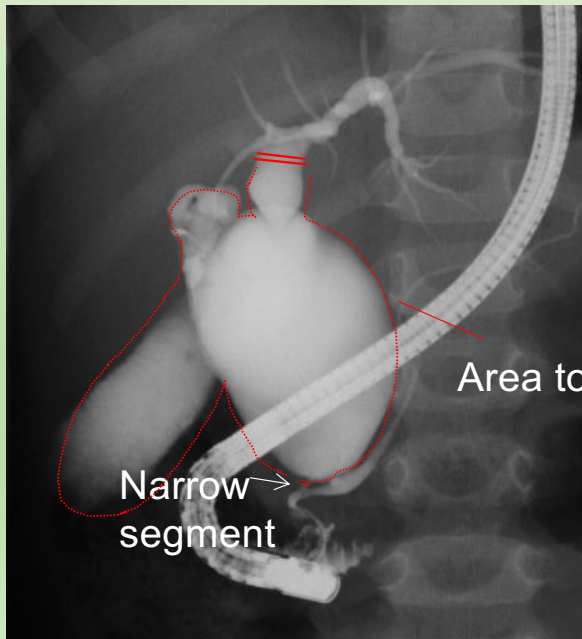
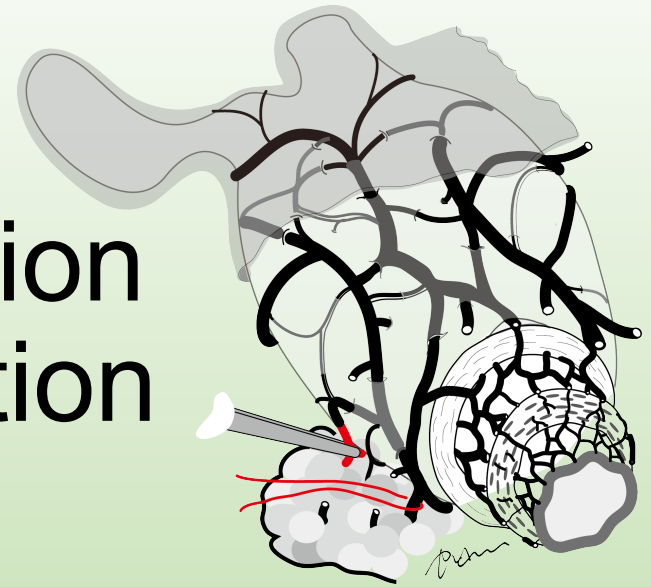


Atlas of Pancreaticobiliary Maljunction & Congenital Biliary Dilatation



Japanese Study Group on Pancreaticobiliary Maljunction (JSGPBM)
Scientific Committee

Editor-in-Chief, Kenitiro Kaneko
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In collaboration with Naoto Urushihara

Preface

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

common type with PBM in Asia as “congenital biliary dilatation”.

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

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

September 2022
Editor-In-Chief, Kenitiro Kaneko

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

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 - Type IV-B really exists?
 - Type V, Caroli syndrome & Caroli disease

Introduction

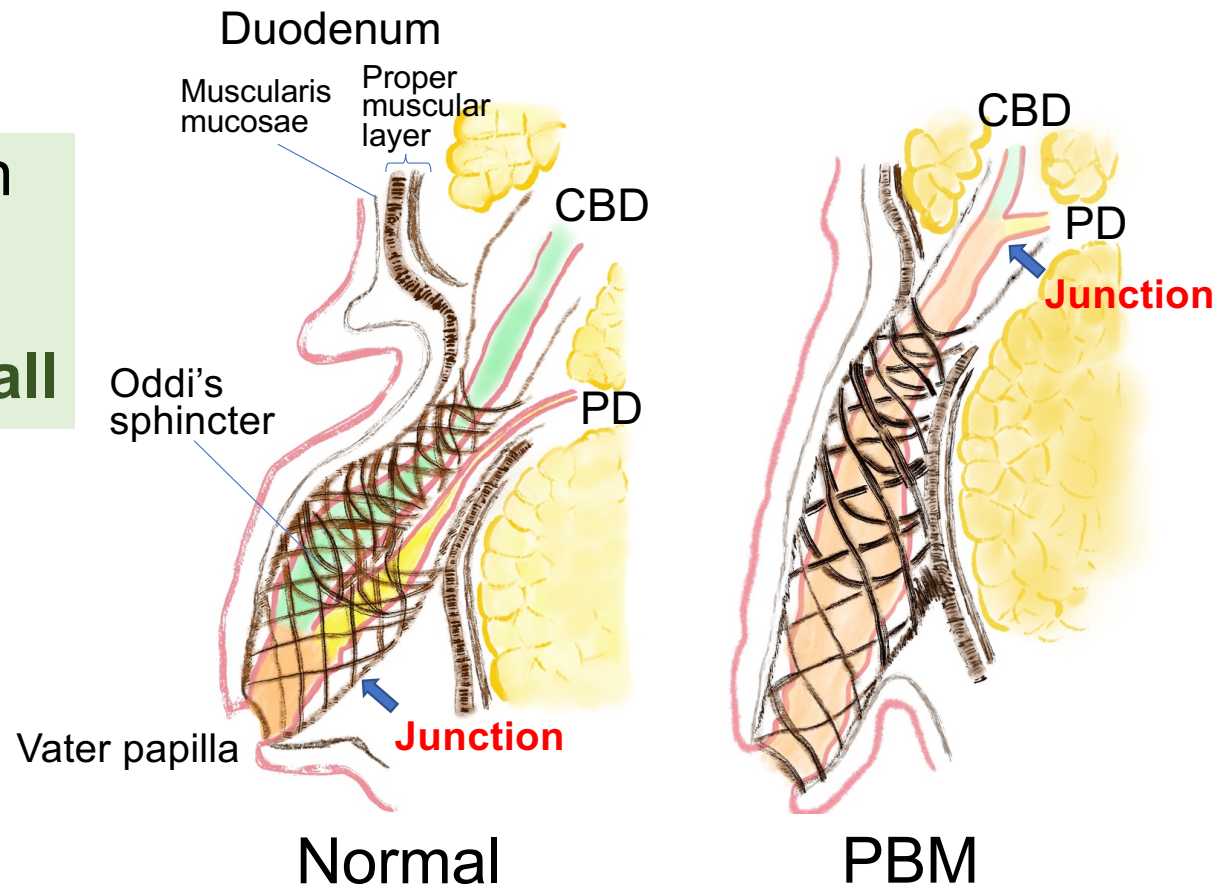
First version, September 2022

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.

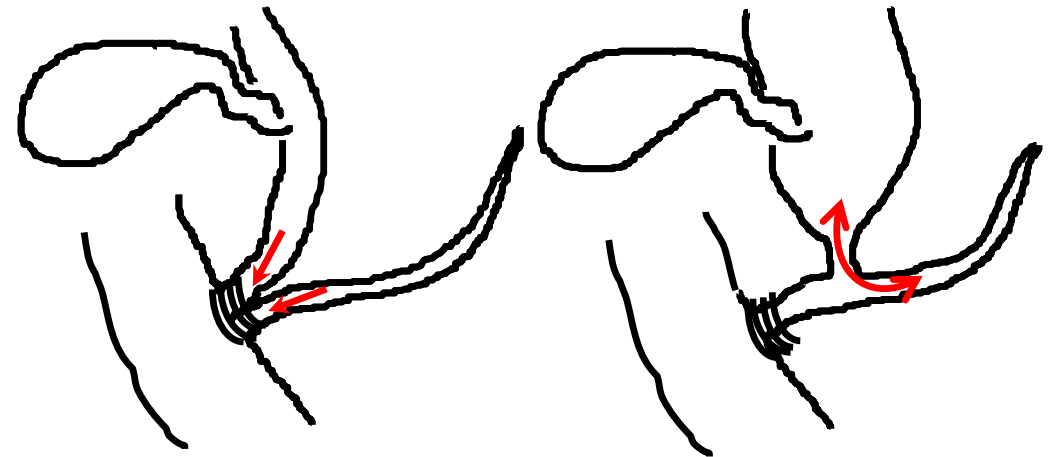


What is **Pancreaticobiliary Maljunction (PBM)**?

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

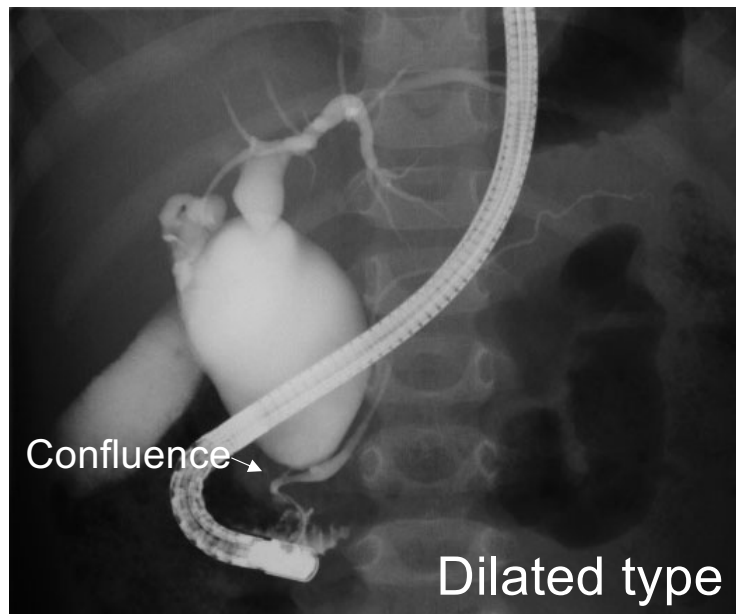
PBM

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*.

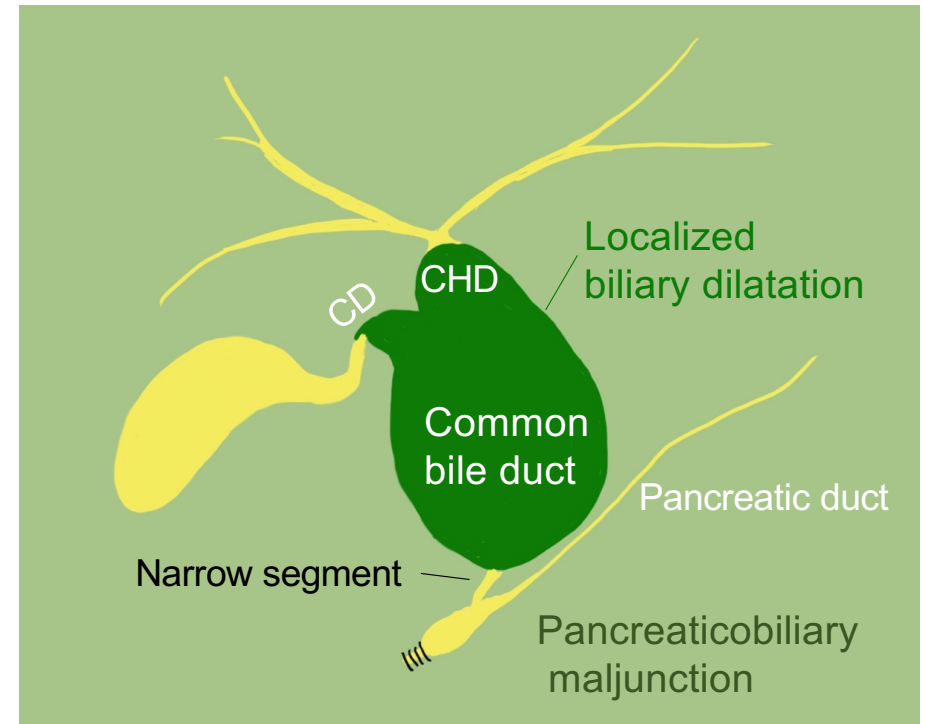


What is Congenital Biliary Dilatation (CBD)?

Definition

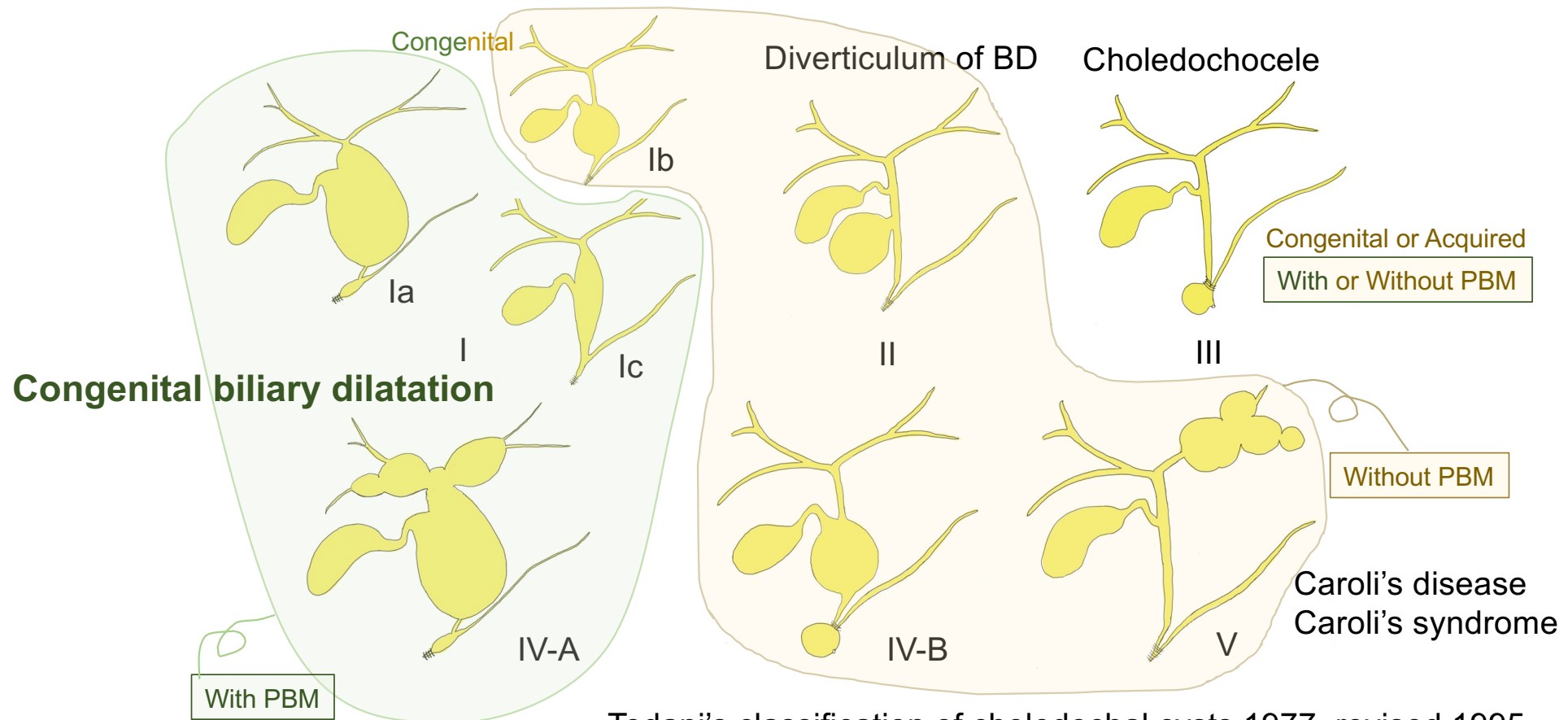
Congenital malformation involving **PBM** and **localized dilatation of the bile duct (BD)** including the common bile duct

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).

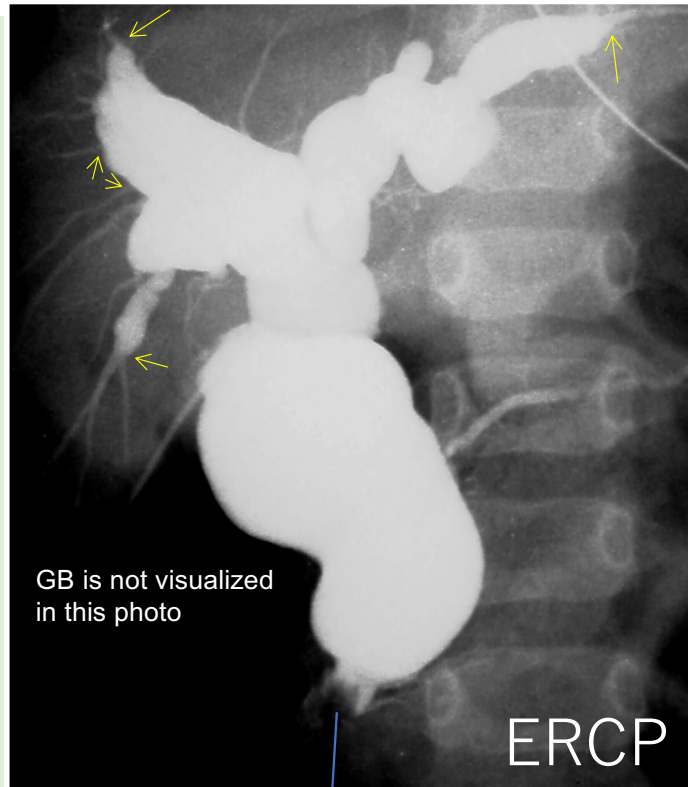
The term choledochal cyst involves different entities



“Localized dilatation of BD” indicates “congenital” disease

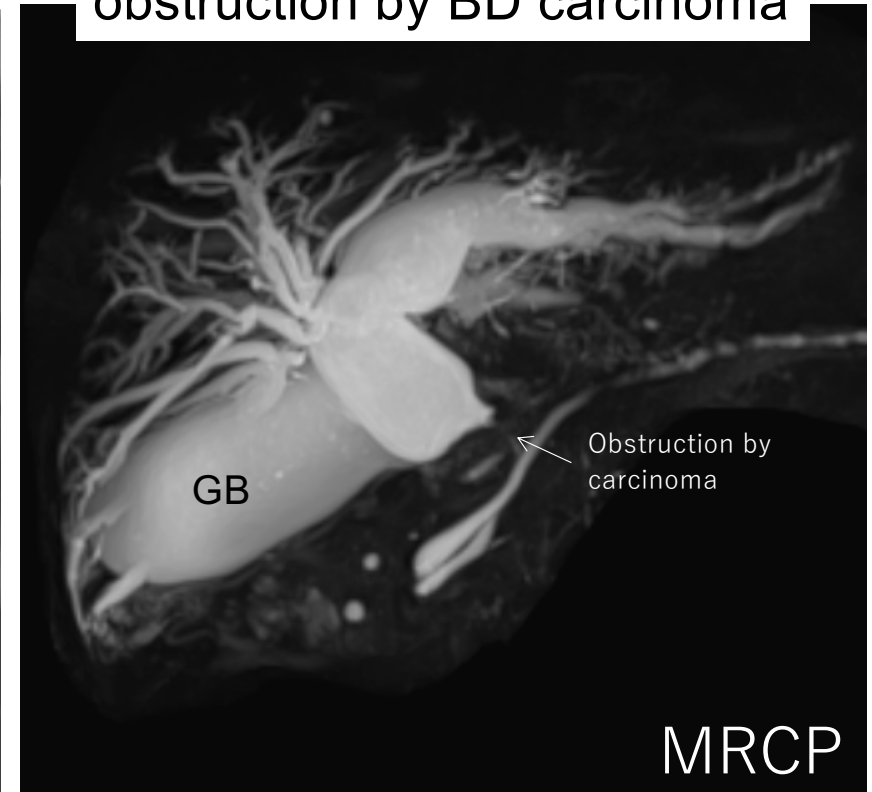
Congenital biliary dilatation

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).



Protein plug
compacted in the common channel

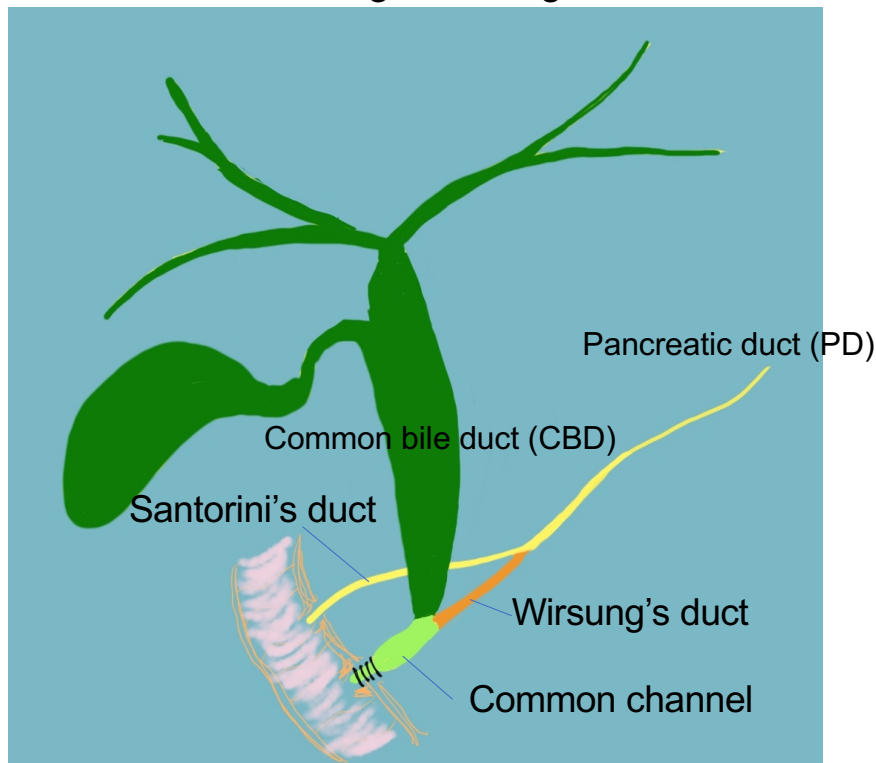
Secondary dilatation due to obstruction by BD carcinoma



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

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

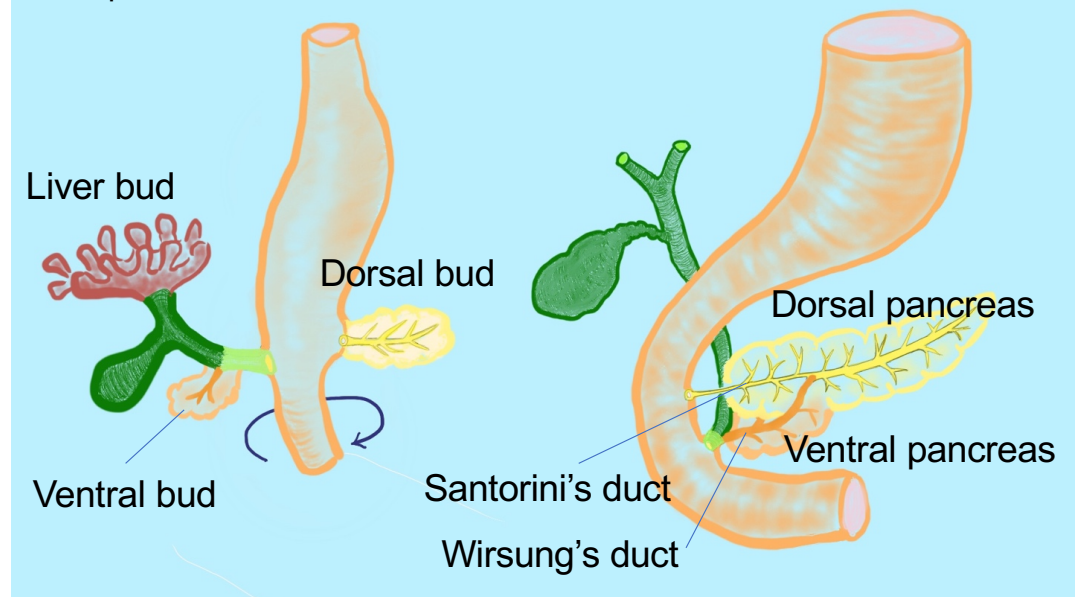
What is wrong in this figure?



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

Pancreas development

Ventral pancreatic bud rotates behind the duodenum and fuses with dorsal bud



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*.

Treatment for pancreaticobiliary maljunction

- Reasons for treatment necessity

PBM causes:

Not merely to mend malformation!

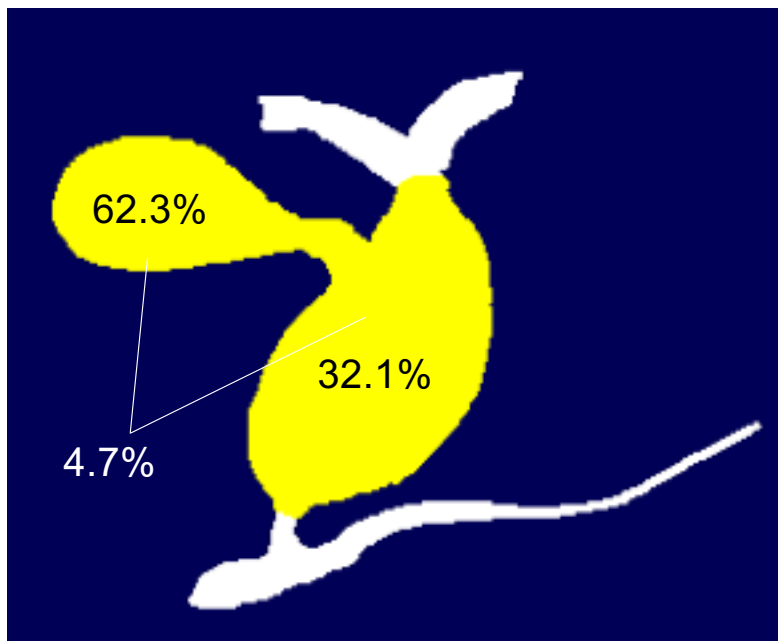
- ✓ Recurrent **symptoms** including abdominal pain, vomiting, and jaundice in children
- ✓ High occurrence of **biliary cancers** in adults see next

- **Aim** of treatment = To **prevent symptoms and cancers**
- The following must be undertaken:
 - **Bile duct resection** for congenital biliary dilatation
 - **At least cholecystectomy** for non-dilated PBM*

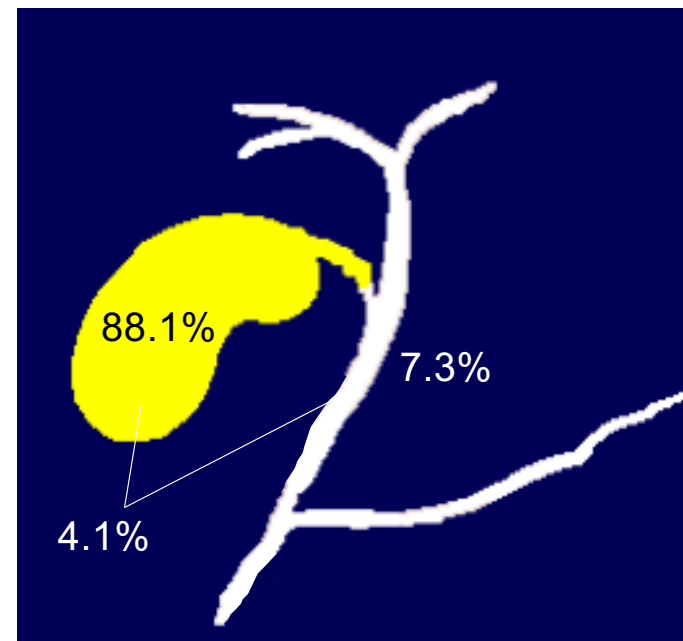
*No consensus on the need for bile duct resection because little incidence of carcinogenesis at the BD

Biliary carcinogenesis

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



Cancer rate CBD 21.6%
215/997 cases



Non-dilated PBM 42.4%
218/514 cases

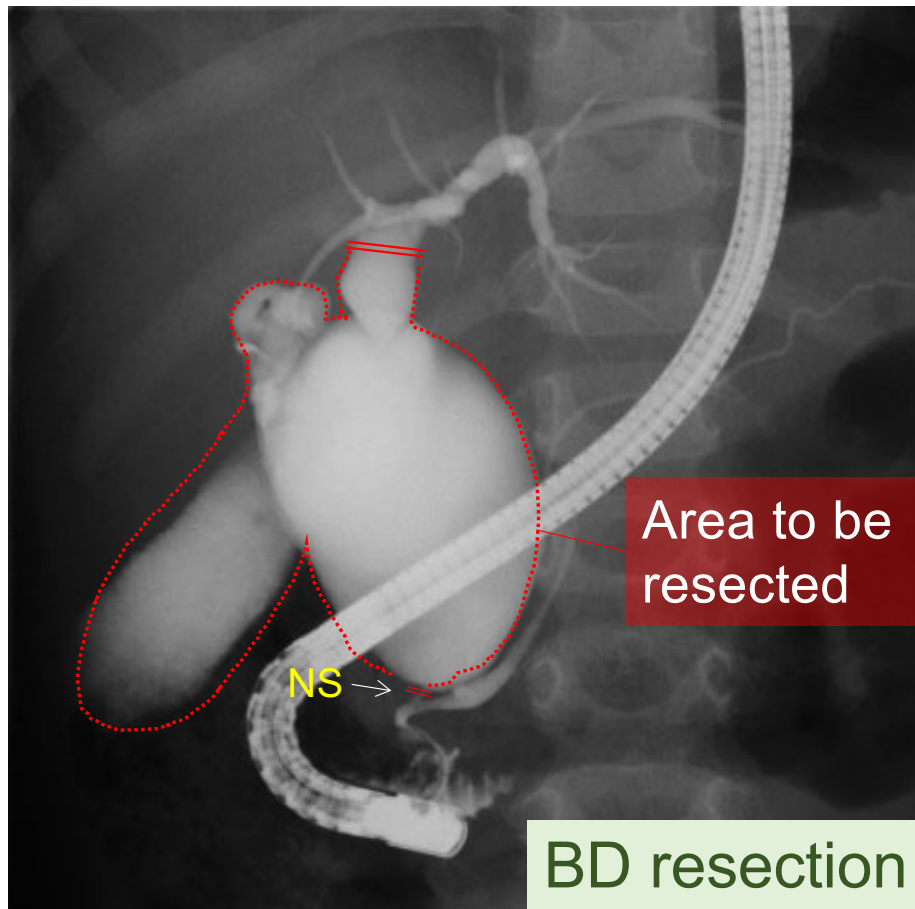
Biliary cancers arise 10–20 years earlier in PBM

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

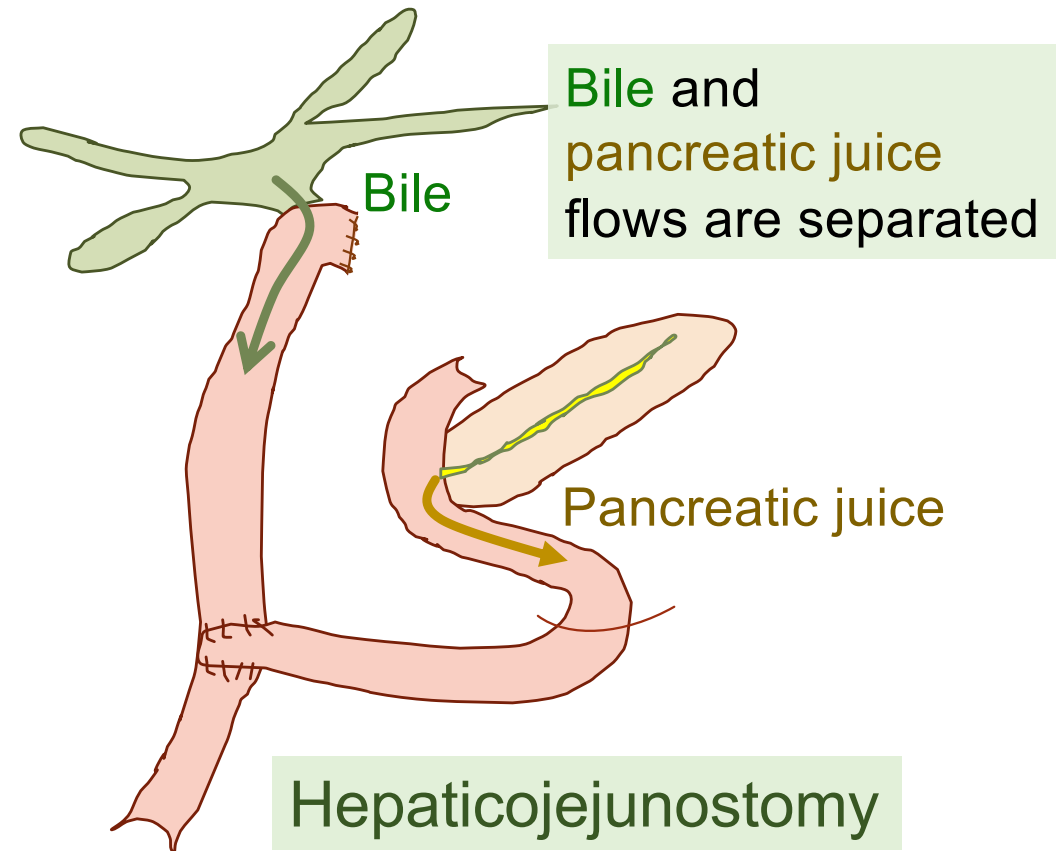
Kamisawa T, Tu Y, Kuwata G, Egawa N, Nakajima H, Tsuruta K, Okamoto A, Matsukawa M. Biliary carcinoma risk in patients with pancreaticobiliary maljunction and the degree of extrahepatic bile duct dilatation. Hepatogastroenterology. 2006 Nov-Dec;53(72):816-8.

Surgery for CBD



NS, narrow segment

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



Recommended terms

- **Pancreaticobiliary maljunction (PBM)**
- **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 Ia, c, and IV-A))

Choledochal cysts involve different entities such as CBD, choledochoceles, and Caroli's disease.

Basics

Pathophysiology, Diagnosis, and Classification

First version, September 2022

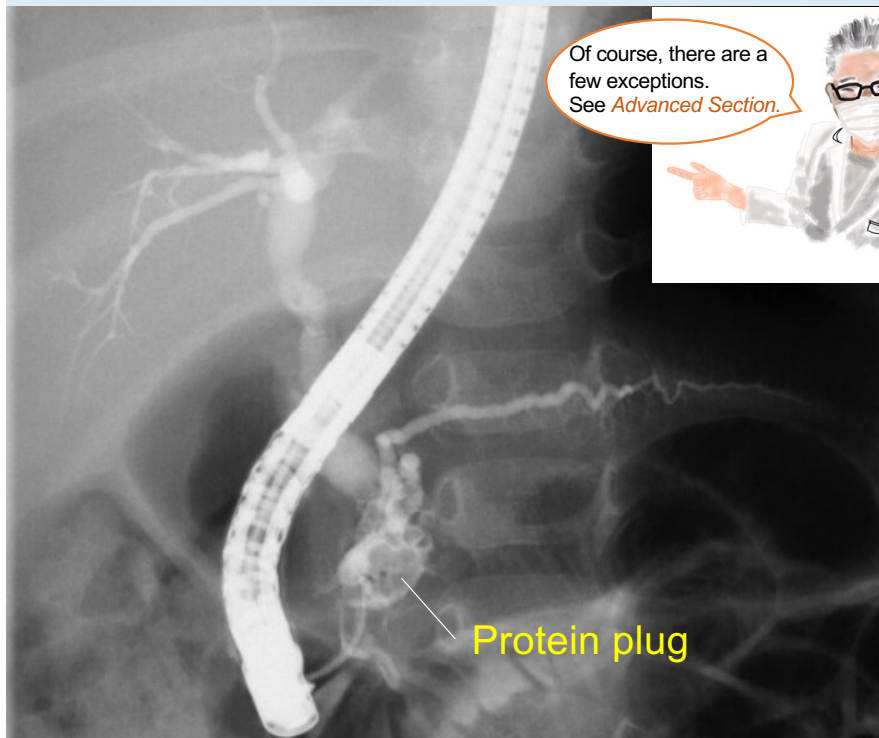
Symptoms in 128 pediatric patients

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

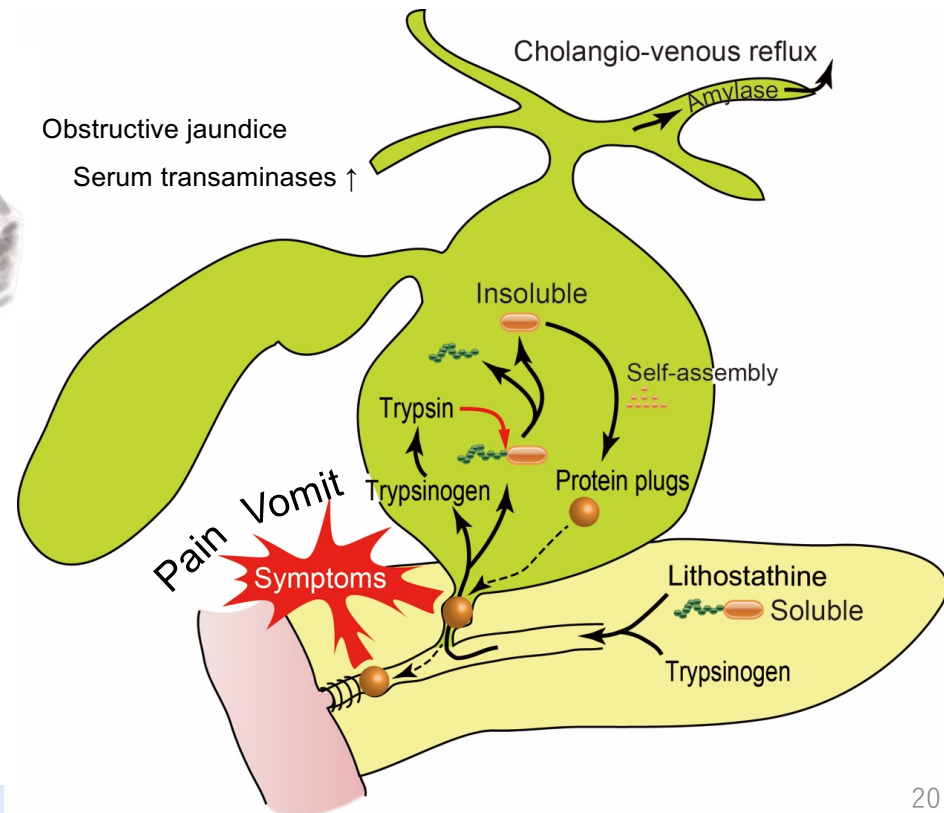
Protein plugs cause symptoms in children.

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

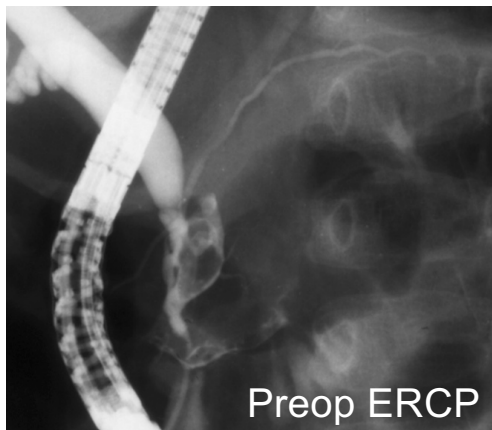


Self-limited

Repeating production



Recurrence



Spontaneous dissolution

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



Case 1

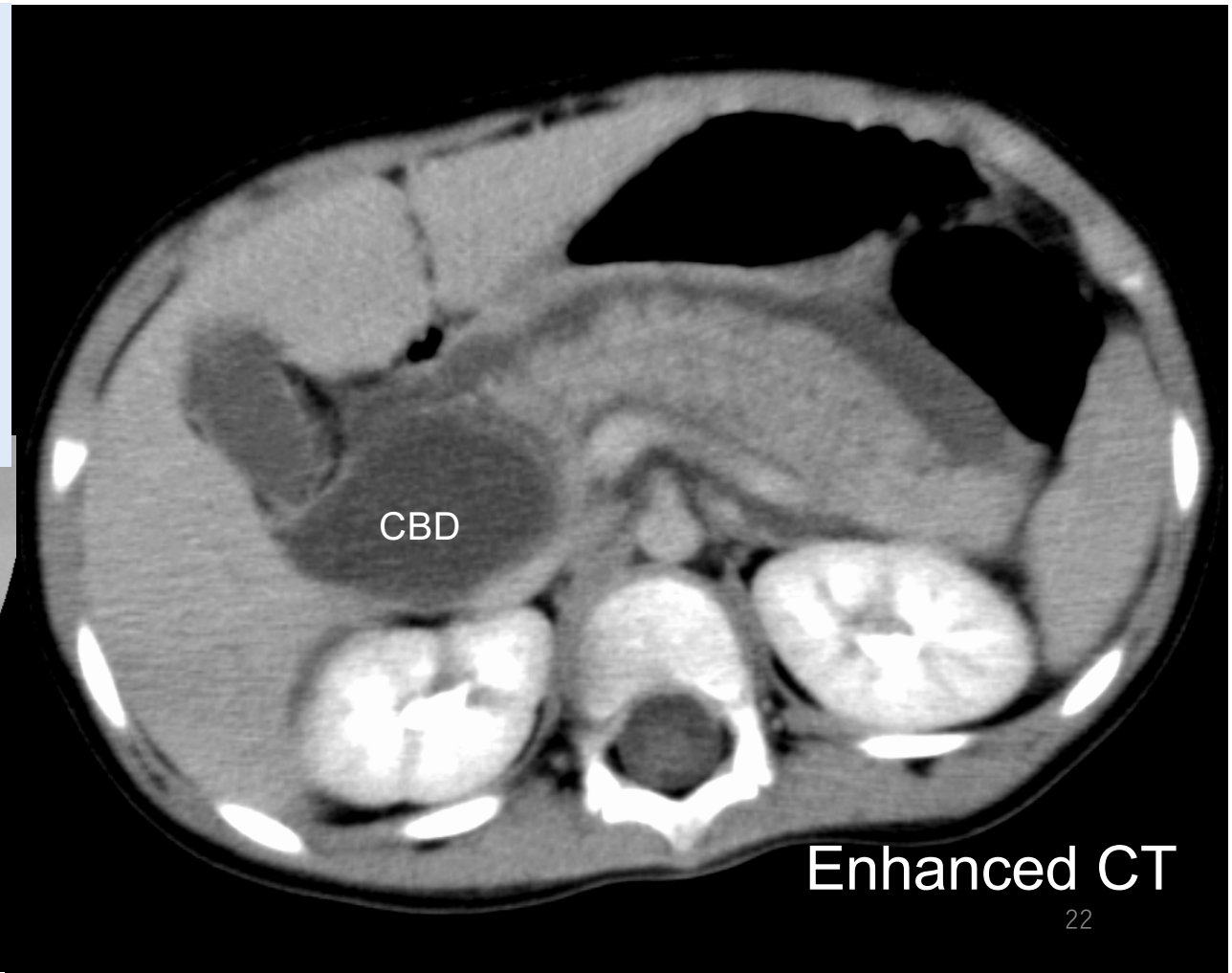
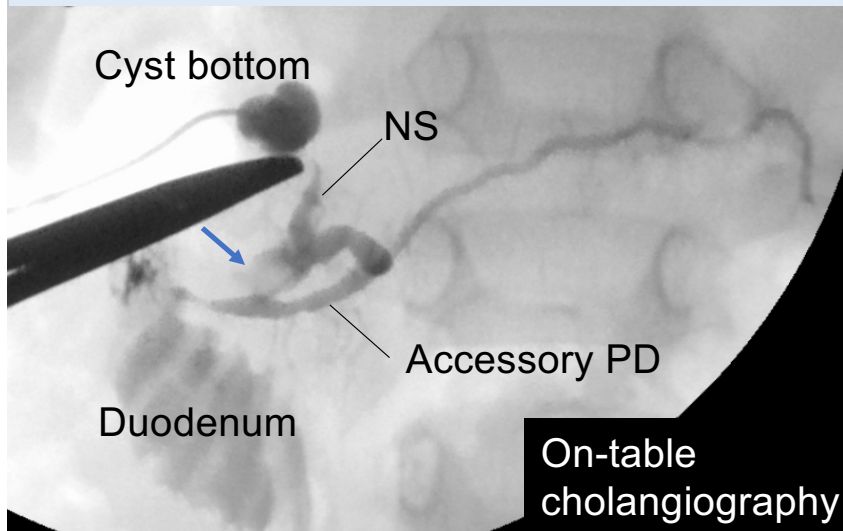
True pancreatitis is observed only in 7% of pediatric patients

Three-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.



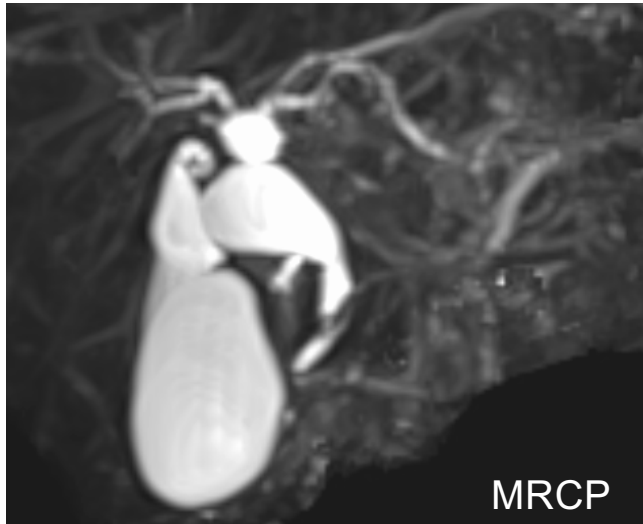
Case 2

Pseudopancreatitis Hyperamylasemia without evidence of pancreatitis

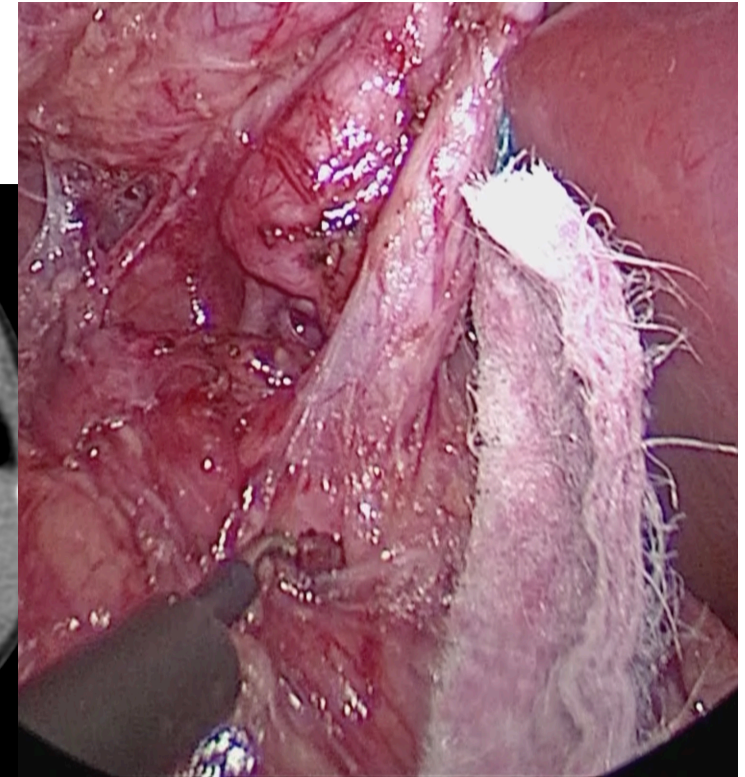
Three-year-old
Repeating **abdominal pain**
and vomiting

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

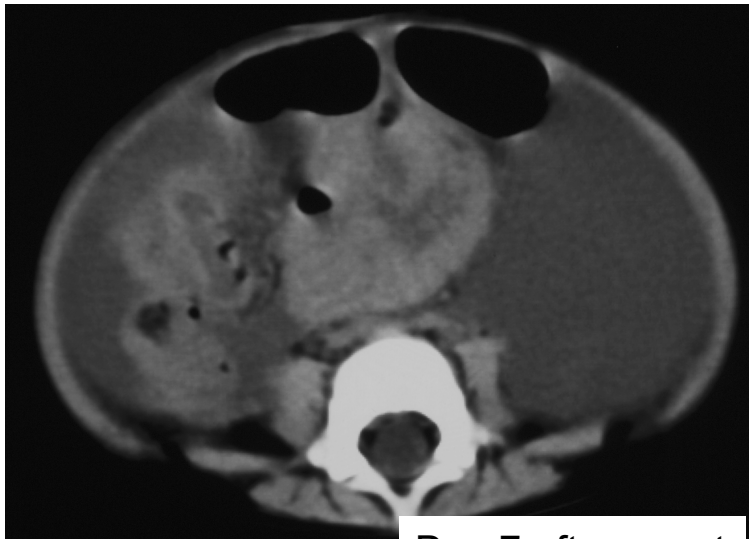
Case 3 Biliary perforation is rare: 3%-6% of children

One-year-old girl

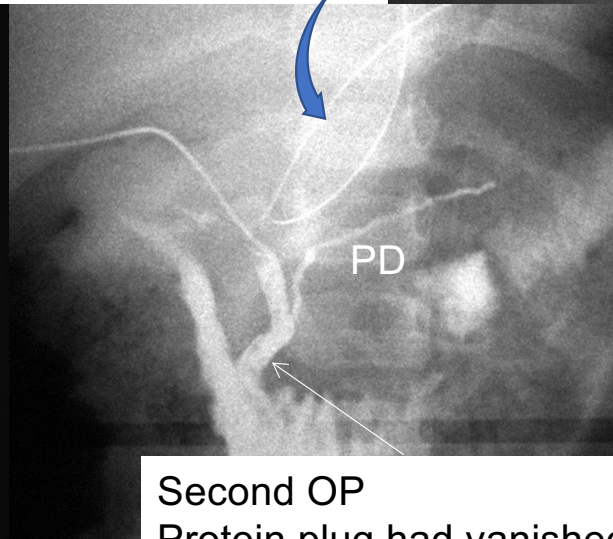
Sudden onset of vomiting and abdominal pain on day 4, diarrhea and abdominal distention on day 5, fever on day 6, and pleural effusion on chest Xp on day 7.

Bile peritonitis causes relatively mild irritation and is often overlooked.

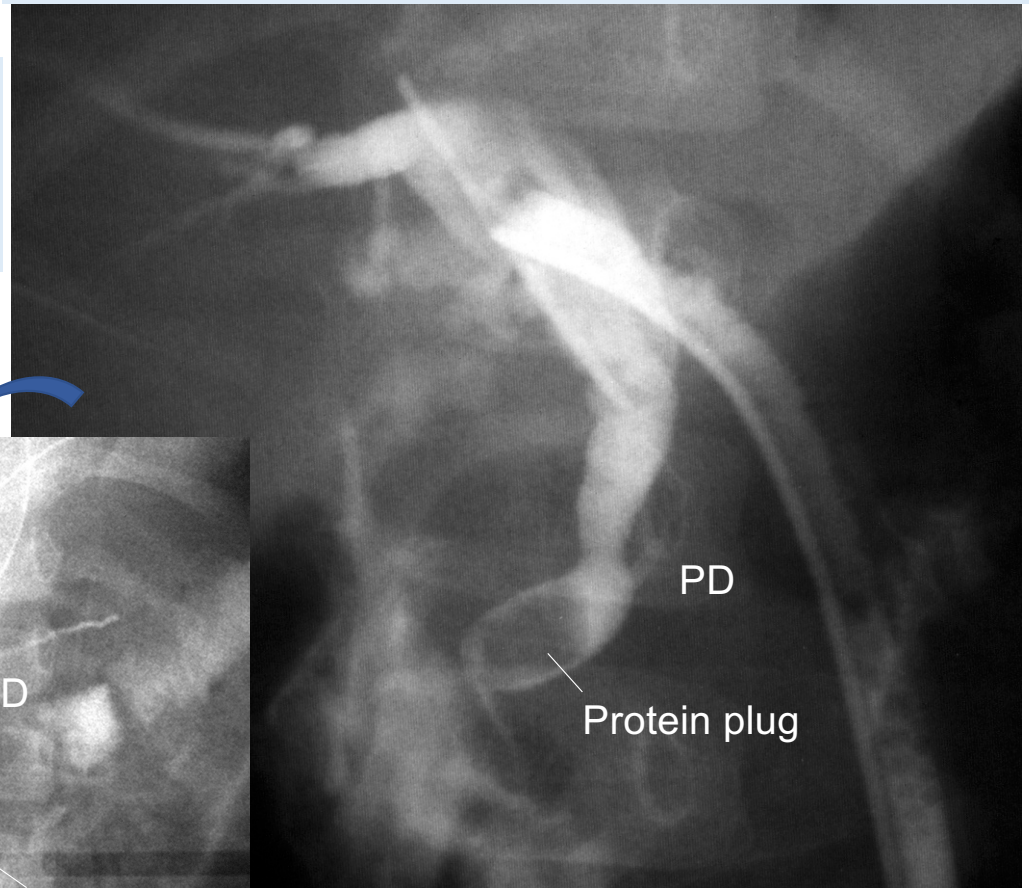
Most protein plugs are fragile and dissolve spontaneously (transient symptoms). However, exceptionally hard protein plugs produce abrupt increases in intraductal pressure, causing a rupture.



Day 7 after onset



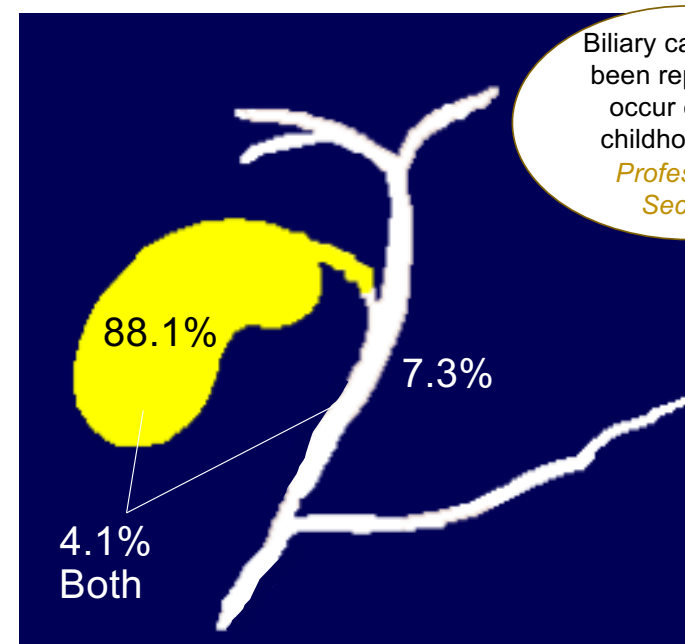
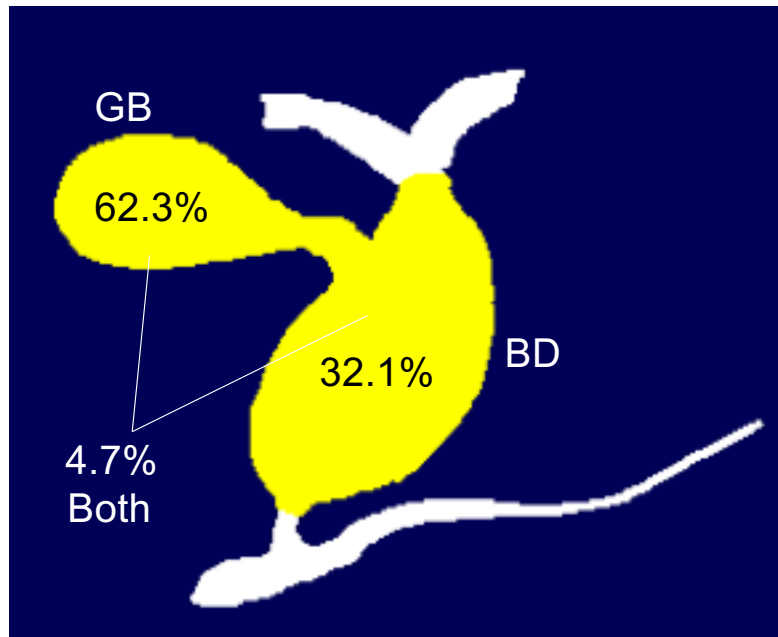
Second OP
Protein plug had vanished



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



Biliary cancer has been reported to occur even in childhood. See *Professional Section*.



Cancer rate

CBD 21.6%

Non-dilated PBM 42.4%

Carcinogenesis in region where bile stagnates

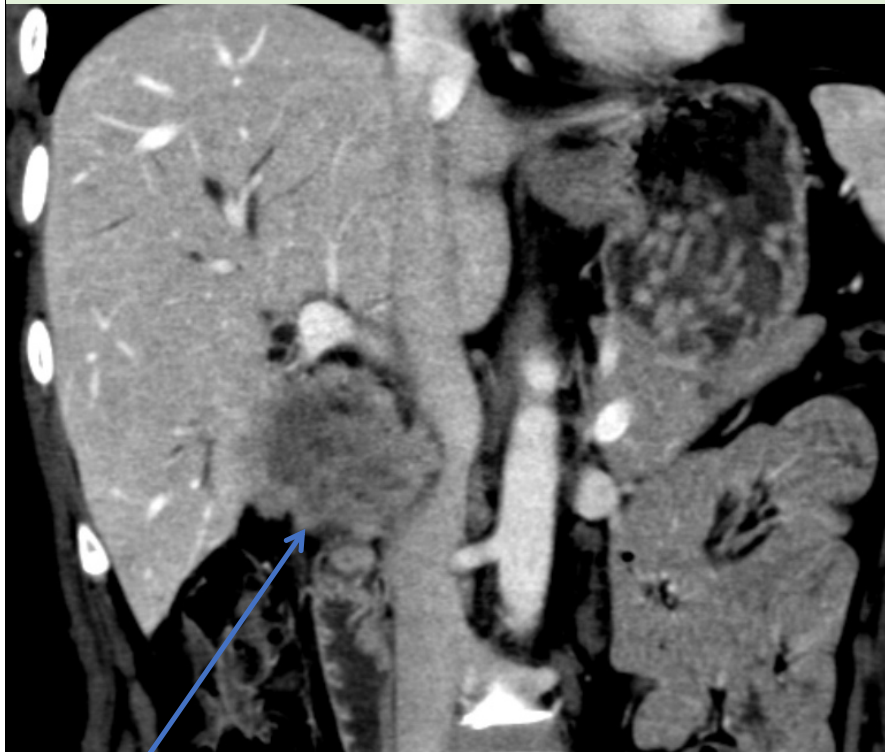


Indicates

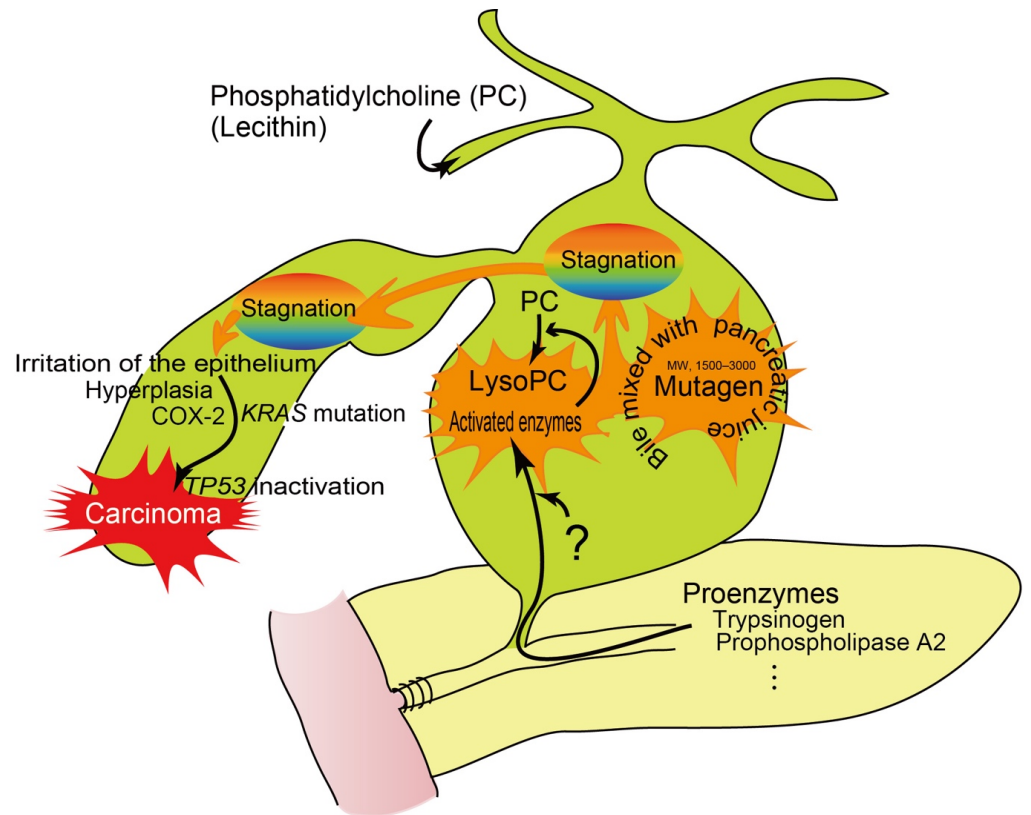
Hazardous substances, produced by a mixture of bile and pancreatic juice, stagnate and cause chronic inflammation, which induces carcinogenesis

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 forties



Hyperplasia–dysplasia–carcinoma sequence

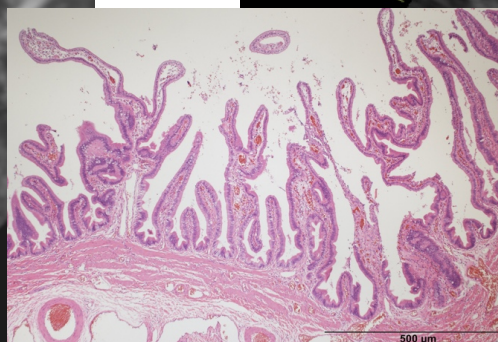
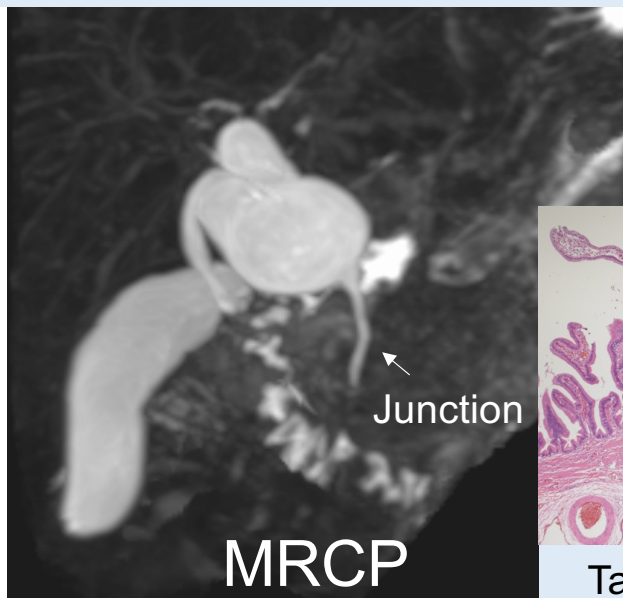
Case 4

Diffuse Hyperplasia of GB mucosa observed in a child

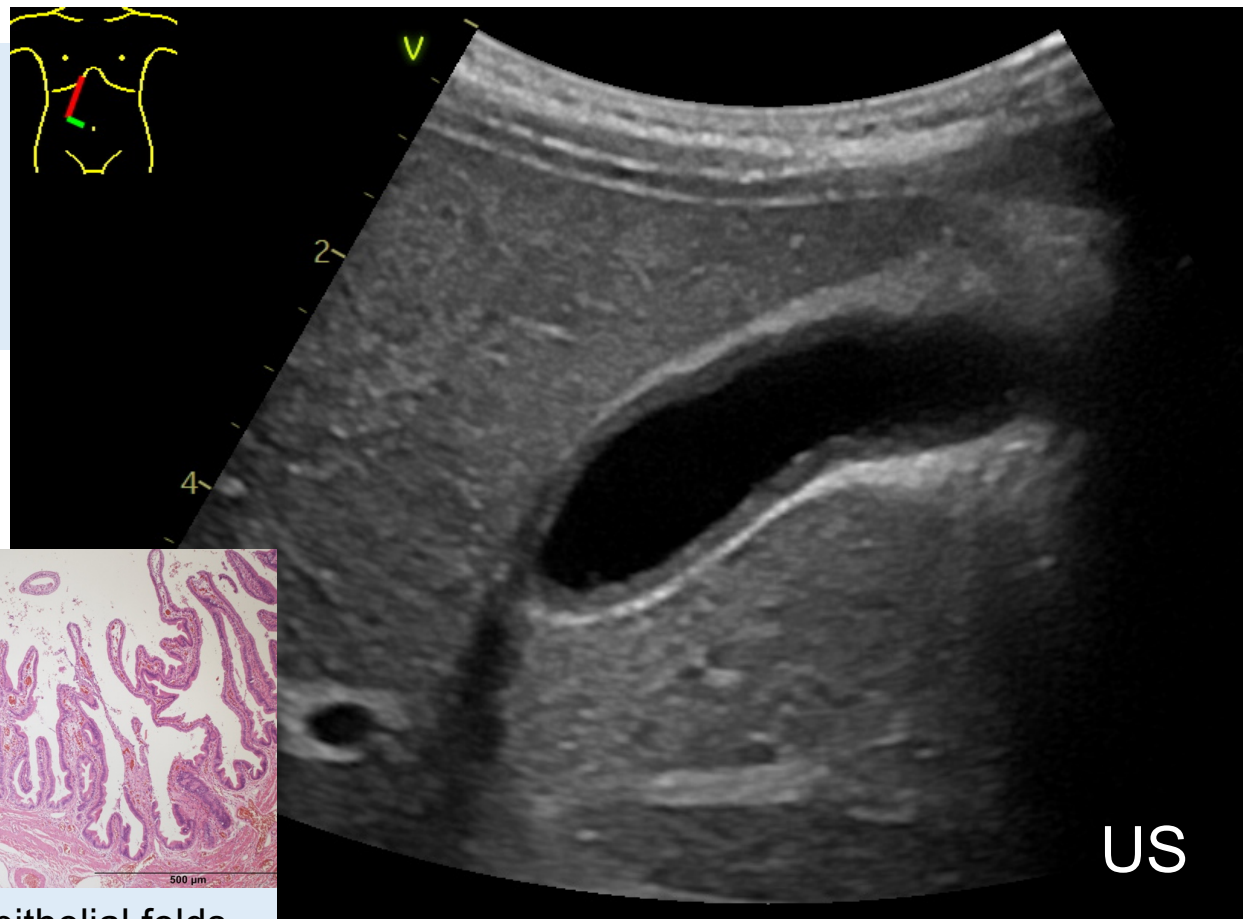
Three-year-old girl
Recurrent vomiting from 1 year of age
Abdominal pain, vomiting, and elevated
serum transaminase levels led to US,
which identified a Todani Type Ia CBD.

Thick wall of GB on ultrasonography (US)

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.

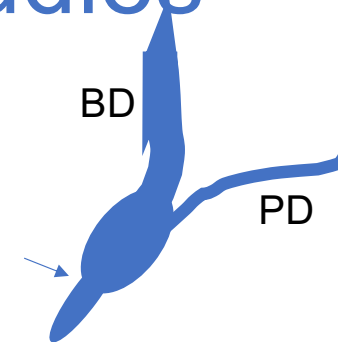


Tall GB epithelial folds



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).

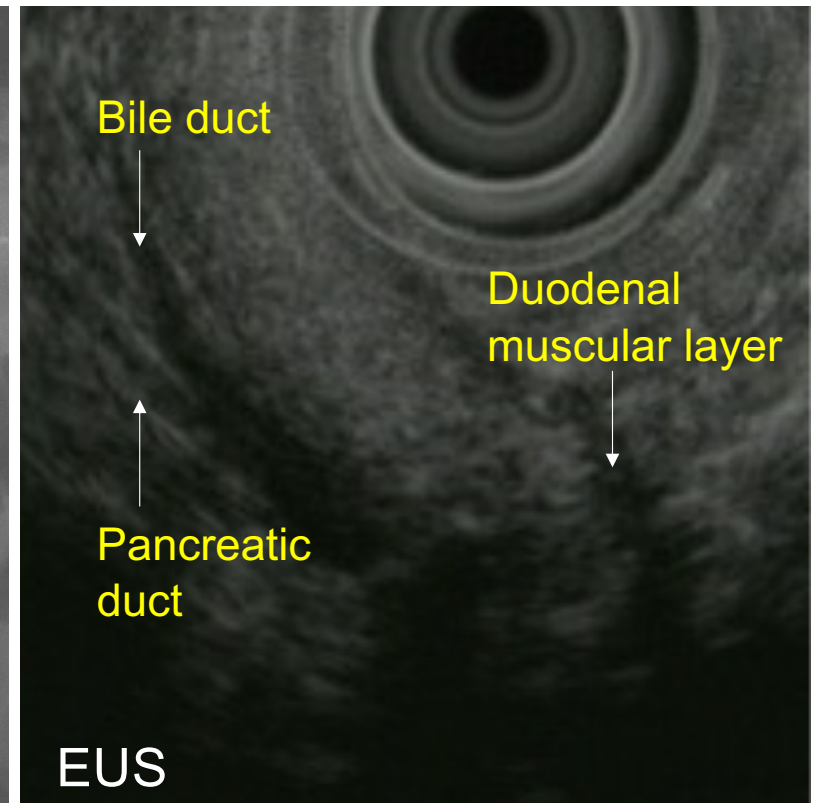
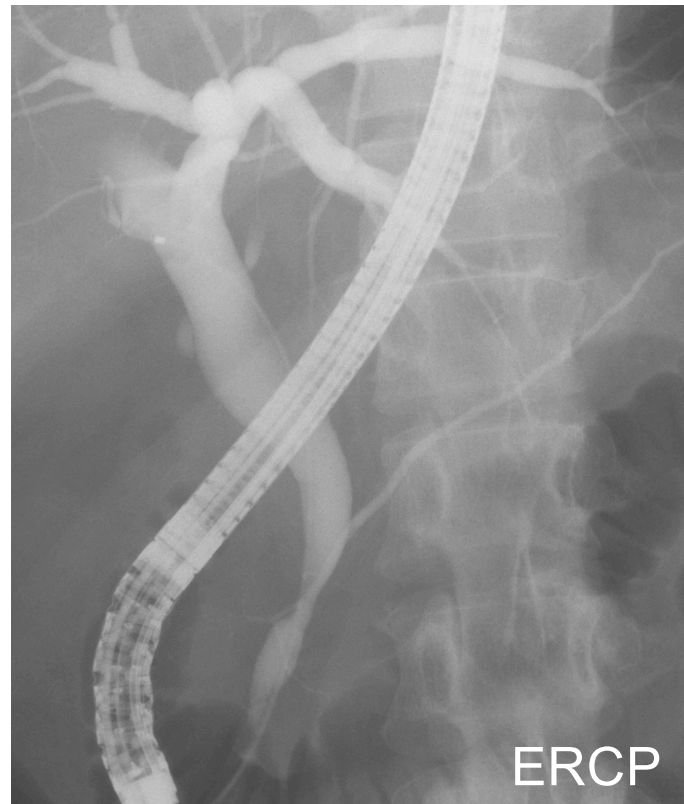
Diagnostic criteria for pancreaticobiliary maljunction 2013
J Hepatobiliary Pancreat Sci 2014;21:159–161

Case 5

EUS directly demonstrates the junction outside the duodenal wall

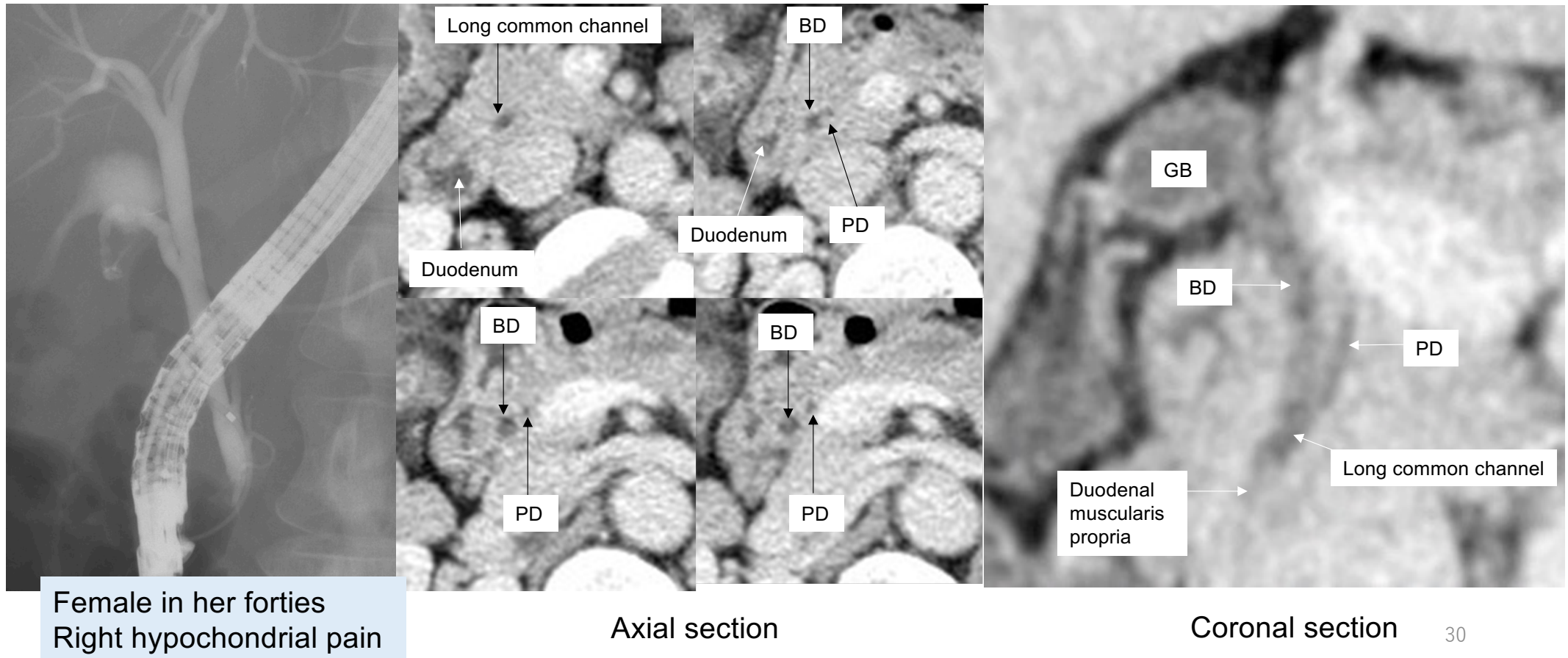
- Female in her forties
Abdominal pain
- EUS shows **union outside the duodenal wall = PBM**

EUS, endoscopic ultrasonography

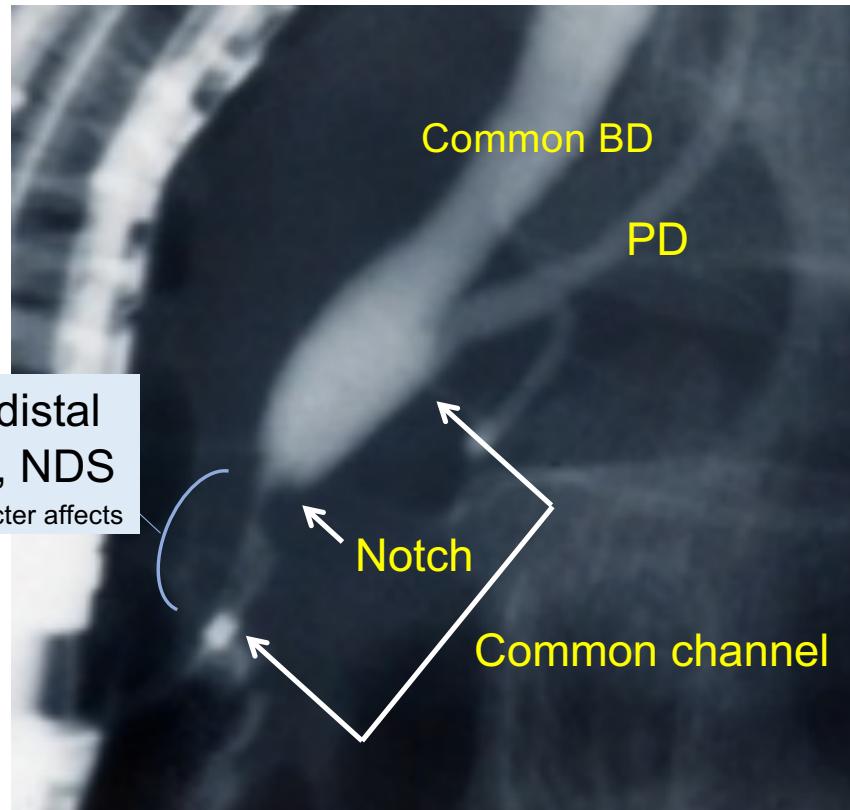


Case 6

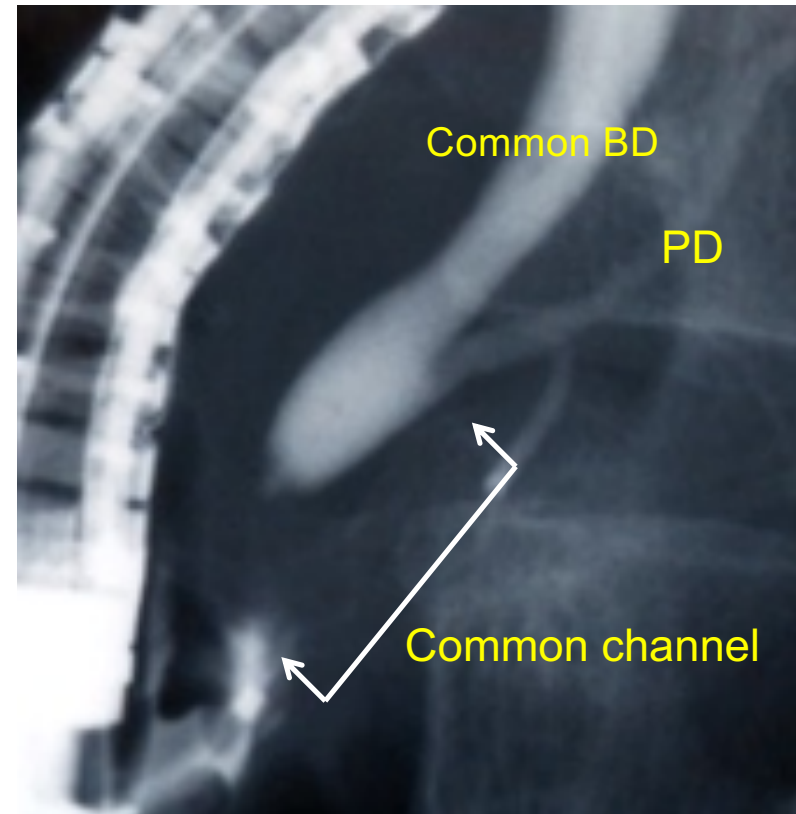
CT directly demonstrates
the junction outside the duodenal wall



Notch on the common channel = PBM



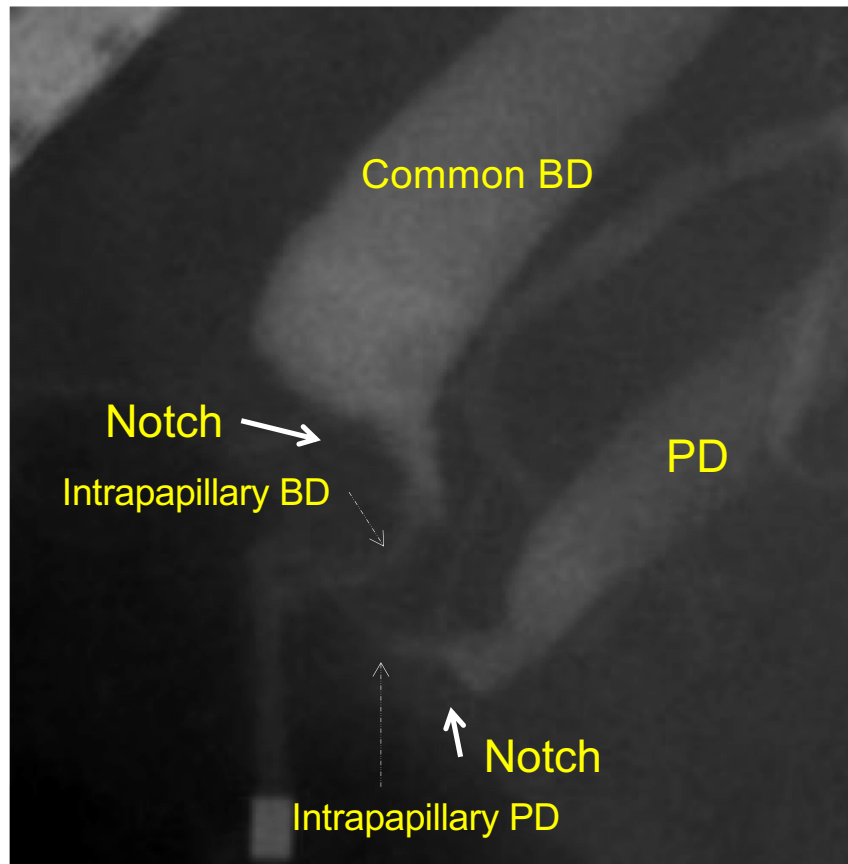
Relaxation phase
of Oddi's sphincter



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

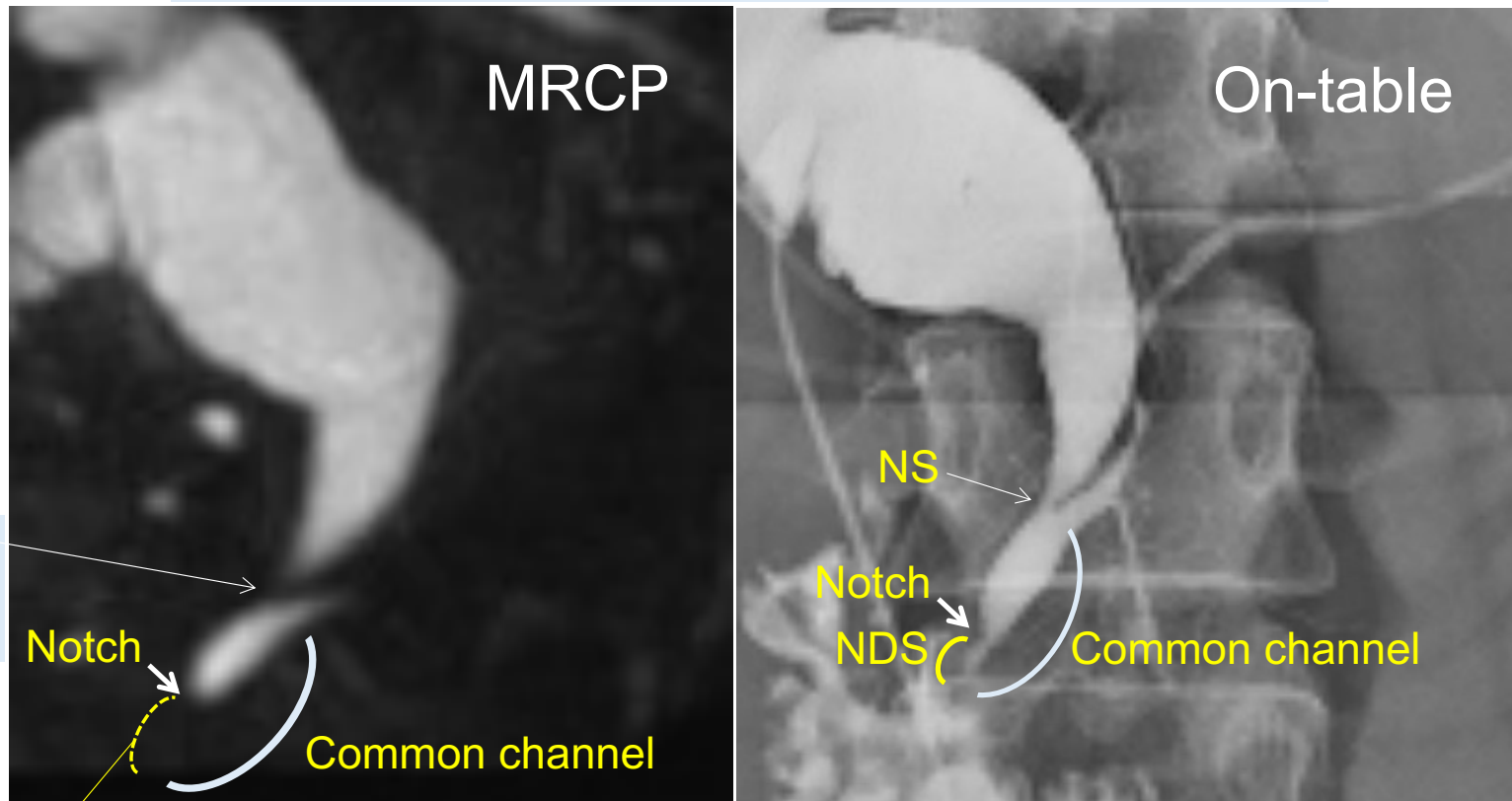


Contraction phase
of Oddi's sphincter

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

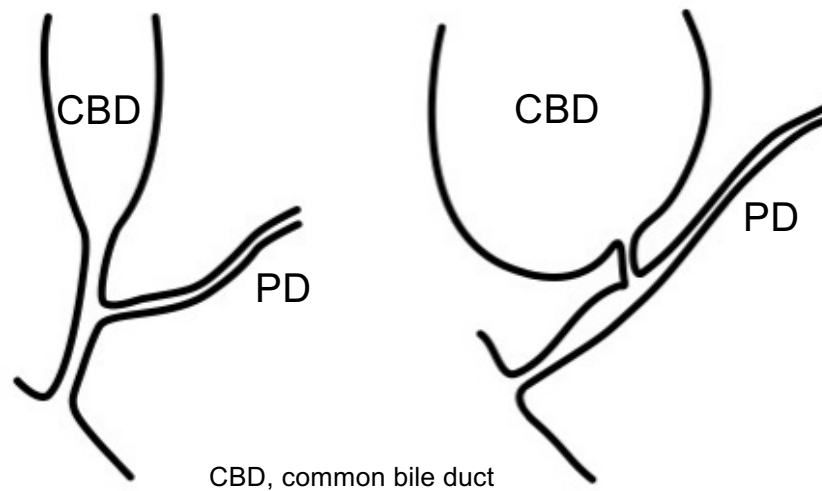


NDS not depicted → MRCP may not elucidate precise length of the common channel

Old Classification of PBM

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

Kimura's classification 1976

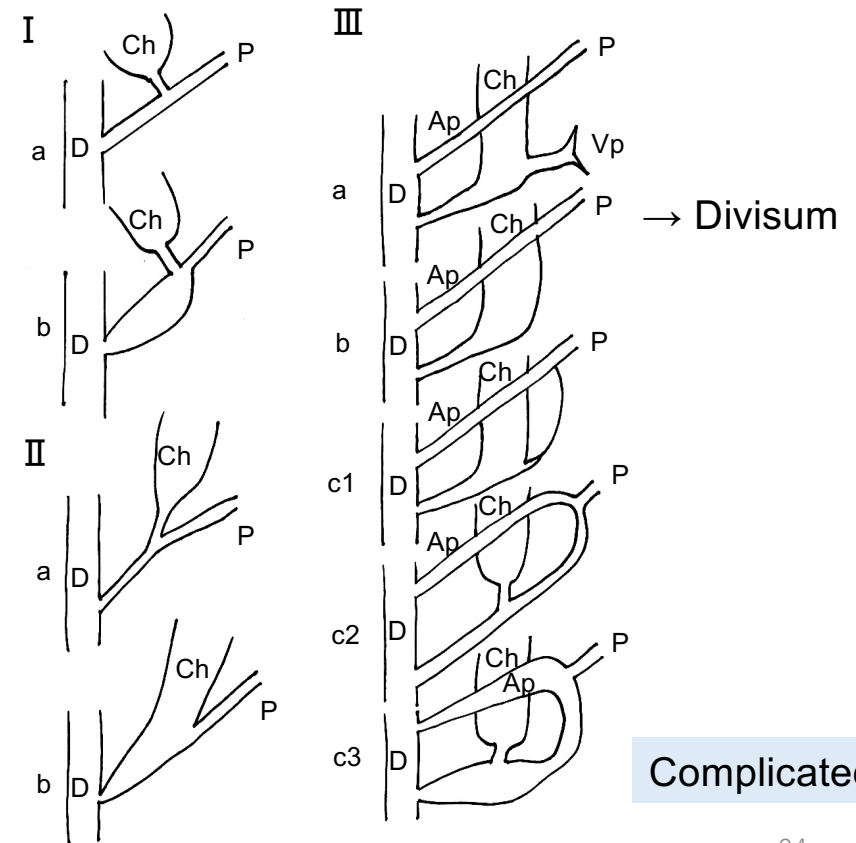


PC type
PD enters CBD

CP type
CBD enters PD

Remains basic even today

Komi's revised classification 1991



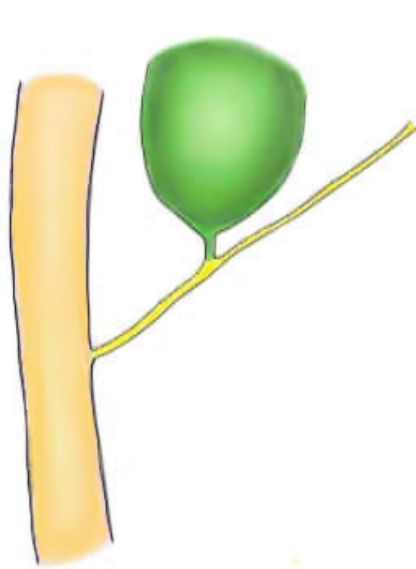
Ch, choledochus; Ap, accessory PD

Complicated

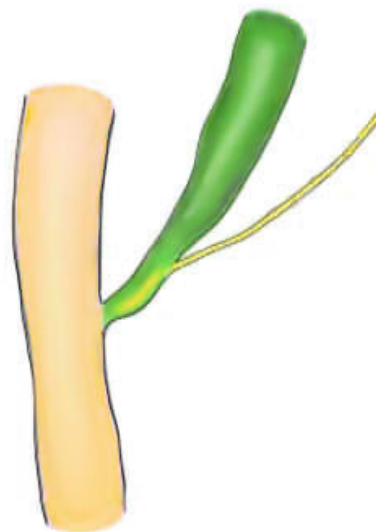
Classification by JSGBPBM

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

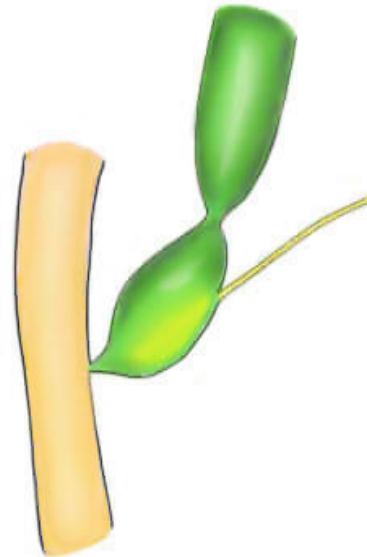
Based on the existence of narrow segment and dilatation of common channel



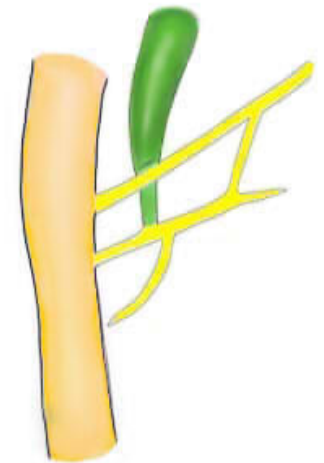
Type A
Stenotic type



Type B
Non-stenotic type



Type C
Dilated channel type

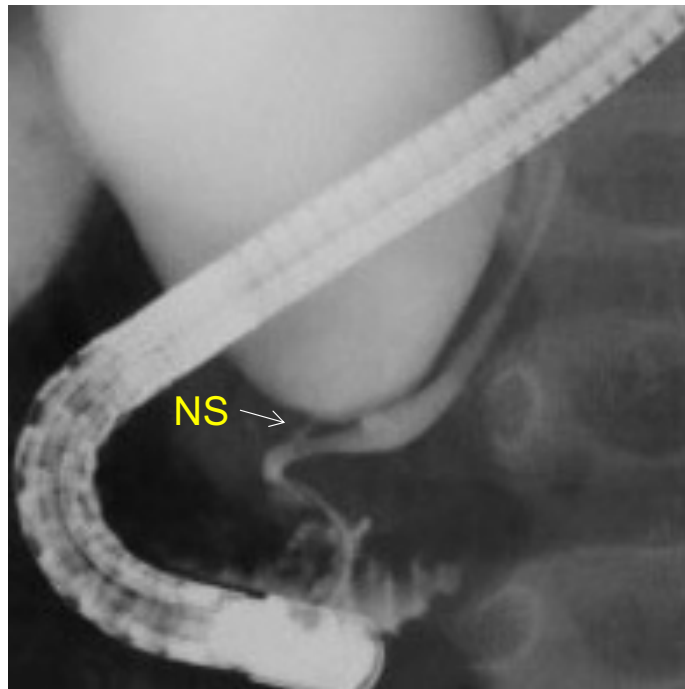


Type D
Complex type

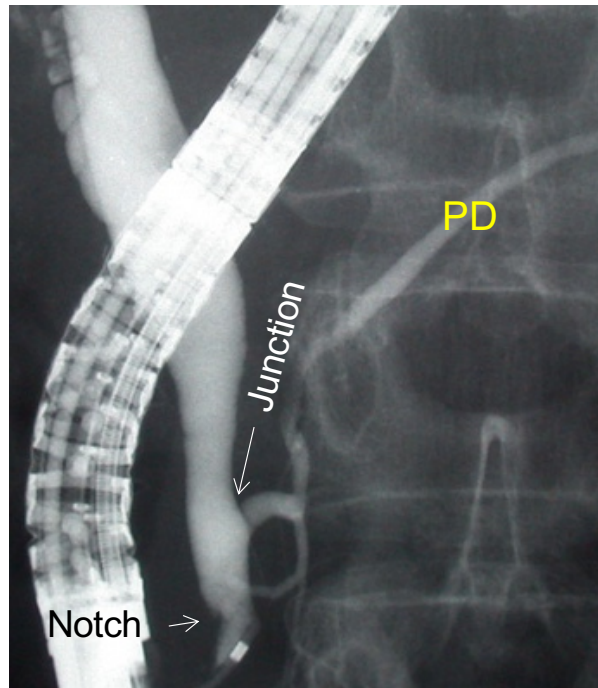
This classification is indifferent to existence of accessory PD (Santorini's duct)

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

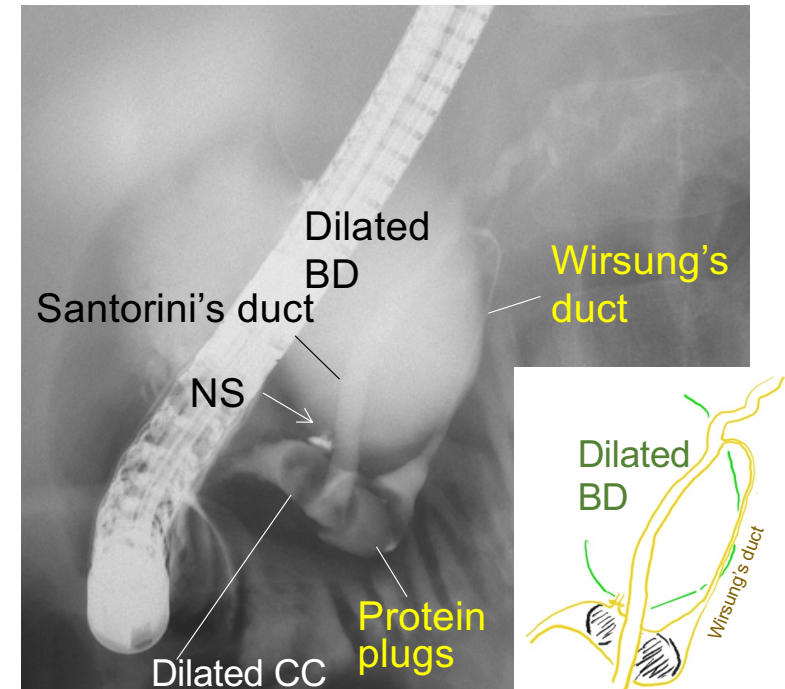
Classification by JSGBPBM



Type A
Stenotic type



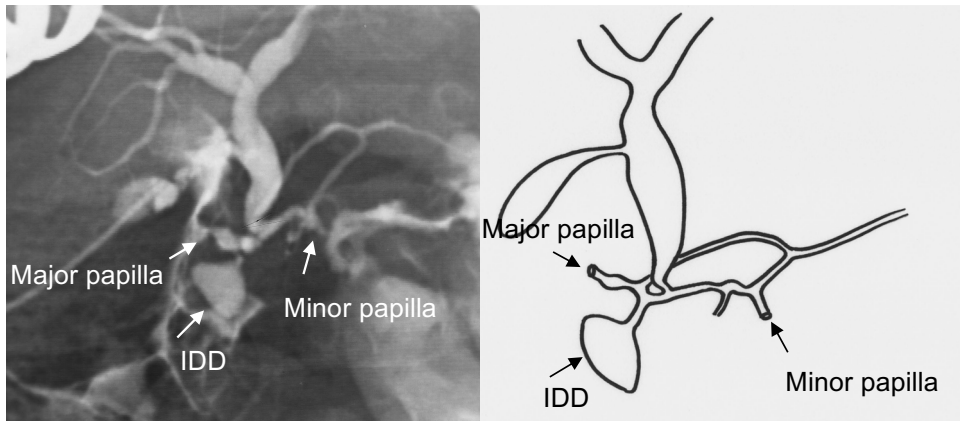
Type B
Non-stenotic type



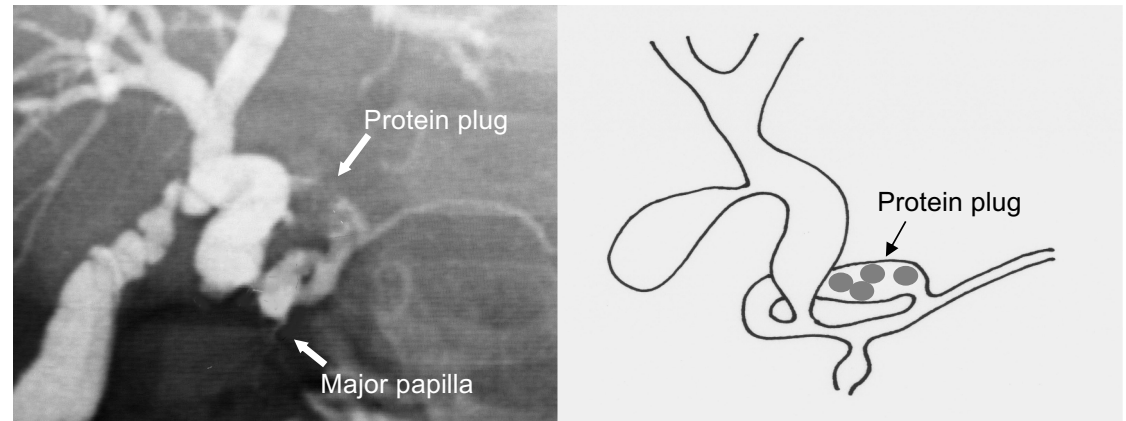
Type C
Dilated channel type

Type D: Complex type

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



IDD, intraluminal duodenal diverticulum

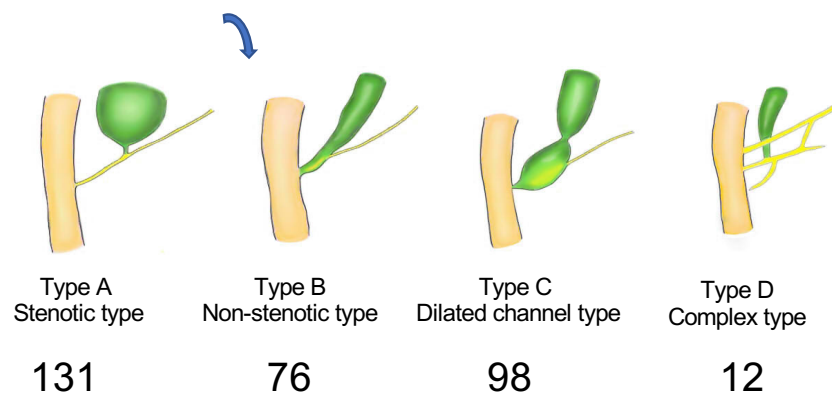


One-year-old girl:
Annular pancreas and
intraluminal duodenal diverticulum

Four-year-old boy:
Annular pancreas

JSGPBM classification reflects clinical characteristics in children

317 pediatric cases



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

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.

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

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?

First version, September 2022

2nd version, September 2024

Imaging features of congenital biliary dilatation

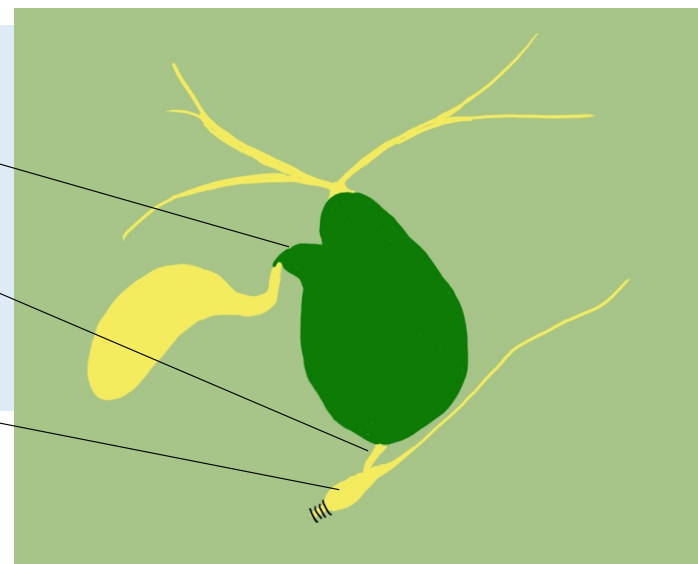
Features that define CBD

- 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 JSGBPBM
See P35–38



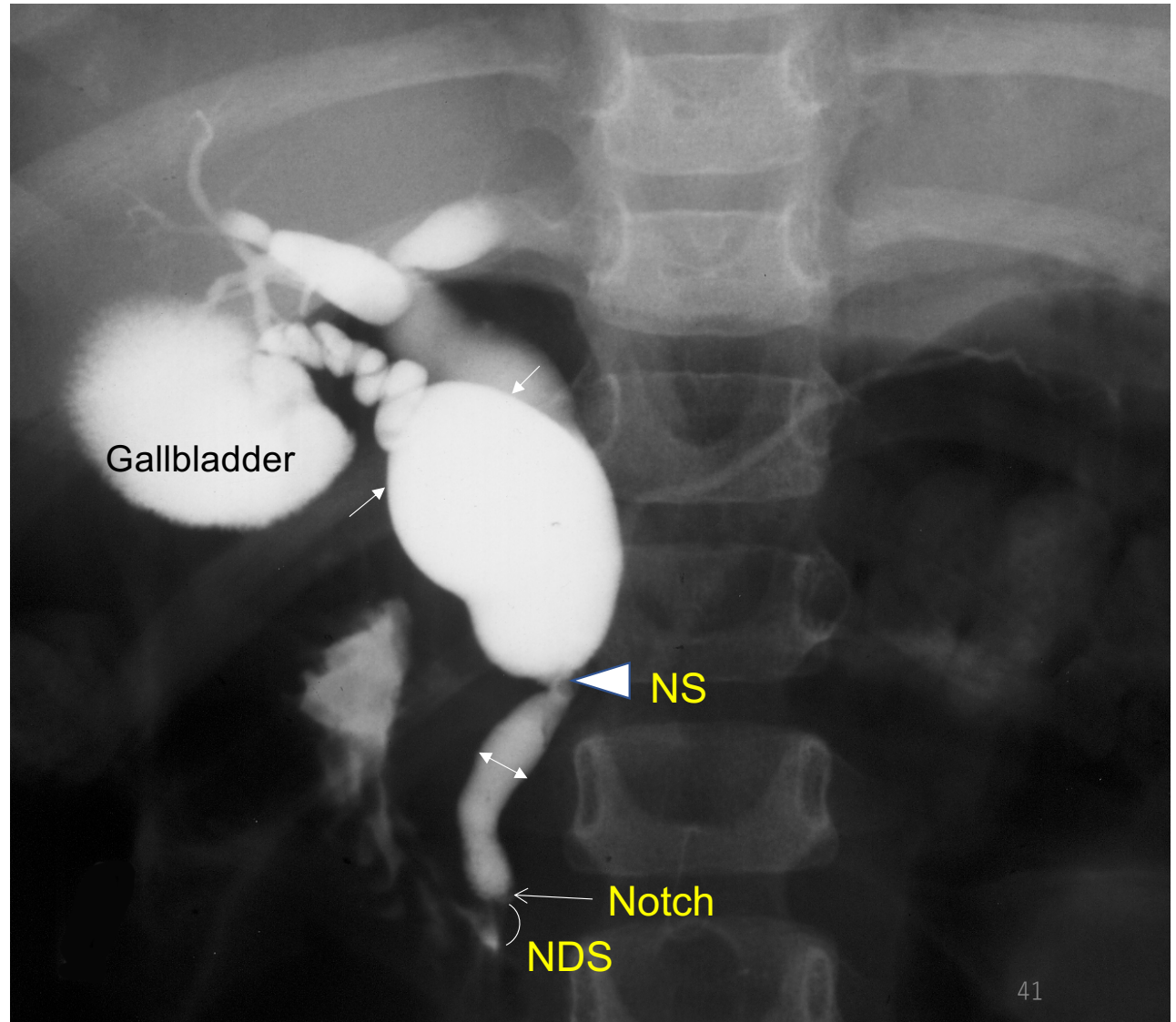
Case 7

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

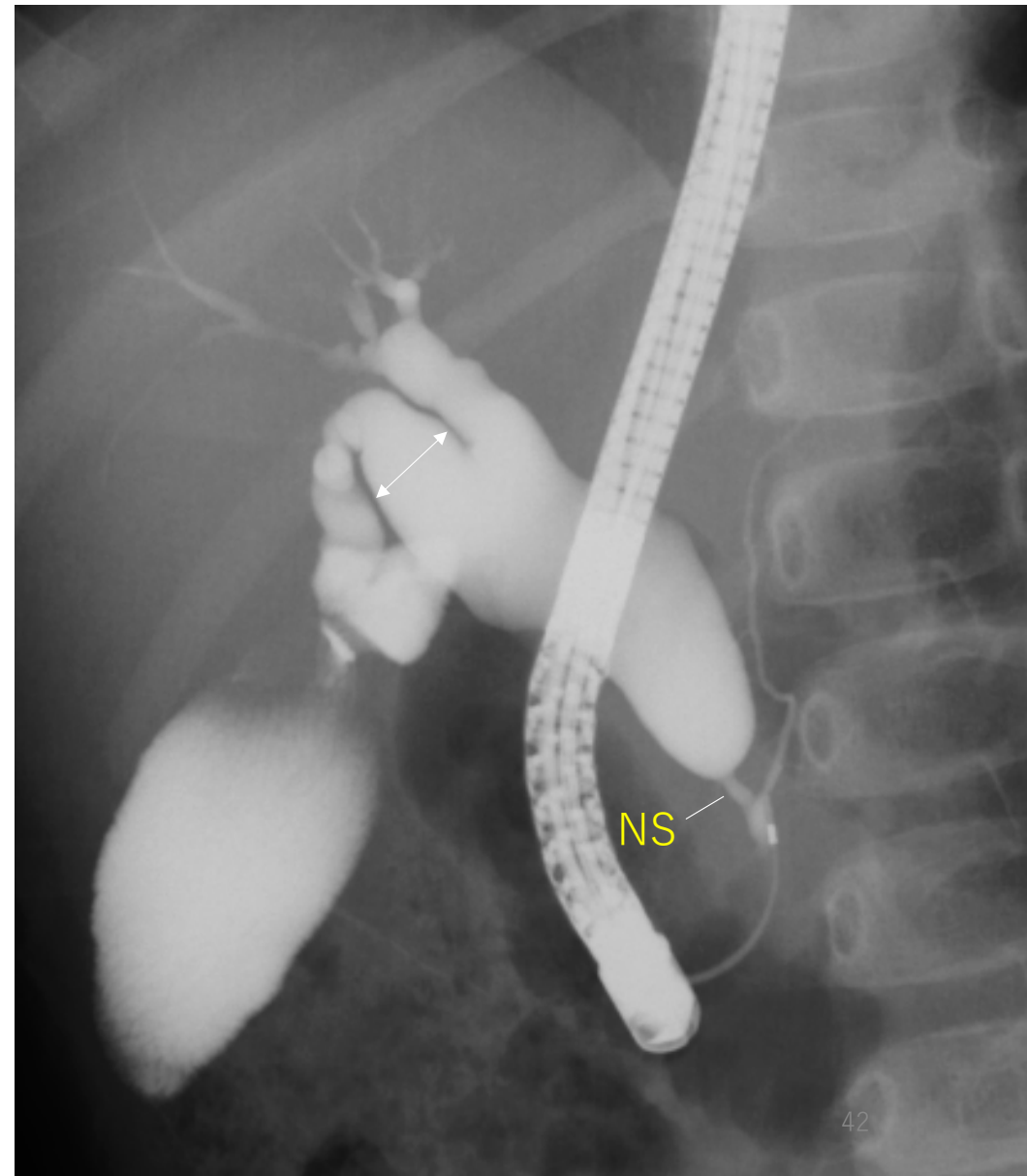


Case 8

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 PD

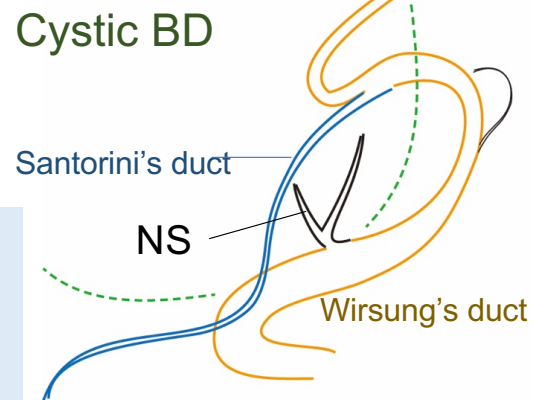
Histopathological demonstration

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
levels of pancreatic
enzymes

Radiological demonstration

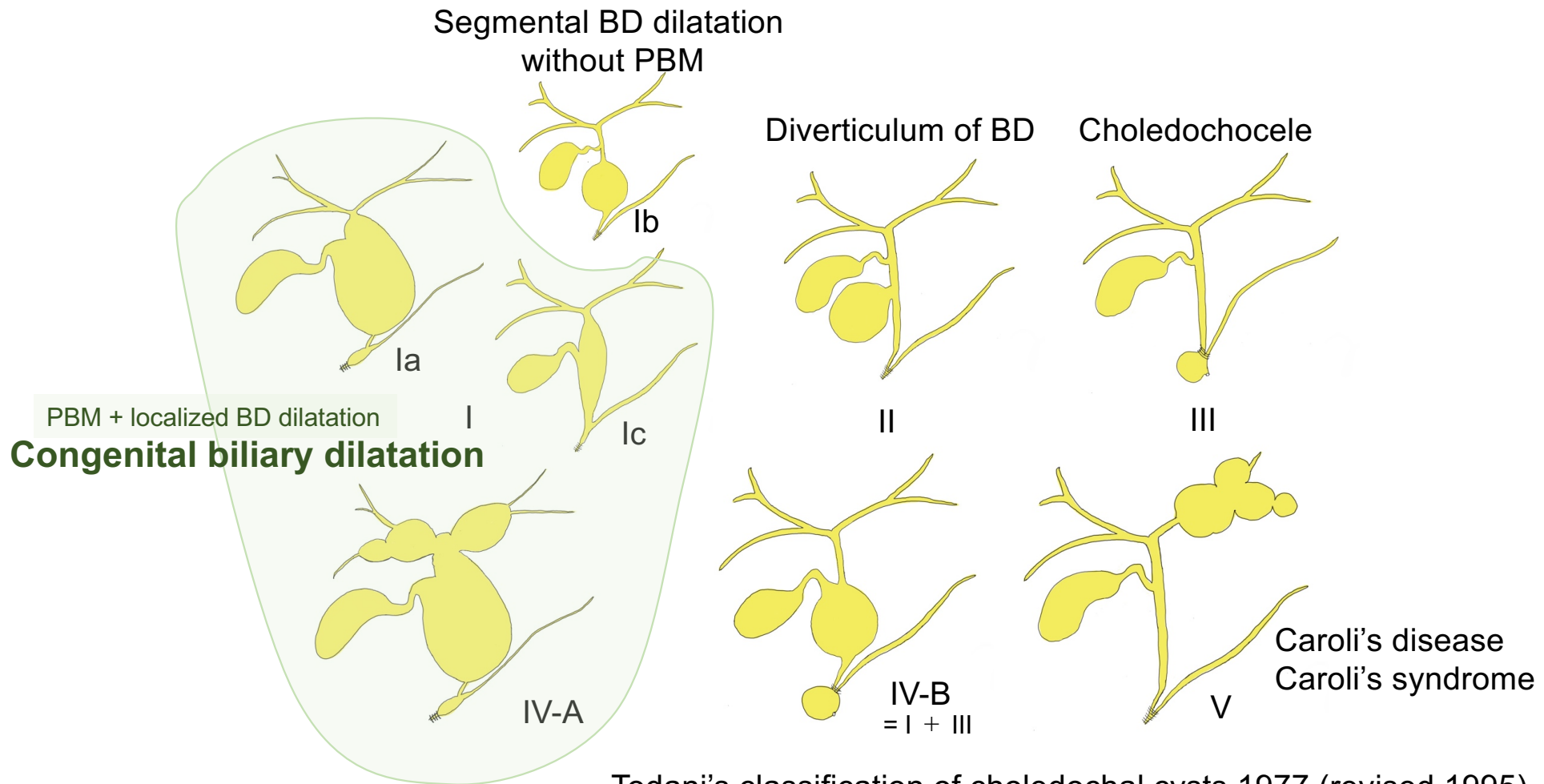


NS is one branch
of PD



Matsumoto's theory of PBM embryogenesis:
PBM is a result of BD misconnecting with
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.

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

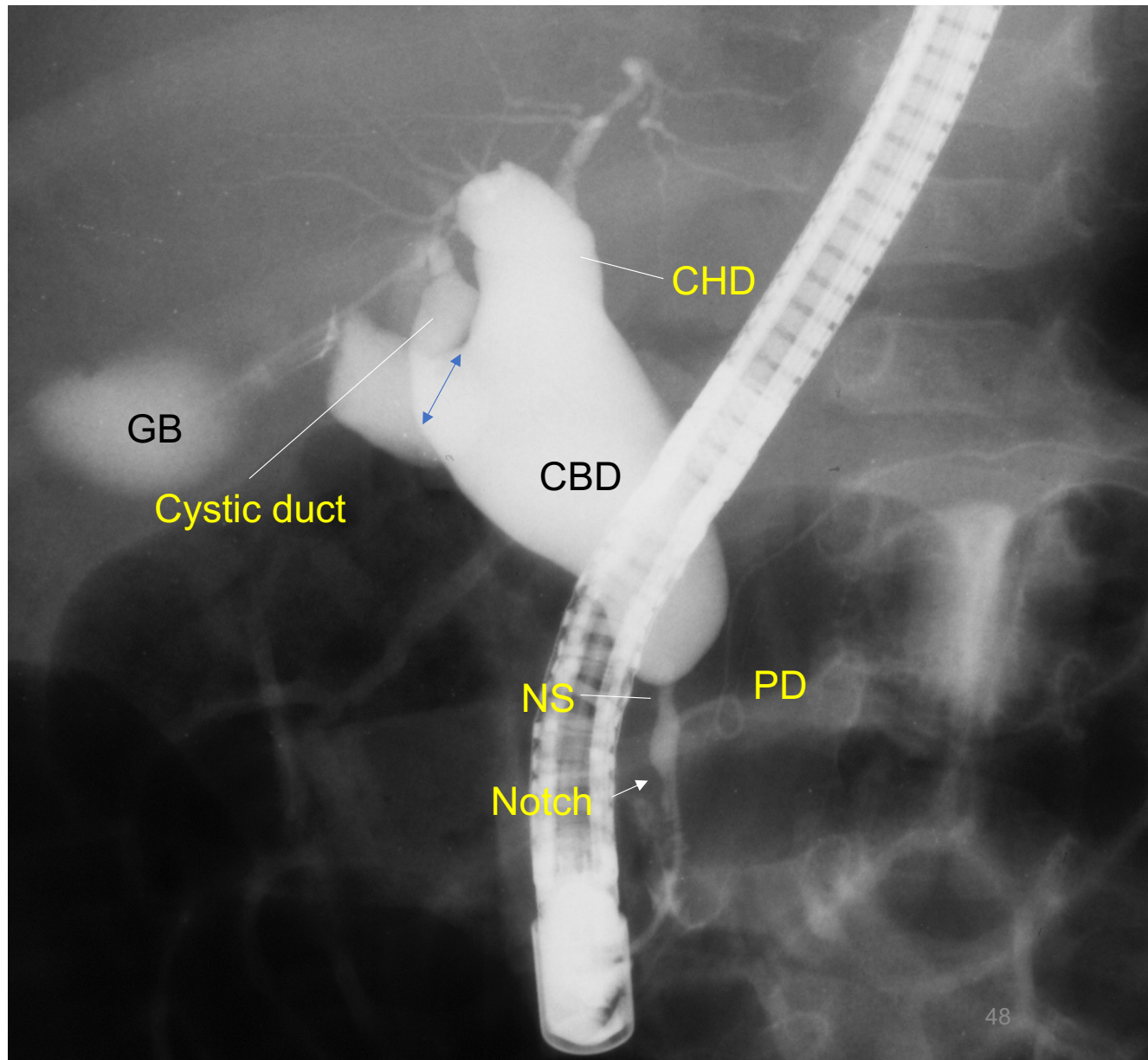
Typical Congenital Biliary Dilatation

First version, September 2022

Case 10 Typical Ia: Pediatric case

ERCP in a 1-year-old 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

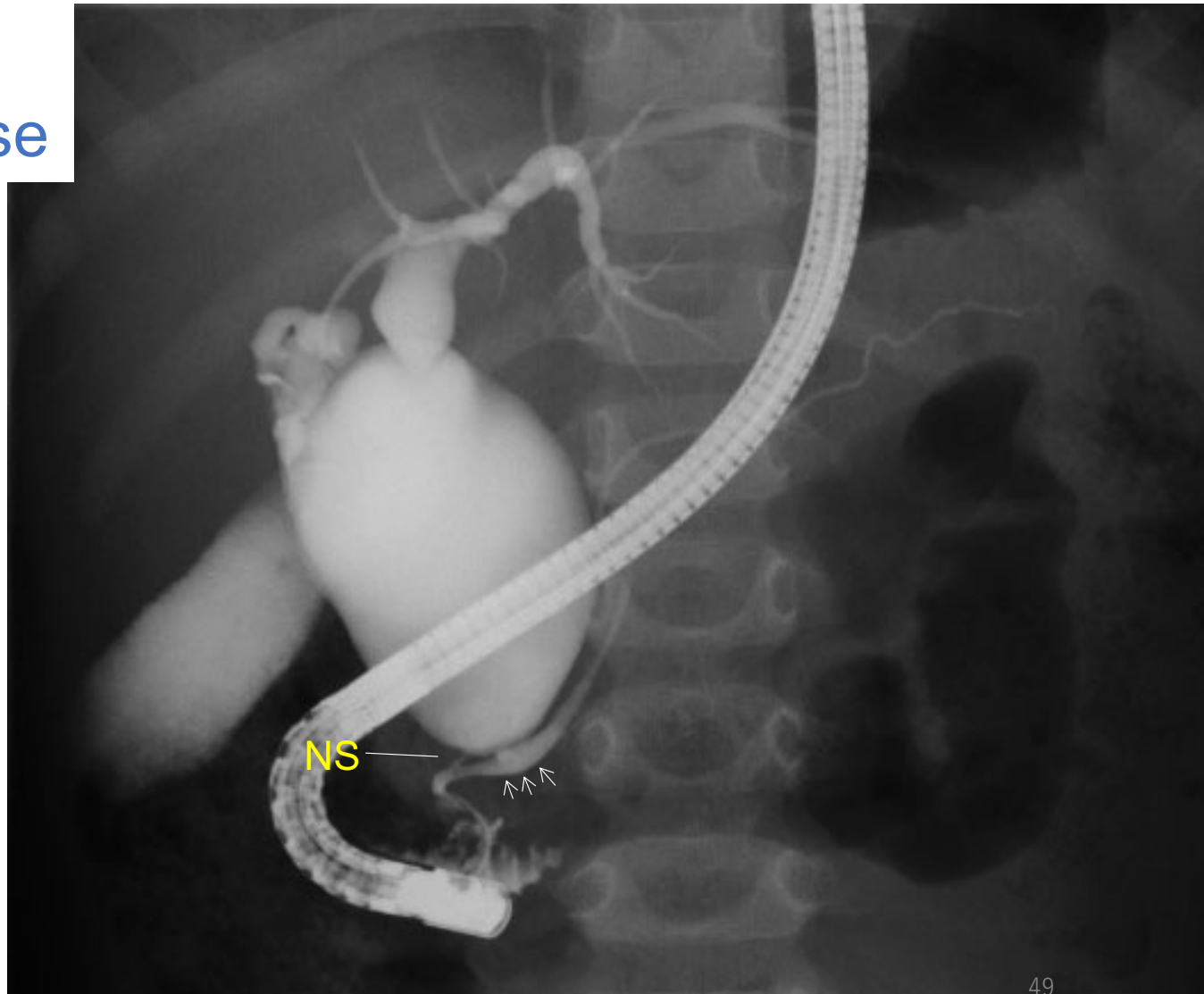


Case 11

Typical Ia: Pediatric case

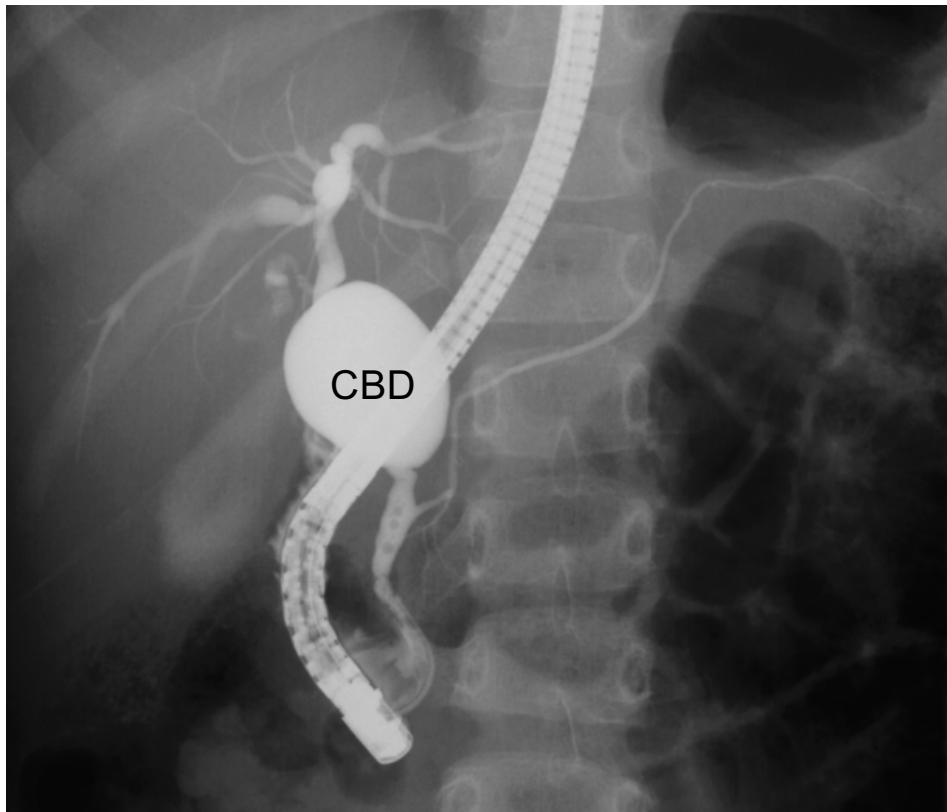
ERCP in a 3-year-old girl
Abdominal pain, vomiting, and elevated serum transaminase levels

- **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

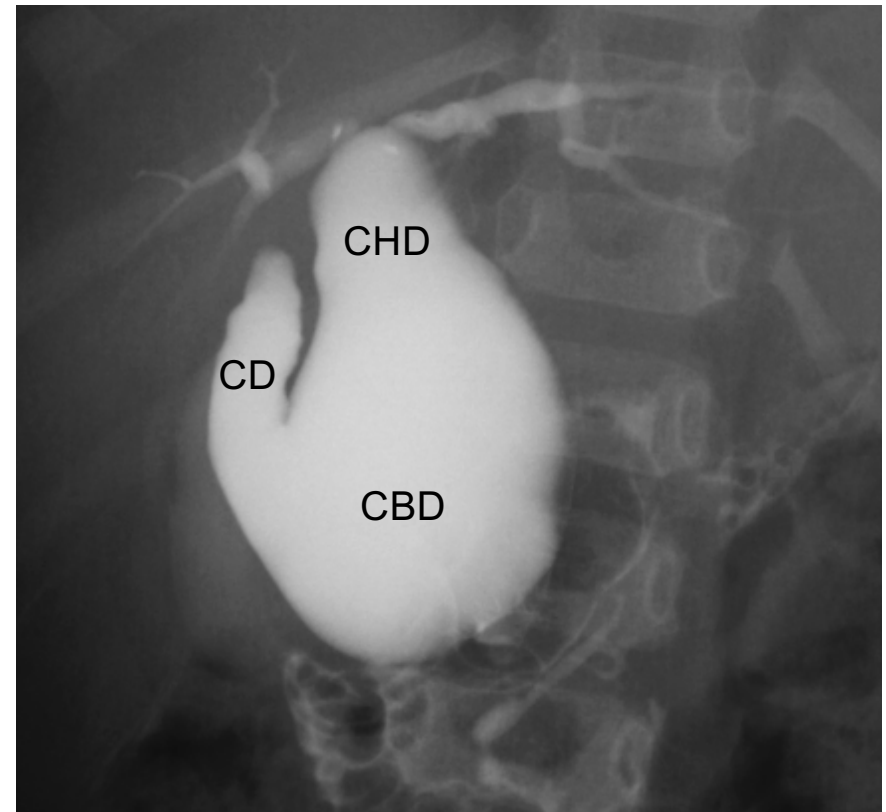


Type Ia: Pediatric case

Localized dilatation to various extents



Dilatation limited to the common bile duct (CBD)



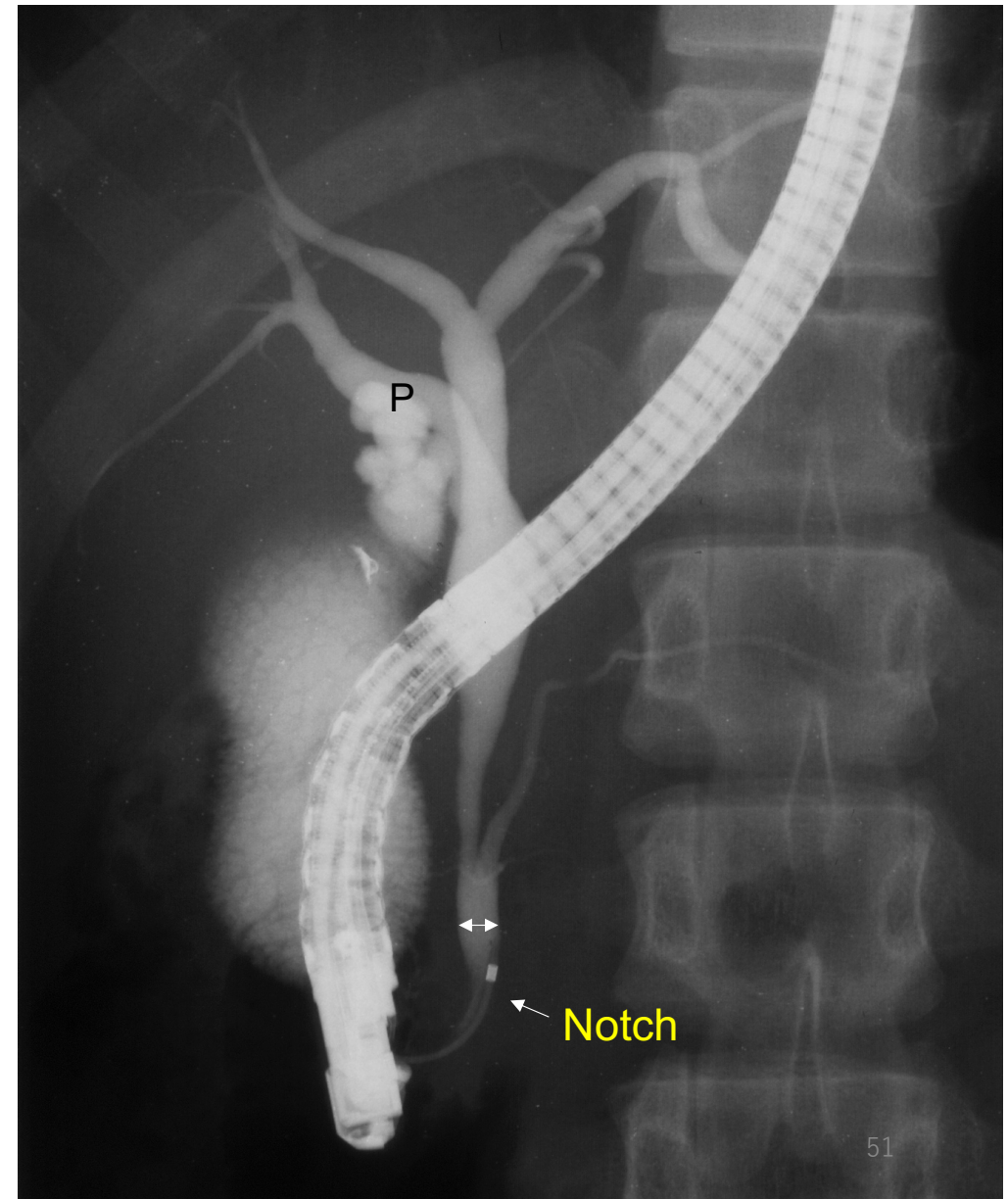
Dilatation involving CBD, CD, and CHD

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** ($\leftrightarrow$)
- Existence of NS is equivocal
- Type C, dilated channel type

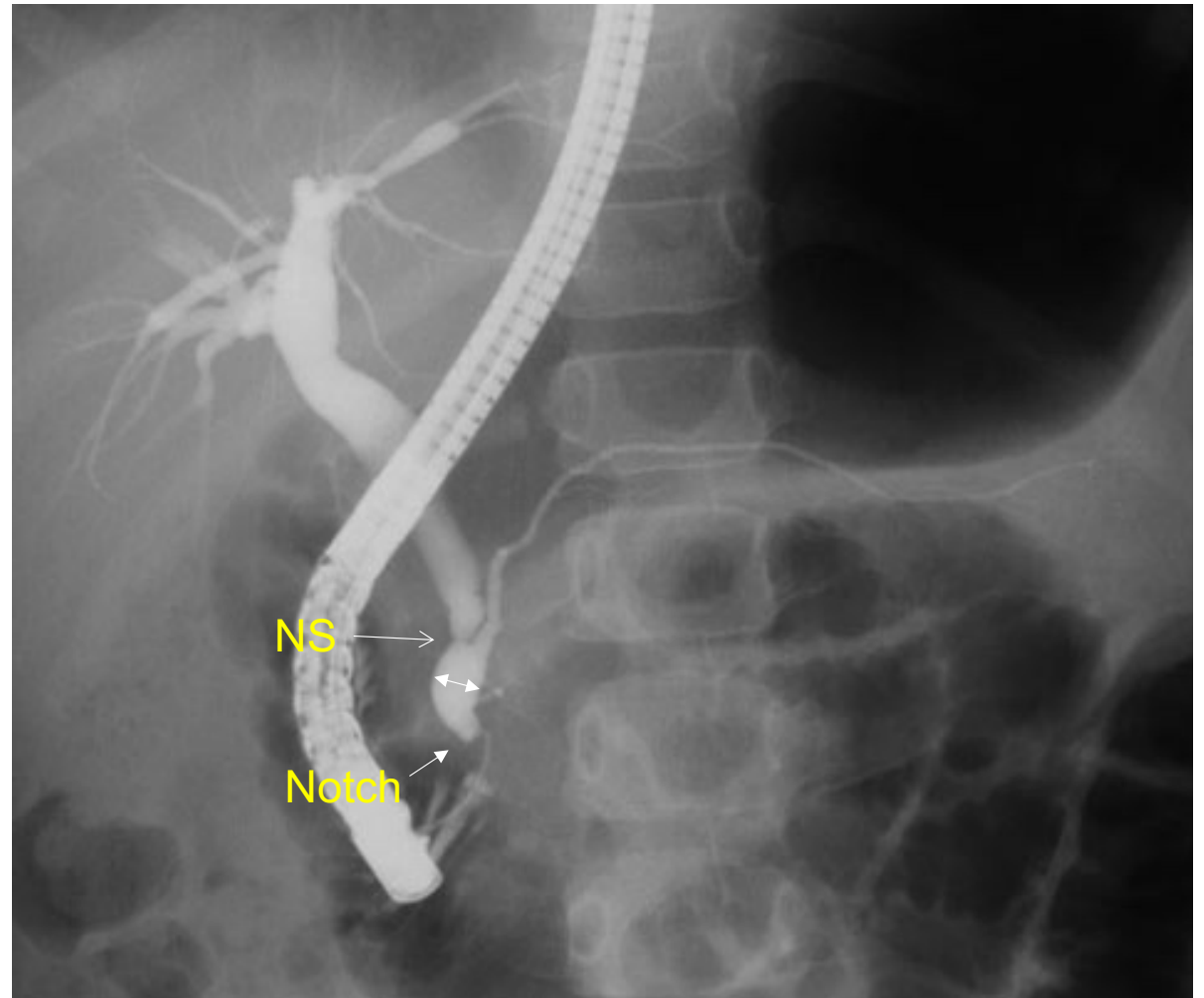


Case 13

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** ($\leftrightarrow$)
- Type C, dilated channel type

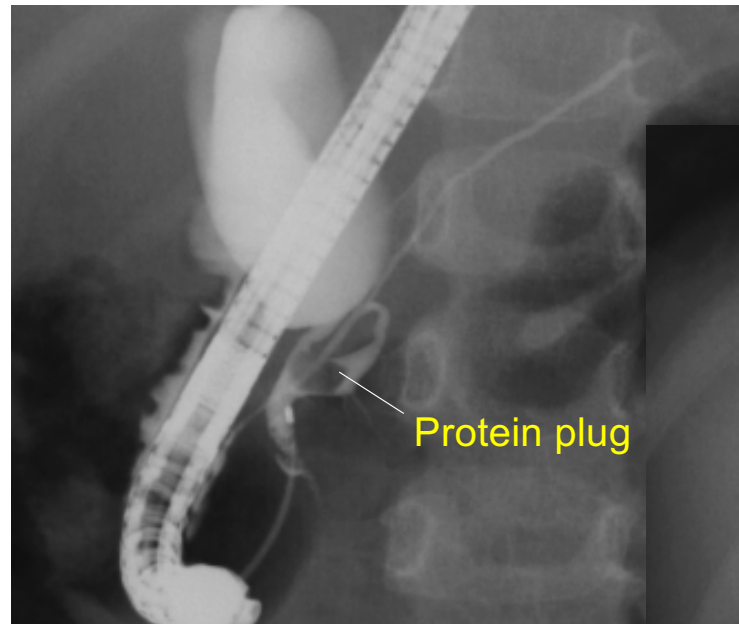


Case 15

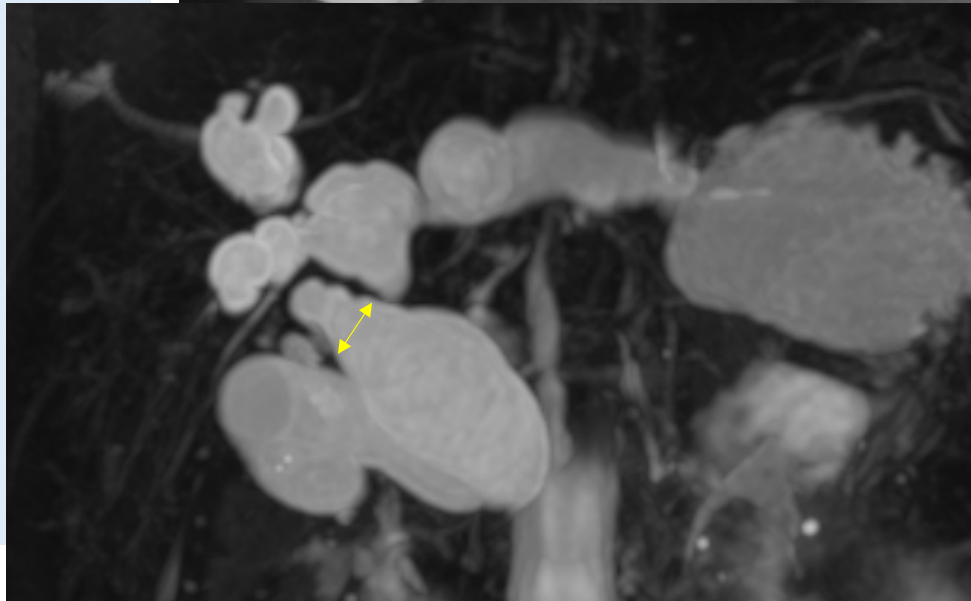
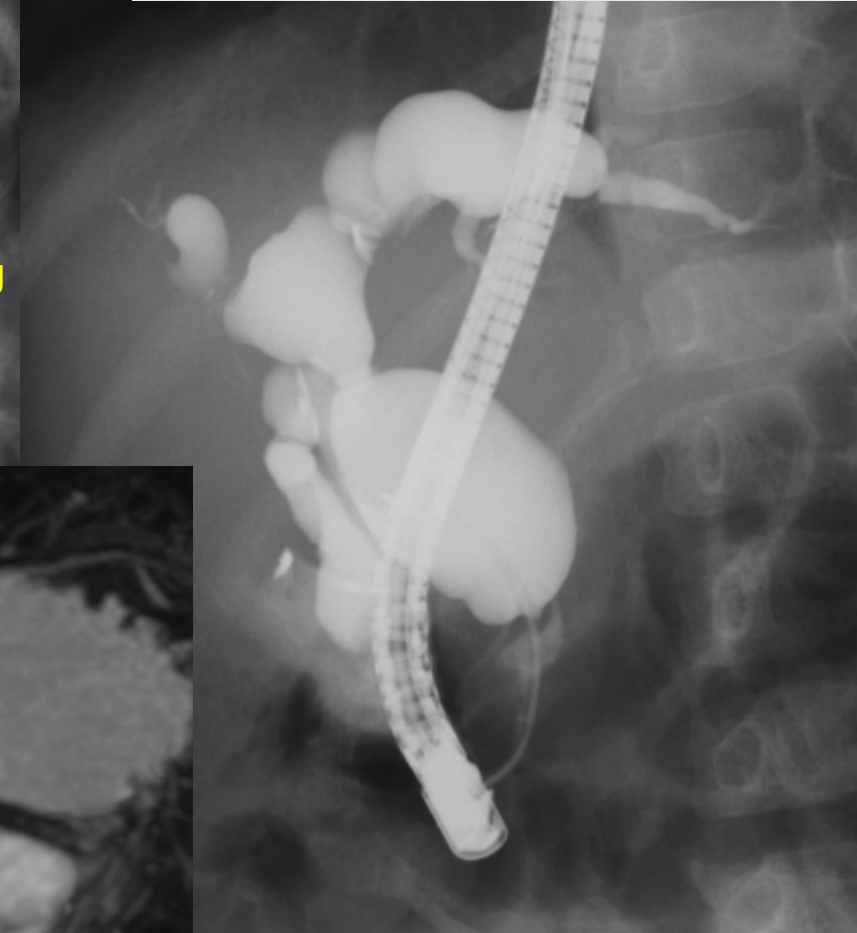
Typical VI-A: Cystic–cystic type pediatric case

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

See P85.



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



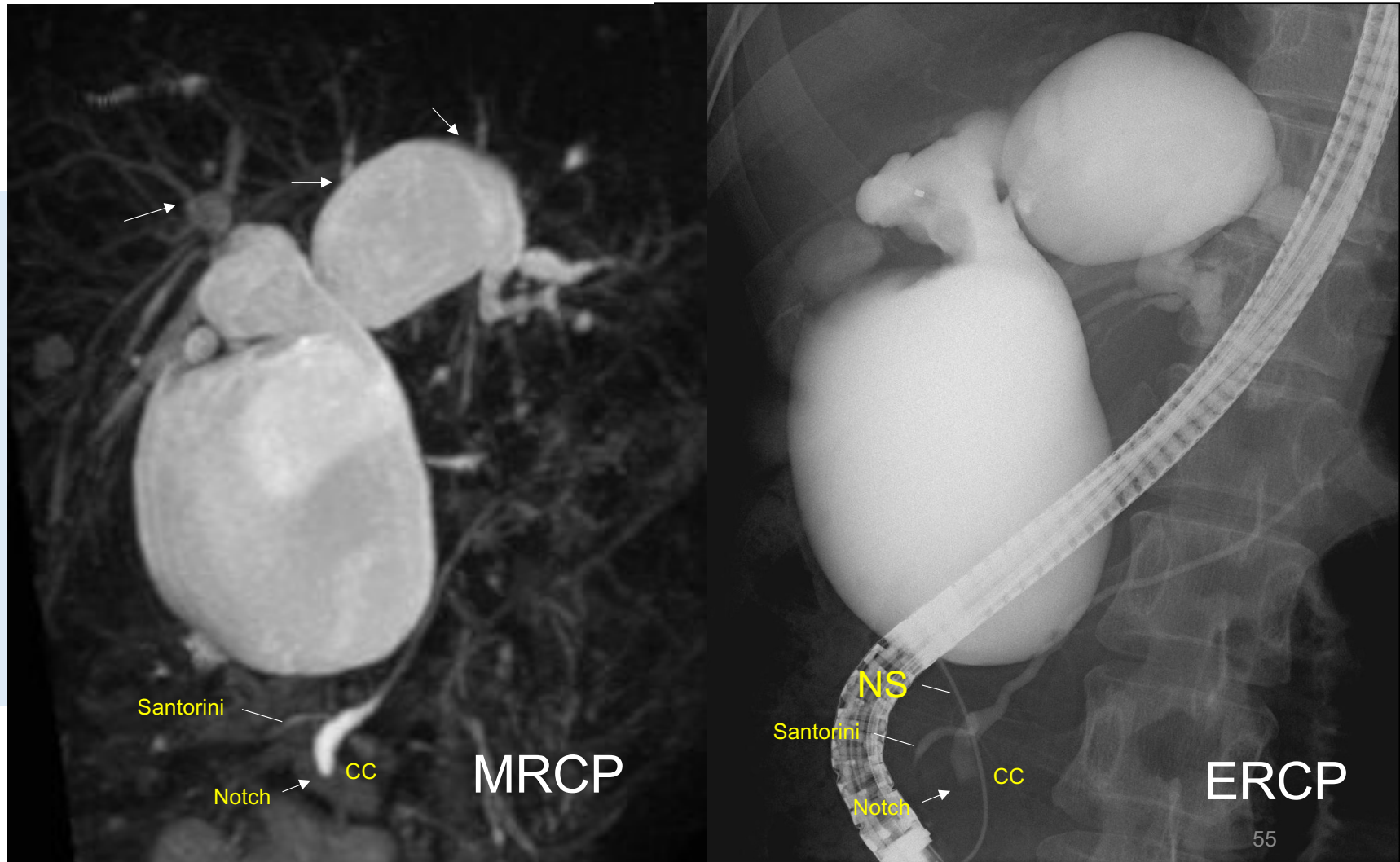
Case 16

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

Female in her thirties
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

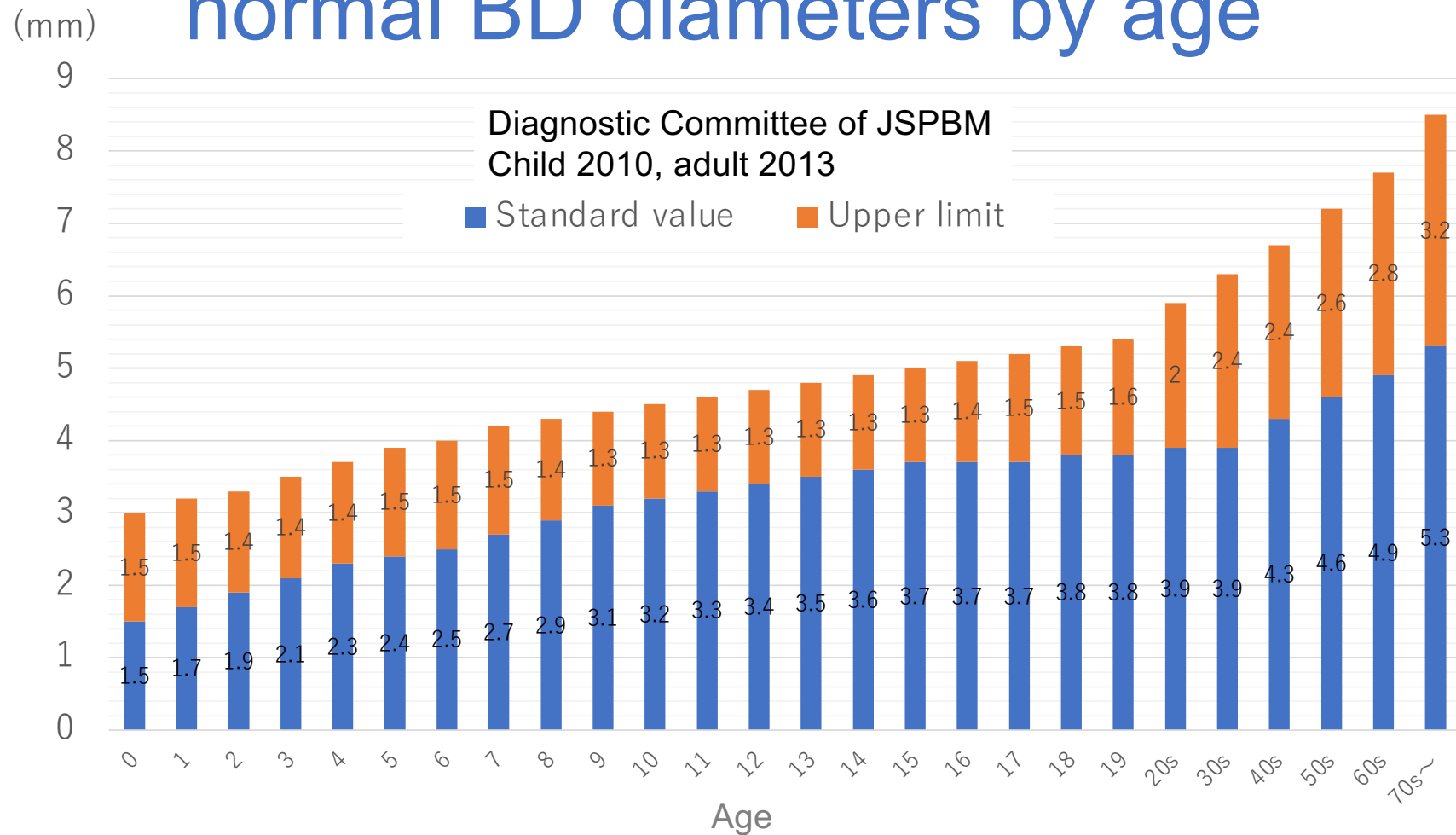
2nd version, September 2024

What is non-dilated type of PBM?

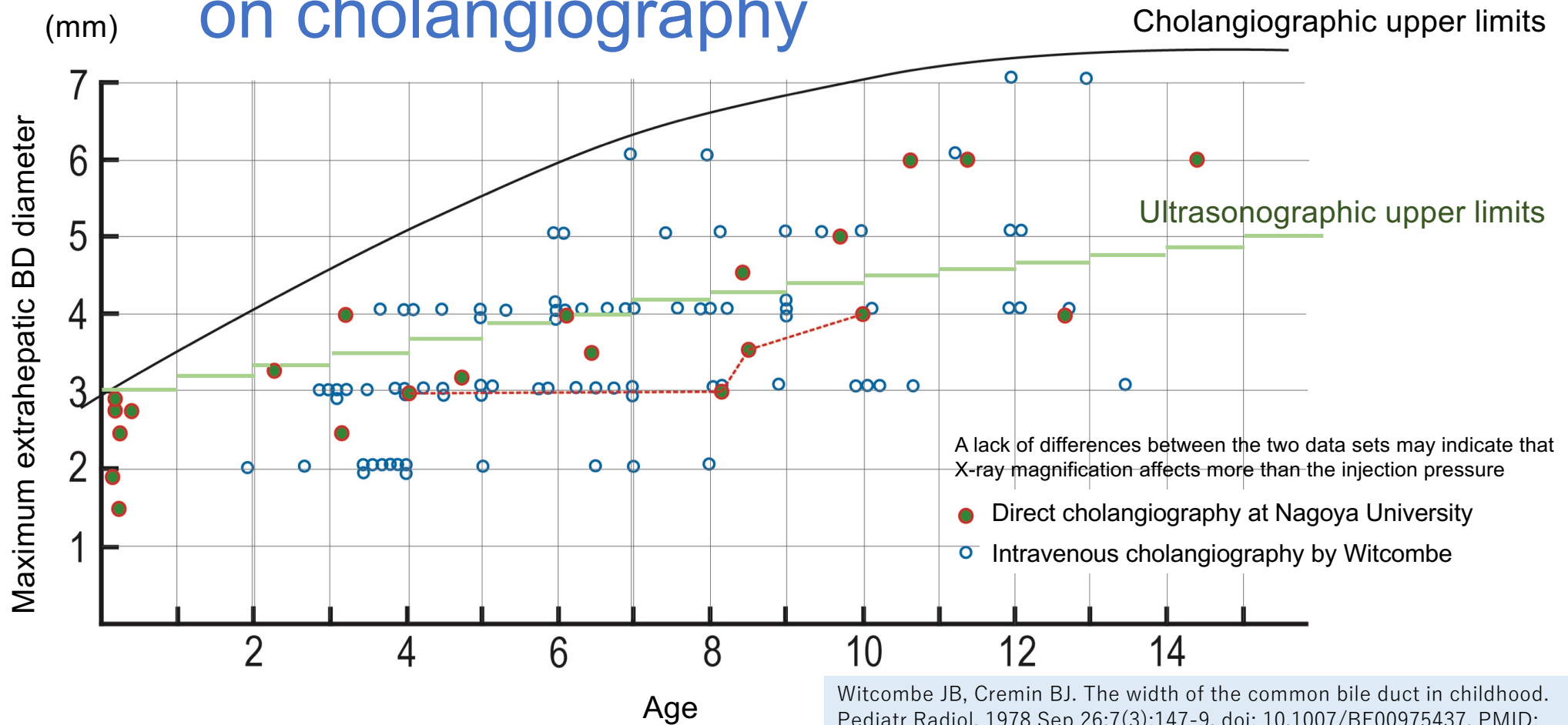
- Definition has not yet been determined.
- BD diameter **increases with age** in both children and adults. Ultrasonographic upper limits of normal BDs by age are determined.
- There is little data of normal BD diameters on direct cholangiography* or MRCP** at present.
- Non-dilated type should have 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



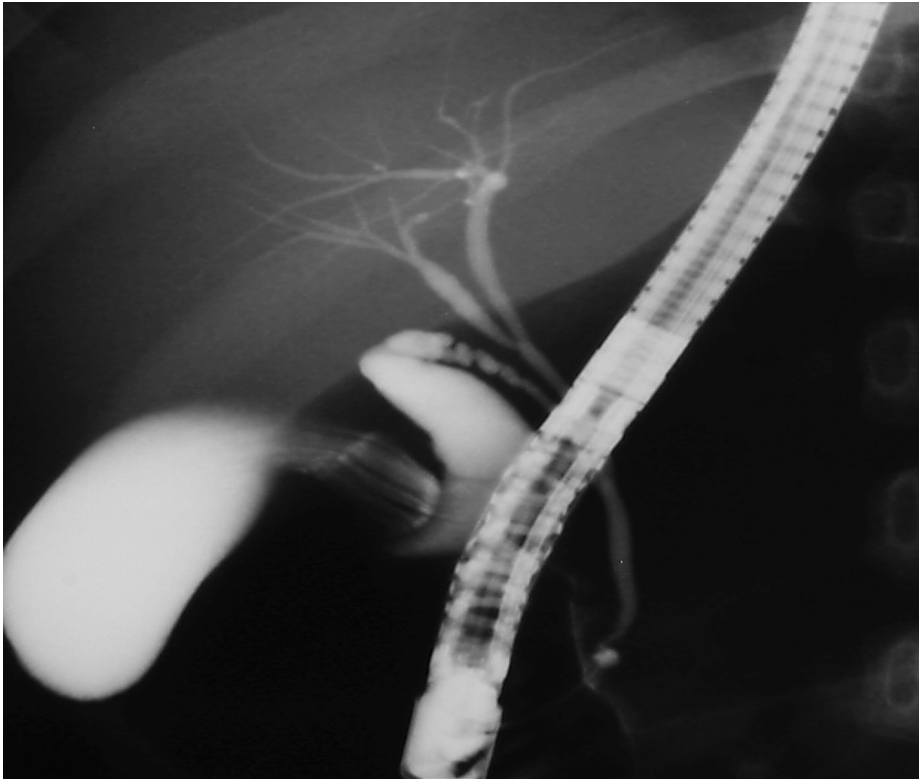
Normal pediatric BD diameters on cholangiography



Witcombe JB, Cremin BJ. The width of the common bile duct in childhood. *Pediatr Radiol*. 1978 Sep 26;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

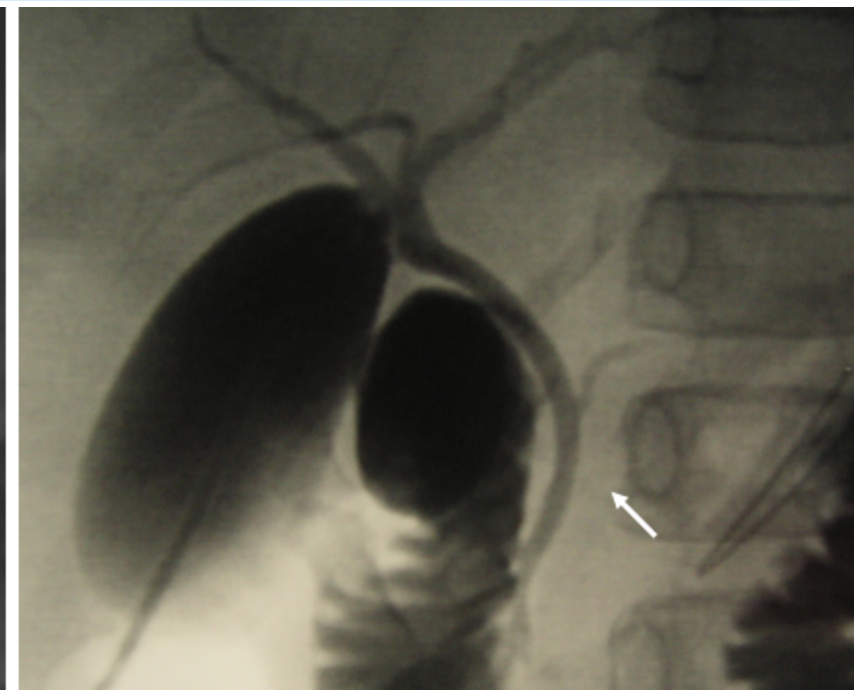
Six-year-old girl
Abdominal pain, vomiting,
hyperamylasemia

- **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

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.



MRCP



On-table cholangiography

NS, dilated CC, dilated CD

See P40.

Case 19

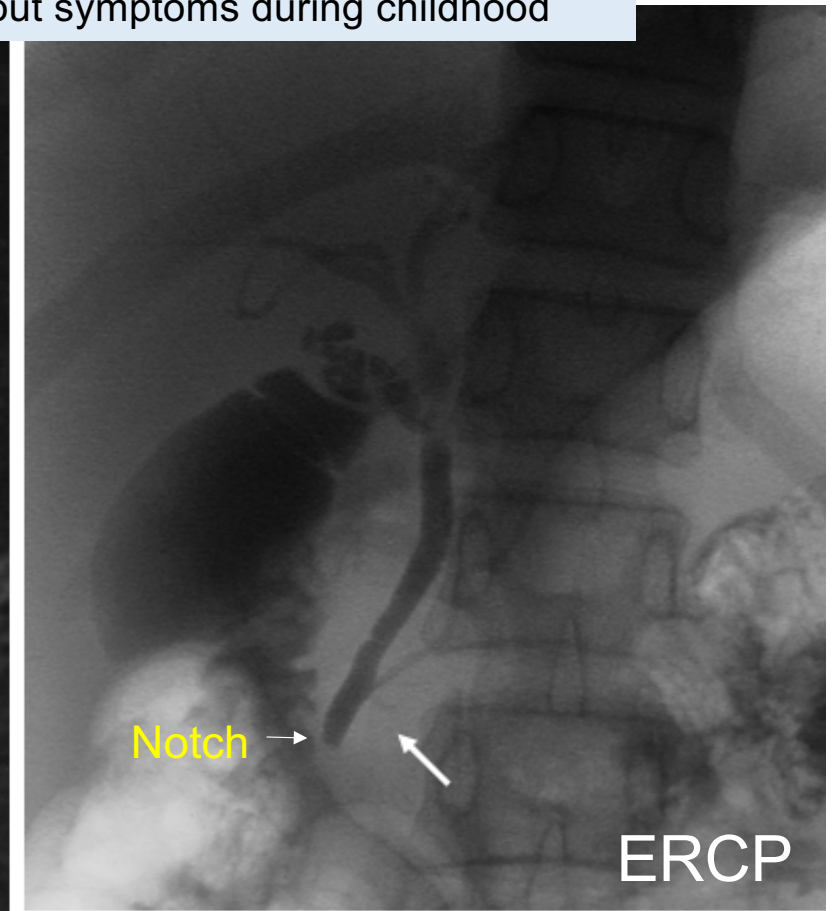
Typical non-dilated type of PBM: Pediatric case

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

Symptoms are the same as those of congenital biliary dilatation

Twelve-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

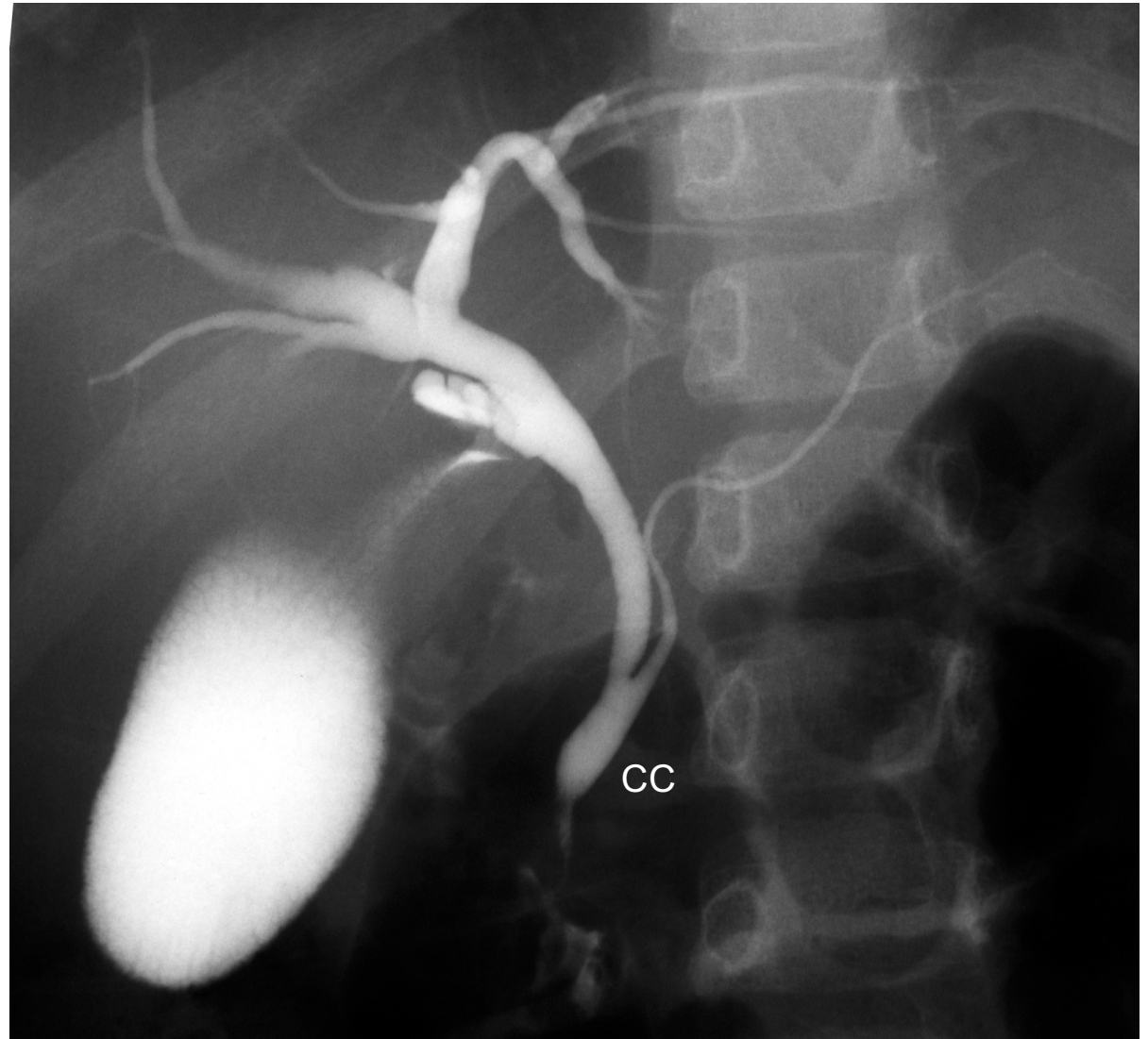
Non-dilated type?

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

Gallbladder cancer often arises
in adult non-dilated type

MRCP in a female in her fifties
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

Non-dilated type is often found without symptoms in adults



Case 6

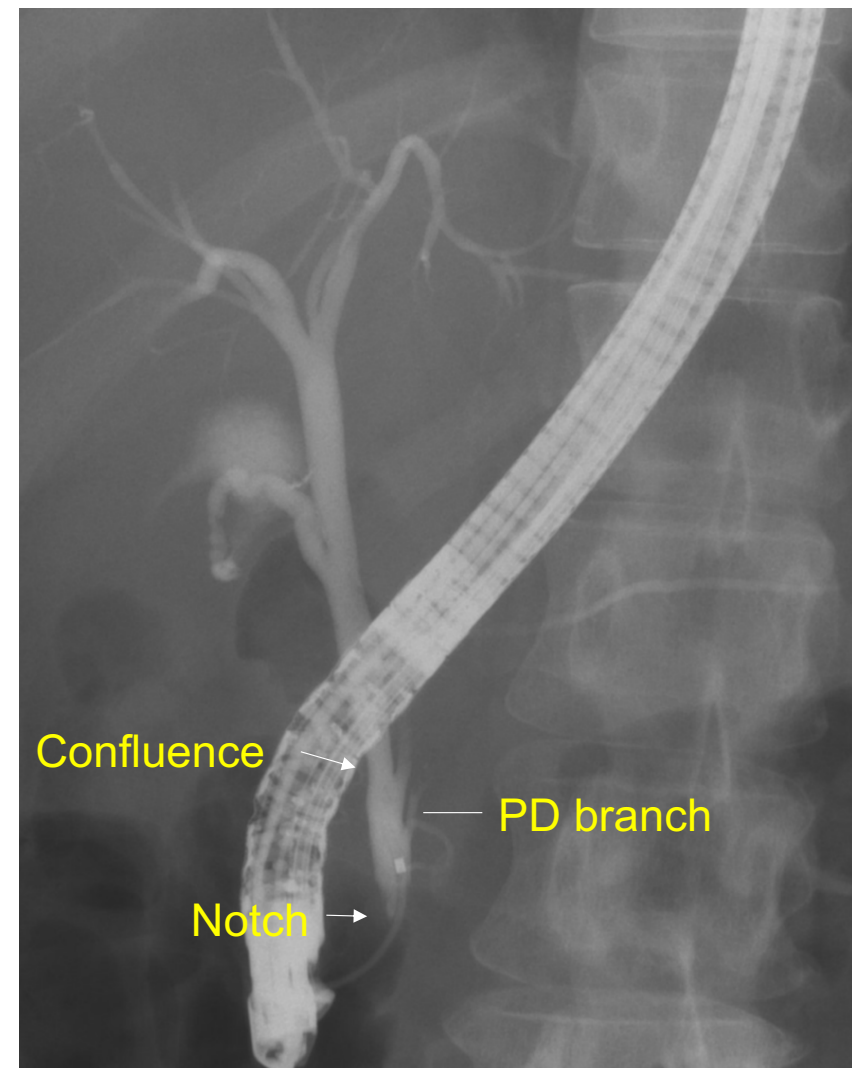
Typical non-dilated PBM: Adult case

See P30.

ERCP in a female in her forties
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.

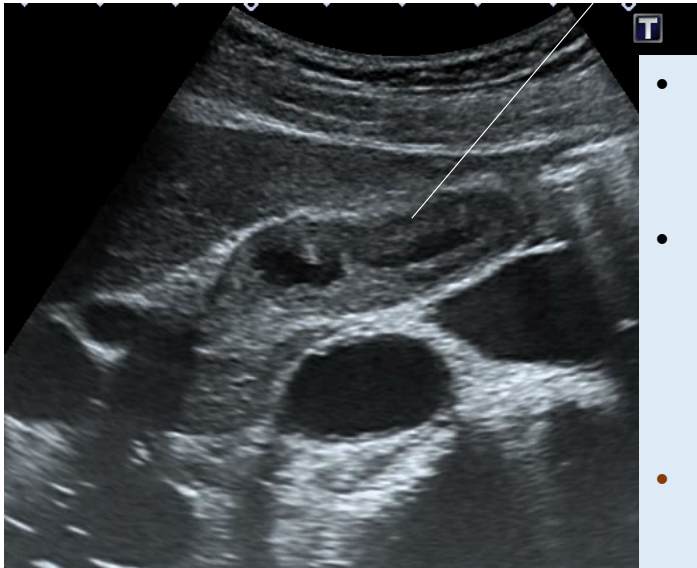


Case 22

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

A female in her fifties

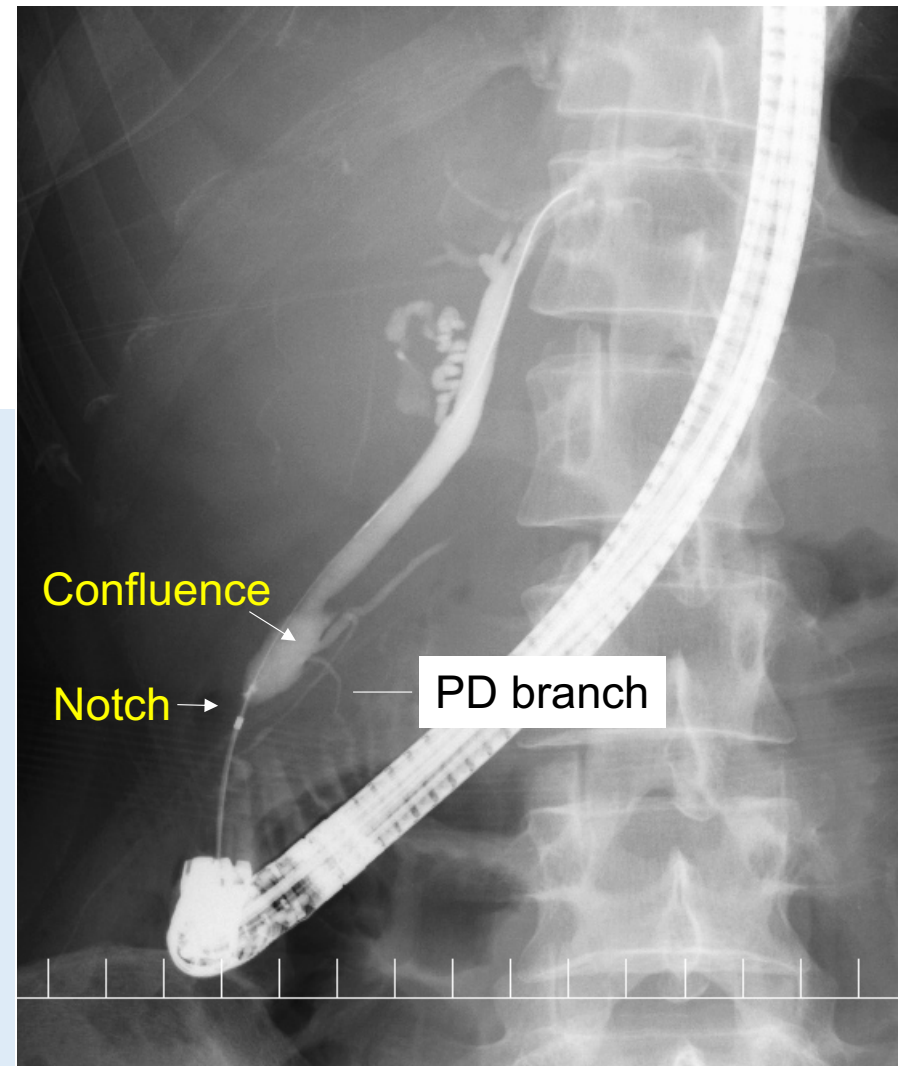
Screening US found GB wall thickness



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

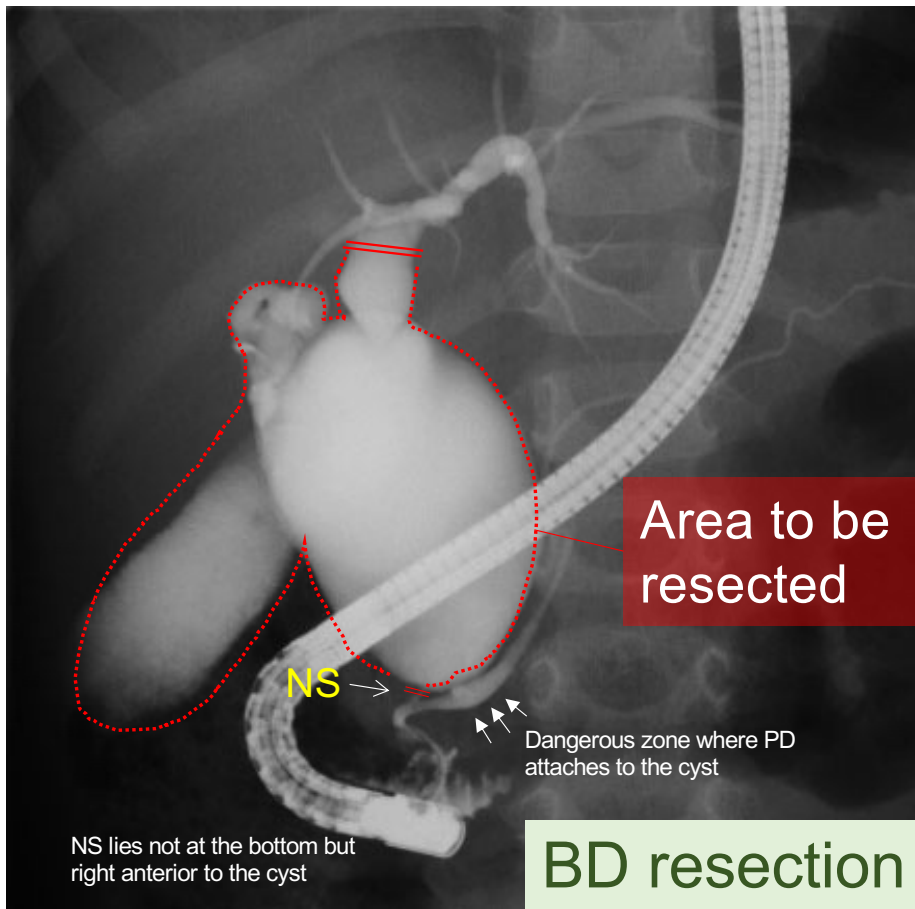
Treatment for pancreaticobiliary maljunction

- Reasons for treatment necessity
- PBM causes:
 - ✓ Recurrent **symptoms** including abdominal pain, vomiting, and jaundice in children
 - ✓ High occurrence of **biliary cancers** in adults see P14.
- **Aim** of treatment = To **prevent symptoms and cancers**
- The following must be undertaken:
 - **Bile duct resection** for congenital biliary dilatation
 - **At least cholecystectomy** for nondilated PBM*

Not merely to mend malformation!

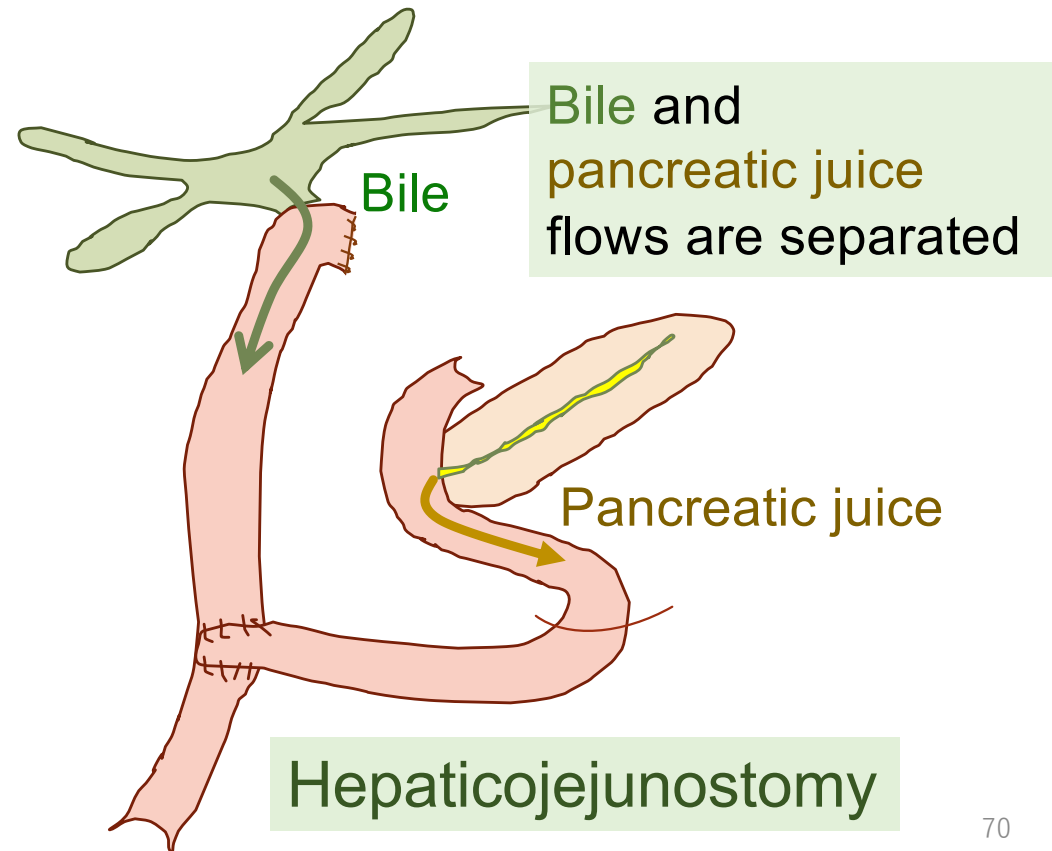
*No consensus on the need for bile duct resection because of little incidence of carcinogenesis at the BD
In pediatric cases, bile duct resection is common because the same symptoms as those of CBD occur due to the same mechanism, i.e. protein plugs.

Surgery for CBD



NS, narrow segment

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



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

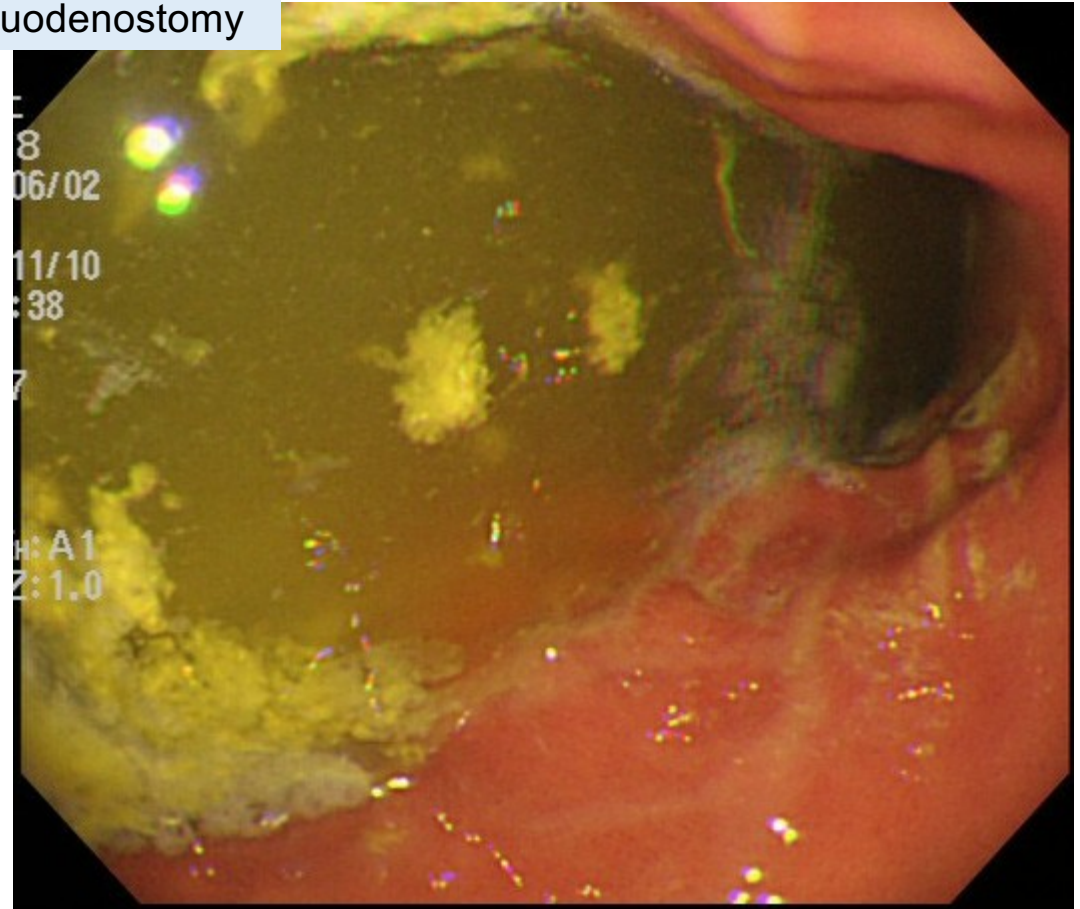
Bile gastritis after hepaticoduodenostomy

One of the reasons not to recommend hepaticoduodenostomy

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

After 7 years, she felt epigastric pain. Endoscopy showed bile regurgitation to the stomach, and redness, edema, and erosion in some areas of gastric mucosa.

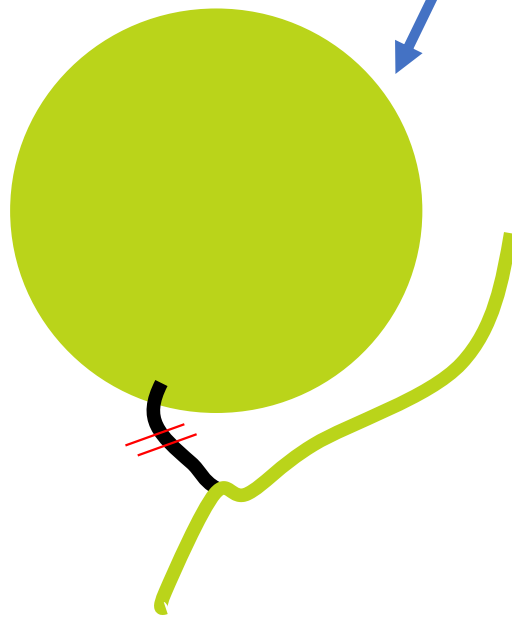
Three months after the examination, she underwent hepaticojejunostomy from duodenostomy, which relieved her epigastric pain and endoscopic abnormality.



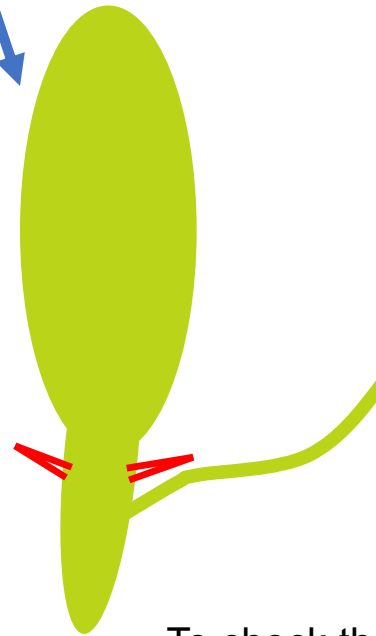
Complete resection of intrapancreatic BD

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

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



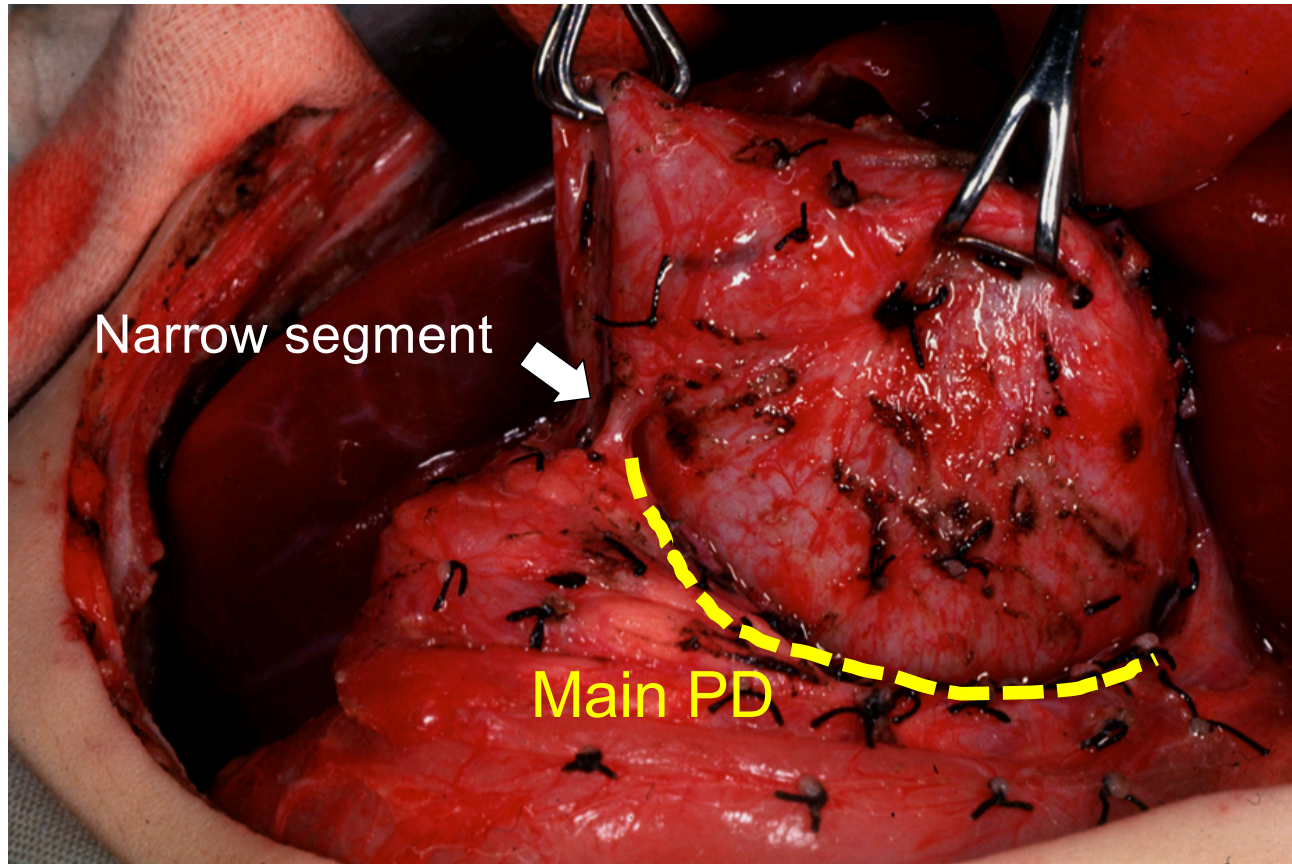
Visual confirmation of narrow segment



To check the distance to PD

Fluoroscopy using clips

Visual confirmation of narrow segment

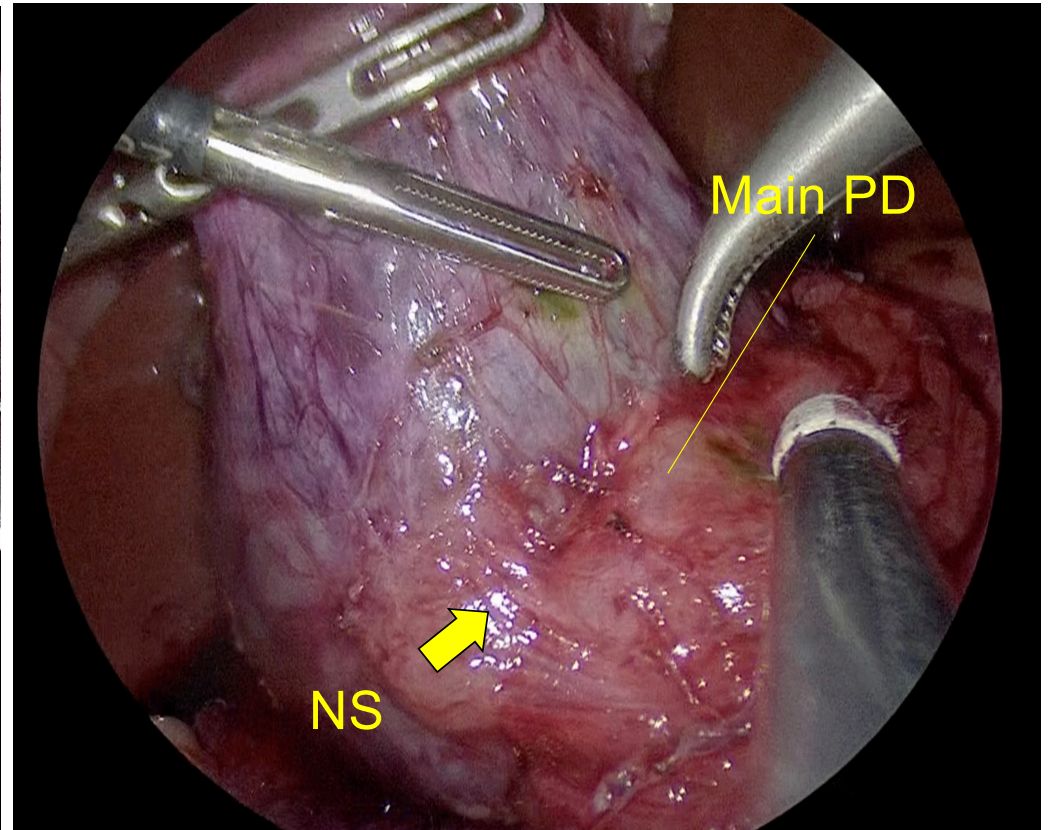
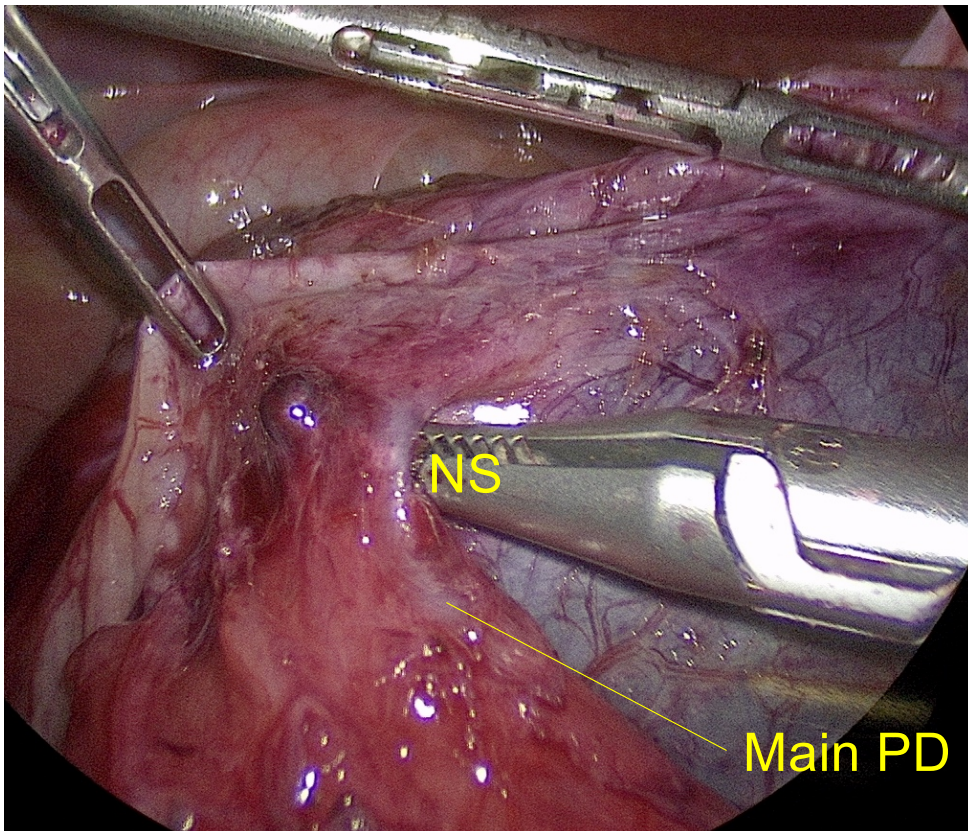


Narrow segment is often located not at the bottom but right-ventral to the cyst.

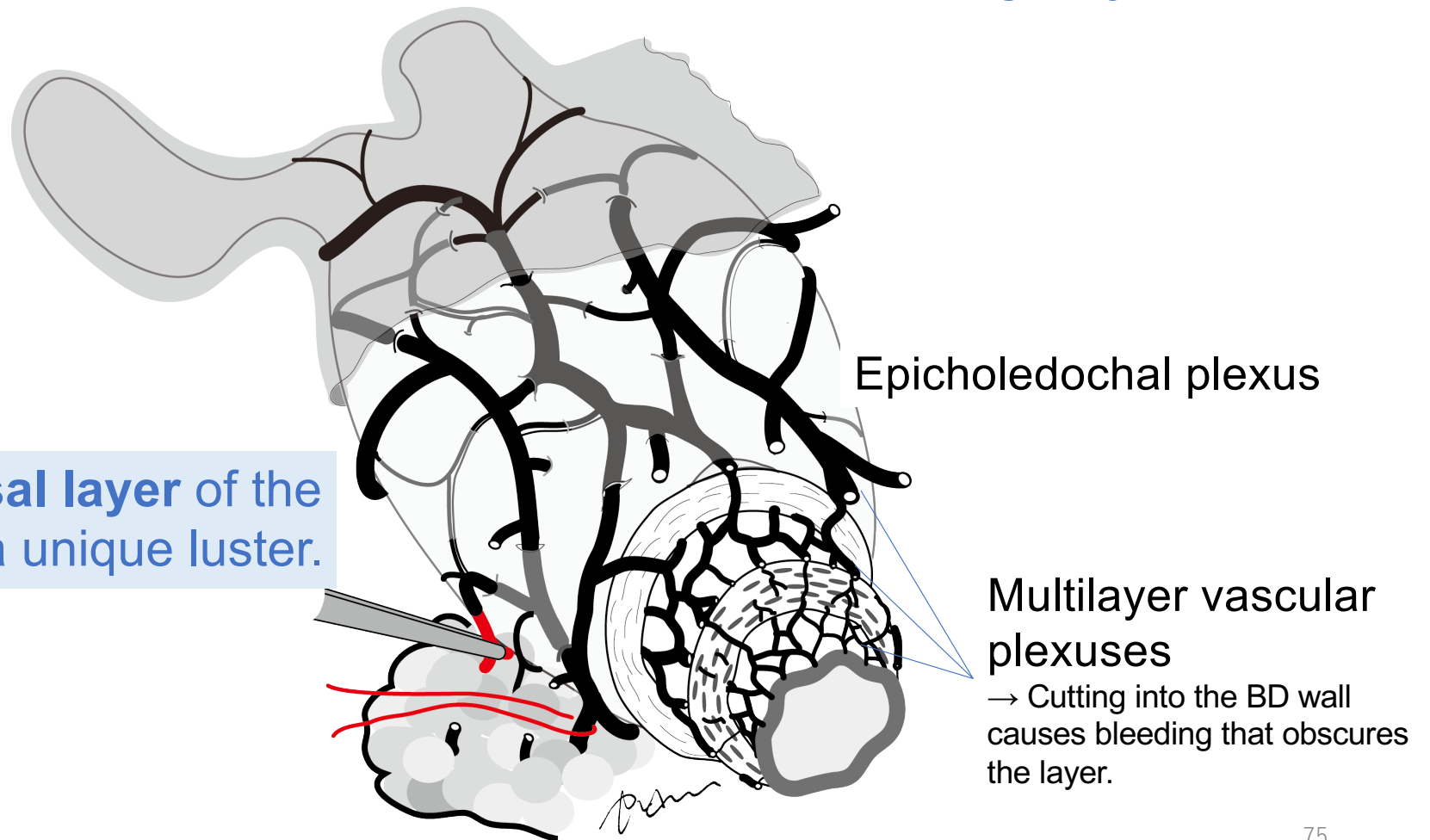
In this case, care must be taken not to injure the PD because the PD runs on the cyst.

PD adheres to the cystically dilated BD

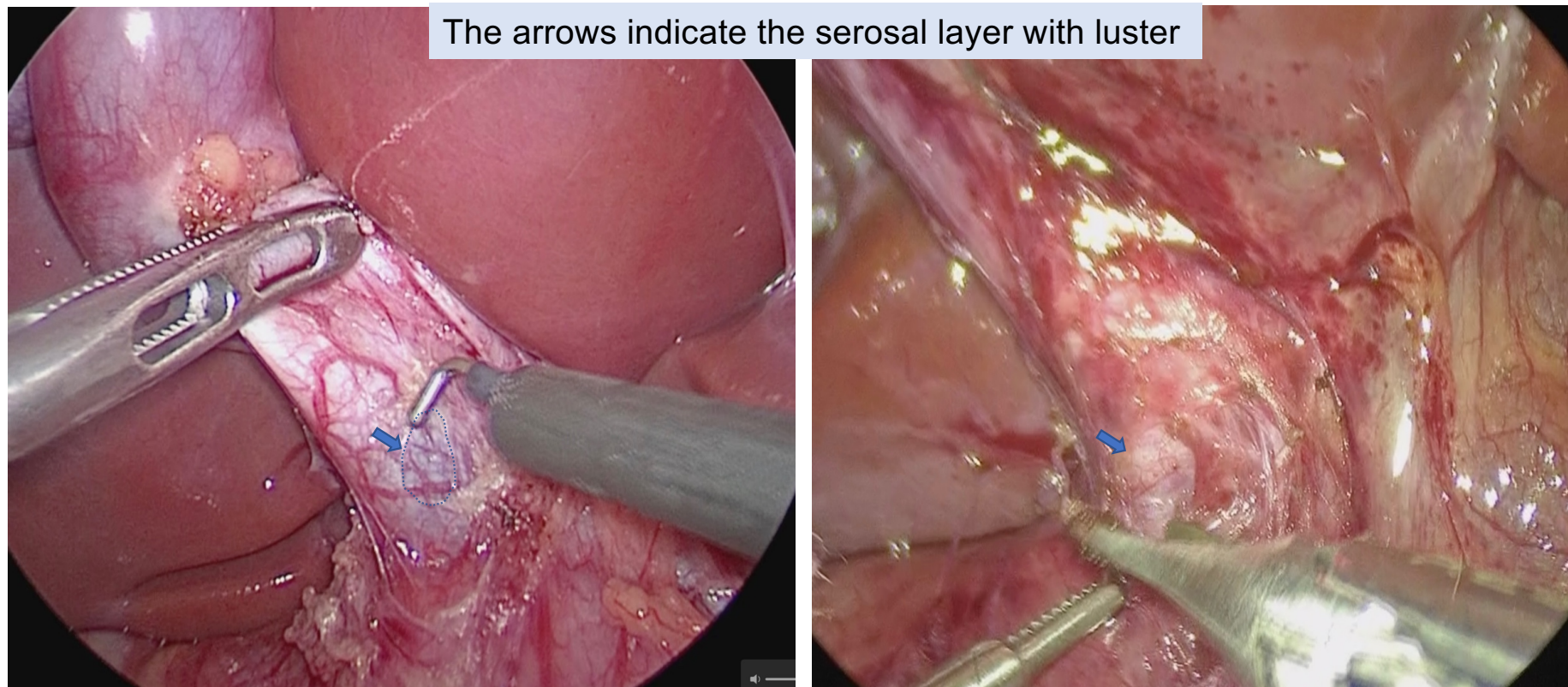
Infant case



How to avoid a pancreatic duct injury



BD serosa has a unique luster

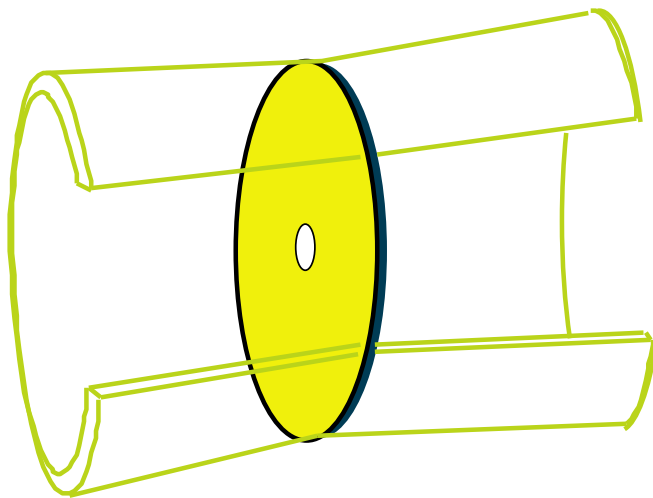


Case without inflammatory adhesion

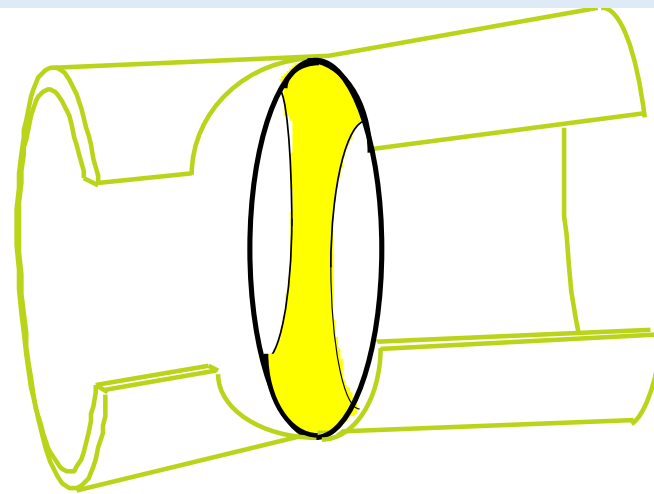
Case with inflammatory adhesion

Two types of congenital BD stenoses around the hepatic hilum

Overlooking these stenoses at BD resection would cause postoperative intrahepatic gall stone formation
Stenoses exist in 85% of IV-A congenital biliary dilatation



Membranous stenosis

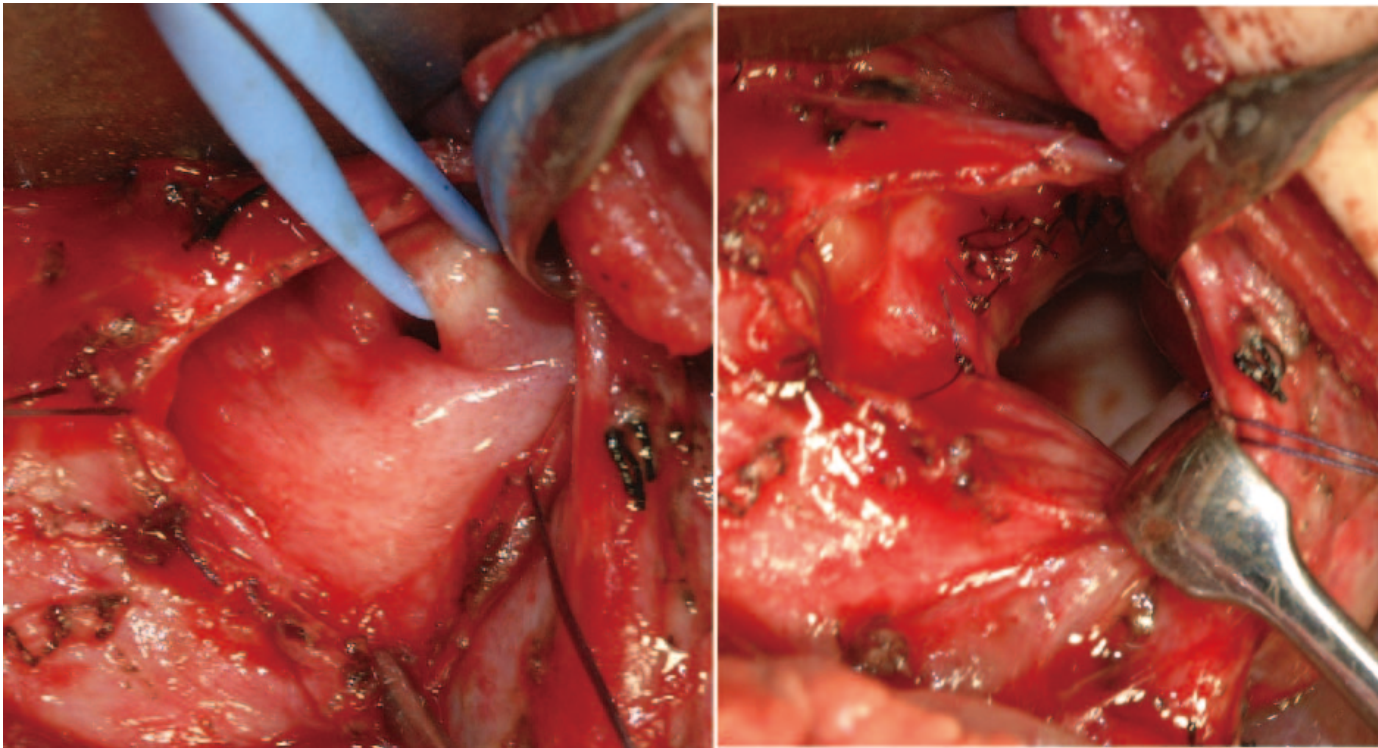


Septal stenosis

Balloon dilatation is not effective for these stenoses.

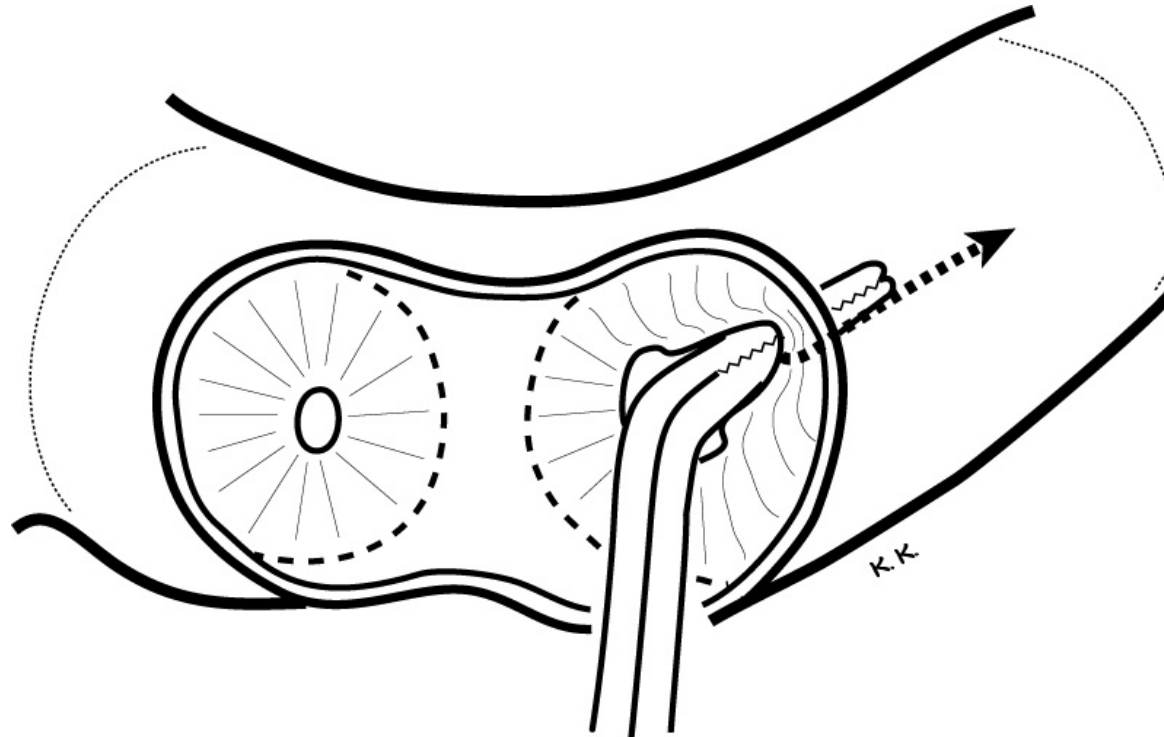
Ando H, Ito T, Kaneko K, Seo T.
Congenital stenosis of the intrahepatic bile duct associated
with choledochal cysts. J Am Coll Surg. 1995
Nov;181(5):426-30. PMID: 7582210.

Resection of septal stenosis



At the time of BD resection

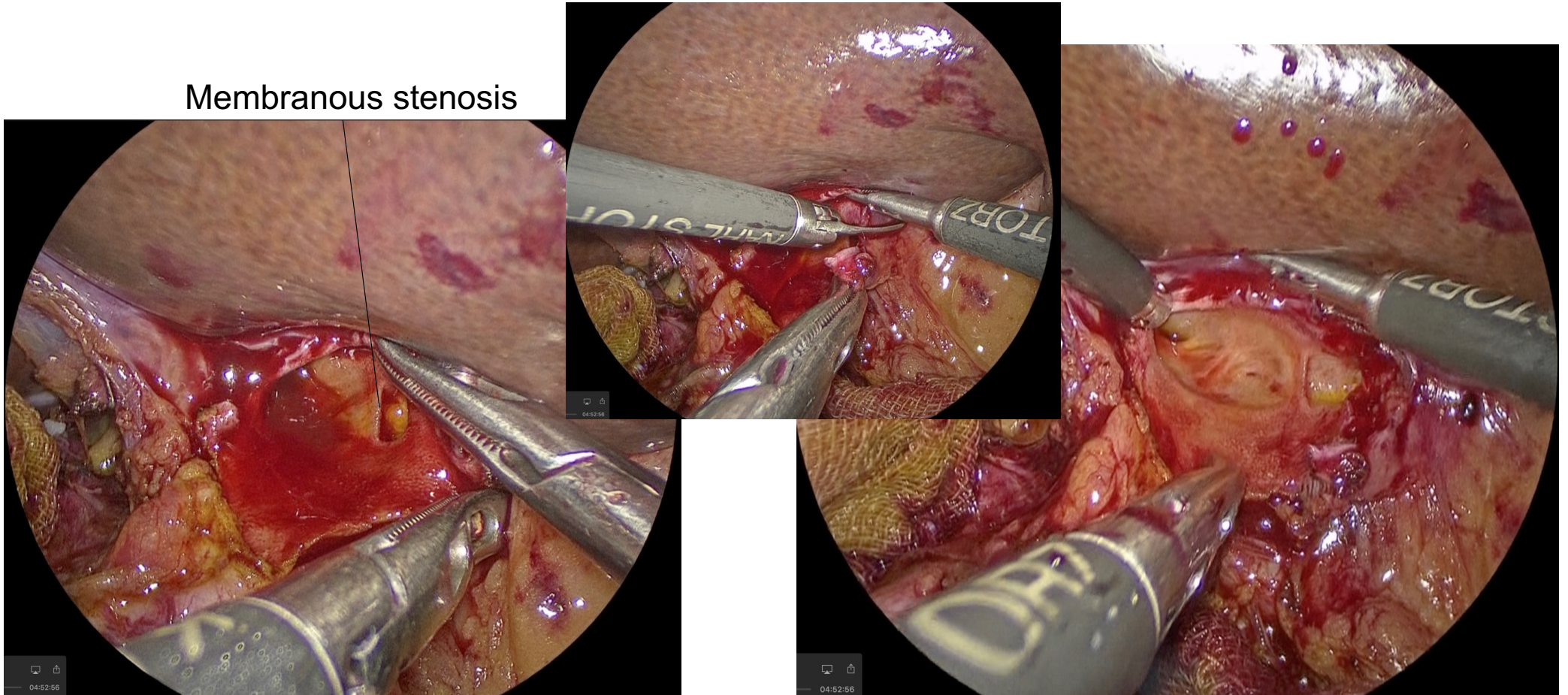
Todani and Lilly's BD plasty for membranous stenosis



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

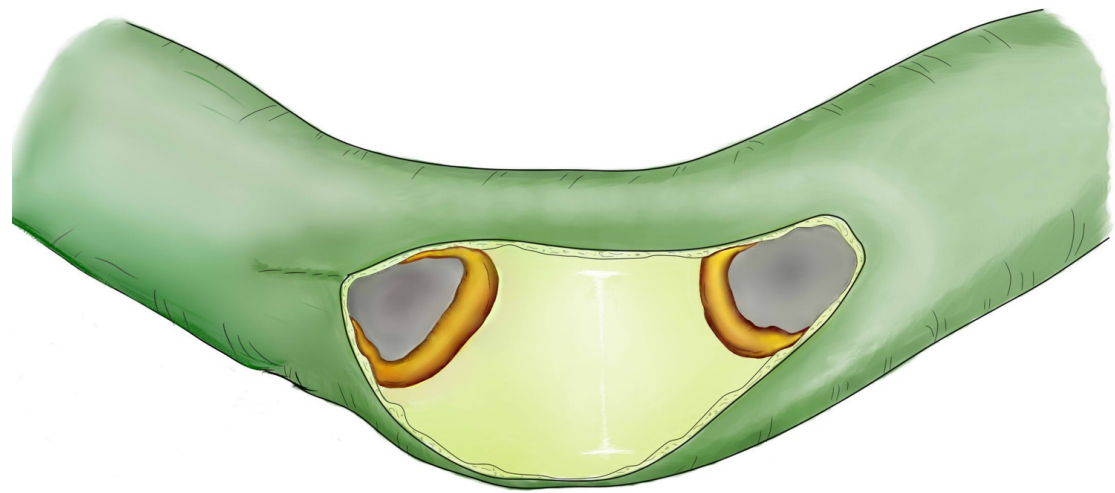
Todani's BD plasty

Membranous stenosis



Hepaticojejunostomy with a wide stoma

Especially important for IV-A cases



Hepaticojejunostomy in a case with membranous stenoses

This procedure prevents anastomotic stenosis and also relieves congenital stenosis

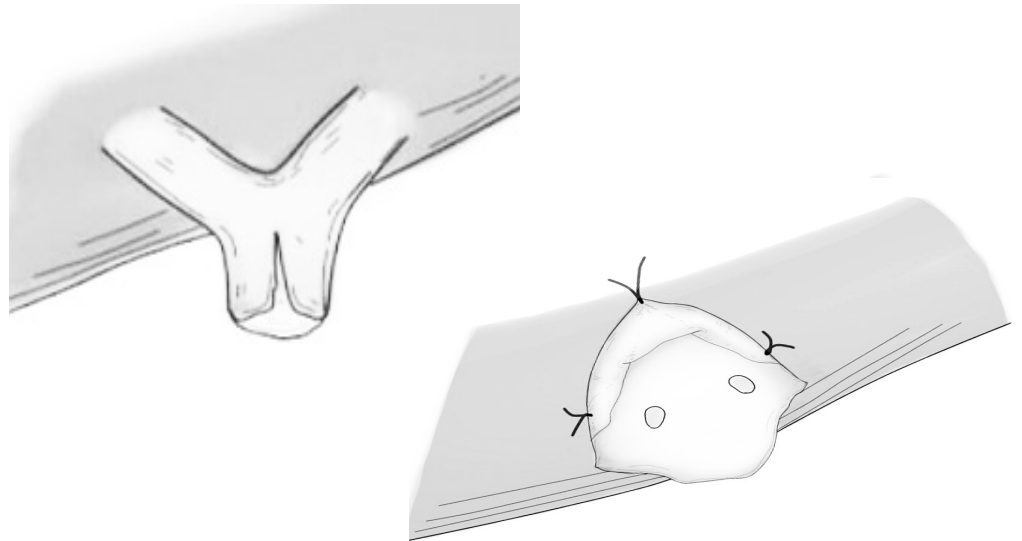
Procedures to prevent anastomotic stenosis in cases with small-caliber hepatic ducts

Open surgery



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

Laparoscopic surgery



Inverted bile duct fixed to the hilar plate provides a good field and certain anastomosis.

Urushihara et al. 2015

Too much dissection causes bad blood flow, resulting in anastomotic stricture

Right hepatic artery runs ventral to BD

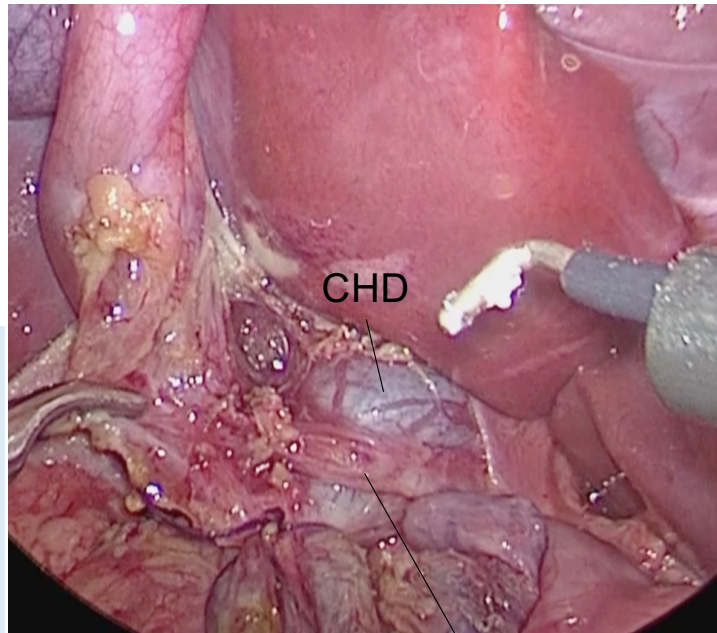
Case 23

Four-year-old boy
Type Ia CBD

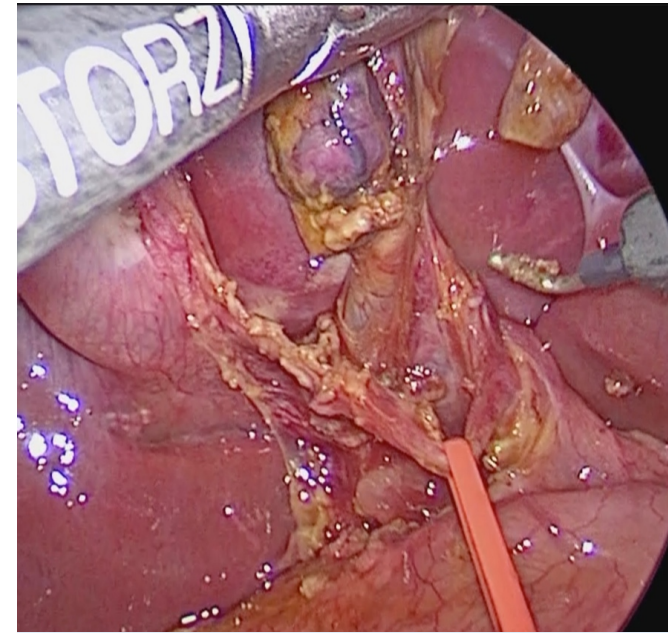
RHA ventral to CHD

Japanese normal 10% ¹⁾
PBM

Japanese adults 21% ¹⁾
Japanese children 26% ²⁾
Chinese children 17% ³⁾



RHA



Hepaticojejunostomy must be created ventral to the RHA.

If not, bile outflow is disturbed 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, Shiota 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.

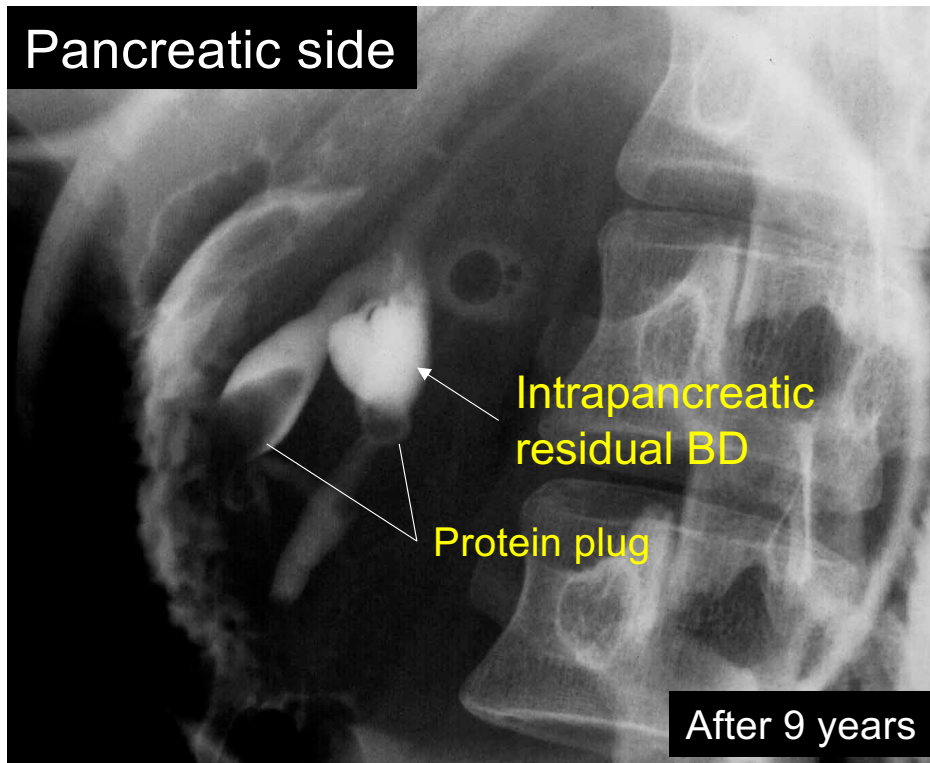
3) Guo WL, Wang J. Preoperative one-stop magnetic resonance imaging evaluation of the pancreaticobiliary junction and hepatic arteries in children with pancreaticobiliary maljunction: a prospective cohort study. Surg Today. 2021 Jan;51(1):79-85. doi: 10.1007/s00595-020-02077-5. Epub 2020 Jul 12. PMID: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

Basics

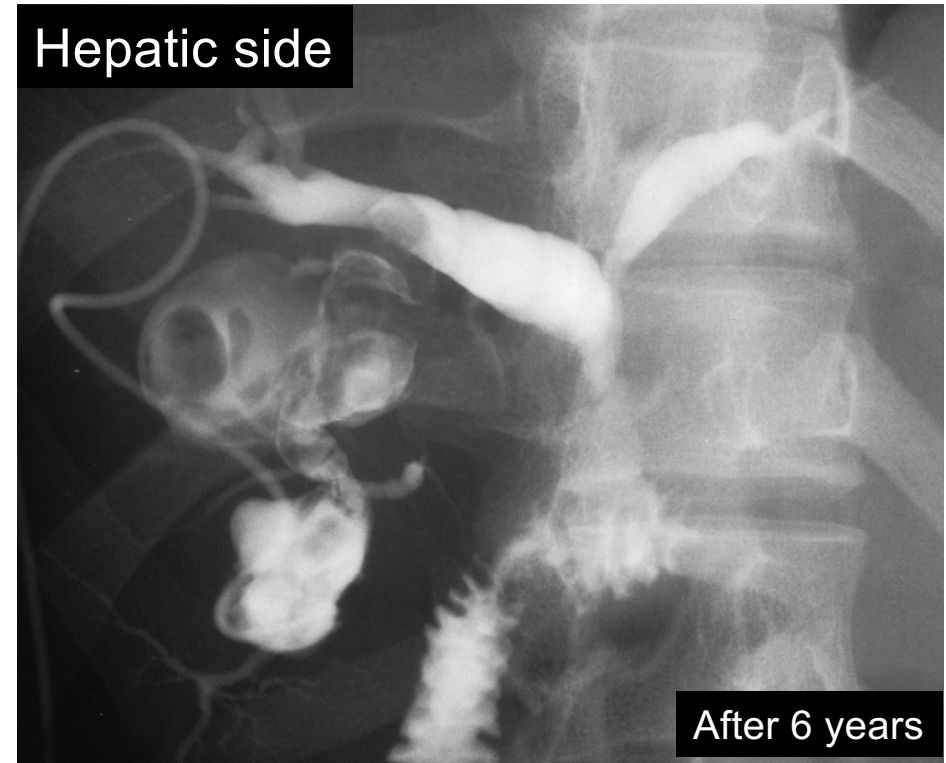
Postoperative Complications

First version, September 2022

Long-term complications after BD resection



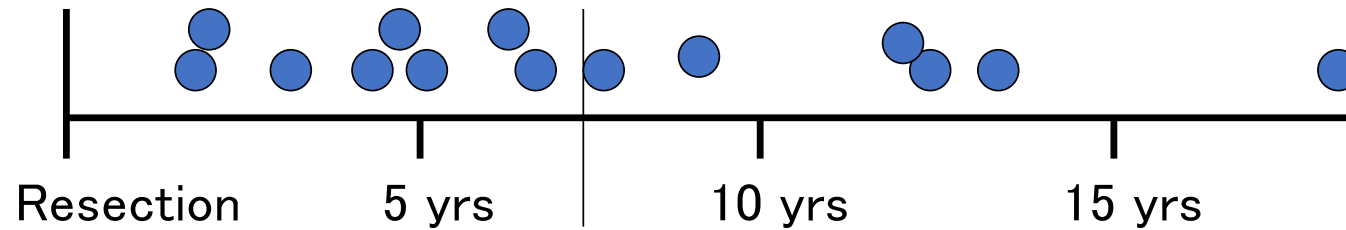
A residual intrapancreatic BD would cause protein plugs or intrapancreatic stones and cancers in the residual BD. Complete excision of intrapancreatic BDs is necessary.



Intrahepatic gallstones and/or cholangitis occurs at a rate of 7% to 8% several years after BD resection.

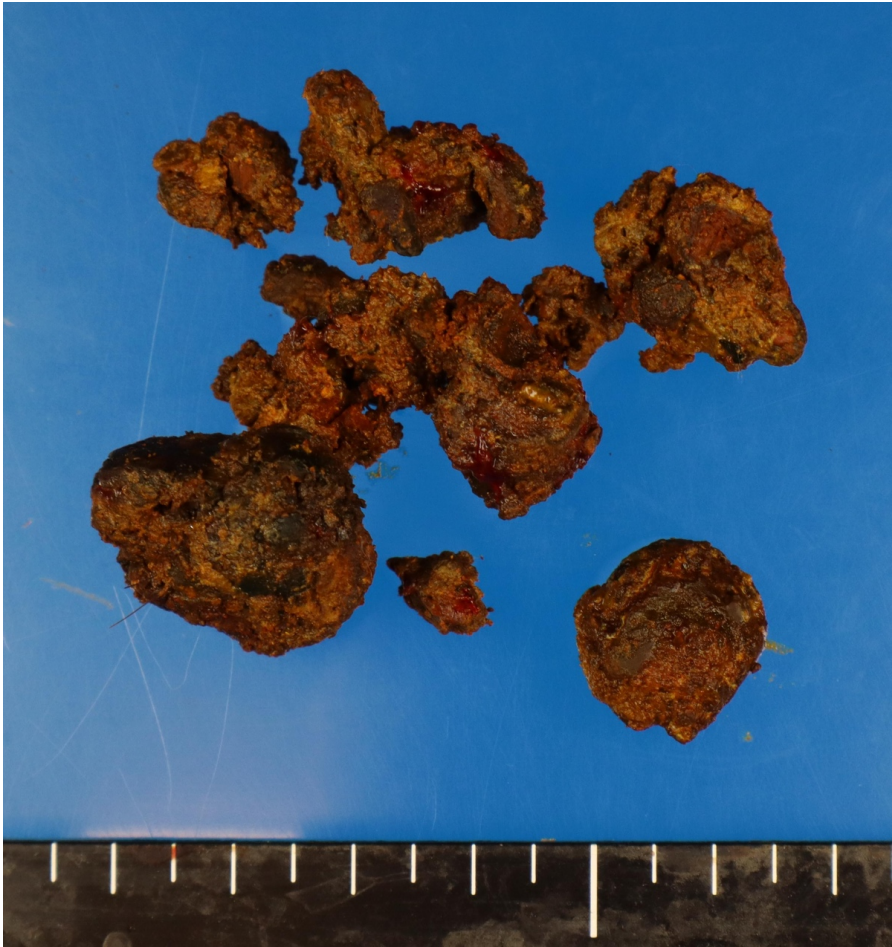
Period from resection to intrahepatic gallstone formation

Pediatric 14 cases



Average 7.3 yrs

Postoperative gallstones are brown pigment stones



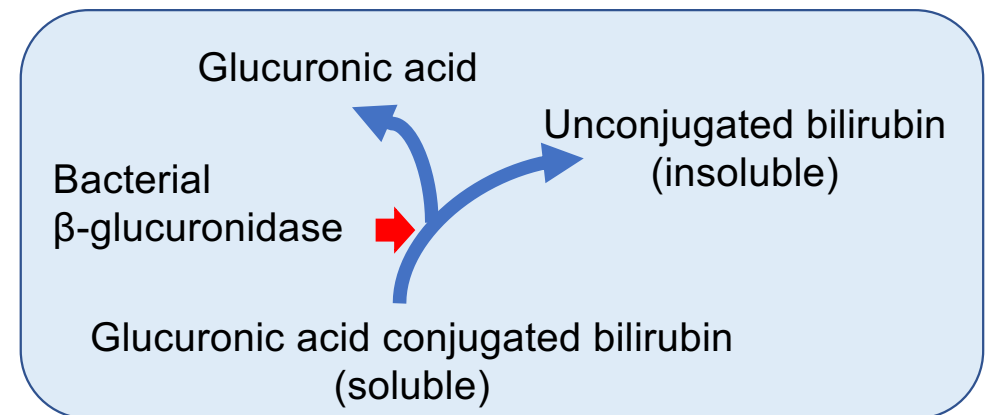
Composition:

Bilirubin calcium + fatty acid calcium



Bacterial β -glucuronidase and phospholipase A1

Bile stasis and bacterial infection



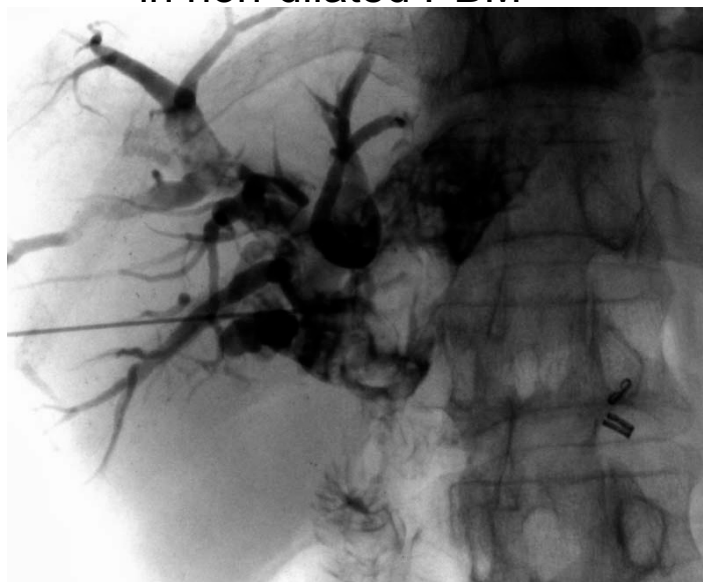
Risk factors of postoperative hepatolithiasis

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

Todani type IV-A



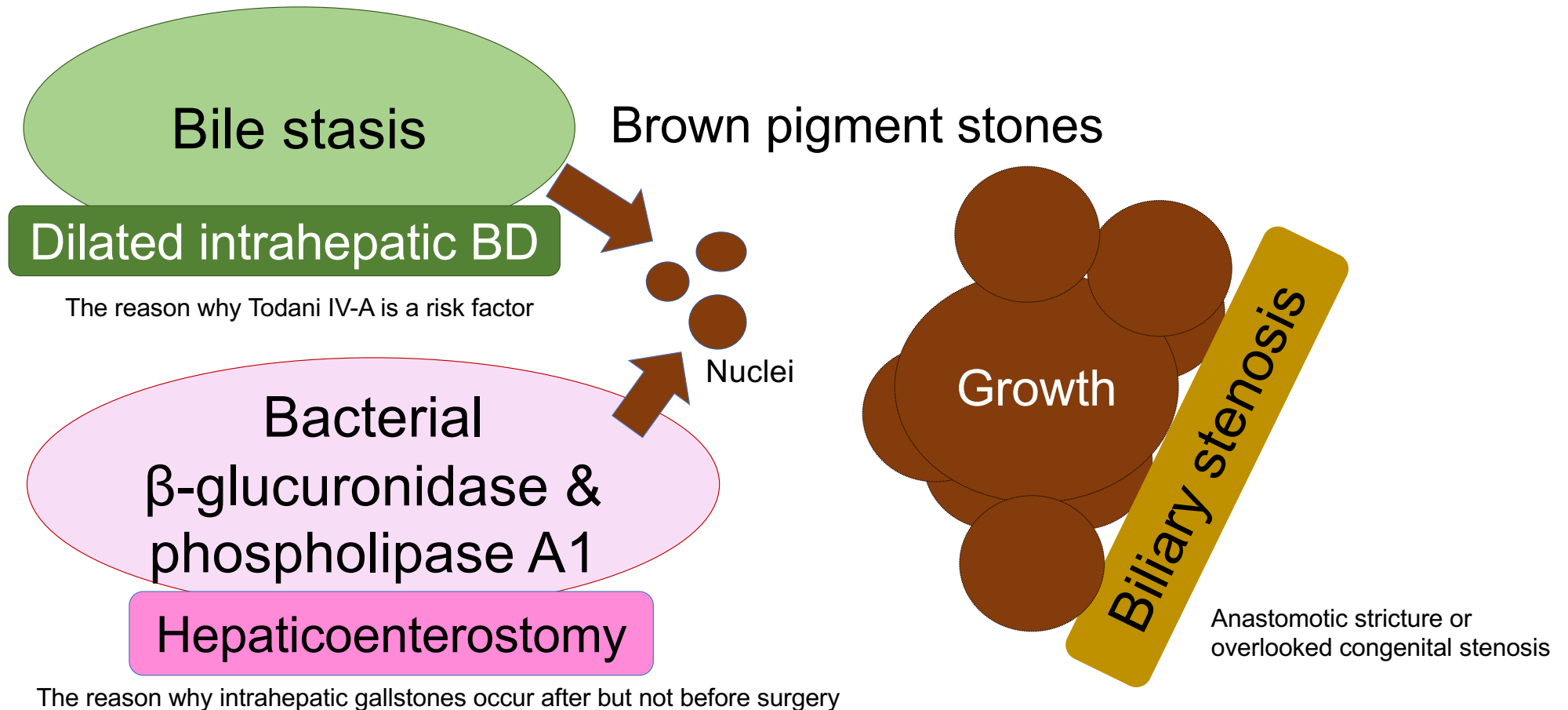
Anastomotic stricture
in non-dilated PBM



Congenital septal stenosis



Mechanism of postoperative gallstone formation

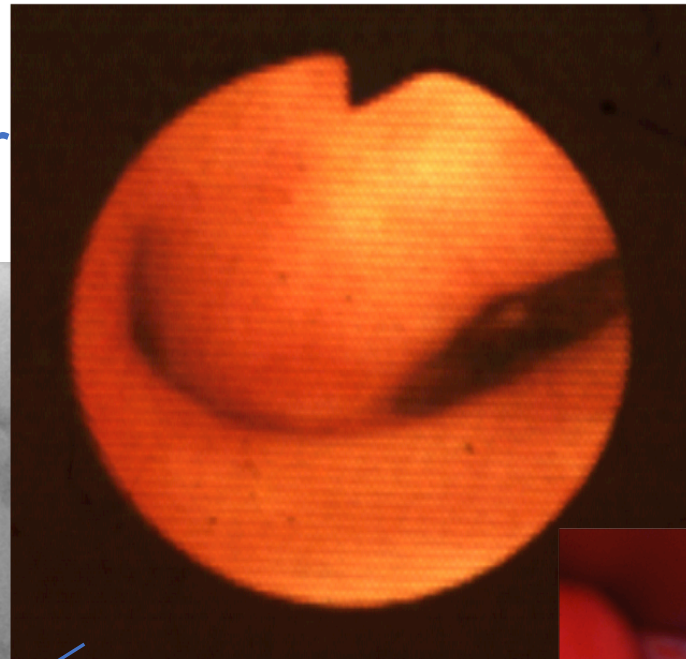
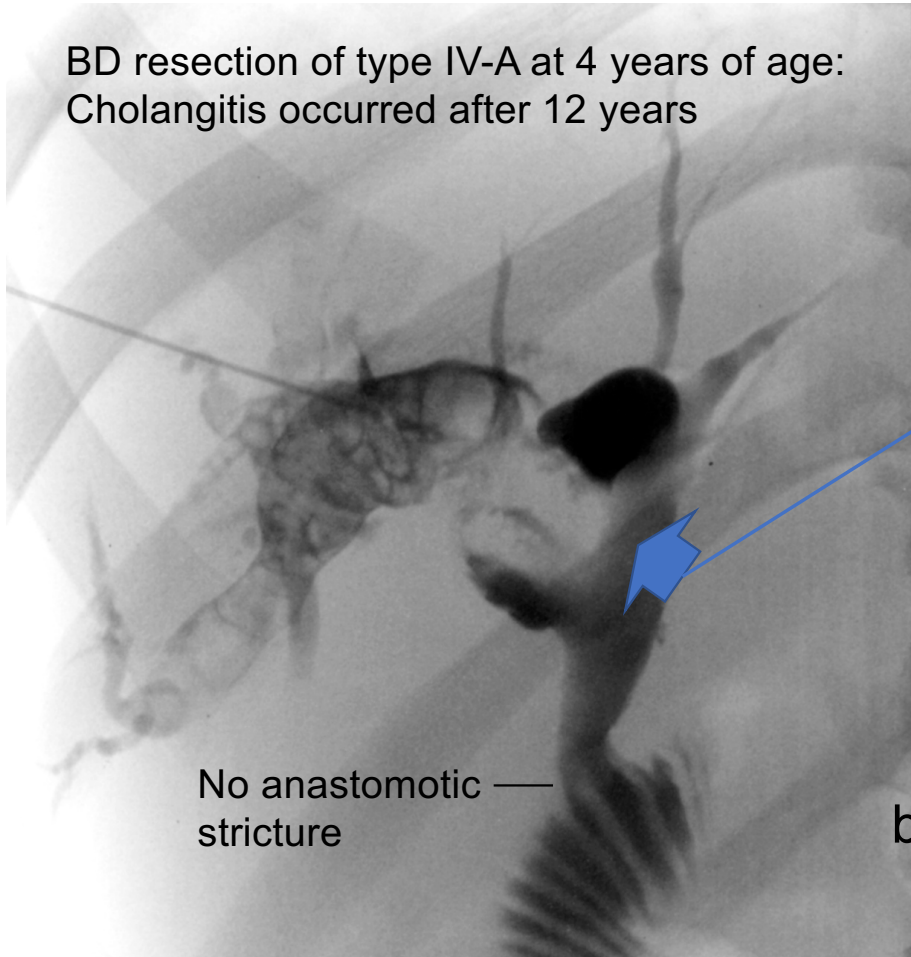


Only removal of stones may be insufficient for a cure; this needs to relieve stenosis or dilated BD (hepatectomy).

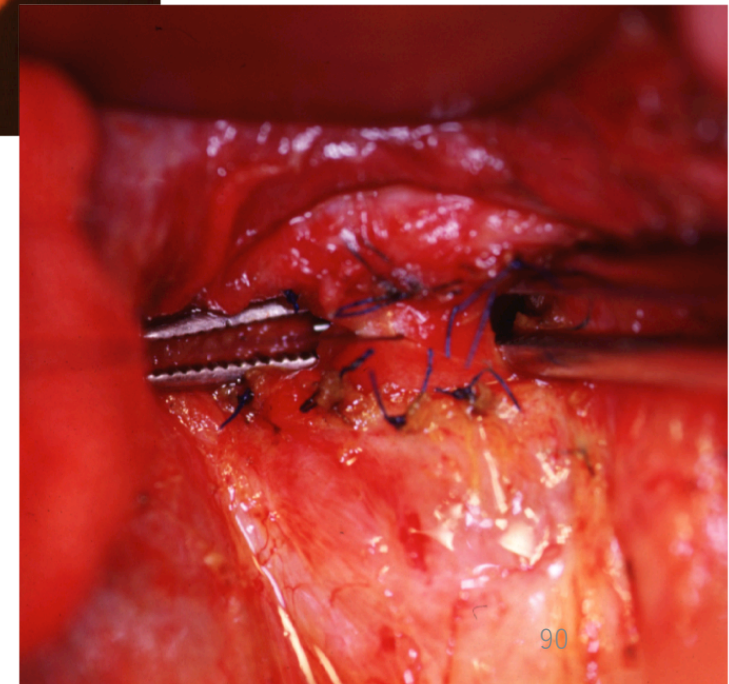
Case 24

Intrahepatic gallstones after BD resection

BD resection of type IV-A at 4 years of age:
Cholangitis occurred after 12 years



Septal stenosis blocking out gallstones

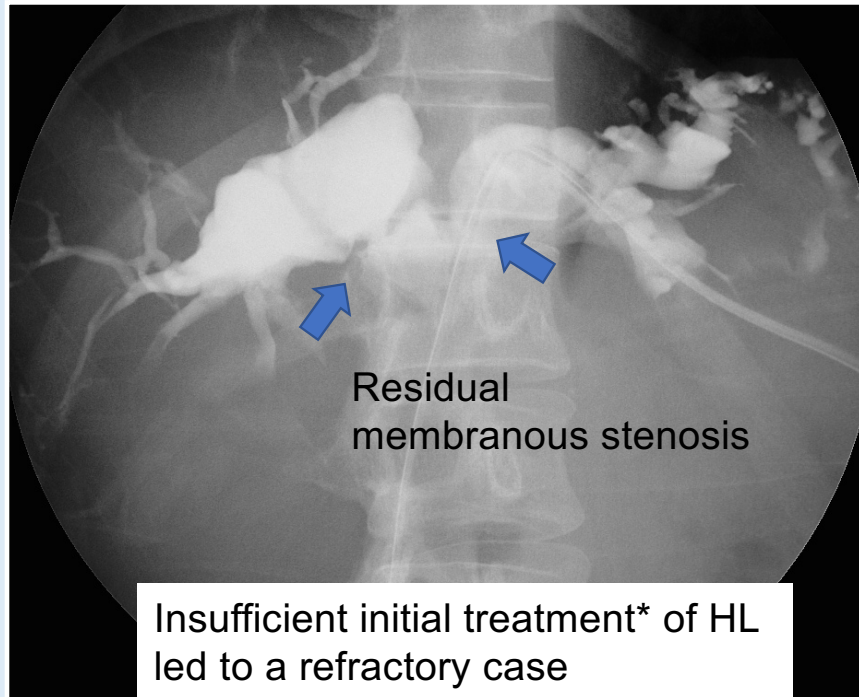


Refractory cholangitis will occur

Case 25

Formation of cavernous transformation of PV

BD resection of type IV-A at 1 year
↓
In her twenties, hepatolithiasis PTCS* was performed
↓
Repeated mild cholangitis but she did not seek advice
↓
In her thirties, hepatolithiasis and severe cholangitis occurred



*PTCS, percutaneous transhepatic cholangioscopic lithotomy



Long-term postoperative complications: Carcinogenesis at residual BD

Higher incidence even after BD resection

- 107 cases collected from the literature
- Carcinogenesis rate 2.0%
- An average of 12 postoperative years at 47 years of age

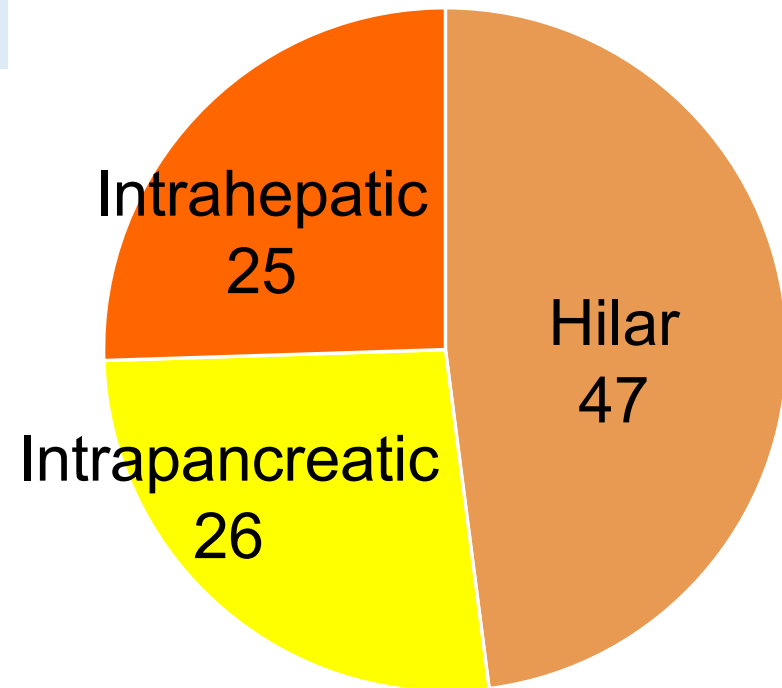
Kamisawa T, Kaneko K, Itoi T, Ando H. Pancreaticobiliary maljunction and congenital biliary dilatation. Lancet Gastroenterol Hepatol. 2017 Aug;2(8):610-618. doi: 10.1016/S2468-1253(17)30002-X. PMID: 28691687.

cf. Survey in USA

Biliary cancer after BD resection 13/394 3.3%

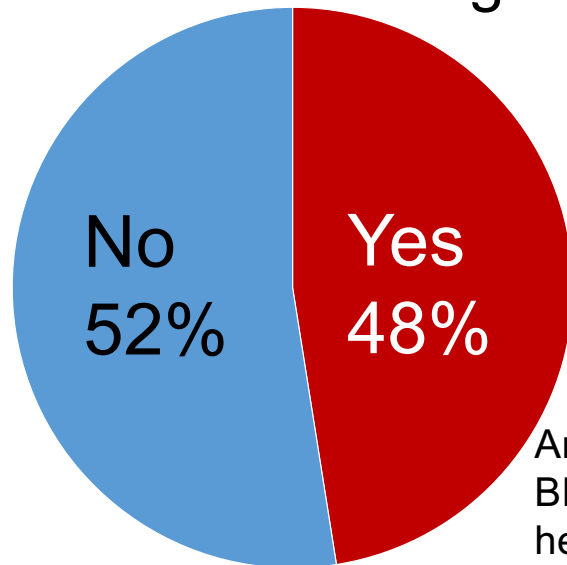
Children 4/135 3.0%, Adults 18/259 6.9%

Soares et al. JAMA Surg 2015



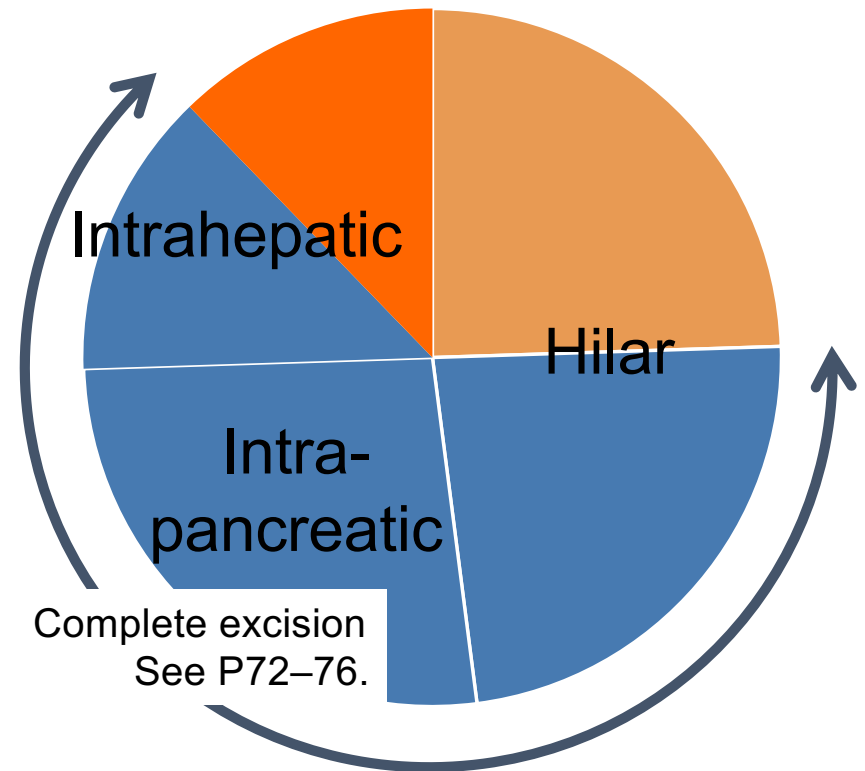
Many residual BD cancers could have been prevented

Hepatolithiasis
and/or cholangitis



Among 80 residual
BD cancers at the
hepatic side

Approximately half of postoperative residual BD cancers have hepatolithiasis and/or cholangitis. Anastomotic stricture or overlooked congenital stenosis leads to hepatolithiasis and/or cholangitis, resulting in carcinogenesis.



Appropriate procedures could have protected against these cancers.

Intrahepatic BD cancer after BD resection

Case 26

Male in his sixties:

Type IV-A

BD resection with

hepaticoduodenostomy

↓ After 4 years

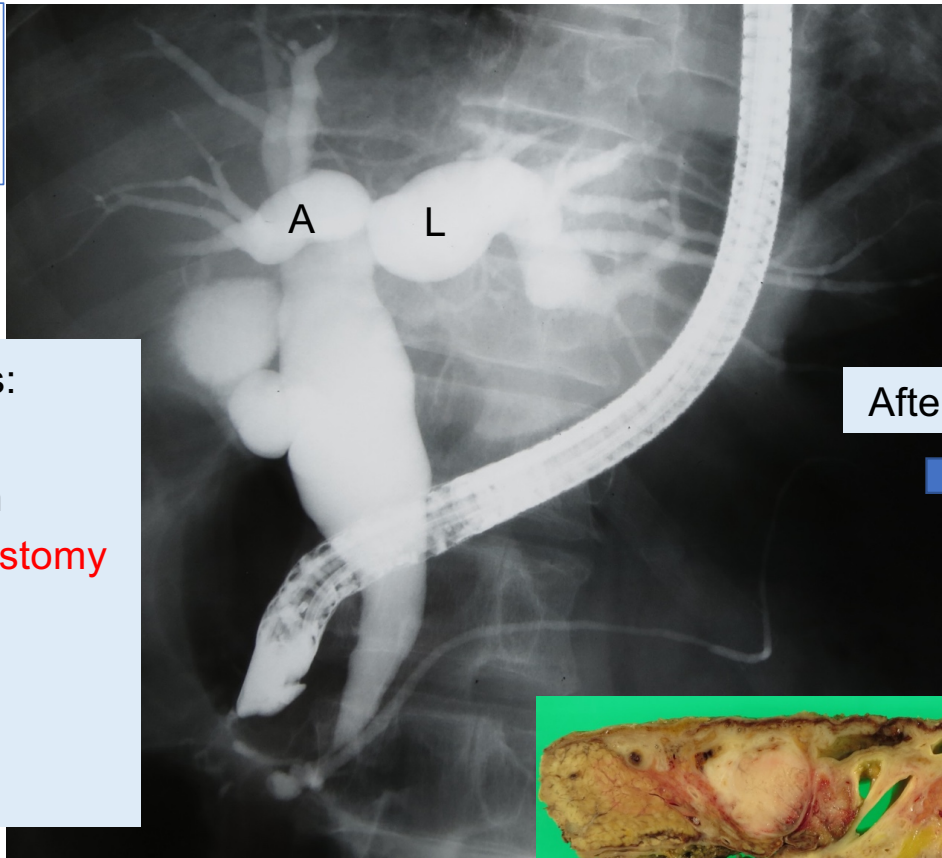
Cholangitis

↓ After 9 years

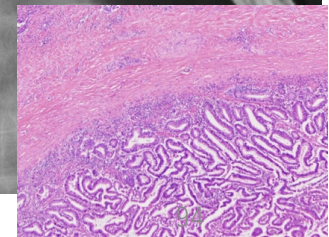
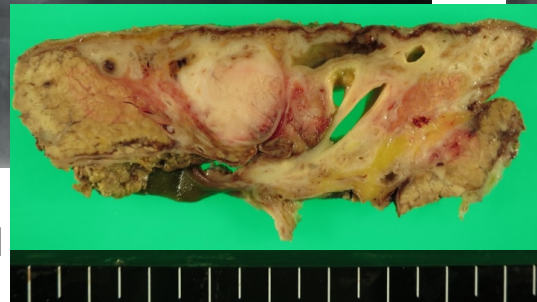
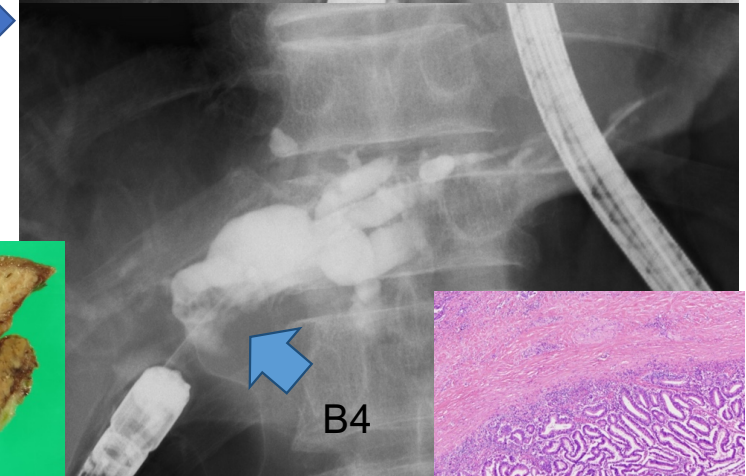
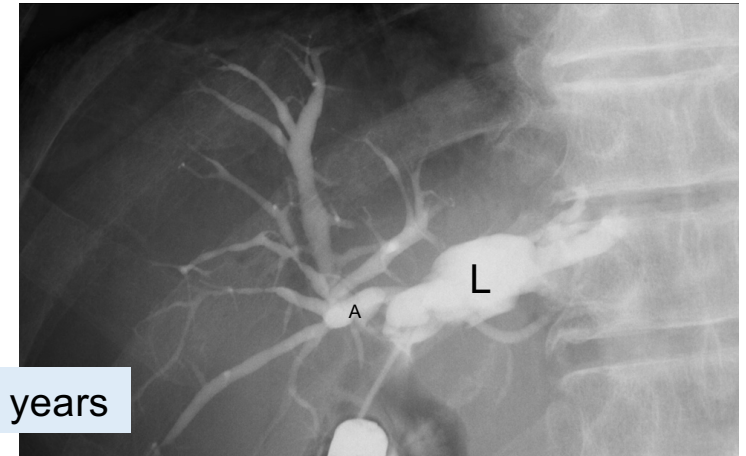
BD cancer (B4)

A, anterior; L, left hepatic duct.

Dilatation of the anterior segment diminished postoperatively.



After 13 years



Basics

High confluence of the pancreaticobiliary ducts

First version, September 2024

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

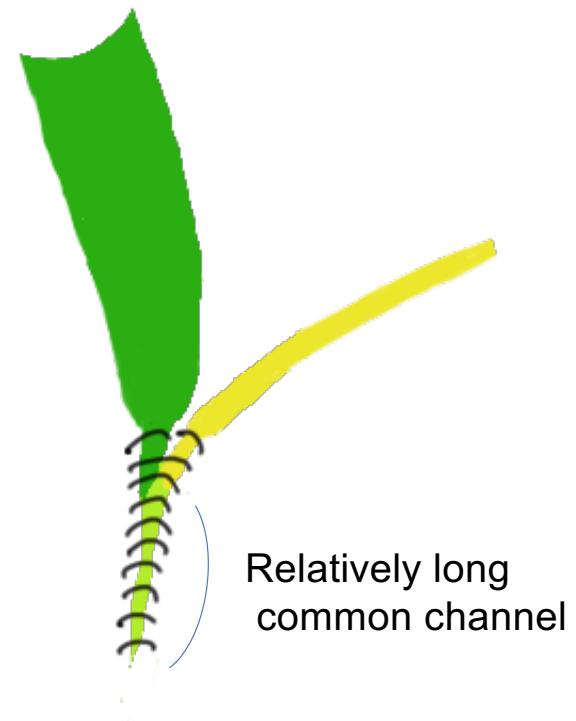
High confluence is the **transitional type between normal and PBM**, and frequently causes **pancreatobiliary reflux**. It is known that patients with high confluence often have gallbladder cancers as is seen in pancreaticobiliary maljunction. Sai et al. proposed a similar concept of **occult pancreatobiliary reflux**, which is defined as the confluence is normal but yields **pancreatobiliary reflux**.

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

Normal

High
confluence

PBM



Spectrum from normal to PBM

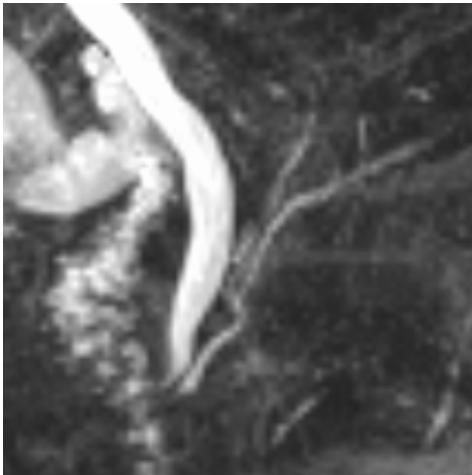
Normal

Occult pancreatobiliary reflux

High confluence

PBM

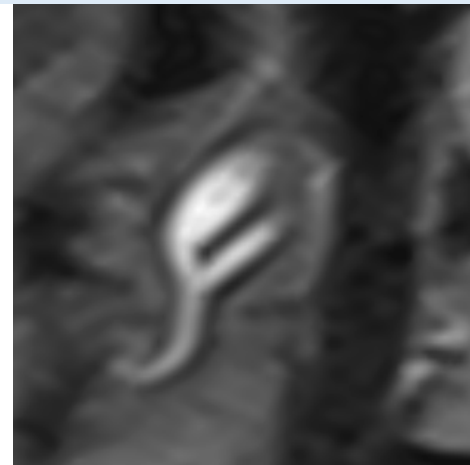
The sphincter function affecting the confluence



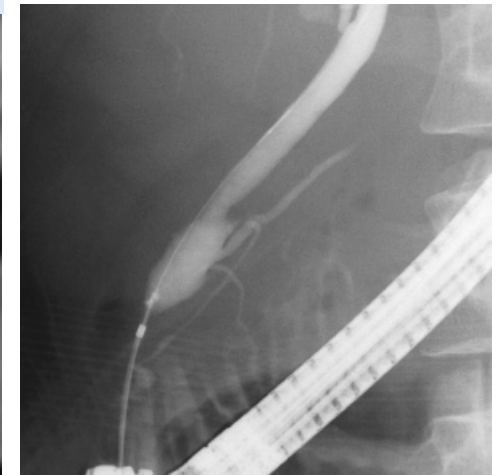
Separate openings



The common channel formed
Cholangiography occasionally
shows the PD.



Relatively long
common channel



Abnormally long
common channel

Advanced

Issues on Todani's classification

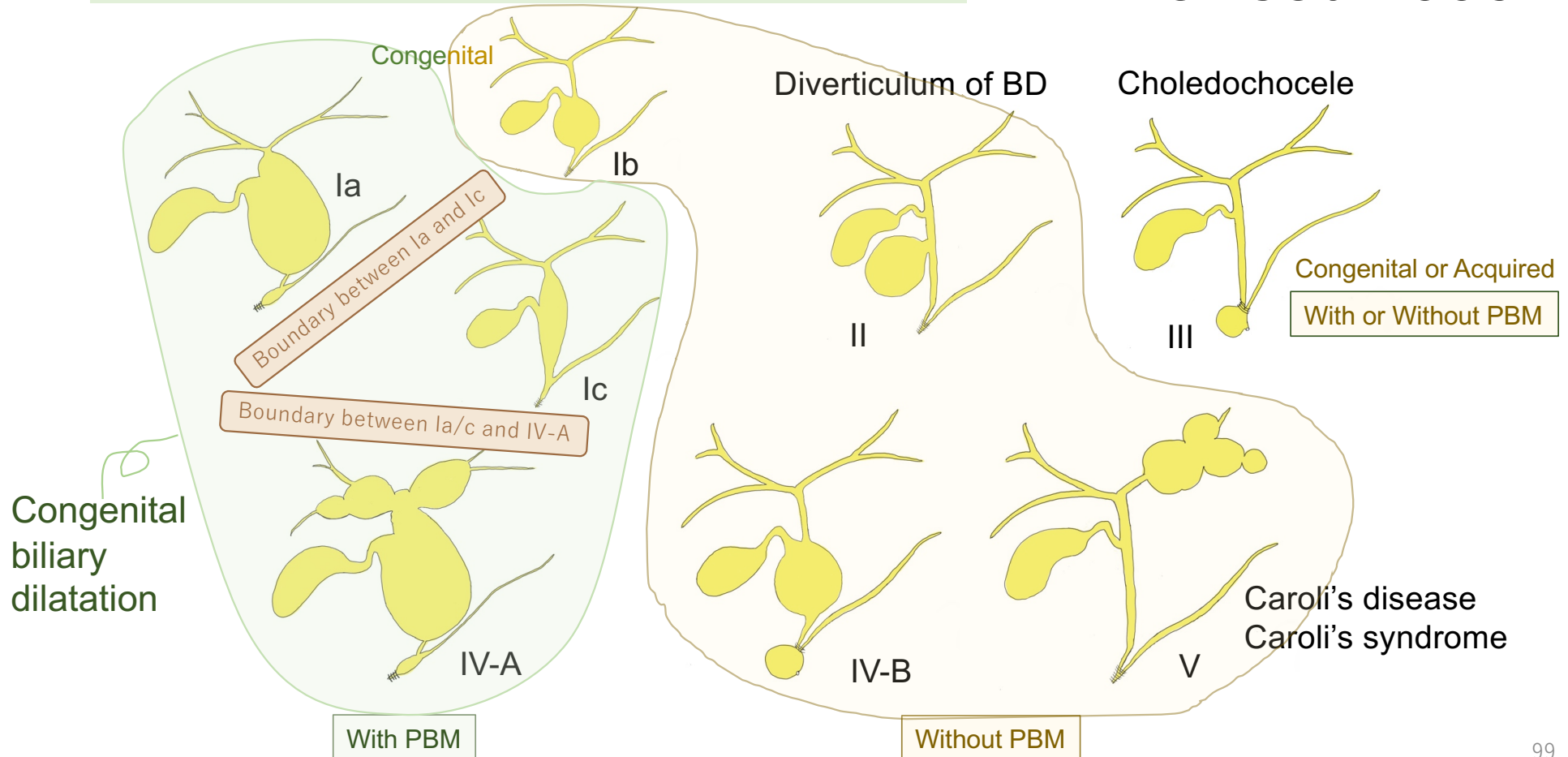
Arbitrary boundaries between Ia/c and IV-A, and between Ia and Ic

First version, September 2024

2nd version, September 2025

Todani's classification of choledochal cysts 1977, revised 1995

The term choledochal cyst involves different entities



Todani type I

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

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

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

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

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

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

Todani type IV

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

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

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

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

- **Type IV** is a multiple cyst

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

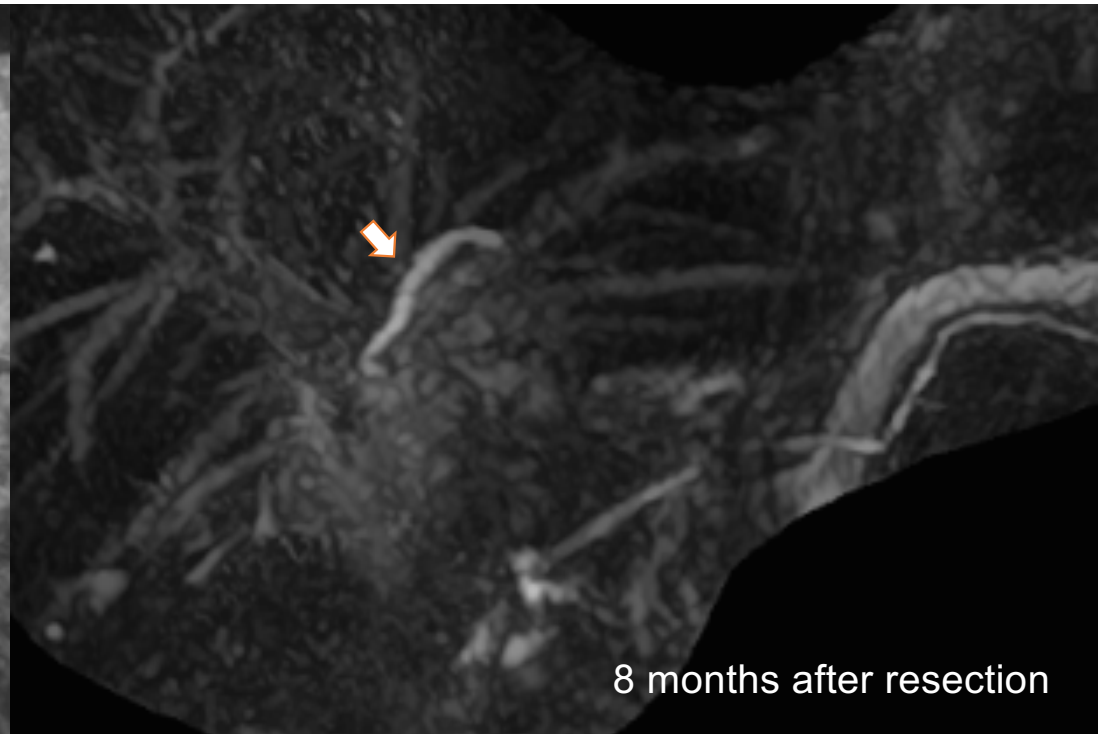
Based on the shape of the extrahepatic and intrahepatic ducts, type IV is subclassified into three forms, cystic–cystic, cystic–fusiform, and fusiform–fusiform.

Necessary condition: BD dilatation upstream from CHD

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

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

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



Literature

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

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

Case 27

Ic>Ia>>IV-A

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

Female in her thirties
Abdominal pain

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

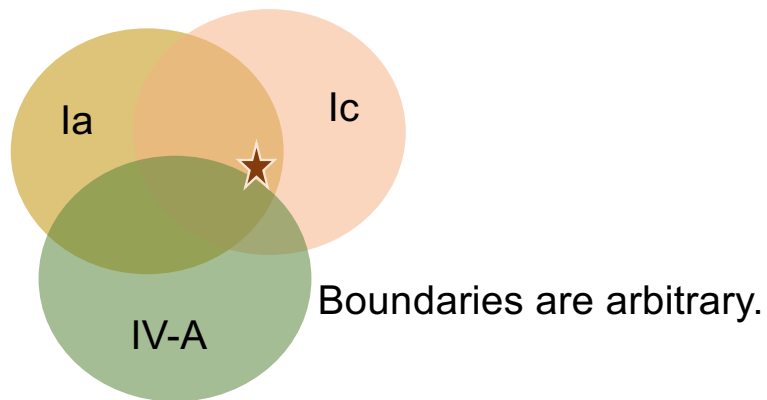


Case 28

Ic>Ia>IV-A

ERCP in a male in his seventies
Abdominal pain and fever

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



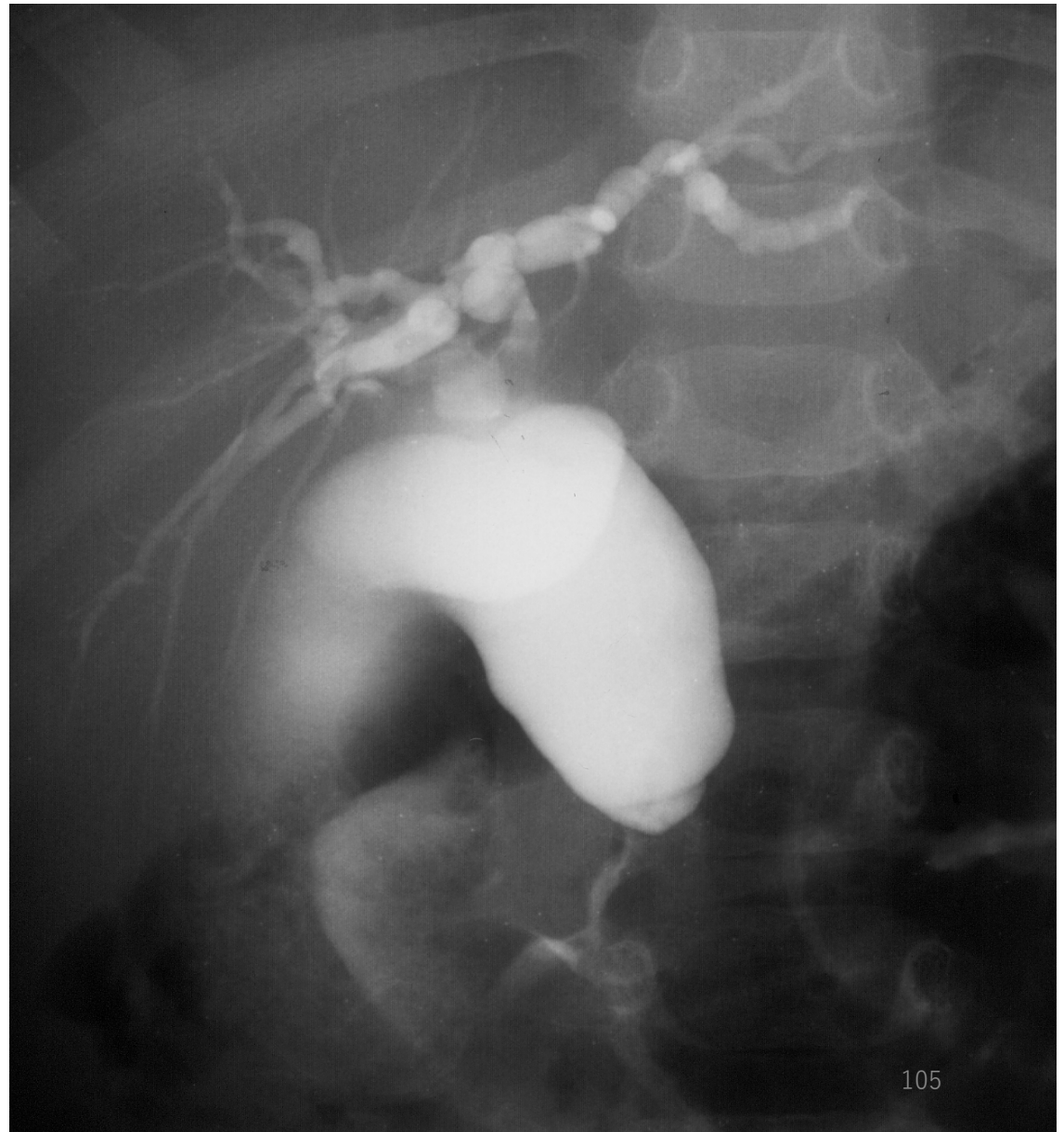
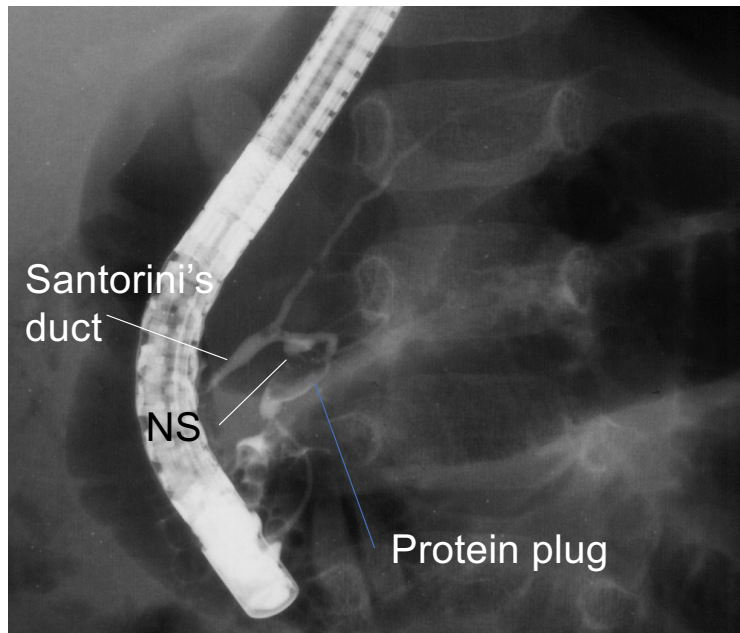
Case 29

Ia>IV-A

ERCP in a 9-year-old child

Abdominal pain, jaundice, acholic stool

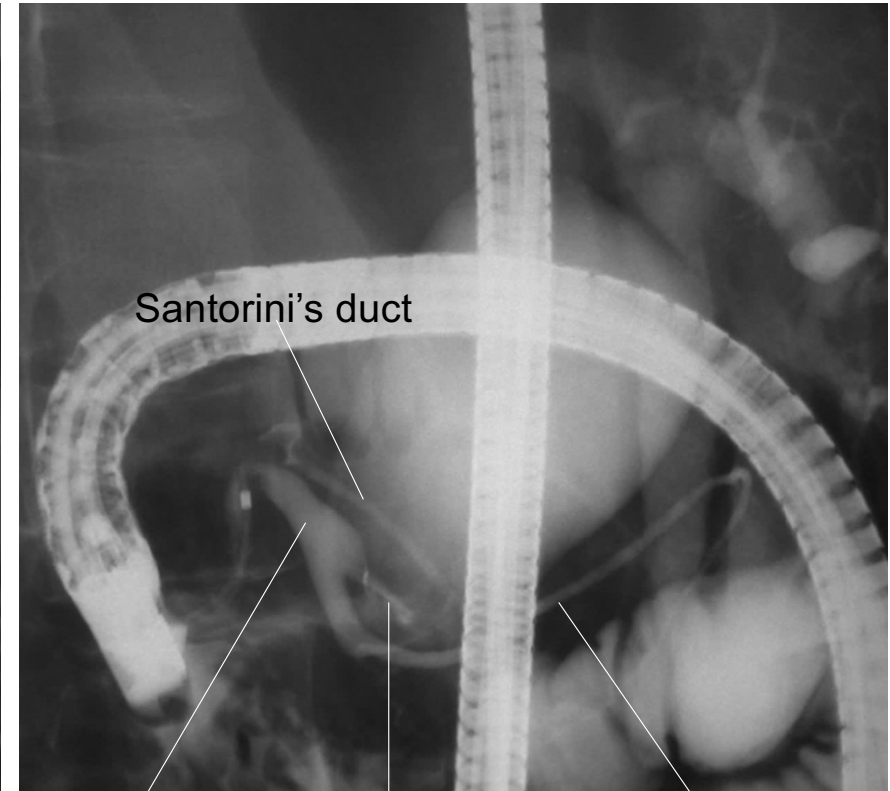
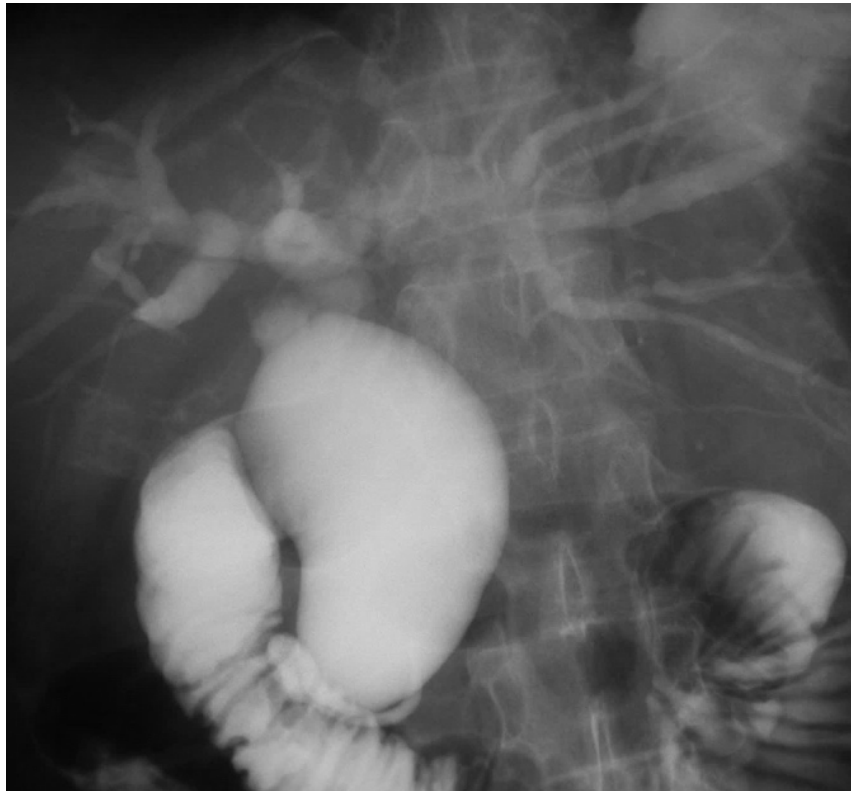
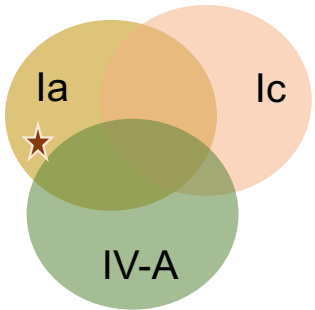
Transient diffuse intrahepatic BD (IHBD) dilatation due to protein plug obstruction
Dilatation diminished after BD resection.



Case 30

la > IV-A

Female in her
fifties
ERCP shows
diffuse dilatation
of IHBD



Common channel

NS

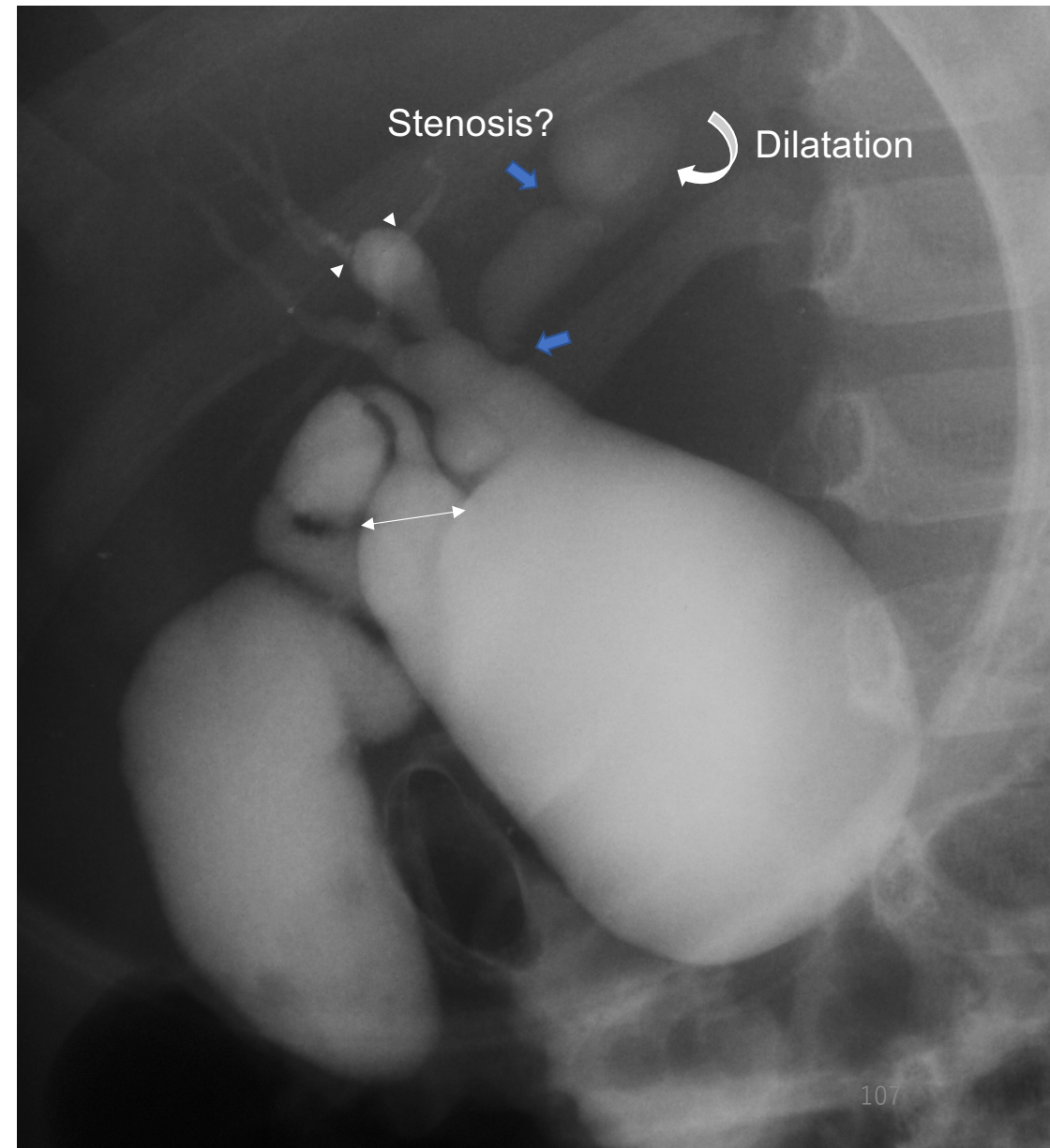
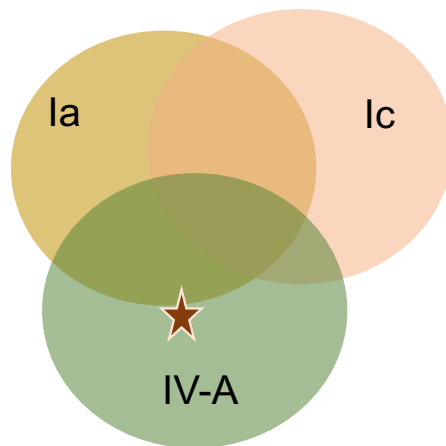
Main PD

Case 31

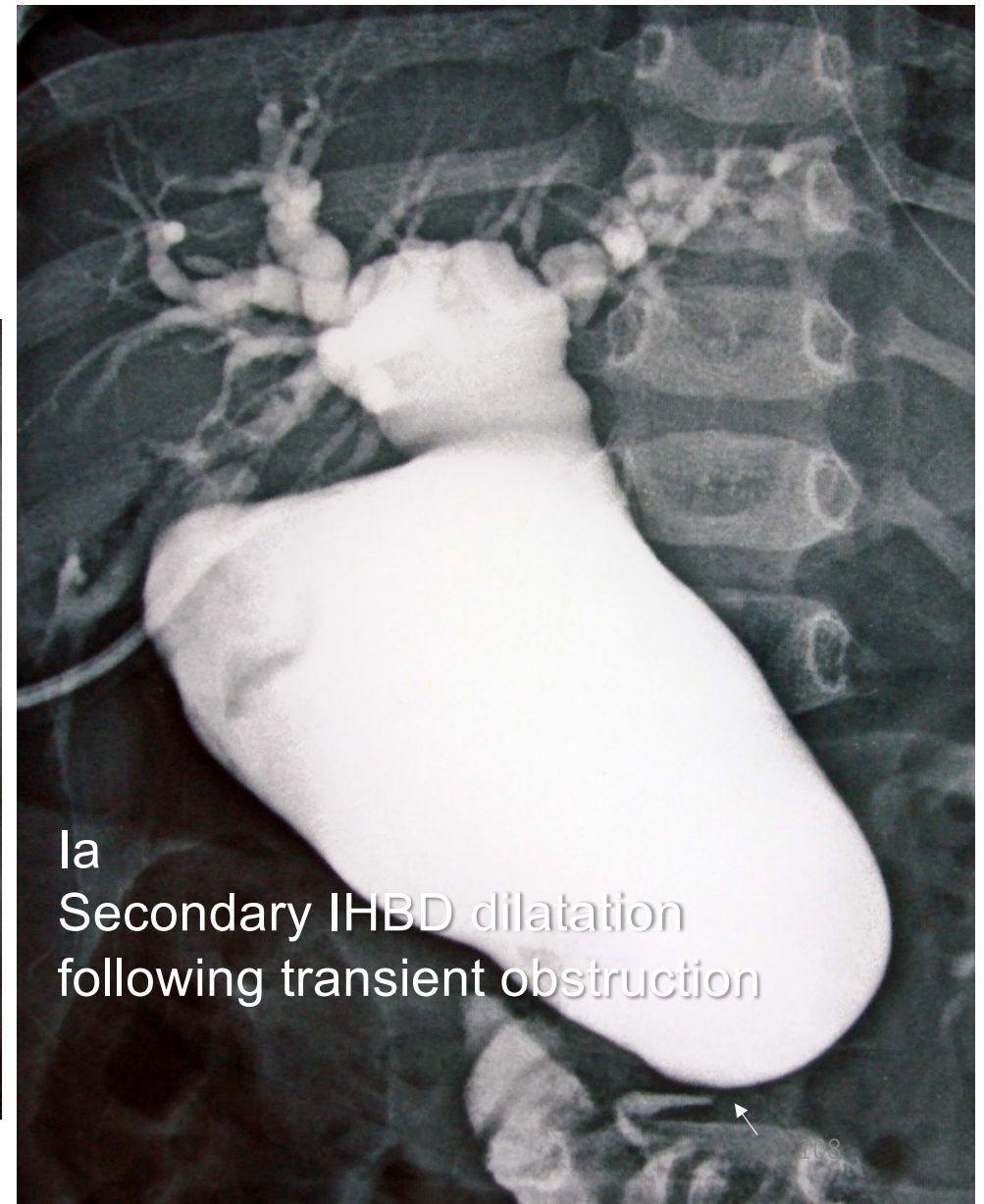
IV-A > Ia

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

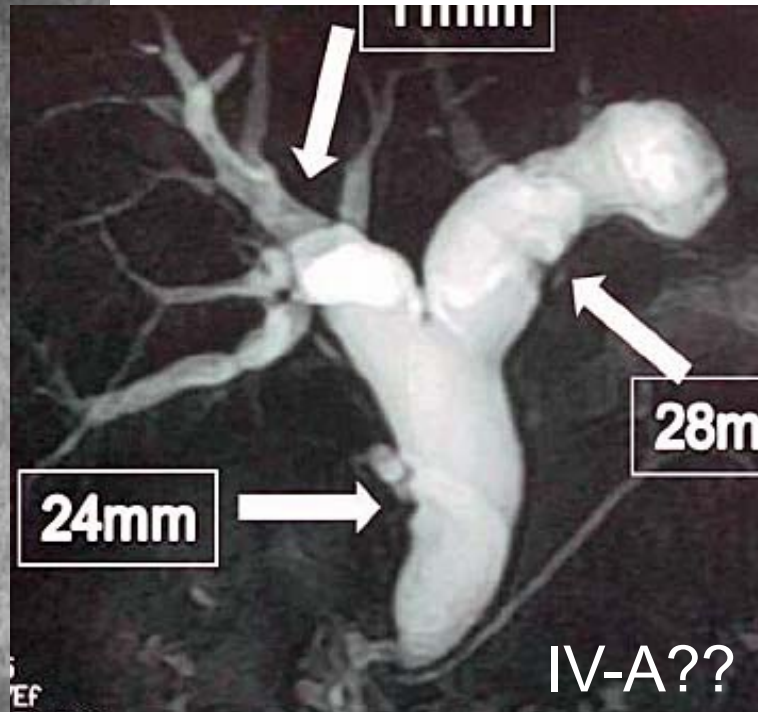
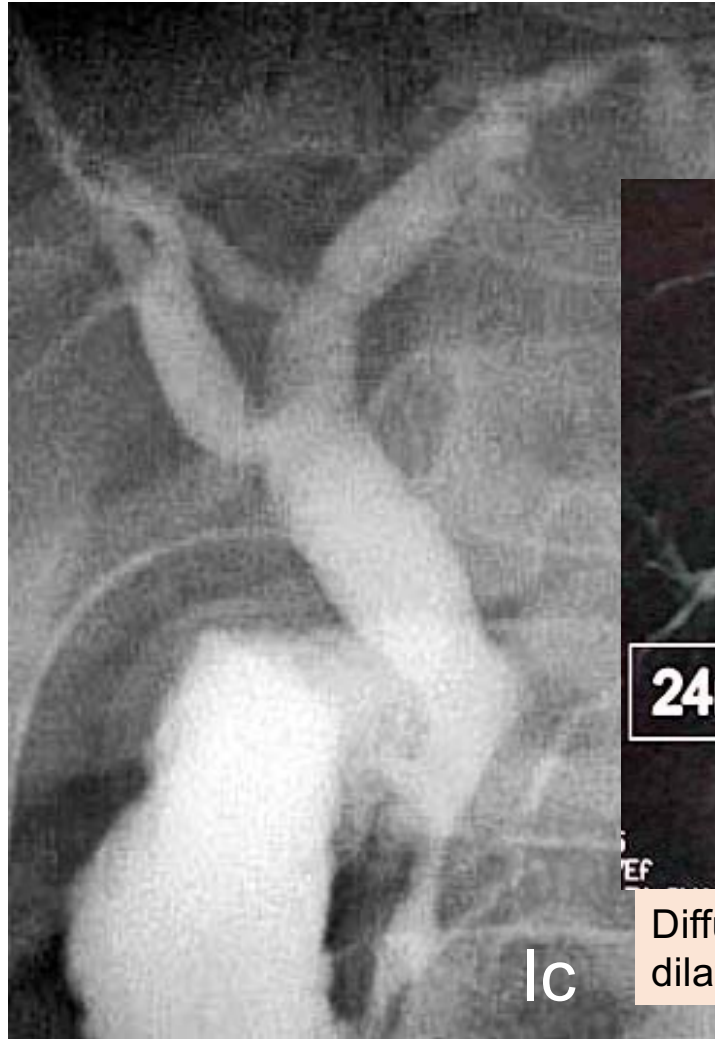
- Cystic dilatation upstream from LHD (curved arrow) with constrictions (arrows)
- Limited dilatation (arrow heads) indicating type IV-A
- Eminent dilatation at the base of the cystic duct (double-headed arrow)



Two cases: Ia or IV-A?

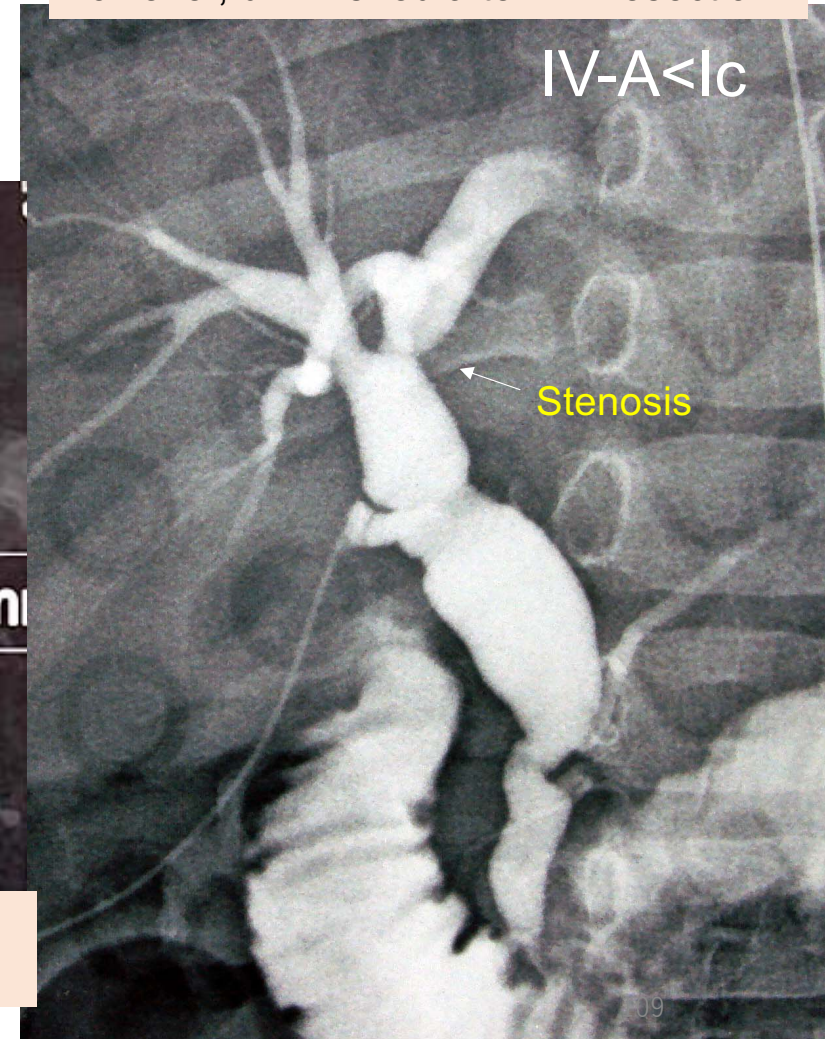


Three cases: Ic or IV-A?



Diffuse dilatation without stenosis but dilatation remaining after BD resection

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



Case 32 Ia or IV-A?

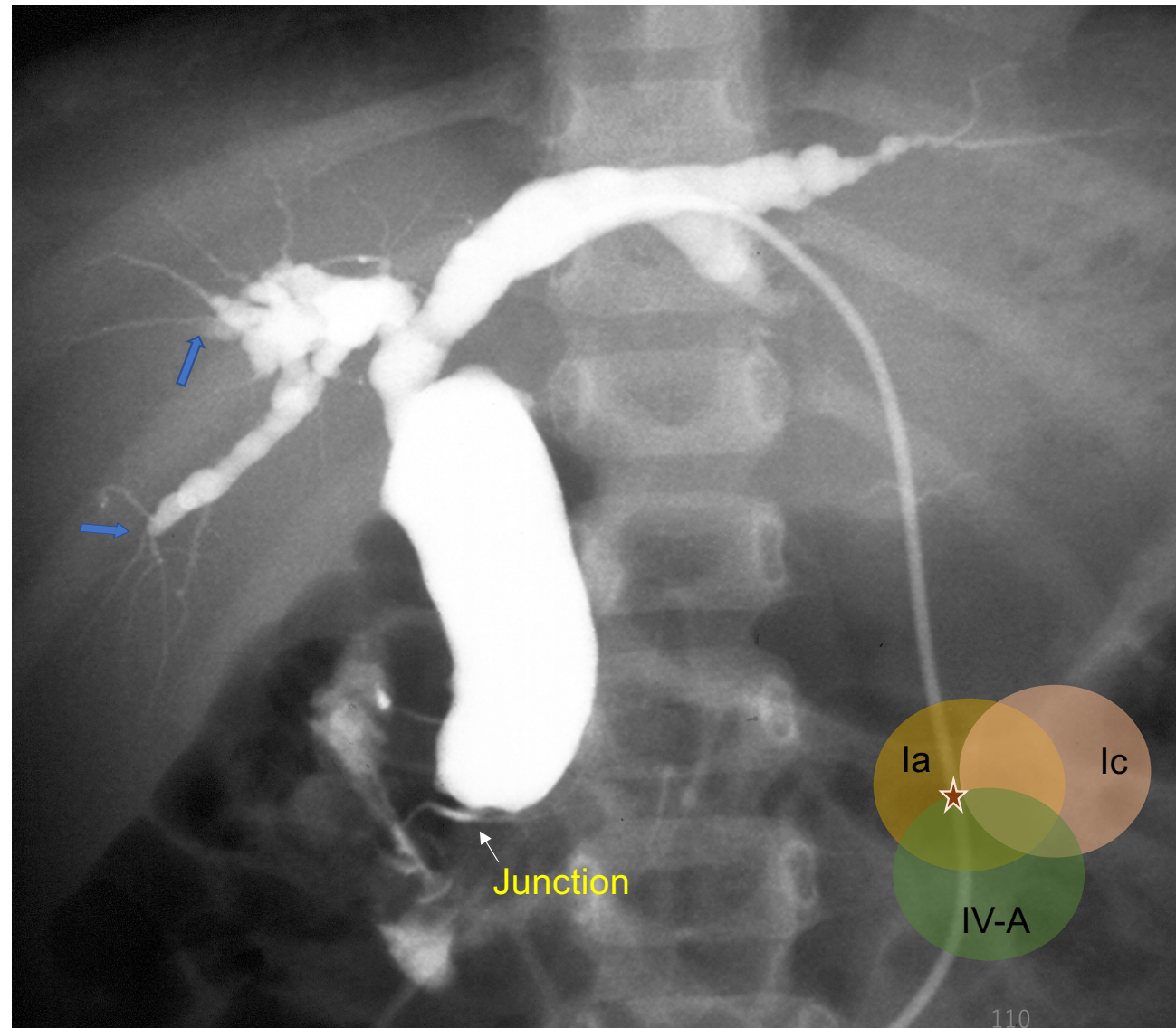
Percutaneous transhepatic cholangiography in a 2-year-old boy
Abdominal pain, jaundice, elevated serum levels of hepato-biliary enzymes

- **Cystic-dilated EHBD** 17 mm
- **Fusiform-dilated IHBDs** (LHD 8mm) abruptly change to those of normal caliber (→), indicating **localized dilatation**.

However, this IHBD dilatation would diminish after BD resection, which contradicts IV-A criteria.

- Very short narrow segment
Type A (stenotic type) PBM

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



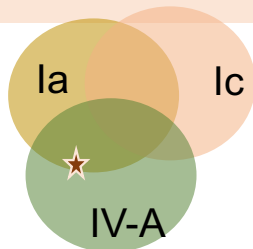
Case 33

Ia or IV-A?

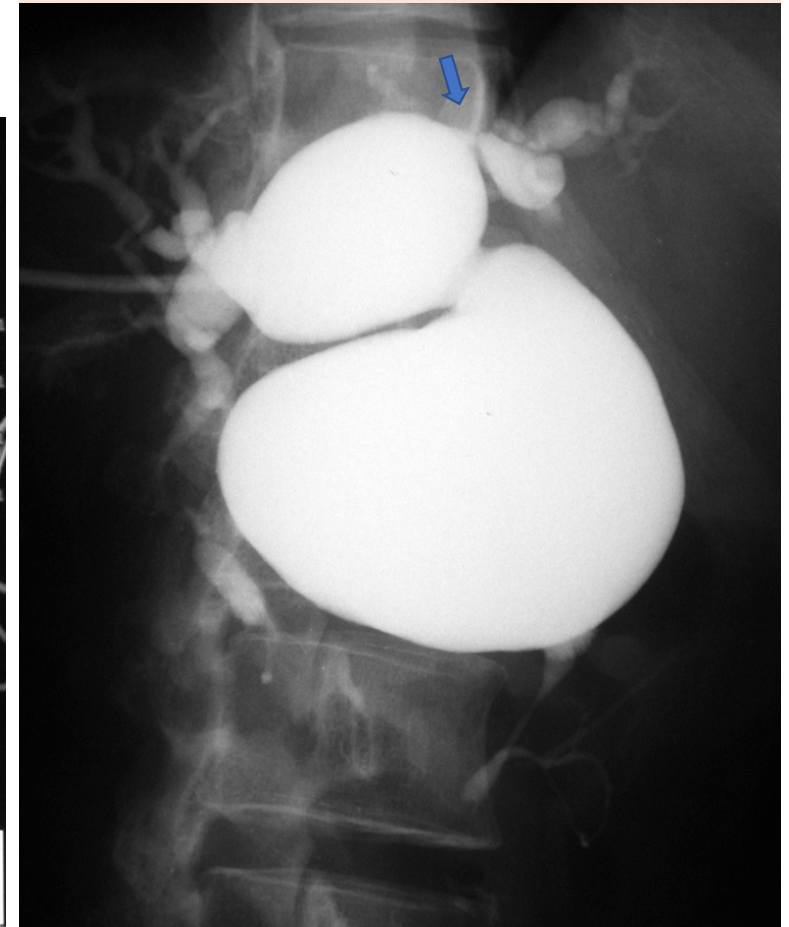
Female in her twenties
Percutaneous transhepatic
biliary drainage due to fever,
jaundice, and abdominal
pain

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

Ia or IV-A, questionable



Stenosis at the orifice but only slightly
dilatated LHD



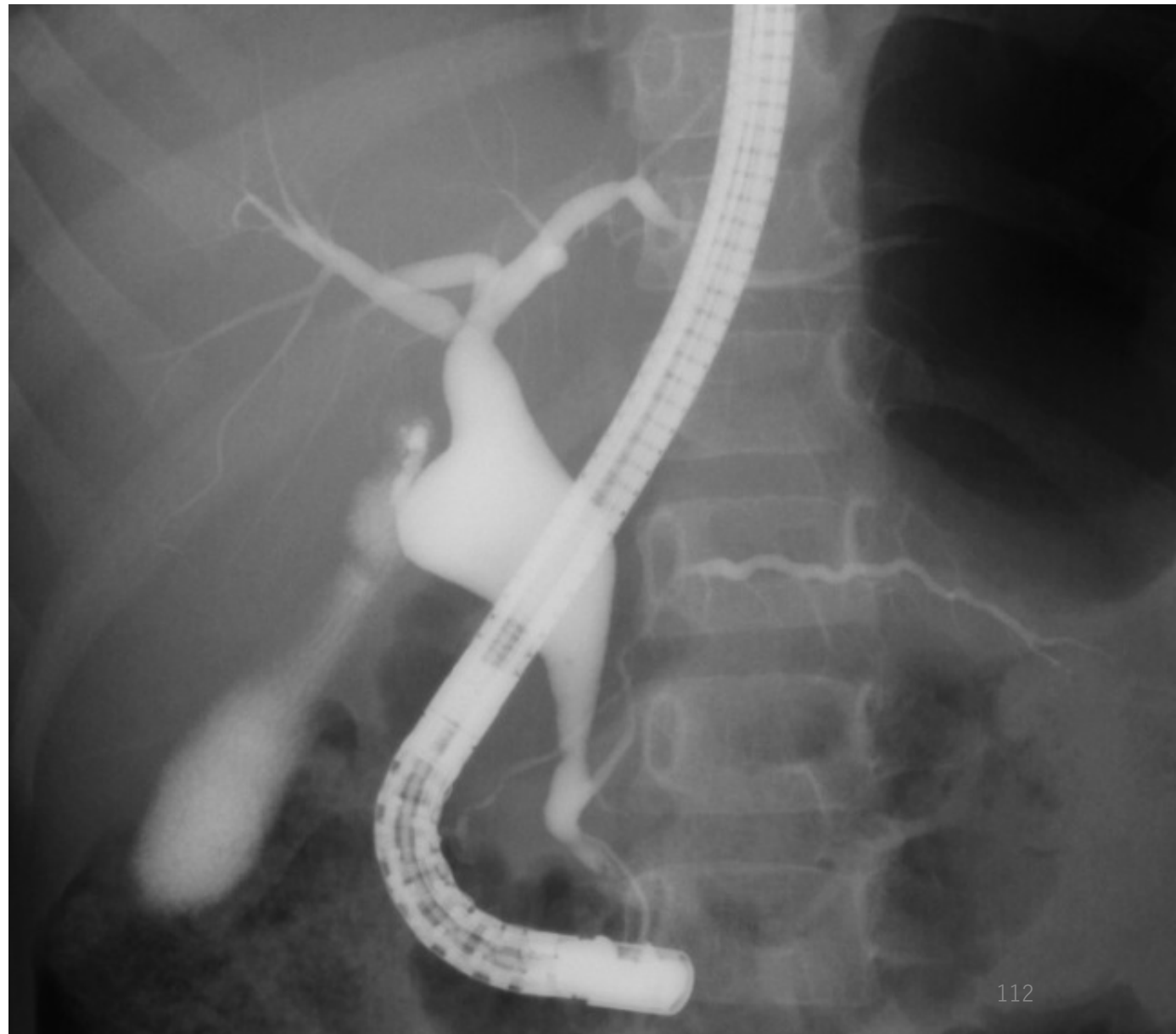
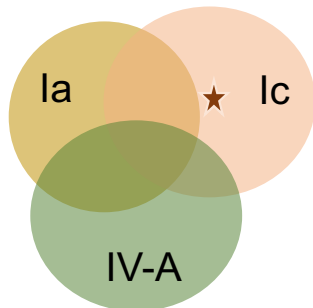
Diffusely dilated RHD but without strictures

Case 34

Ia<Ic

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

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



Advanced

Nondilated type of PBM

Definition, pathophysiology, and diagnosis

First version, September 2025

Imaging features of congenital biliary dilatation

Features that define CBD

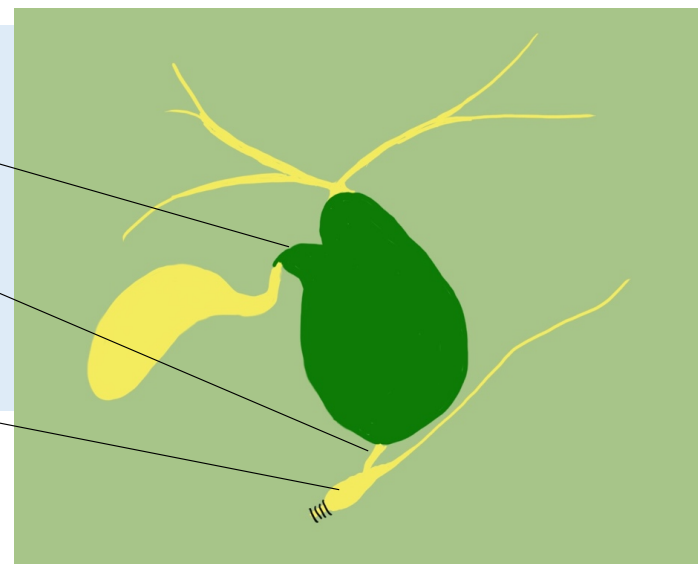
Introduction section

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

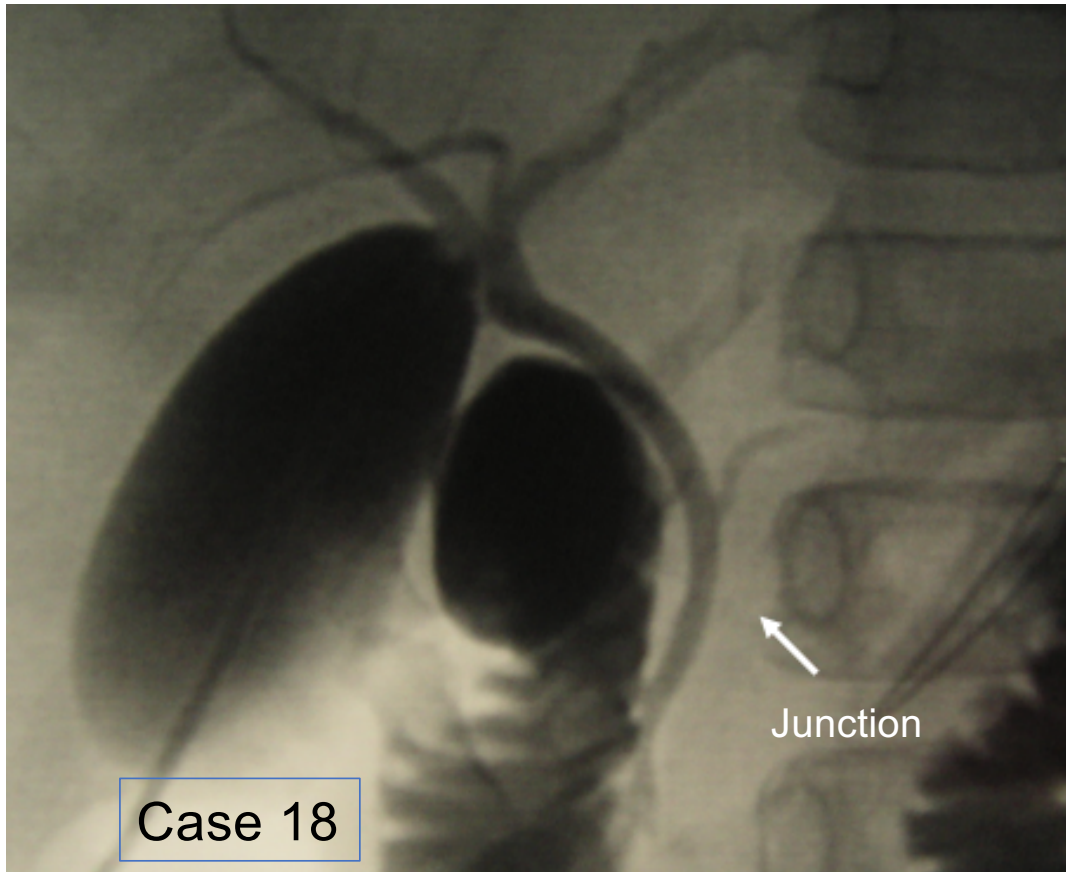
Features (but not in all cases)

Basic section

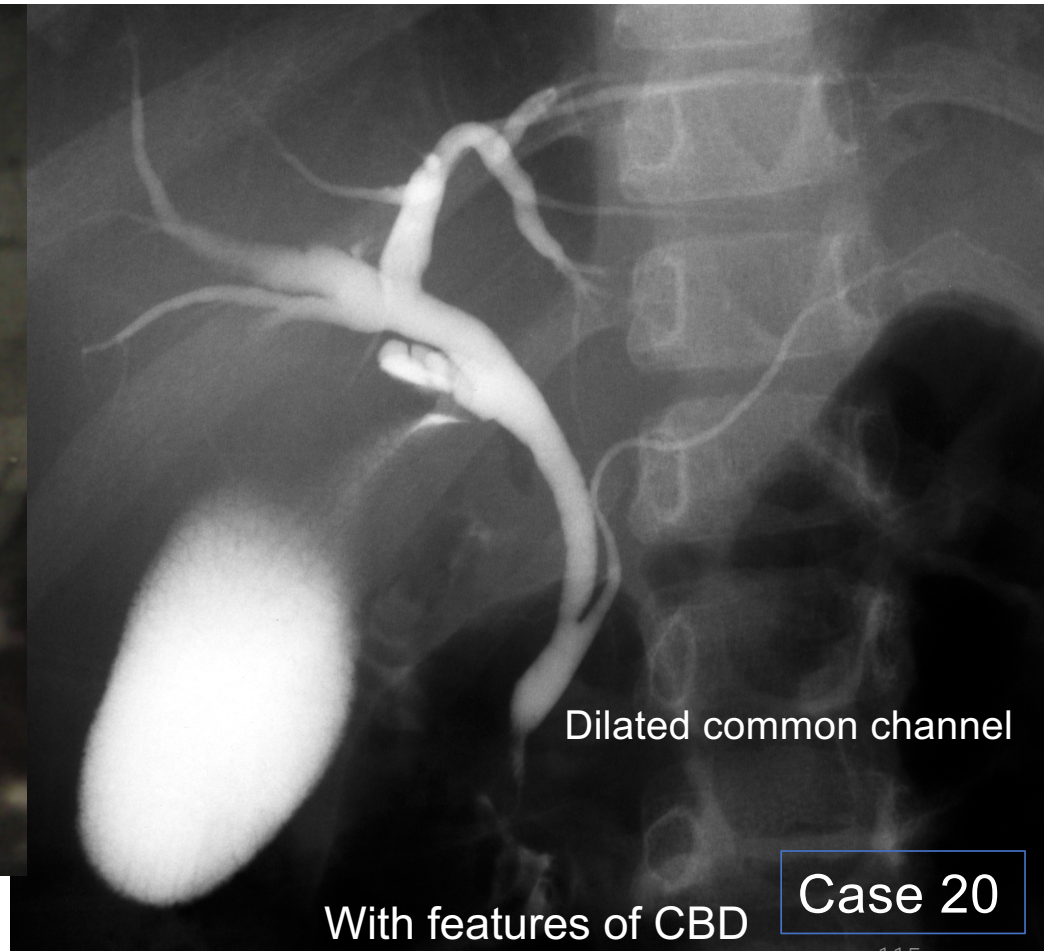
- Dilatation at the base of the cystic duct
- Narrow segment below the bile duct
- Dilated common channel



PBM with normal BD diameter Pediatric cases

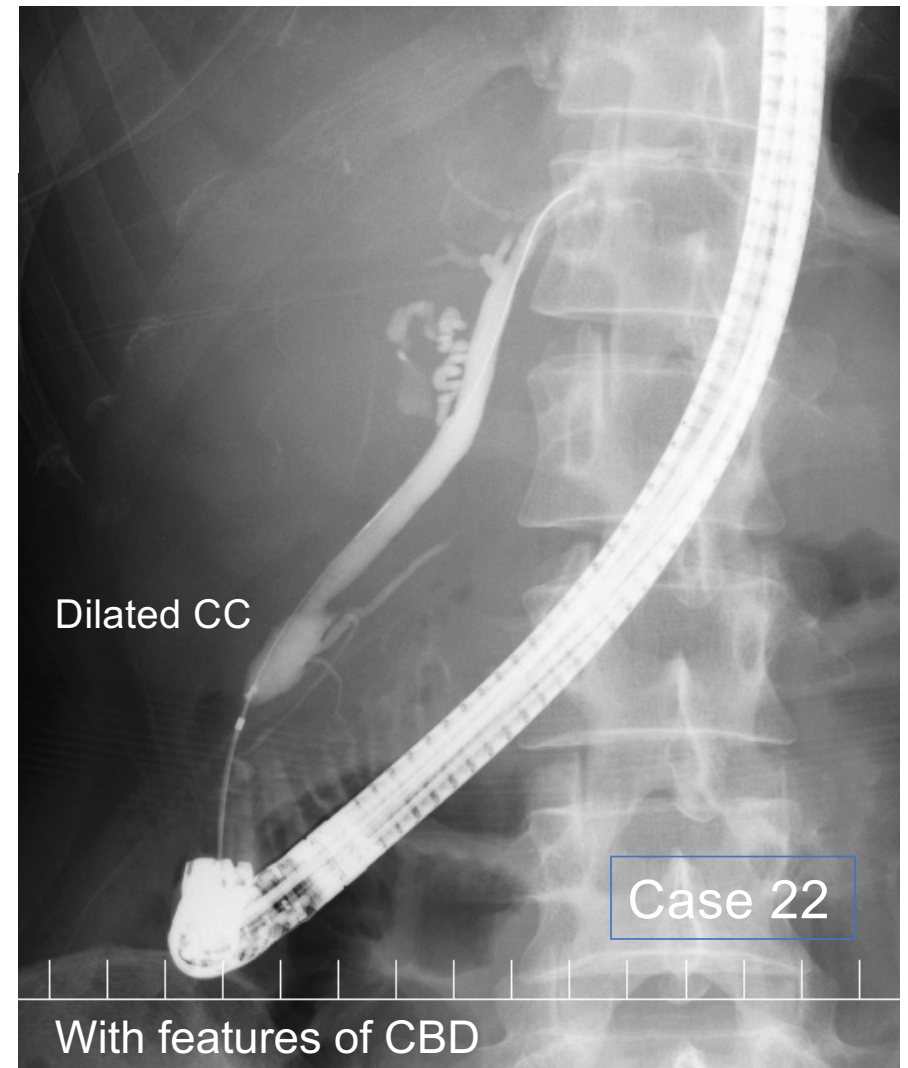
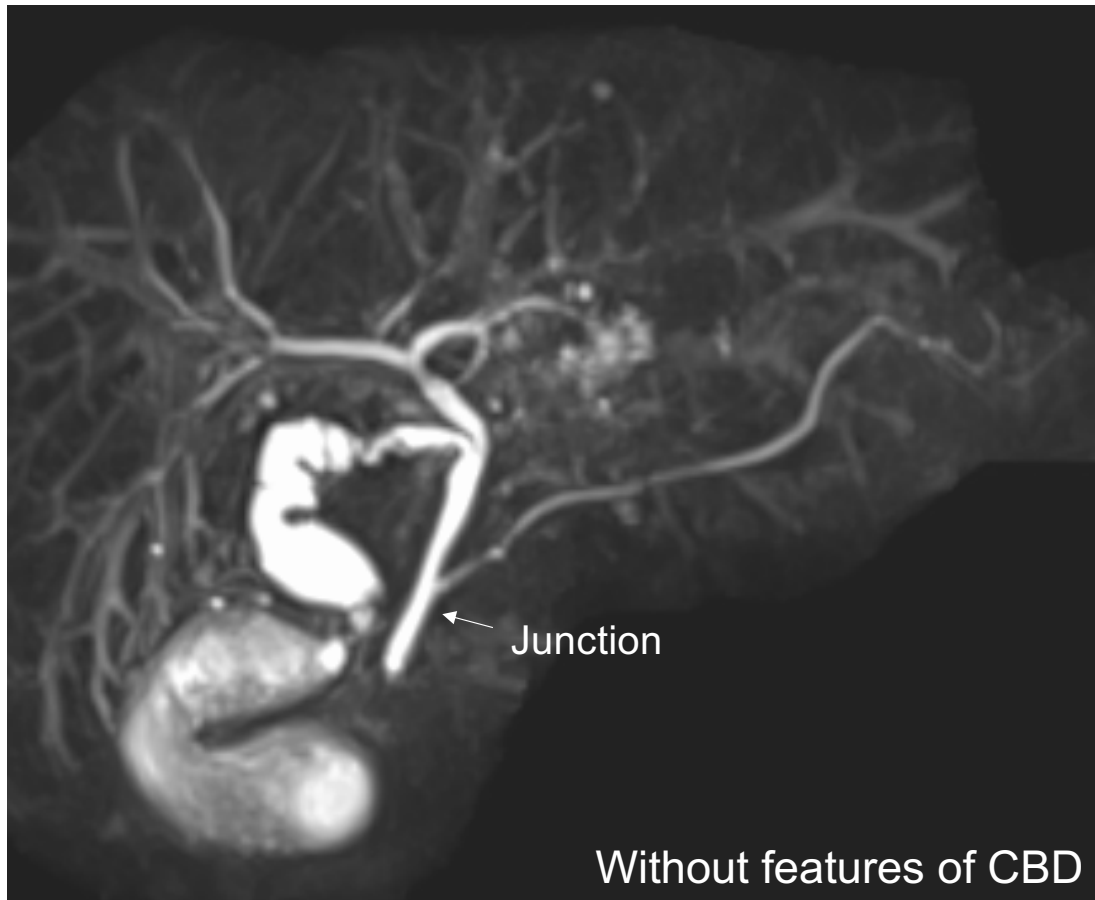


Without features of CBD



With features of CBD

PBM with normal BD diameter Adult cases



Case 35

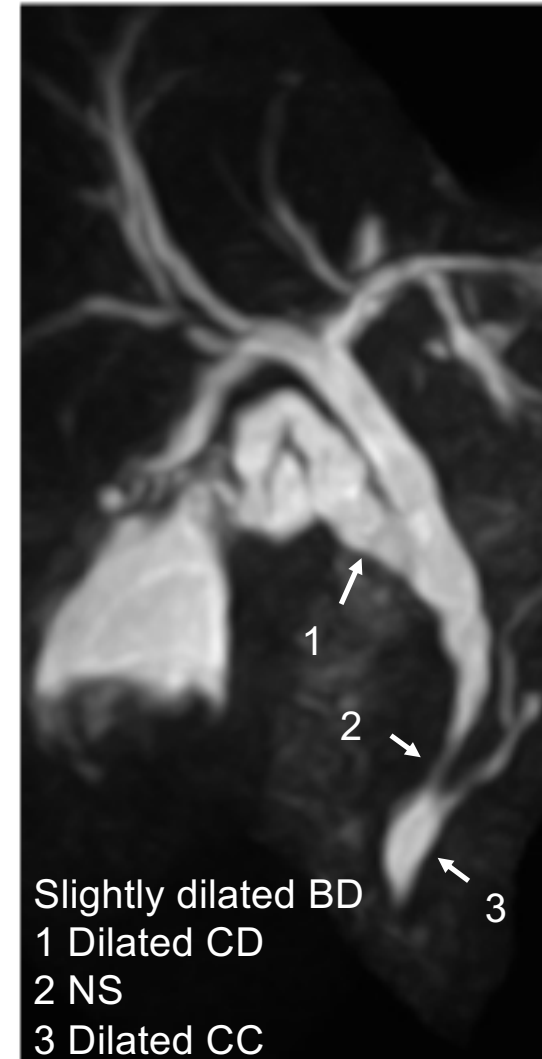
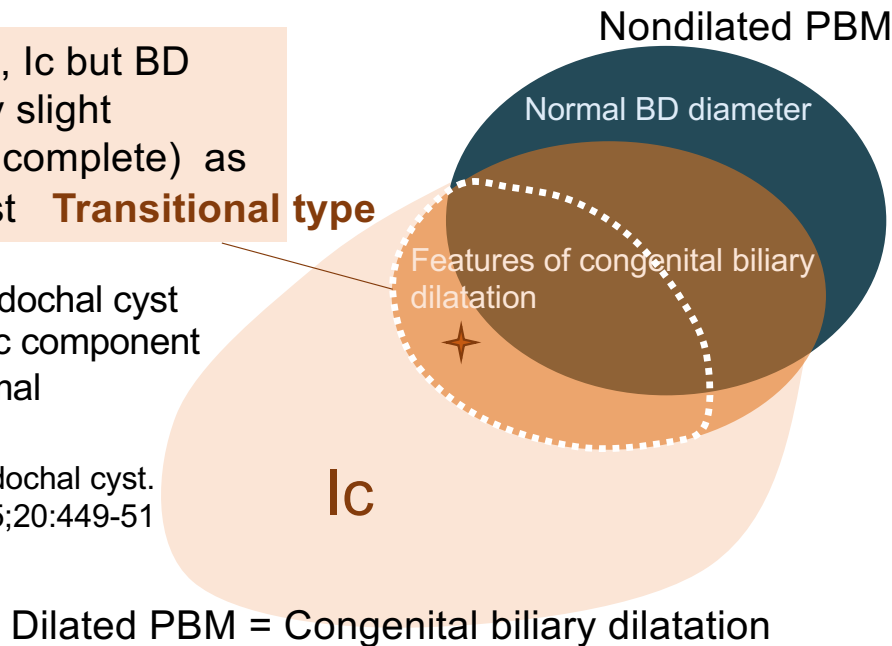
Forme fruste (Ic) Adult case

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

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

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

Lilly JR et al.
Forme fruste choledochal cyst.
J Pediatr Surg 1985;20:449-51



Features of congenital biliary dilatation

Case 36

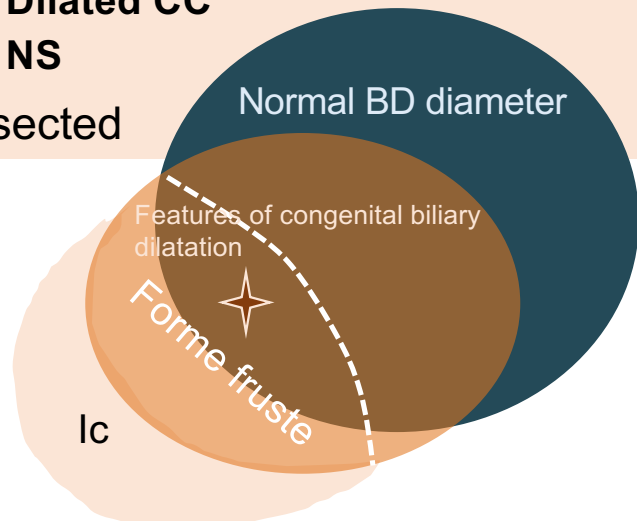
Pediatric case Normal BD diameter

Nondilated PBM < Forme fruste

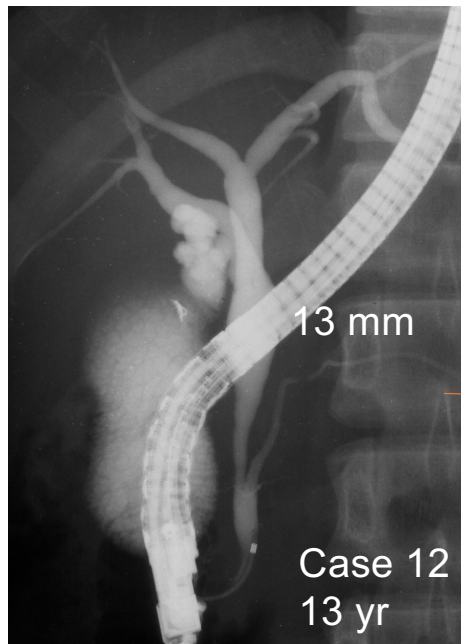
15-year-old girl
Ultrasound due to abdominal pain detected
gallbladder wall thickness

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

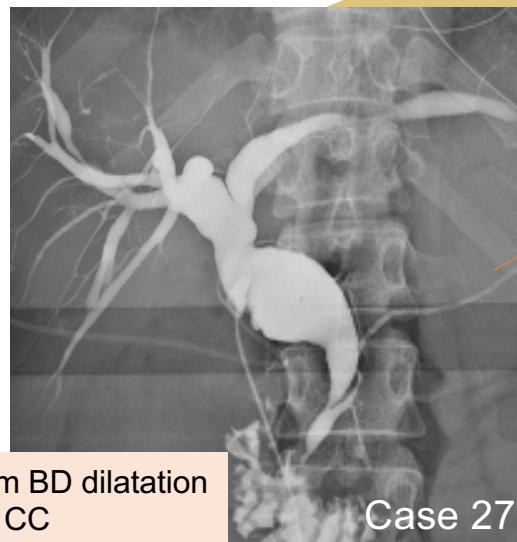
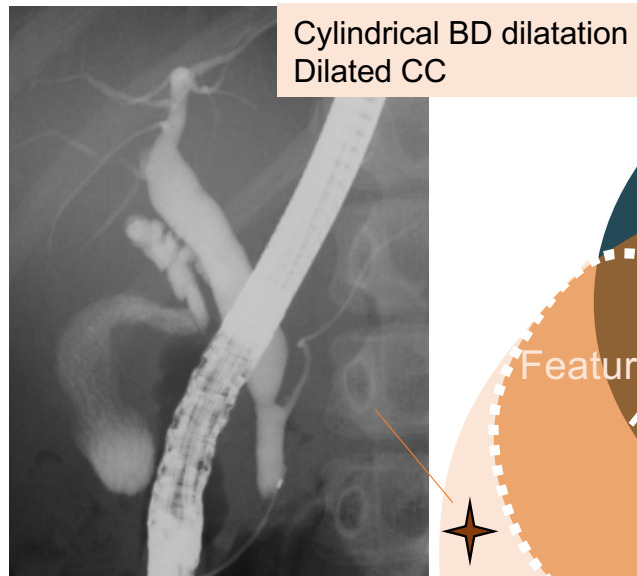
BD resected



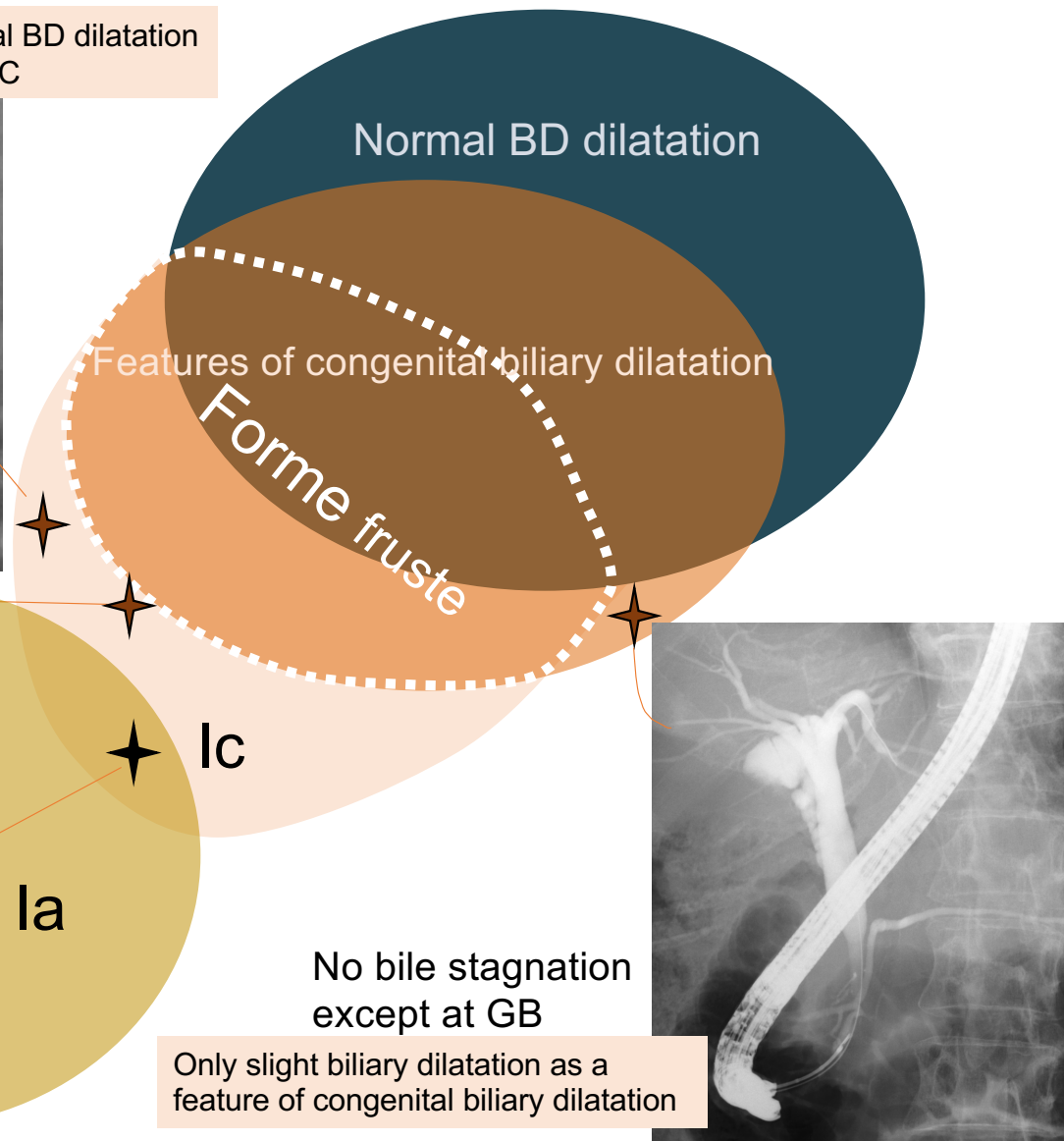
Around Forme fruste



Fusiform BD dilatation
Dilated CC
NS

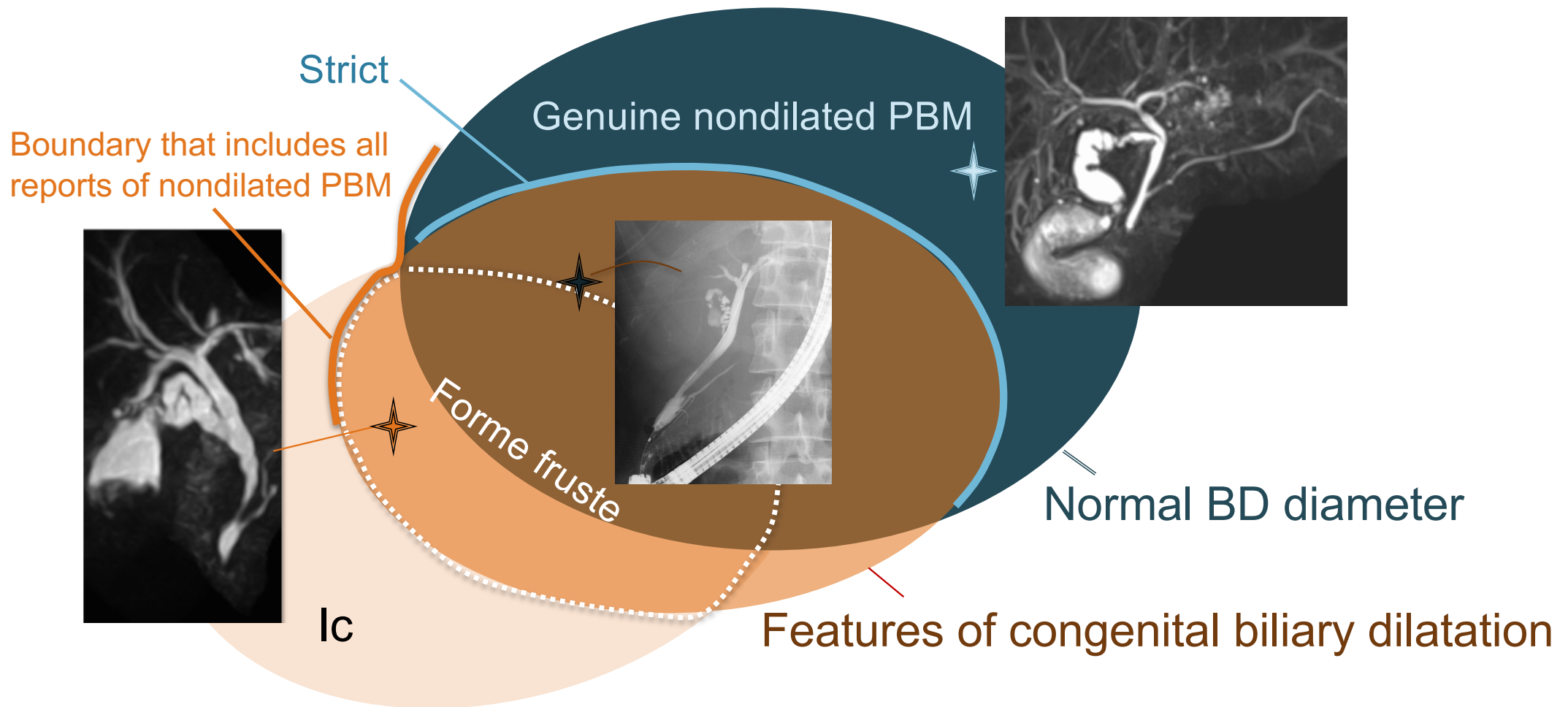


Fusiform BD dilatation
Dilated CC



Definition of Nondilated PBM

Where to draw the boundary on this spectrum?

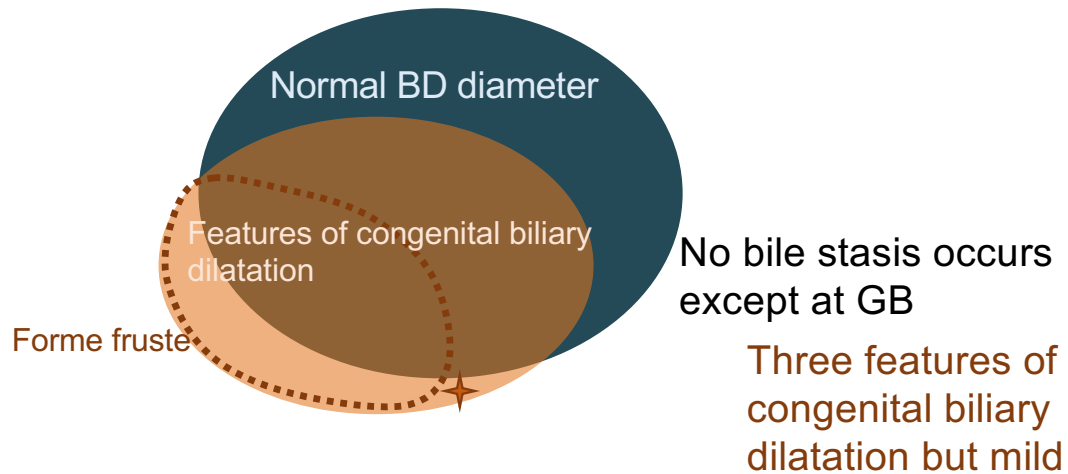


Case 37

Nondilated PBM >Forme fruste Pediatric case

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

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

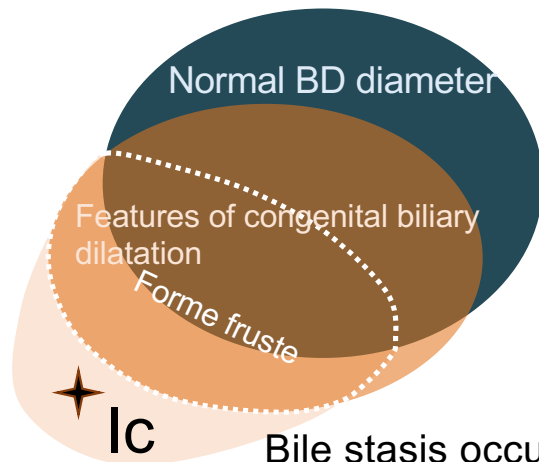


From basic section

Case 14

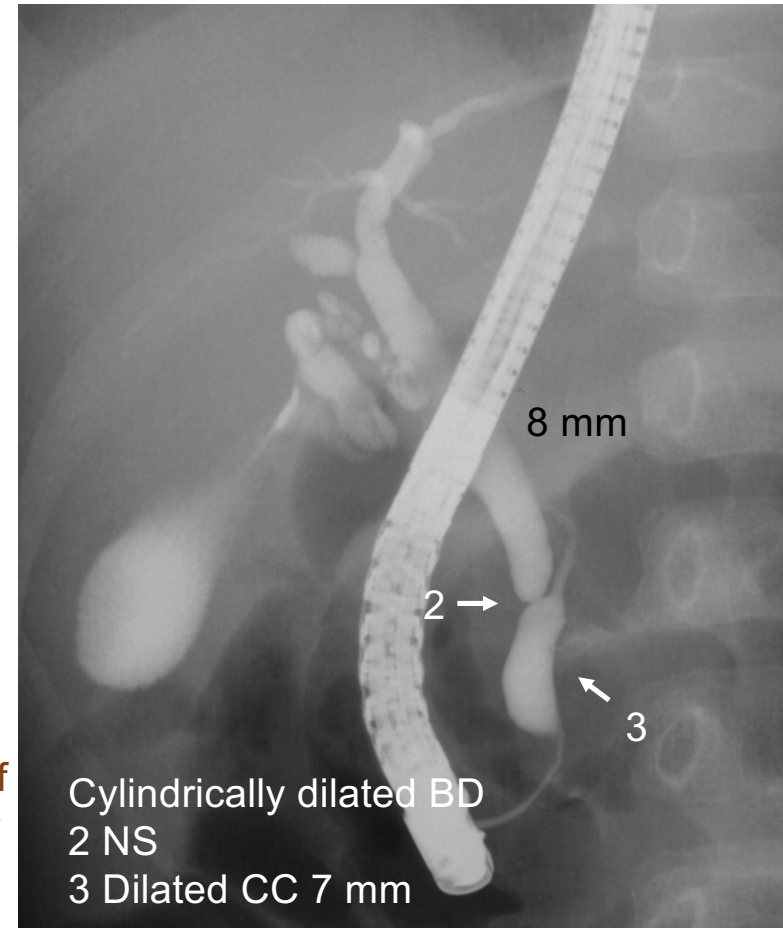
Ic Cylindrical dilatation Pediatric case

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



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

Three features of
congenital biliary
dilatation



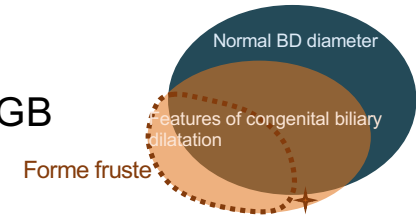
2 yr old, US upper limit 3.3 mm

Case 38

Adult case

Nondilated PBM > forme fruste >> Ic

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



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

BD diameter
MRCP 8 mm

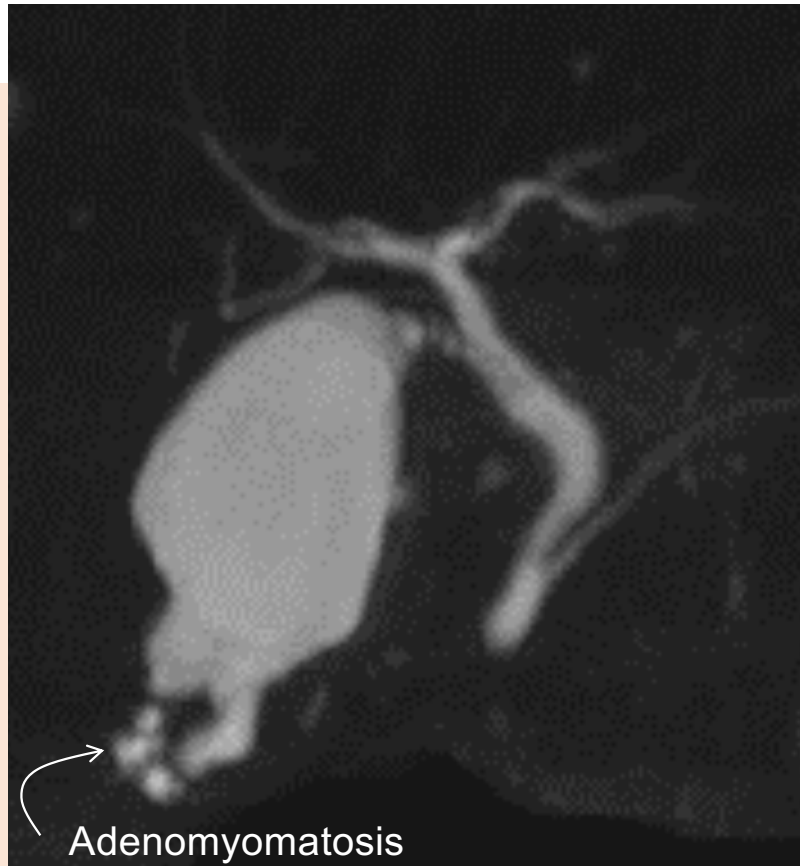
ERCP 9 mm

US upper limit 7.2 mm

Features of congenital
biliary dilatation are
mild

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

Cholecystectomy



Advanced

PBM seemingly complicated

Rare type of PBM

First version, September 2024

Case 39

Protein plugs complicate PBM

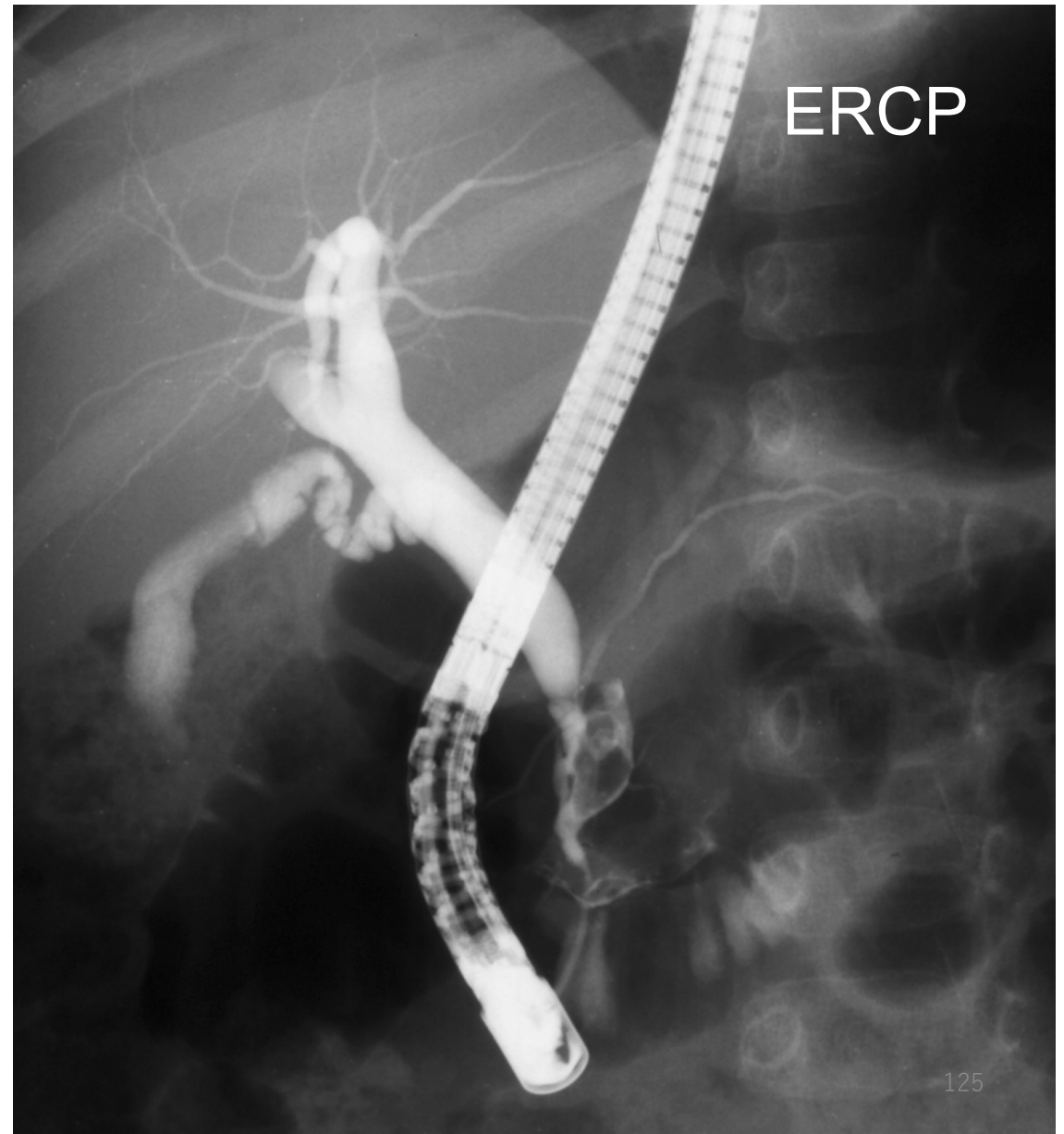
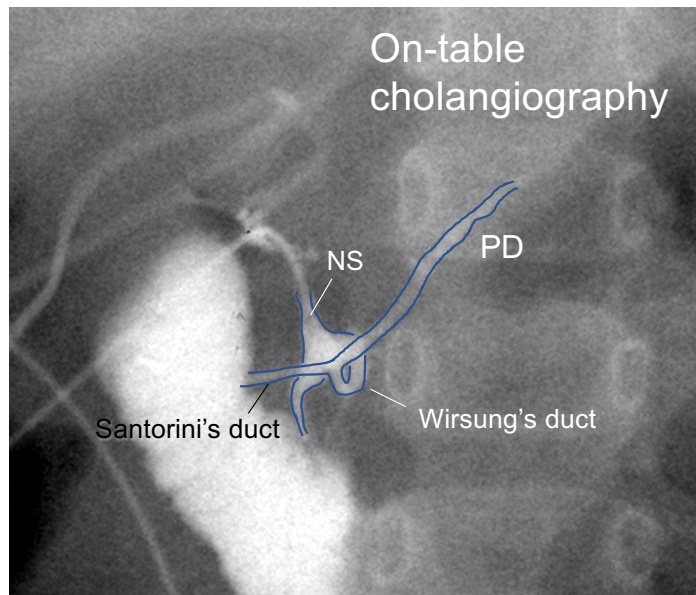
A 3-year old girl developed abdominal pain and vomiting, and jaundice after five days

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

She underwent BD resection after symptoms subsided.

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

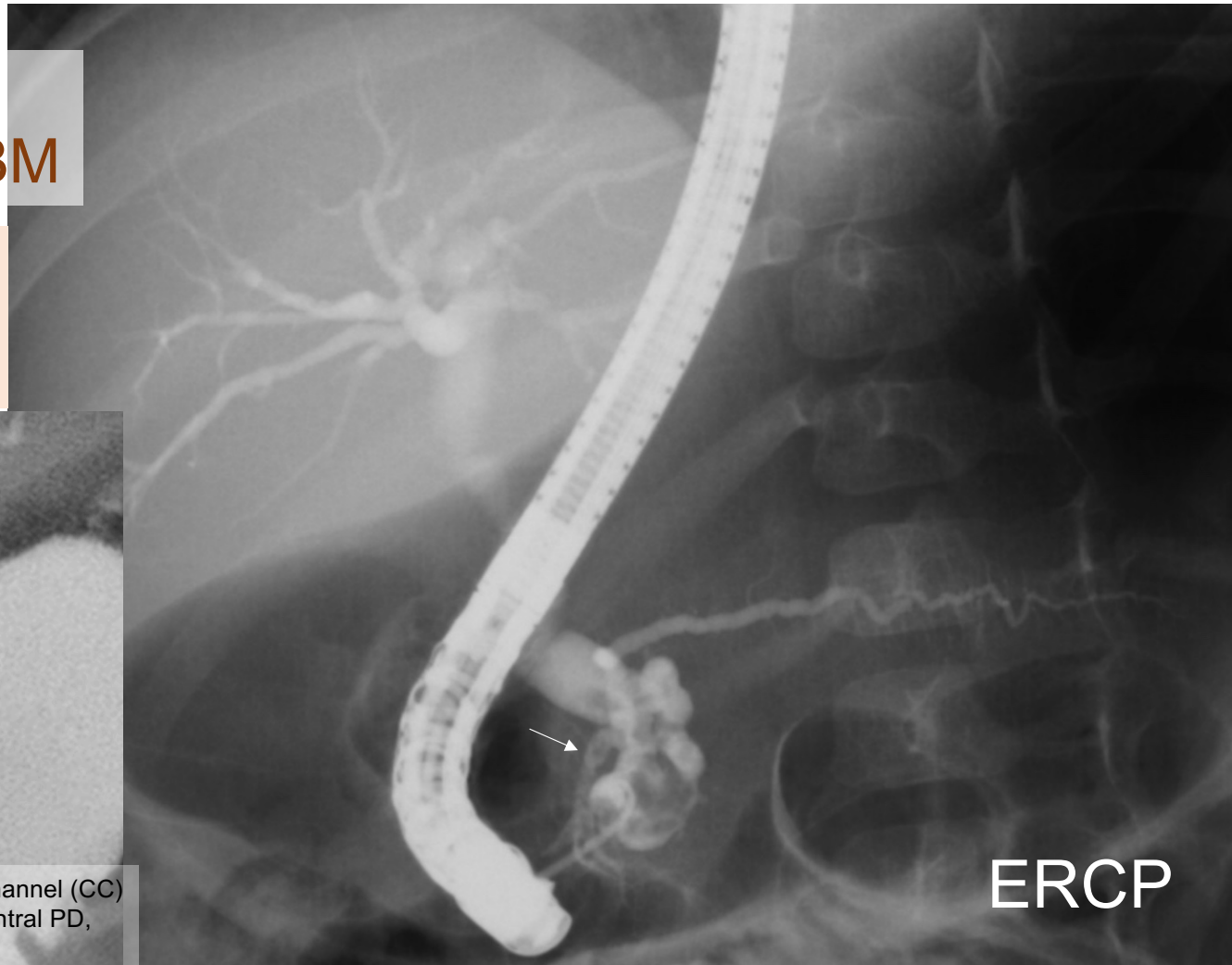
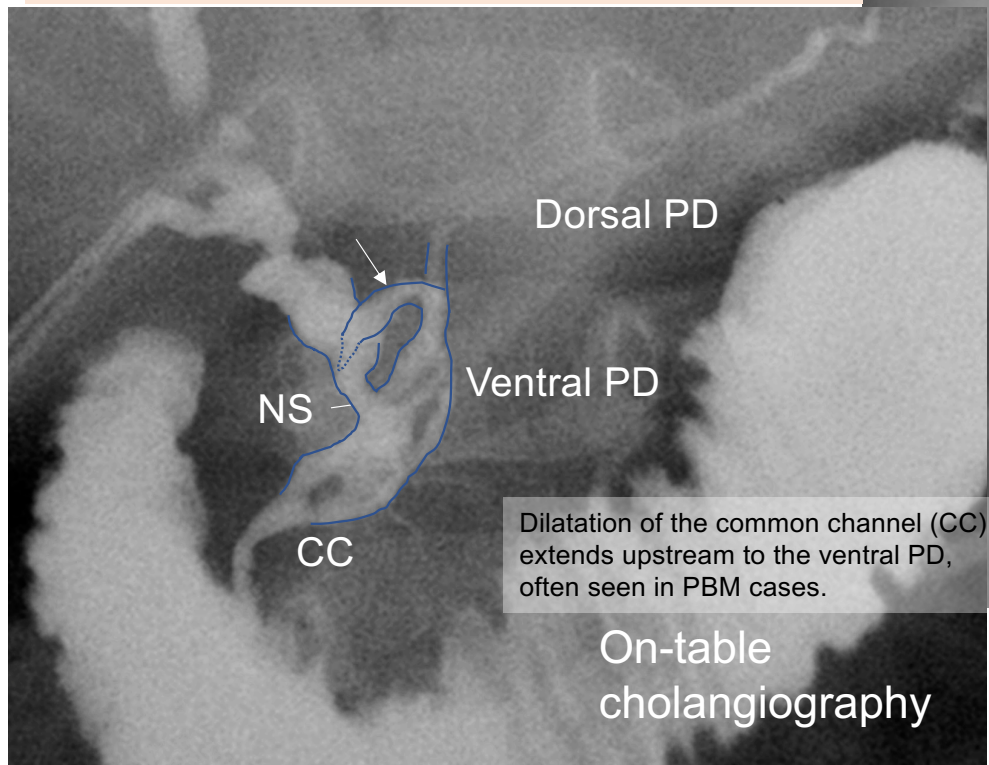
Dilatation of the common channel had disappeared.



Case 40

Protein plugs complicate PBM

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



Arrows indicate the same PD branch.

Case 41

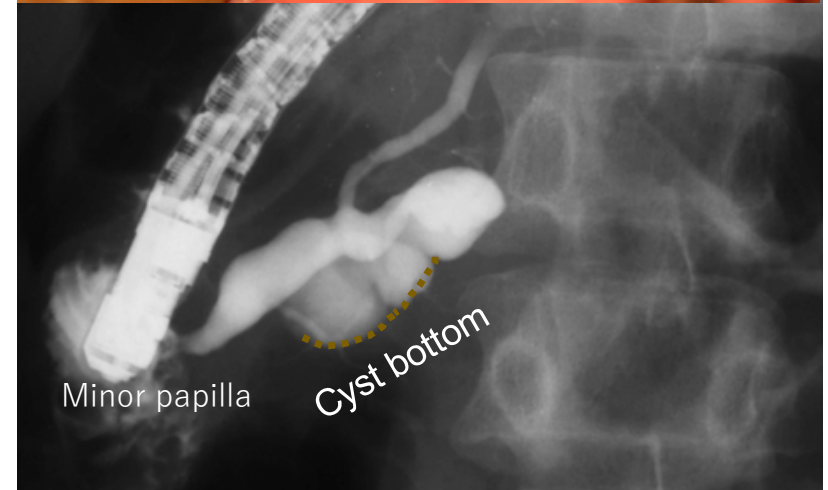
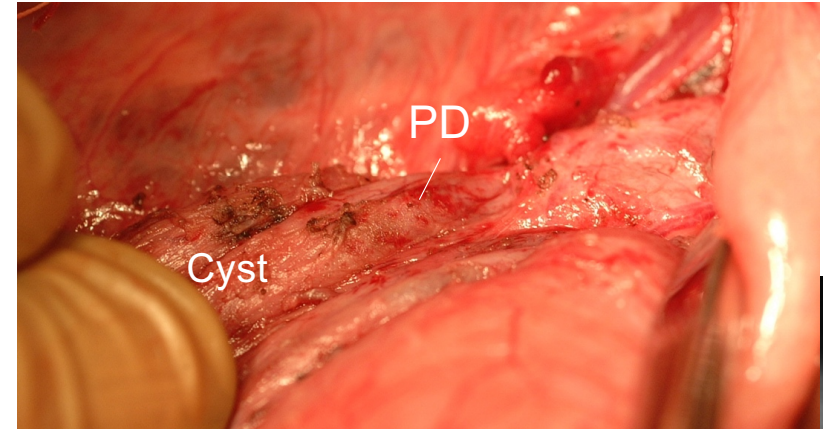
A female in her twenties developed abdominal pain with hyperamylasemia caused by protein plugs.

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

Main (Ventral) pancreatic duct forms cyst bottom



Dotted line of the cyst was composed of PD



Pancreatography through the minor papilla after cyst excision

Case 42

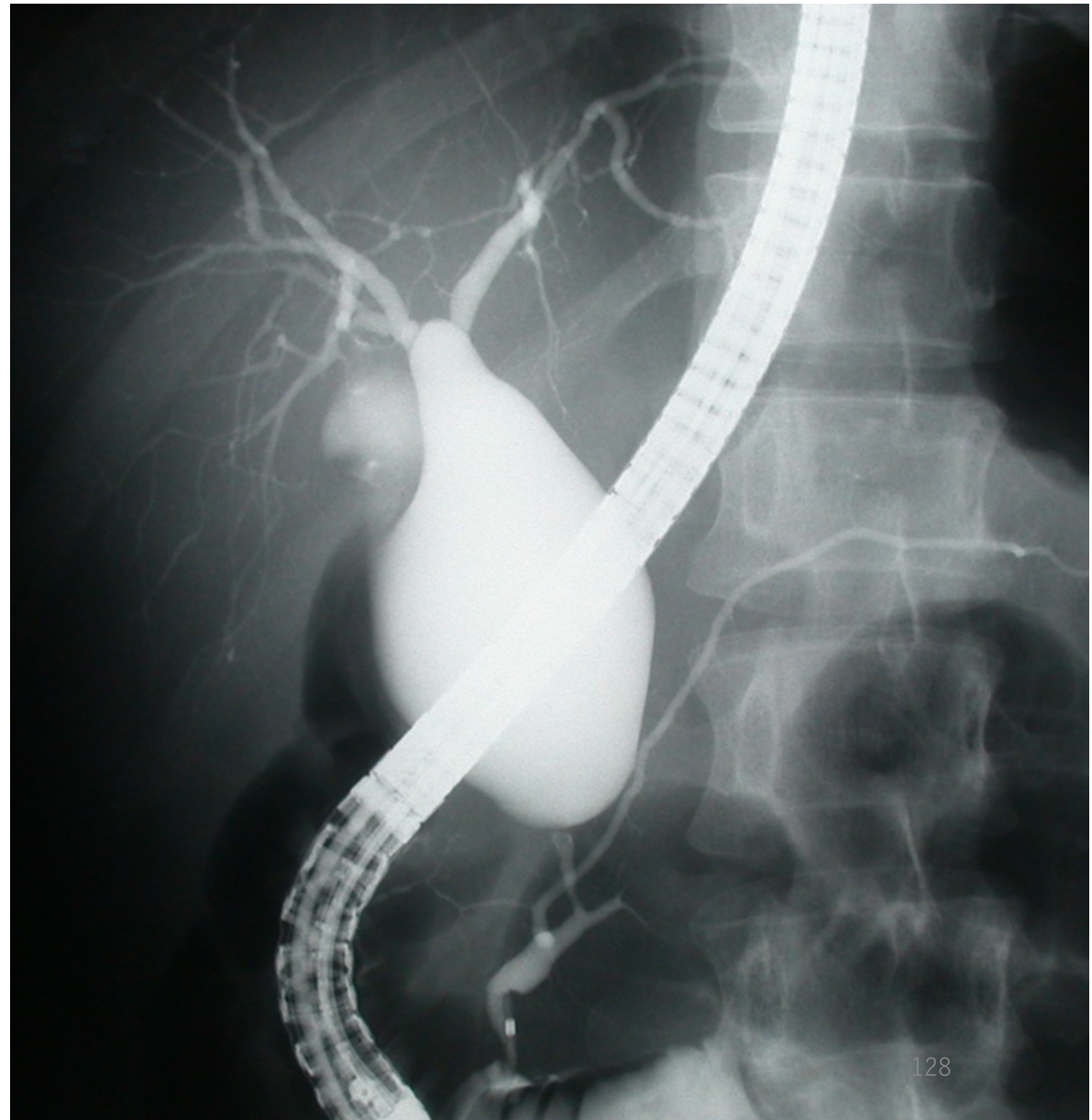
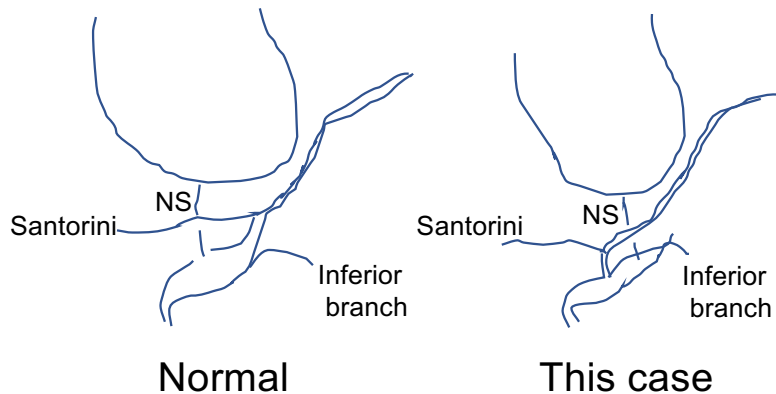
Rare type Bile duct joins the inferior branch of PD

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

Type Ia congenital biliary dilatation

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

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

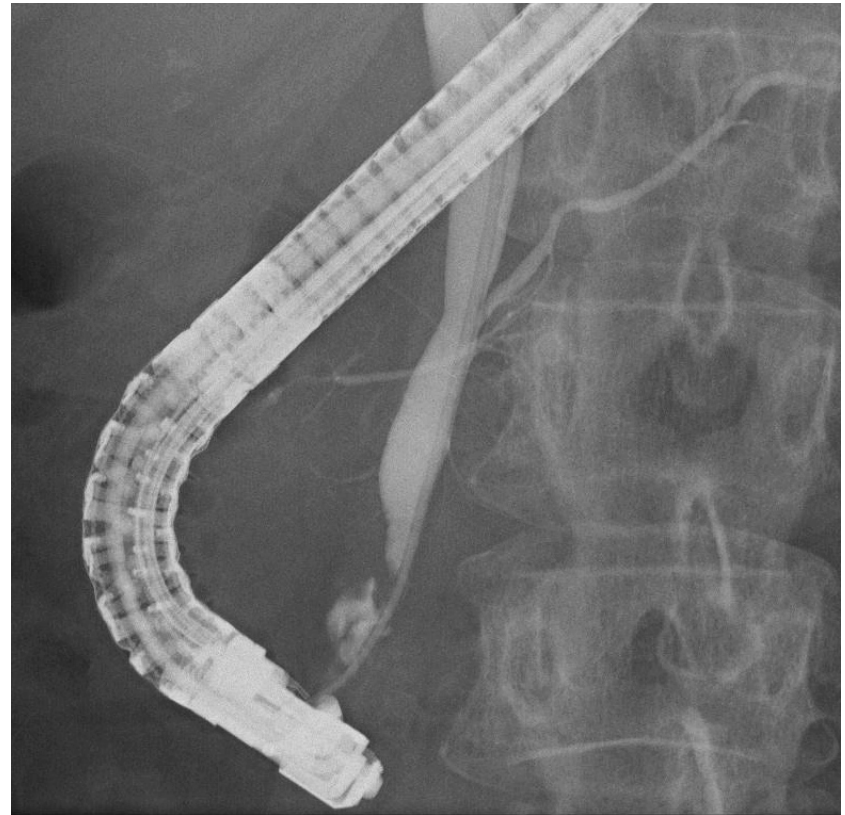


Advanced

Common channel & Narrow segment

First version, September 2024

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



PD branches coming out of the common channel support it is a PD.

Dilated common channel composed of both PD and BD

Proposed by Ando H in 1999

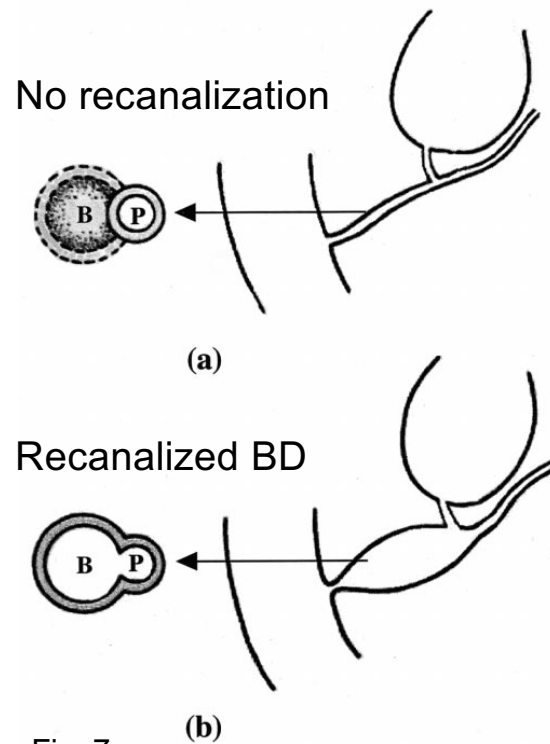
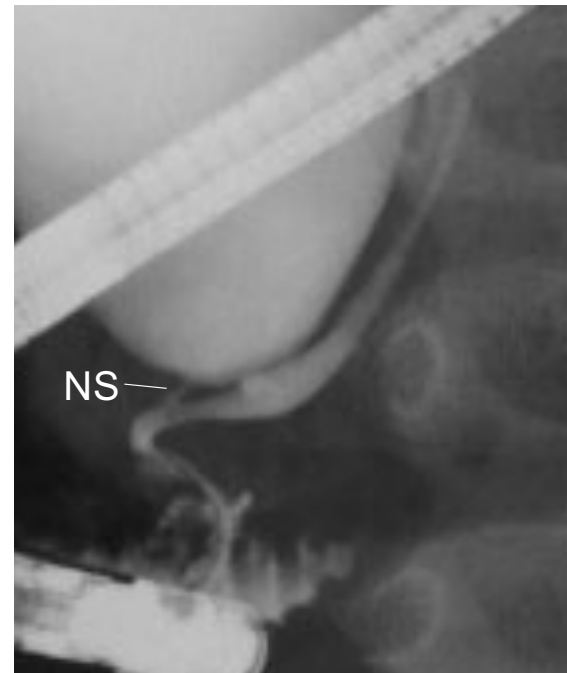
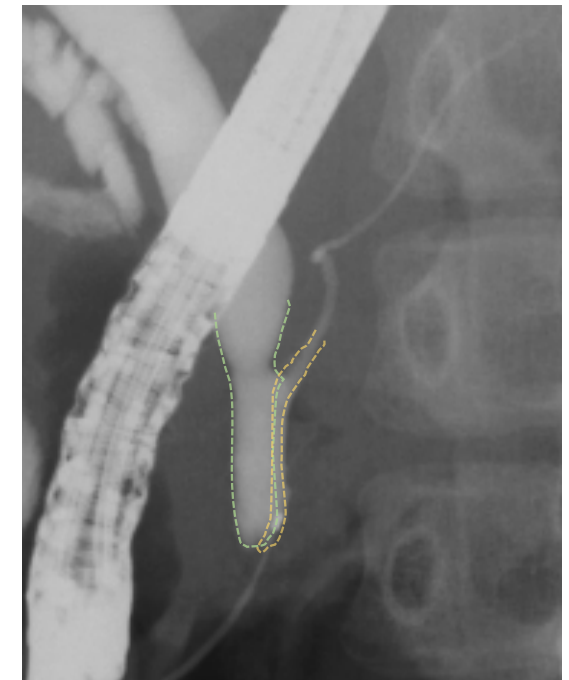


Fig. 7

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):50-4



PD alone



BD+PD

Case from *Basic Section*

See P43.

NS is a branch of 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
levels of pancreatic
enzymes

Radiological demonstration

Cystic BD

Santorini's duct

NS

Wirsung's duct

NS is one branch
of PD



Is NS really a PD branch?

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

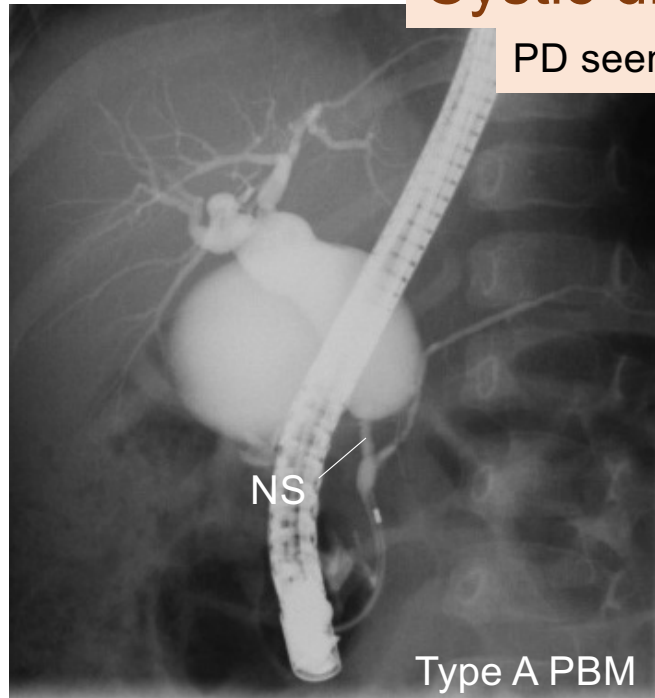
- JSGBPBM classification (See P33.):
Type B, non-stenotic type rather than Type A, stenotic type
- The BD should be resected completely down to the PD using fluoroscopy with clips. See Next and P70.



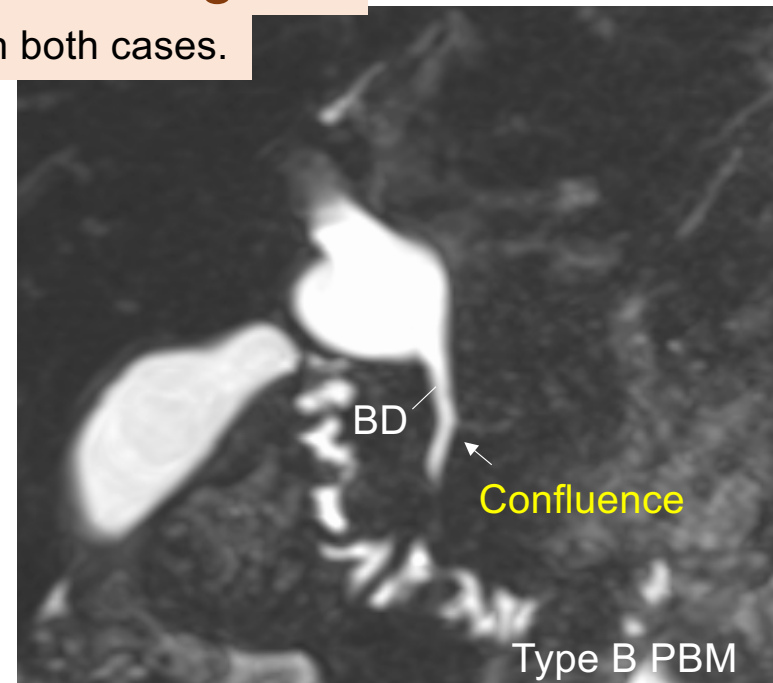
Is NS really a PD branch?

Cystic dilatation with a long NS

PD seems to enter BD in both cases.

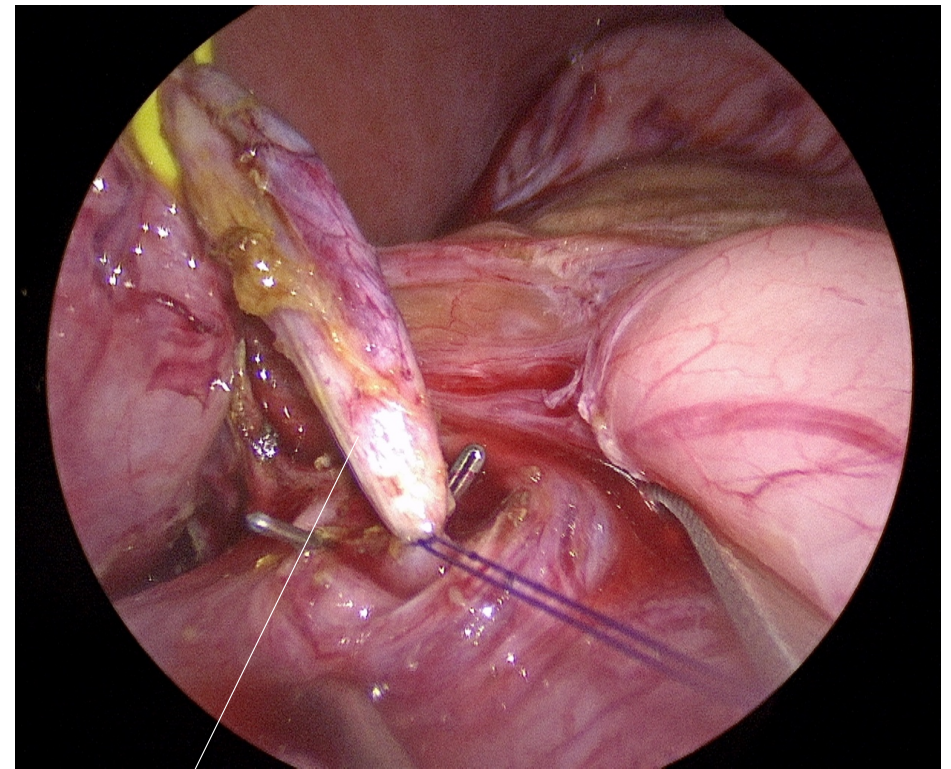
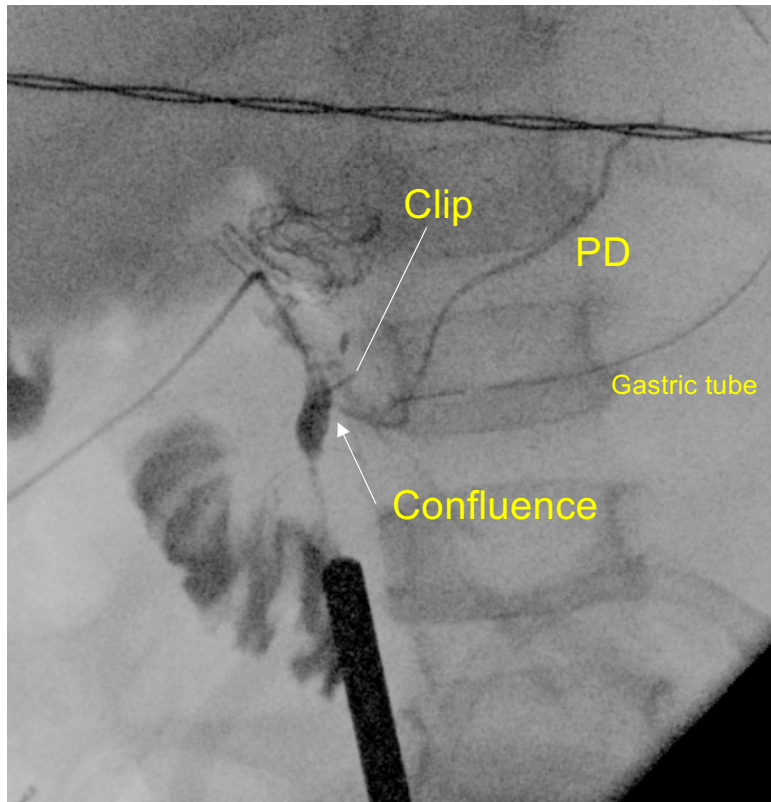


One-year-old child
Vomiting, elevated serum levels of liver enzymes and amylase
BD resected at the obvious NS.



Three-year-old girl
Abdominal pain, vomiting, and elevated serum levels of liver and biliary enzymes
No abrupt narrowing needed using fluoroscopy with clips. See next.

Surgery showed no NS.
BD was resected just above the PD using fluoroscopy with clips.



The narrowing portion on MRCP was 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

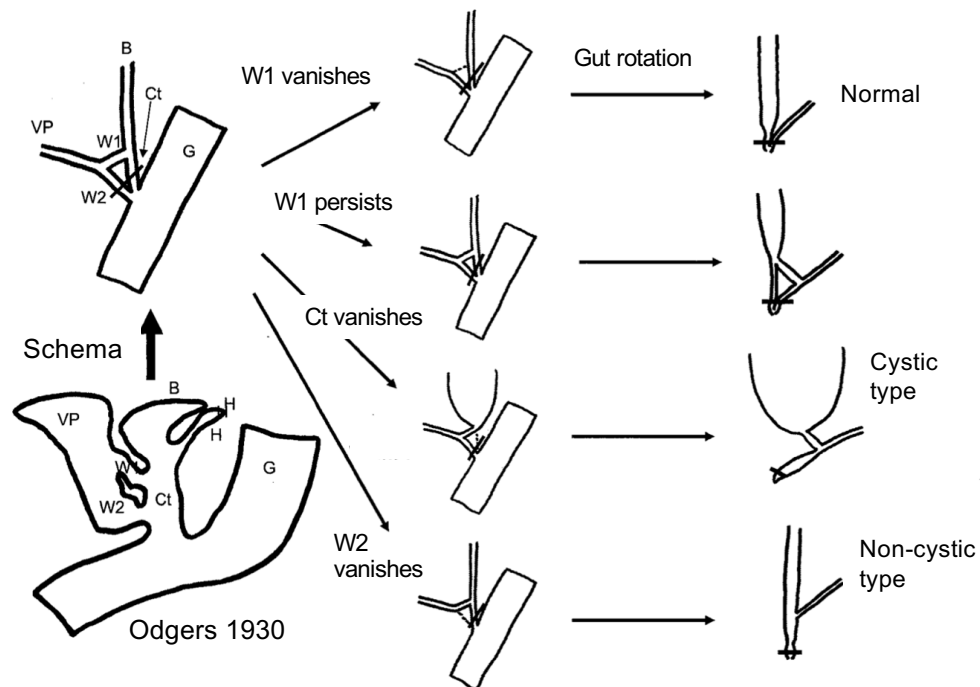
Adult case
Loop type of PD

Portion of BD
Portion of PD branch

Surgery confirmed NS

Matsumoto's theory of PBM embryogenesis:
PBM is the malformation where the choledochus misconnects with the ventral PD or its branches outside the duodenal wall. See next.

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 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 BD never enters a dorsal PD in PBM

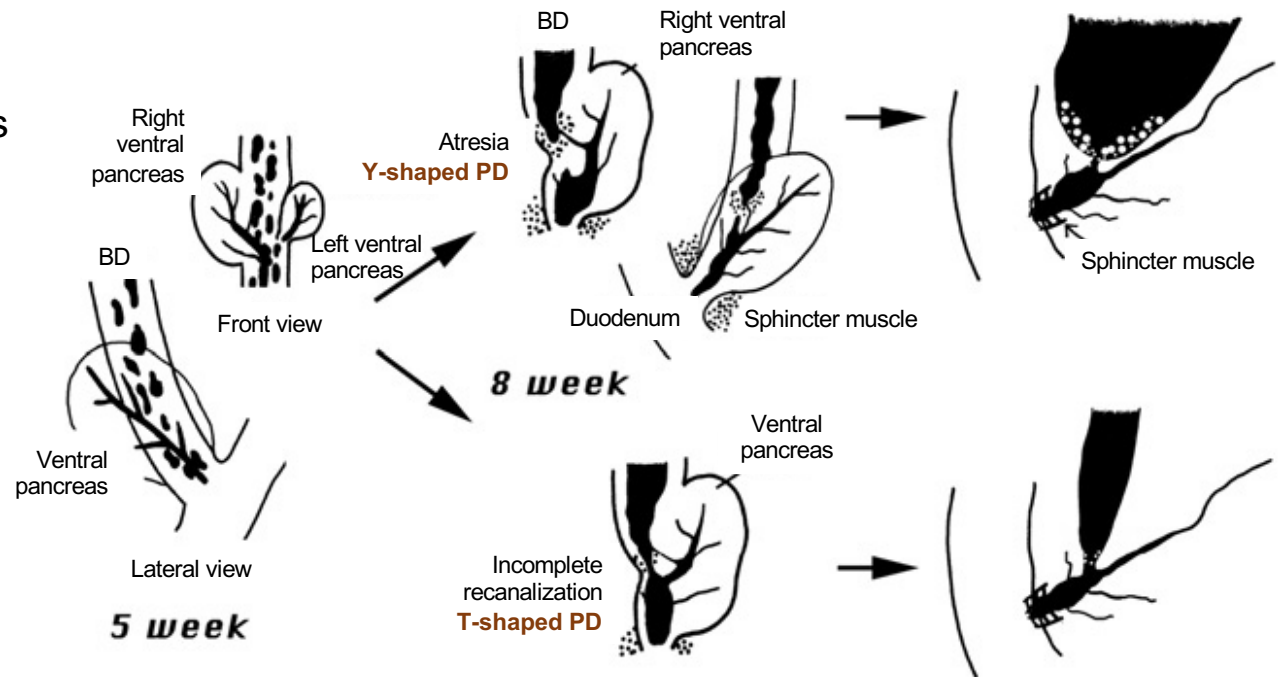
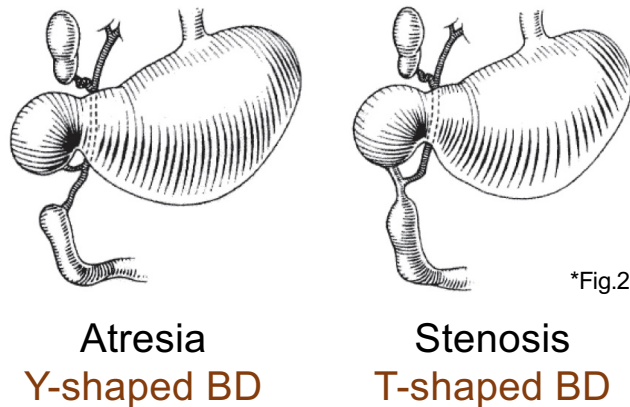
He thought that PD branches coming out of the common channel (CC) on cholangiopancreatography (See P115.) suggested that the CC was a PD. He estimated that PBM is the malformation where the choledochus misconnects with the ventral PD or its branches outside the duodenal wall.

See Cases 9* and 42**.

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



Cited from Hisami Ando, Tando
(Biliary Tract) 2008;22:175-80

Inferred from embryogenesis of congenital duodenal atresia and stenosis
BD has a solid stage during its development as well as the duodenum,
while PD does not have a solid stage.

Because almost all congenital diseases with limited BD dilatation have PBM, there must be embryological relationship between them. However, does regurgitation due to PBM really cause biliary dilatation?



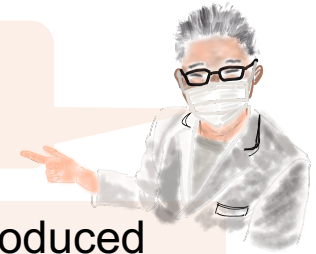
Advanced

What causes biliary dilatation?

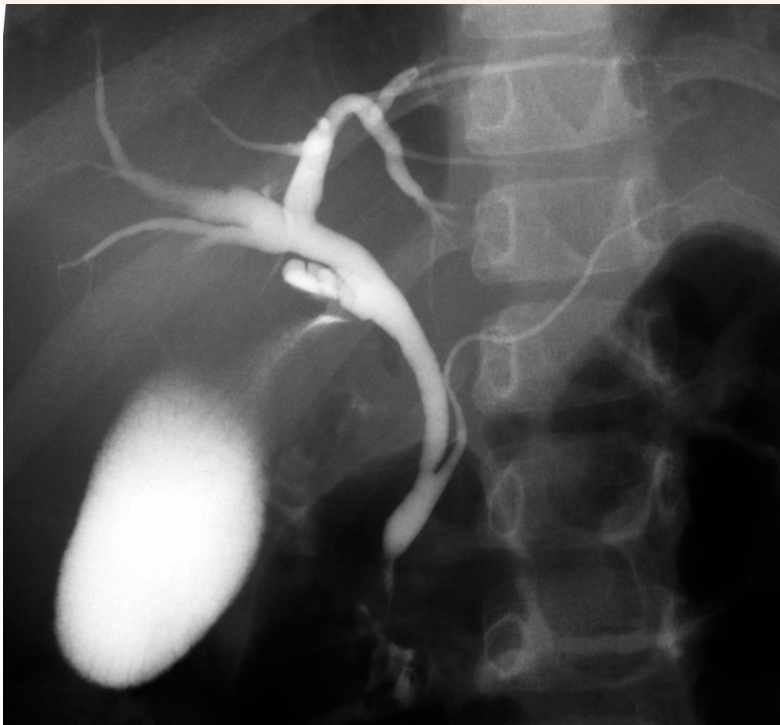
First version, September 2024

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.



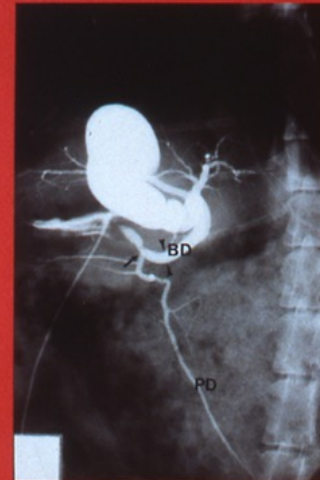
Non-dilated type of PBM exists



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

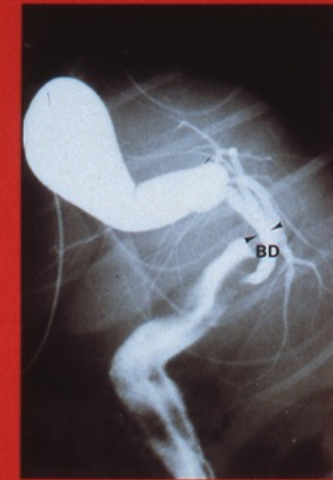
Cystic dilatation was not produced in animal PBM models

Cholangiography



PBM model

2.72 ± 0.60 (mm)



Control

2.33 ± 0.76 (mm)

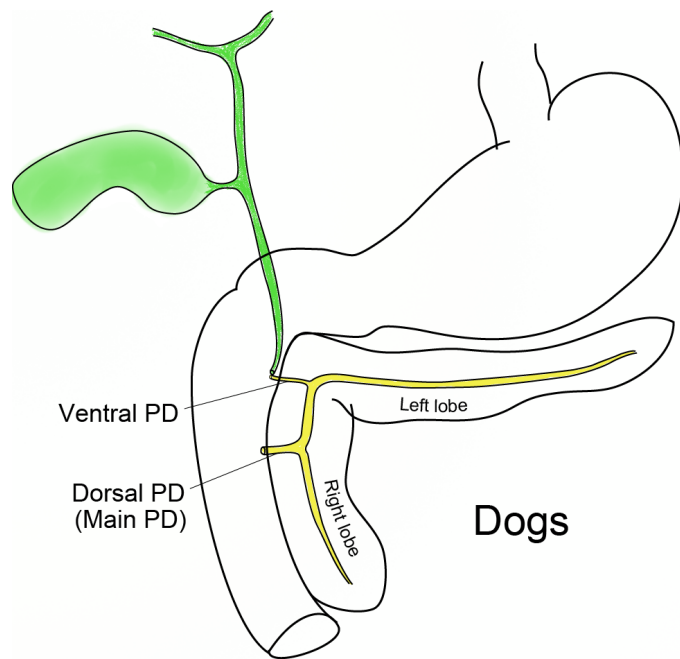
Cat model of PBM
BD did not dilate.

Pancreatobiliary anatomy differs among animals

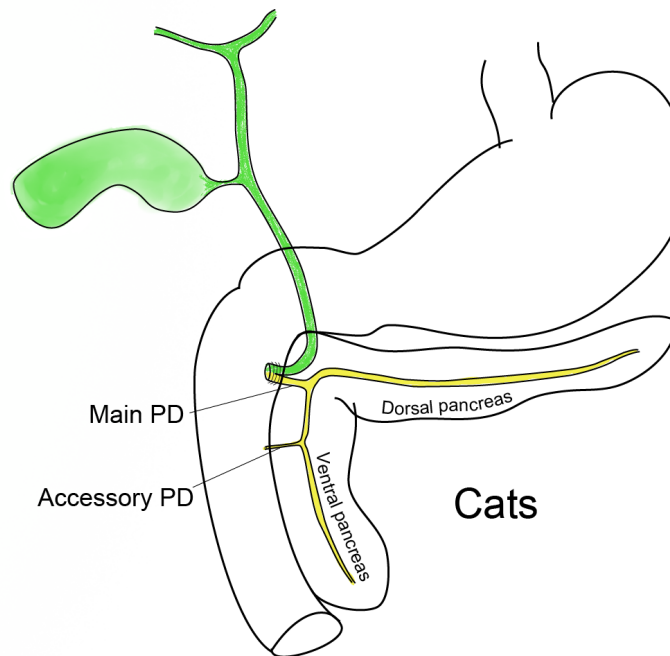
BD joins the accessory PD

Similar to human anatomy

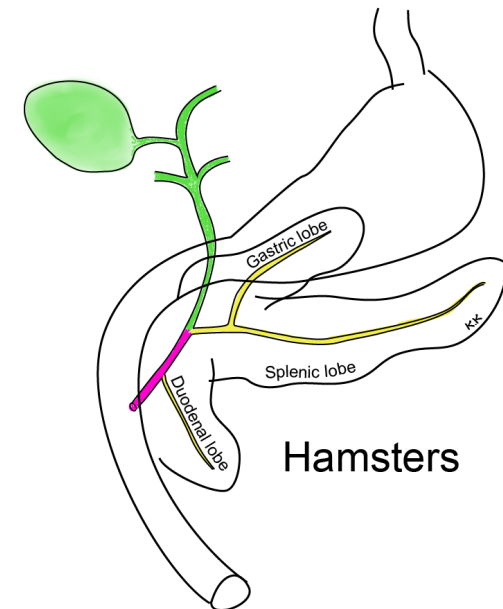
PBM is normal in
rodents, goats, and sheep



Dogs



Cats



Hamsters

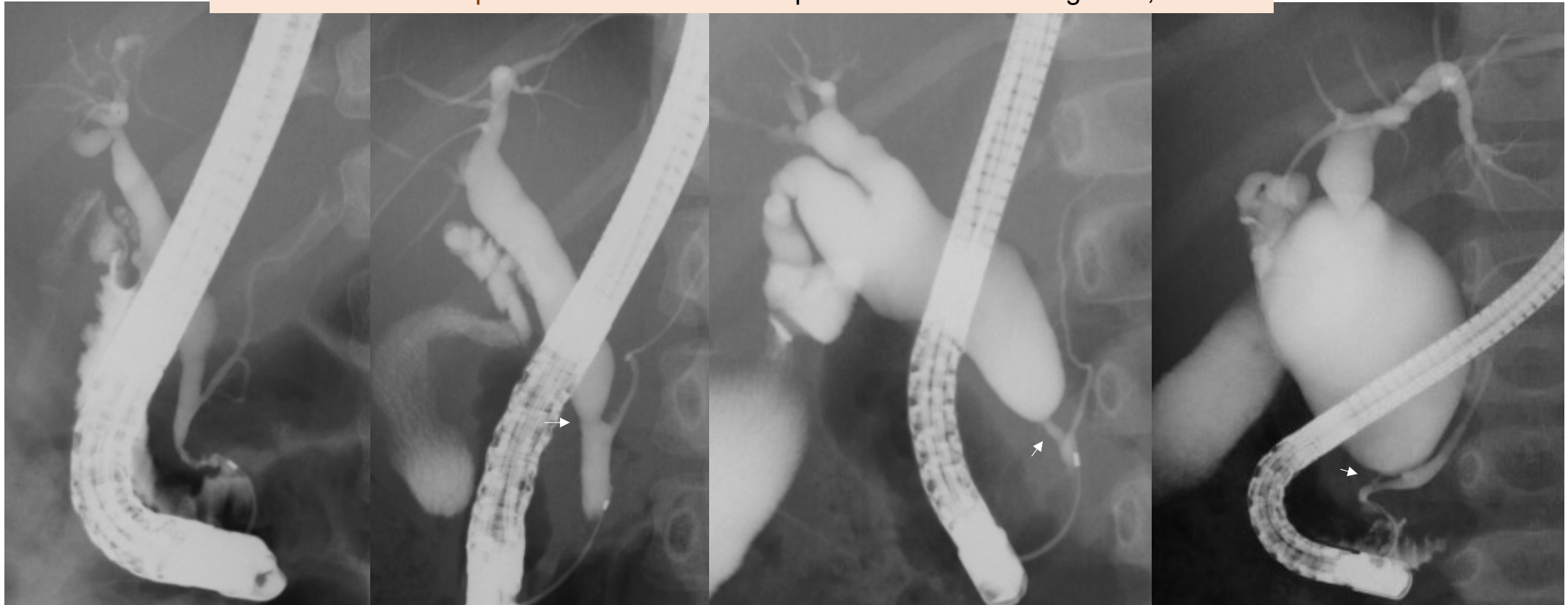
The cause may be the narrow segment (NS)

Dilatation related with NS length

Increased intraductal pressure due to NS

Ando H. Jpn J Surgery 1983;84:1174-85

Davenport et al. J Pediatr Surg 2005;40:331-5



No NS

Slight constriction

Thick NS

Thin NS

NS alone cannot explain characteristic dilatation

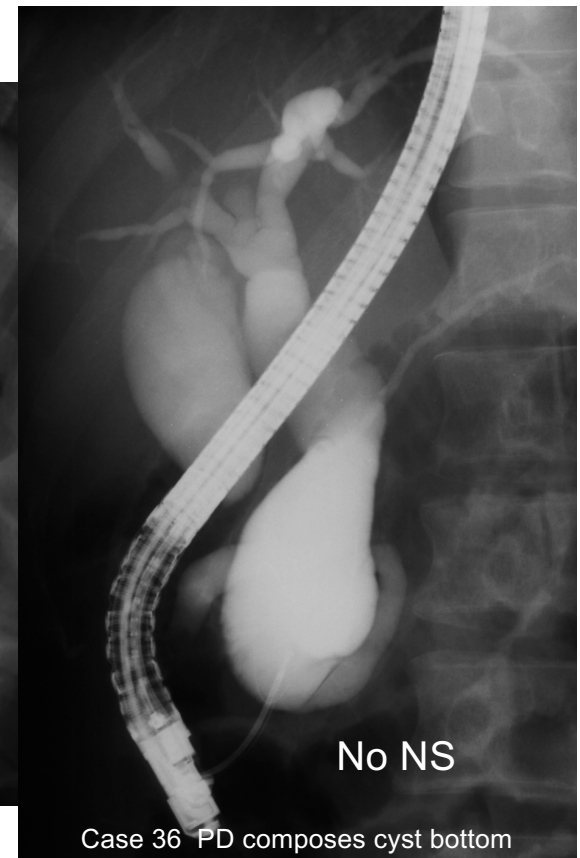
Increased ductal pressure cannot cause limited dilatation See P11.



Limited dilatation

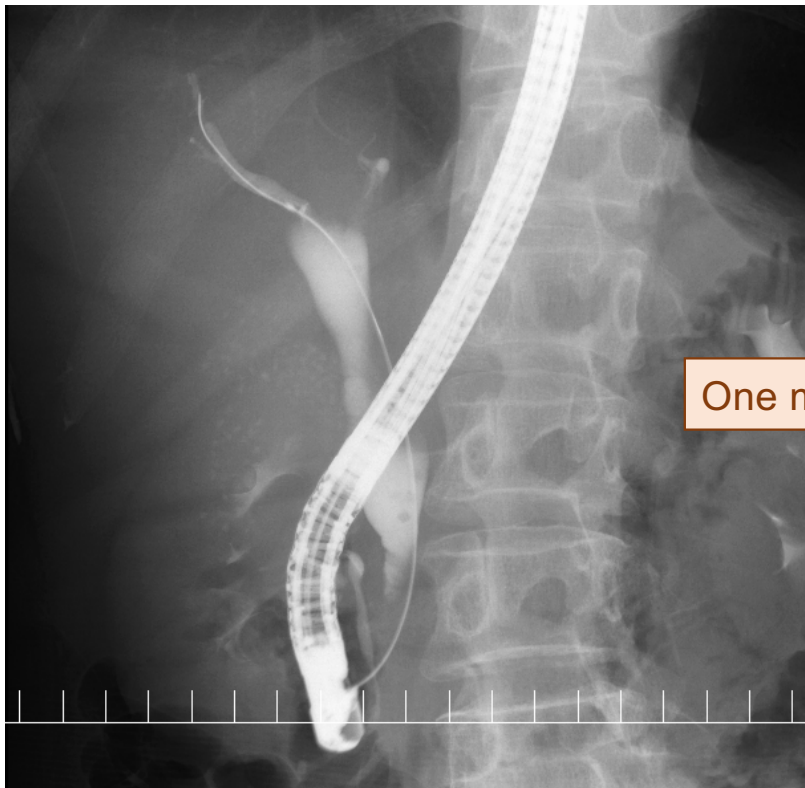


Limited dilatation
Intrahepatic bile ducts are more dilated

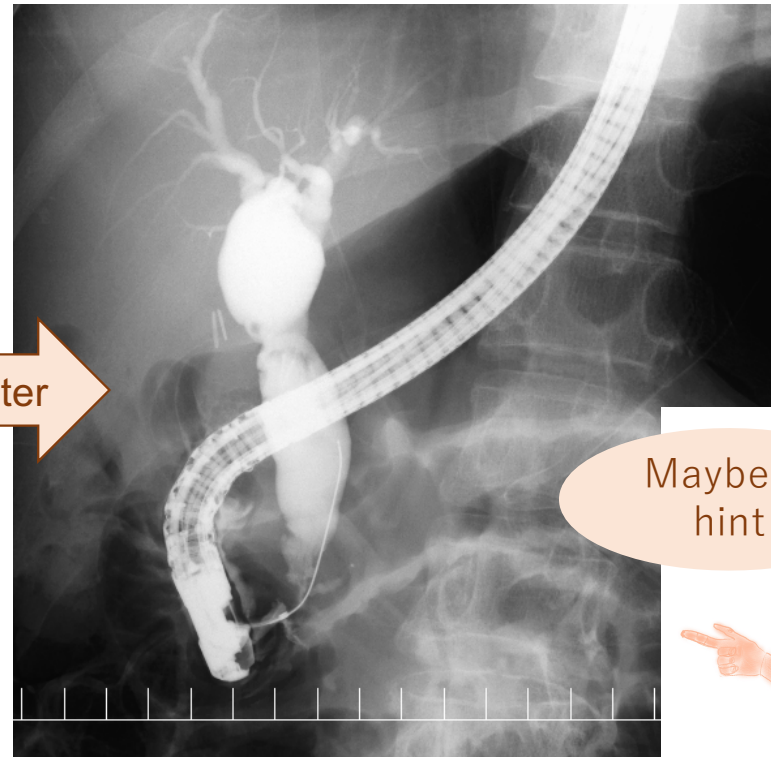


Acquired limited BD dilatation

Chronic intermittent increase of intraductal pressure



One month later



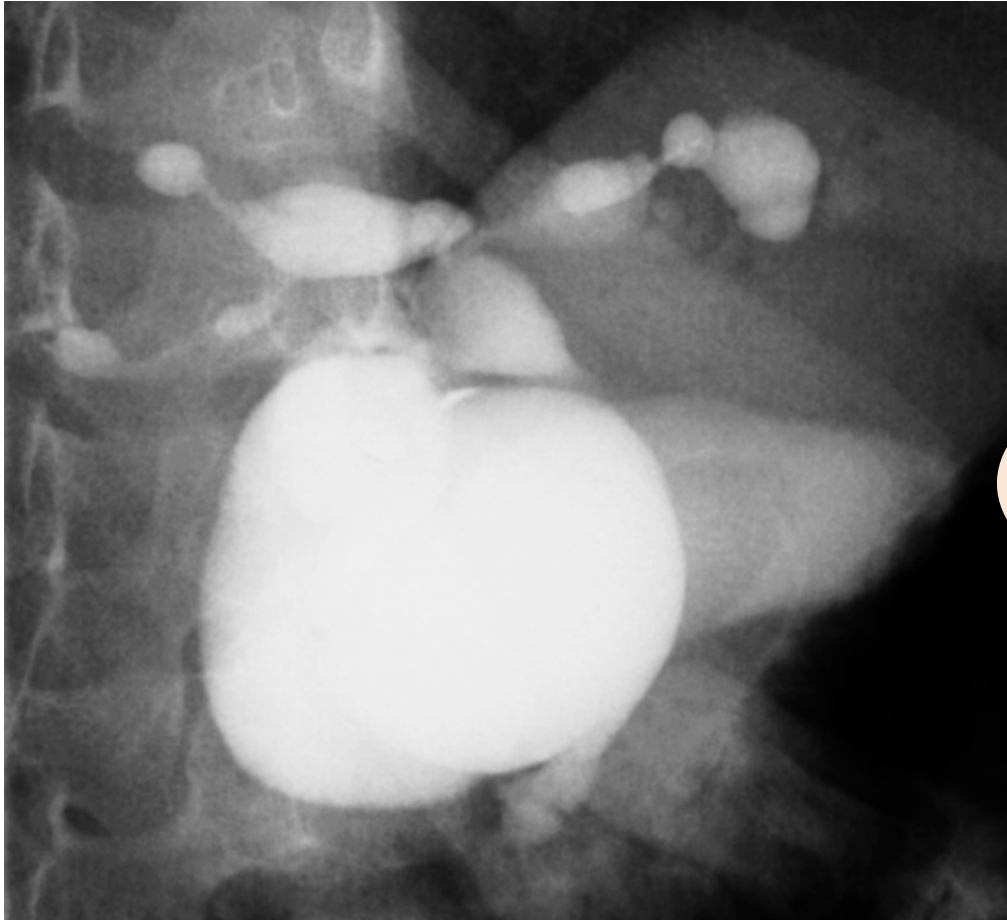
Maybe, a hint



Male in his fifties, Bile duct stones

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

Peripheral cystic dilatation of Intrahepatic BDs

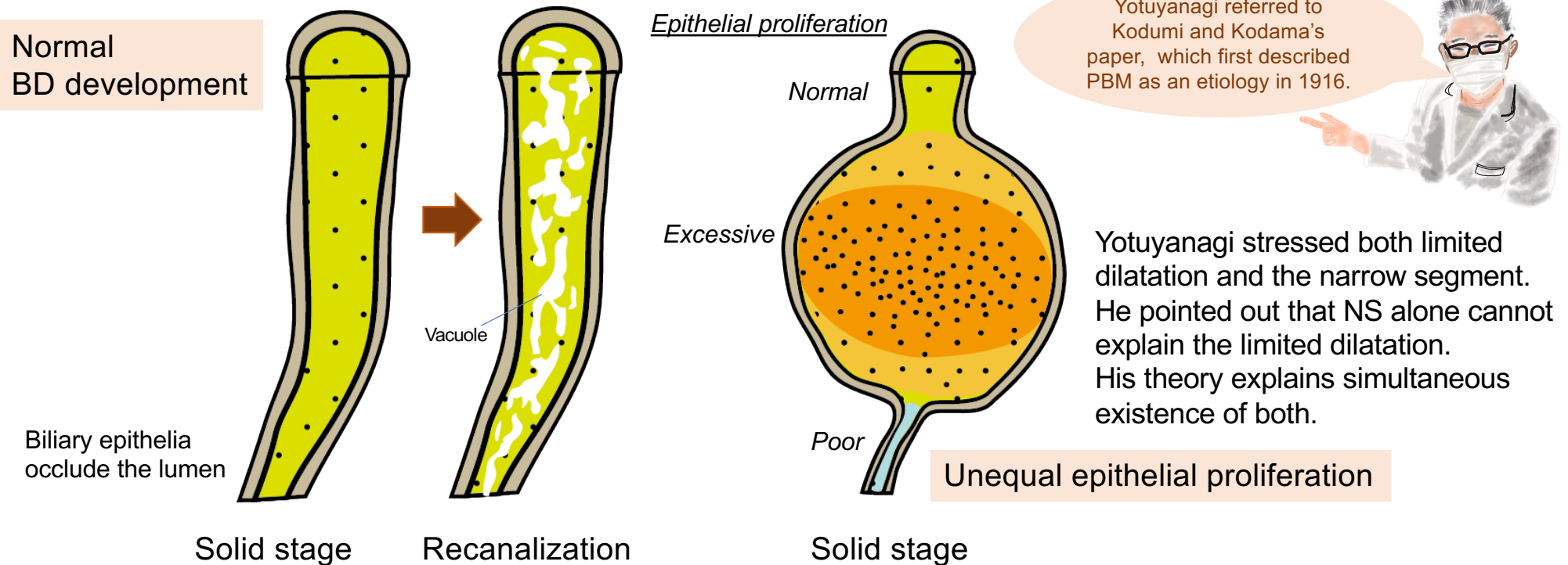


However, it is difficult to explain this phenomenon only by high pressure.



Yotuyanagi's etiological theory in 1936

Limited dilatation in congenital biliary dilatation



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

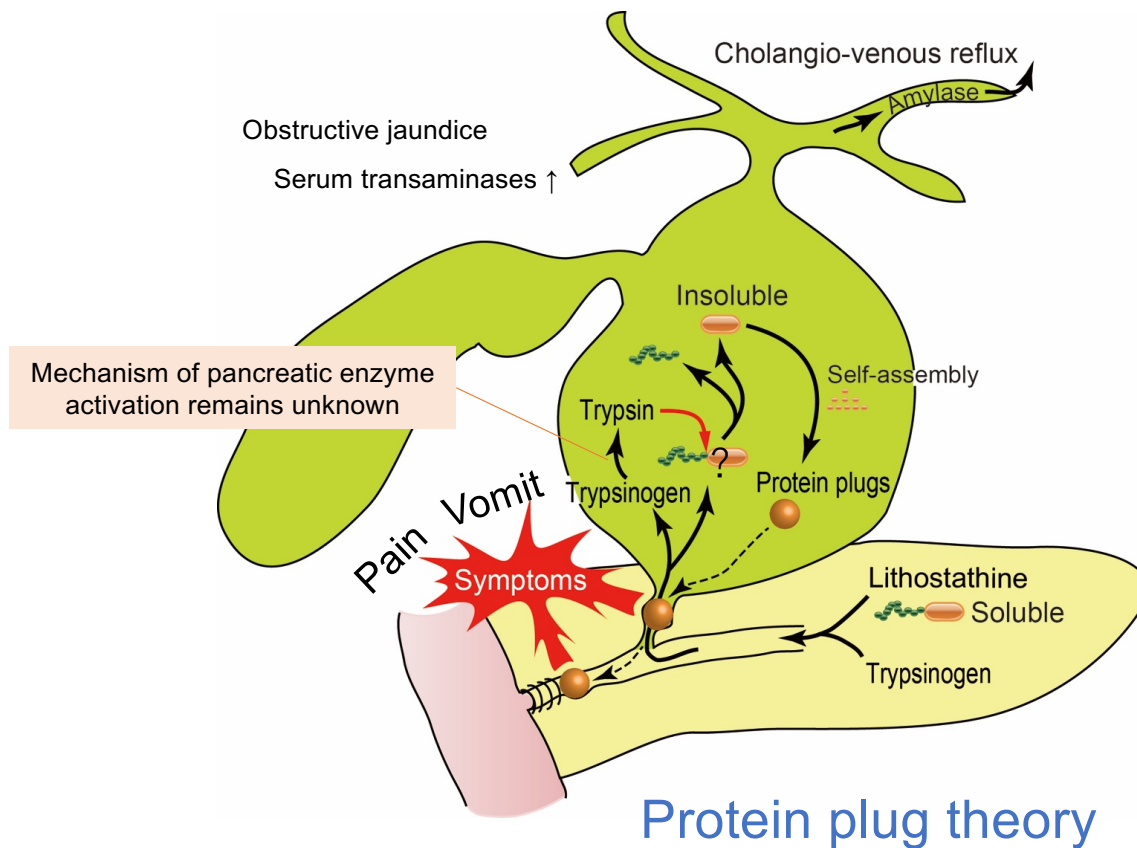
Advanced

Pathophysiology in pediatric patients

First version, September 2024

Symptomatology in childhood

See Basic Section, P18-23.



Fragile protein plugs occlude the narrow segment or the common channel, producing a transient increase of intraductal pressure and cause the symptoms.

Findings supporting protein plug theory

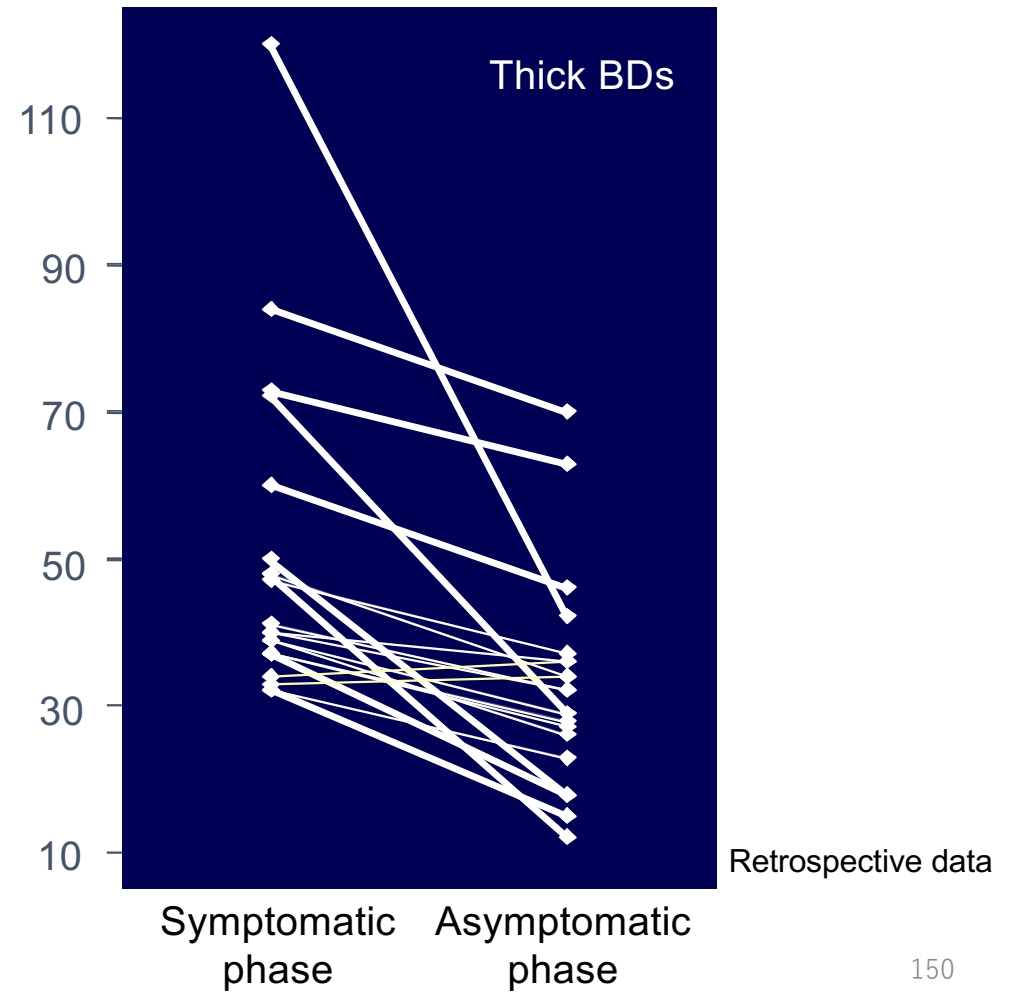
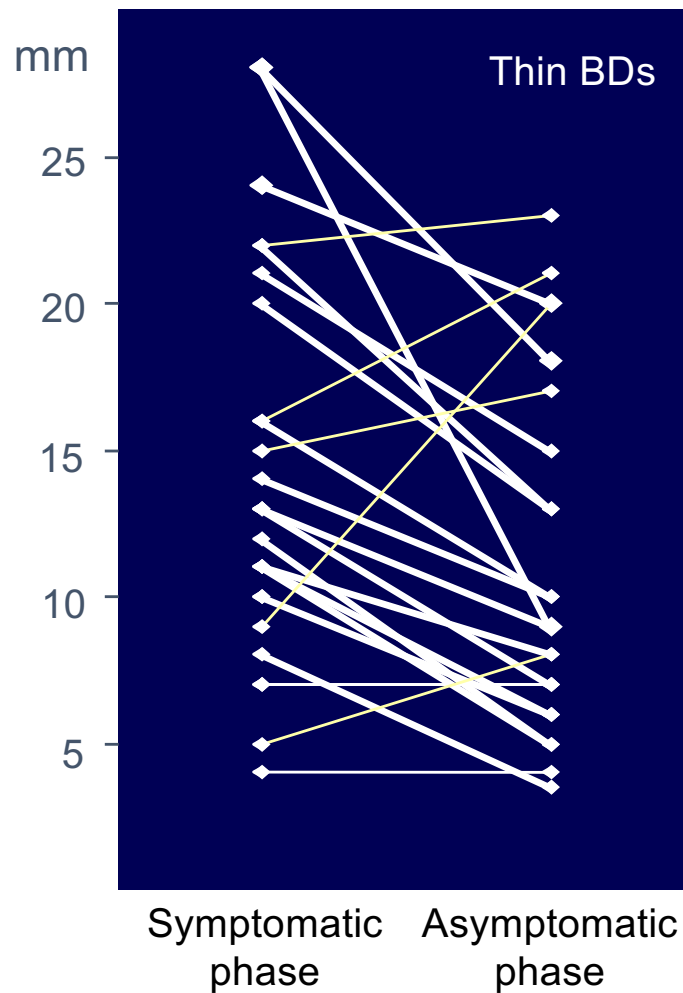
Endoscopic drainage promptly relieves persistent or exacerbated symptoms



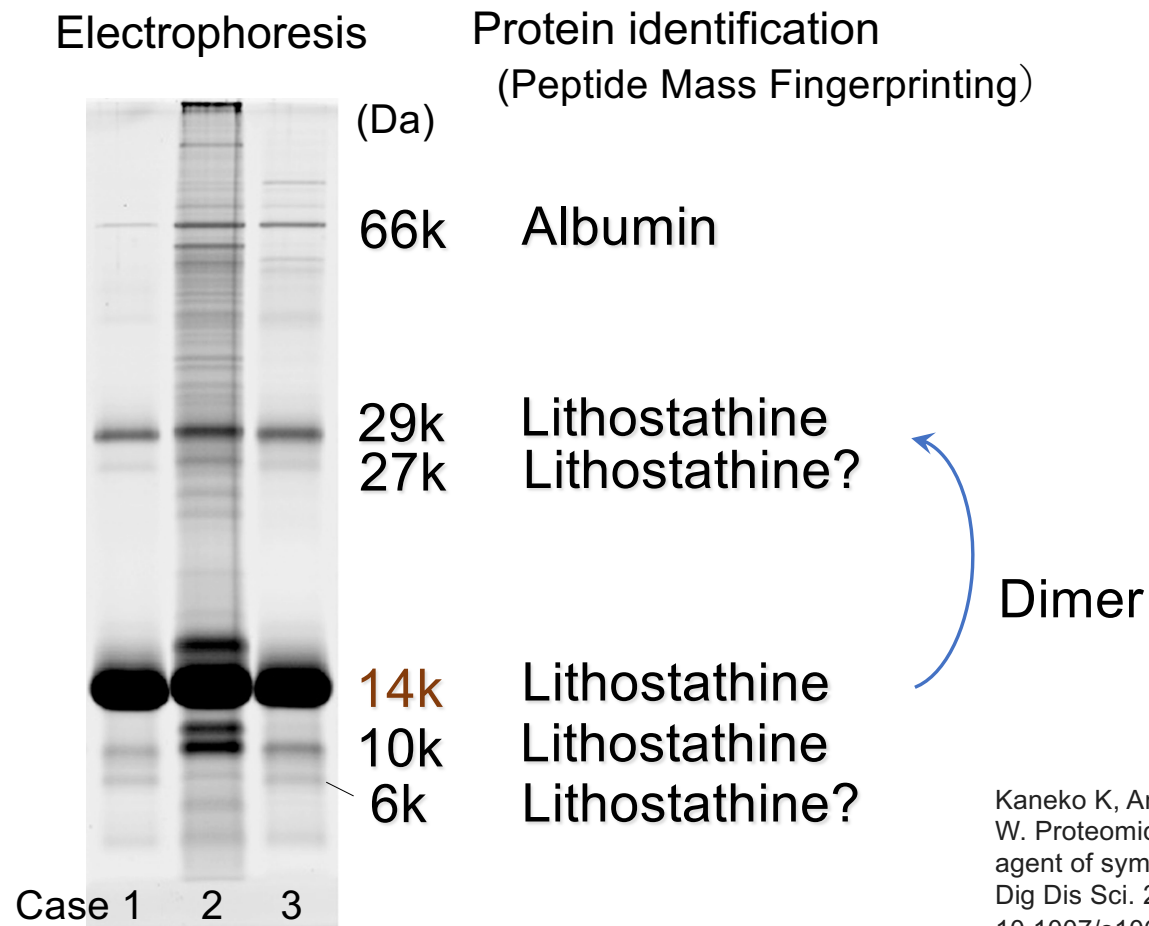
Tsuchiya H, Kaneko K, Itoh A, Kawashima H, Ono Y, Tainaka T, Murase N, Ando H. Endoscopic biliary drainage for children with persistent or exacerbated symptoms of choledochal cysts. J Hepatobiliary Pancreat Sci. 2013 Mar;20(3):303-6. doi: 10.1007/s00534-012-0519-5. PMID: 22581057. Fig. 1

Findings supporting protein plug theory

Diameter of the common bile duct is larger in symptomatic phase



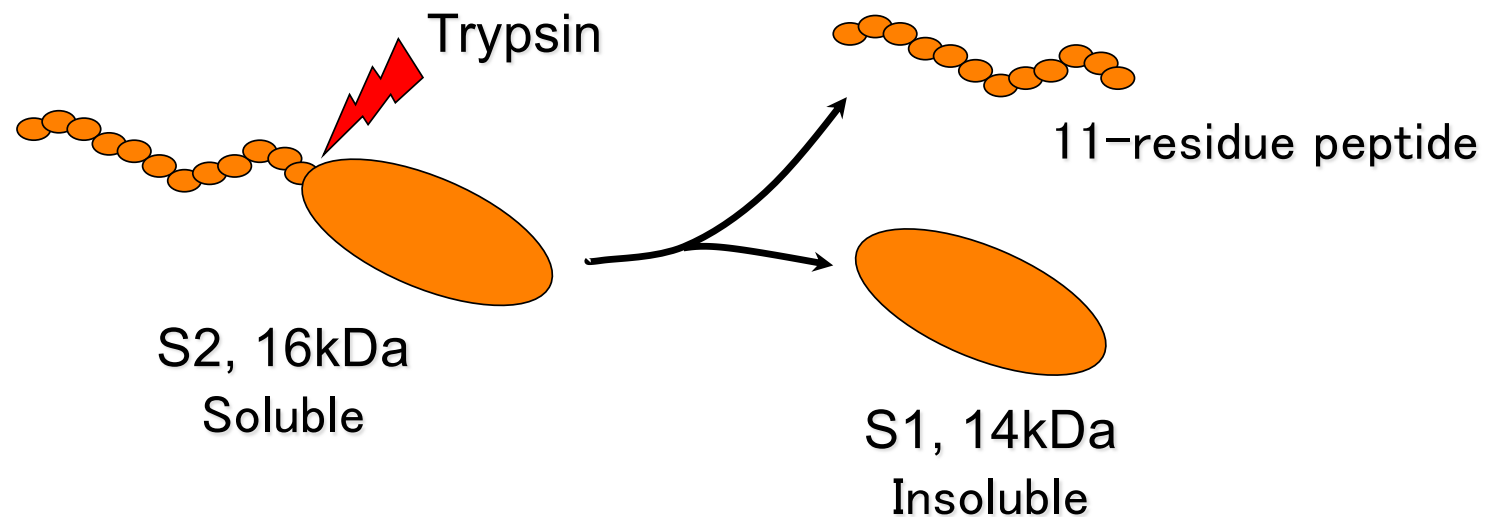
Protein plugs are composed of lithostathine



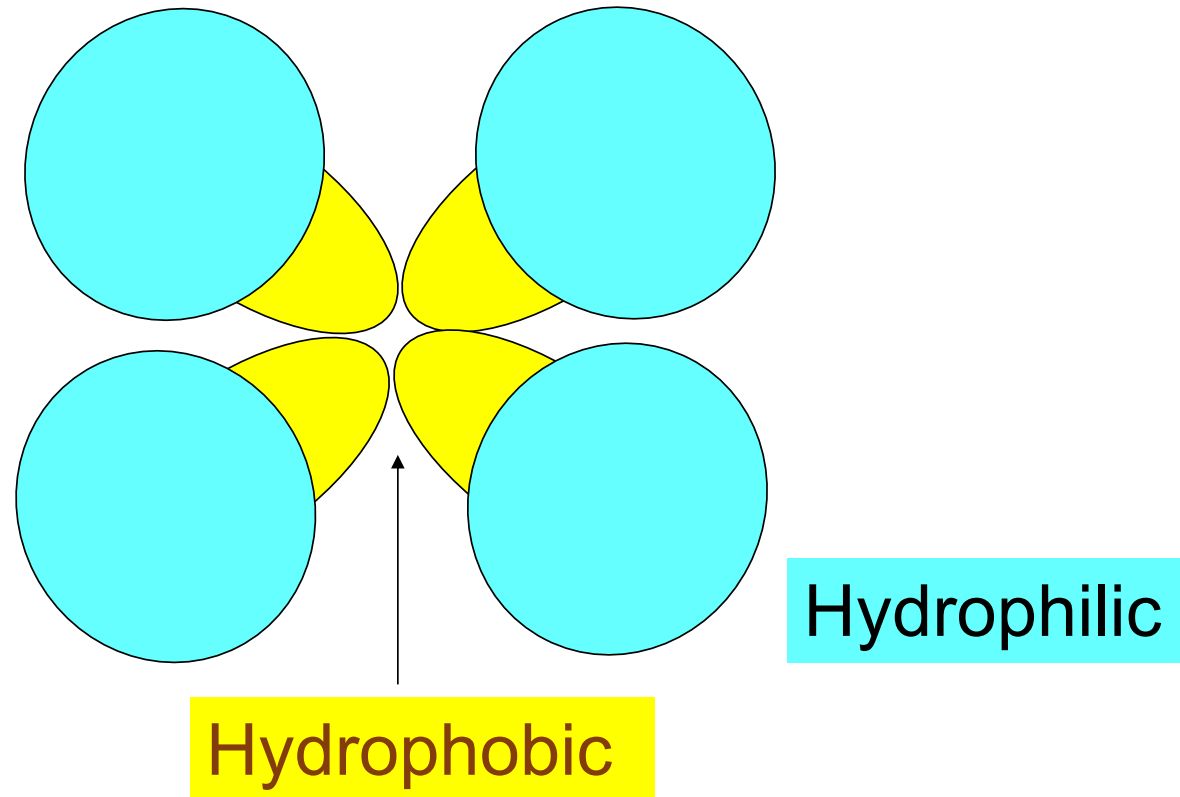
Kaneko K, Ando H, Seo T, Ono Y, Tainaka T, Sumida W. Proteomic analysis of protein plugs: causative agent of symptoms in patients with choledochal cyst. Dig Dis Sci. 2007 Aug;52(8):1979-86. doi: 10.1007/s10620-006-9398-4. Epub 2007 Apr 6. PMID: 17415647.

Lithostathine

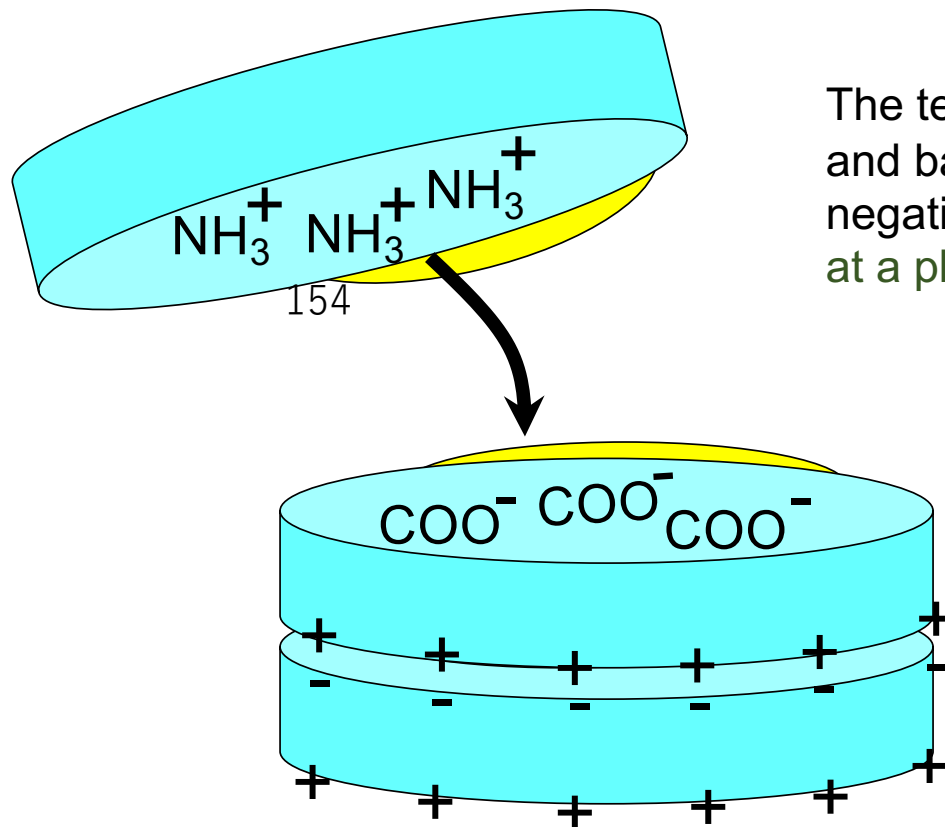
- Soluble 144-amino acid glycoprotein secreted by pancreatic acinar cells
- Accounts for 5%-10% of proteins in pancreatic juice but unknown function
- Many other names such as pancreatic stone protein (PSP)



Lithostathine S1 forms tetramers by lateral hydrophobic interactions



Tetramers self-assemble by longitudinal electrostatic interactions

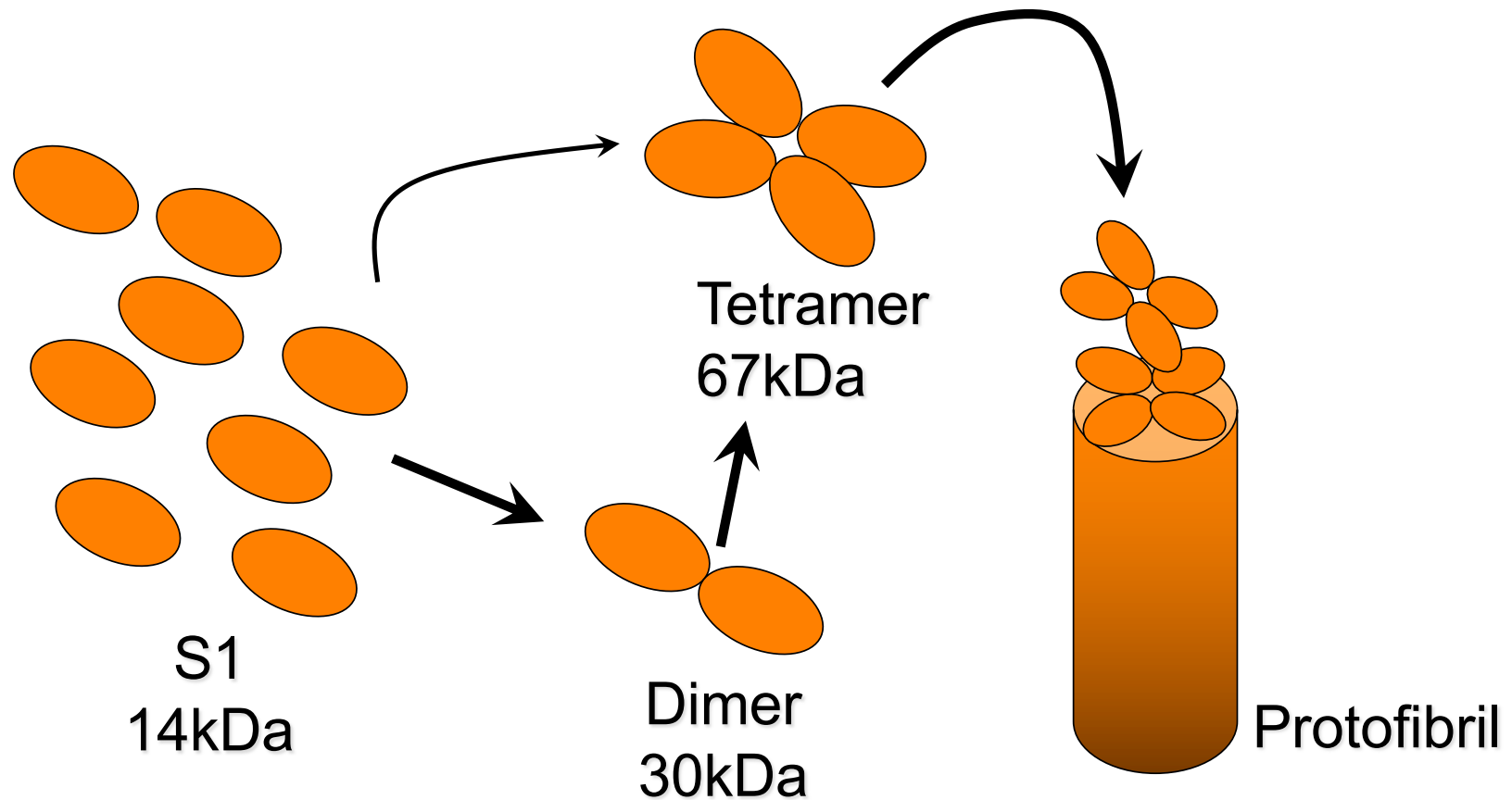


The tetramer has acidic residues on one side and basic residues on the other, contributing negative and positive charges, respectively, at a physiologic pH.

Low pH	Physiologic PH	High pH
$-\text{NH}_3^+$	$-\text{NH}_3^+$	$-\text{NH}_2$
$-\text{COOH}$	$-\text{COO}^-$	$-\text{COO}^-$

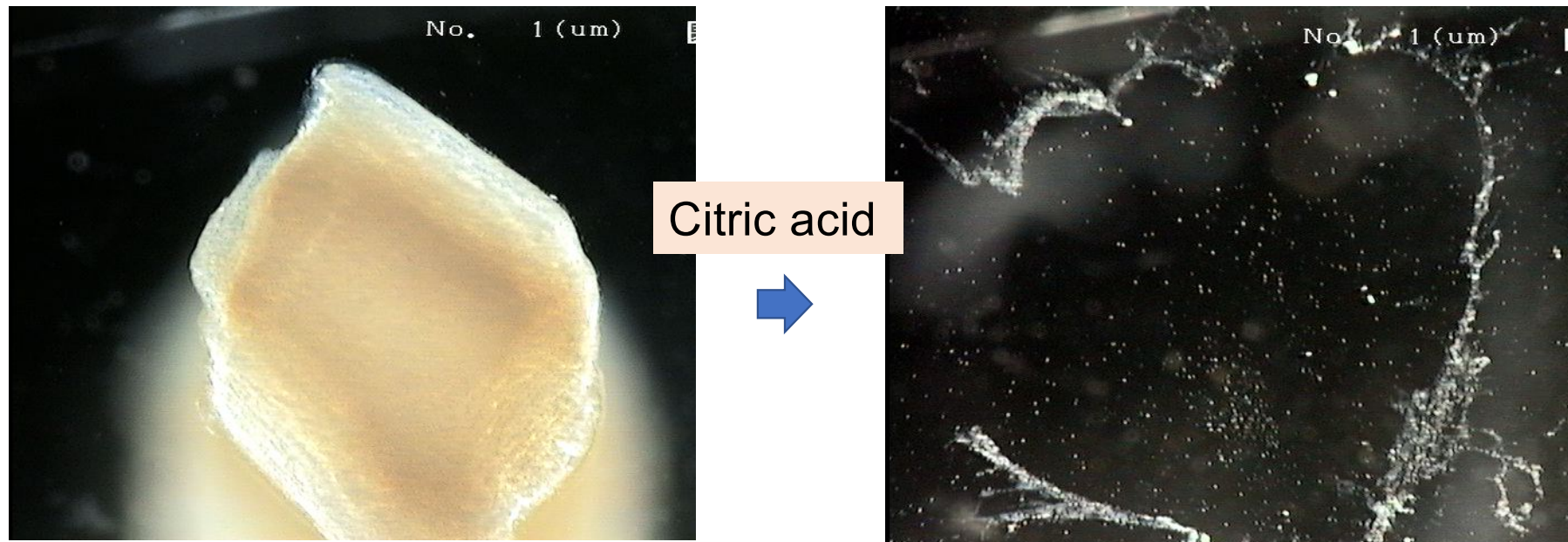
Gregoire et al. EMBO J 2001;20:3313–3321

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



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

Acidic and basic solutions dissolve protein plugs



Proof that electrostatic interactions work only at a physiologic pH

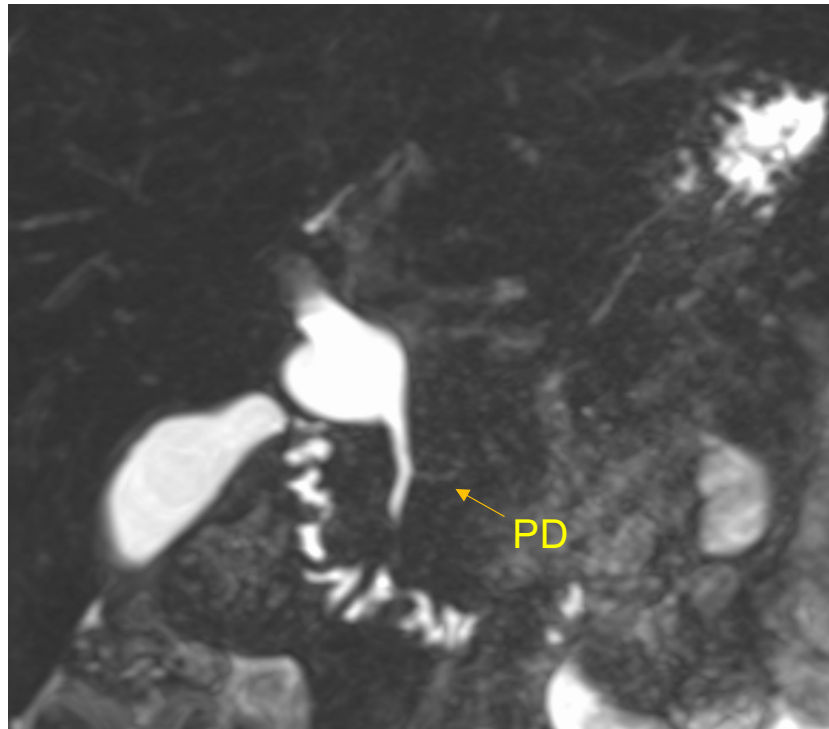
Kaneko K, Ono Y, Tainaka T, Sumida W, Ando H. Acidic and basic solutions dissolve protein plugs made of lithostathine complicating choledochal cyst/pancreaticobiliary maljunction. Dig Dis Sci. 2009 Jul;54(7):1475-80. doi: 10.1007/s10620-008-0503-8. Epub 2008 Nov 7. PMID: 18989781.

Where protein plugs form?

Case 4 from Basic Section

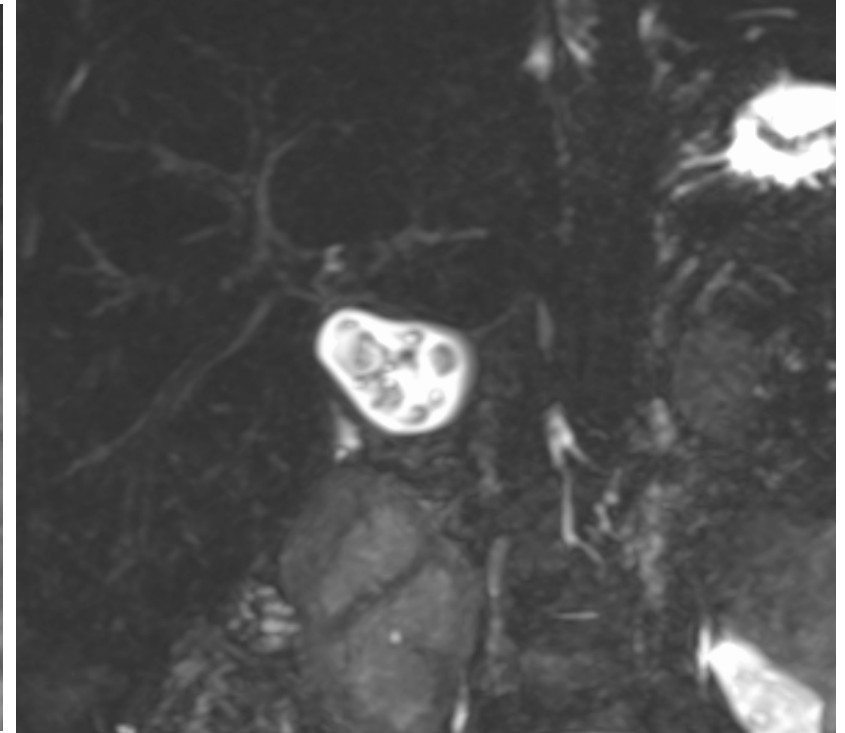
- Onset with abdominal pain and vomiting
- Elevated serum levels of liver and biliary enzymes
- No elevation of serum levels of pancreatic enzymes

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



No dilatation at the common channel

Probably, here



Many filling defects at the dorsal side in the choledochus

Advanced

Symptomatology besides protein plugs

First version, September 2024

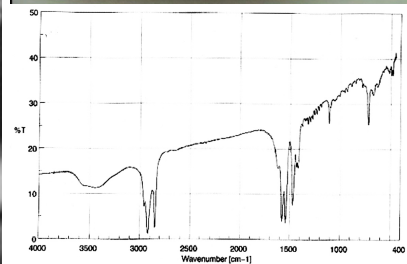
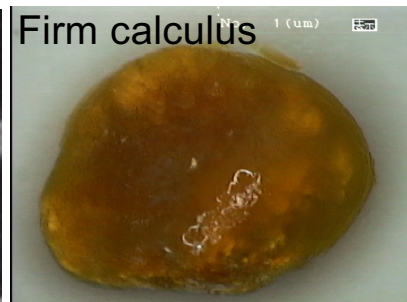
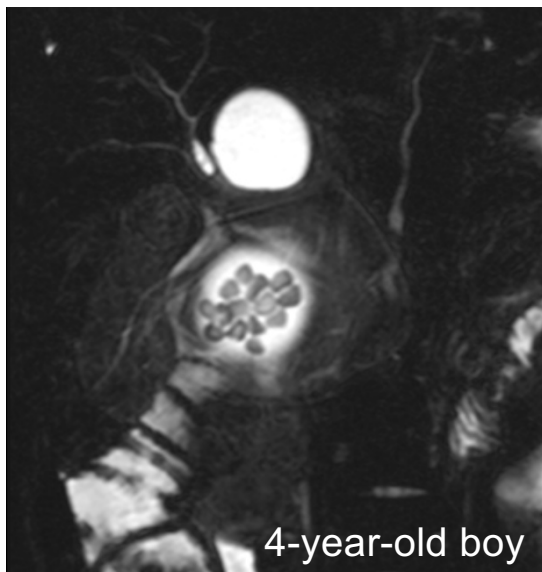
There is no theory without exceptions.



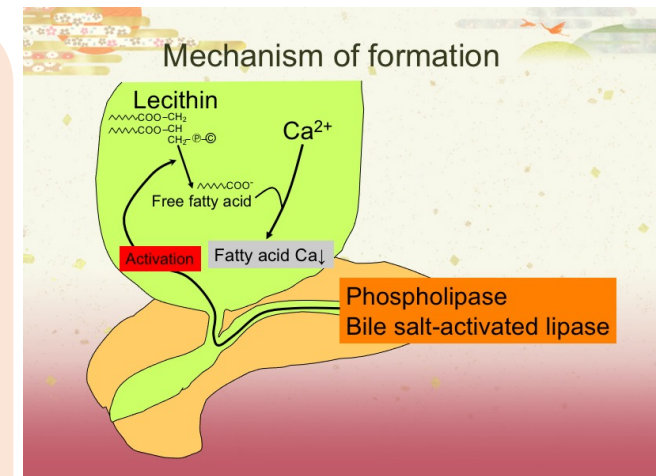
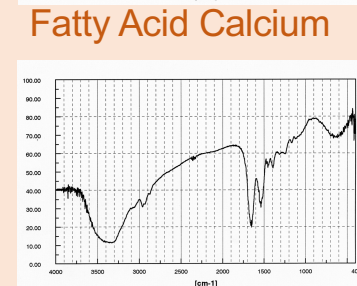
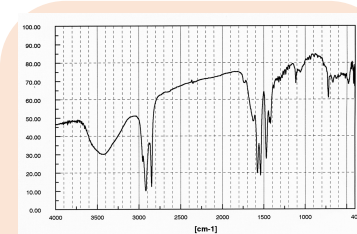
Symptomatology besides protein plug

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

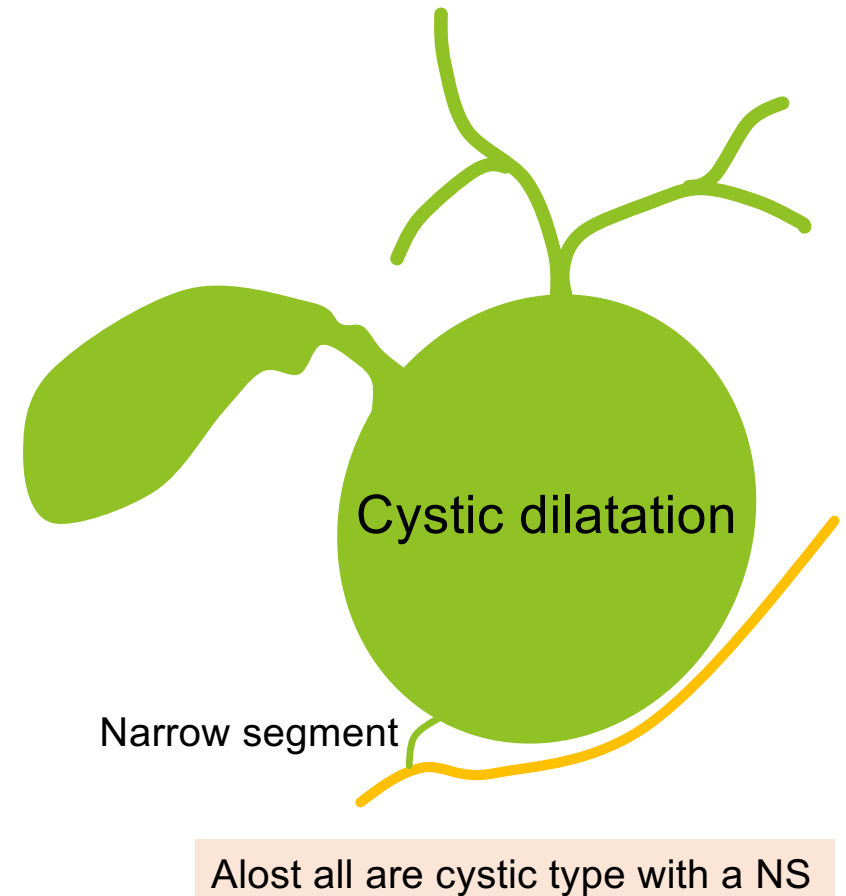
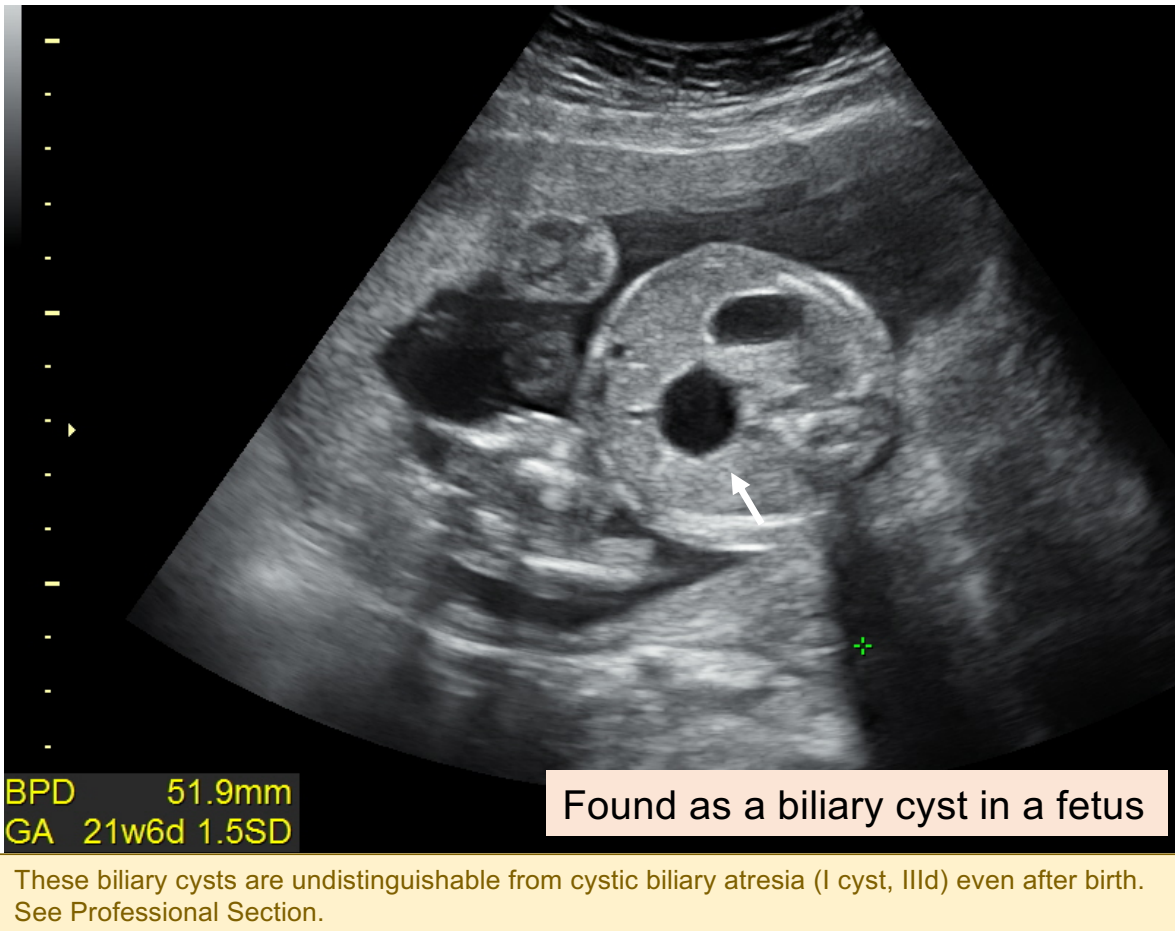


cf. Spectra of standard materials

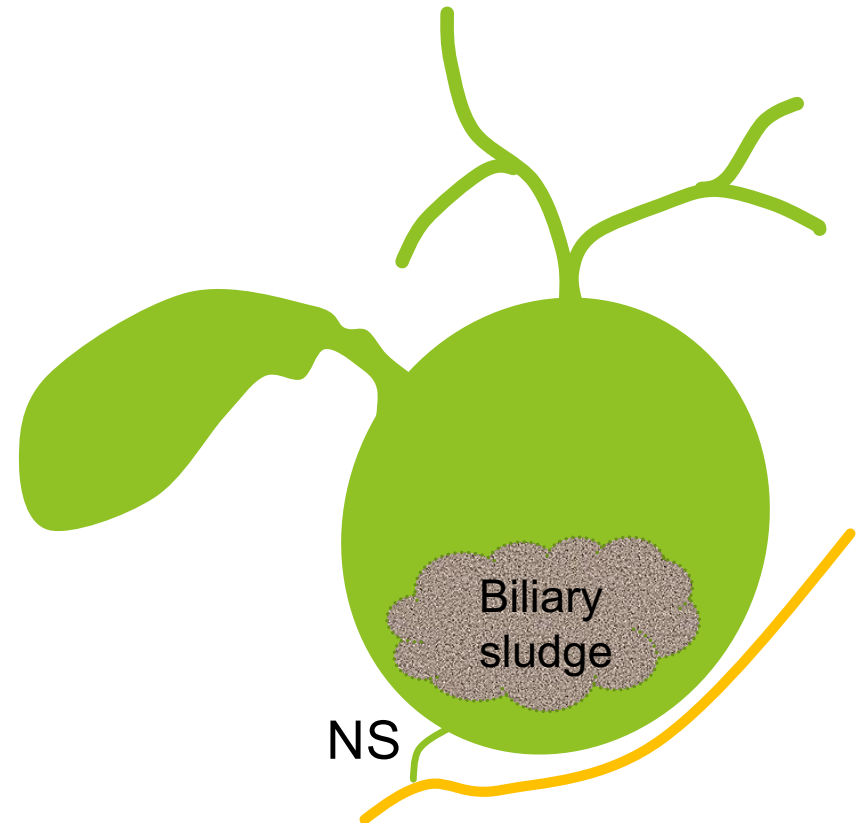
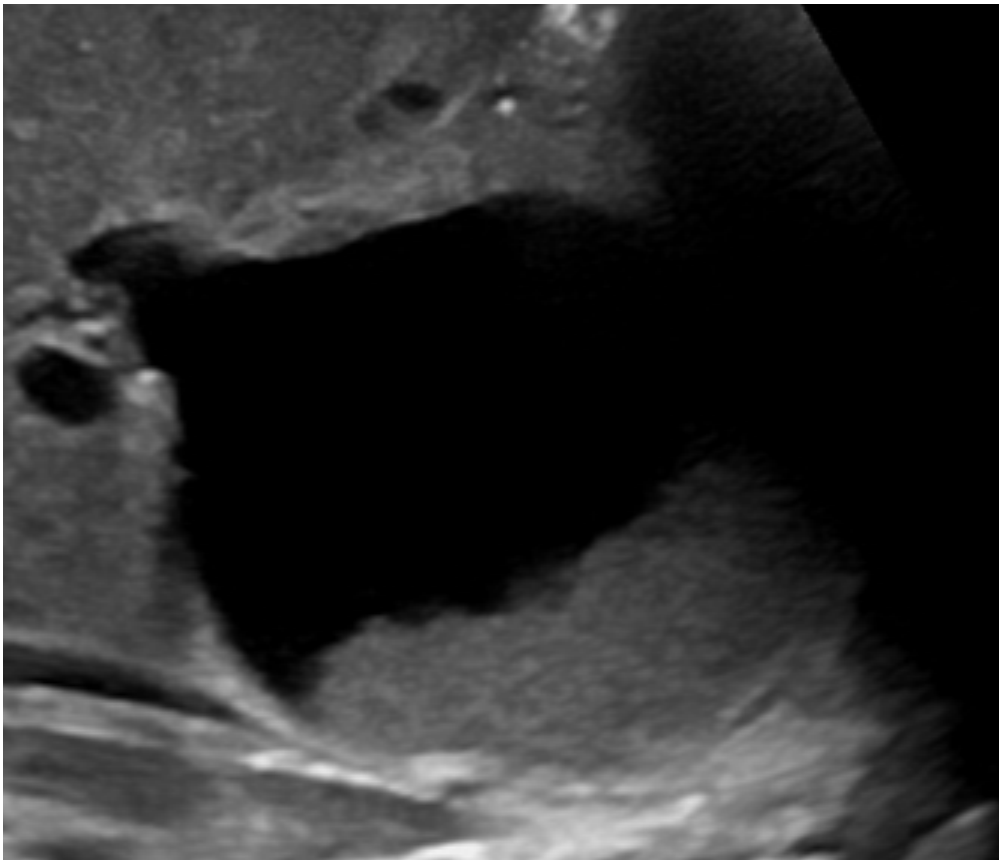


Kaneko K, Ono Y, Tainaka T, Sumida W, Ando H. Fatty acid calcium stones in patients with pancreaticobiliary maljunction/choledochal cyst as another cause of obstructive symptoms besides protein plugs. J Pediatr Surg. 2008 Mar;43(3):564-7. doi: 10.1016/j.jpedsurg.2007.11.004. PMID: 18358304.

Onset in neonates and early infants



Debris appears in the cyst before symptoms occur

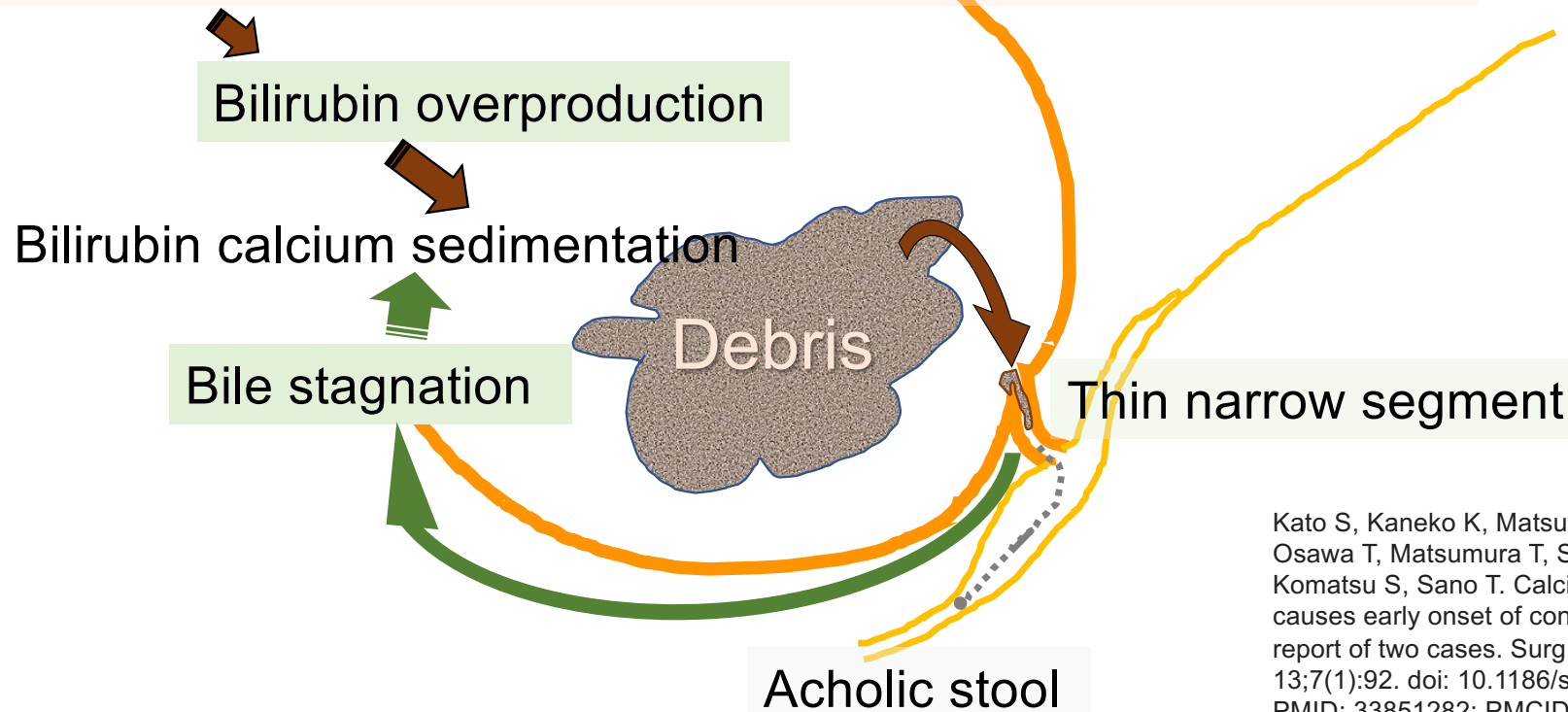


Infrared absorption spectrometry shows the sludge is composed of **Bilirubin Calcium**.

Proposed mechanism of early onset

Accelerated **hemolysis** due to neonatal hematologic features

- Physiological **polycythemia** of red blood cells (RBC)
- High percentage of **Hb F** RBC 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 Apr 13;7(1):92. doi: 10.1186/s40792-021-01175-x. PMID: 33851282; PMCID: PMC8044275.

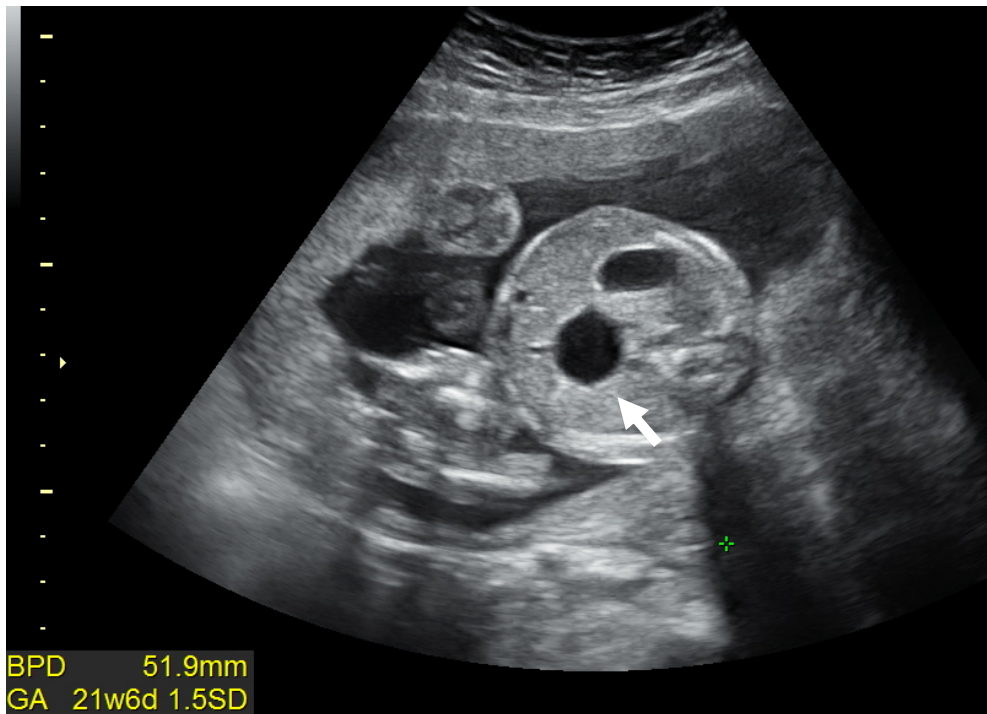
Professional

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



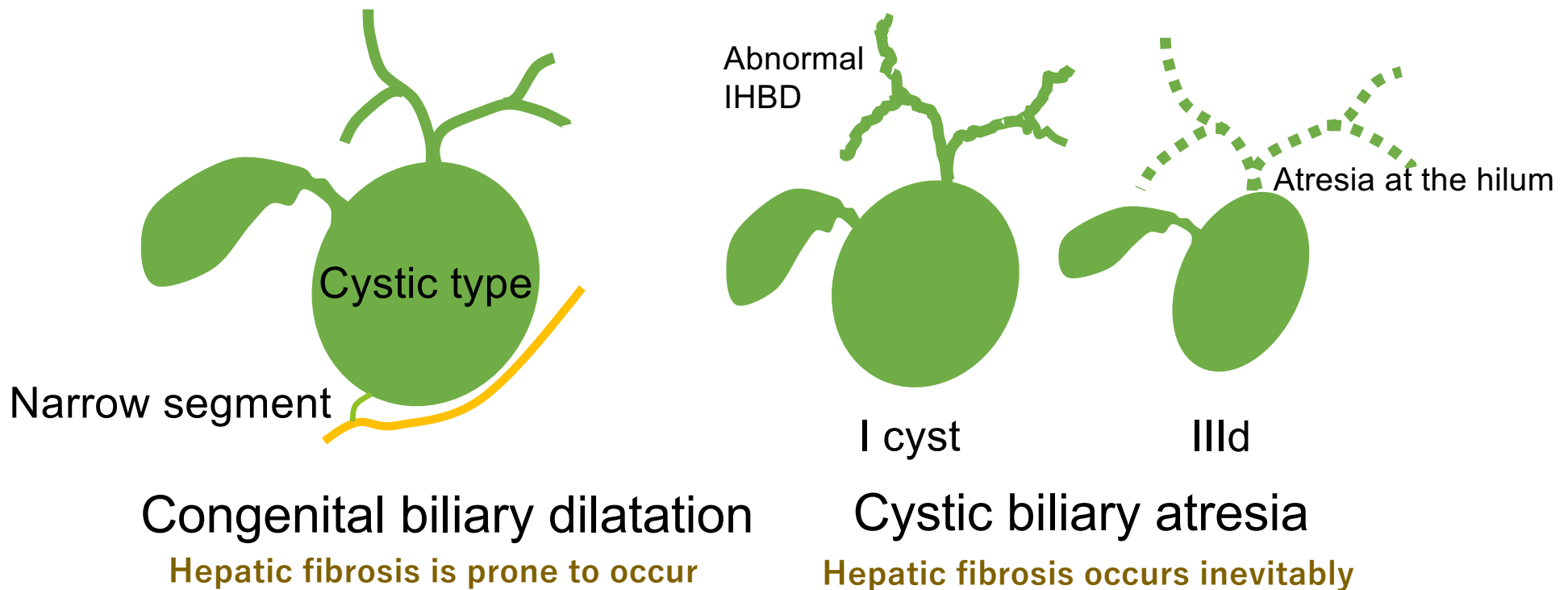
Differential diagnosis is difficult even after birth

→ Congenital biliary dilatation ?
or
Biliary atresia ?

Why?

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

Fetal biliary cysts present a similar form regardless of final diagnoses



Biliary obstruction causes cholangiopathy specific to neonates and early infants

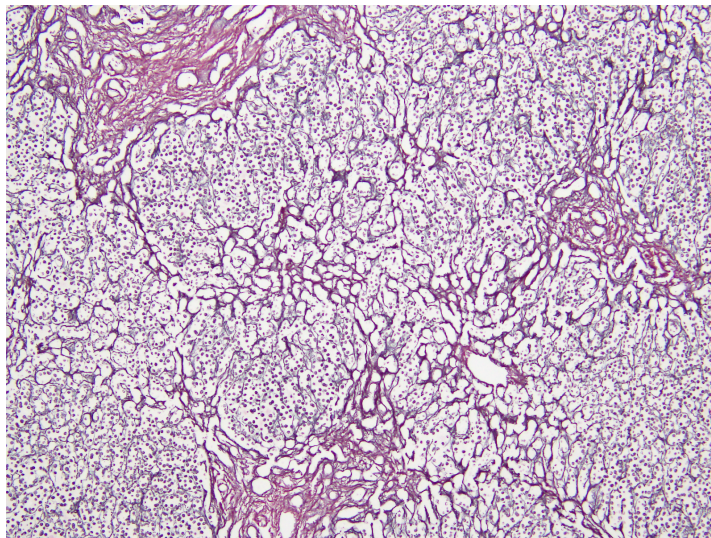
Prenatally diagnosed CBD and hepatic fibrosis

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

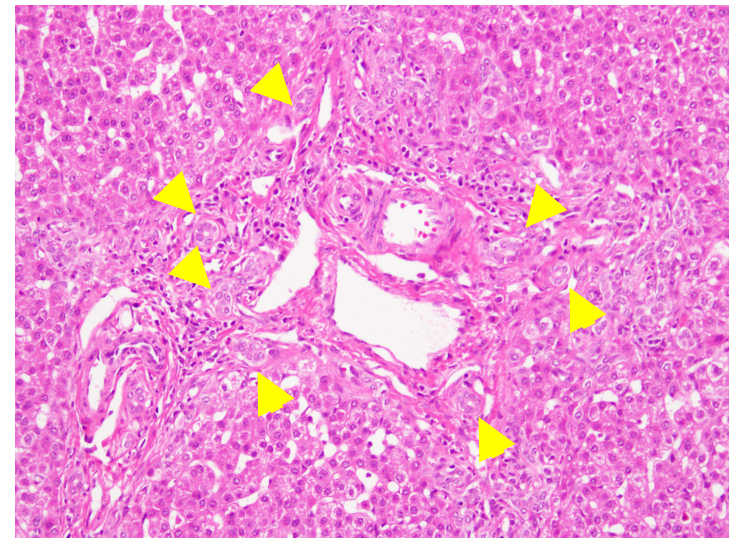
Diagnosis at 29 weeks' gestation

BD resection when acholic stool occurred at 6 months

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

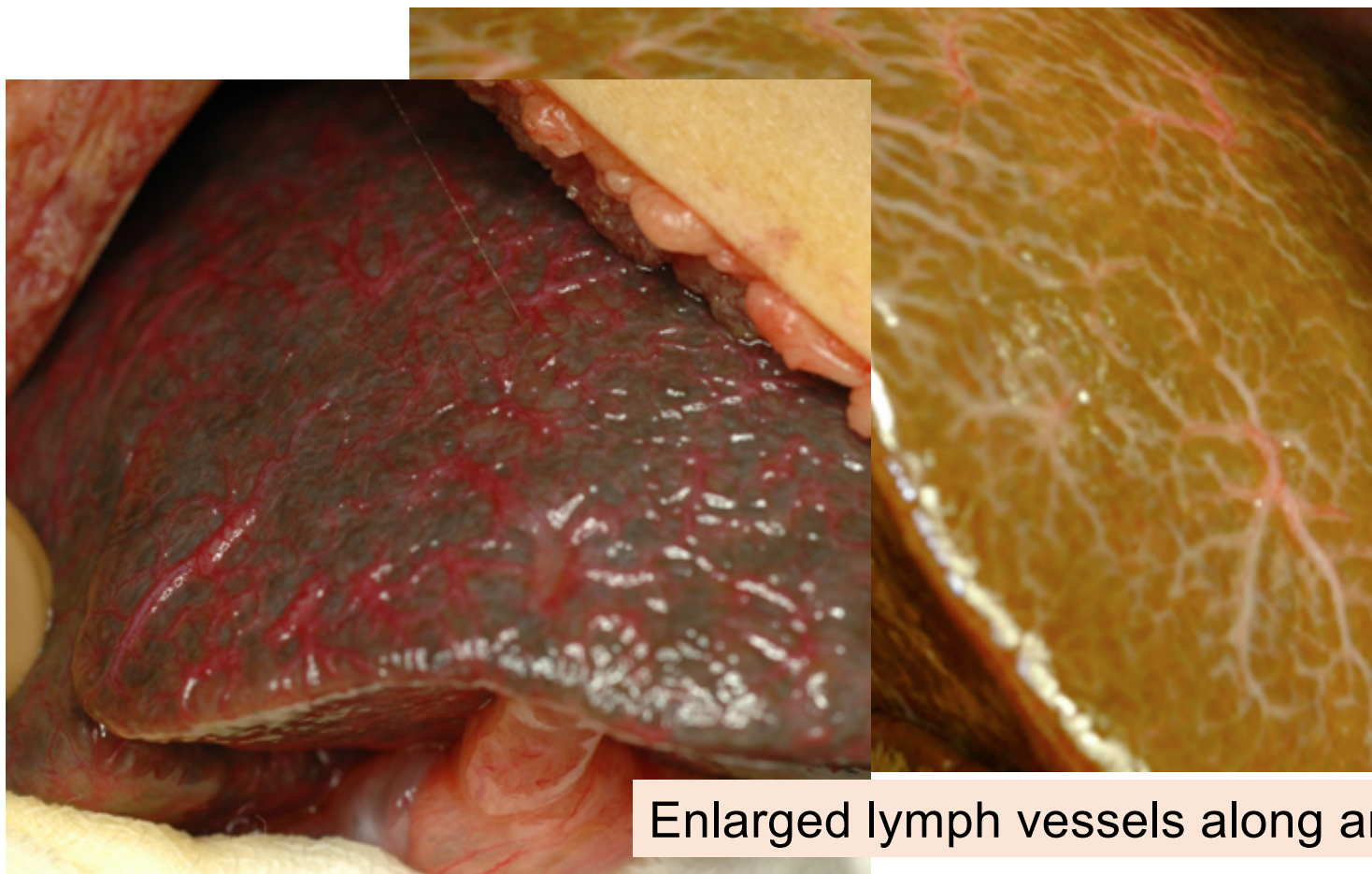


Bridging F1-2

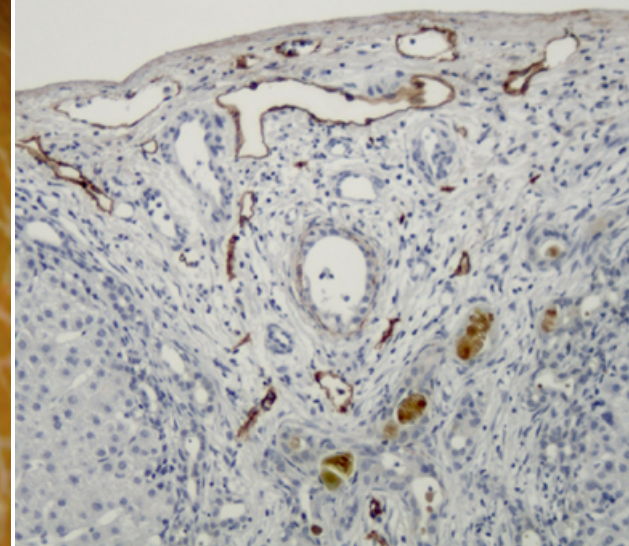


Cholangiocyte proliferation

Makibishi sign specific to Biliary Atresia



Presented by Dr Tainaka, Nagoya Univ.

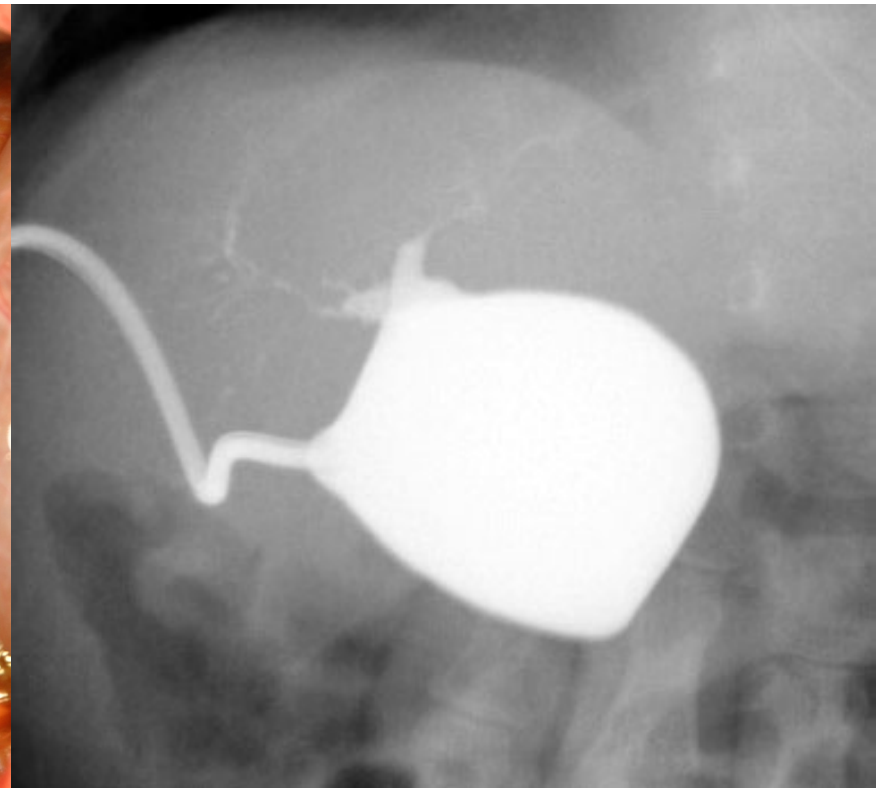
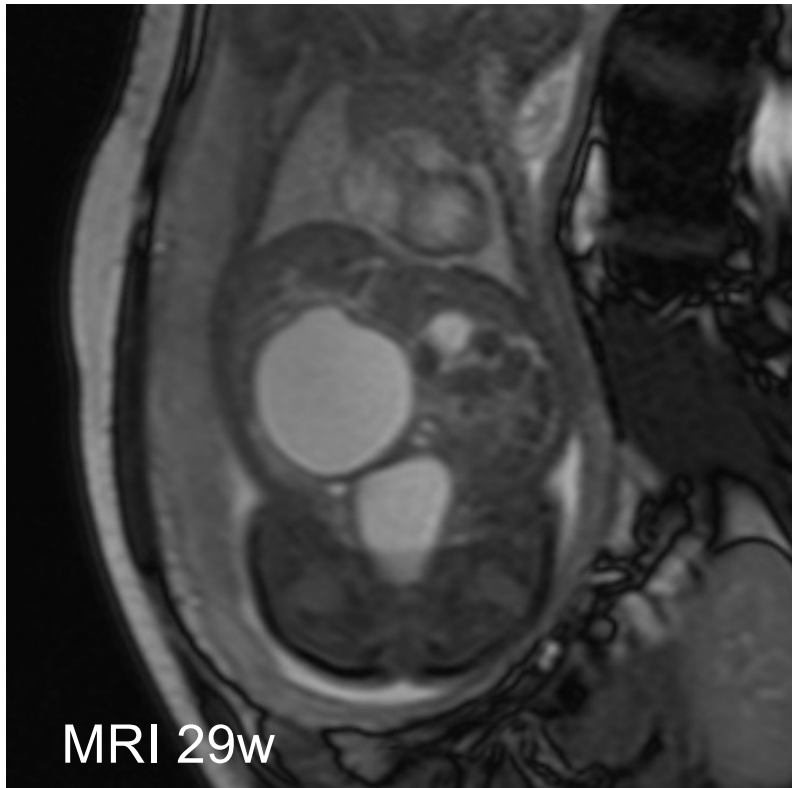


Case **43**

Type I cyst Biliary Atresia

Makibishi sign

Treelike intrahepatic BDs

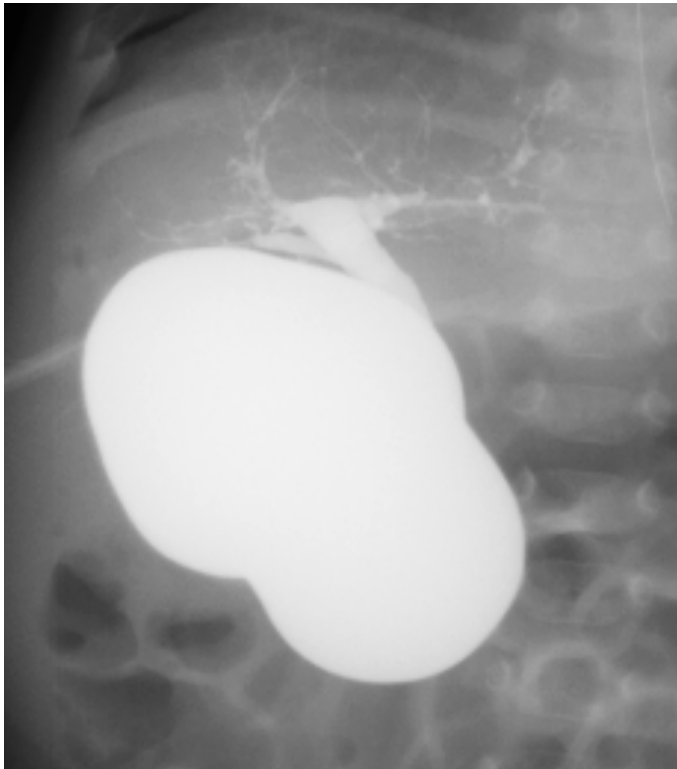


Type I cystic Biliary Atresia

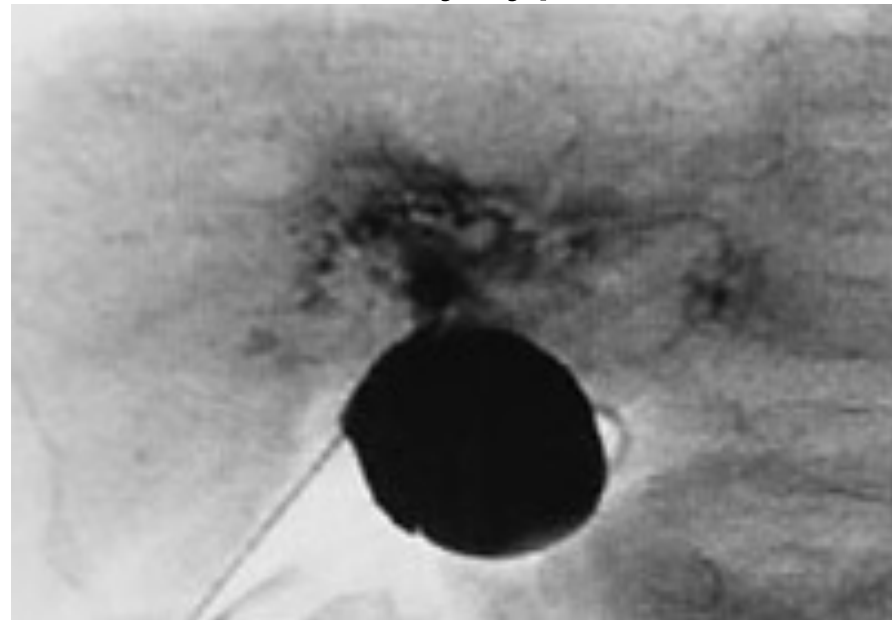
Cholangiography shows

- No flow into the duodenum
- Abnormal IHBD shapes

Treelike type



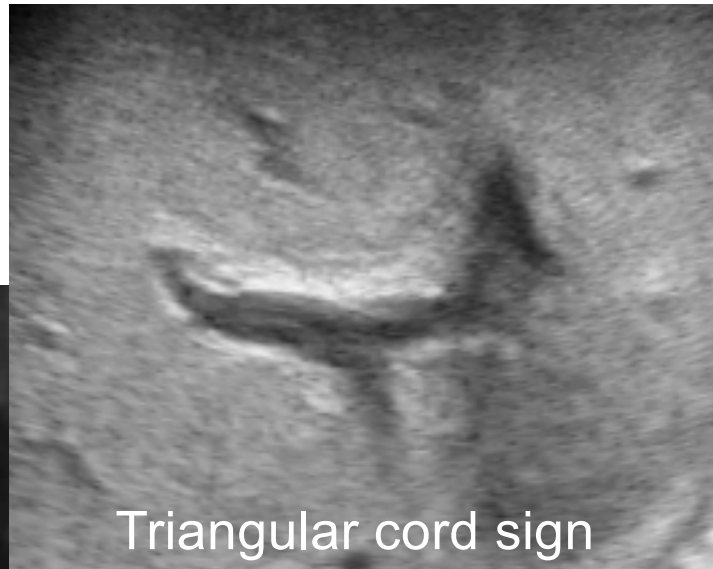
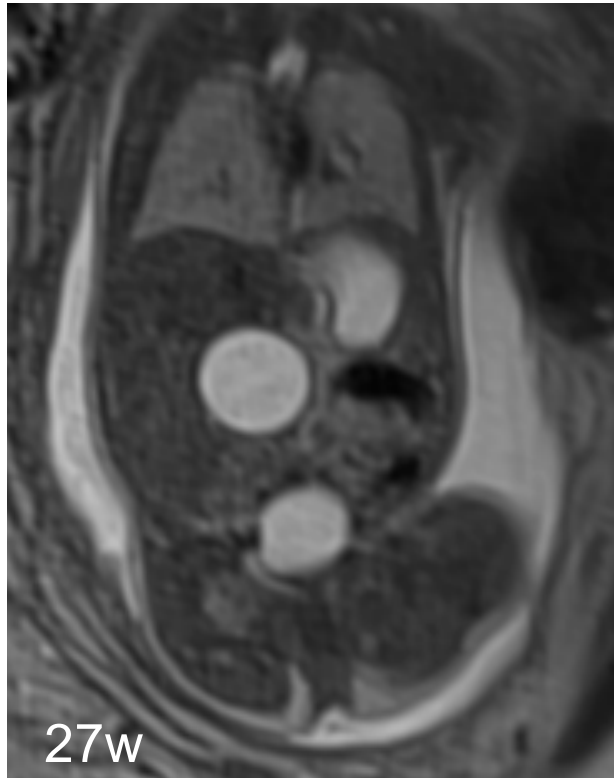
Cloudy type



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

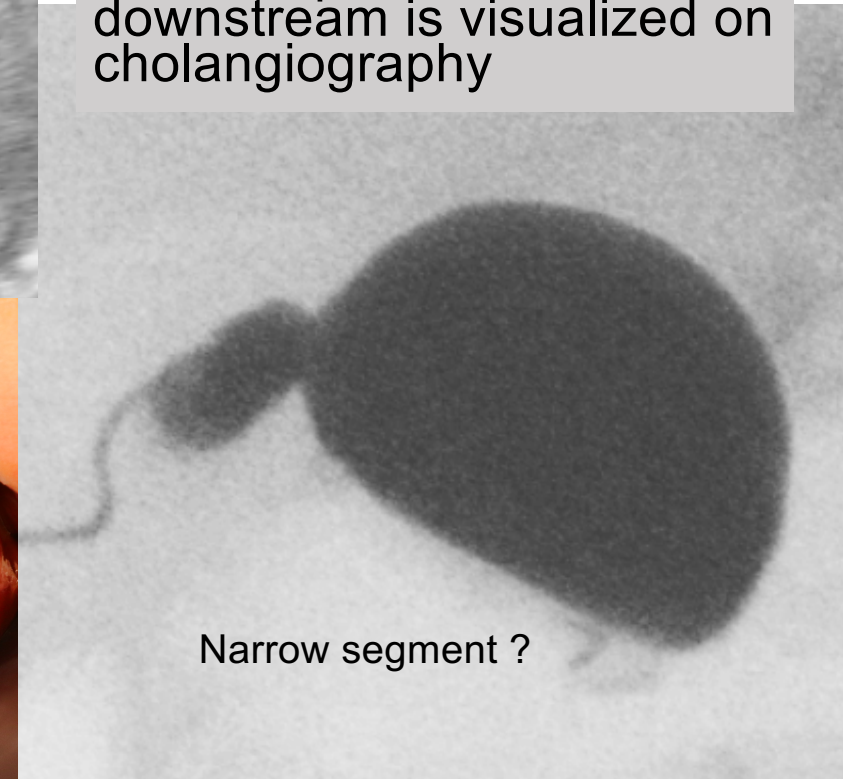
Case **44**

Type IIId BA



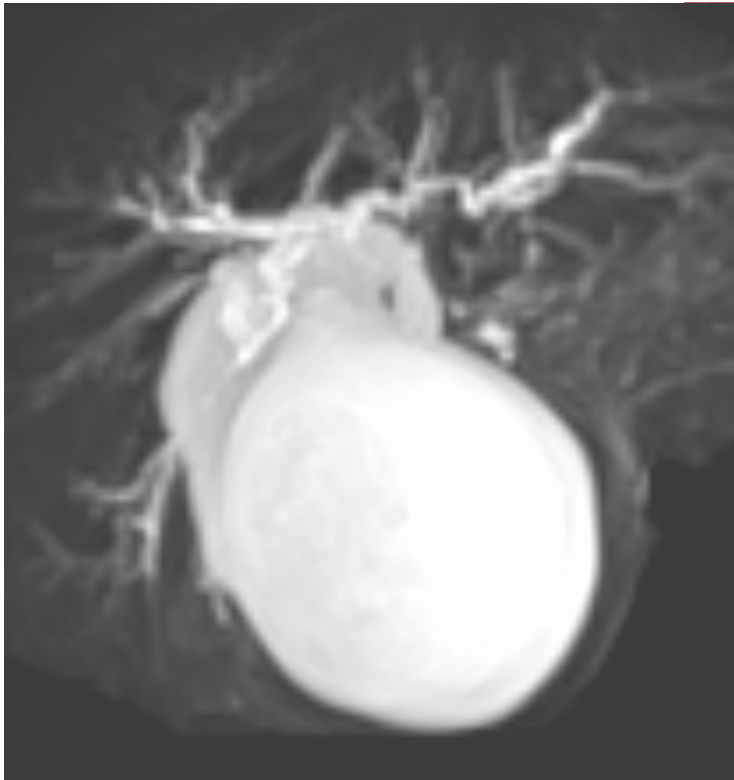
Atresia at the hepatic hilum
requires the Kasai operation

Neither upstream nor
downstream is visualized on
cholangiography



Case **45**

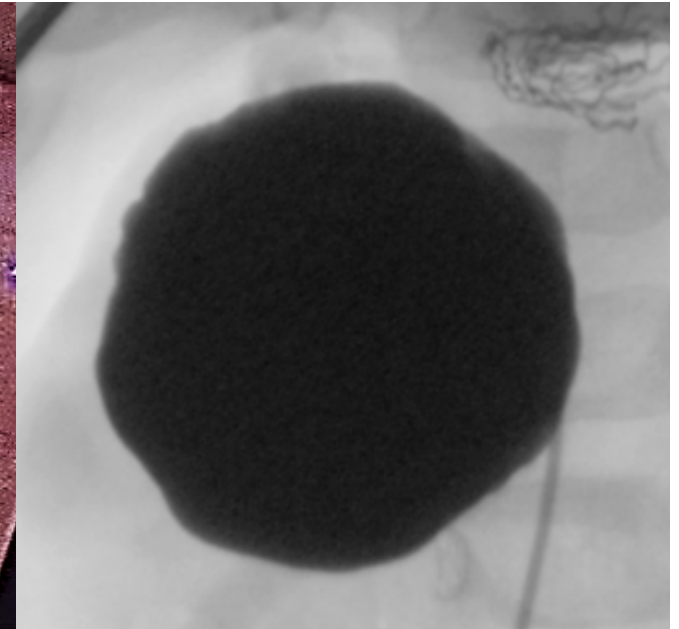
Type Ia CBD > Type I cyst BA



Almost normal IHBD



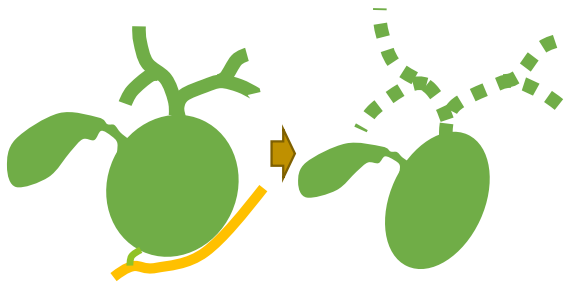
Makibishi sign



Contrast material did not enter
the duodenum

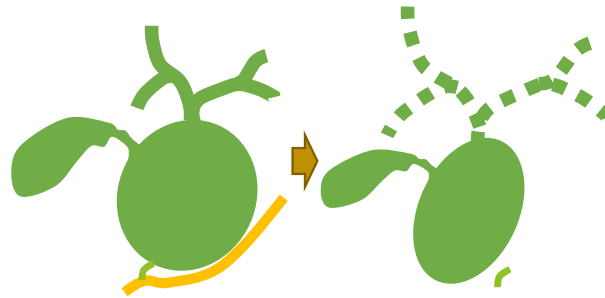
CBD that has features of BA
Intermediate / Transitional Type
Bile duct resection yields good result

There exists a transitional type from CBD to BA



CBD → IIIId BA

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



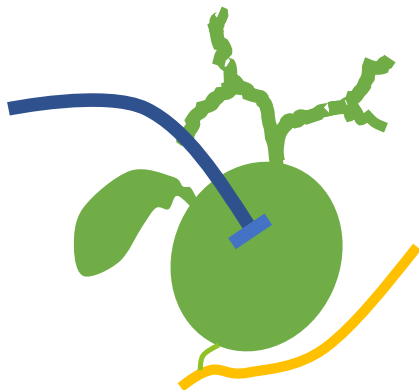
CBD → IIIId BA (+NS)

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

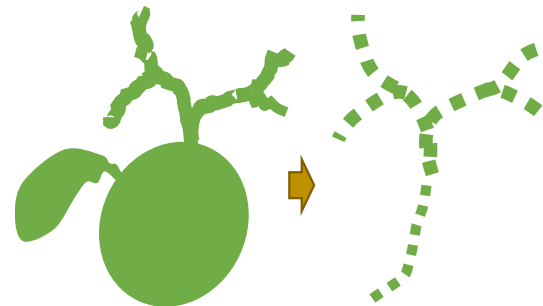


I cyst BA with a dilated LHD
Rapid progress of liver cirrhosis
that results in liver plantation
(The authors describes a type IV-A CBD)

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



I cyst BA
After biliary drainage,
contrast material began to
enter the duodenum,
revealing a NS and PBM.
Suzuki T, et al.
J Hepatobiliary Pancreat Sci
2013;20:465

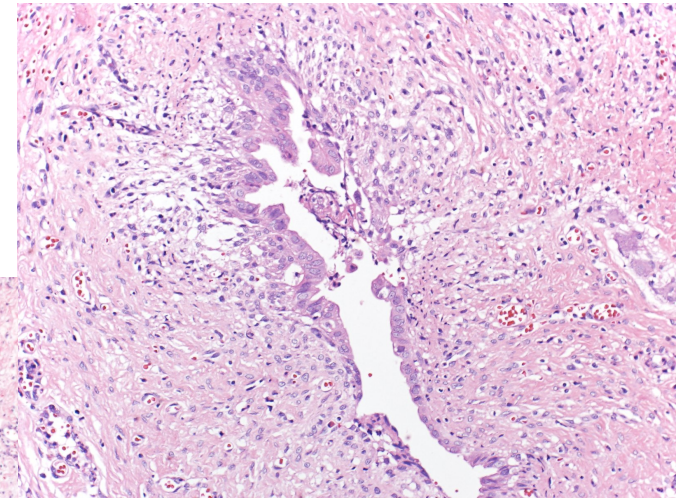


I cyst → III or IIIId

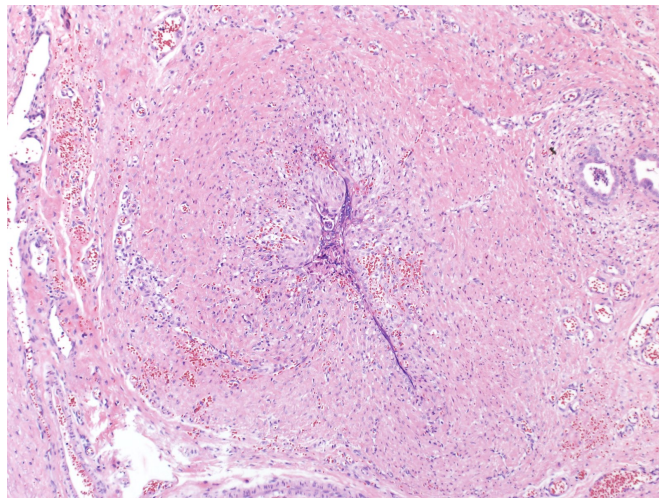
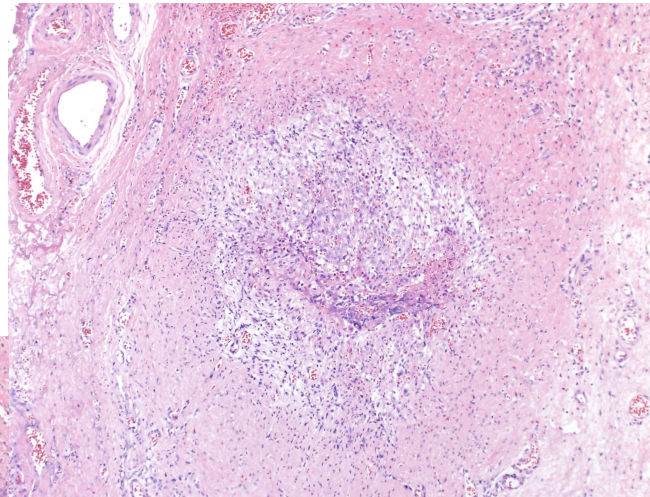
Nakata et al.
J Jpn Pediatr Surg 2000;36:332
Fujishiro J, et al.
Pediatr Sur Int 2005;21:116
Kawasaki et al.
J Jpn Pediatr Surg 2009;45:729
Masumoto K, et al.
Pediatr Sur Int 2011;27:99
Odashima et al.
J Jpn Pediatr Surg 2016;52:1042

Pathology of hilar remnant in type IIIId BA

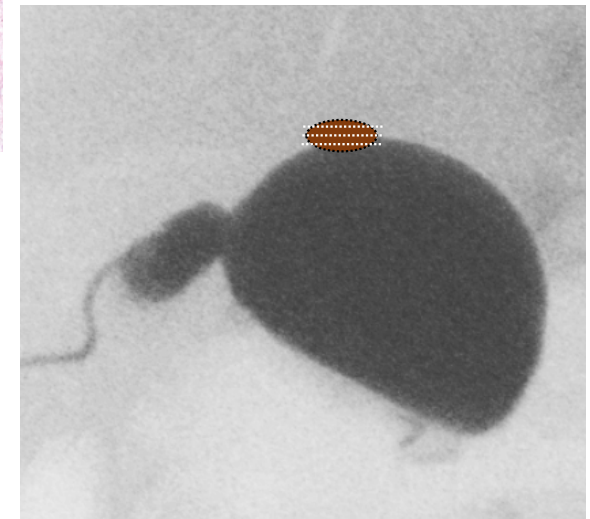
Obstruction progresses from downstream to upstream?



Upstream



Downstream



Hypothesis

Cystic BA originates from CBD with a NS

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

