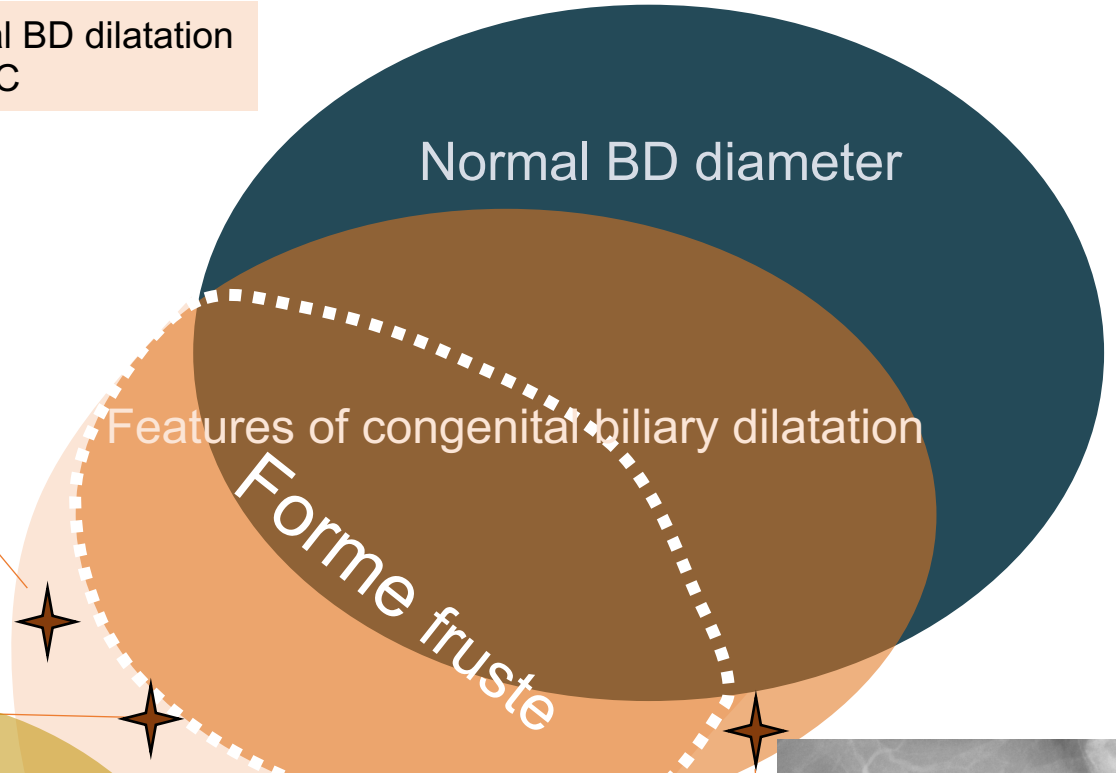
Fusiform BD dilatation
Dilated common channel (CC)
NS



Cylindrical BD dilatation
Dilated CC



Fusiform BD dilatation
Dilated CC



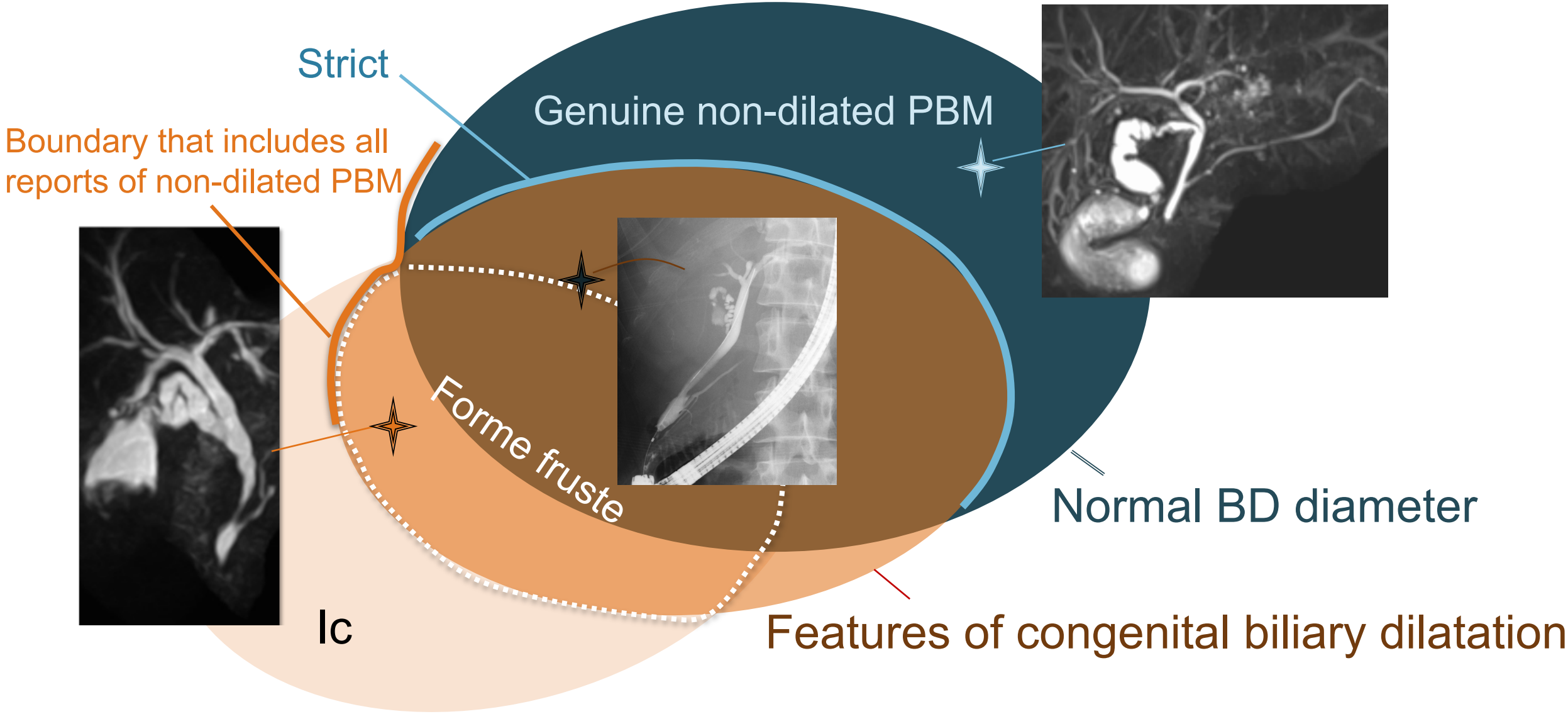
No bile stasis
except in the GB



Only slight biliary dilatation as a
feature of congenital biliary dilatation

Definition of non-dilated PBM

Where to draw the boundary on this spectrum?

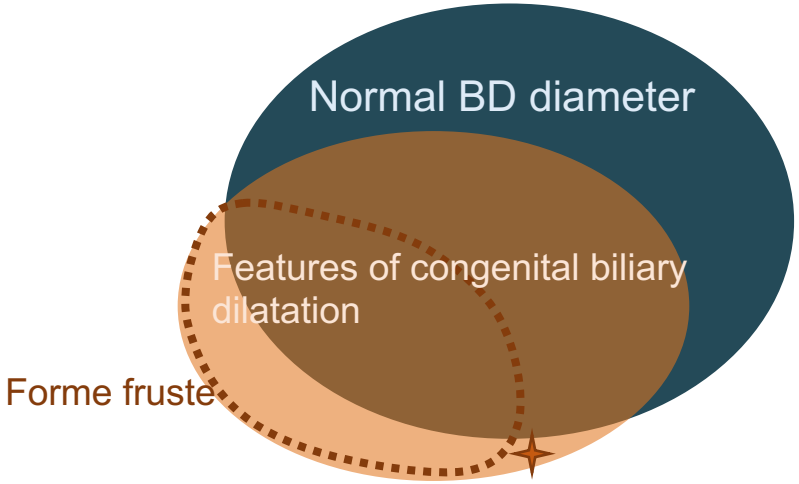


Case 37

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

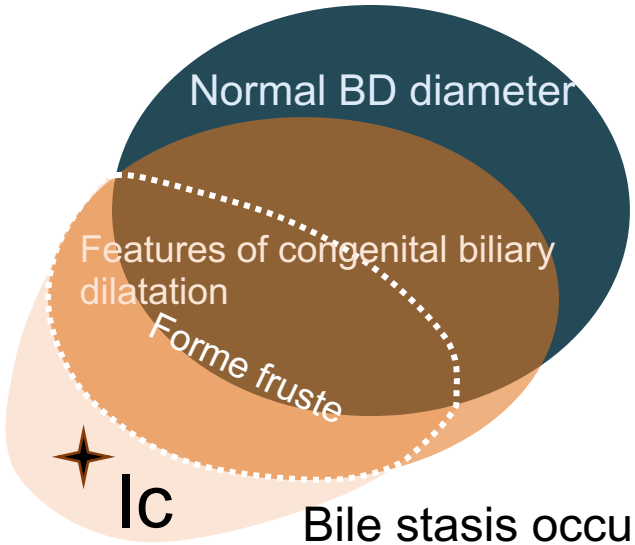


From *Basic Section*

Case 14

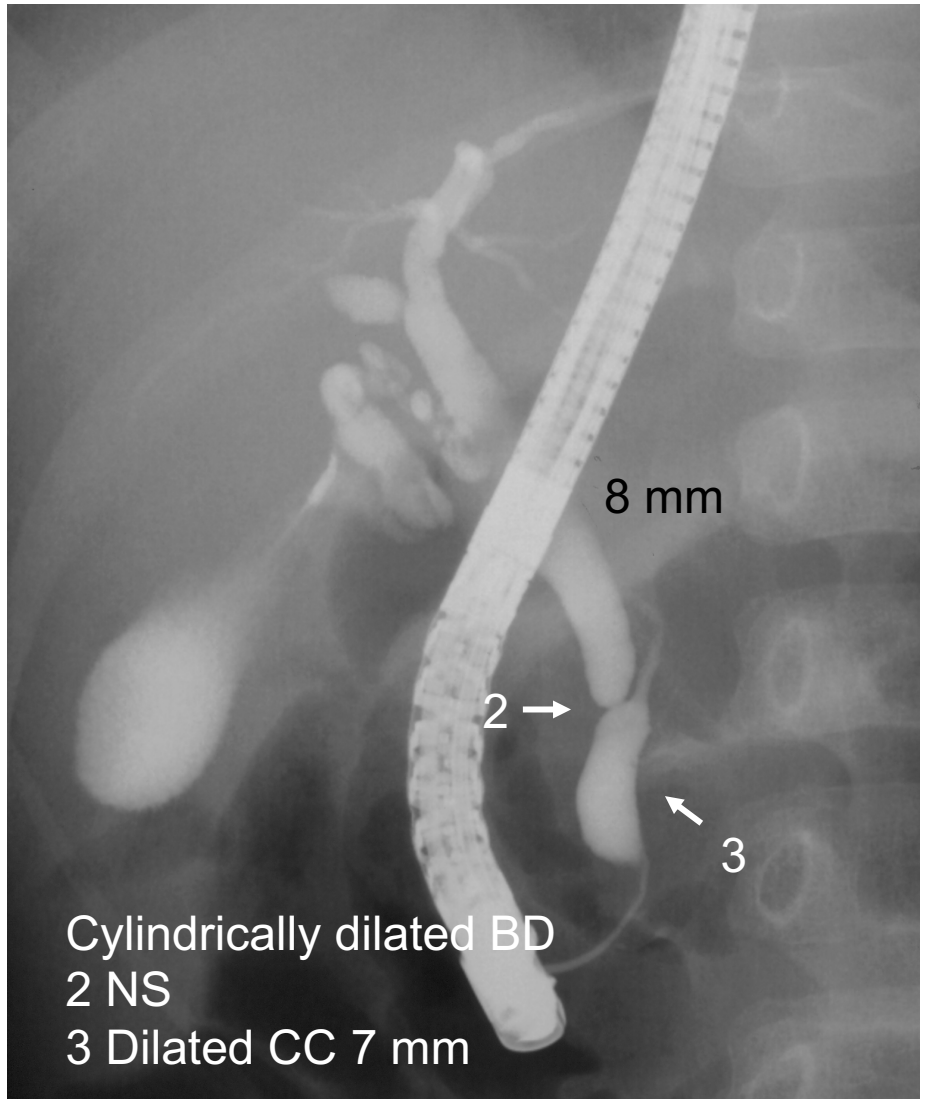
Type Ic: cylindrical dilatation Pediatric case

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



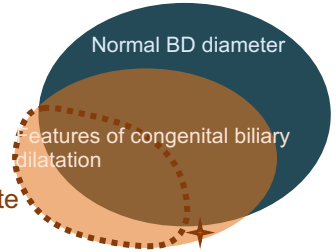
2 years old (US upper limit: 3.3 mm)

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.



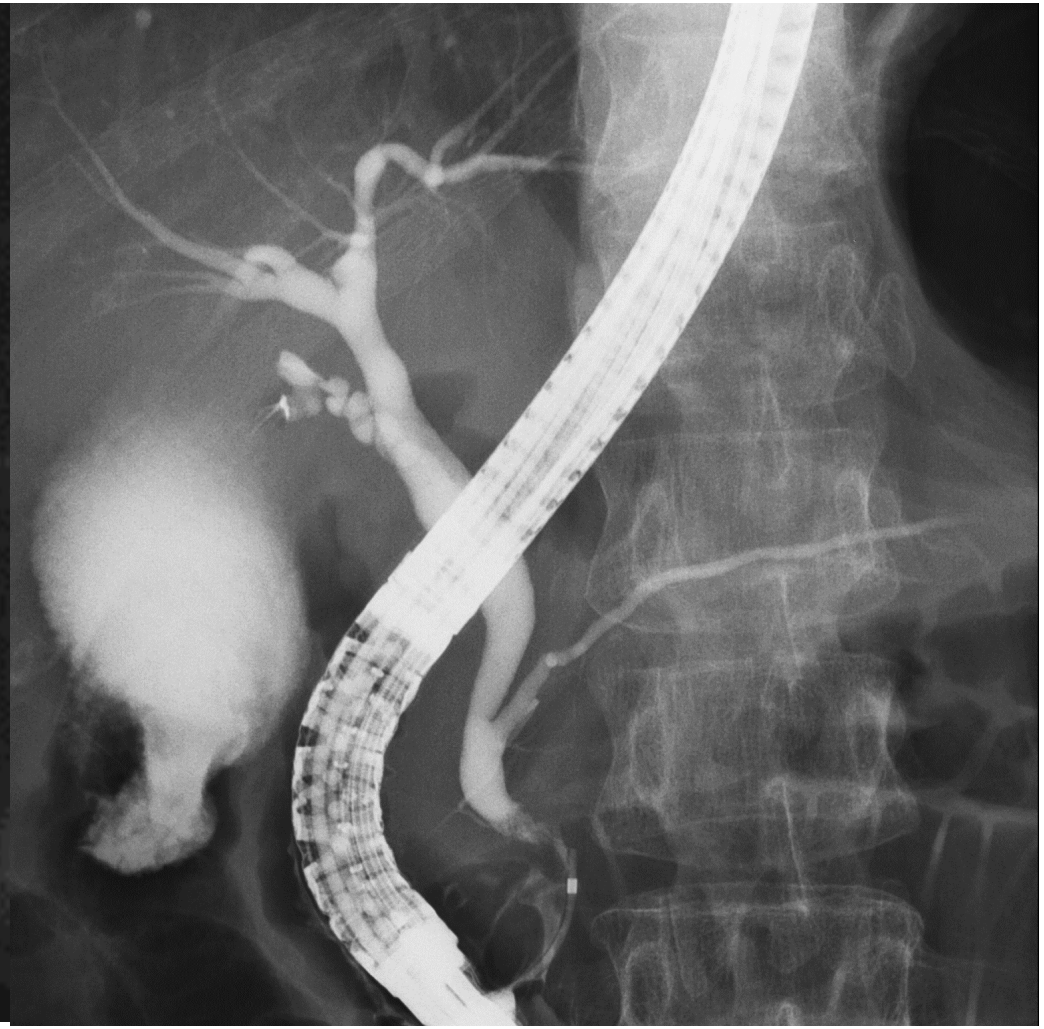
A female in her 50s.
US during medical checkup detected GB wall thickening.

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

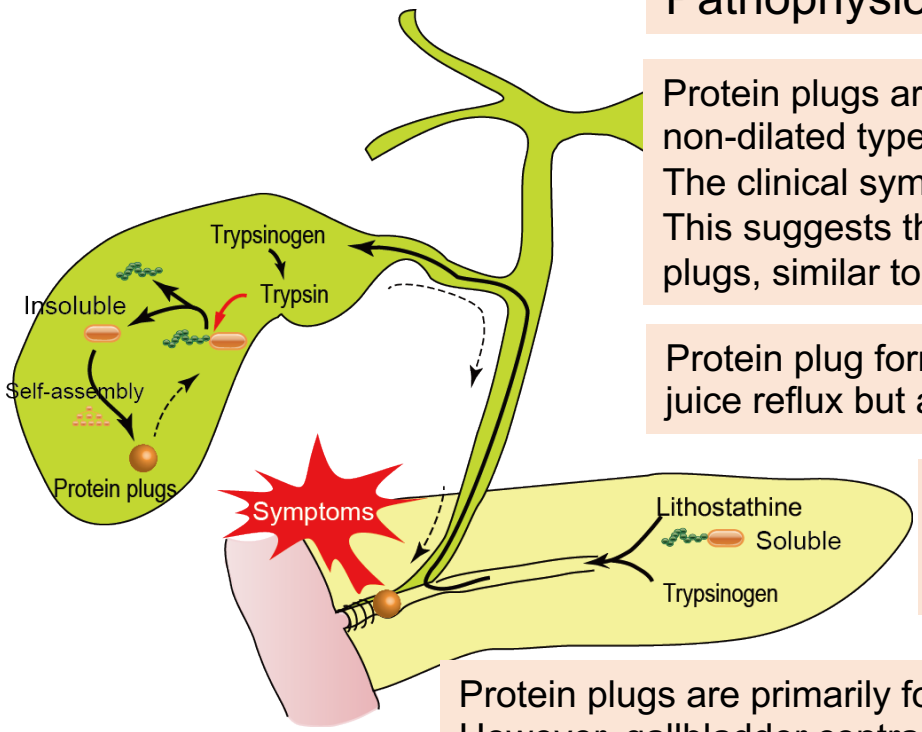
First version, September 2026

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

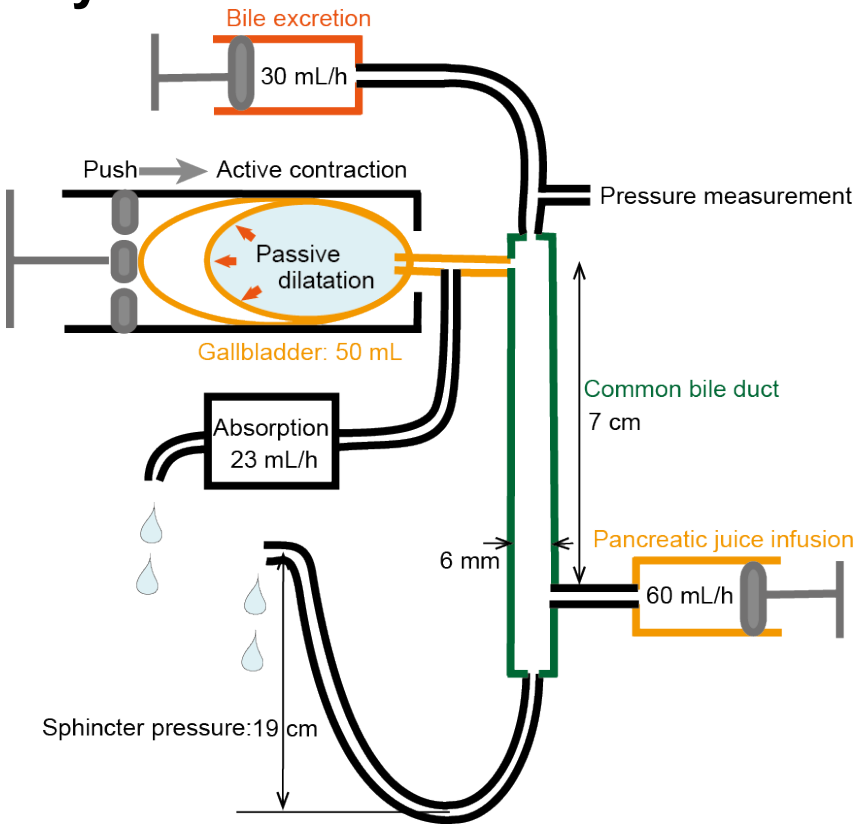
Protein plugs are primarily formed within the gallbladder. However, gallbladder contraction acts to inhibit this formation, which explains the lower overall incidence.



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

Cholecystectomy eliminates pancreatic juice reflux in the non-dilated type

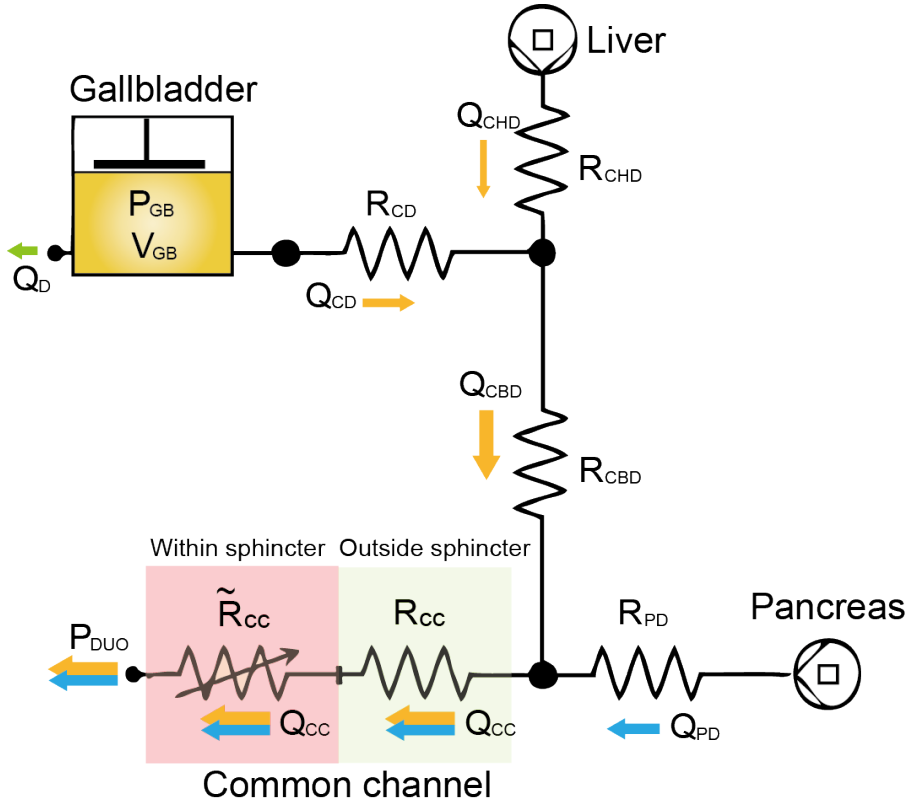
Fluid dynamics model of non-dilated PBM



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: $\tilde{R}_{CC}Q_{PD} > P_{GB} - P_{DUO}$

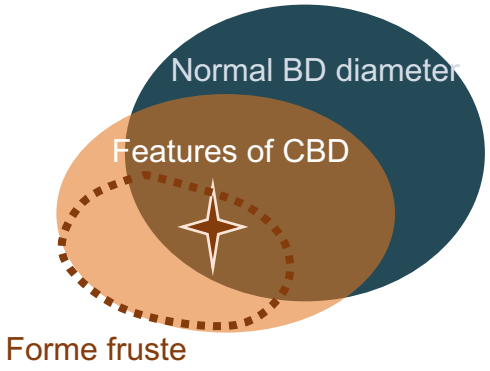
Tajikawa T, et al. *J Biorheol* 2023;37(2):44–55
Biomed J Sci & Tech Res 2024;55:47447–56

Case 39

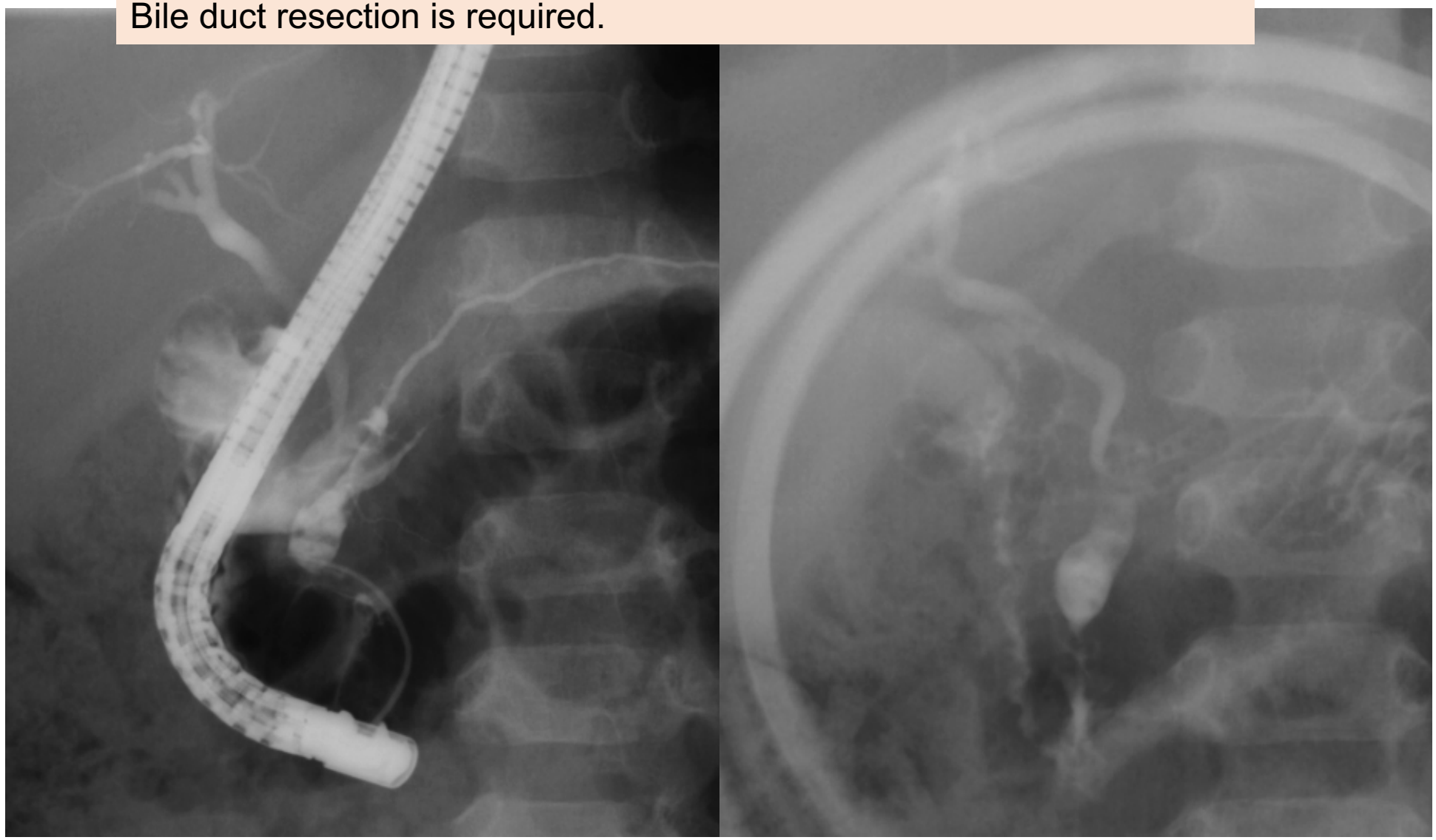
PBM with a normal BD diameter and prominent common channel dilatation, Forme fruste > Non-dilated

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.

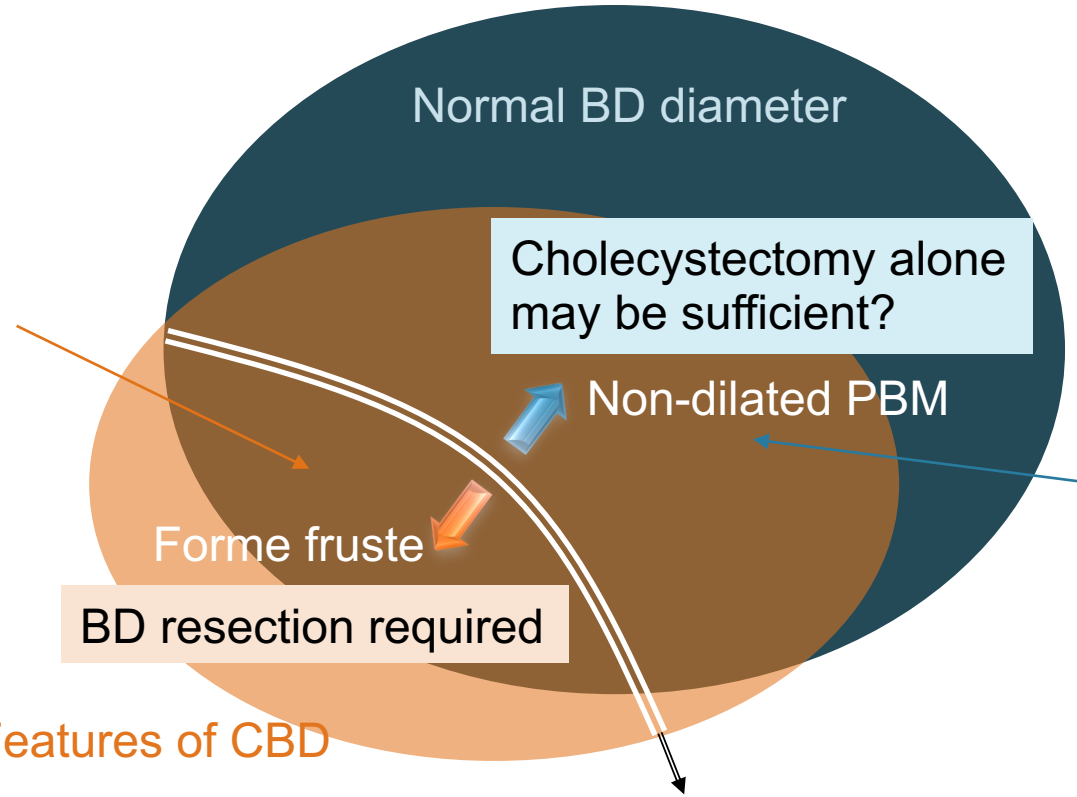


Therapeutic considerations in children
Varying features of common channel dilatation



Normal BD diameter but prominent common channel dilatation

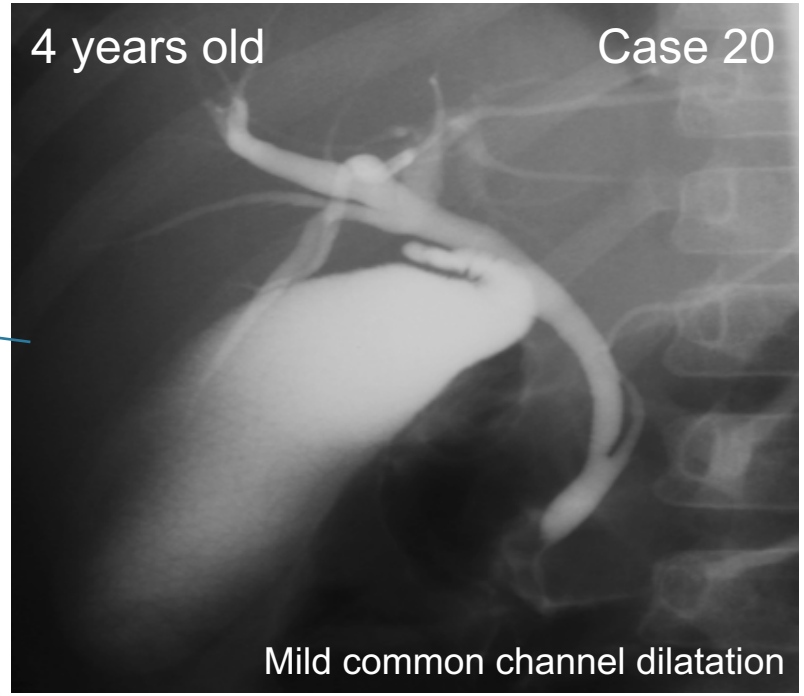
Bile stasis in both the GB and common channel



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.

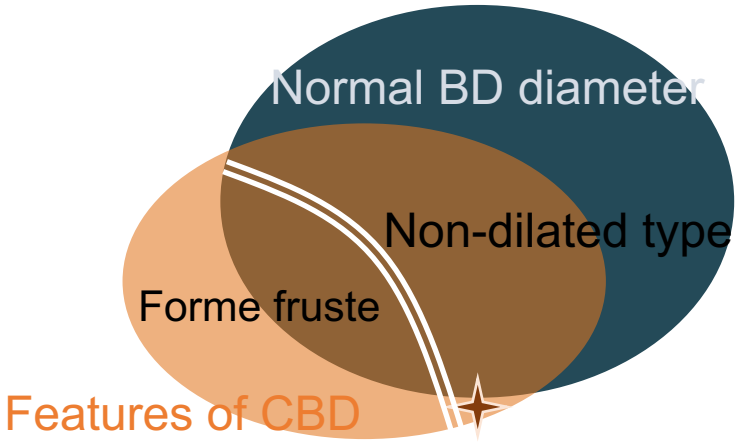
Case 40

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.

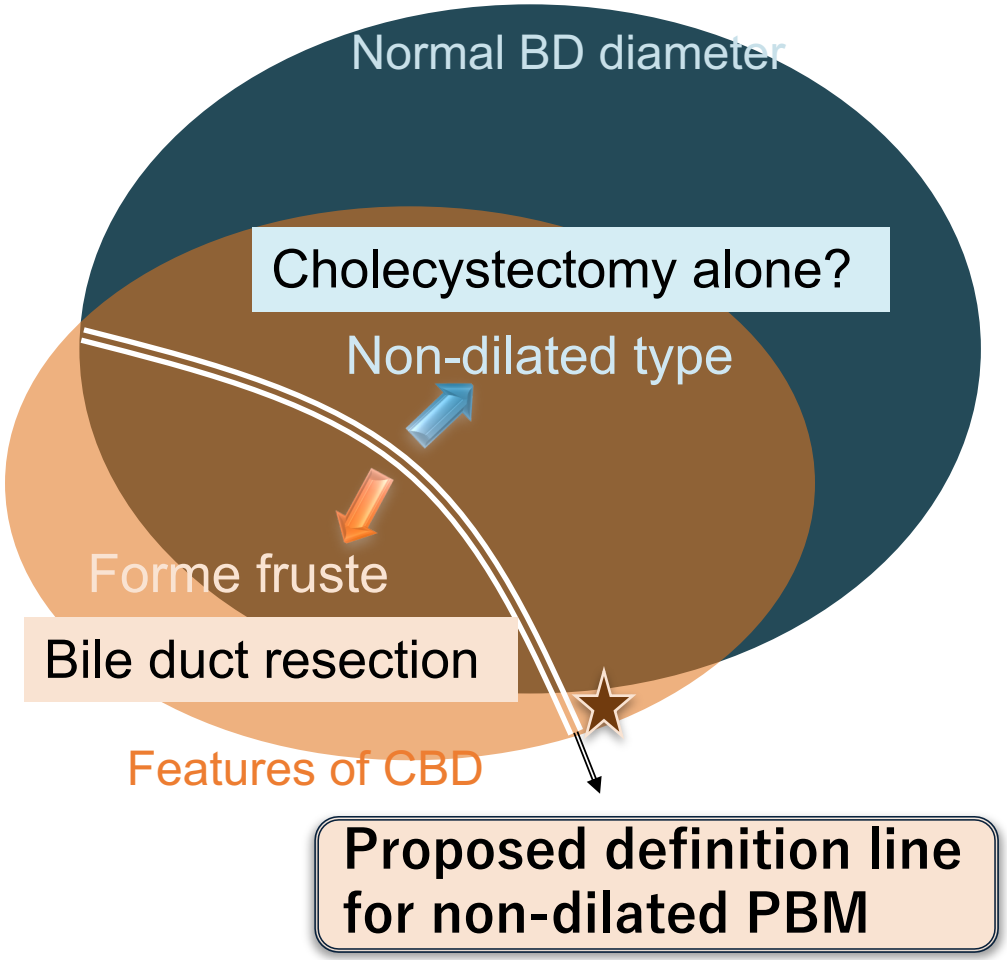






Strictly speaking, the BD is dilated,
but no bile stasis occurs except in the GB.

Slight BD dilatation and mild features of CBD



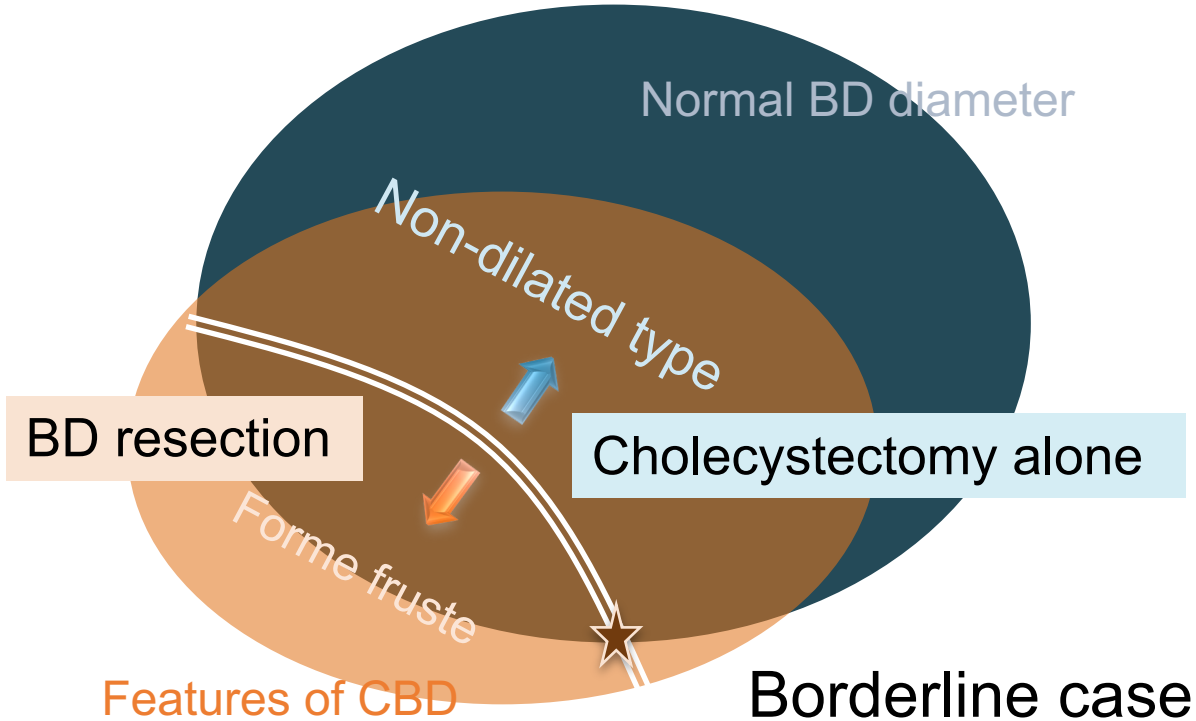




The definition and treatment may depend on the existence of bile stasis except in the GB.

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



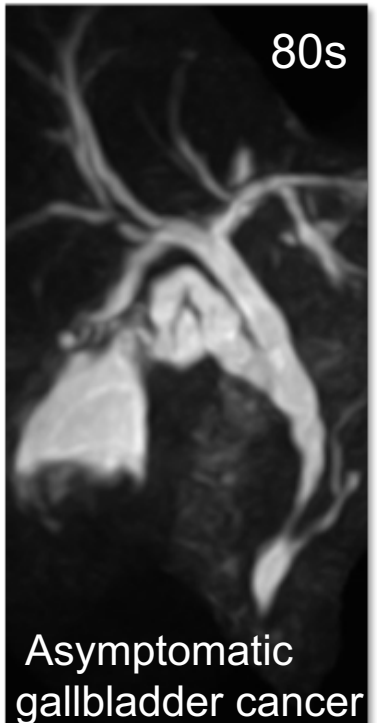
Advanced

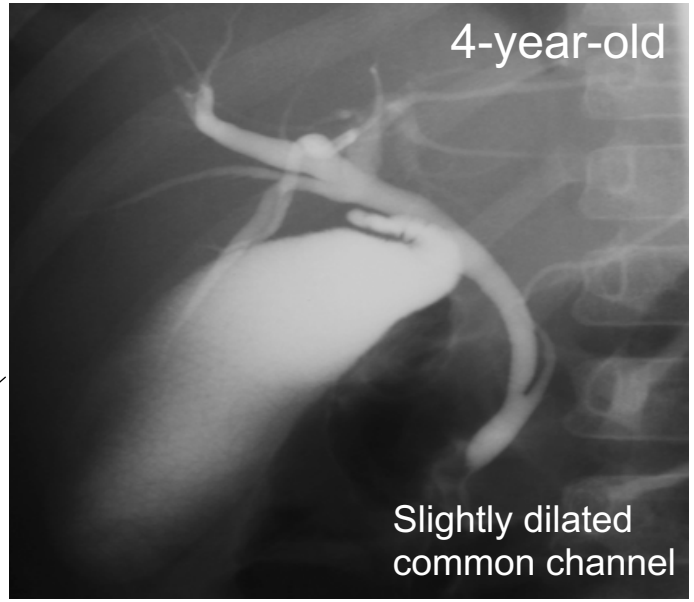
Non-dilated type of PBM

Pathophysiology and treatment in adults

First version, September 2026

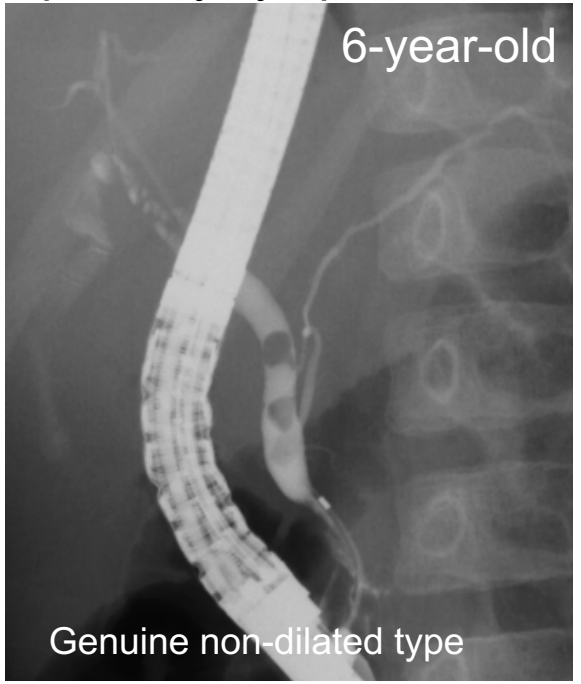
Non-dilated type
Why are so many cases asymptomatic?

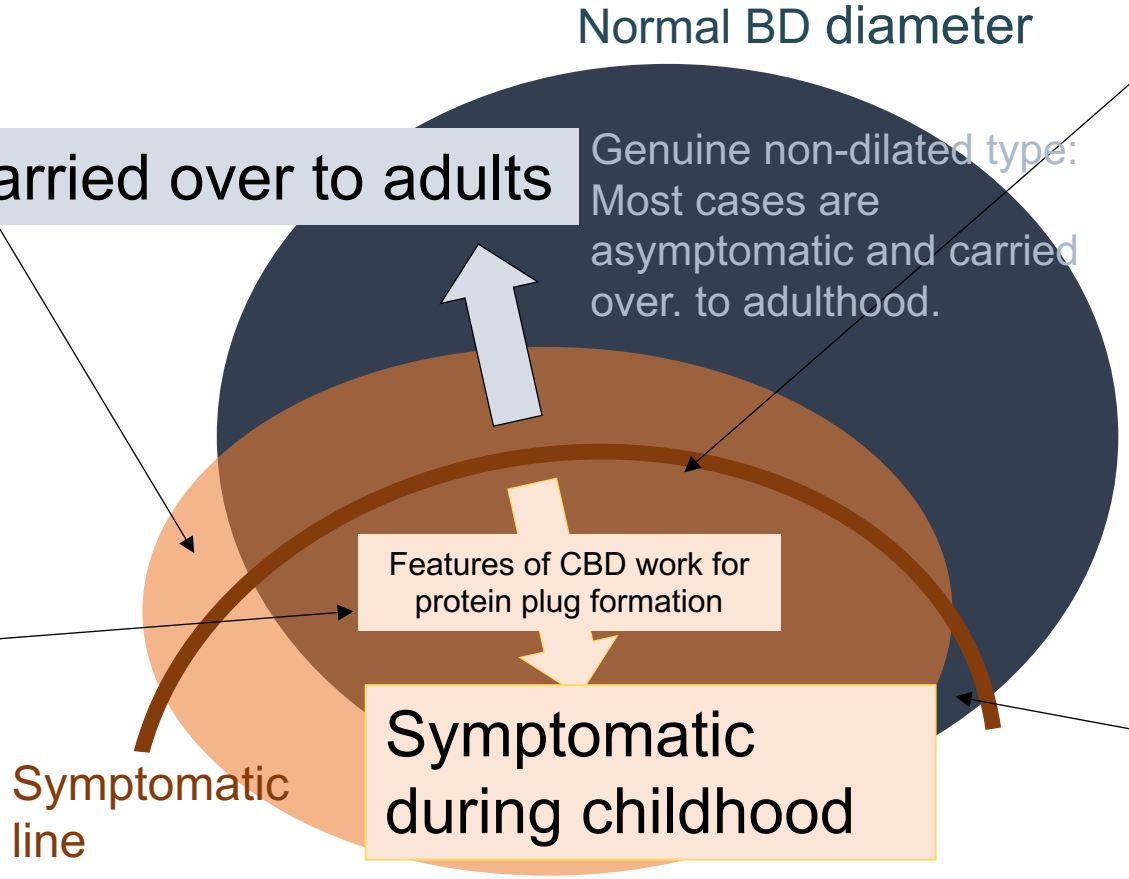




Exceptionally symptomatic cases



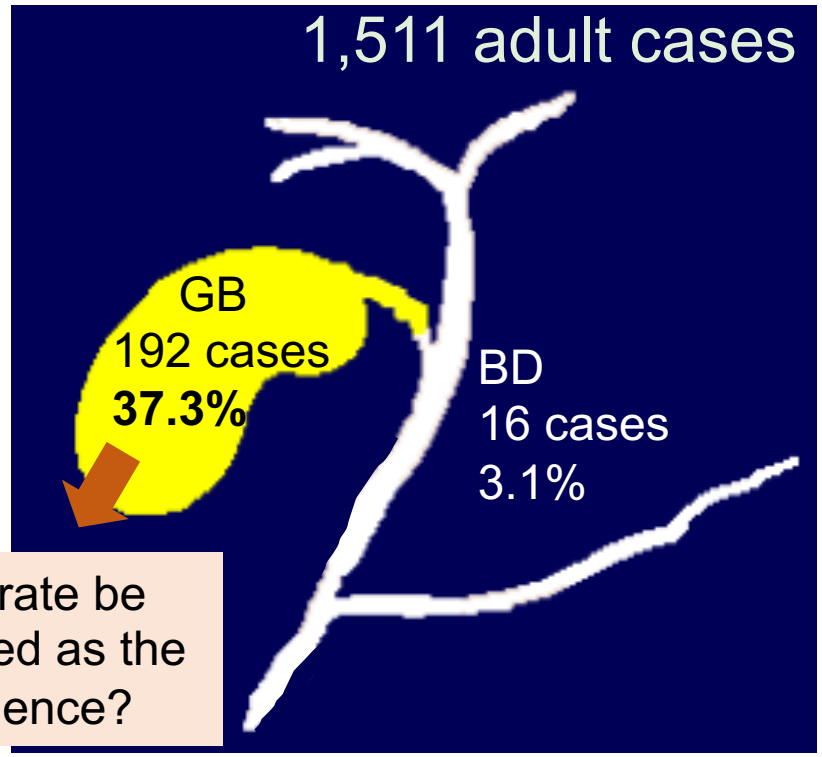




Is the incidence of gallbladder cancer in non-dilated type really high?

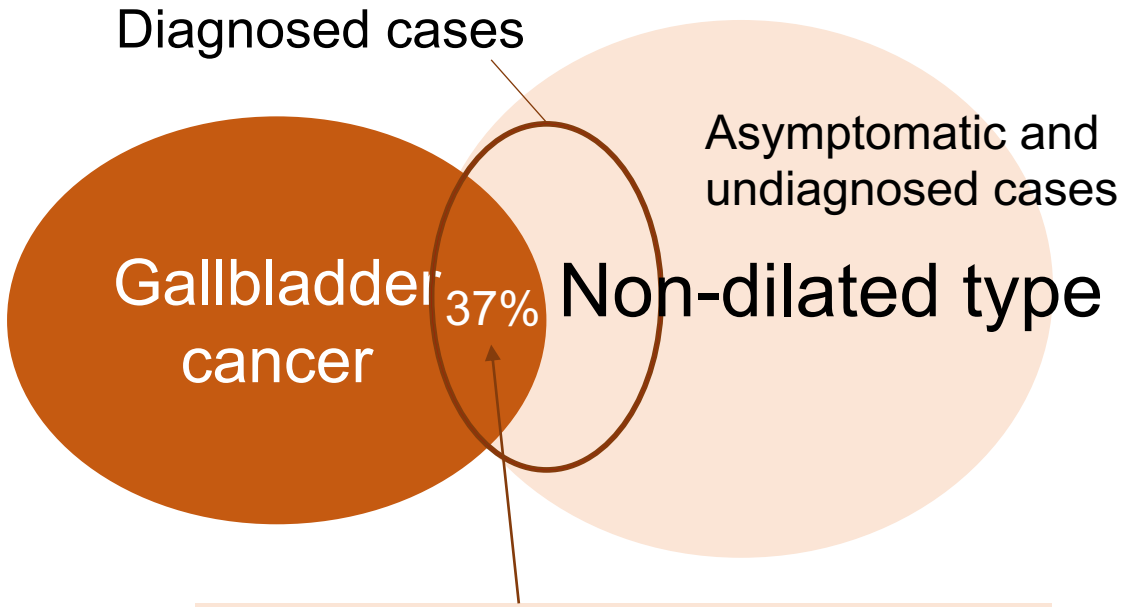
From *Introduction Section*

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.

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

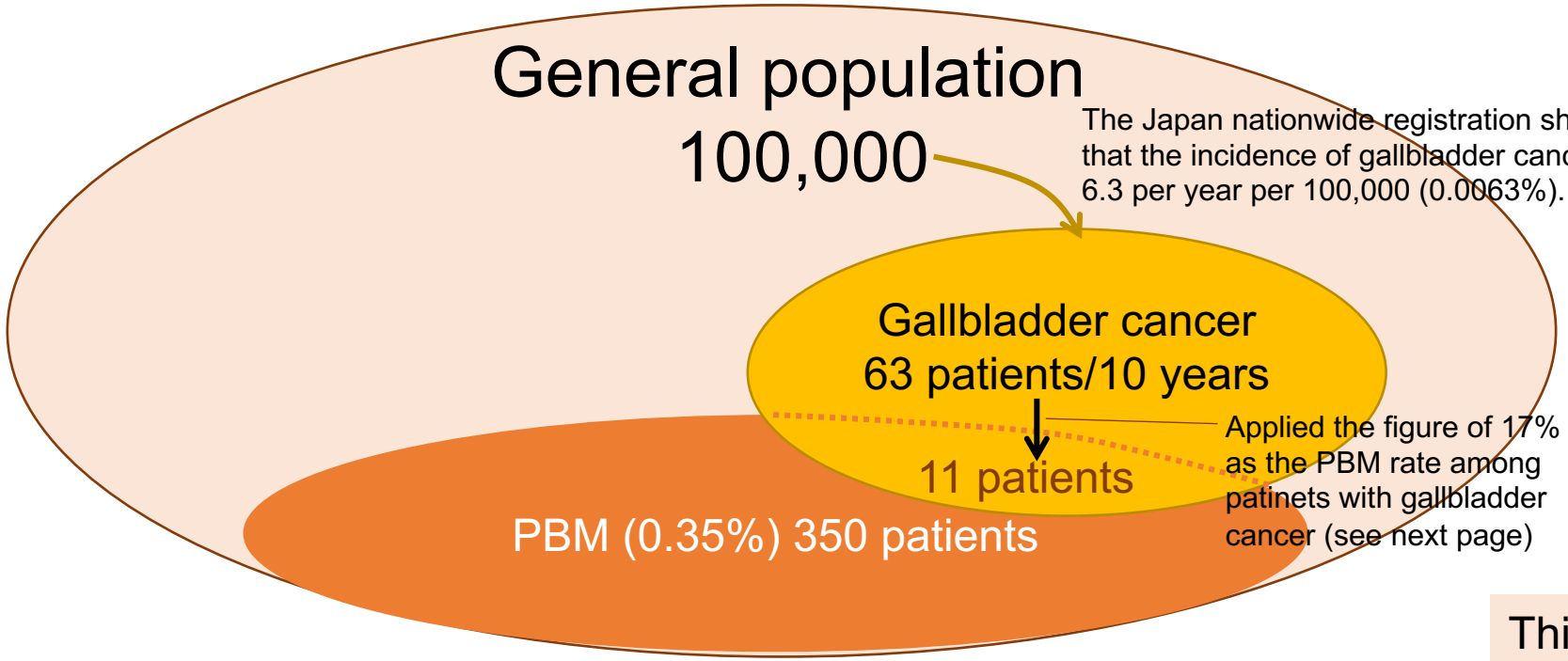
Yamao K, et al.
Prospective study of the detection of anomalous connections of pancreatobiliary ducts during routine medical examinations.
Hepatogastroenterology 1996; 43: 1238–1245.

Estimation of the incidence of gallbladder cancer

This is just a rough estimate.



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



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.

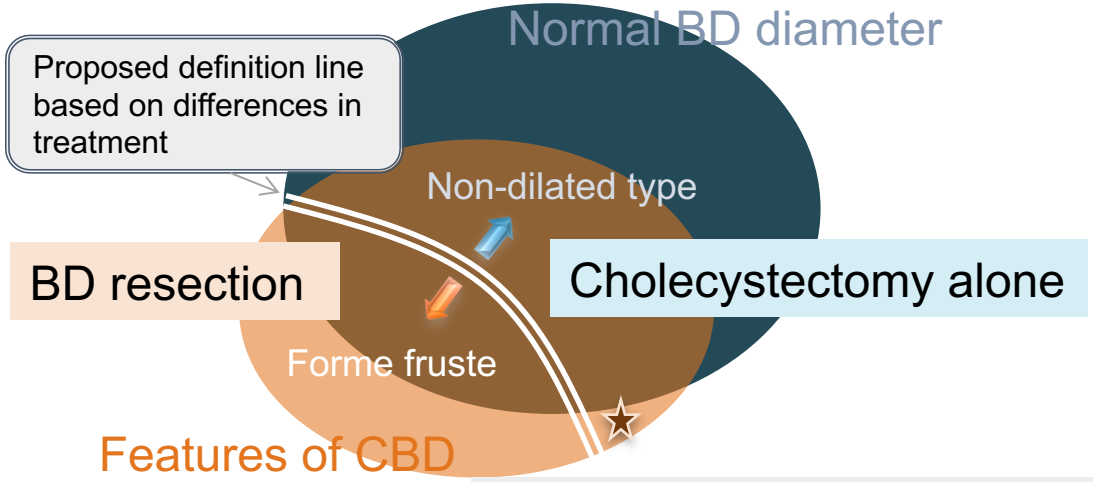
~ 2%

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

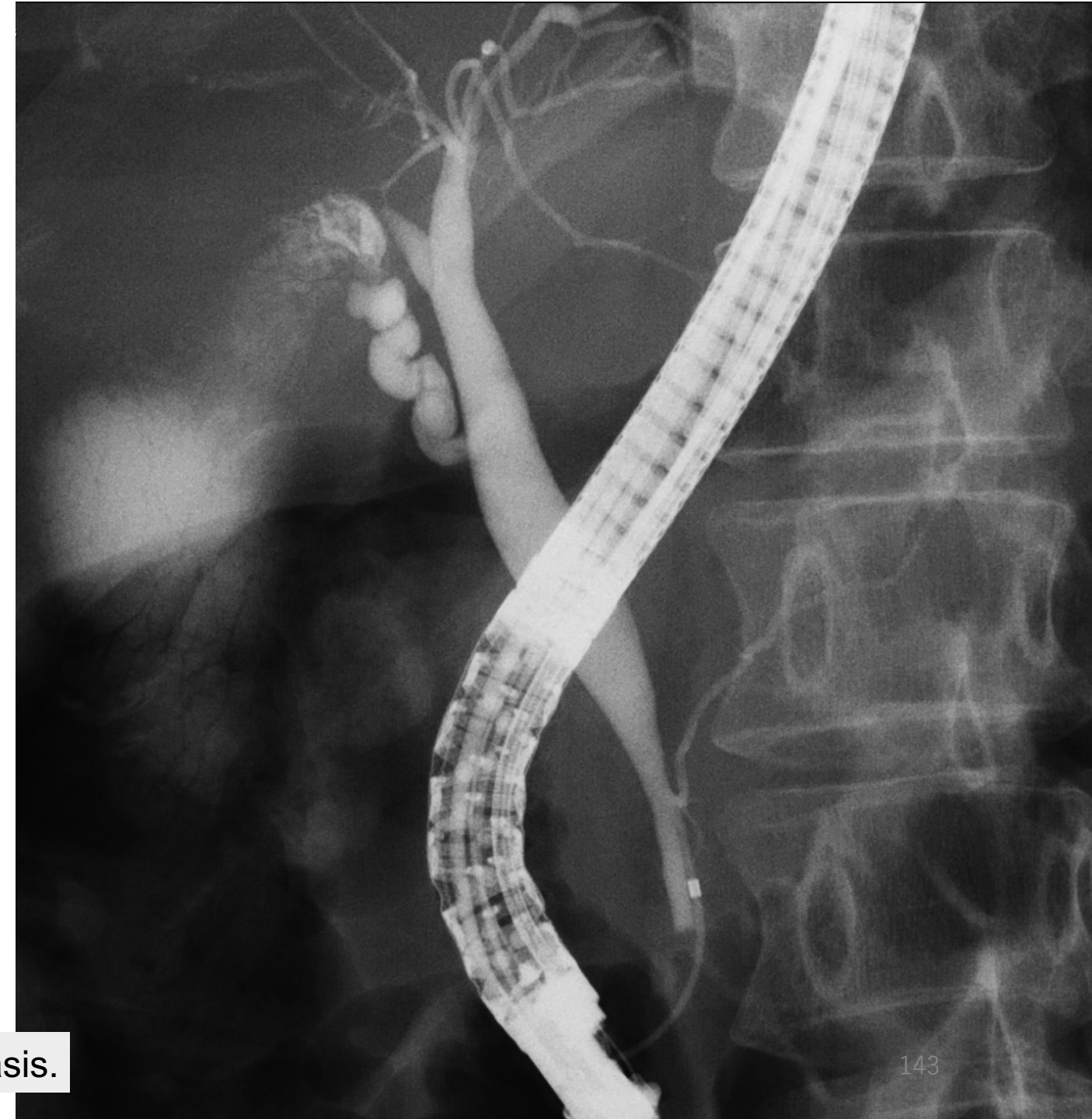
Case 42 Non-dilated type > Forme fruste (Ic)

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.



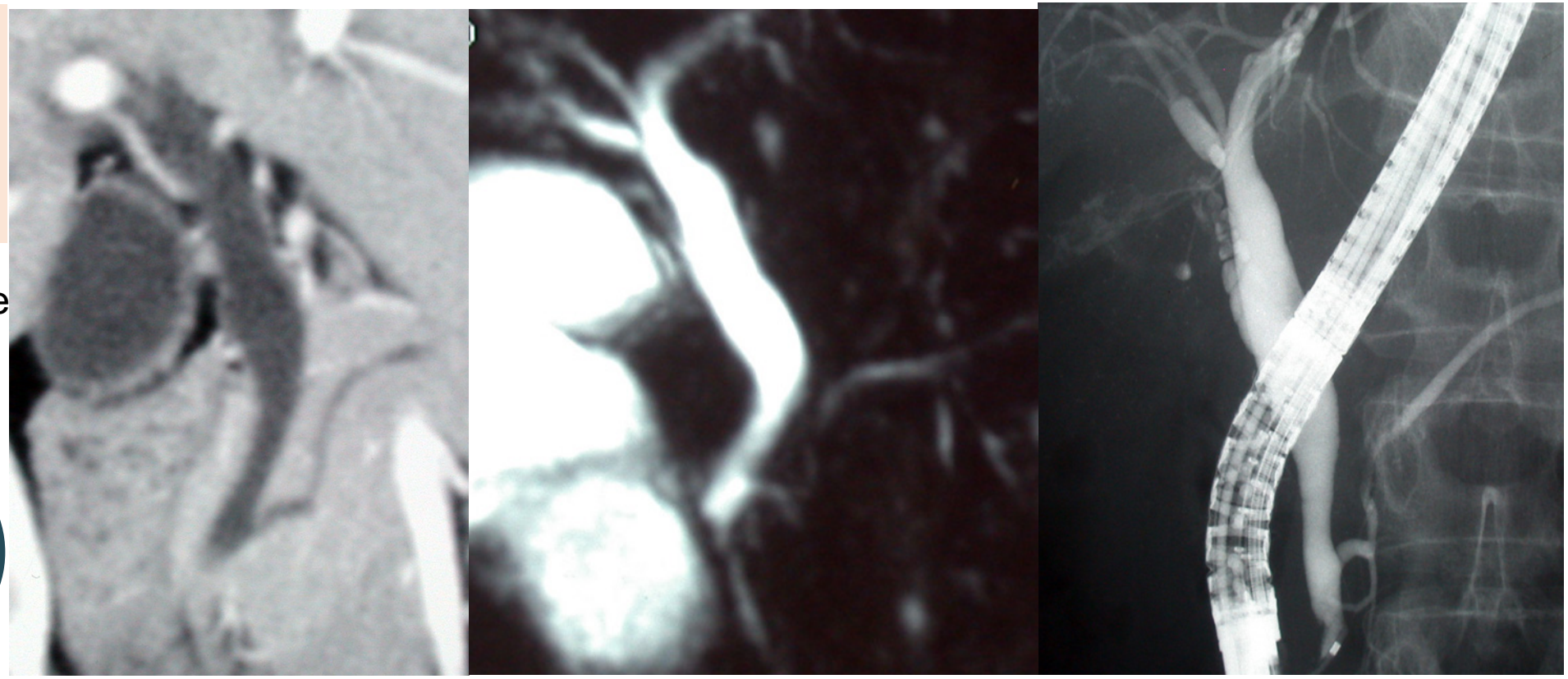
Carcinogenesis needs bile stasis.



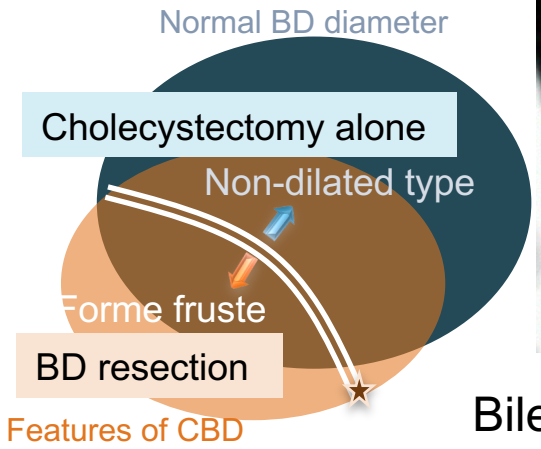
Case 43

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.

Advanced

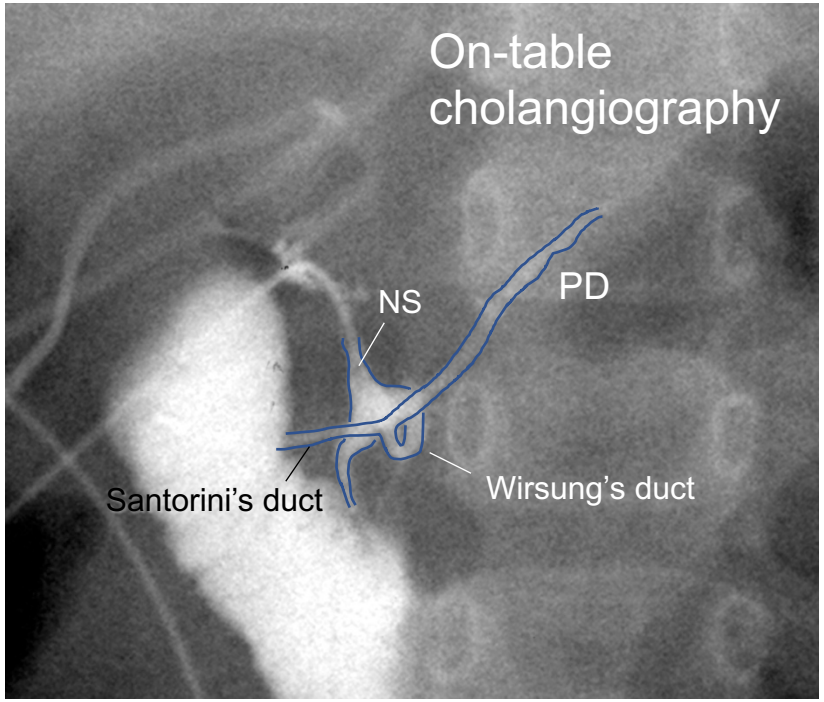
PBM seemingly complicated and rare type of PBM

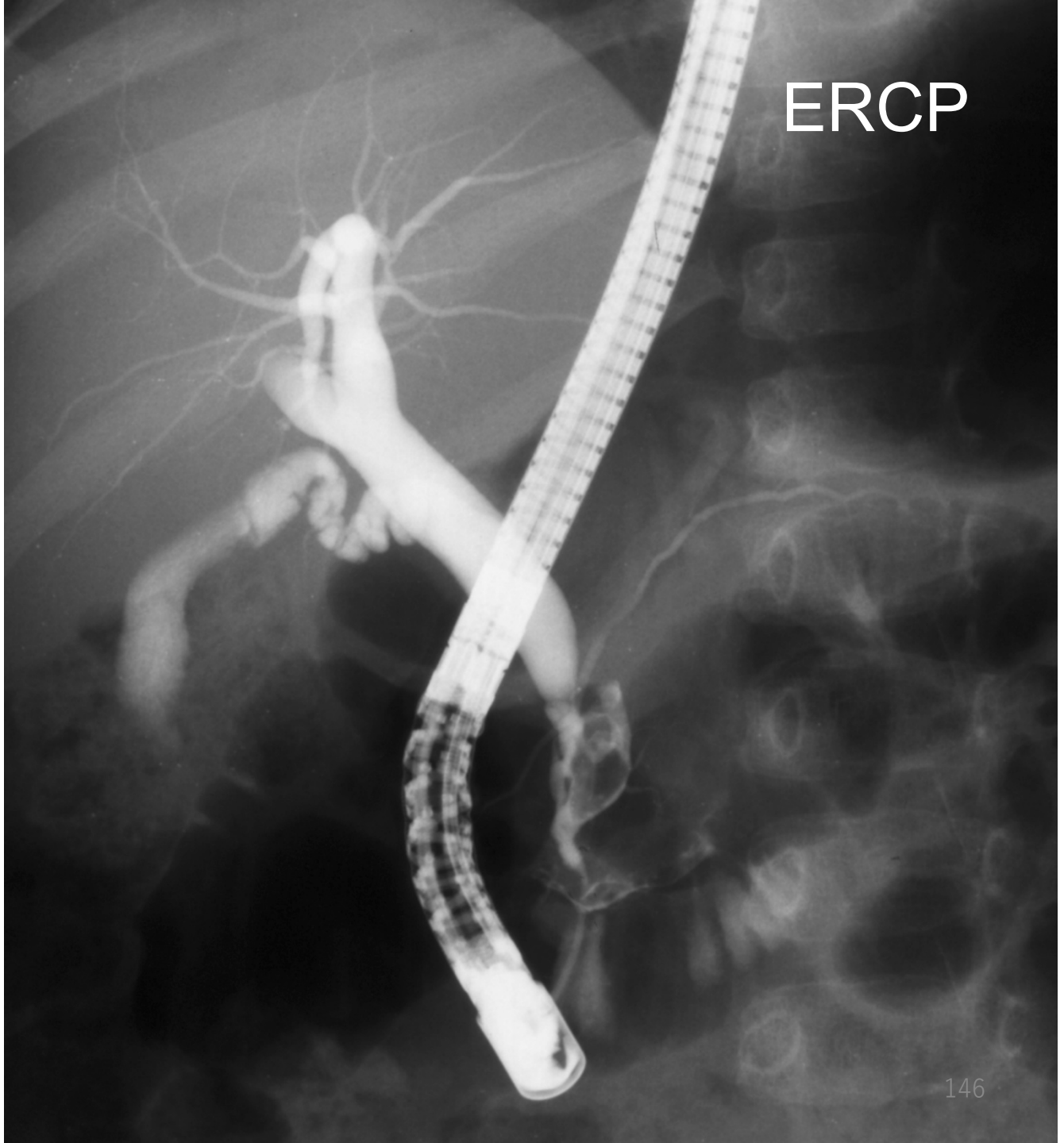
First version, September 2024

Case 44 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 its dilatation, complicating the appearance of PBM.

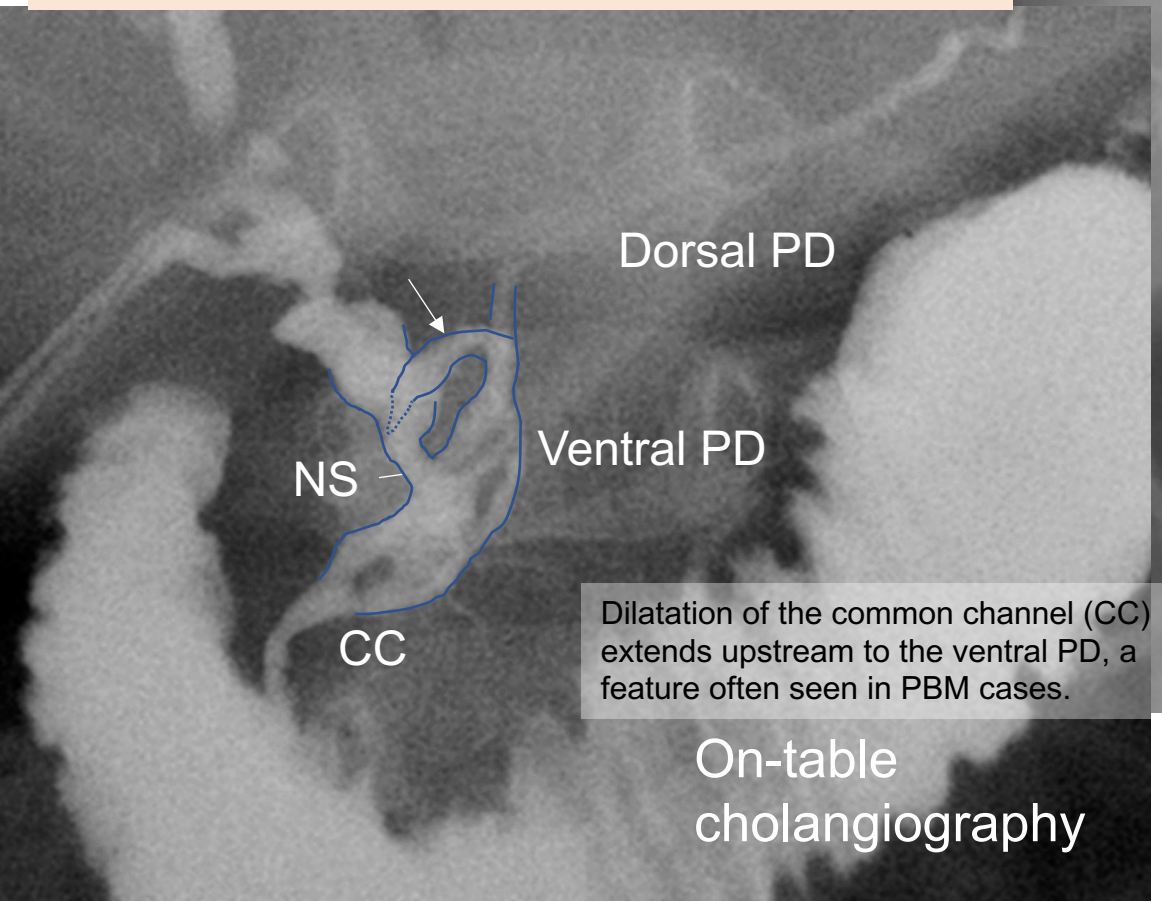
She underwent BD resection after her symptoms subsided.
On-table cholangiography showed the disappearance of the protein plugs, which disclosed the structure of PBM.
The dilatation of the common channel had completely disappeared.





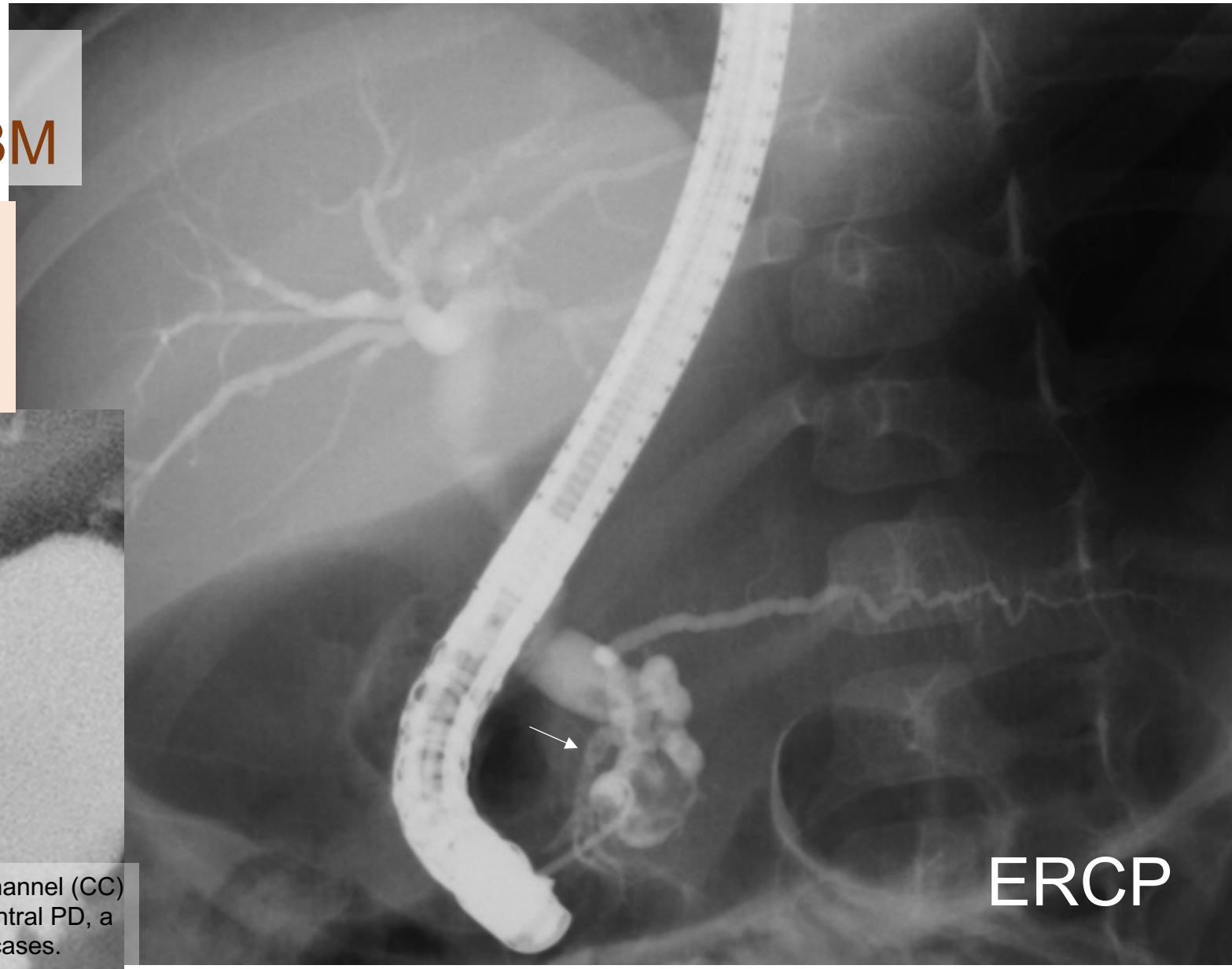
Case 45 Protein plugs complicate PBM

A 1-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 the appearance of PBM.
Most of the plugs had disappeared by the time of surgery.



Dilatation of the common channel (CC) extends upstream to the ventral PD, a feature often seen in PBM cases.

On-table cholangiography



Arrows indicate the same PD branch

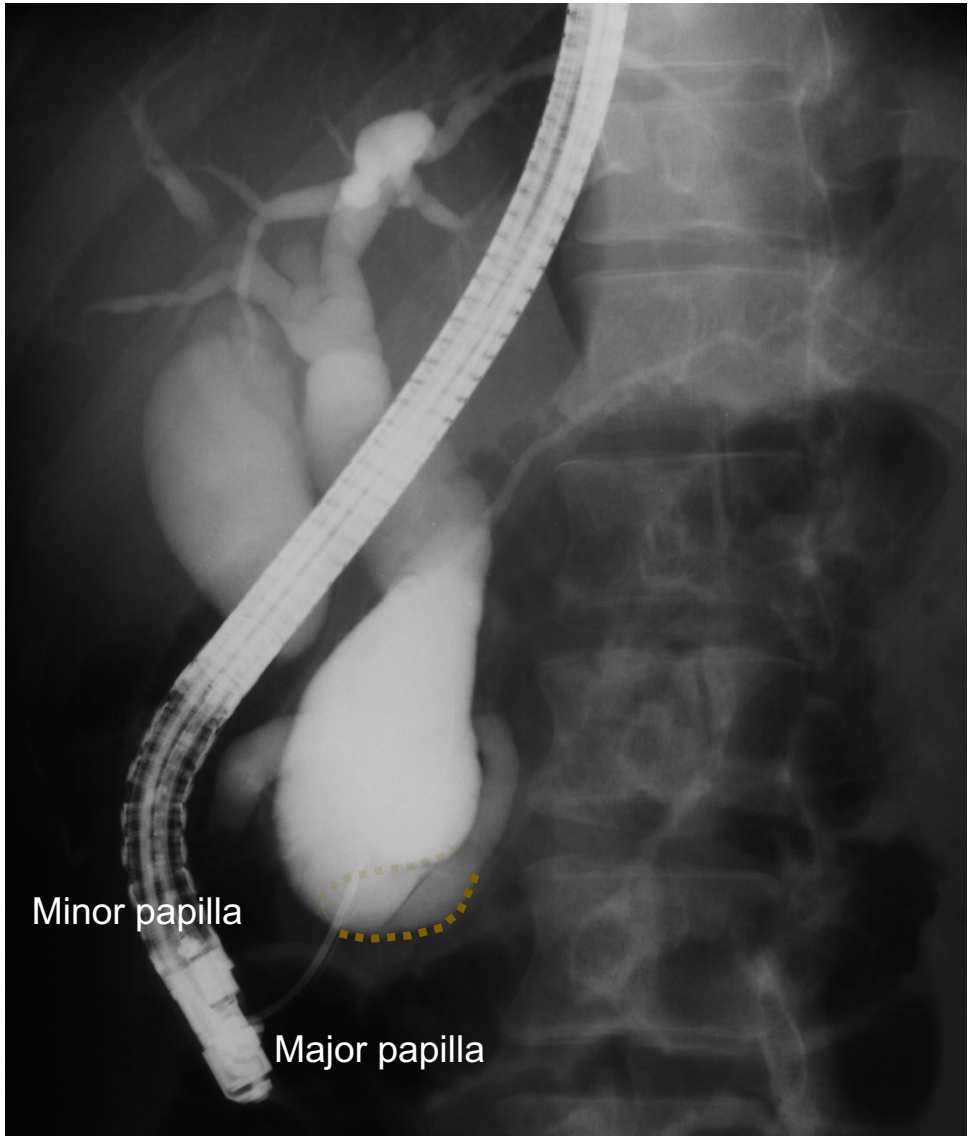
Case 46

Main (ventral) pancreatic duct forms the cyst bottom

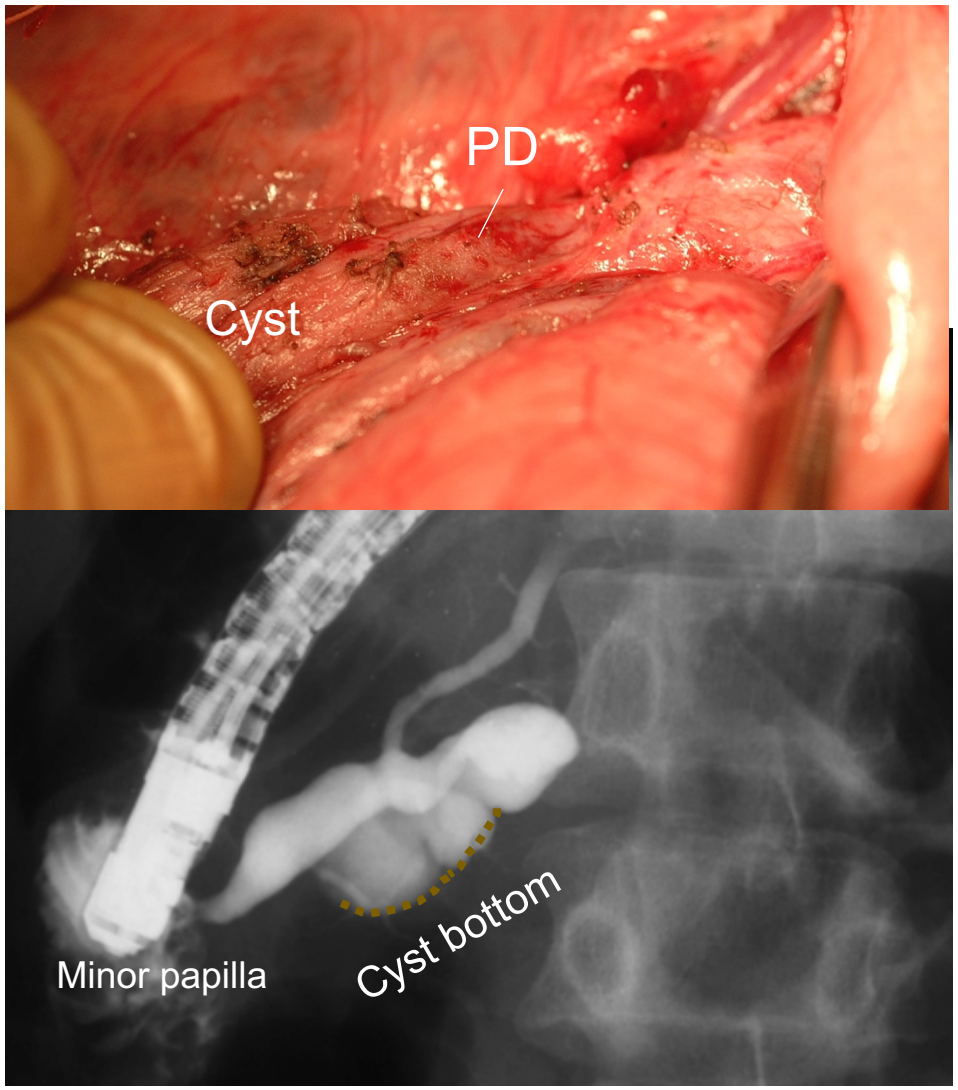
A female in her 20s developed abdominal pain with hyperamylasemia caused by protein plugs.

Because the narrow segment, which the cyst was expected to have, was not found during surgery, ERCP was performed to disclose the relationship with PD.

The guidewire through the main papilla was observed running along the bottom of the cyst.



Dotted line indicates where the cyst bottom was composed of the PD



Pancreatography through the minor papilla after cyst excision

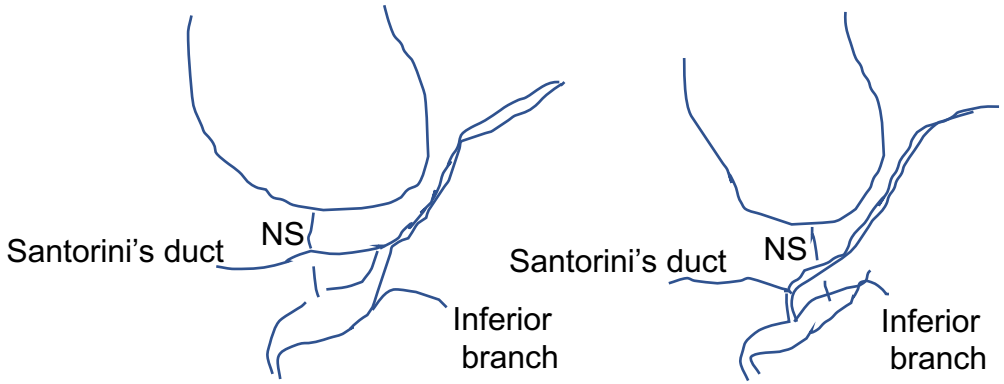
Case 47

Rare type
Bile duct joins the inferior branch of the PD

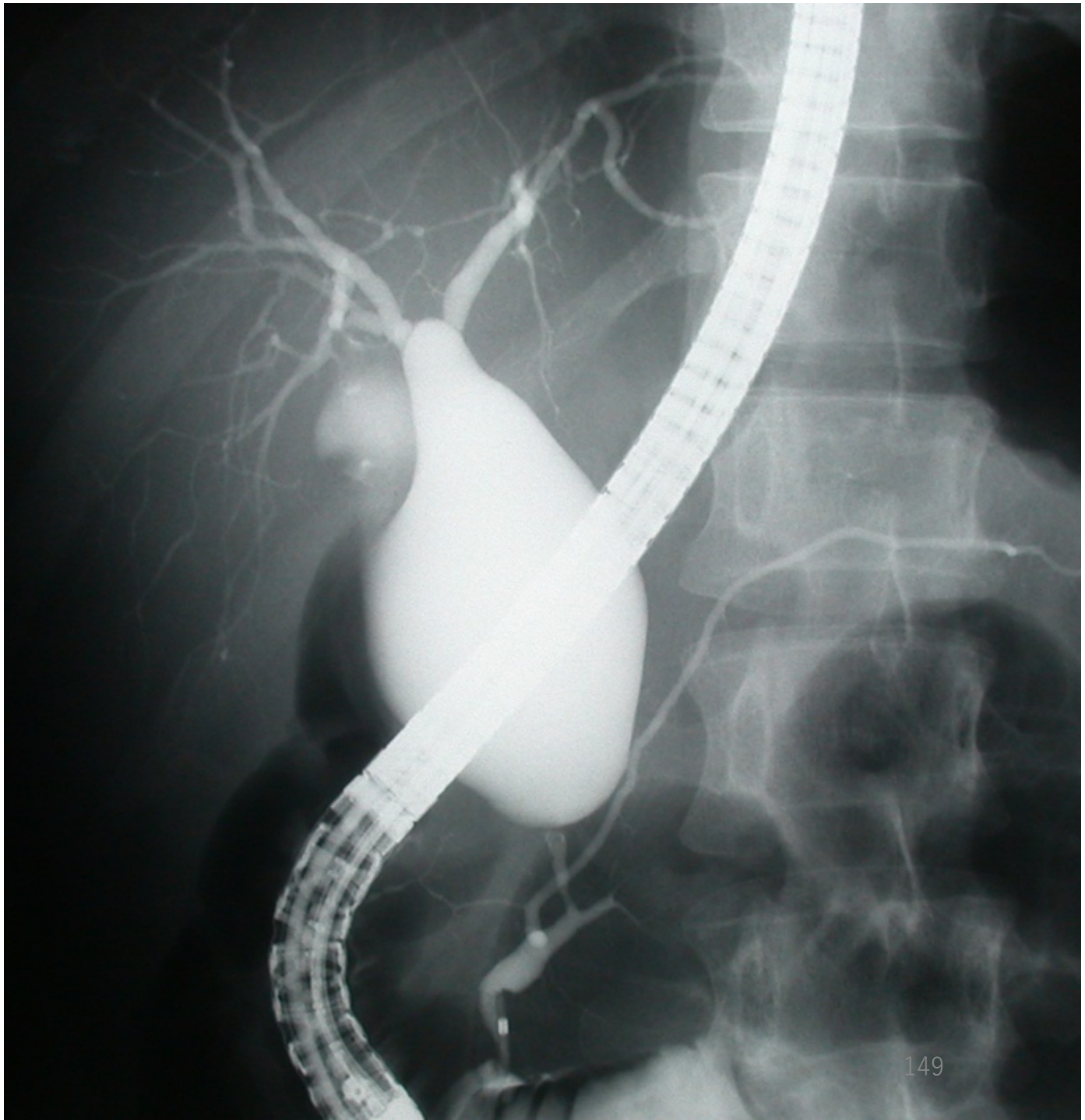
A male in his 20s.
Ultrasonography detected a dilated BD during a medical checkup.

Type Ia congenital biliary dilatation.

- **The BD joins the inferior branch of the PD**
  This case supports Matsumoto's theory that PBM is a malformation in which the choledochus misconnects with the ventral PD or its branches outside the duodenal wall (see p158).



Normal　　This case

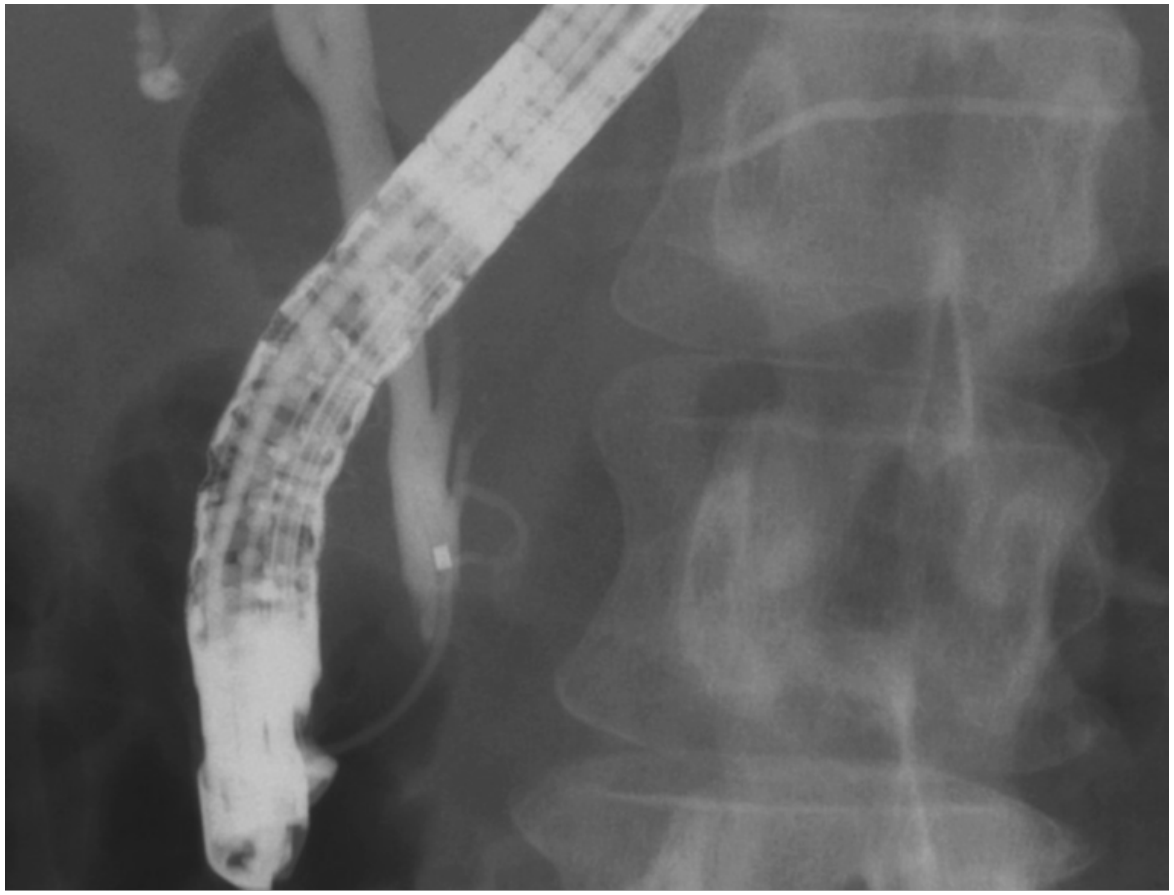


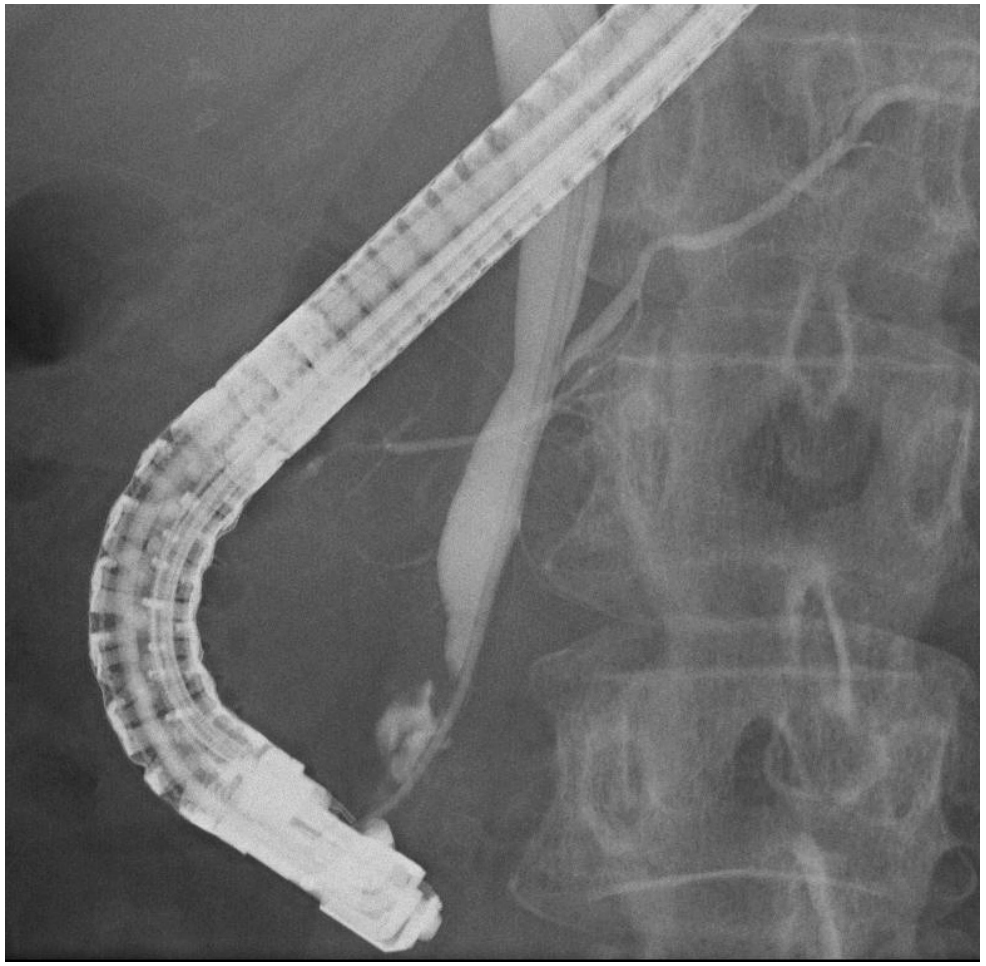
Advanced

Common channel and narrow segment

First version, September 2024

Is the common channel a pancreatic duct or a bile duct?





PD branches arising from the common channel indicate that it is a pancreatic duct

Dilated common channel composed of both PD and BD

Proposed by Ando H, in 1999

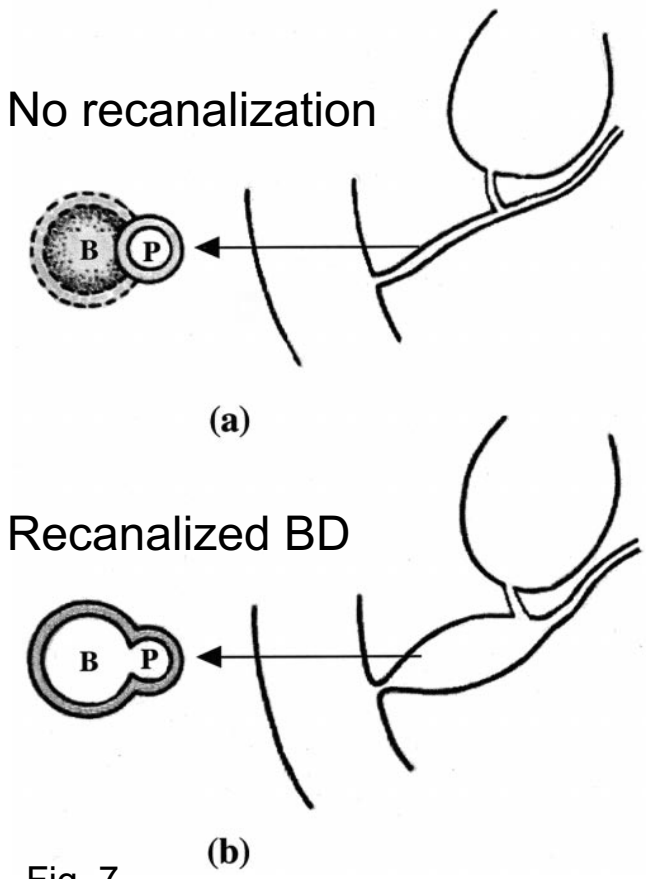
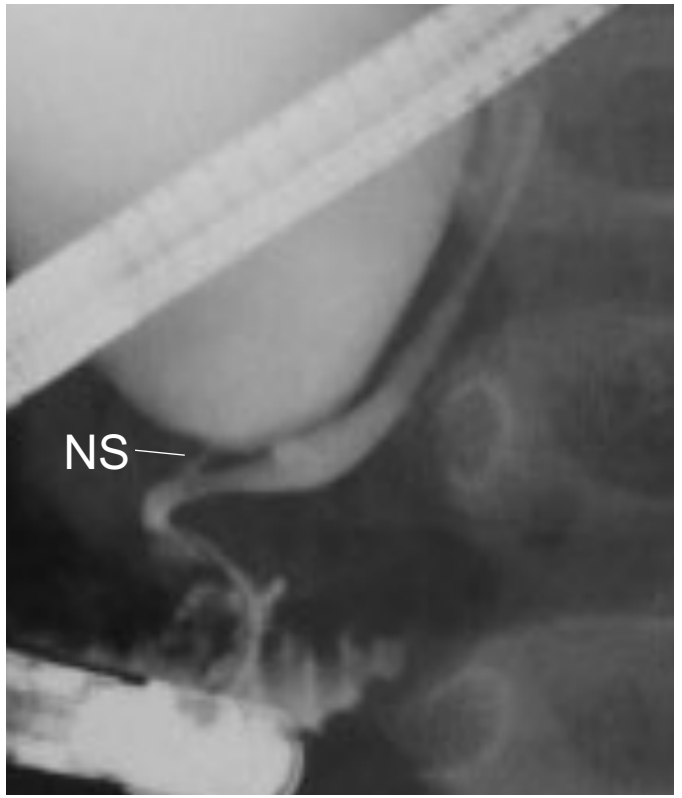
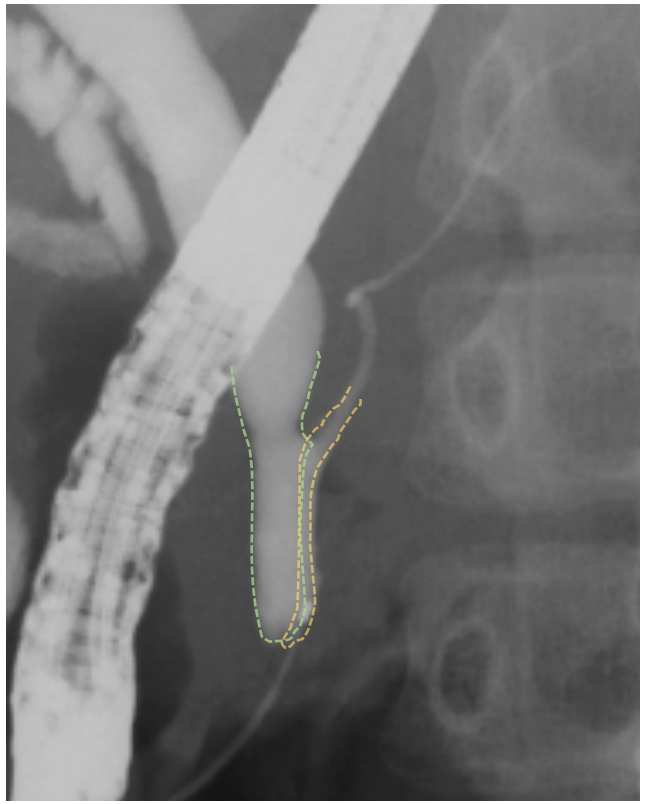


Fig. 7

Ando H, Kaneko K, Ito F, Seo T, Harada T, Watanabe Y. Embryogenesis of pancreaticobiliary maljunction inferred from development of duodenal atresia. *J Hepatobiliary Pancreat Surg.* 1999;6(1):50-54



PD alone



BD + PD

Case from *Basic Section* See p44.

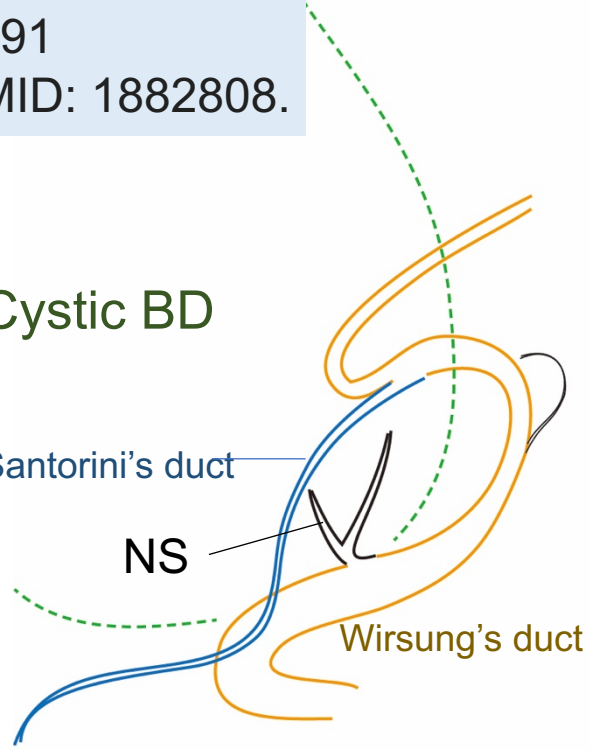
NS is a branch of the PD

Histopathological demonstration

Suda K, Matsumoto Y, Miyano T. Narrow duct segment distal to choledochal cyst. *Am J Gastroenterol*. 1991 Sep;86(9):1259–63. PMID: 1882808.

Case 9

ERCP in a 1-year-old girl.
Vomiting and elevated pancreatic enzymes.



NS is one branch of the PD

Radiological demonstration

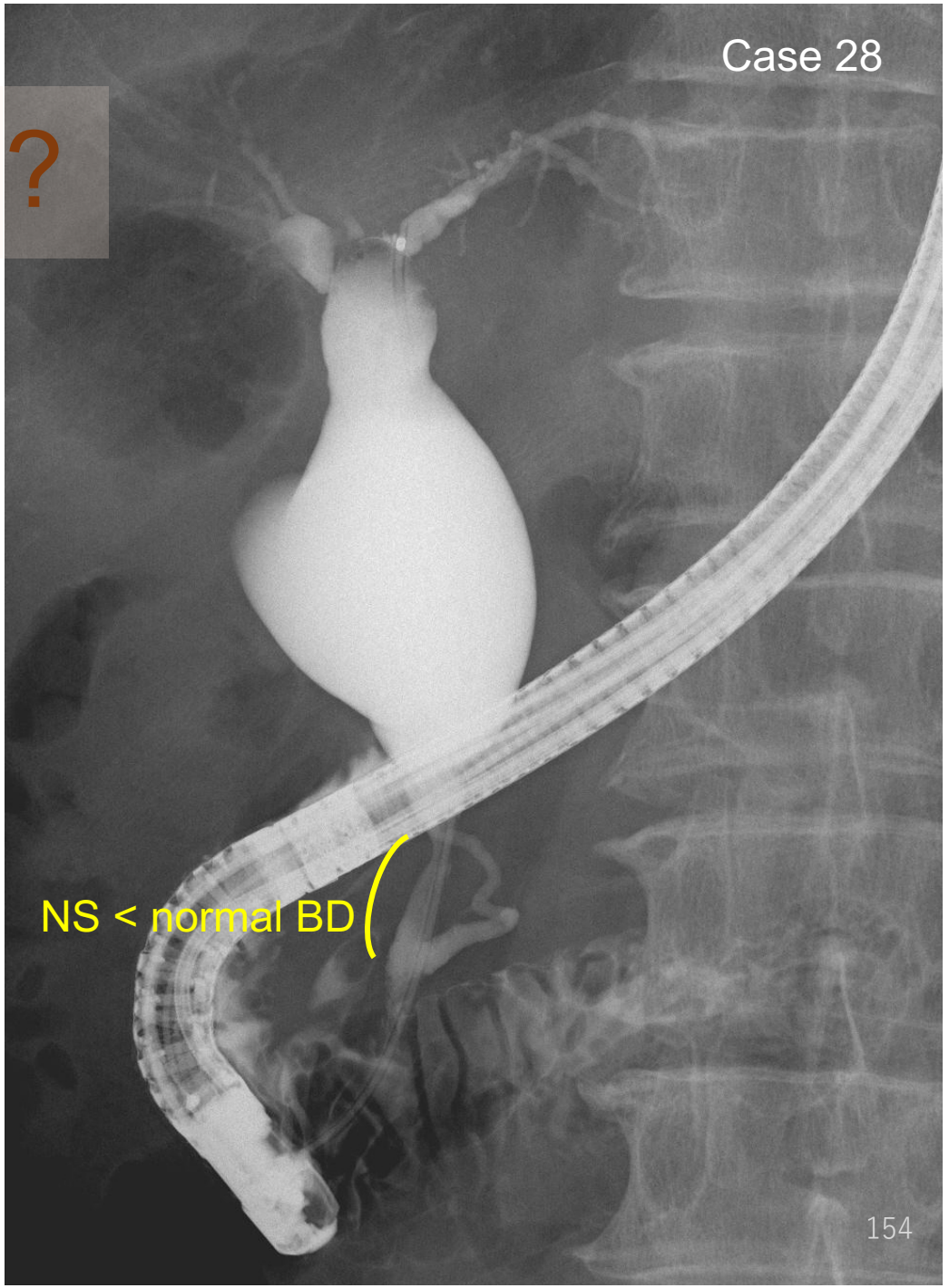


Matsumoto's theory of PBM embryogenesis:
PBM results from the BD misconnecting with the ventral PD or its branches (see p158).

Is NS really a PD branch?

In some cases of type Ic fusiform dilatation, the narrowed portion beneath the dilated BD may be considered a normal BD rather than a narrow segment (NS).
There is no abrupt narrowing in these cases.

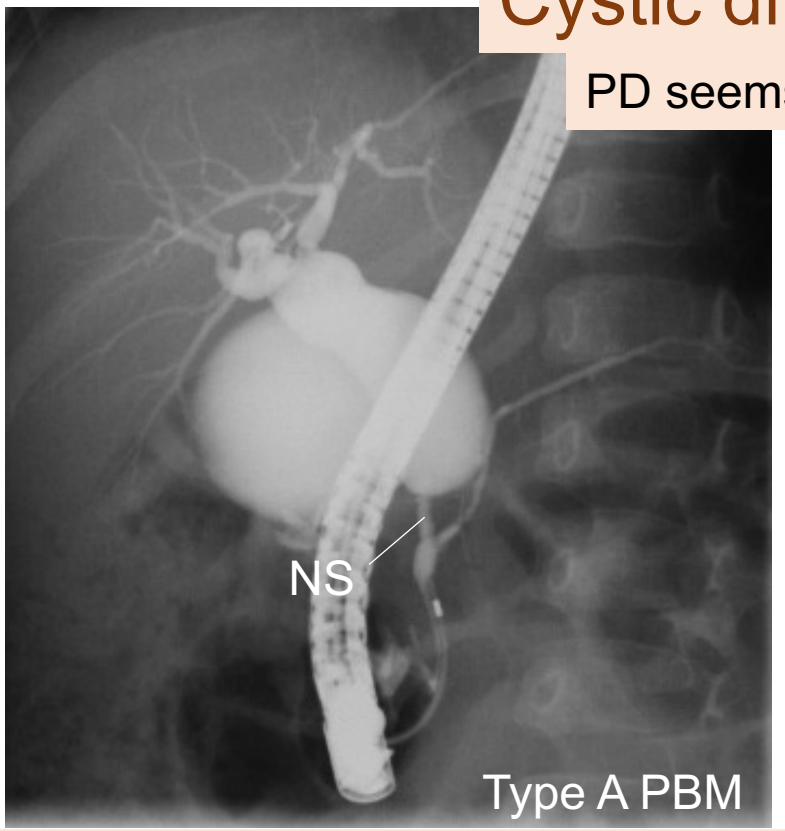
- JSGPBM classification (see p33): **Type B (non-stenotic type)** rather than Type A (stenotic type)
- The BD should be resected completely down to the PD using fluoroscopy with clips (see next page and p73).

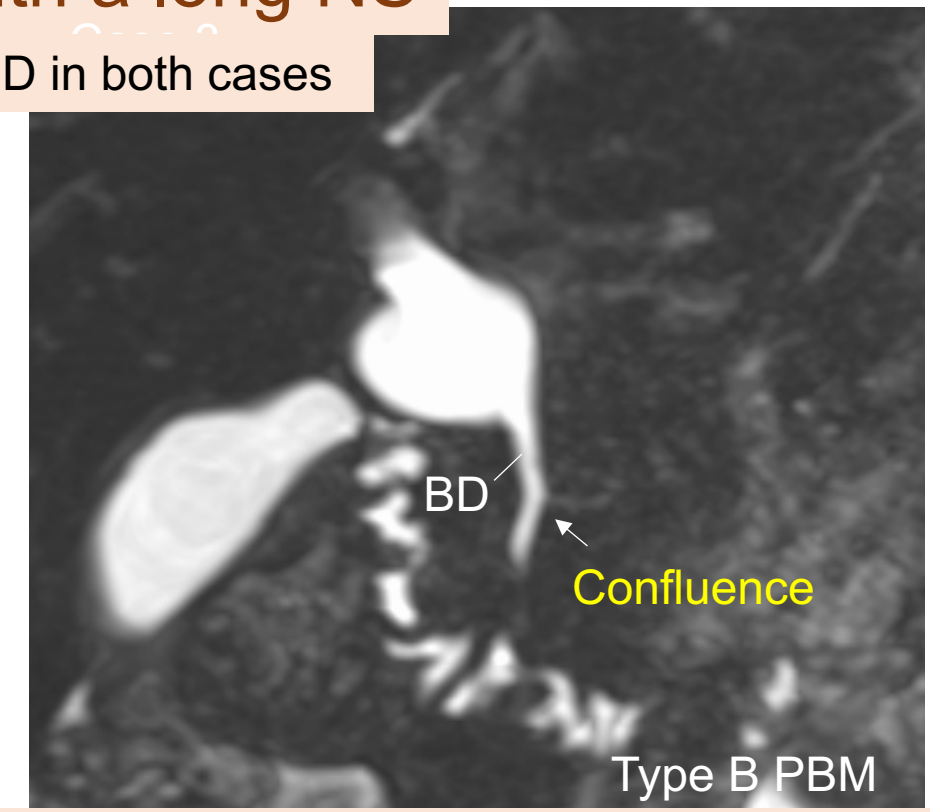


Is NS really a PD branch?

Cystic dilatation with a long NS

PD seems to enter the BD in both cases

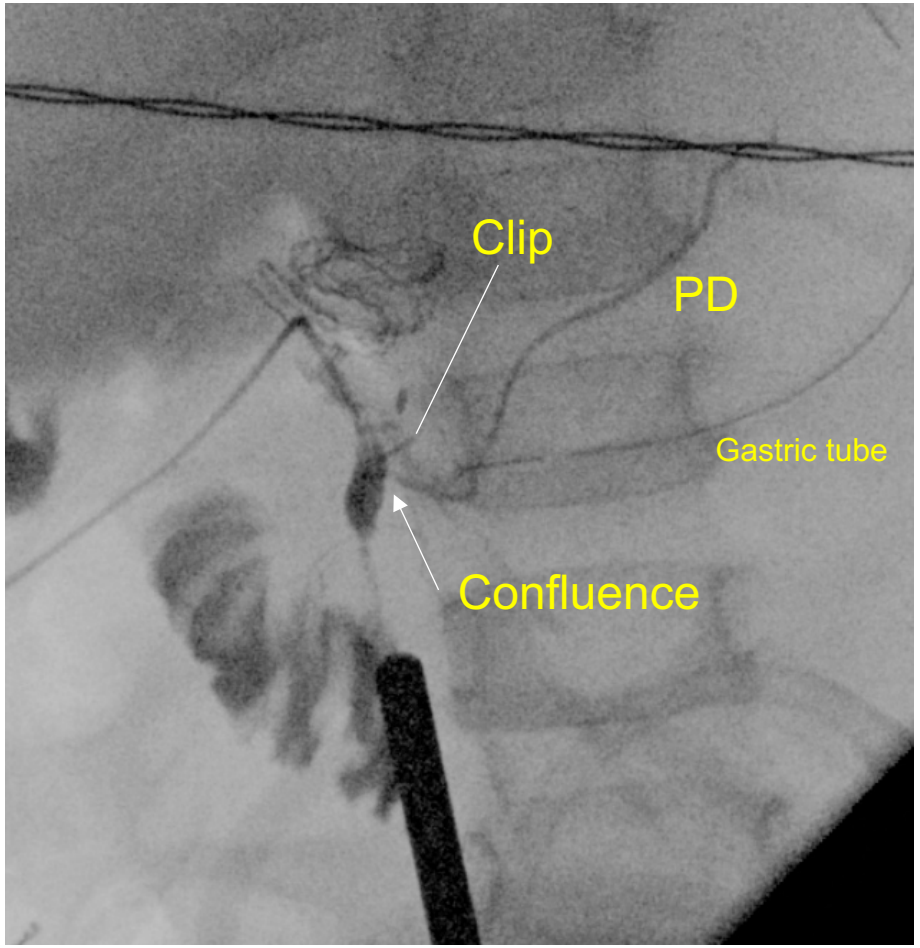


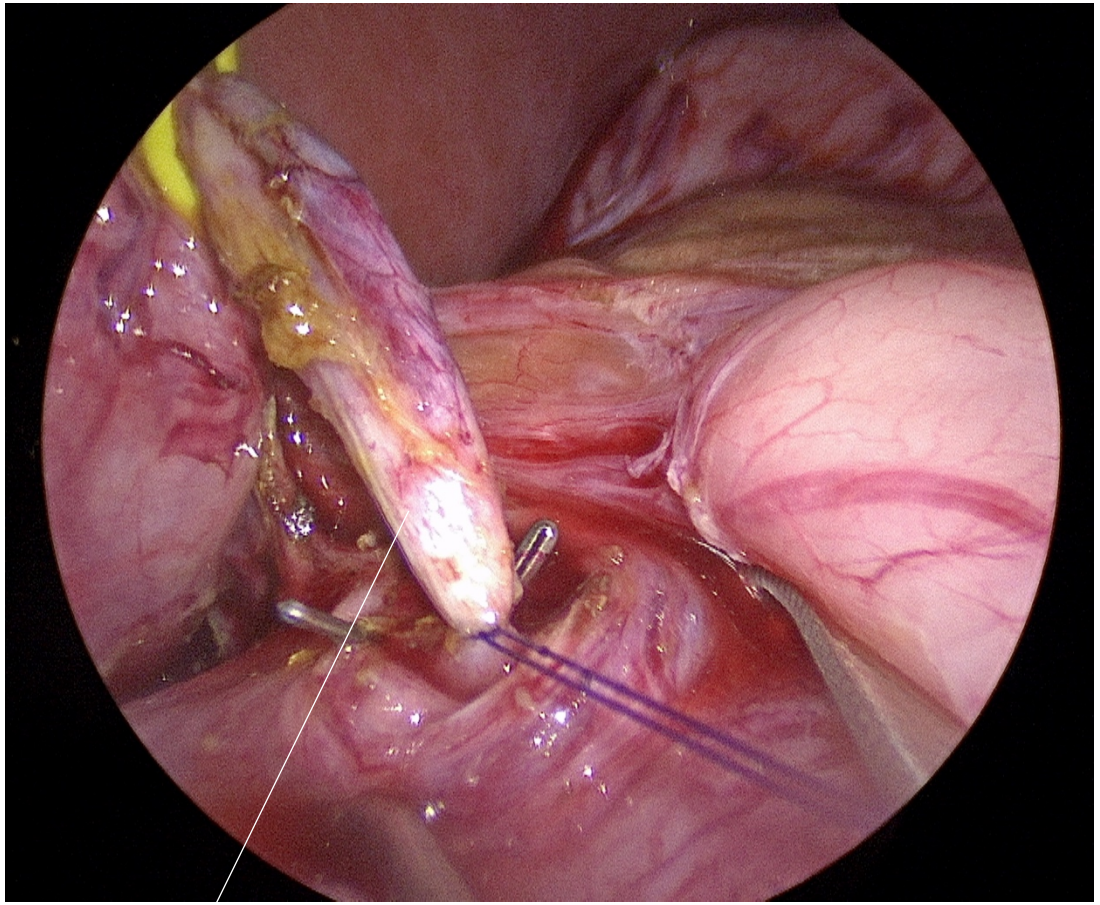


A 1-year-old child.
Vomiting, with elevated serum liver enzymes and amylase.
The BD was resected at the obvious NS.

A 3-year-old girl.
Abdominal pain, vomiting, and elevated serum liver and biliary enzymes.
No abrupt narrowing was identified, requiring dissection down to the PD using fluoroscopy with clips (see next page).

Surgery confirmed the absence of a true NS.
The BD was resected just above the PD using fluoroscopy with clips.





The narrowing portion on MRCP was actually a normal BD.

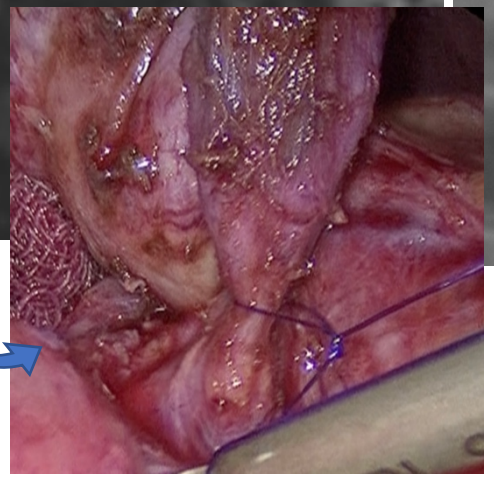
Is NS really a PD branch?

A PD branch and a narrowed BD may compose the NS

Pediatric case

Portion of BD

Portion of PD branch



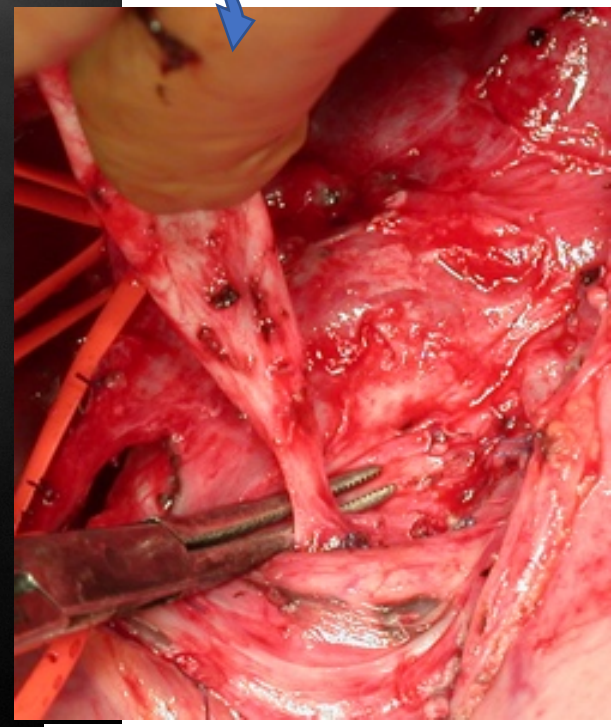
Surgery-confirmed NS

Adult case

Loop type of PD

Portion of BD

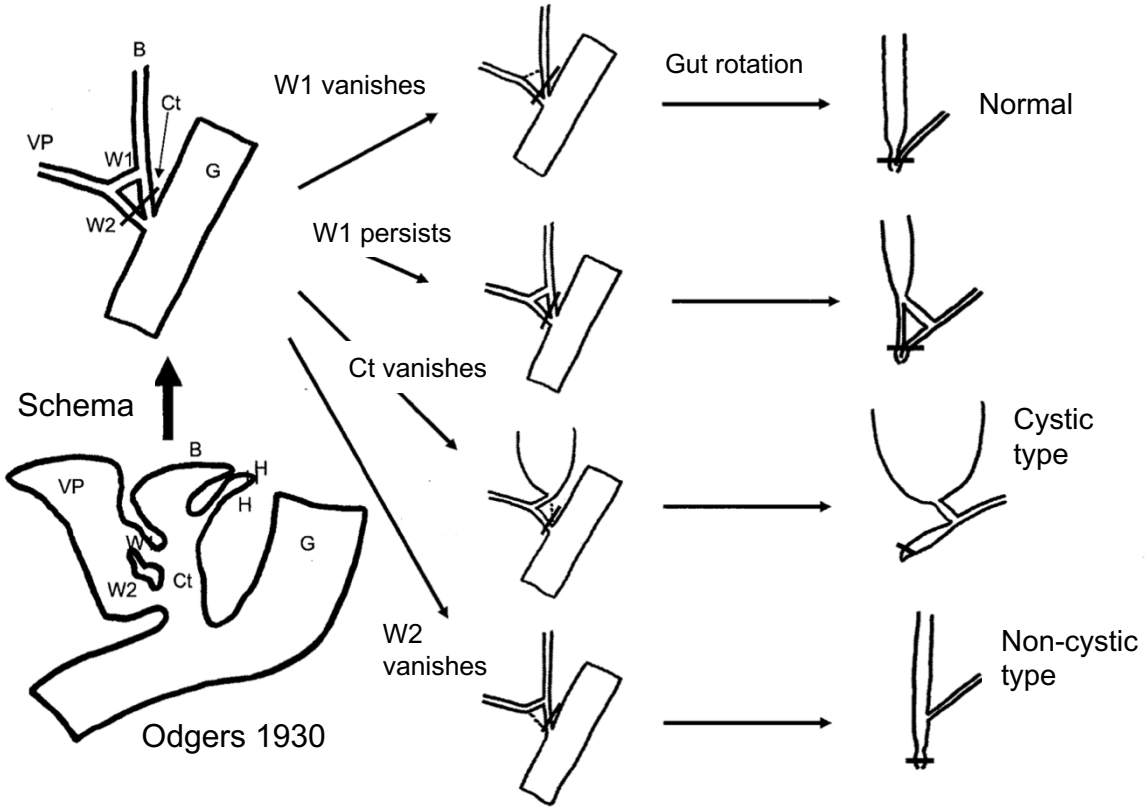
Portion of PD branch



Surgery-confirmed NS

Matsumoto's theory of PBM embryogenesis:
PBM is a malformation in which the choledochus misconnects with the ventral PD or its branches outside the duodenal wall (see next page).

PBM embryogenesis



Cited from Itaru Oi. *Tokyo Women's Medical University Journal* 2008;78:55-60.

Oi's theory of PBM embryogenesis in 1977

Estimation based on Odgers's embryogenesis of PD (1930)

W1, Left ventral pancreas duct; W2, right VP duct; Ct, terminal choledochus

Two classic theories

Confluence site of the BD and ventral PD in congenital biliary dilatation



Cited from Yoshiro Matsumoto. *Nippon Shokakibyo Gakkai Zasshi* 1986;19:614-23.

Matsumoto's theory of PBM embryogenesis in 1986

Based on the fact that the BD never enters the dorsal PD in PBM

He proposed that PD branches arising from the common channel (CC) on cholangiopancreatography (see p151) indicated that the CC was functionally a pancreatic duct. He estimated that PBM is a malformation in which the choledochus misconnects with the ventral PD or its branches outside the duodenal wall.

See Cases 9* and 47**.

PBM embryogenesis

Ando H, Kaneko K, Ito F, Seo T, Harada T, Watanabe Y. Embryogenesis of pancreaticobiliary maljunction inferred from development of duodenal atresia. *J Hepatobiliary Pancreat Surg.* 1999;6(1):50-4*

Congenital duodenal atresia and stenosis

Disorder of recanalization from the solid stage

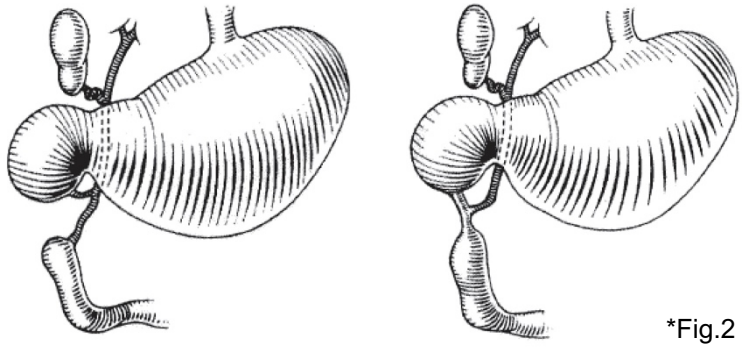
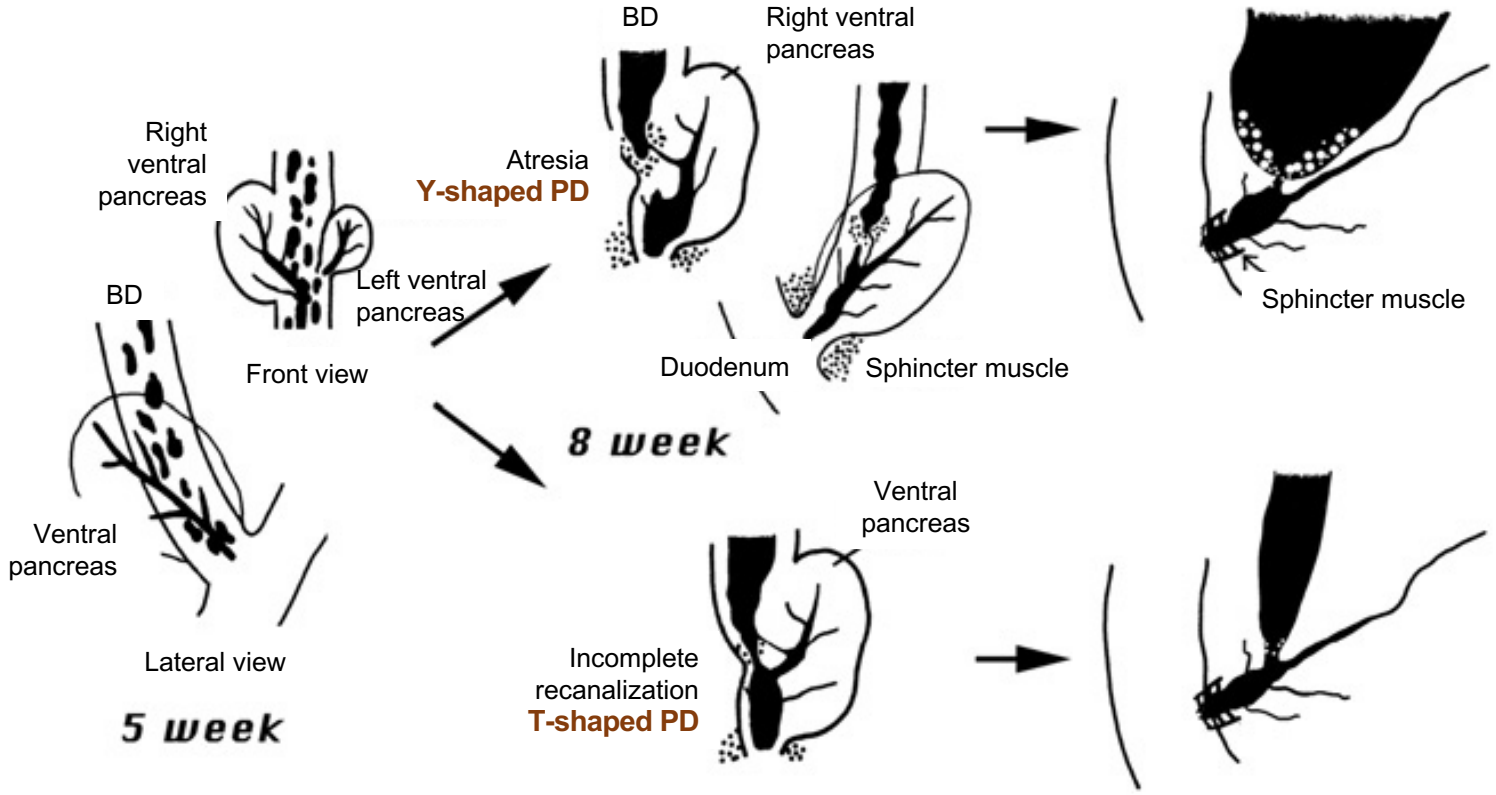


*Fig.2

Atresia
Y-shaped BD

Stenosis
T-shaped BD



Cited from Hisami Ando. *Tando (Biliary Tract).* 2008;22175–80.

Inferred from the embryogenesis of congenital duodenal atresia and stenosis, the BD passes through a solid stage during its development, similar to the duodenum, whereas the PD does not experience a solid stage.



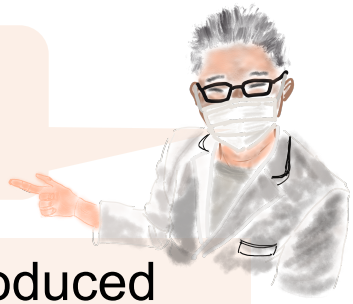
Advanced

What causes biliary dilatation?

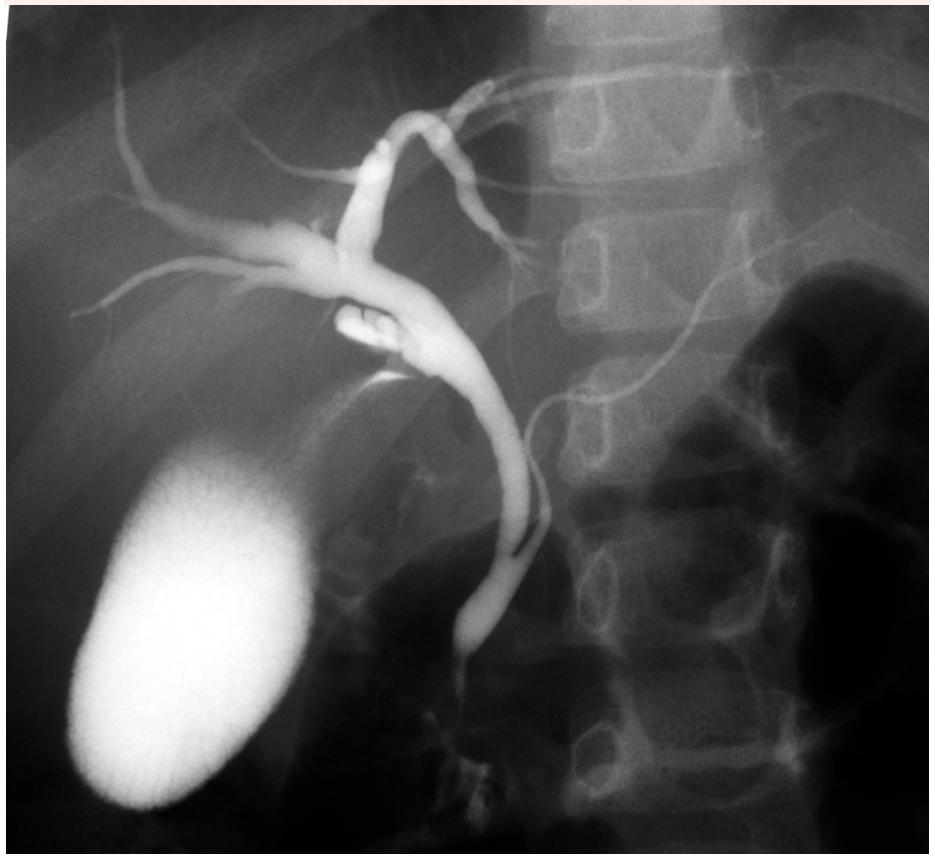
First version, September 2024

Regurgitation of pancreatic juice does not cause biliary dilatation

When the idea of PBM became popular in Japan, it was thought that bile duct injury due to pancreatobiliary reflux was the cause of dilatation

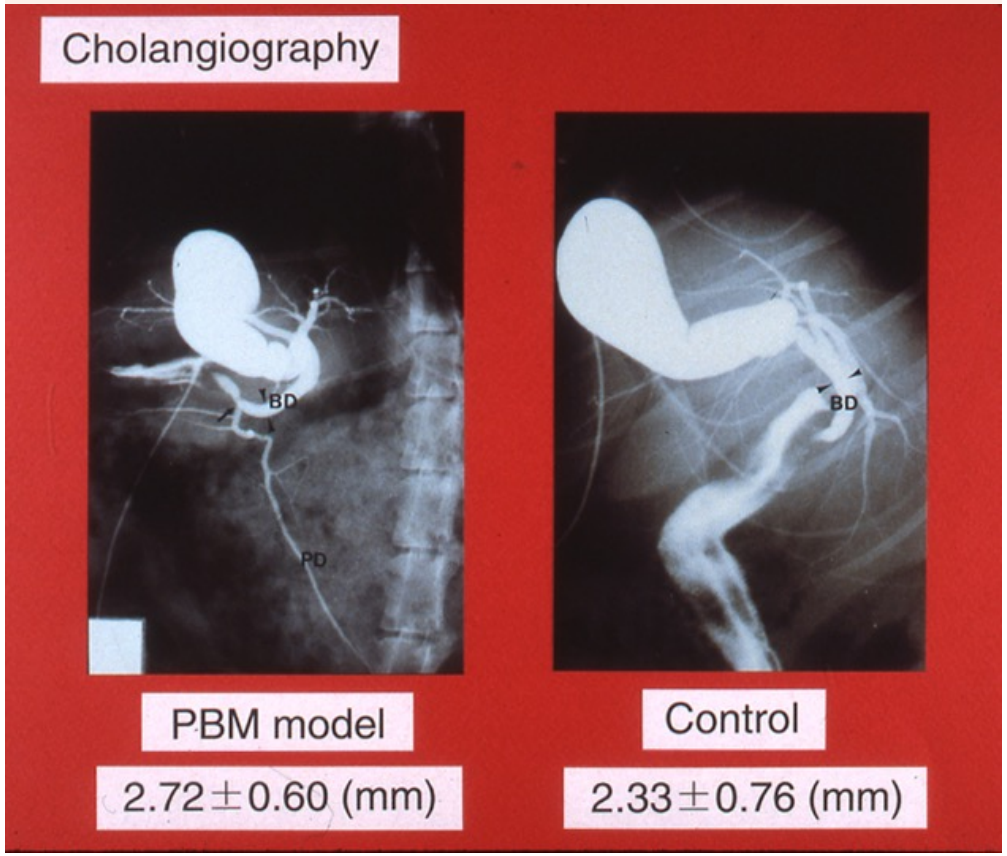


Non-dilated type of PBM exists



Pediatric case of non-dilated PBM
The common channel is dilated but the BD is not.

Cystic dilatation was not produced in animal PBM models

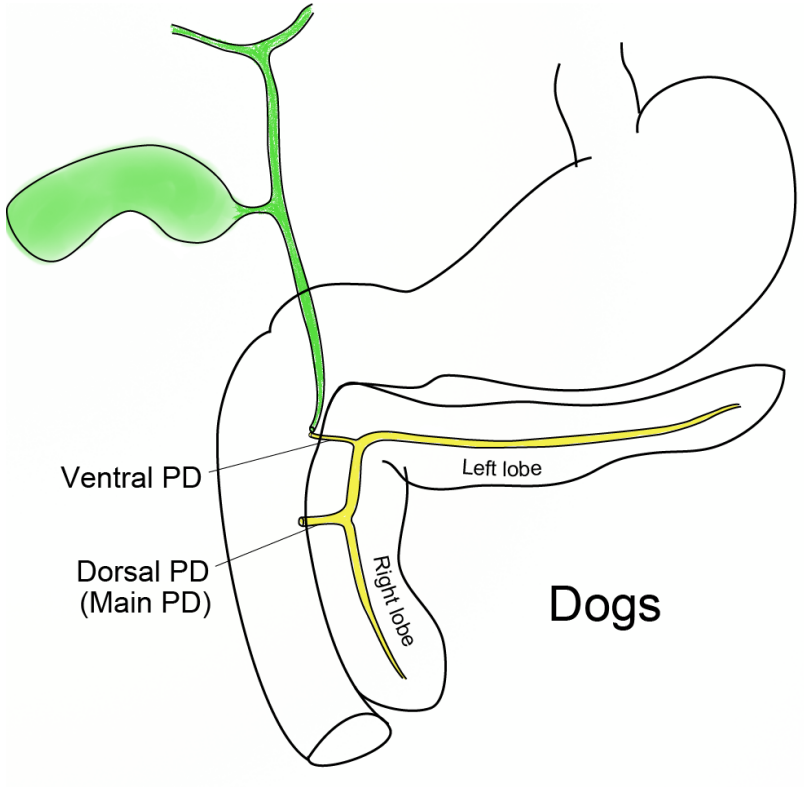


Cat model of PBM
The BD did not dilate.

Kaneko K, et al. *J Surg Res.* 1996;60:115-21.

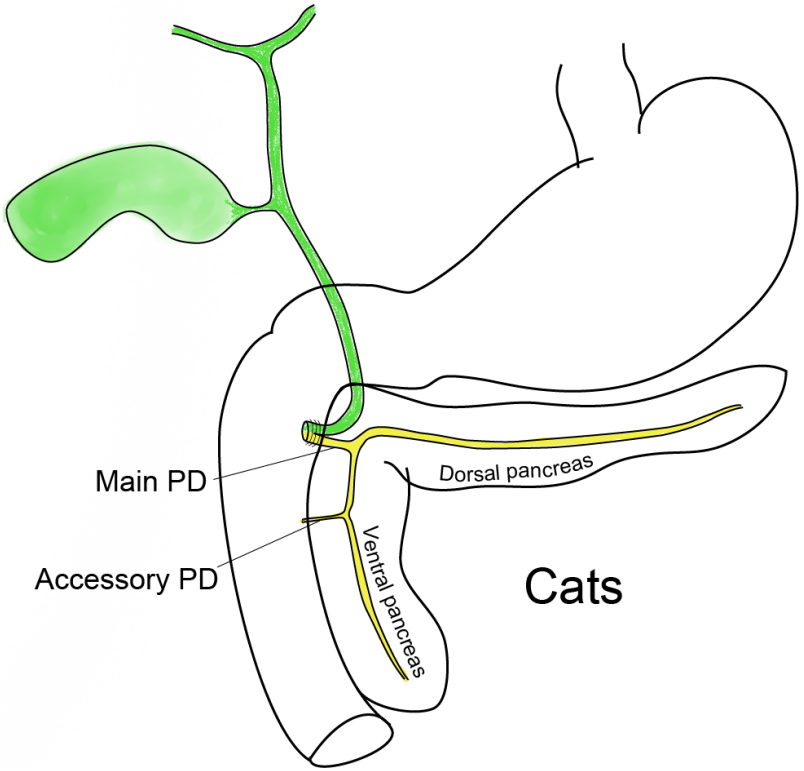
Pancreatobiliary anatomy differs among animals

BD joins the accessory PD



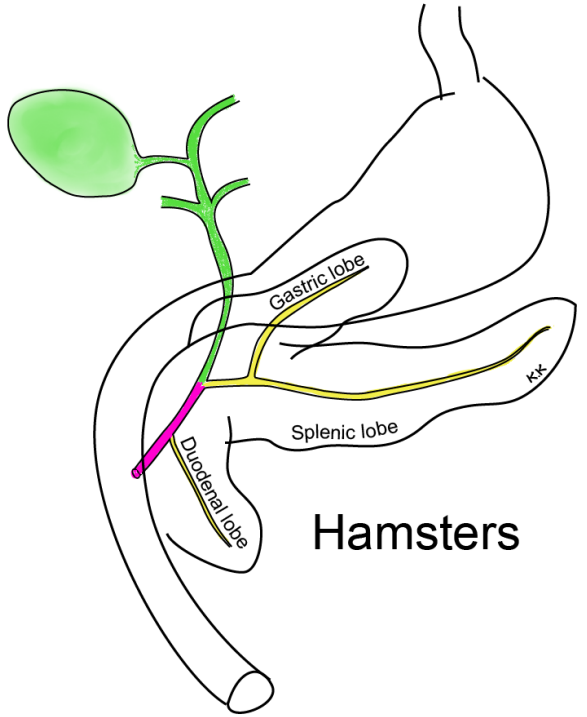
Dogs

Similar to human anatomy



Cats

PBM is normal in rodents, goats, and sheep.



Hamsters

The cause may be the narrow segment (NS)

Dilatation related with NS length — Ando H. *Jpn J Surgery.* 1983;84:1174–85
Increased intraductal pressure due to NS — Davenport et al. *J Pediatr Surg.* 2005;40:331–5



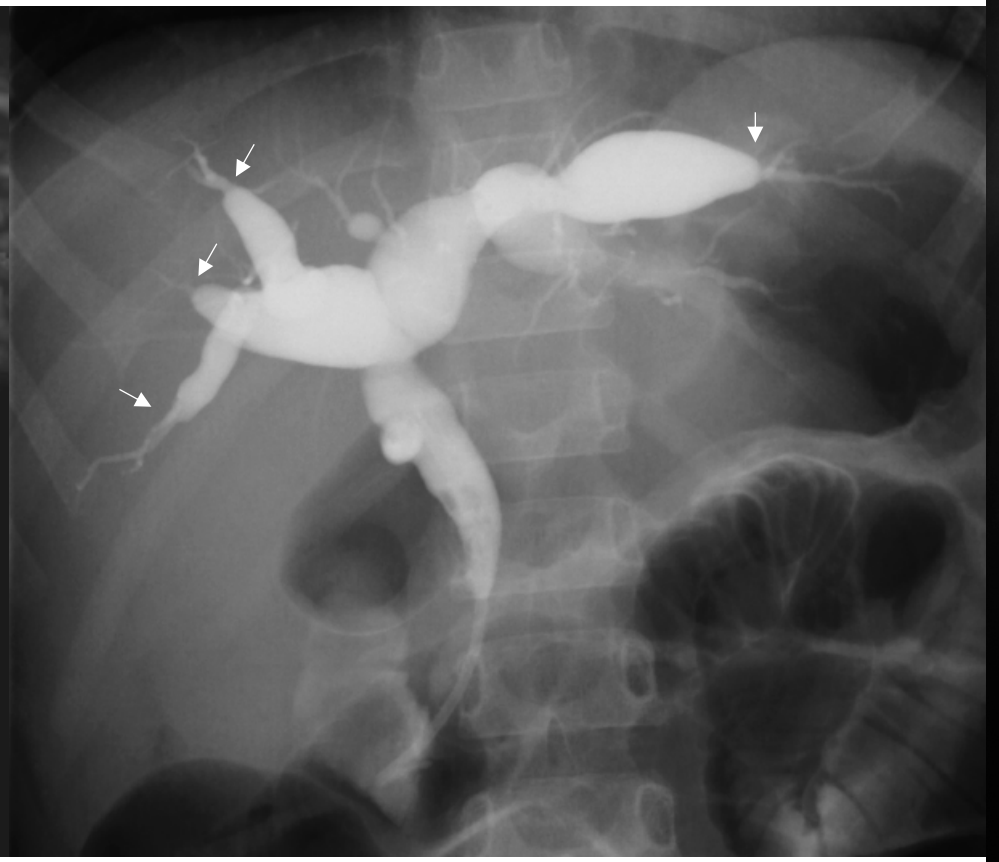
No NS | Slight constriction | Thick NS | Thin NS

The NS alone cannot explain characteristic dilatation

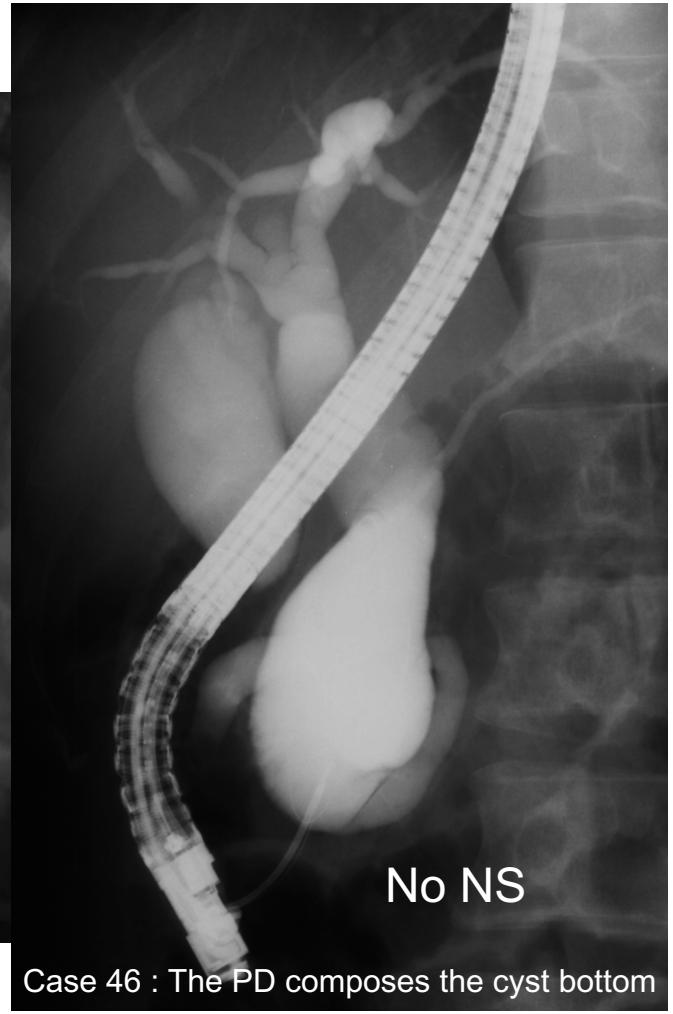
Increased ductal pressure cannot cause limited dilatation (see p11).



Localized dilatation



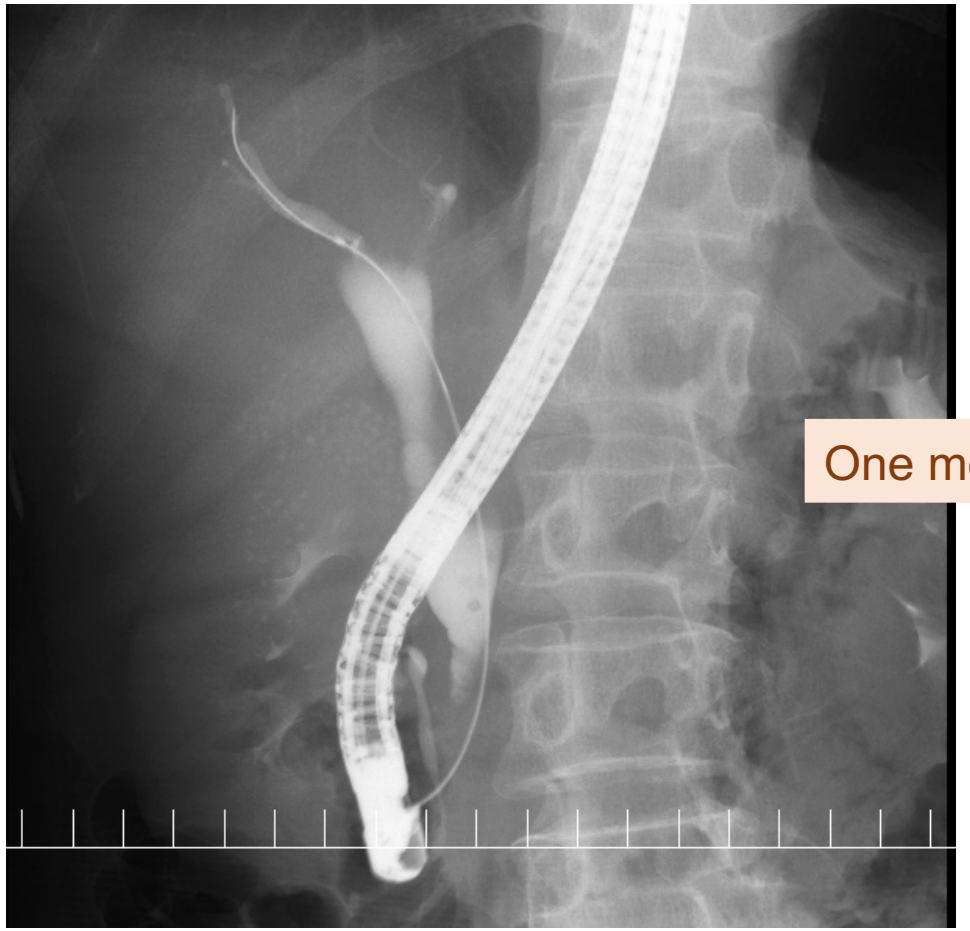
Localized dilatation
Intrahepatic bile ducts are more dilated



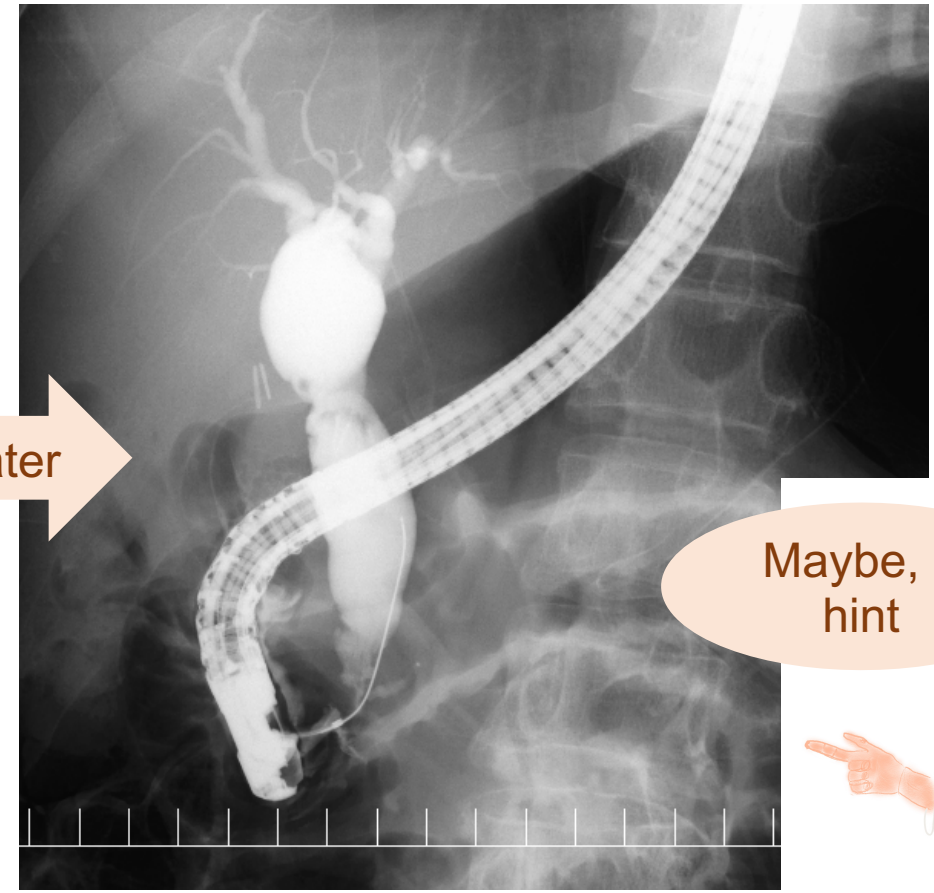
Case 46 : The PD composes the cyst bottom

Acquired localized BD dilatation

Chronic intermittent increase in intraductal pressure



One month later

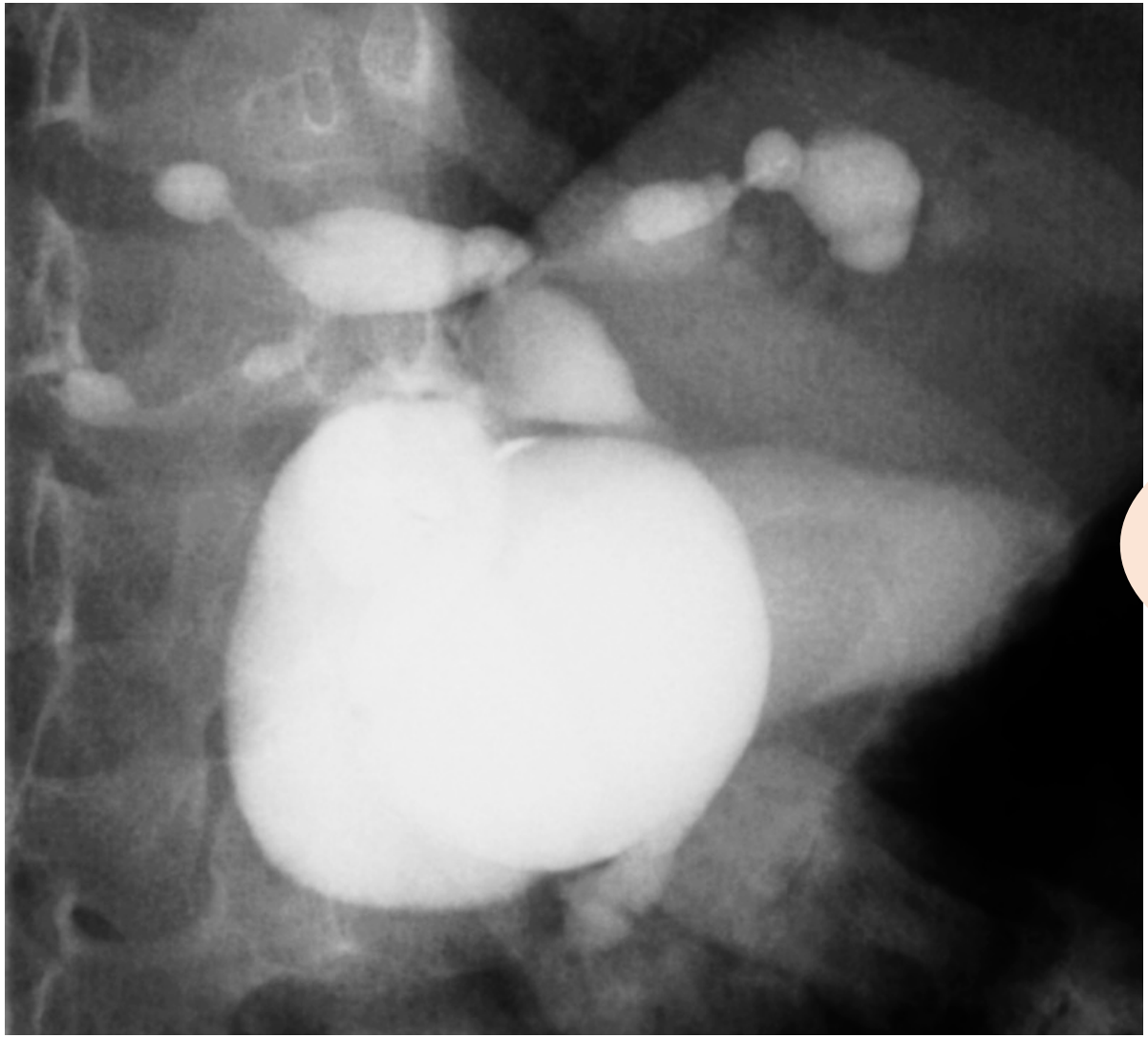


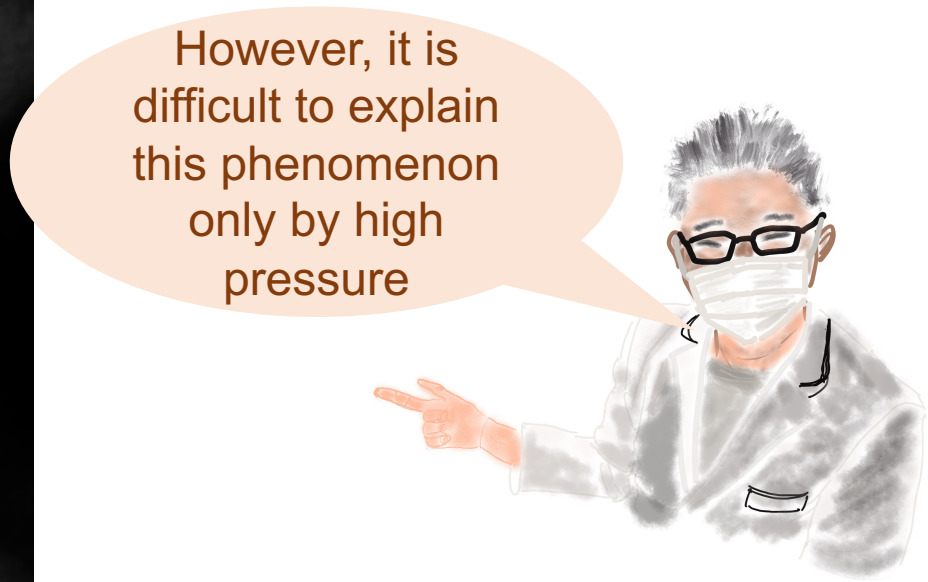


The common hepatic duct became larger but the upstream BDs did not dilate.

A male in his 50s with bile duct stones

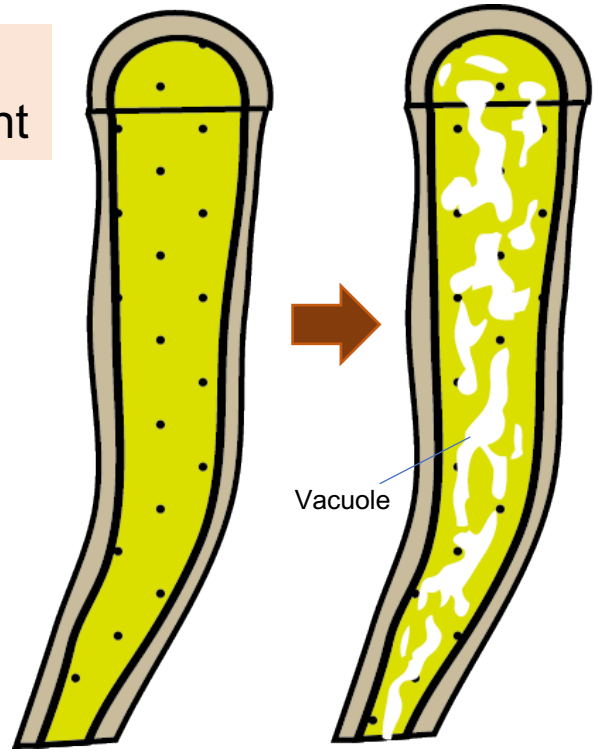
Peripheral cystic dilatation of intrahepatic BDs





Yotuyanagi's etiological theory in 1936
Localized dilatation in congenital biliary dilatation

Normal
BD development

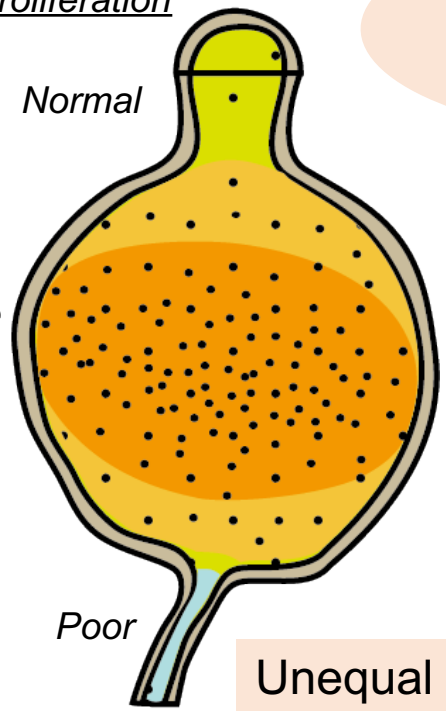


Biliary epithelia occlude the lumen

Solid stage

Recanalization

Epithelial proliferation



Solid stage

Yotuyanagi referred to Kodumi and Kodama's paper, which first described PBM as an etiology in 1916.



Yotuyanagi stressed both localized dilatation and the narrow segment. He pointed out that the NS alone cannot explain localized dilatation. His theory explains simultaneous existence of both.

Unequal epithelial proliferation

Yotuyanagi S. Contributions to the aetiology and pathogeny of idiopathic cystic dilatation of the common bile-duct with report of three cases; A new aethiological theory based on supposed unequal epithelial proliferation at the stage of the physiological epithelial occlusion of the primitive choledochus. *Gann* 1936;30:601–52.

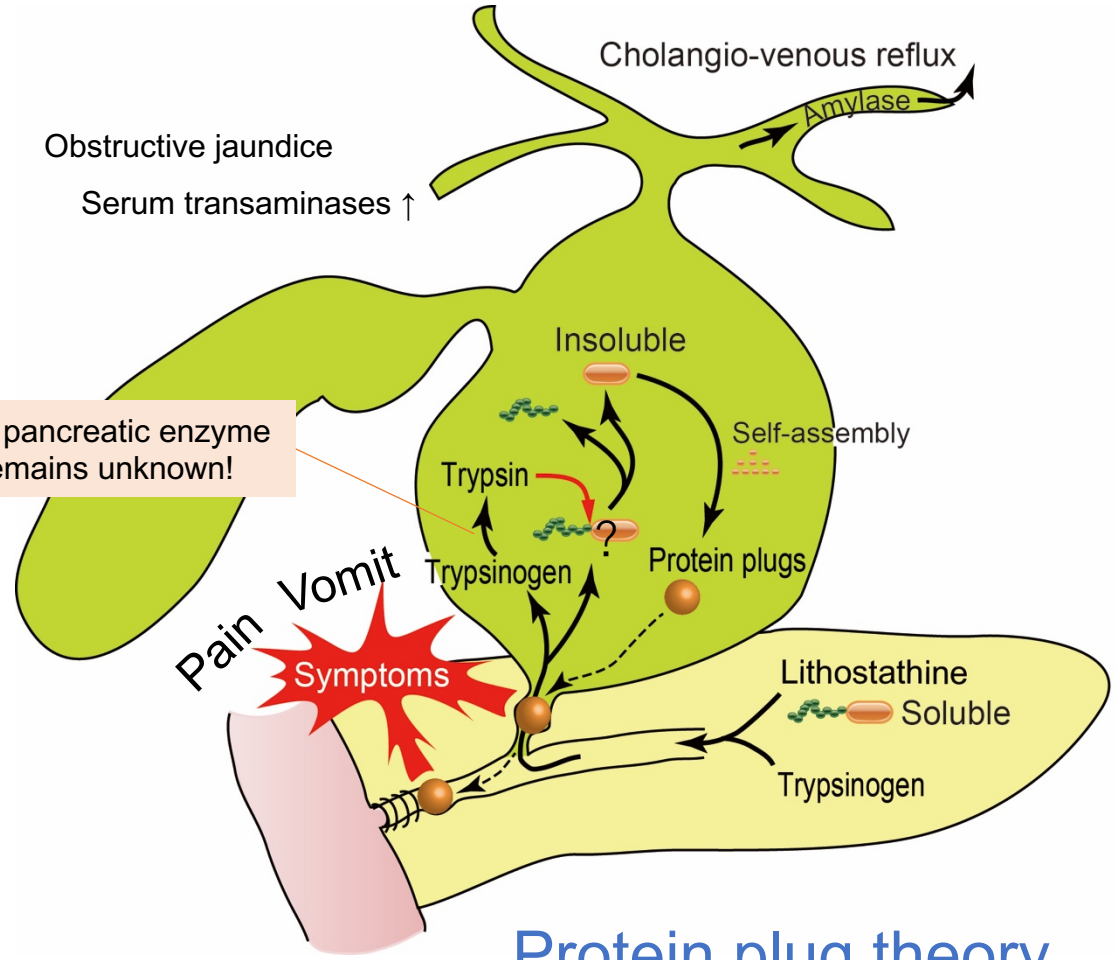
Advanced

Pathophysiology in pediatric patients

Mechanism of protein plug formation

First version, September 2024

Symptomatology in childhood (see *Basic Section*, p20–25)



Mechanism of pancreatic enzyme activation remains unknown!

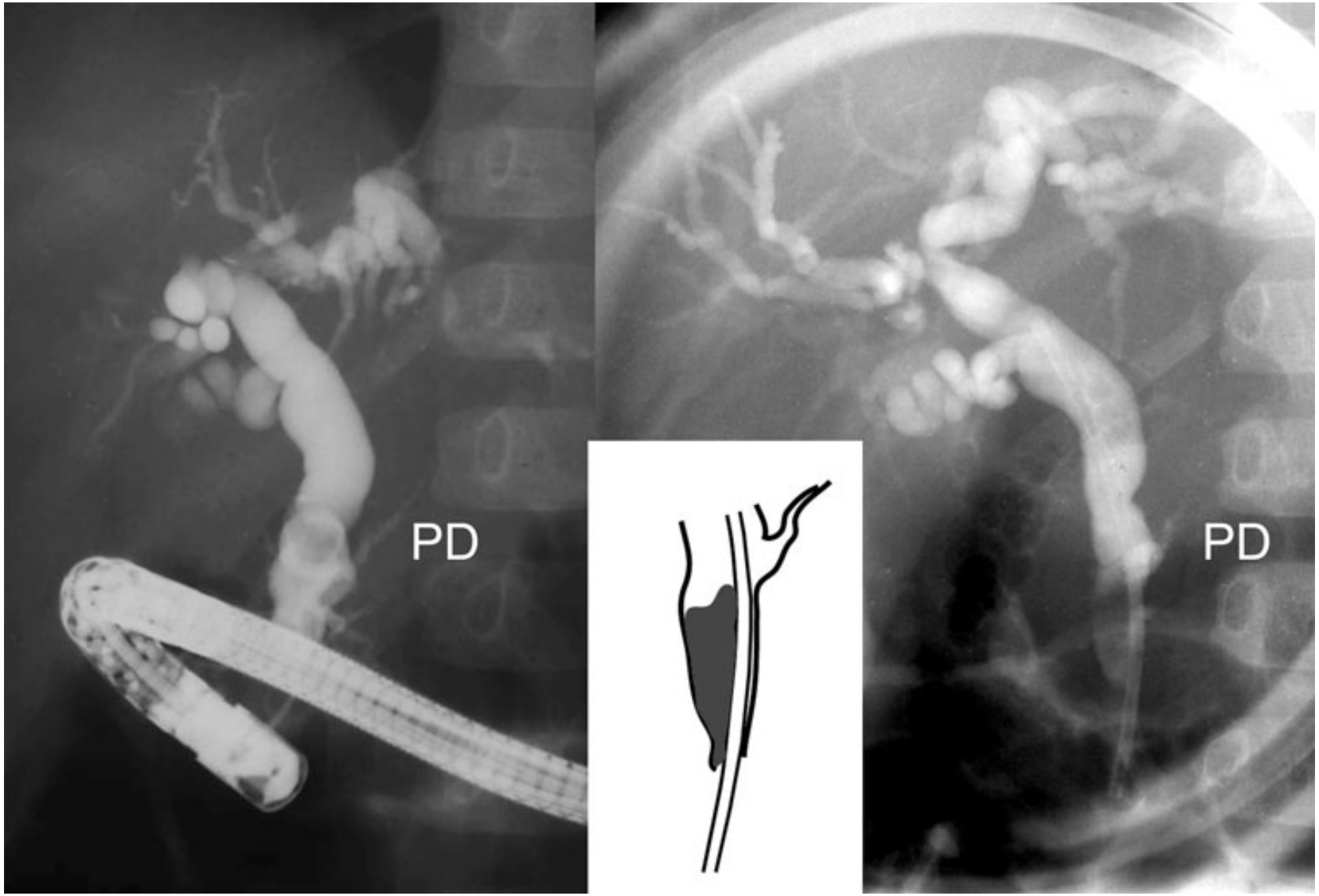
Protein plug theory



Fragile protein plugs occlude the narrow segment or the common channel, producing a transient increase in intraductal pressure and causing the symptoms

Findings supporting the protein plug theory

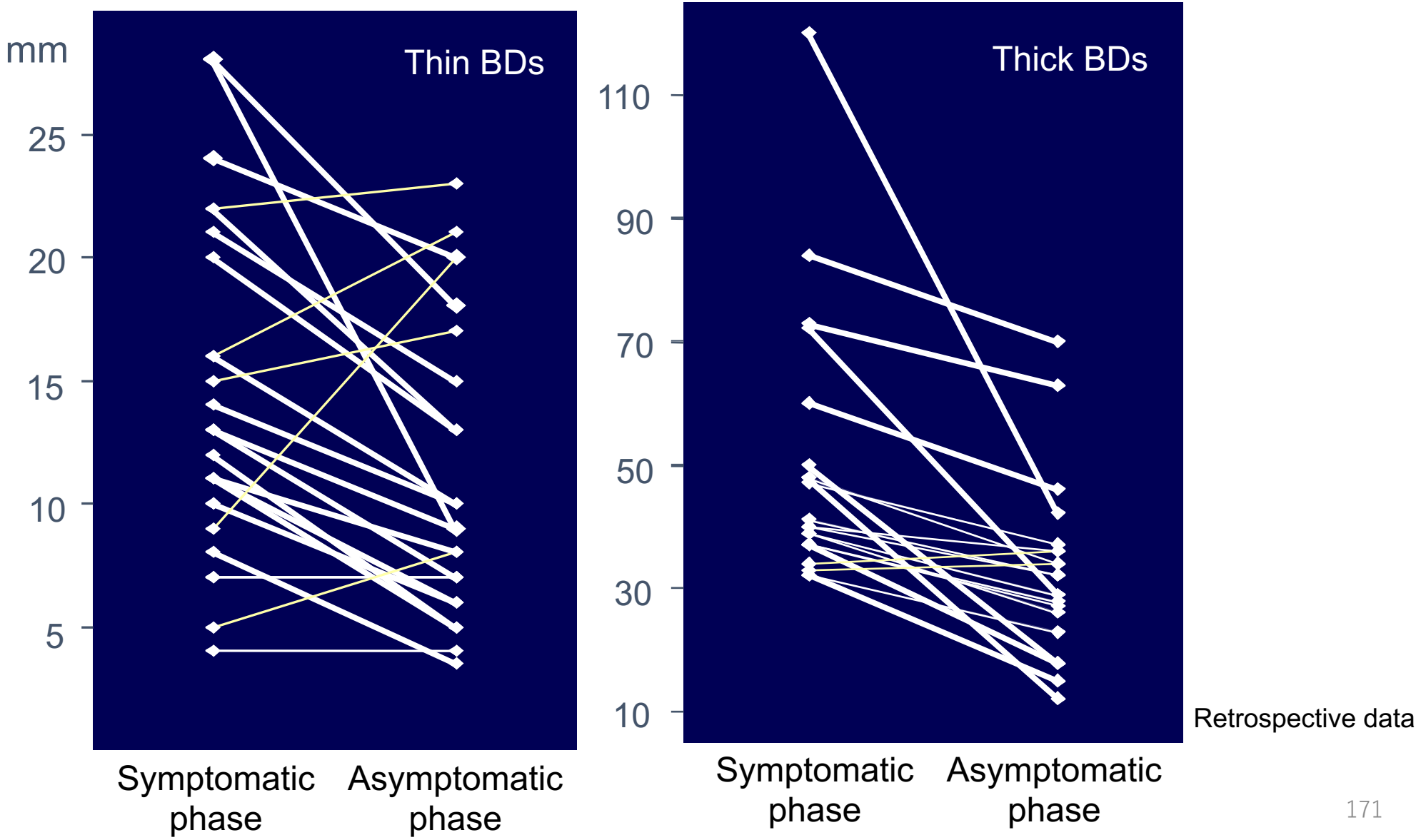
Endoscopic drainage promptly relieves persistent or exacerbated symptoms



Tsuchiya H, Kaneko K, Itoh A, Kawashima H, Ono Y, Tainaka T, Murase N, Ando H. Endoscopic biliary drainage for children with persistent or exacerbated symptoms of choledochal cysts.
J Hepatobiliary Pancreat Sci. 2013 ;20(3):303-6. doi: 10.1007/s00534-012-0519-5. PMID: 22581057.
Fig. 1

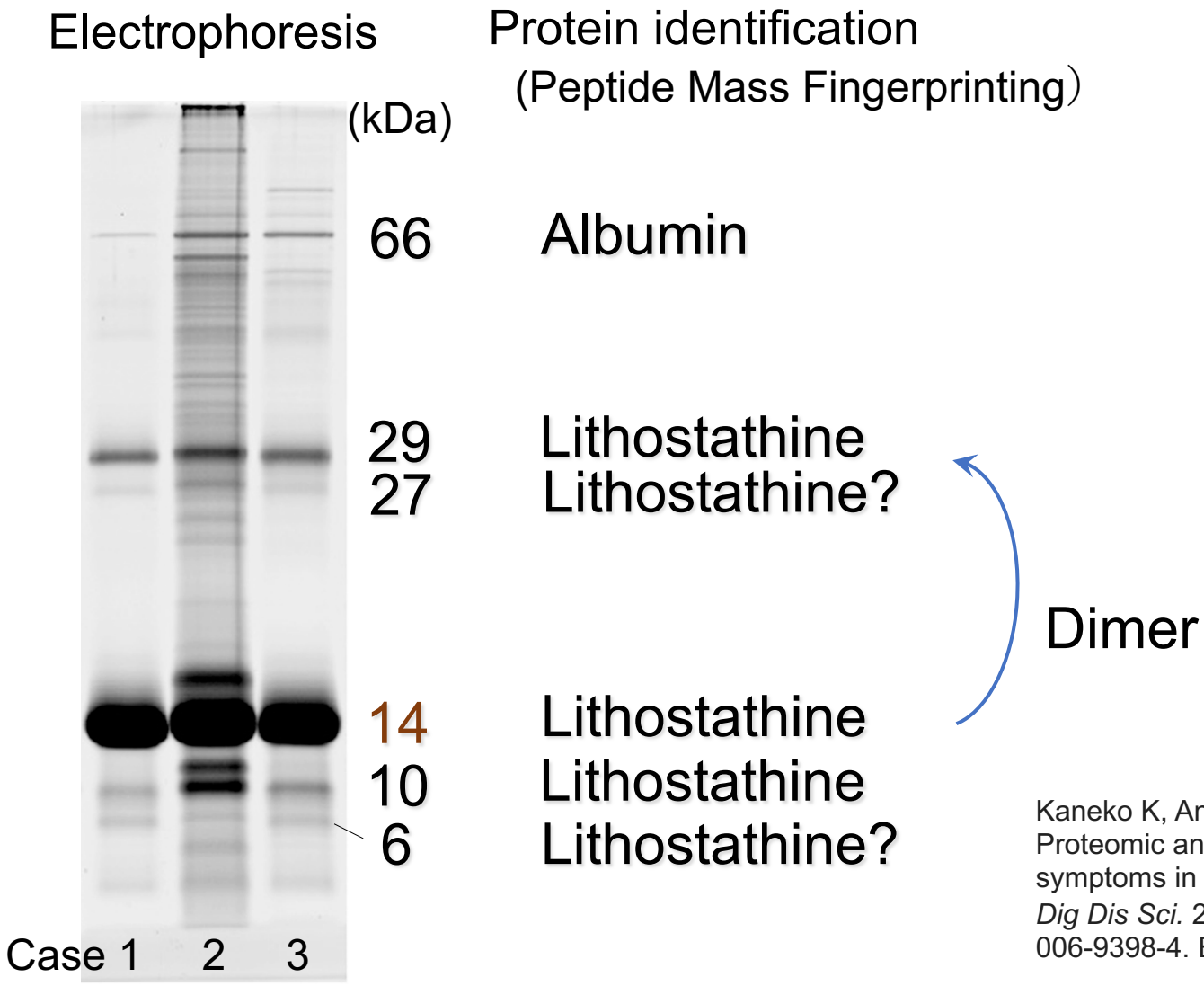
Findings supporting the protein plug theory

The common bile duct diameter is larger during the symptomatic phase



Retrospective data

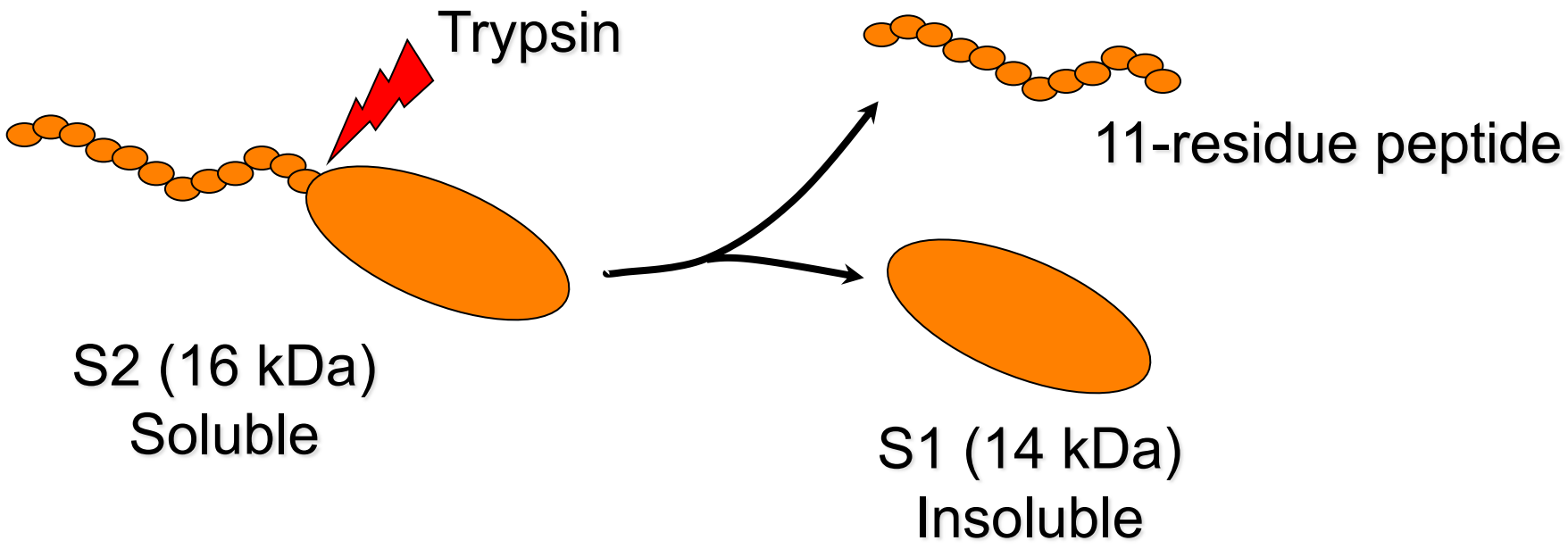
Protein plugs are composed of lithostathine



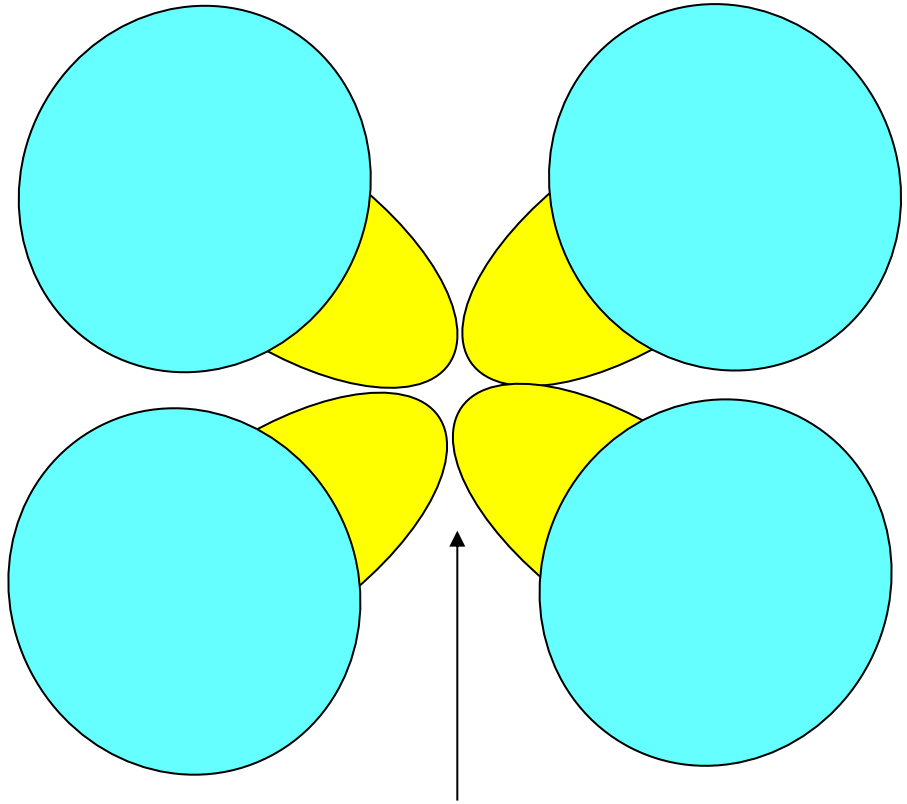
Kaneko K, Ando H, Seo T, Ono Y, Tainaka T, Sumida W. Proteomic analysis of protein plugs: causative agent of symptoms in patients with choledochal cyst. *Dig Dis Sci.* 2007;52(8):1979-86. doi: 10.1007/s10620-006-9398-4. Epub 2007 Apr 6. PMID: 17415647.

Lithostathine

- It is a soluble 144-amino acid glycoprotein secreted by pancreatic acinar cells.
- It accounts for 5%–10% of proteins in pancreatic juice, though its function remains unknown.
- It has many other names, such as pancreatic stone protein (PSP).



Lithostathine S1 forms tetramers by lateral hydrophobic interactions

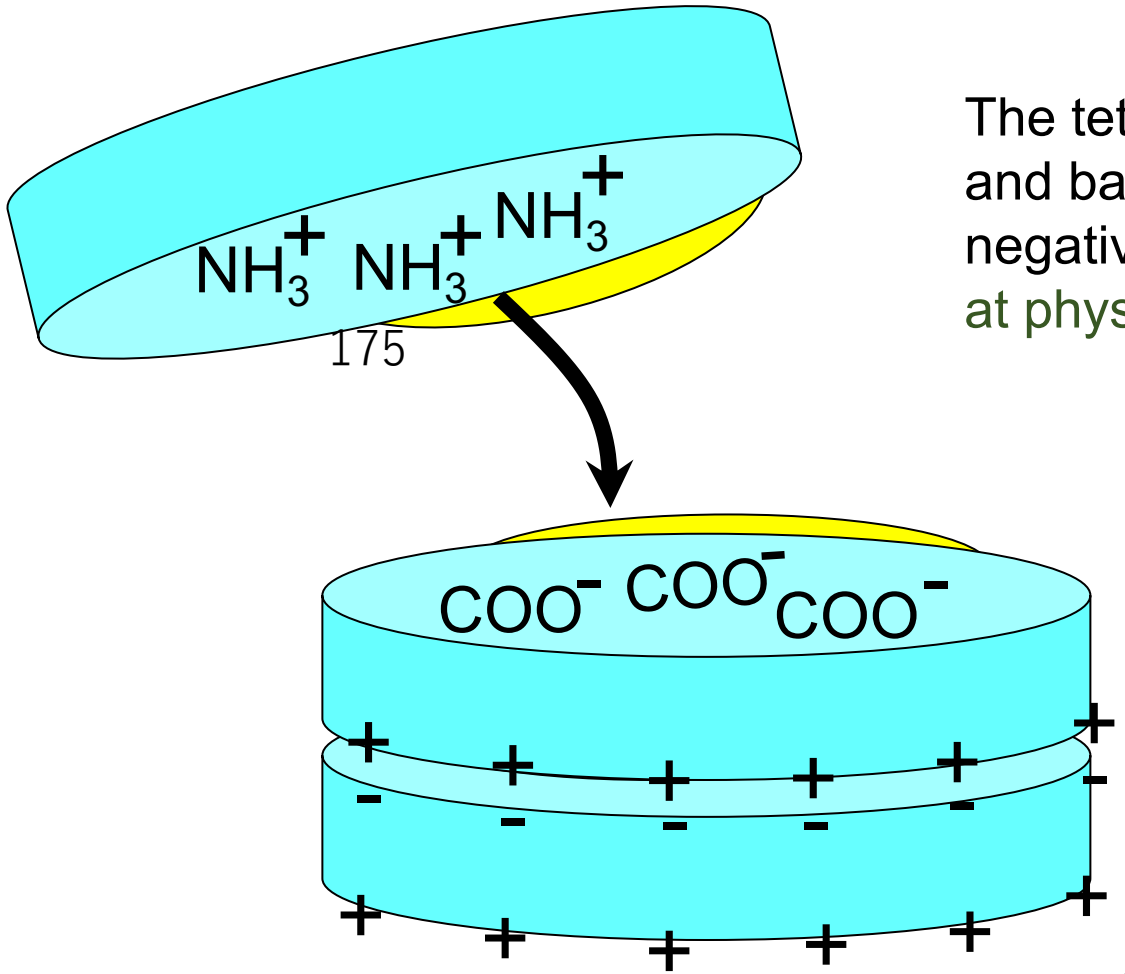


Hydrophilic

Hydrophobic

Gregoire, et al. *EMBO J.* 2001;20:3313–3321

Tetramers self-assemble by longitudinal electrostatic interactions

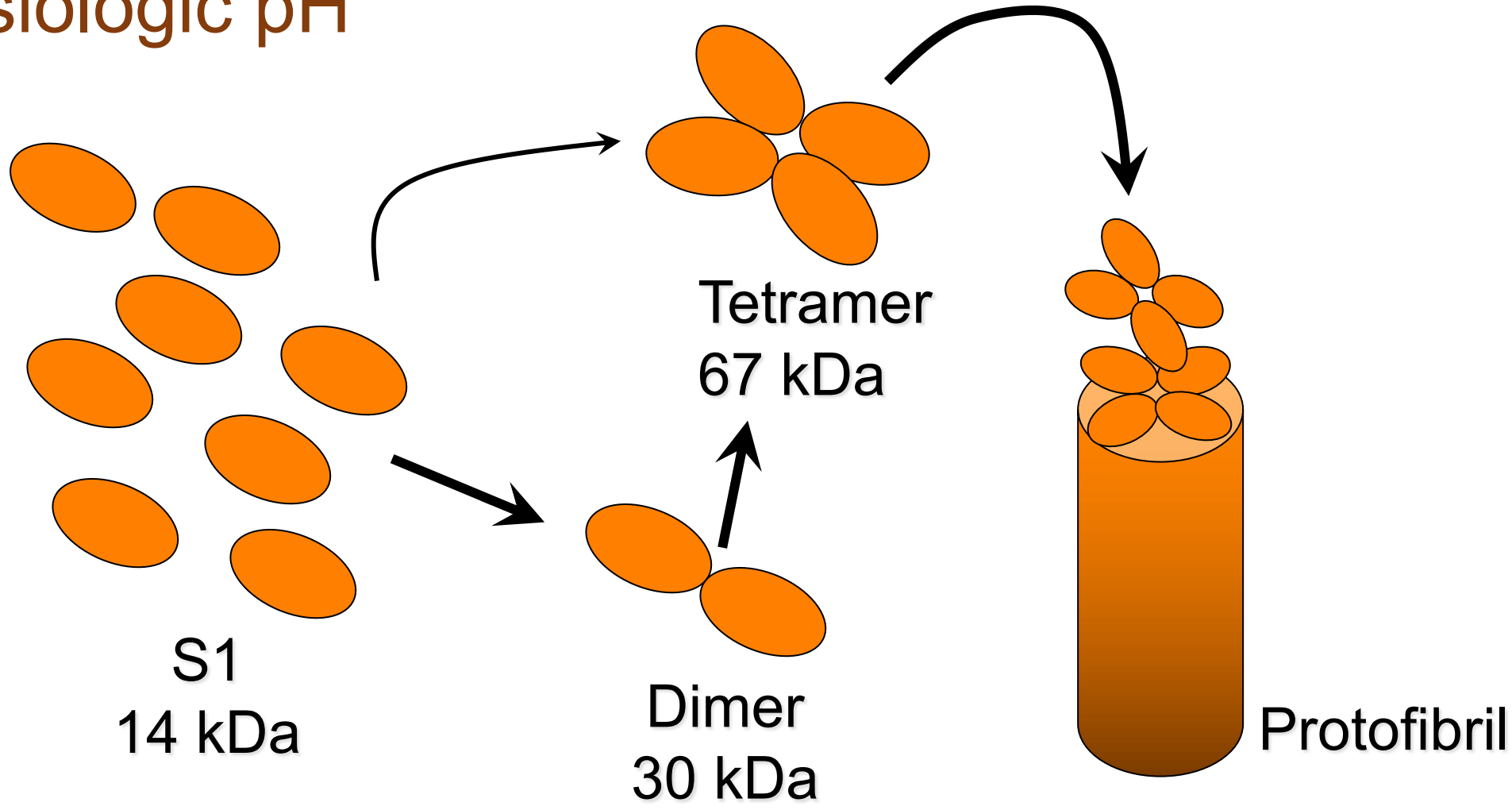


The tetramer has acidic residues on one side and basic residues on the other, contributing negative and positive charges, respectively, at physiologic pH.

Low pH	Physiologic pH	High pH
$-NH_3^+$	$-NH_3^+$	$-NH_2$
-COOH	$-COO^-$	$-COO^-$

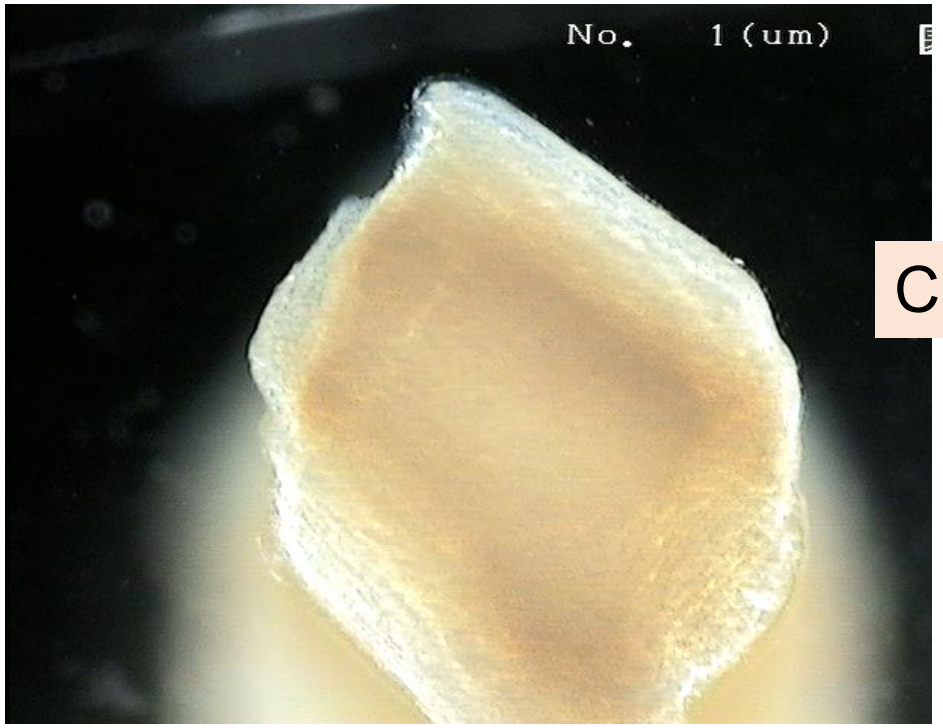
Gregoire, et al. *EMBO J* 2001;20:3313–3321

Cleaved lithostathine (S1) self-assembles into protofibrils at physiologic pH

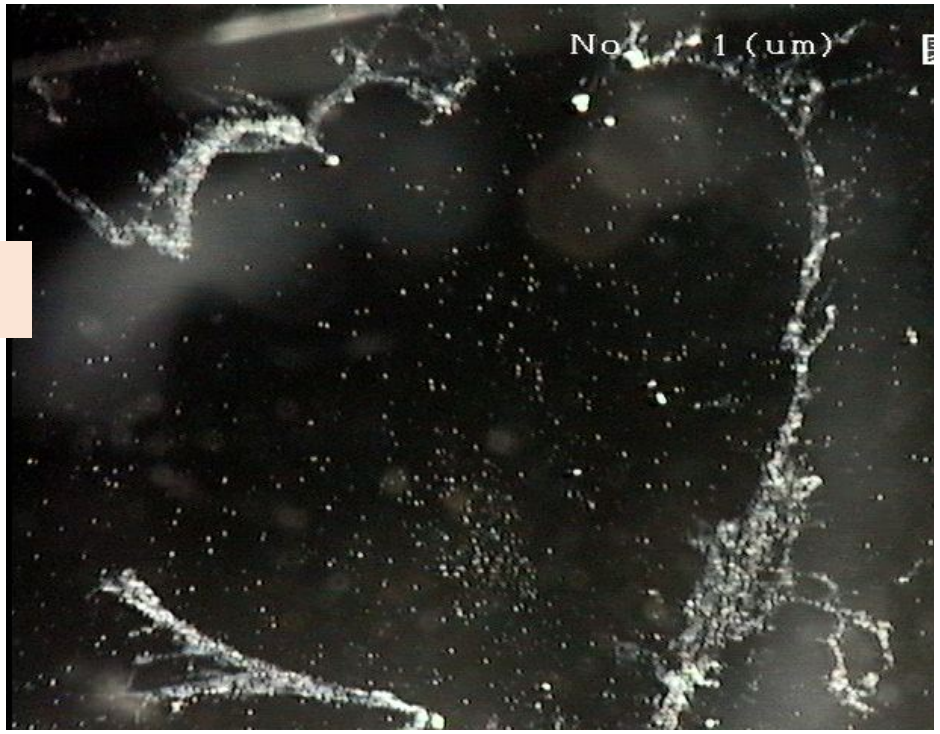


Gregoire, et al. *EMBO J* 2001;20:3313–3321

Acidic and basic solutions dissolve protein plugs



Citric acid



Proof that electrostatic interactions work only at physiologic pH

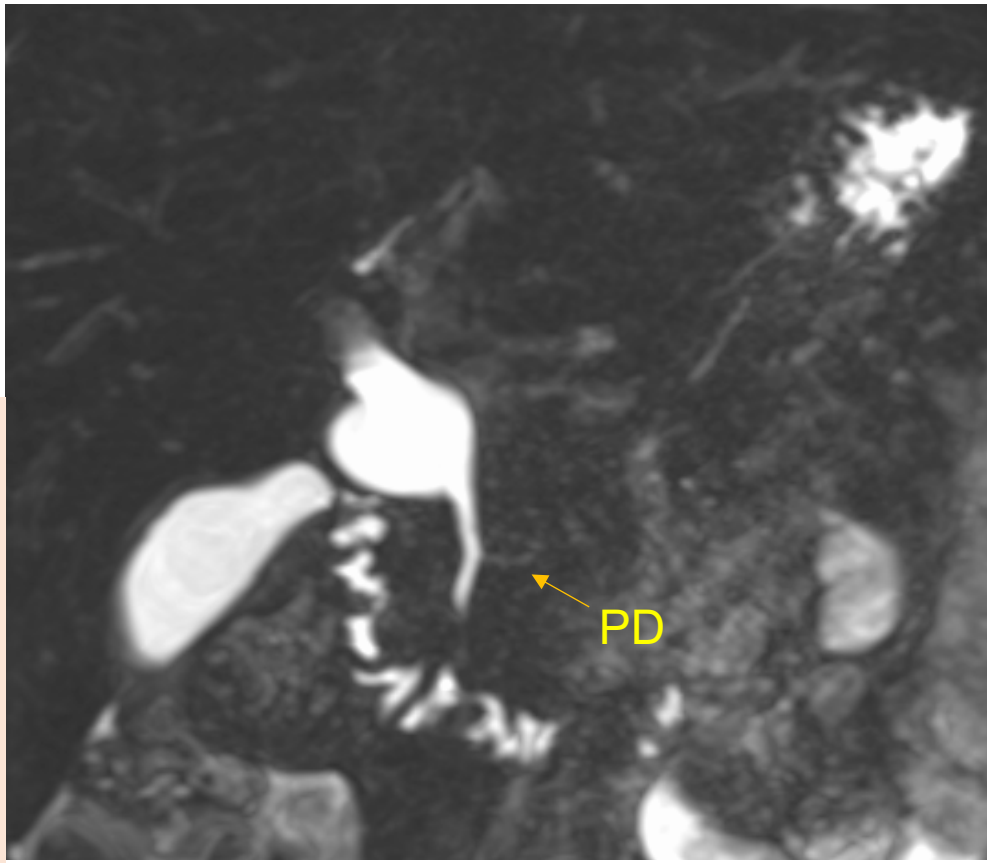
Kaneko K, Ono Y, Tainaka T, Sumida W, Ando H.
Acidic and basic solutions dissolve protein plugs made of lithostathine complicating choledochal cyst/pancreaticobiliary maljunction.
Dig Dis Sci. 2009;54(7):1475–80. doi: 10.1007/s10620-008-0503-8. Epub 2008 Nov 7. PMID: 18989781.

Where do protein plugs form?

Case 4 from *Basic Section* (see p28)

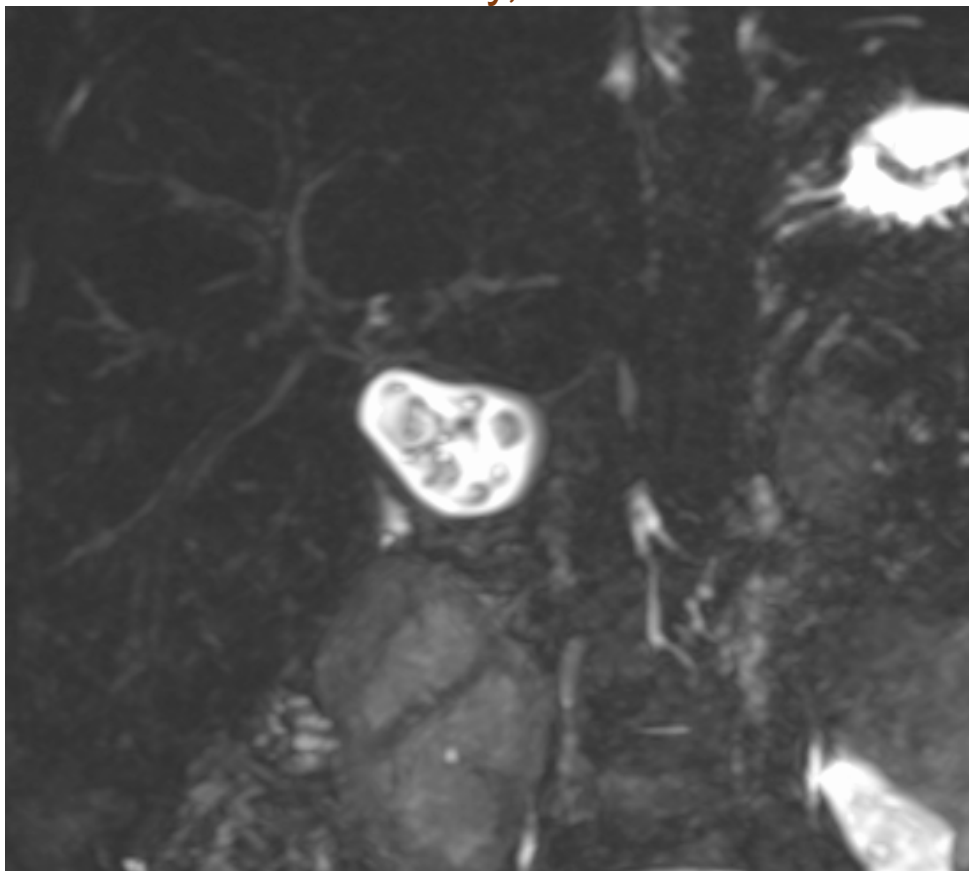
- A pediatric case with abdominal pain and vomiting.
- Elevated serum liver and biliary enzymes.
- No elevation of serum pancreatic enzymes.

Protein plugs are often found in the dilated common channel. However, plugs are formed as well in cases without common channel dilatation, as demonstrated here. Most plugs are presumed to form in the dilated BD or the gallbladder, where bile stagnates.



No dilatation of the common channel

Probably, here



Multiple filling defects on the dorsal side of the choledochus



Advanced

Pathophysiology in pediatric patients

Symptomatology besides protein plugs

First version, September 2024

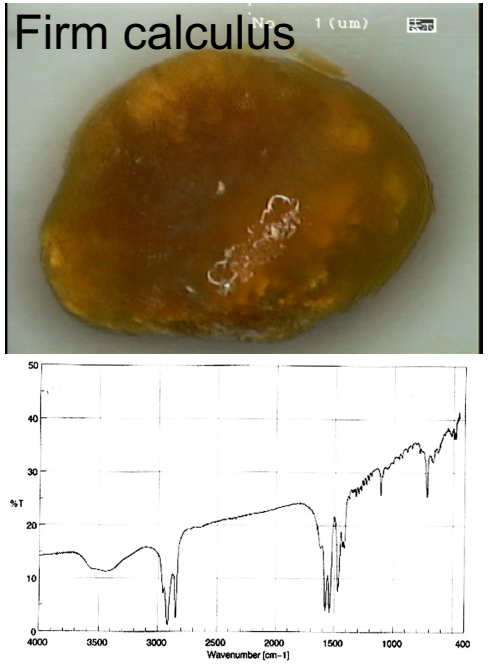
Symptomatology besides protein plugs

The protein plug theory explains symptoms in most pediatric cases. However, there are a few exceptions.

- Fatty acid calcium stones causing occlusion, but extremely rare
- Early onset in neonates and early infants

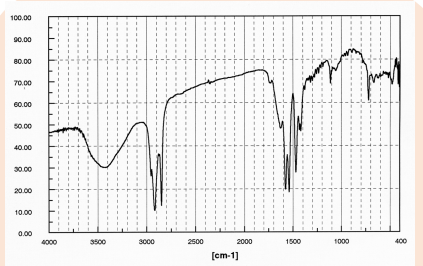


Uniformly shaped filling defects, a feature of gallstones different from nonuniform shape of protein plugs (compare with p178)

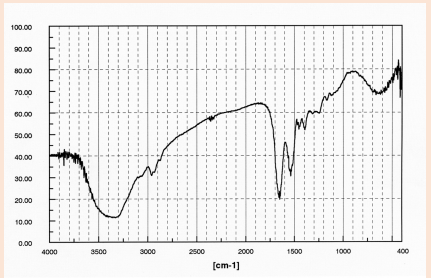


Infrared absorption spectrometry identifying fatty acid calcium

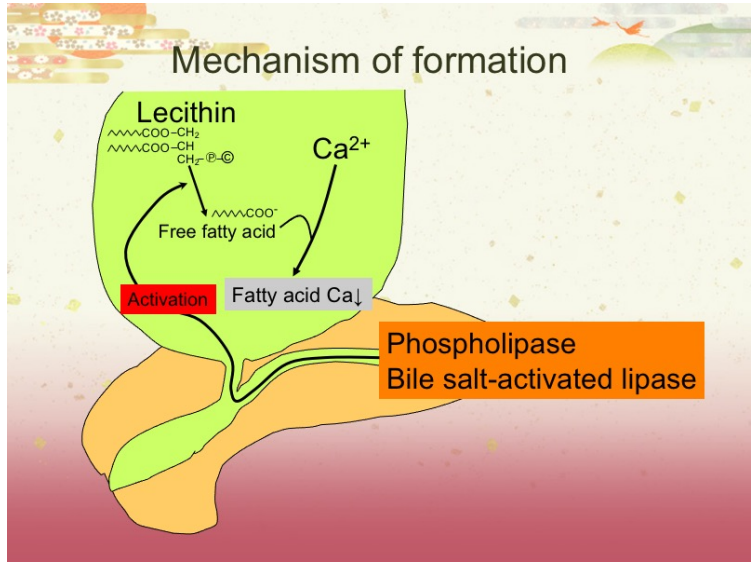
cf. Spectra of standard materials



Fatty Acid Calcium

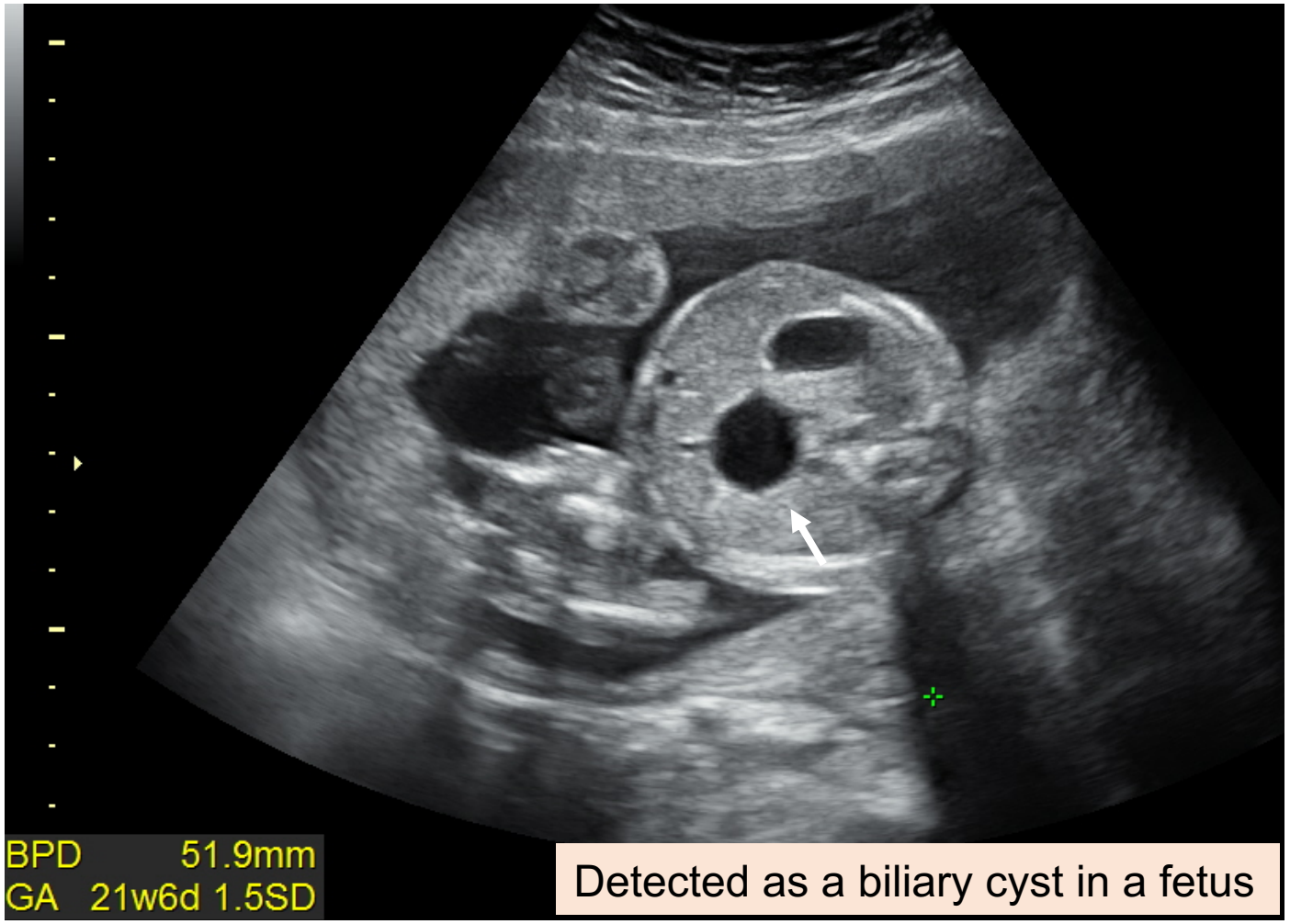


Protein

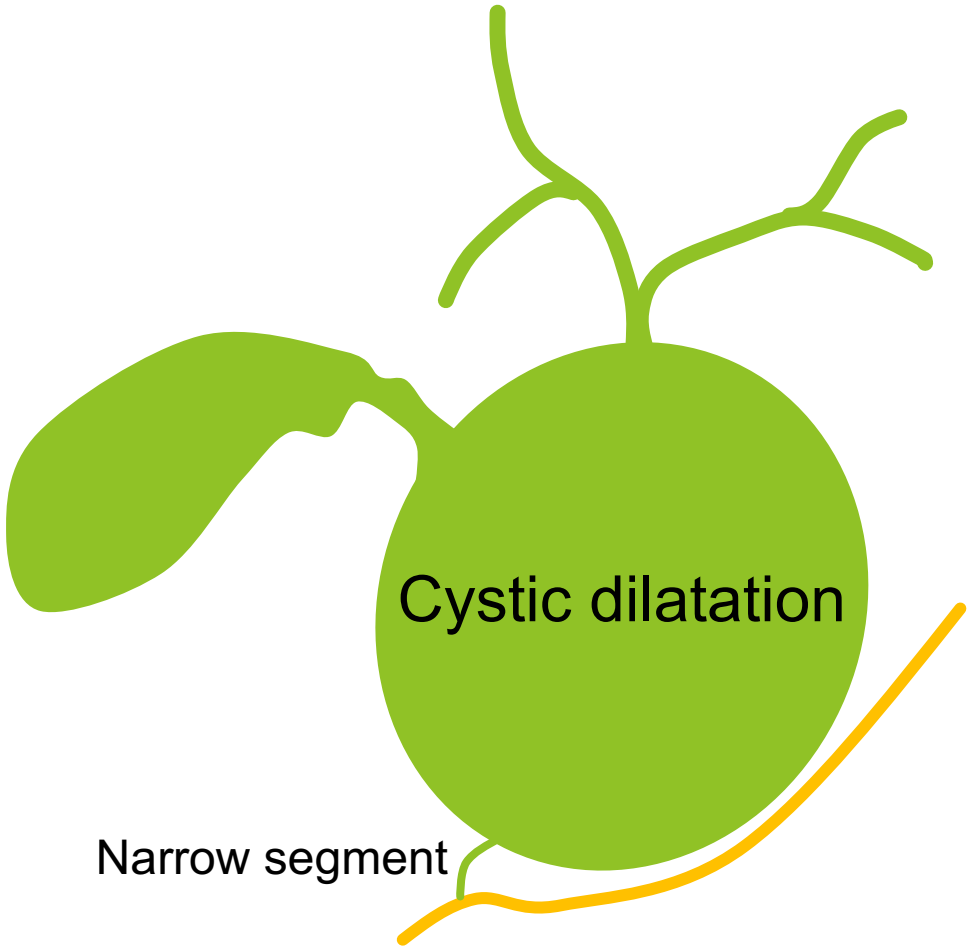


Kaneko K, Ono Y, Tainaka T, Sumida W, Ando H.
Fatty acid calcium stones in patients with pancreaticobiliary maljunction/choledochal cyst as another cause of obstructive symptoms besides protein plugs.
J Pediatr Surg. 2008 Mar;43(3):564-7. doi: 10.1016/j.jpedsurg.2007.11.004.
PMID: 18358304.

Onset in neonates and early infants



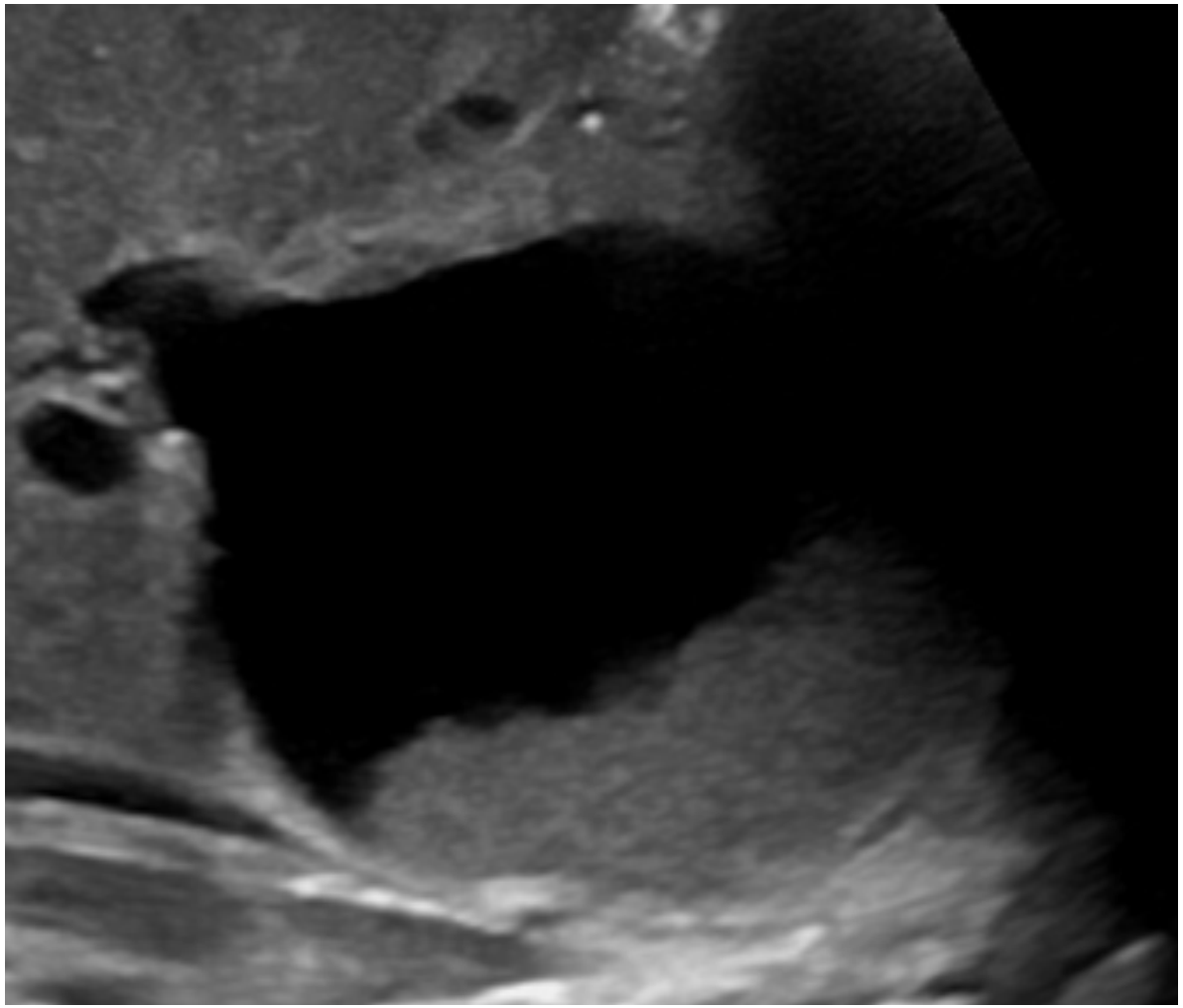
Detected as a biliary cyst in a fetus

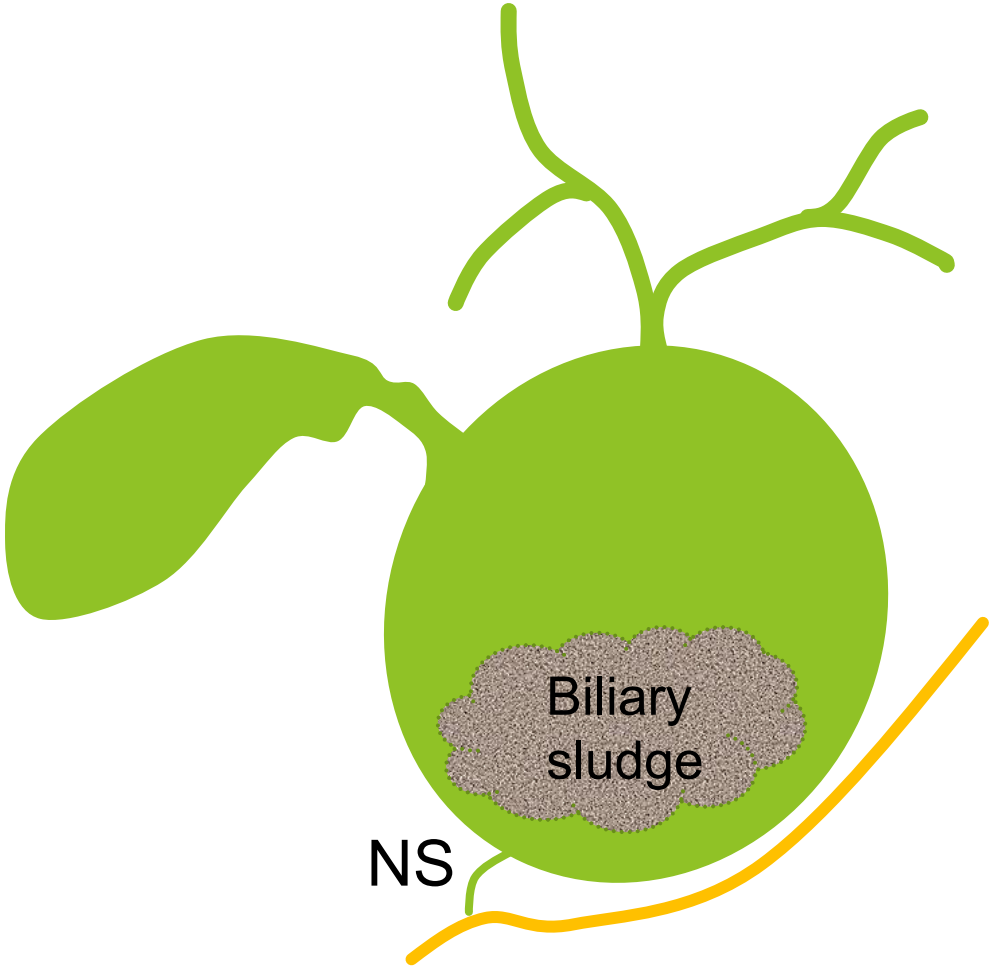


Almost all cases are of the cystic type with an NS

These biliary cysts are indistinguishable from cystic biliary atresia (type I cyst and IIId) even after birth (see *Expert Section*).

Debris appears in the cyst before symptoms occur

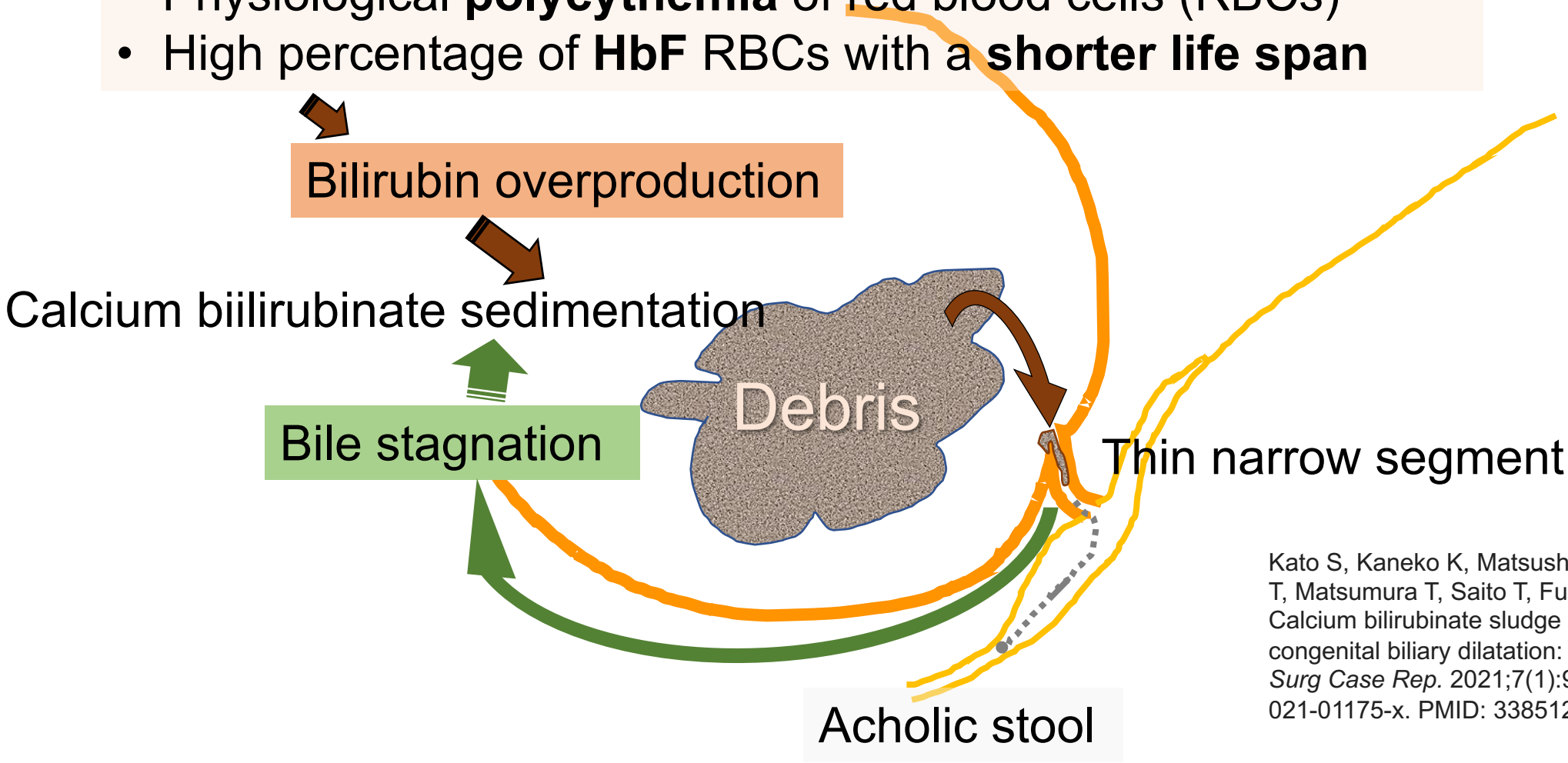




Infrared absorption spectrometry shows that the sludge is composed of calcium bilirubinate.

Proposed mechanism of early onset

Accelerated **hemolysis** due to neonatal hematologic features
- Physiological **polycythemia** of red blood cells (RBCs)
- High percentage of **HbF** RBCs with a **shorter life span**



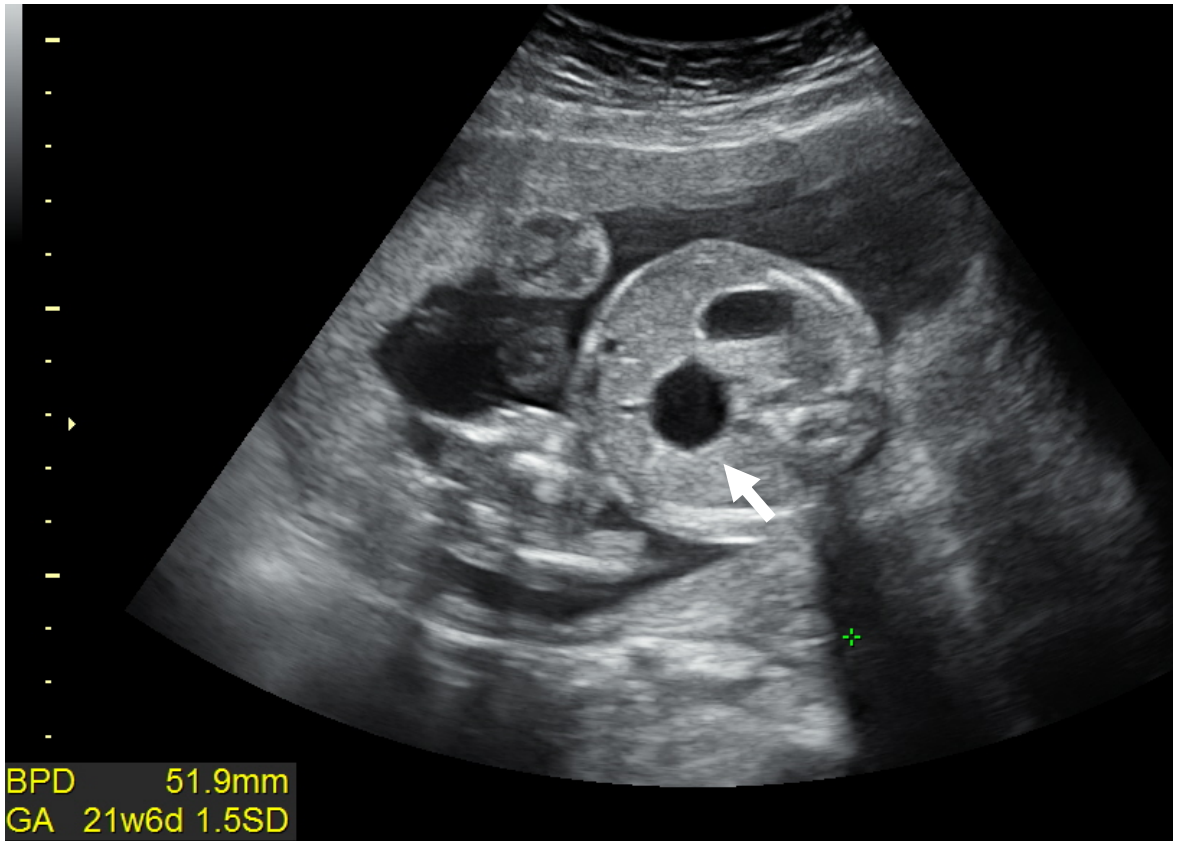
Kato S, Kaneko K, Matsushita N, Kurahashi S, Osawa T, Matsumura T, Saito T, Fukami Y, Komatsu S, Sano T. Calcium bilirubinate sludge causes early onset of congenital biliary dilatation: a report of two cases. *Surg Case Rep.* 2021;7(1):92. doi: 10.1186/s40792-021-01175-x. PMID: 33851282; PMCID: PMC8044275.

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



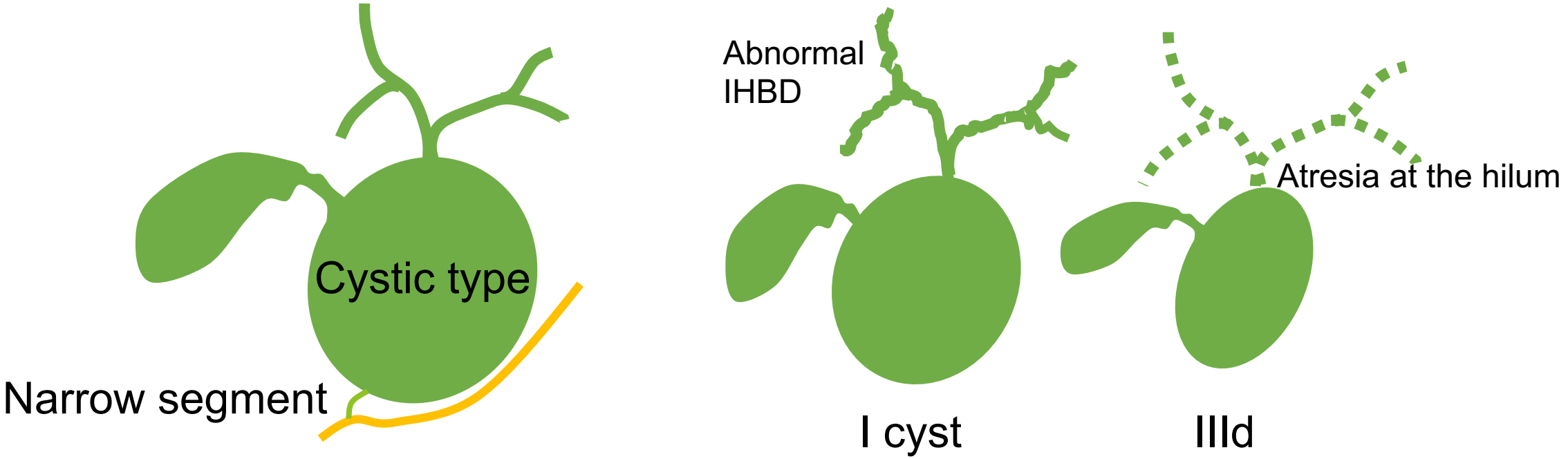
Differential diagnosis is difficult even after birth

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

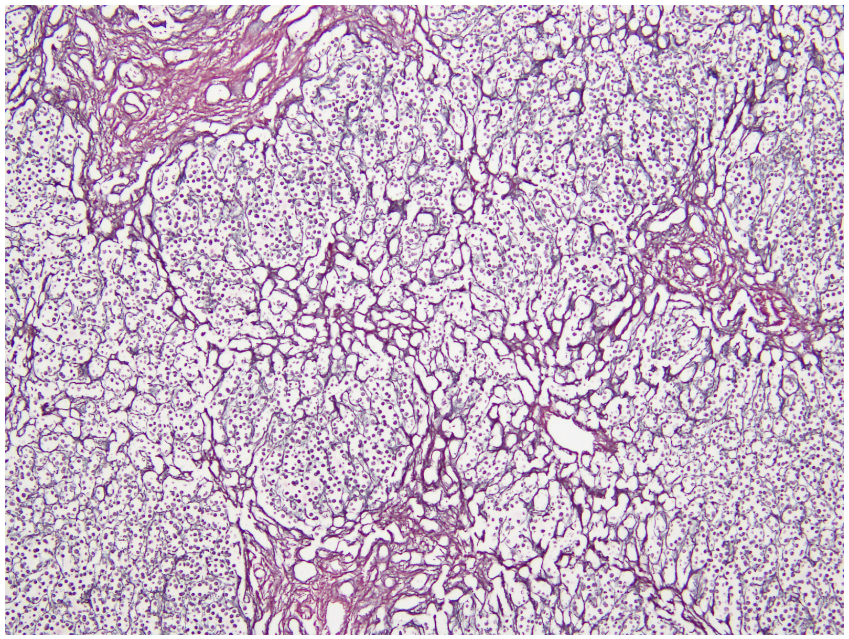
Biliary obstruction causes cholangiopathy specific to neonates and early infants.

Prenatally-diagnosed CBD and hepatic fibrosis

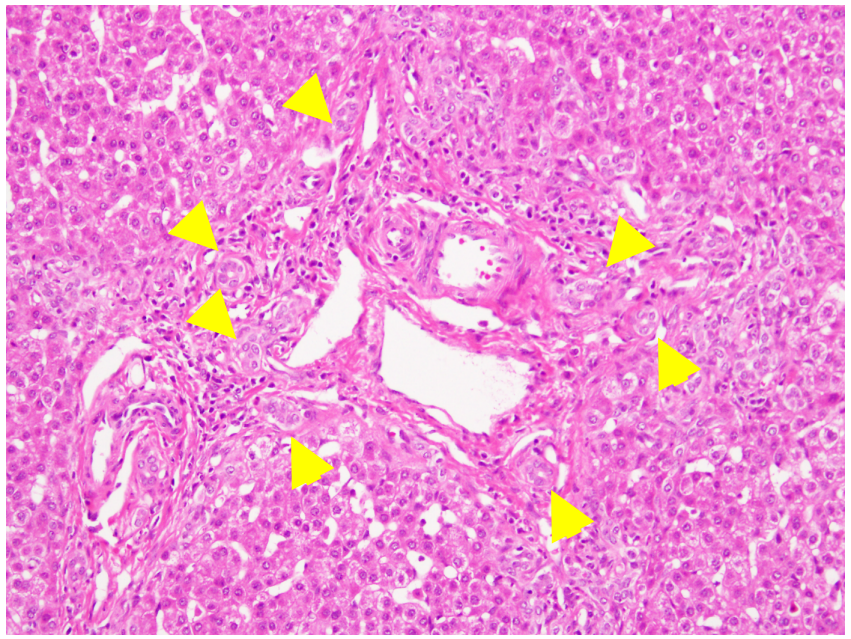
- These cases are prone to hepatic fibrosis.
- Fibrosis can occur even in asymptomatic cases.
- The fibrosis improves after bile duct resection, suggesting that the fibrosis is reversible.

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.

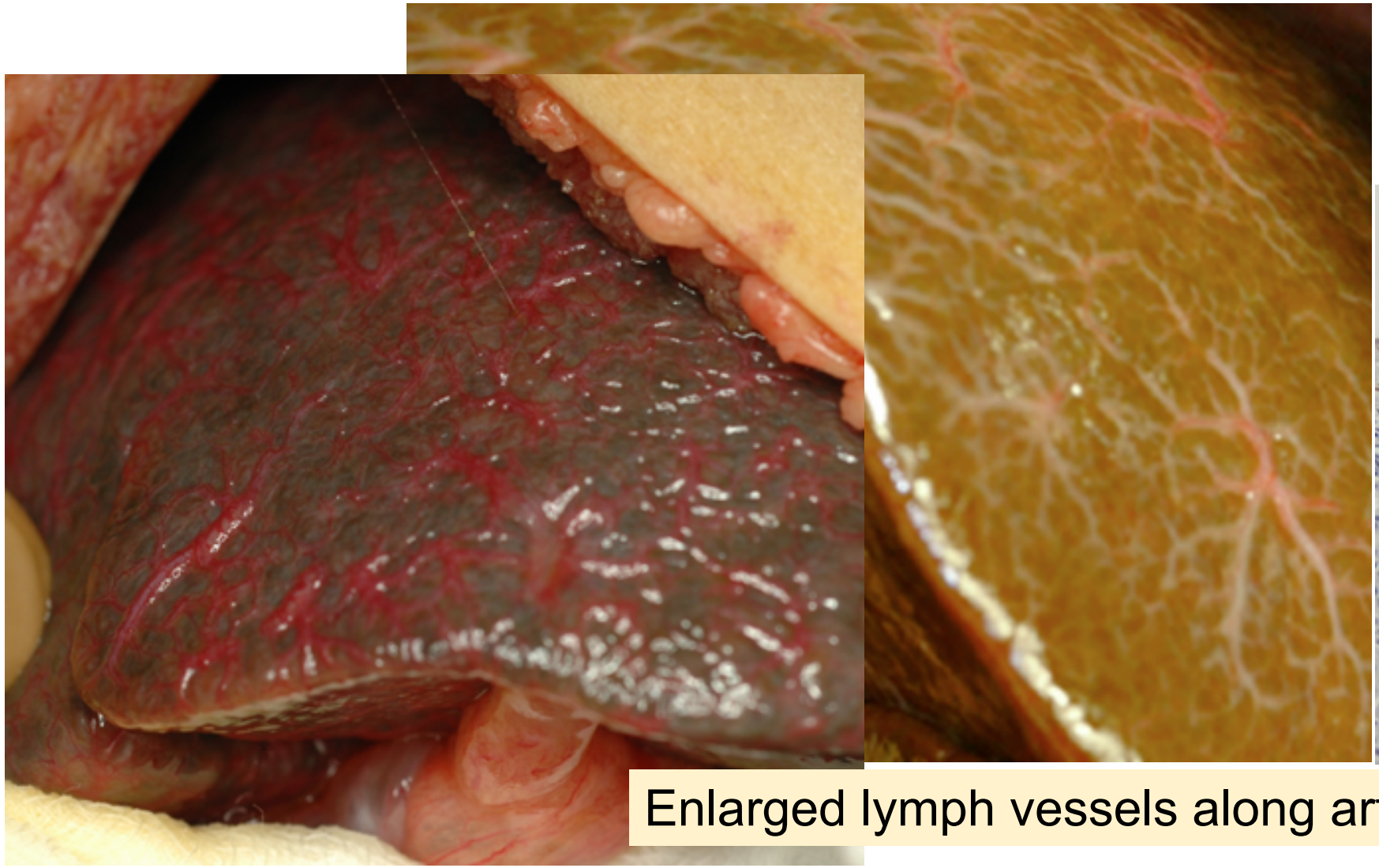


Bridging F1-2



Cholangiocyte proliferation

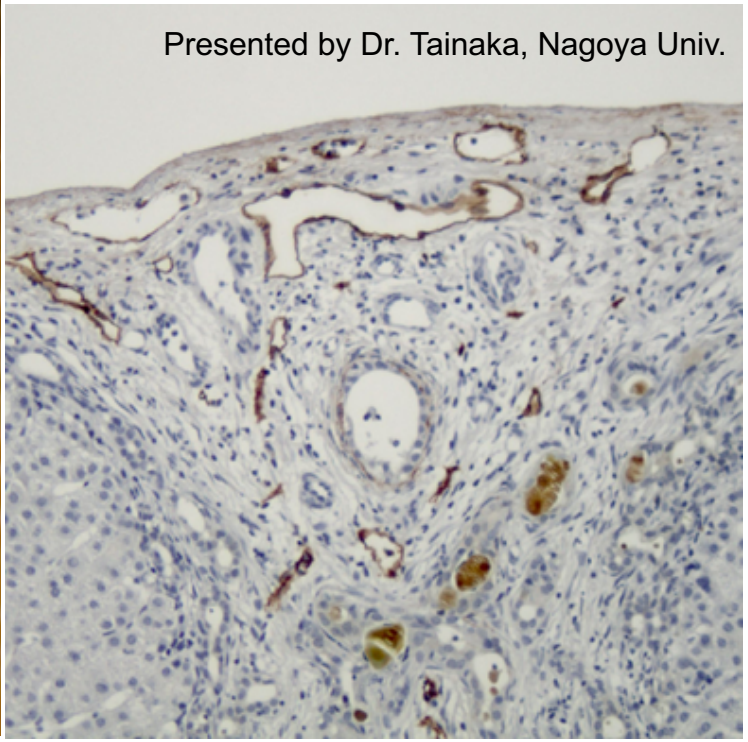
Makibishi sign specific to biliary atresia





Makibishi, ninjas' tool

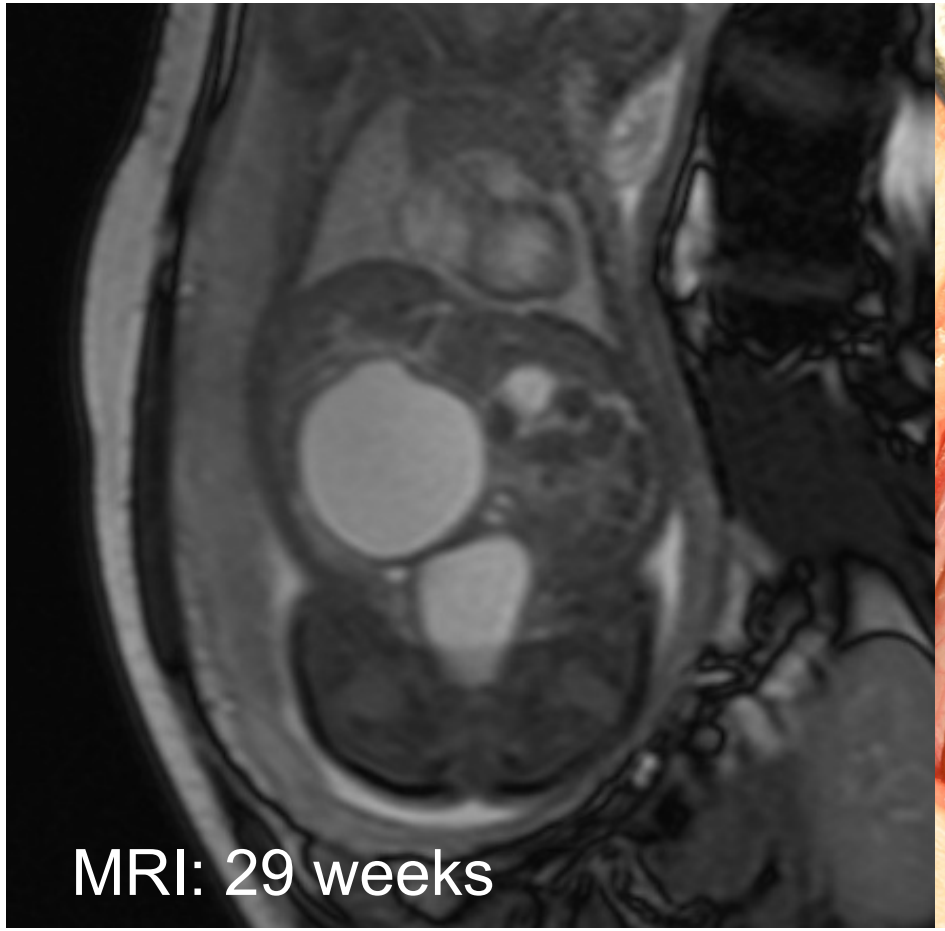
Presented by Dr. Tainaka, Nagoya Univ.



Enlarged lymph vessels along arteries on the liver surface

Case 48

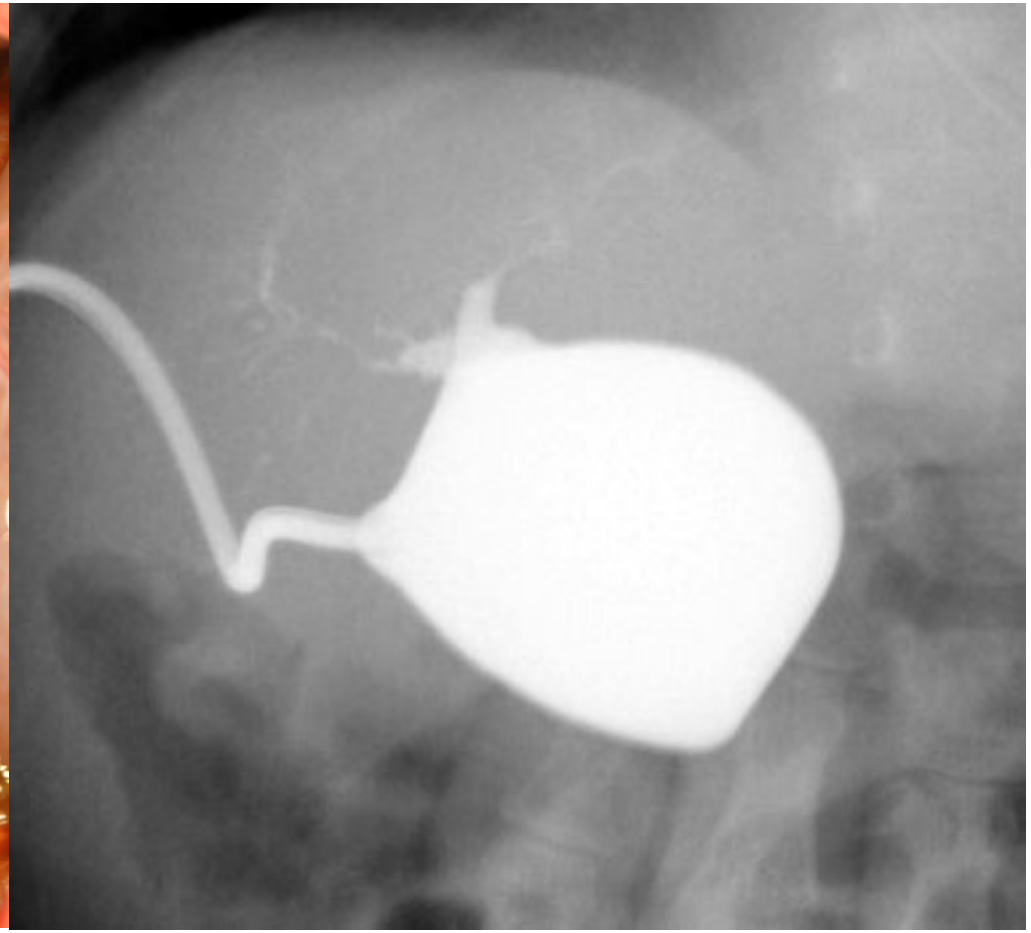
Type I cyst biliary atresia



Makibishi sign



Treelike intrahepatic BDs

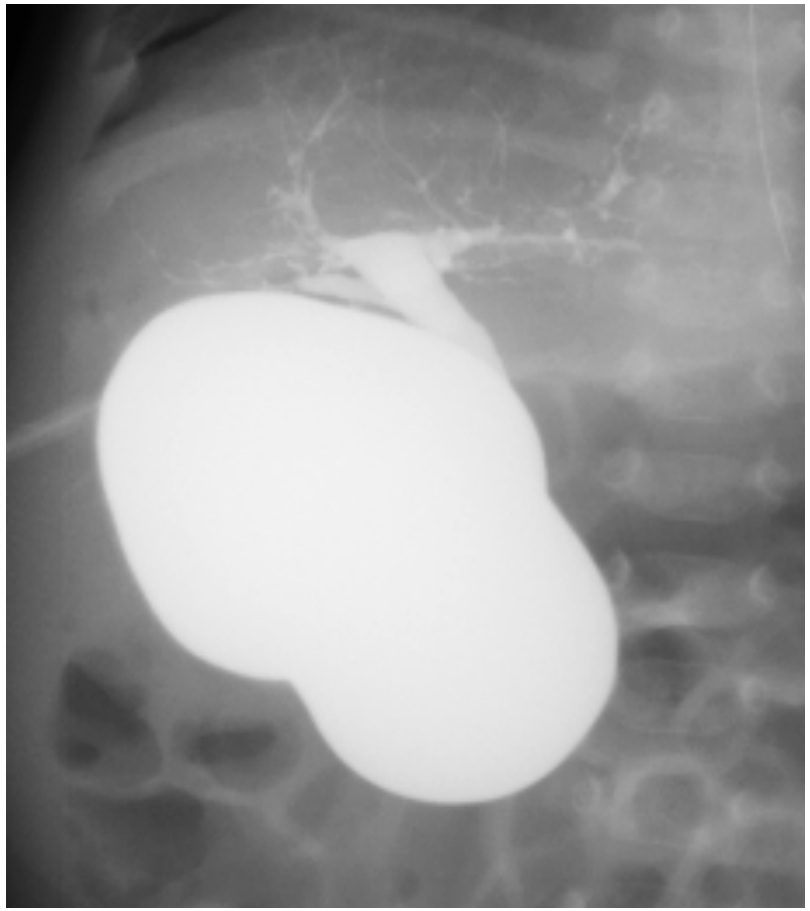


Contrast material did not go out into the duodenum.

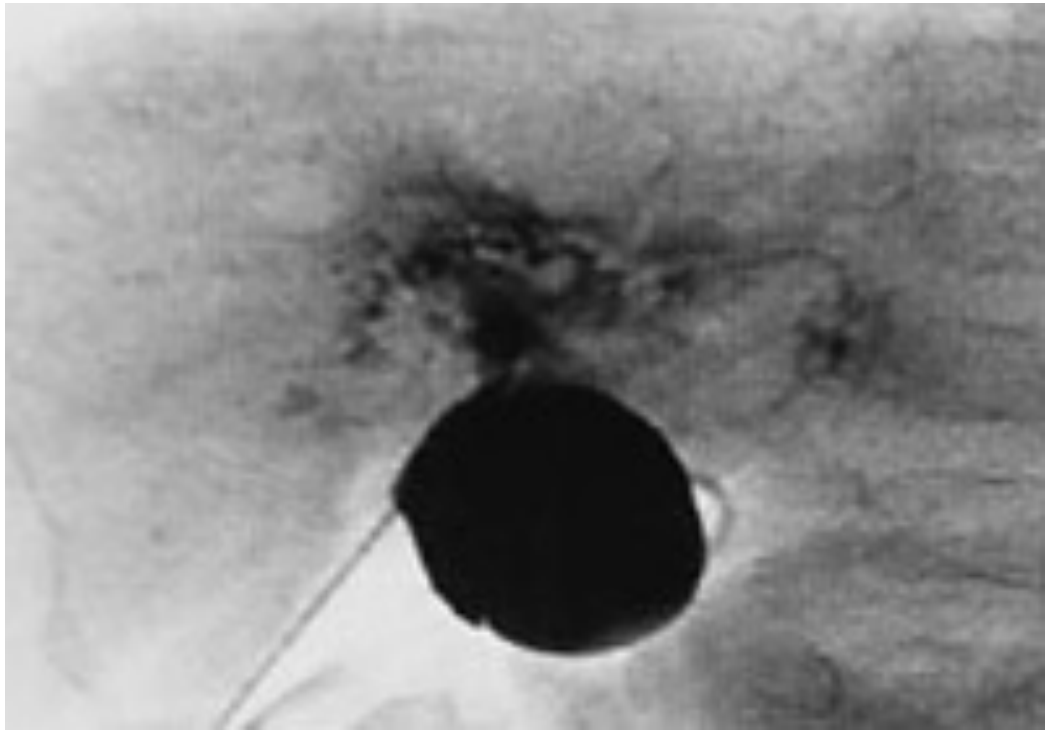
Type I cyst biliary atresia

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

Treelike type



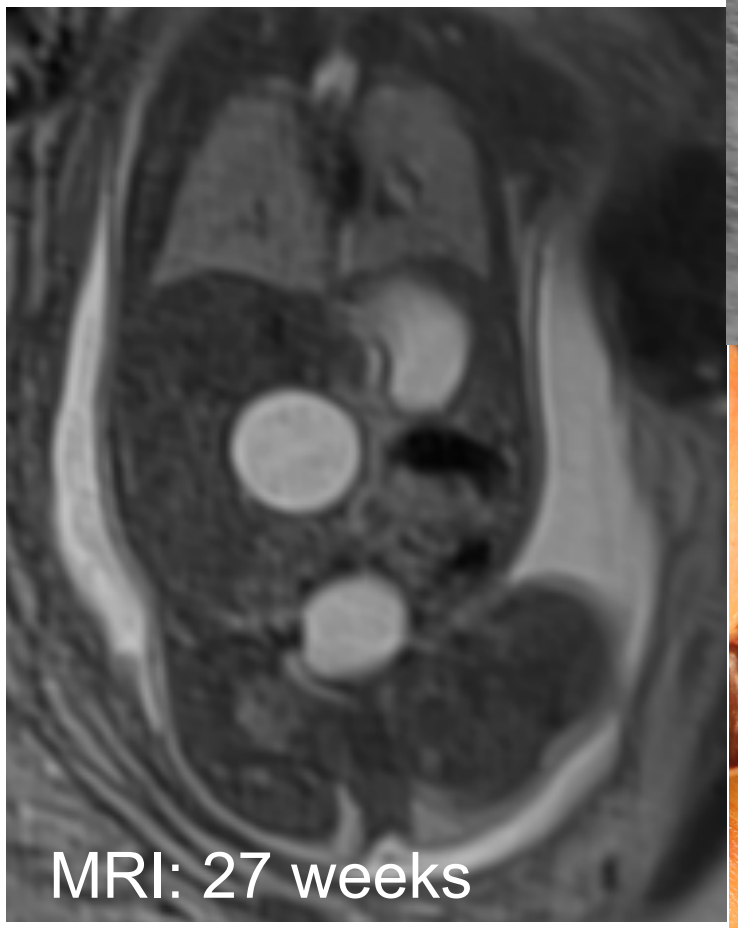
Cloudy type

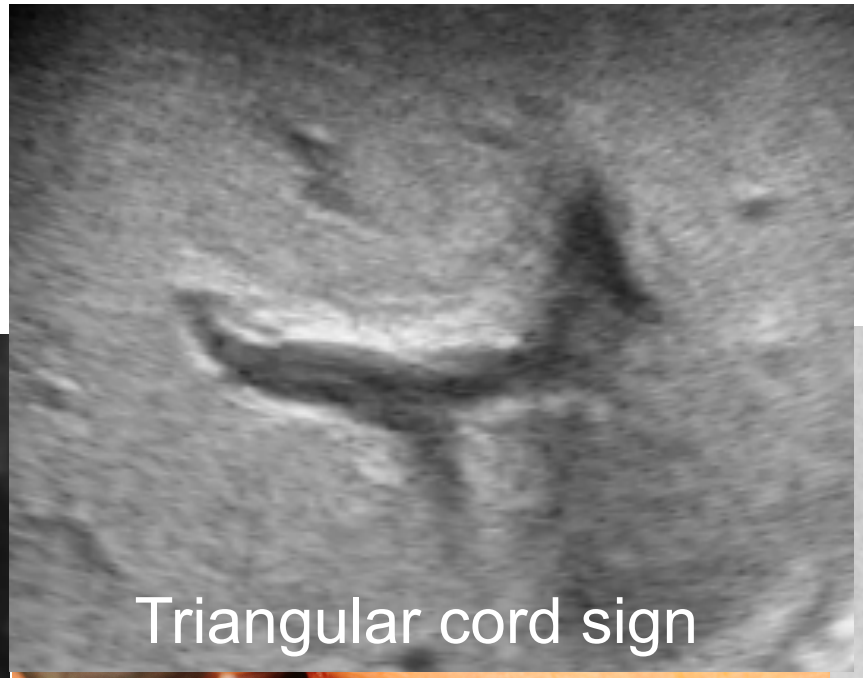


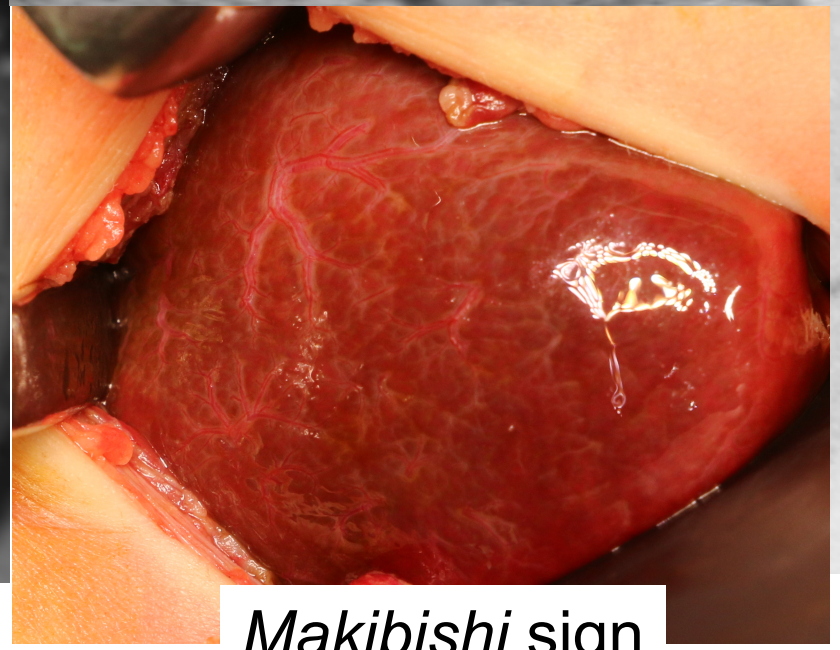
Nio M, et al. Long-term outcome in type I biliary atresia. *J Pediatr Surg.* 2006;41:1973–5.

Case 49

Type IIId BA



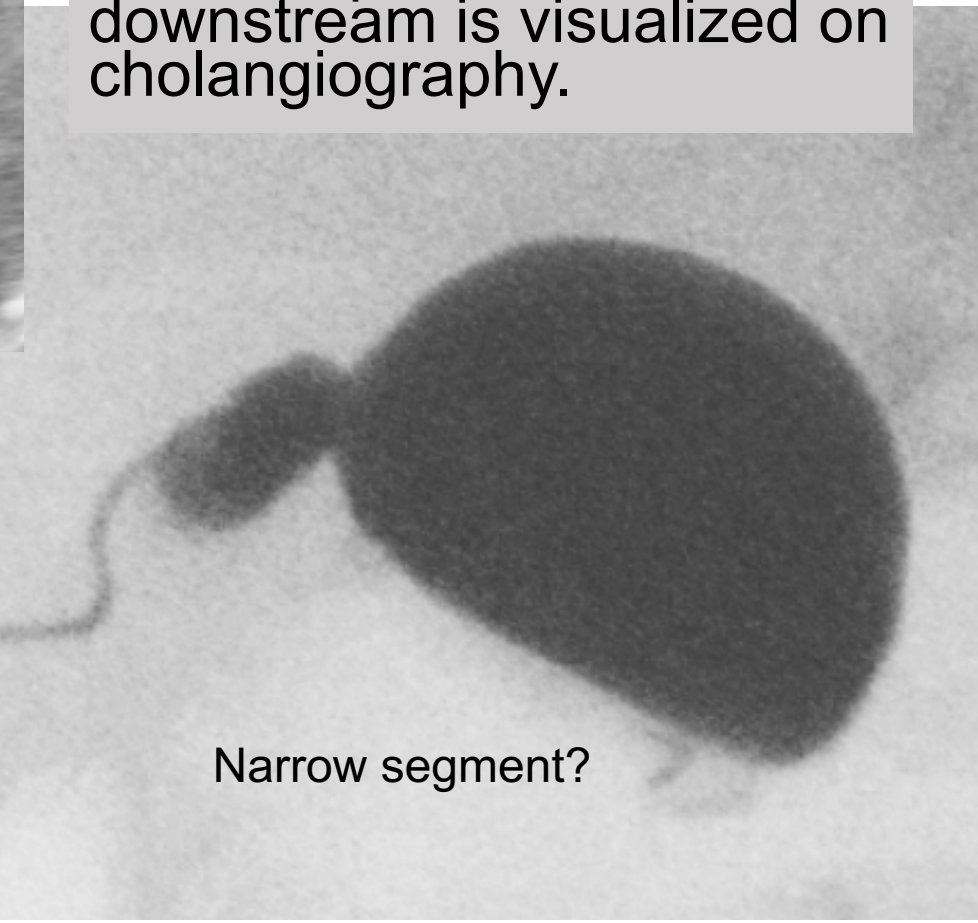




Makibishi sign

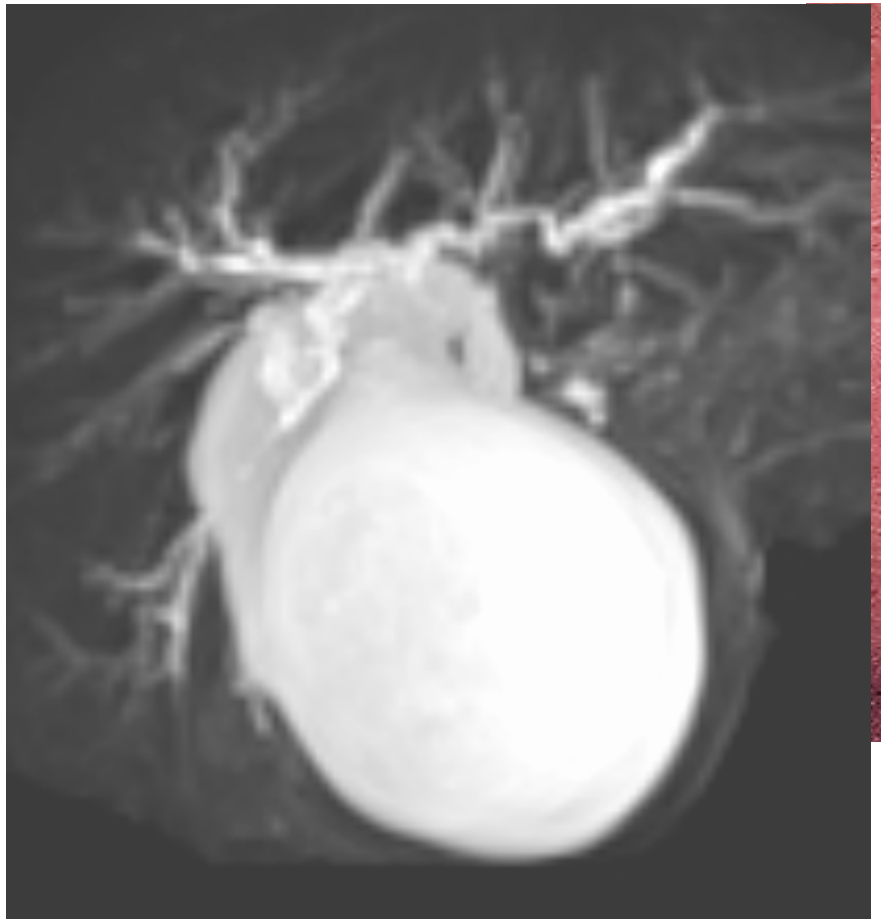
Atresia at the hepatic hilum requires the Kasai operation.

Neither upstream nor downstream is visualized on cholangiography.



Case 50

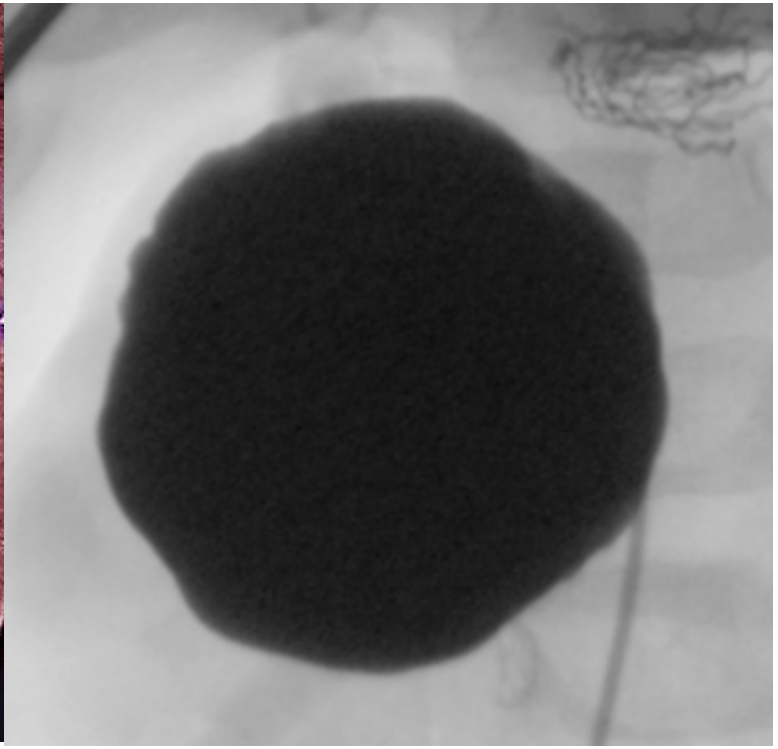
Type Ia CBD > Type I cyst BA



Almost normal IHBD



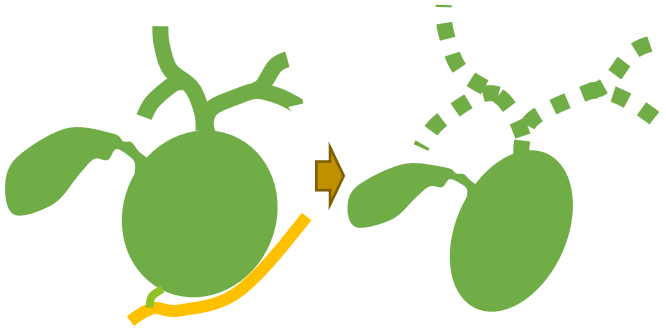
Makibishi sign



Contrast material did not enter the duodenum

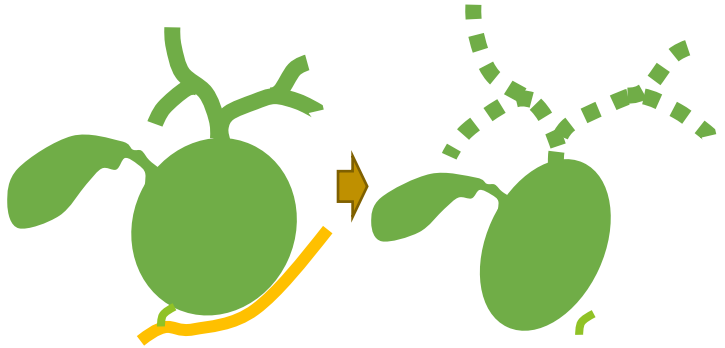
CBD featuring elements of BA
Intermediate/Transitional type
Bile duct resection yields good result.

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



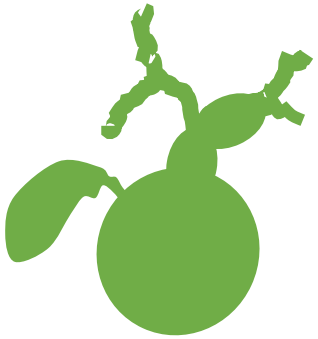
CBD → IIId BA

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



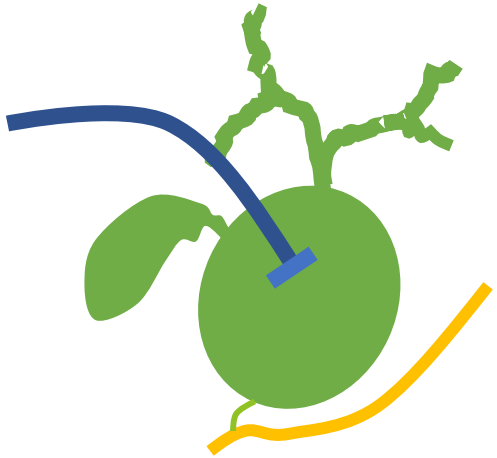
CBD → IIId BA (+NS)

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



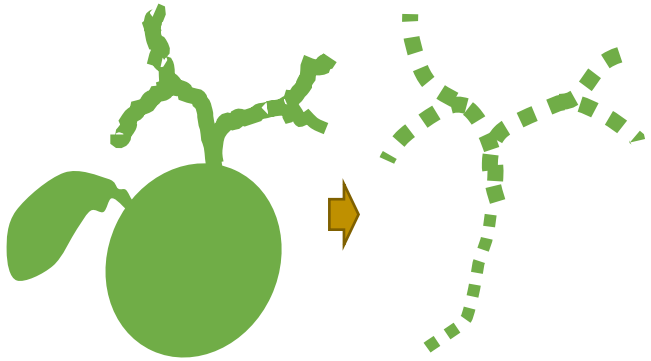
I cyst BA with a dilated LHD
Rapid progression of liver cirrhosis resulting in liver transplantation.
(The authors described a type IV-A CBD).

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

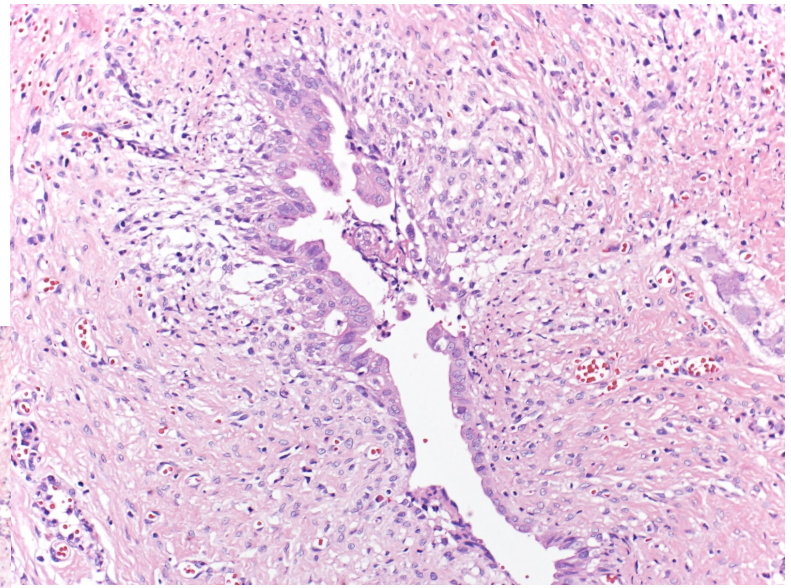


I cyst → III or IIId

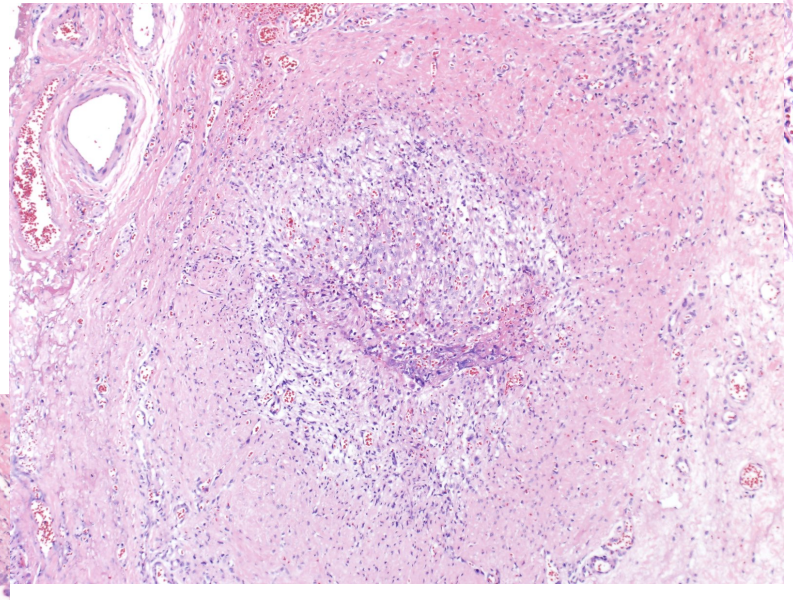
Nakata, et al.
J Jpn Pediatr Surg. 2000;36:332
Fujishiro J, et al.
Pediatr Sur Int. 2005;21:116
Kawasaki, et al.
J Jpn Pediatr Surg. 2009;45:729
Masumoto K, et al.
Pediatr Sur Int. 2011;27:99
Odashima, et al.
J Jpn Pediatr Surg. 2016;52:1042

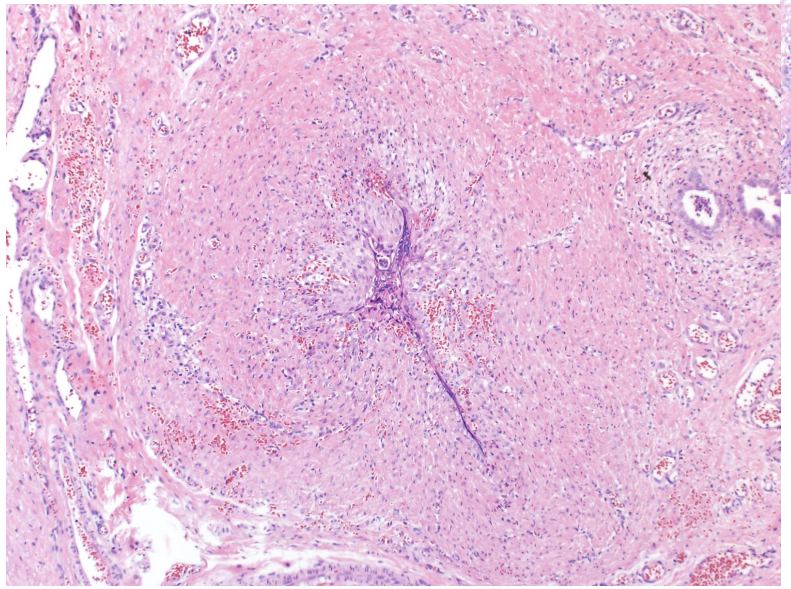
Pathology of hilar remnant in type IIId BA

Obstruction progresses from downstream to upstream?

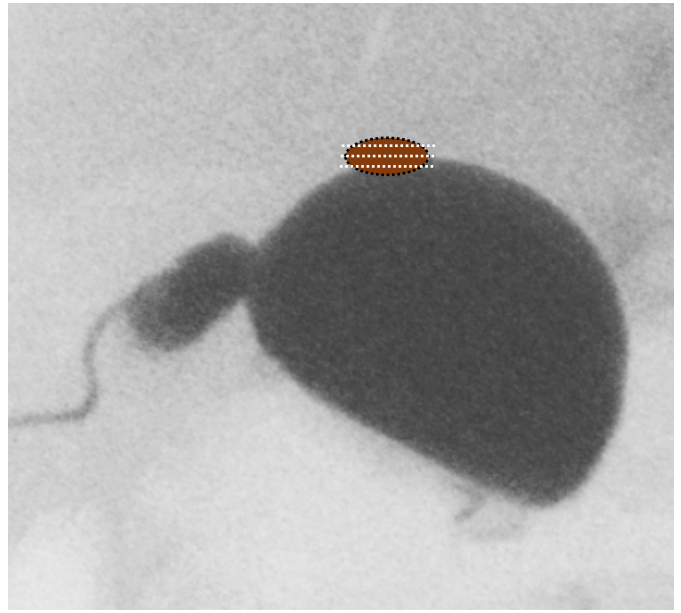


Upstream





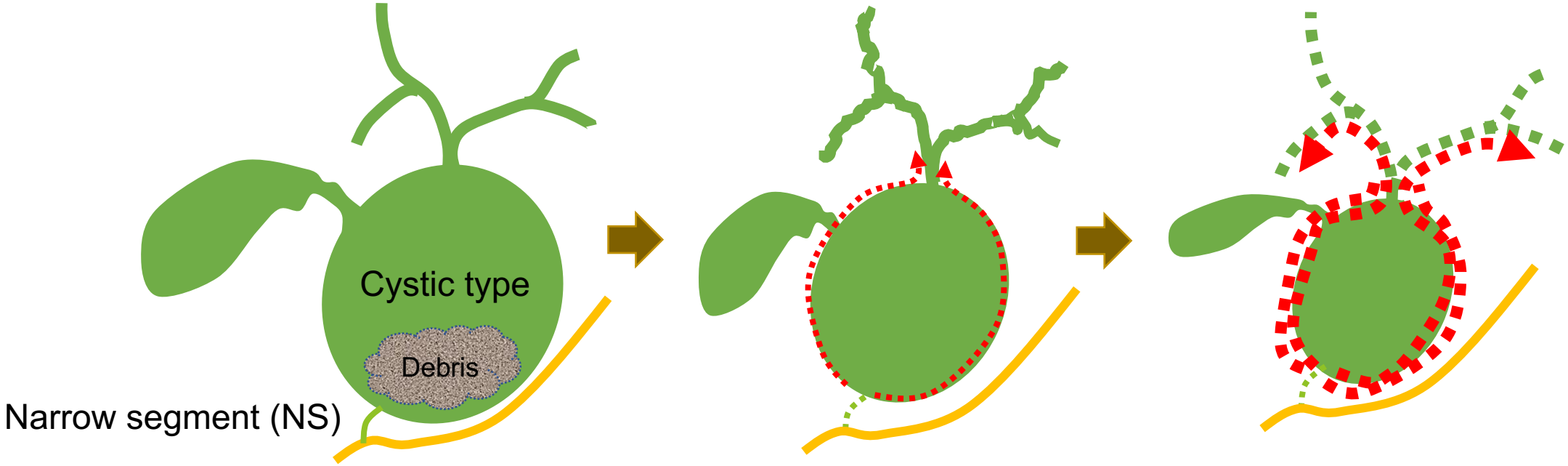
Downstream



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.

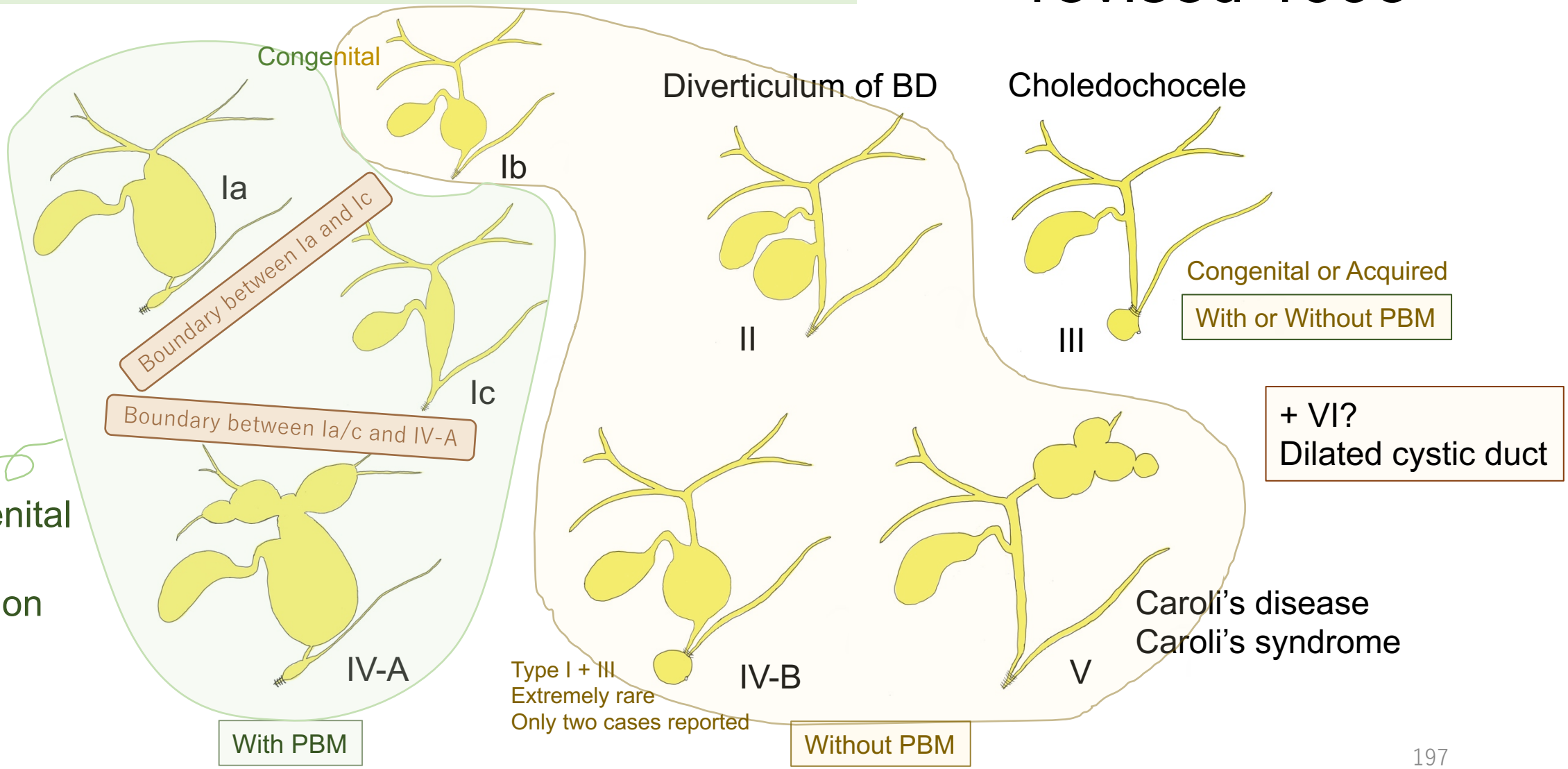
Expert

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



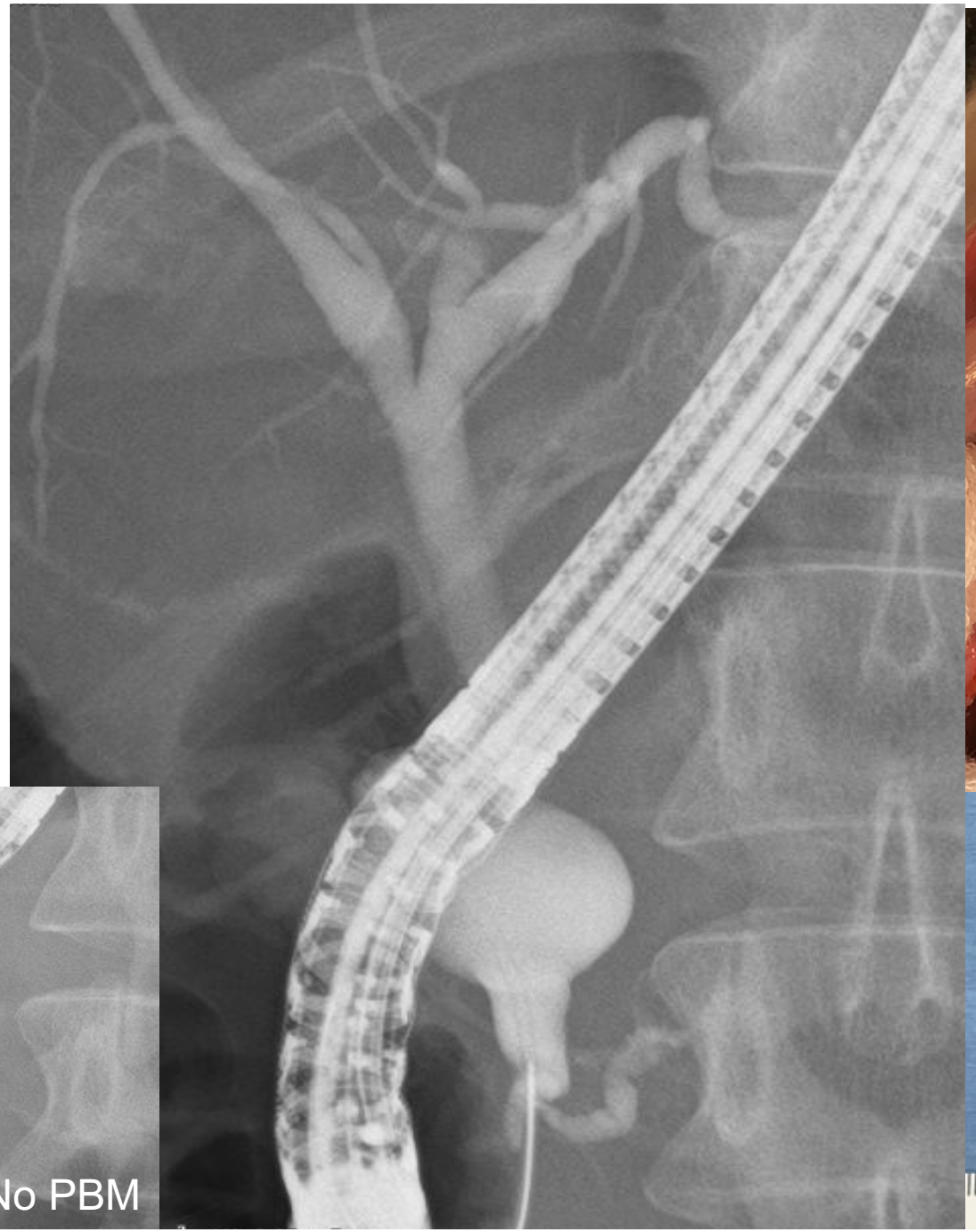
Case 51

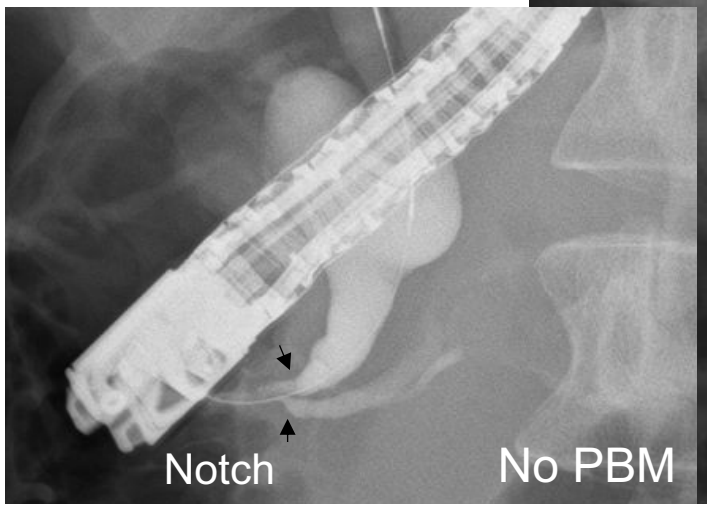
Type Ib:
segmentally dilated common bile duct without PBM

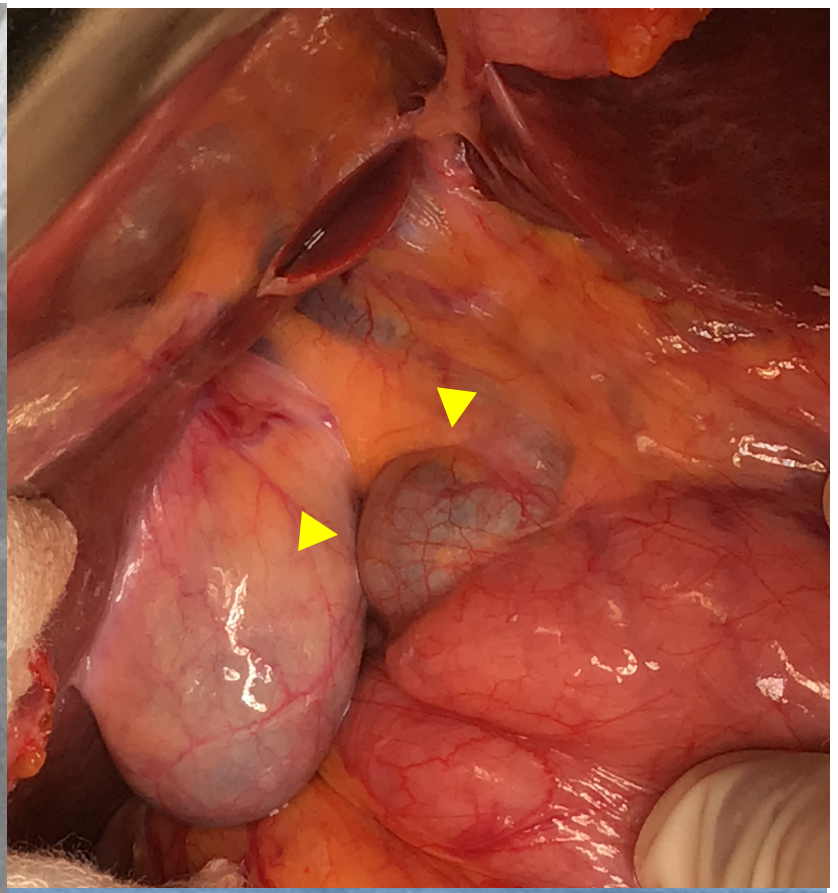
A female in her 30s.

She experienced repeated episodes of abdominal pain.

The BD was resected.

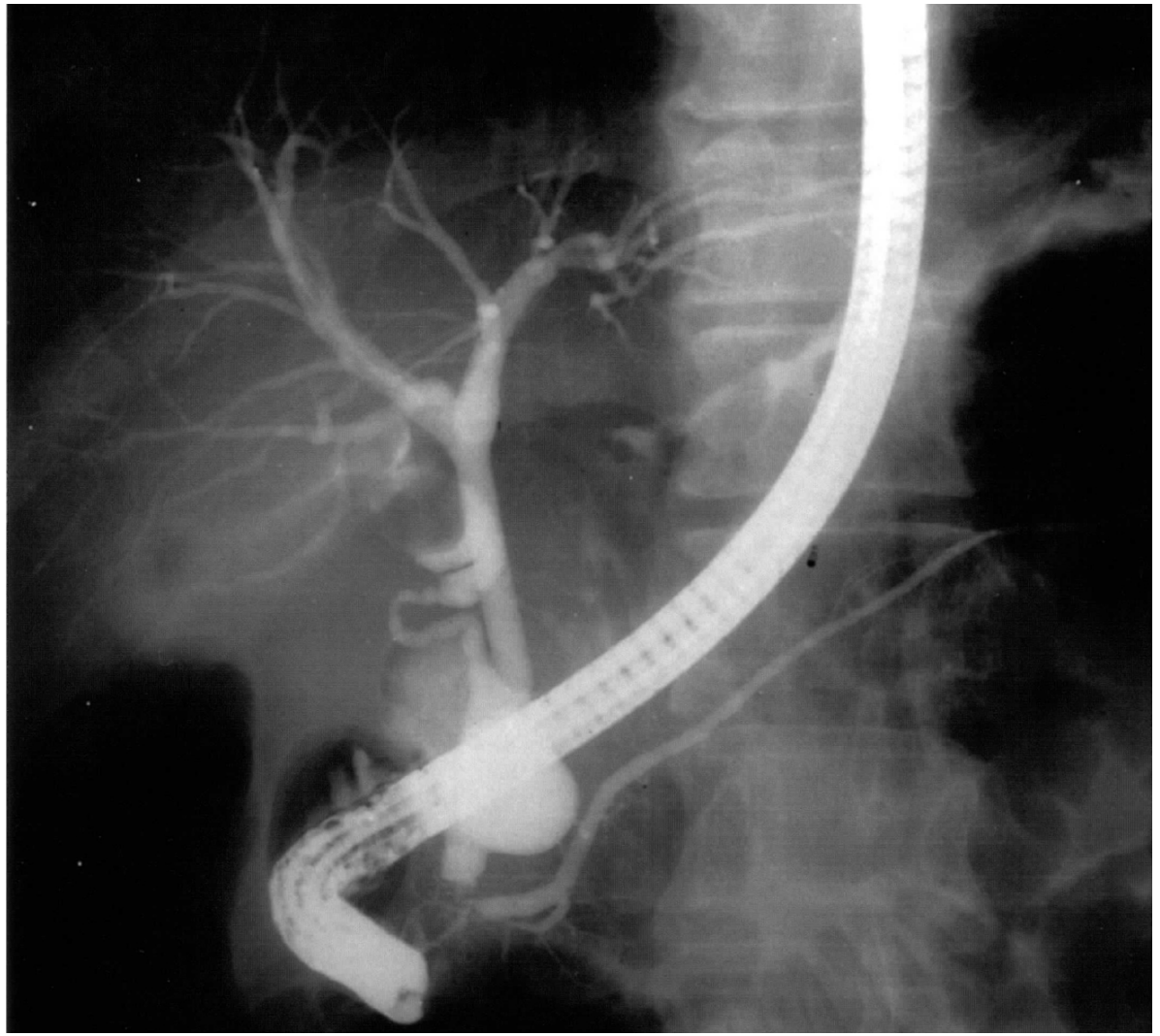








Type Ib



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

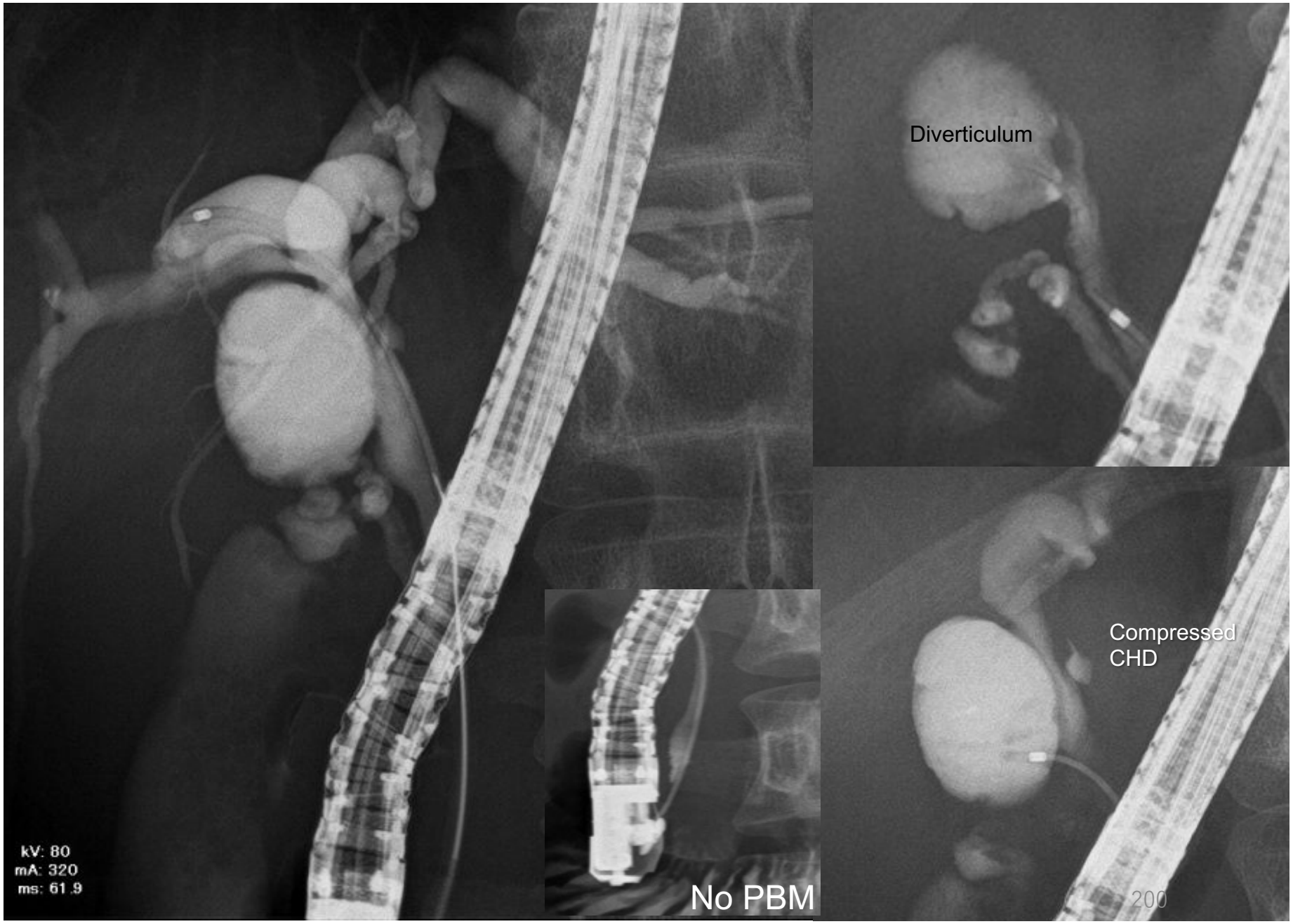
Case 52

Type II: diverticulum

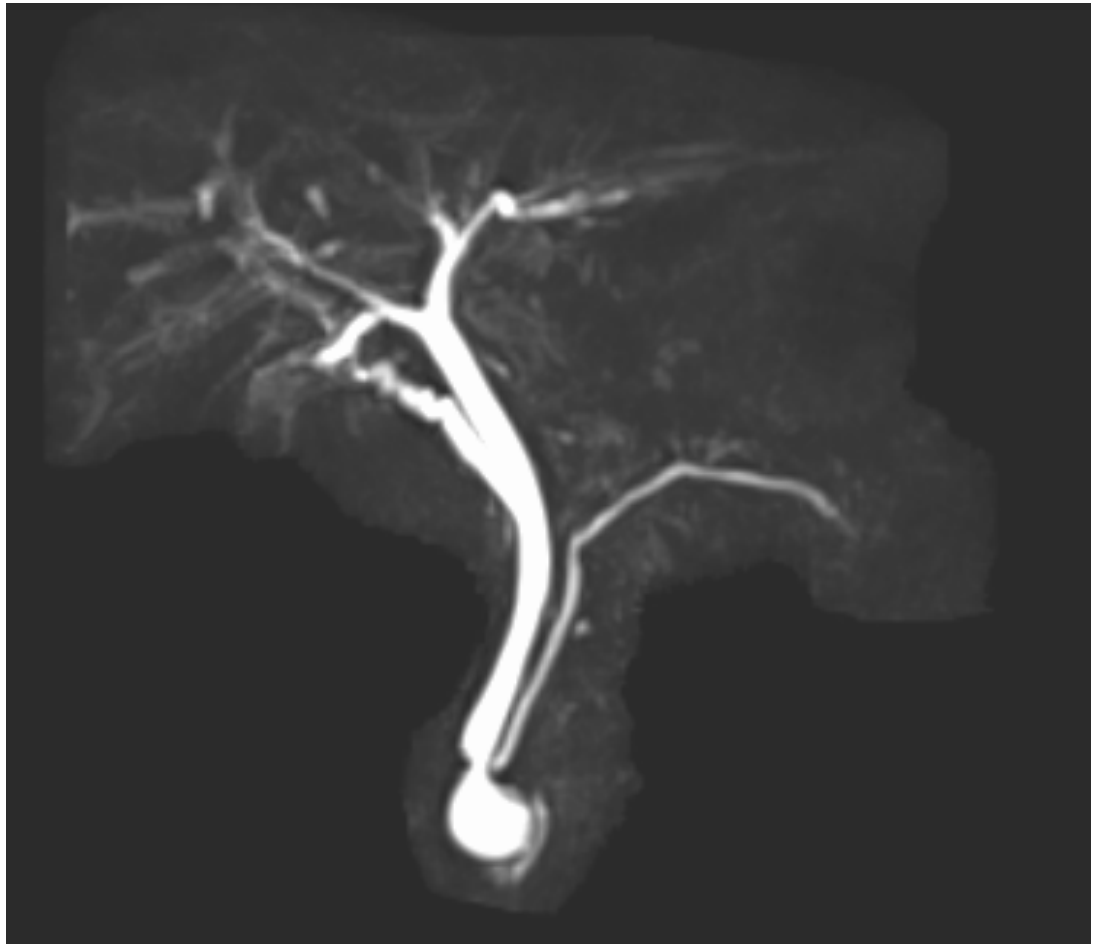
A male in his 20s.
He presented with epigastric pain and obstructive jaundice (T-bil 5.8 mg/dL, D-bil 4.2 mg/d, AST 195 U/L, ALT 595 U/L, γ-GT 733 U/L).

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

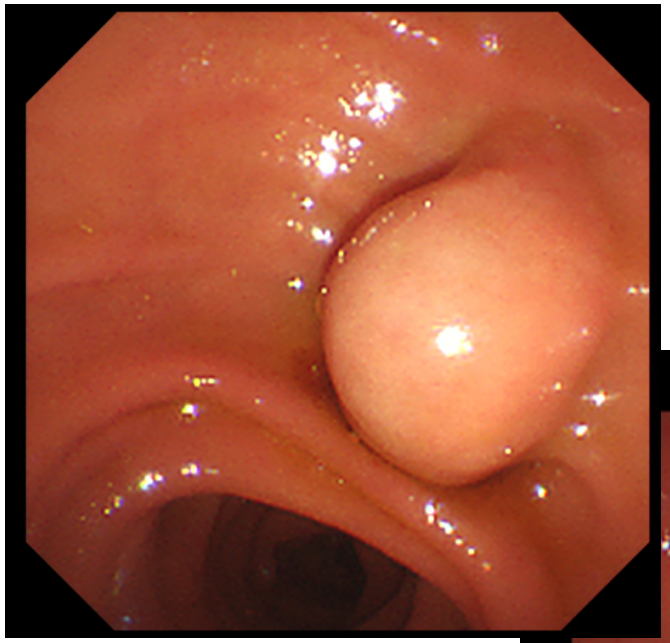
The BD was resected.

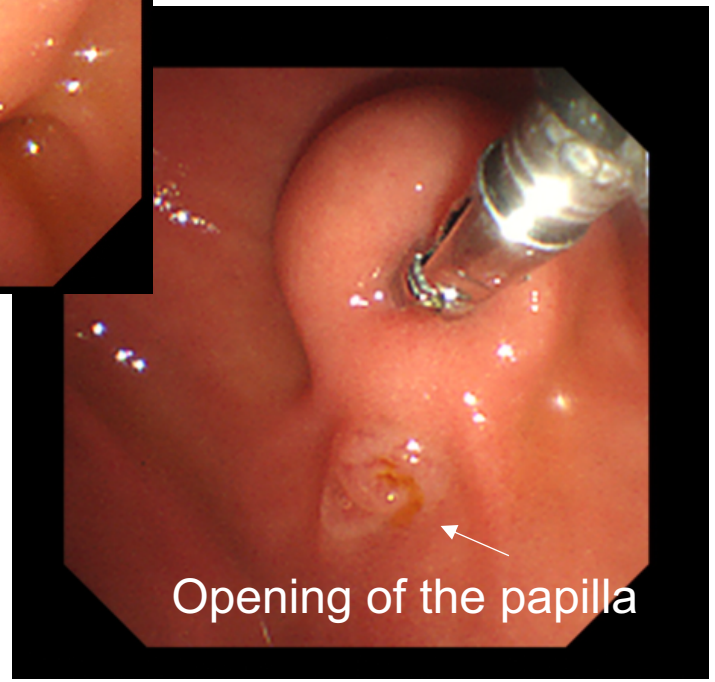


Case 53 Type III: Choledochocele



Kamisawa's classification: cystic dilatation of the intramural BD



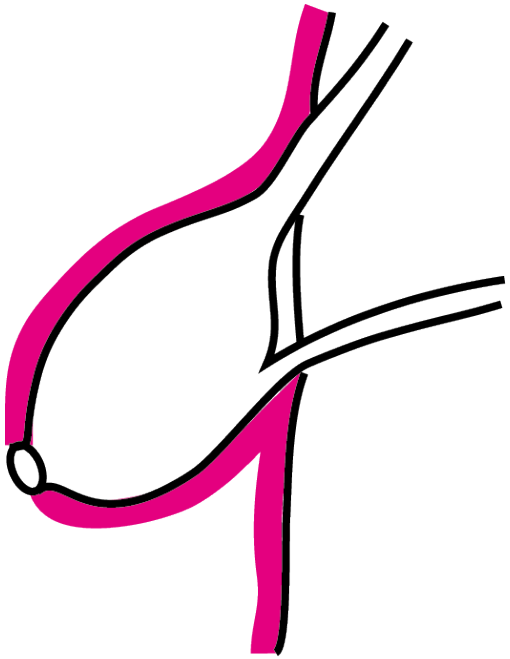


A male in his 60s. A choledochocele was detected during endoscopic examination for reflux esophagitis, and was managed by observation alone due to the absence of symptoms.

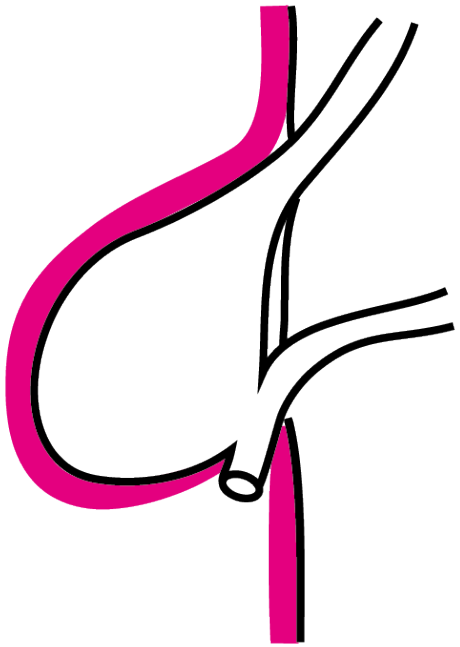
Classification of choledochocele

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

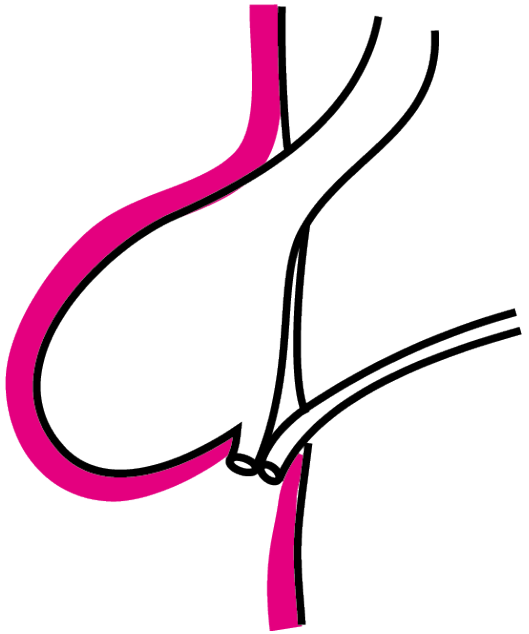
This type contains acquired cases or cases without PBM.



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.