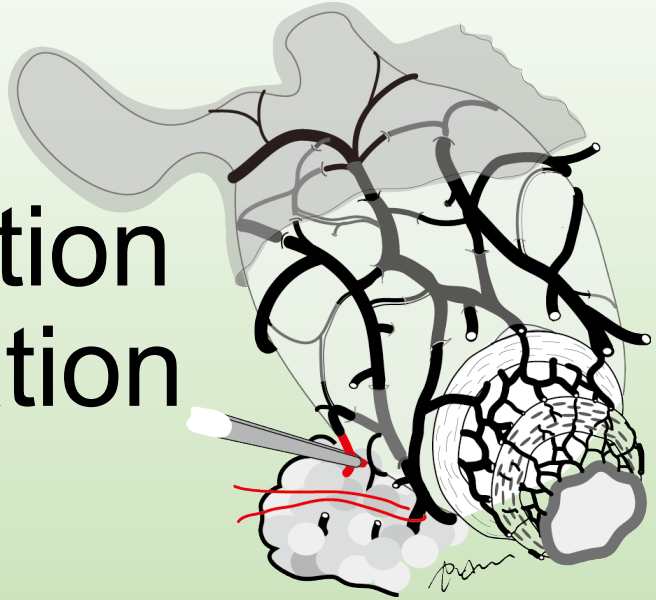
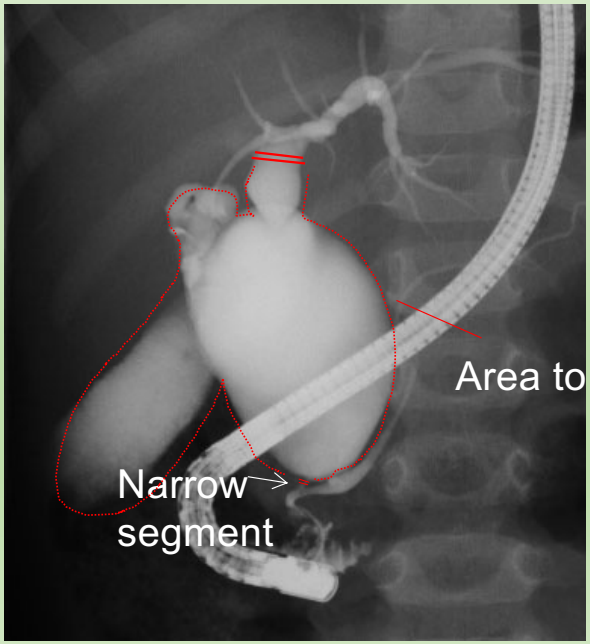
Atlas of Pancreaticobiliary Maljunction & Congenital Biliary Dilatation





Japanese Study Group on Pancreaticobiliary Maljunction (JSGPBM)
Scientific Committee

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In collaboration with Naoto Urushihara

Preface

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

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

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

September 2022
Editor-In-Chief, Kenitiro Kaneko

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Basics

Pathophysiology, diagnosis, and classification

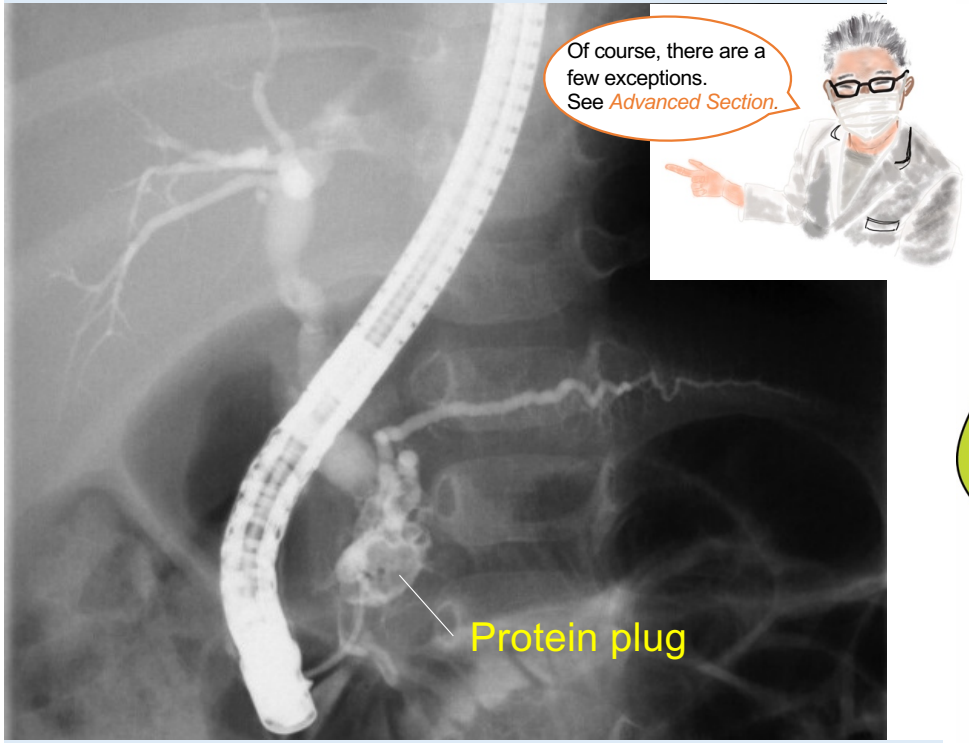
First version, September 2022

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

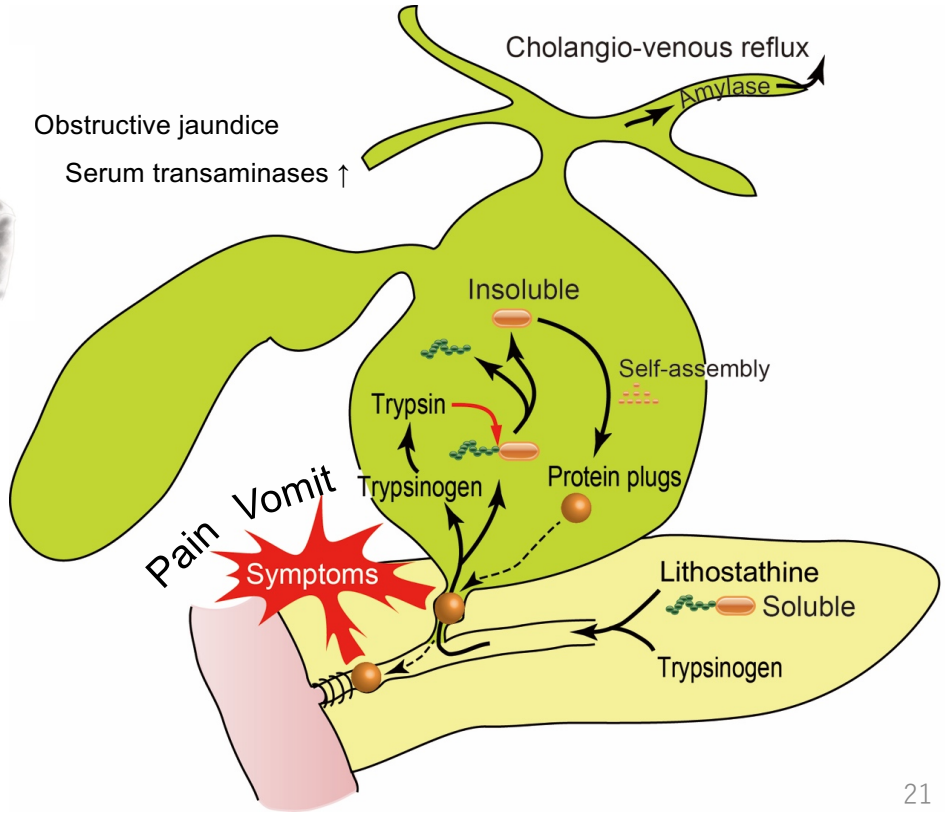
Symptomatology in childhood

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.

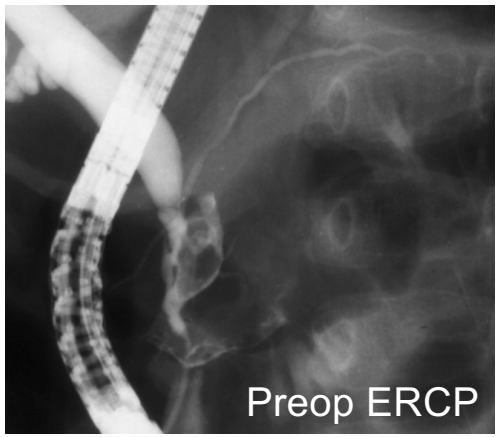
Hyperamylasemia is observed in 60% of pediatric cases but is caused by **cholangiovenous reflux** in most cases. True pancreatitis is rare (7%). See two slides ahead.

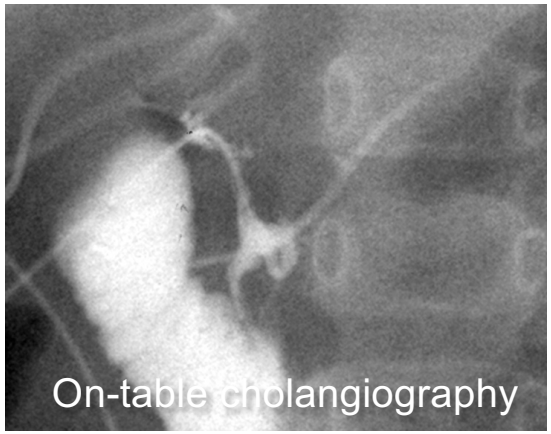


Kaneko K, et al. *Dig Dis Sci* 2007;52:1979-86

Protein plug theory explains intermittent symptoms

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


Preop ERCP


On-table cholangiography

Spontaneous dissolution

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

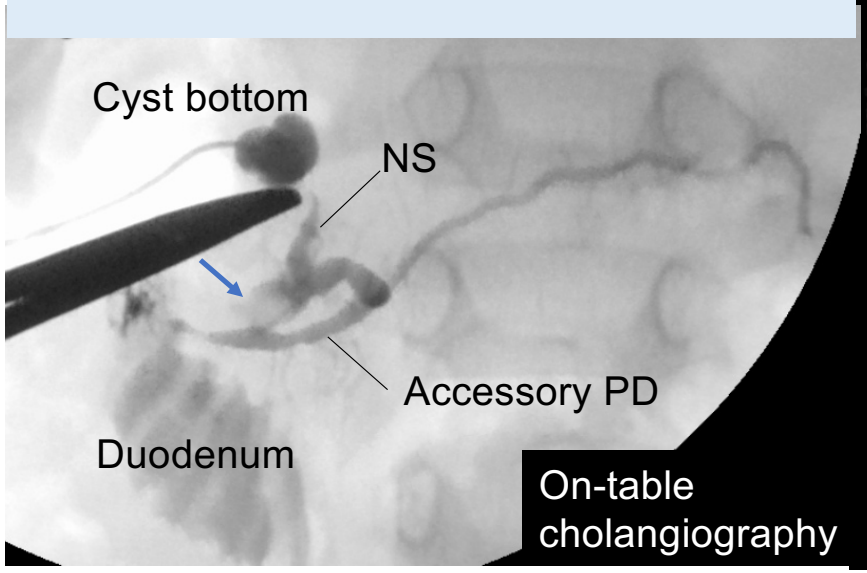


Case 1

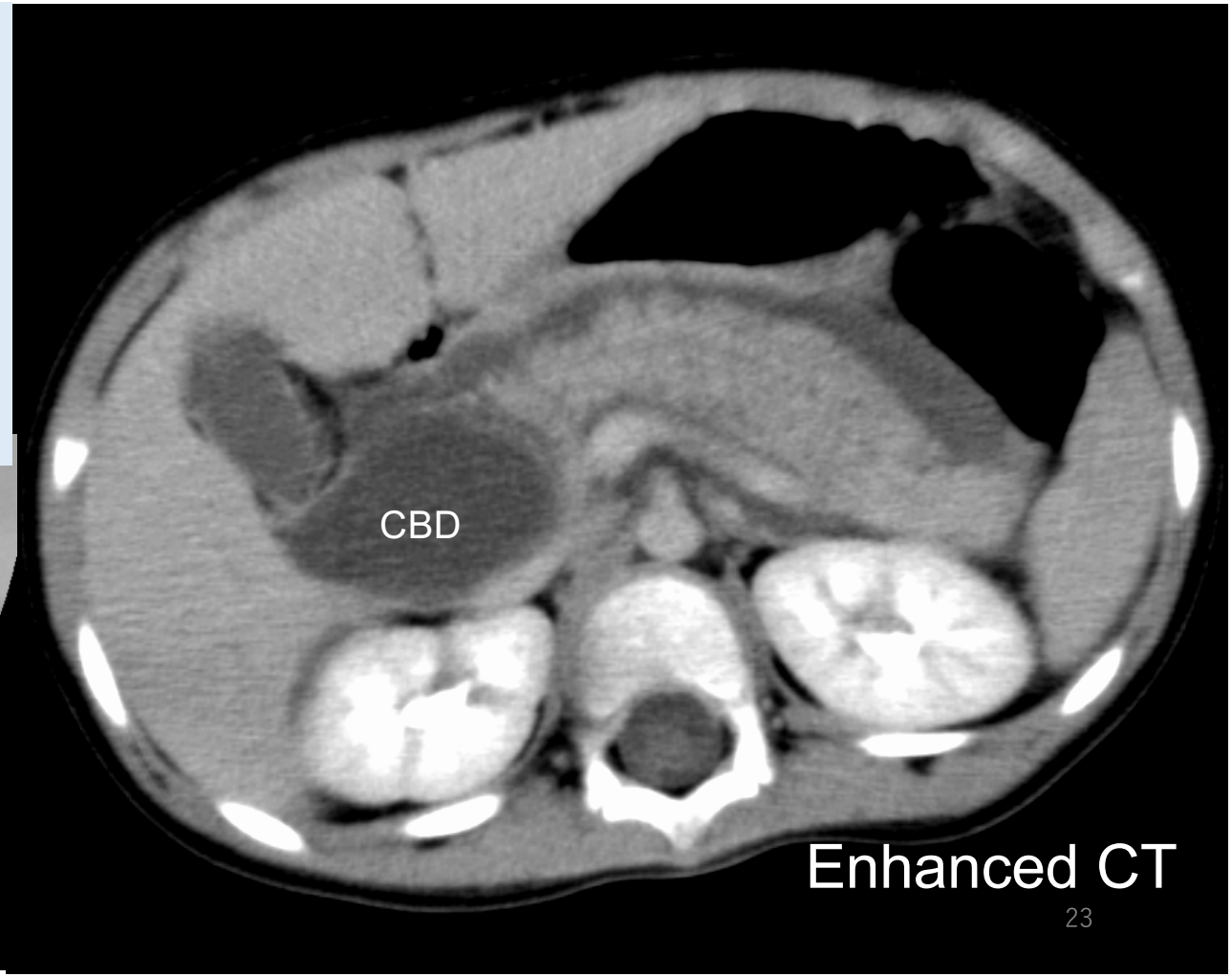
True pancreatitis is observed only in 7% of pediatric patients

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

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



On-table cholangiography

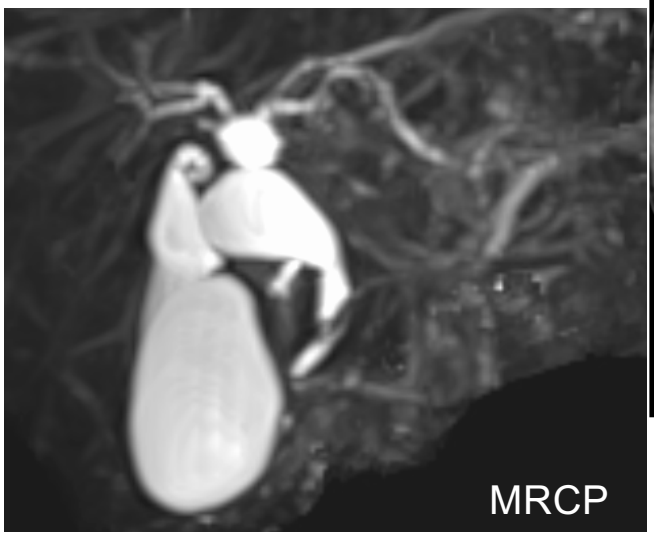


Enhanced CT

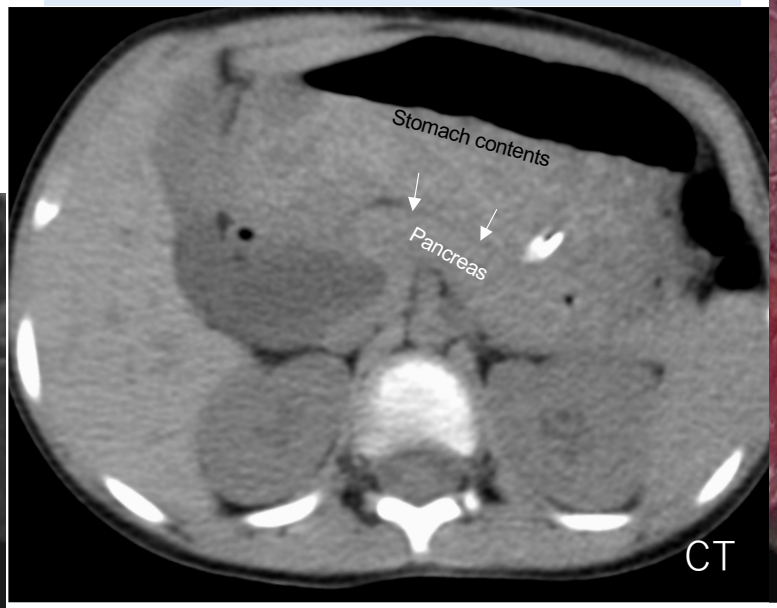
Case 2 Pseudopancreatitis Hyperamylasemia without evidence of pancreatitis

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

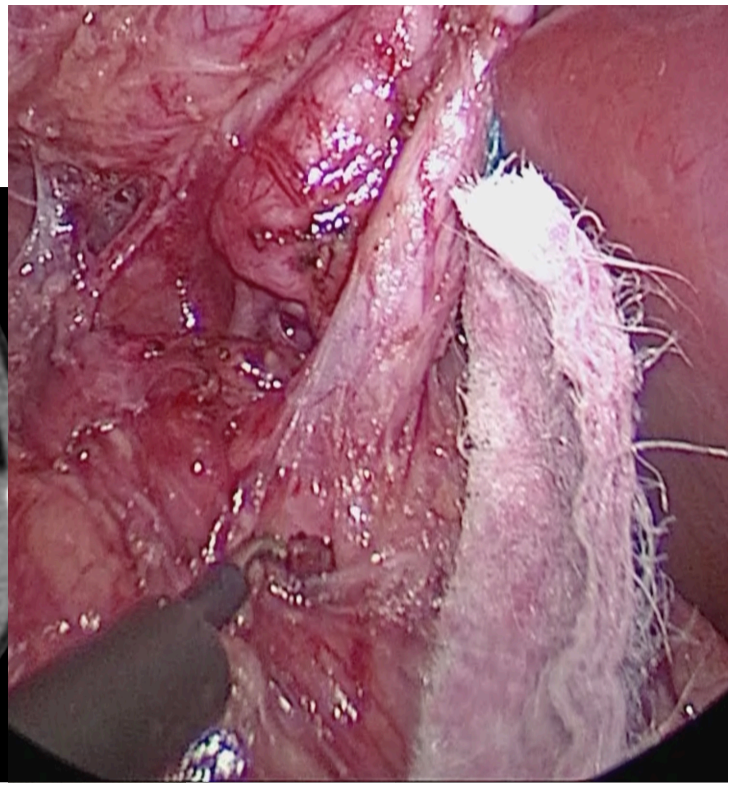
T.bil 0.7 D.bil 0.1 mg/dL
AST 176 ALT 235 IU/L
Amylase 1385 U/L



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



No swollen pancreas

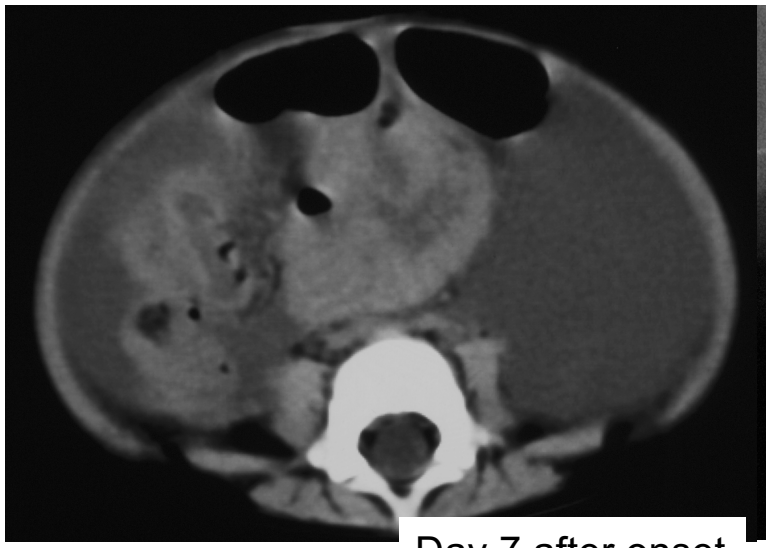


No adhesion
around the intrapancreatic BD

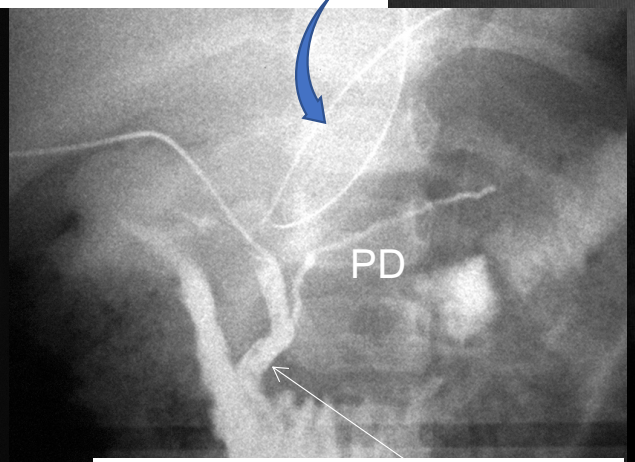
Case 3 Biliary perforation is rare: 3%–6% of children

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

Bile peritonitis causes relatively mild irritation and is often overlooked.

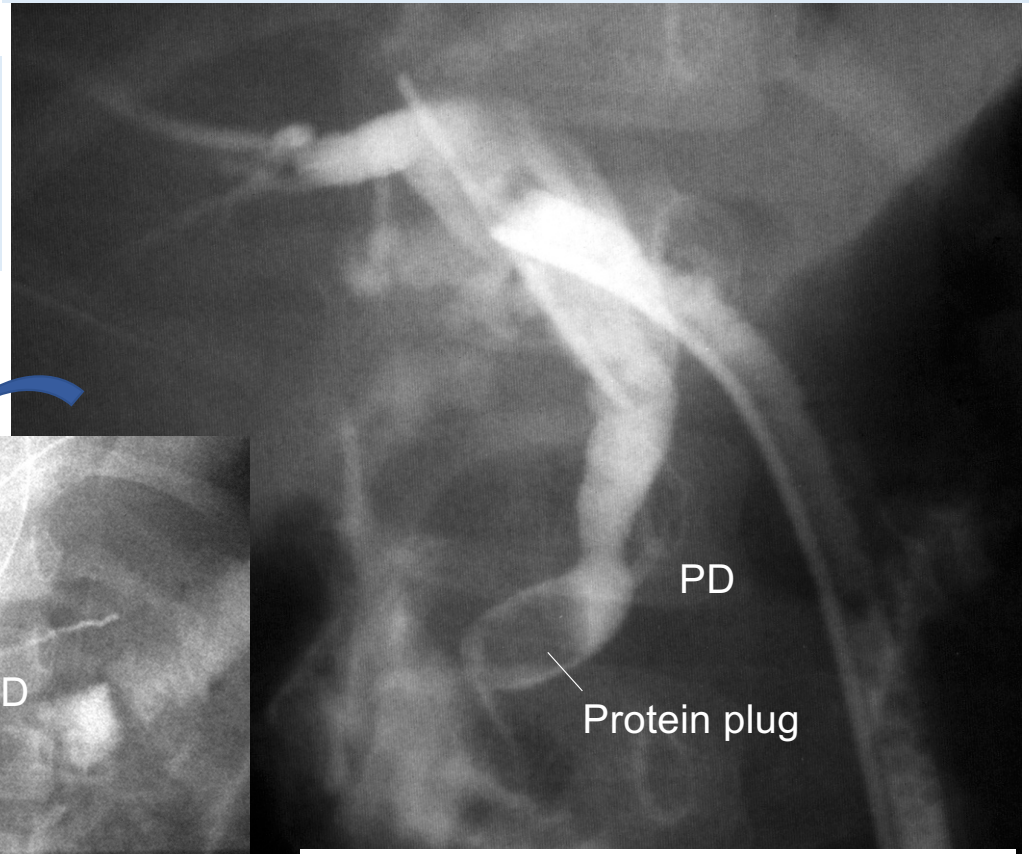


Day 7 after onset



Second OP
Protein plug had vanished

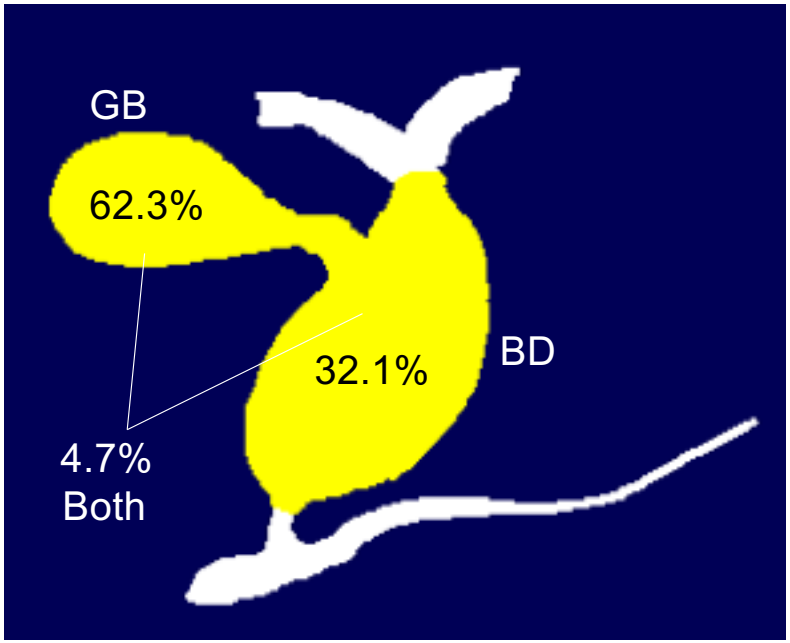
Most protein plugs are fragile and dissolve spontaneously (transient symptoms). However, exceptionally hard protein plugs produce abrupt increases in intraductal pressure, causing a rupture.

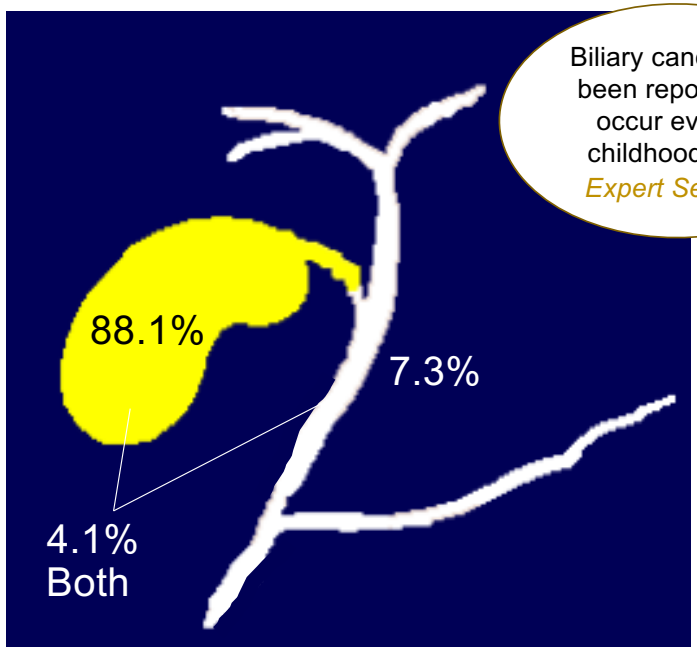


First OP on day 8 after onset
T-tube drainage through a perforation

Biliary carcinogenesis

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







Cancer rate	CBD 21.6%	Non-dilated PBM 42.4%

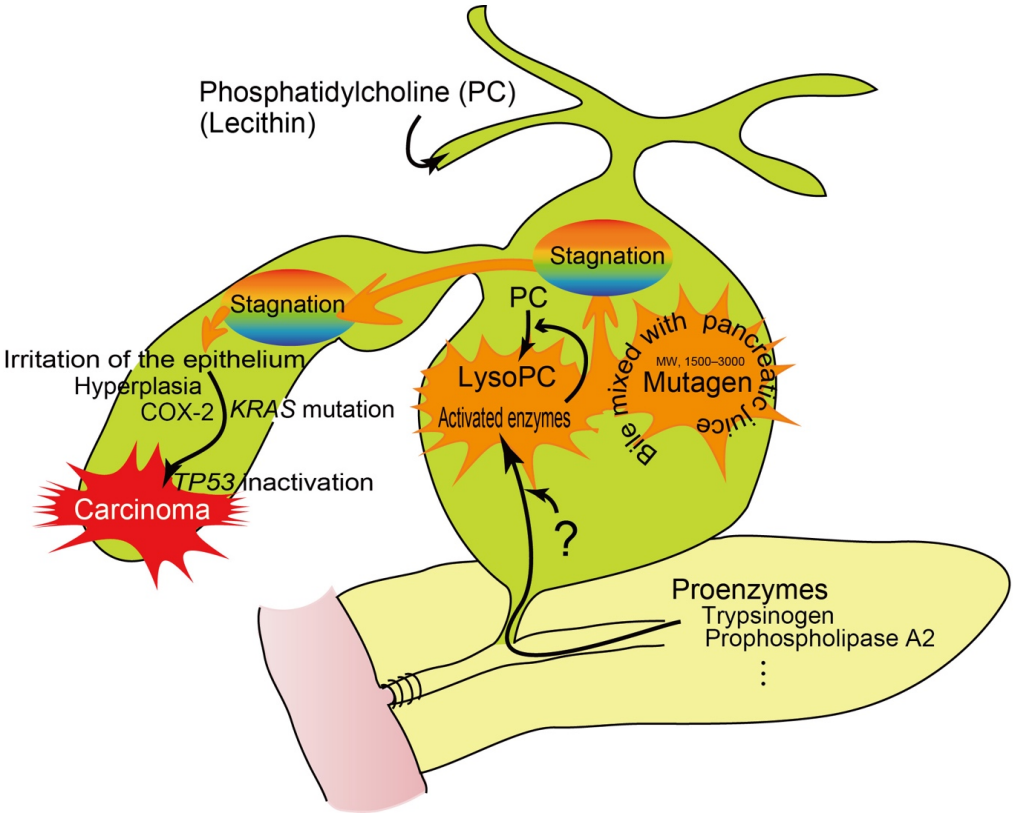
Carcinogenesis in region where bile stagnates → Indicates → Hazardous substances, produced by a mixture of bile and pancreatic juice, stagnate and cause chronic inflammation, which induces carcinogenesis

Cancer develops during adulthood

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



BD cancer in a CBD patient in her 40s



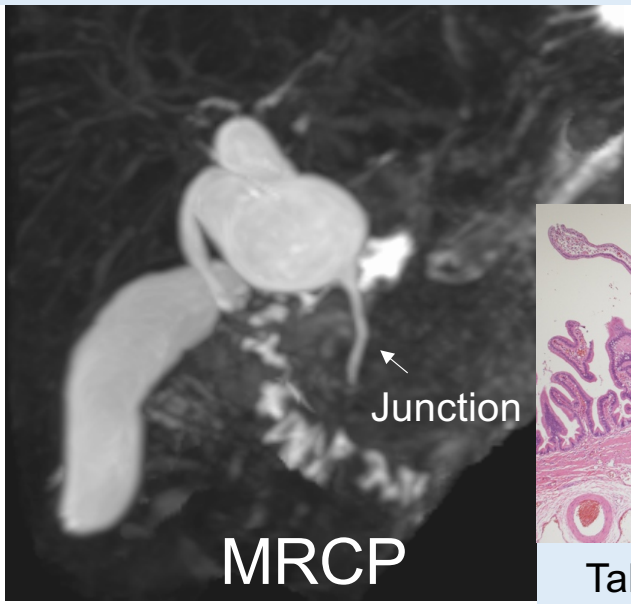
Hyperplasia–dysplasia–carcinoma sequence

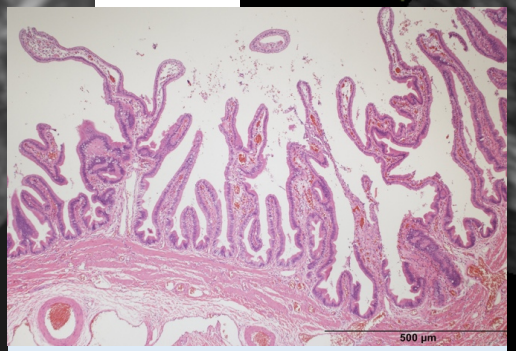
Case 4 Diffuse Hyperplasia of GB mucosa observed in a child

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

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

Thick wall of GB on ultrasonography (US)

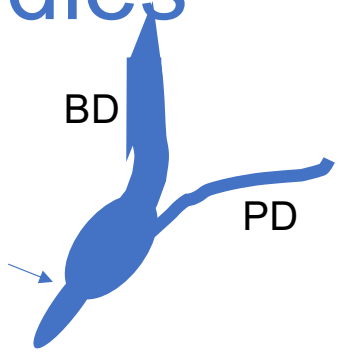




Tall GB epithelial folds

Diagnosis of PBM by imaging studies

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



Notch, indentation made by sphincter action

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

Imaging diagnosis

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

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

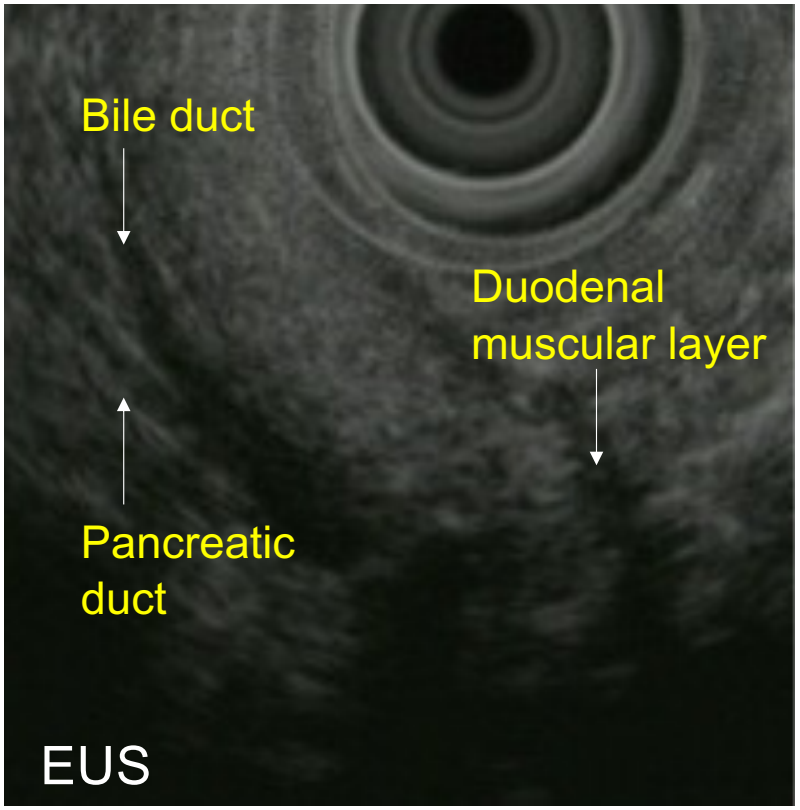
Case 5

EUS directly demonstrates the junction outside the duodenal wall

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

EUS, endoscopic ultrasonography



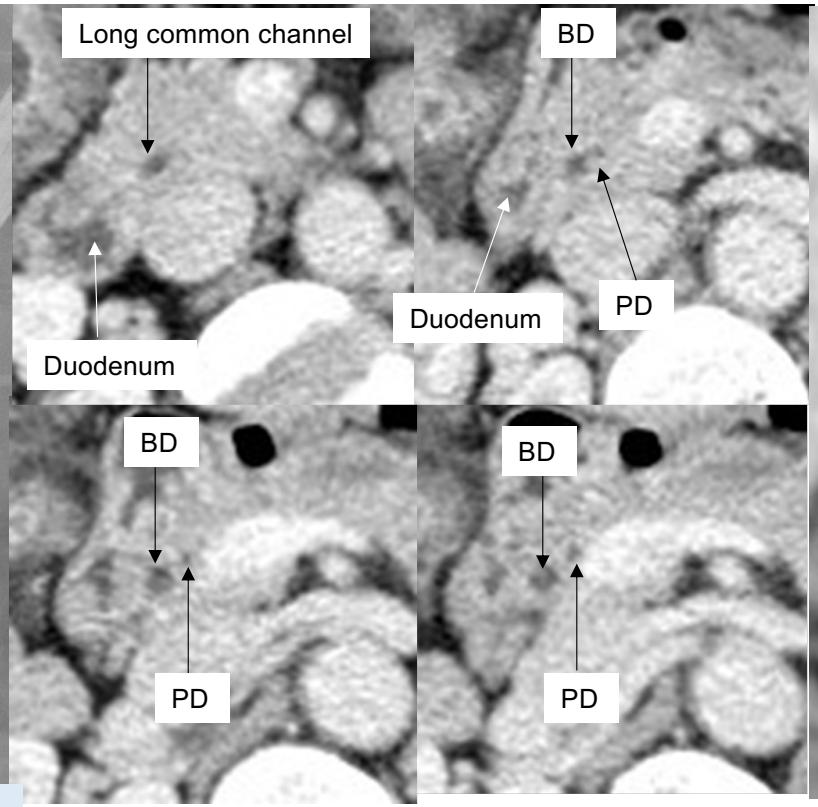


Case 6

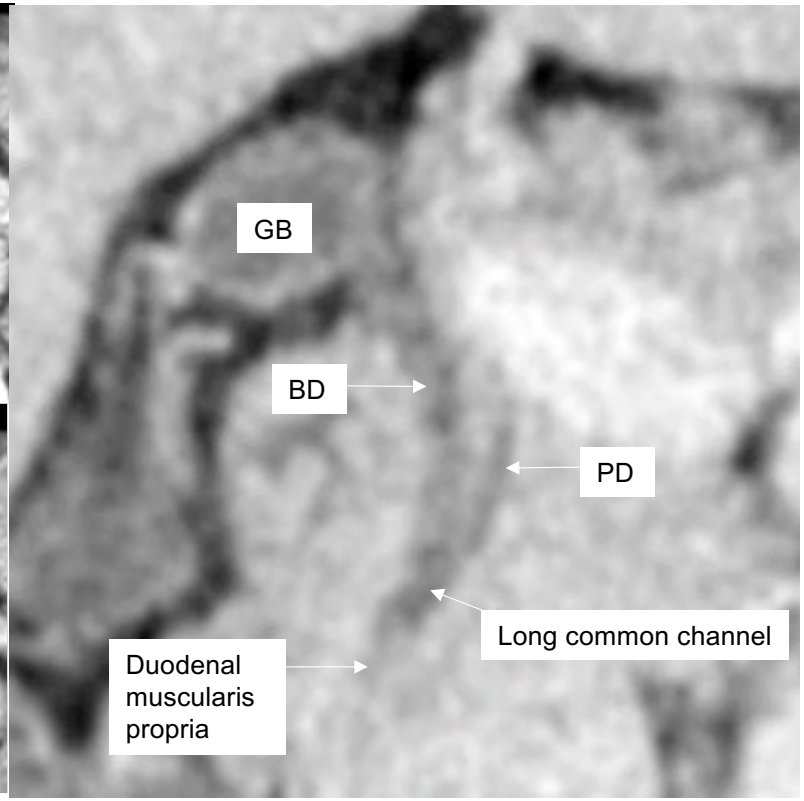
CT directly demonstrates the junction outside the duodenal wall



A female in her 40s
Right hypochondrial pain

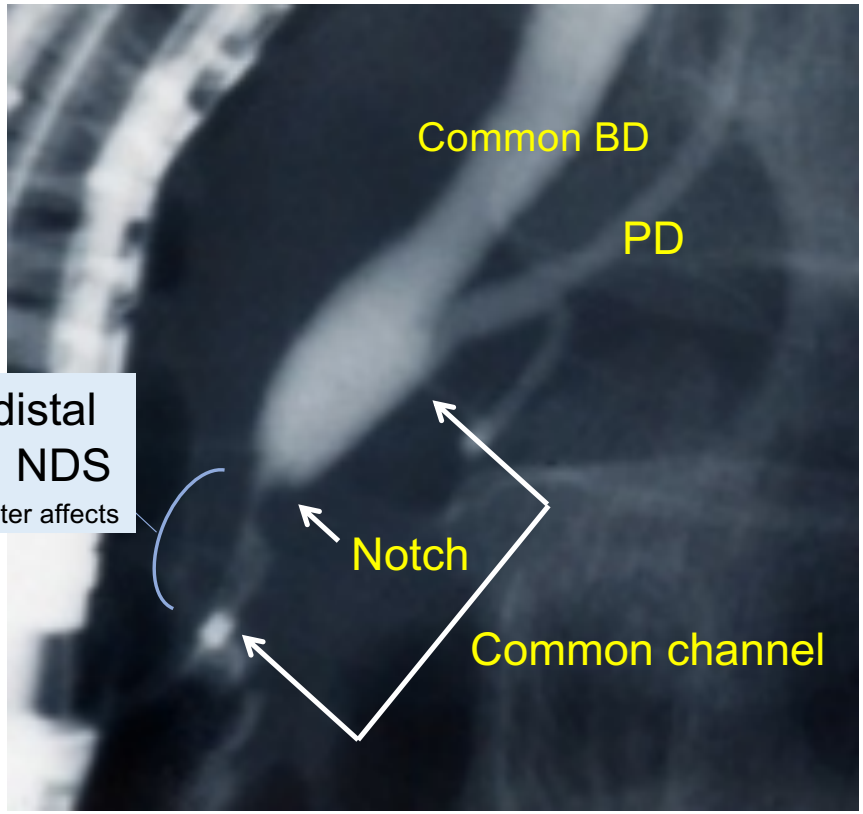


Axial section



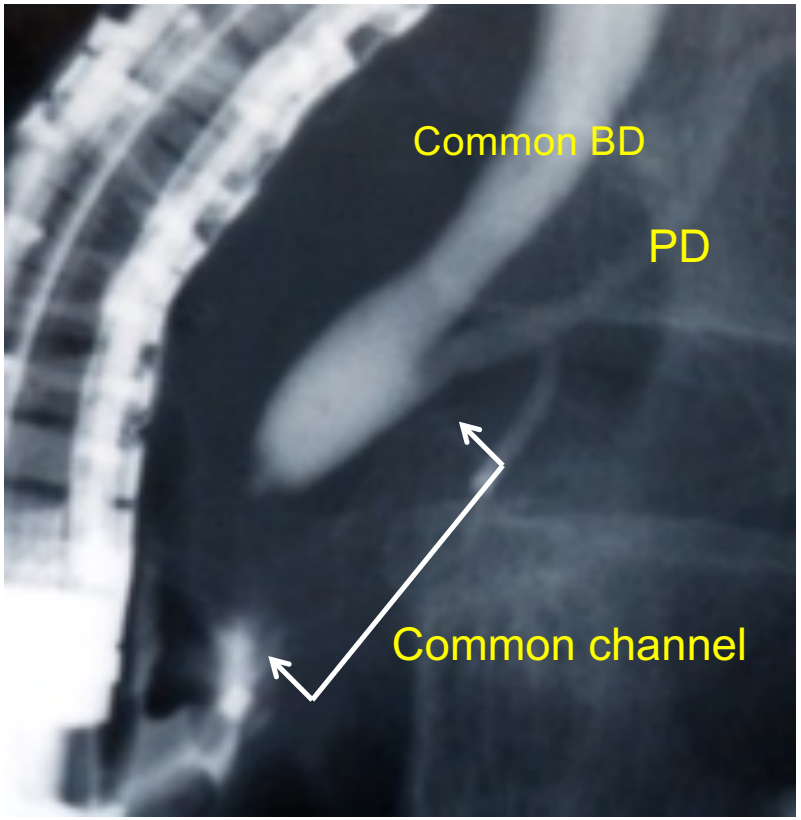
Coronal section

Notch on the common channel = PBM



Narrow distal segment, NDS
Area the sphincter affects

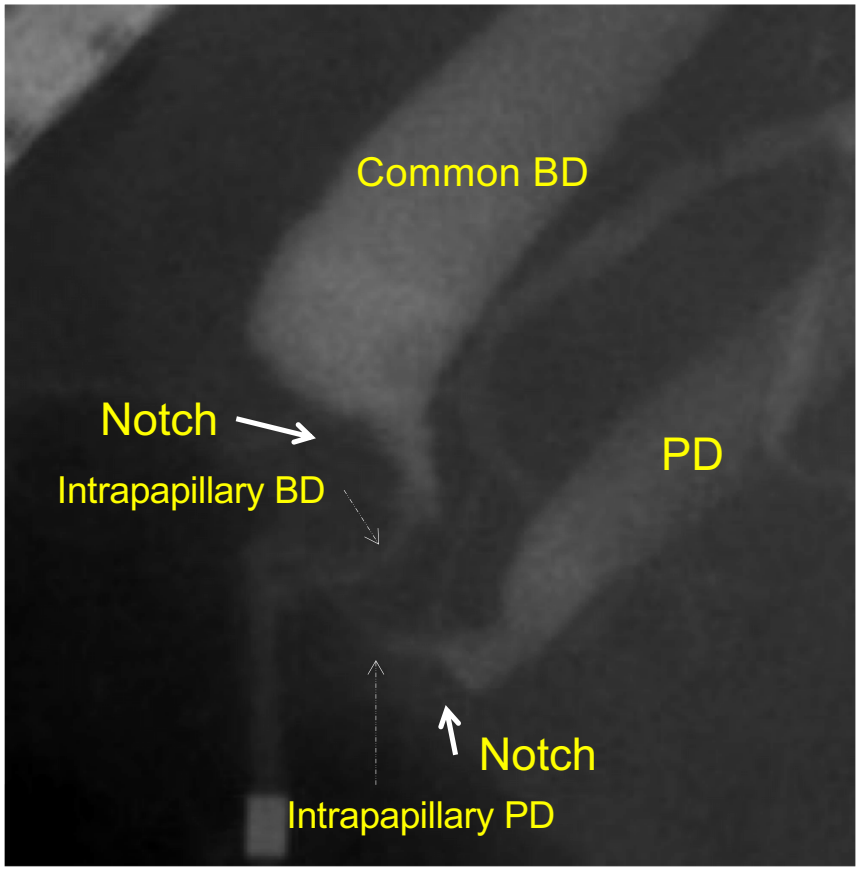
Relaxation phase of Oddi's sphincter



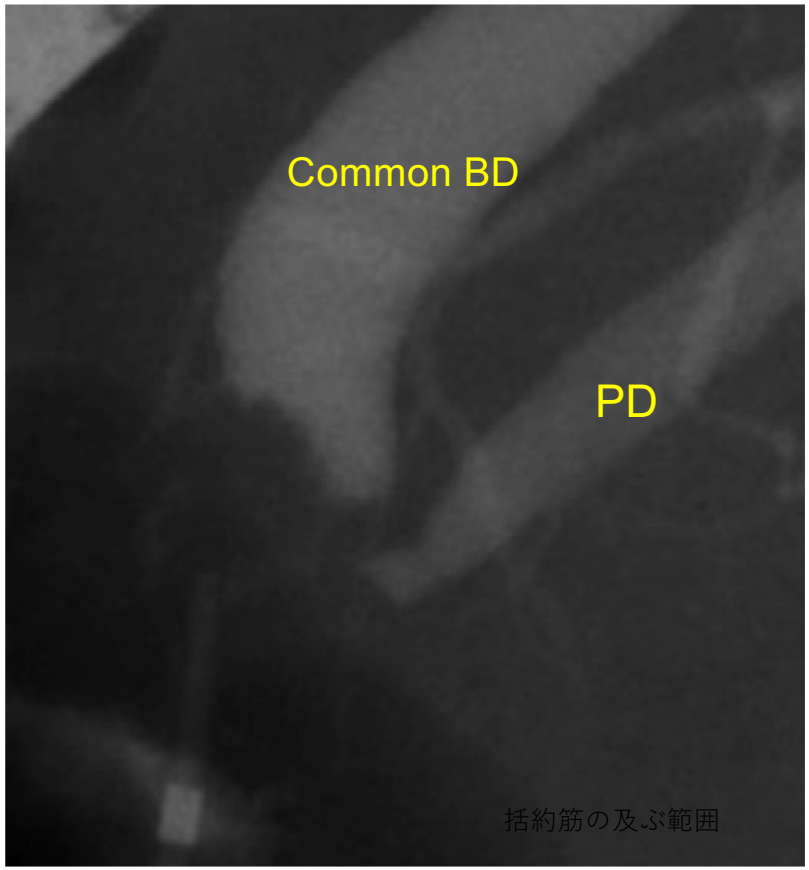
Contraction phase of Oddi's sphincter

Normal control

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



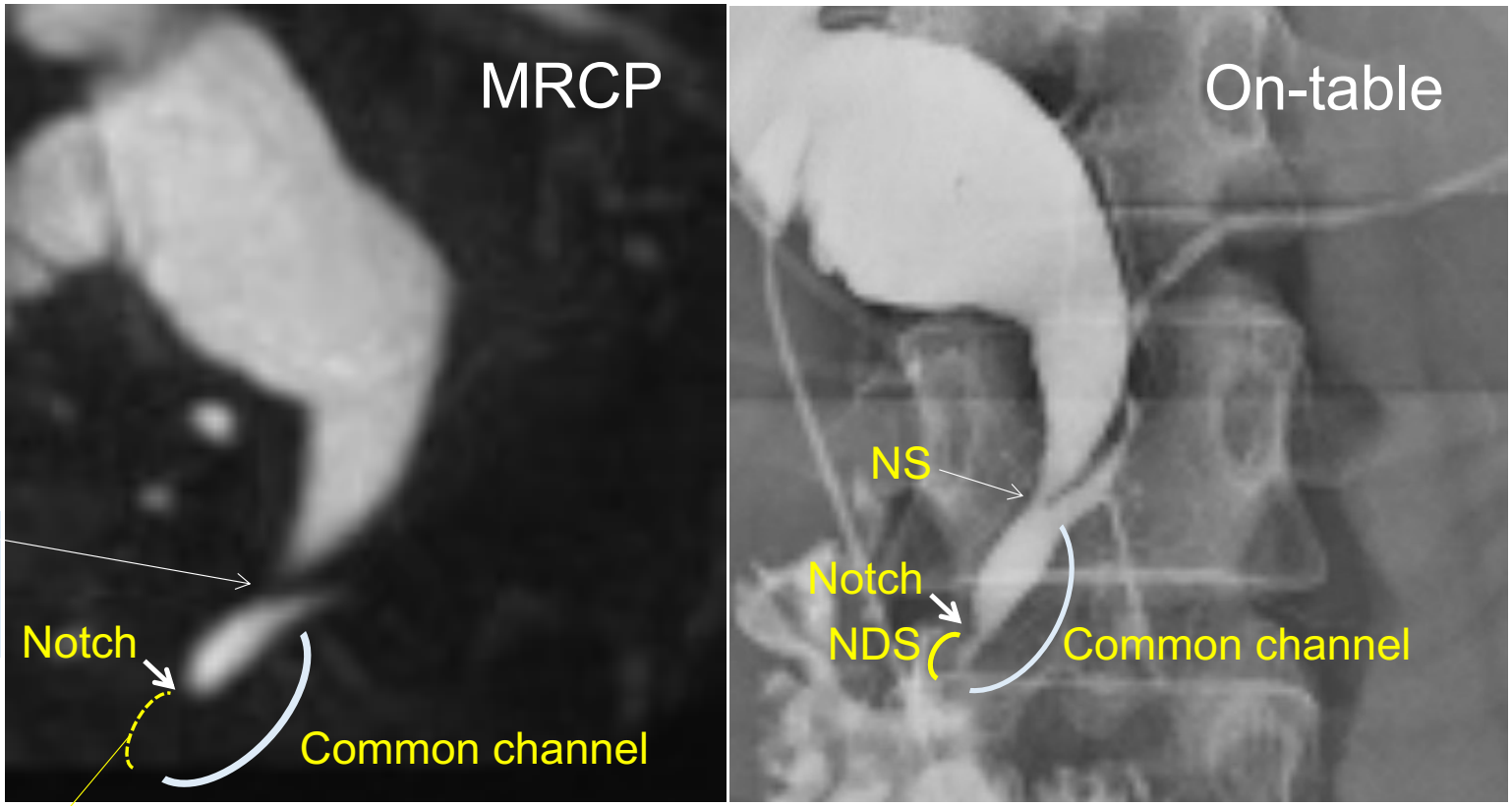
Relaxation phase
of Oddi's sphincter



Contraction phase
of Oddi's sphincter

MRCP often does not depict NDS

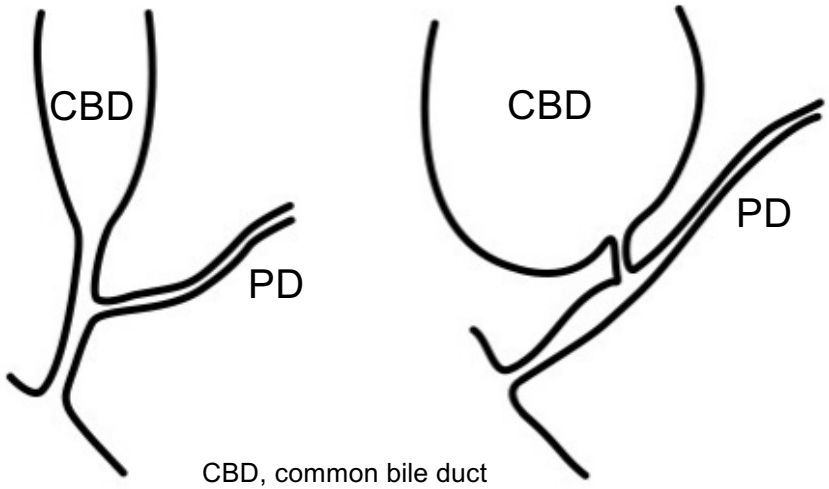
Narrow distal segment (NDS) is the area the sphincter affects



Old Classification of PBM

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

Kimura's classification 1976



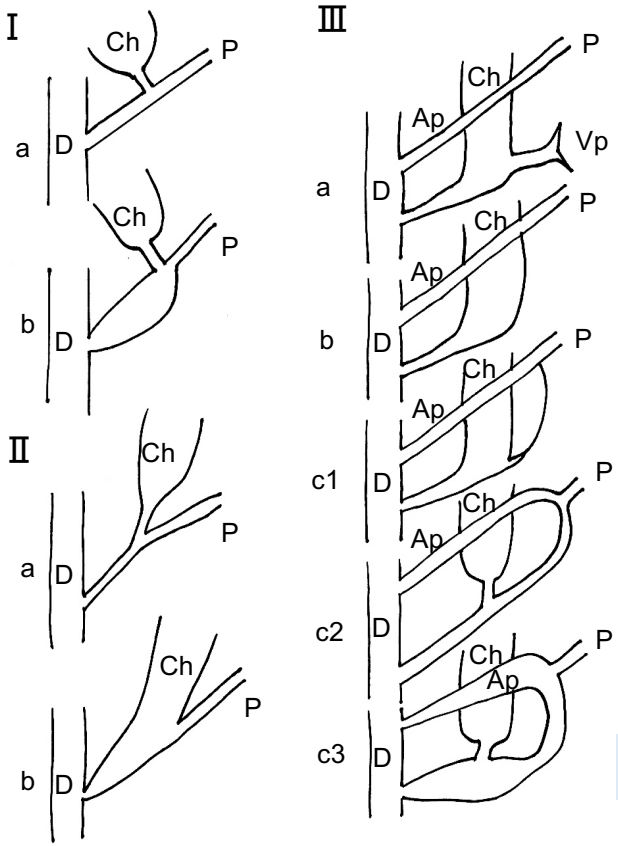
CBD, common bile duct

PC type
PD enters CBD

CP type
CBD enters PD

Remains basic even today

Komi's revised classification 1991



→ Divisum

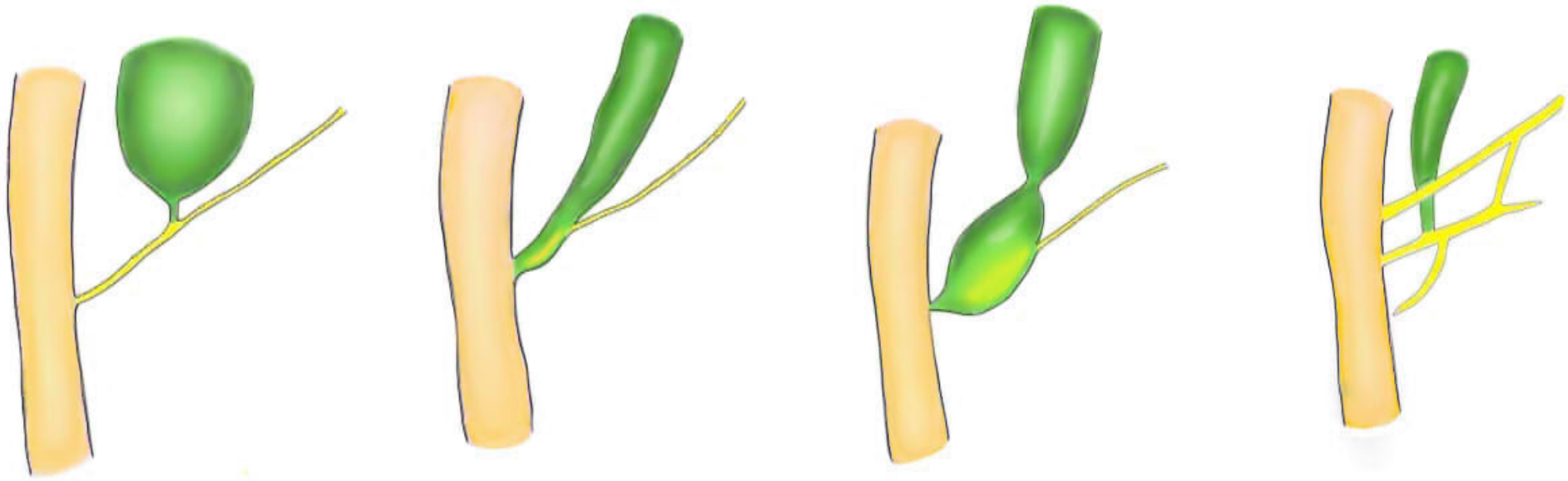
Complicated

Ch, choledochus; Ap, accessory PD

Classification by JSGPBM

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

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



Type A
Stenotic type

Type B
Non-stenotic type

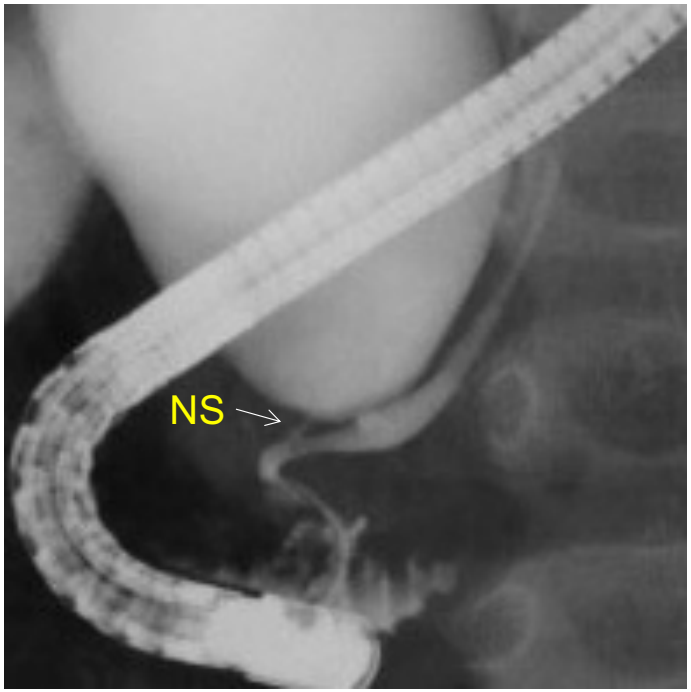
Type C
Dilated channel type

Type D
Complex type

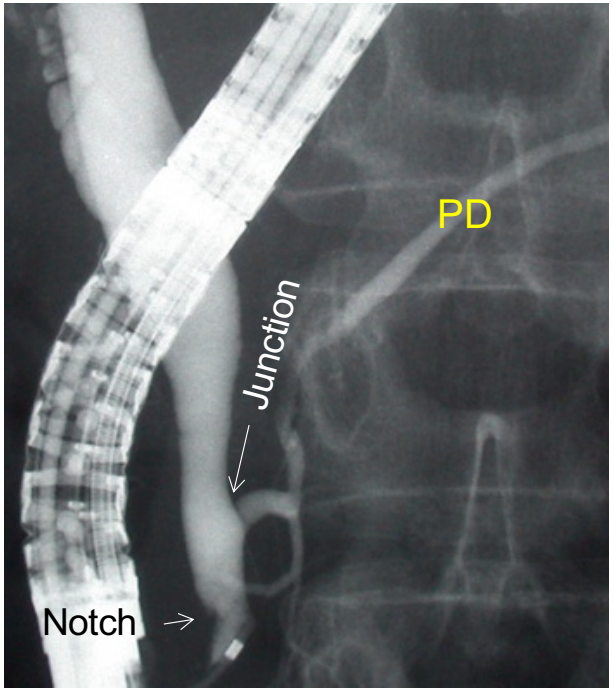
This classification is
indifferent to existence of accessory PD (Santorini's duct)

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

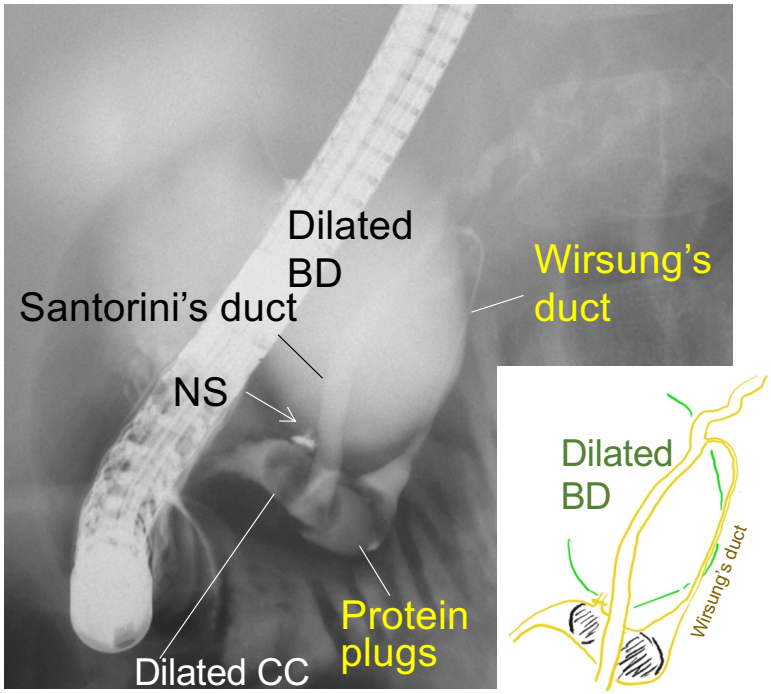
Classification by JSGPBM



Type A
Stenotic type



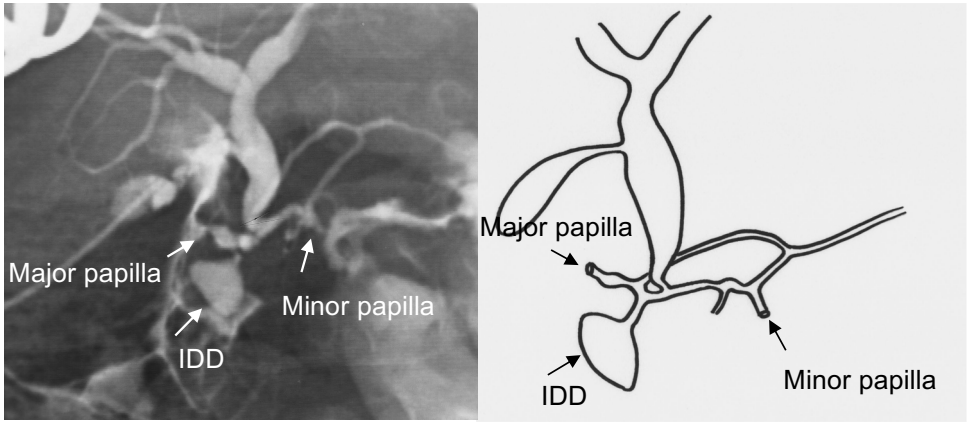
Type B
Non-stenotic type

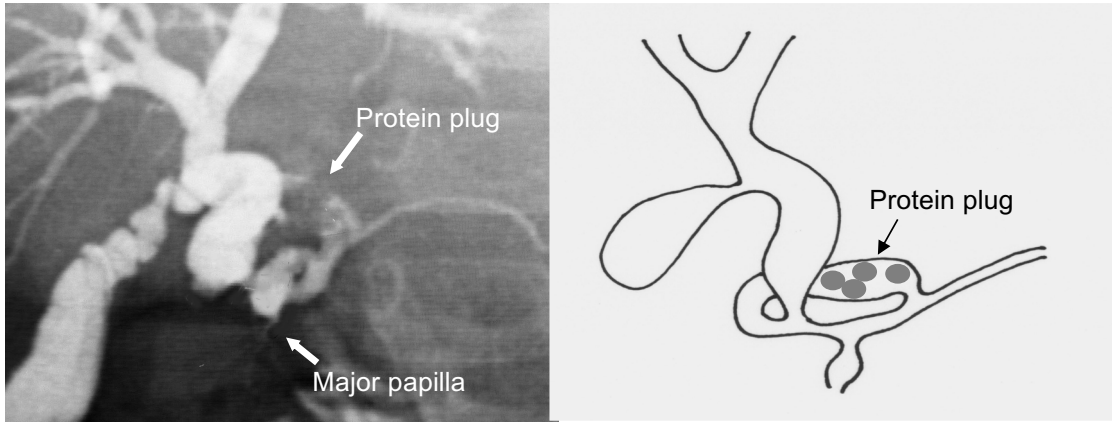


Type C
Dilated channel type

Type D: Complex type

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





IDD, intraluminal duodenal diverticulum

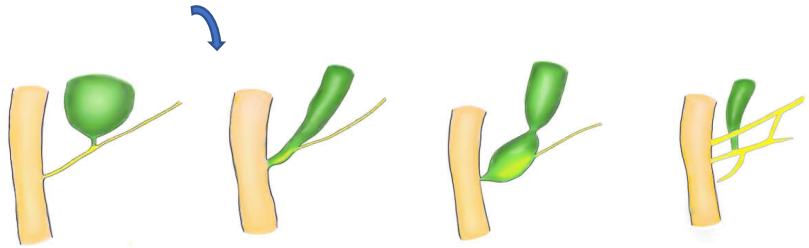
A 1-year-old girl:
Annular pancreas and intraluminal duodenal diverticulum

A 4-year-old boy:
Annular pancreas

JSGPBM classification reflects clinical characteristics in children

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

317 pediatric cases



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

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

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

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

Basics

What is congenital biliary dilatation (CBD)?

First version, September 2022
2nd version, September 2024

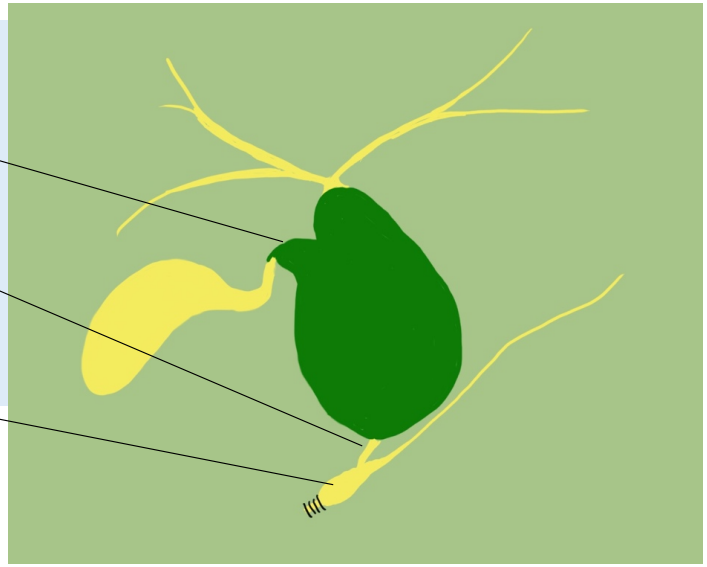
Imaging features of congenital biliary dilatation

Features that define CBD

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

Features (but not in all cases)

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



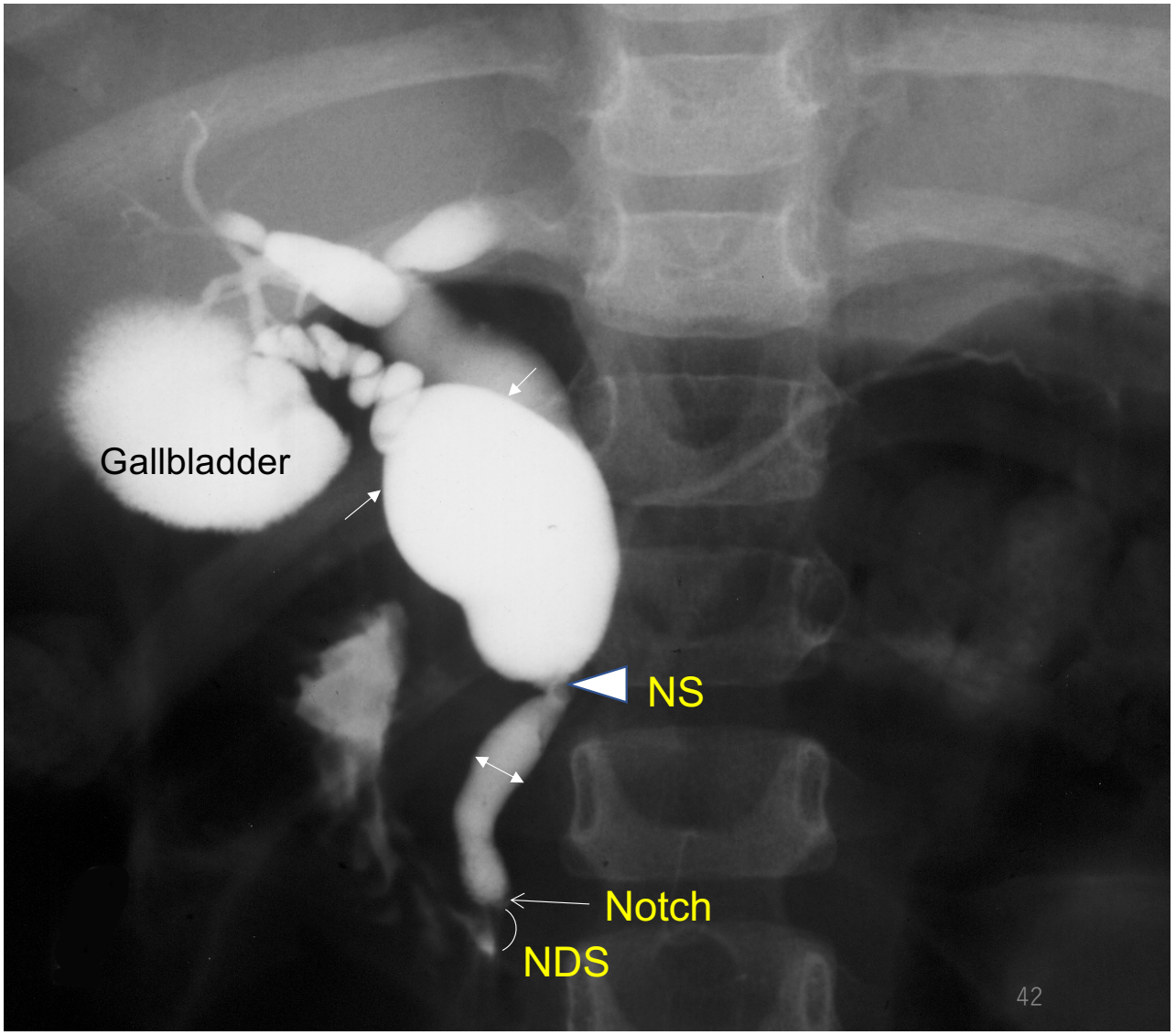
Criteria of
PBM classification by JSGPBM
See pp36–39

Case 7

Dilated cystic duct
Narrow segment
Dilated common channel

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

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

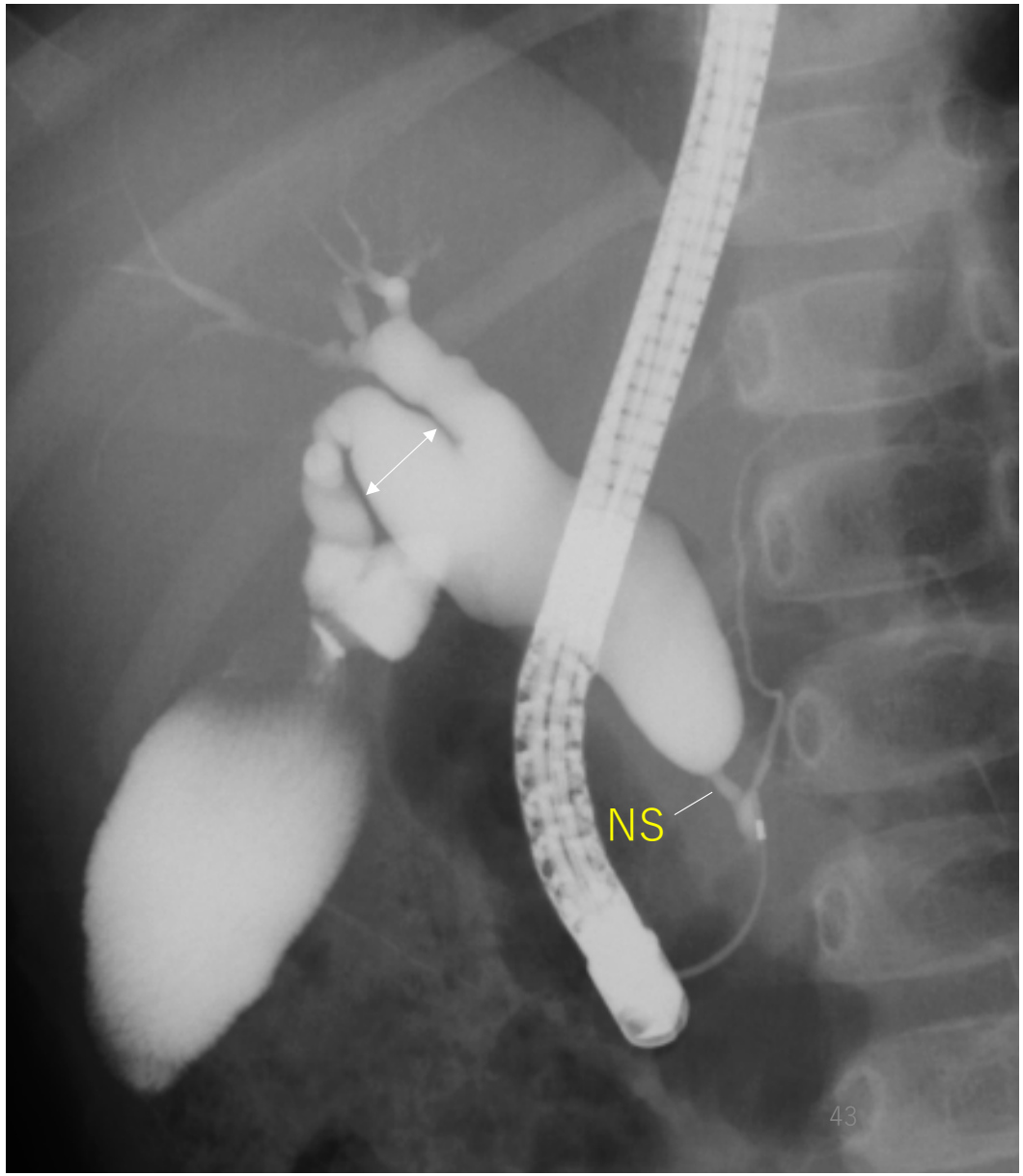


Case 8

Dilated cystic duct & long narrow segment

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

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



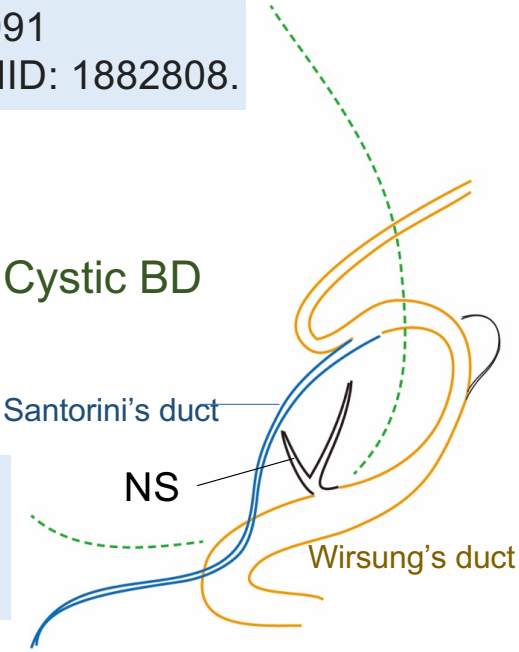
NS is a branch of the PD

Histopathological demonstration

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

Case 9

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



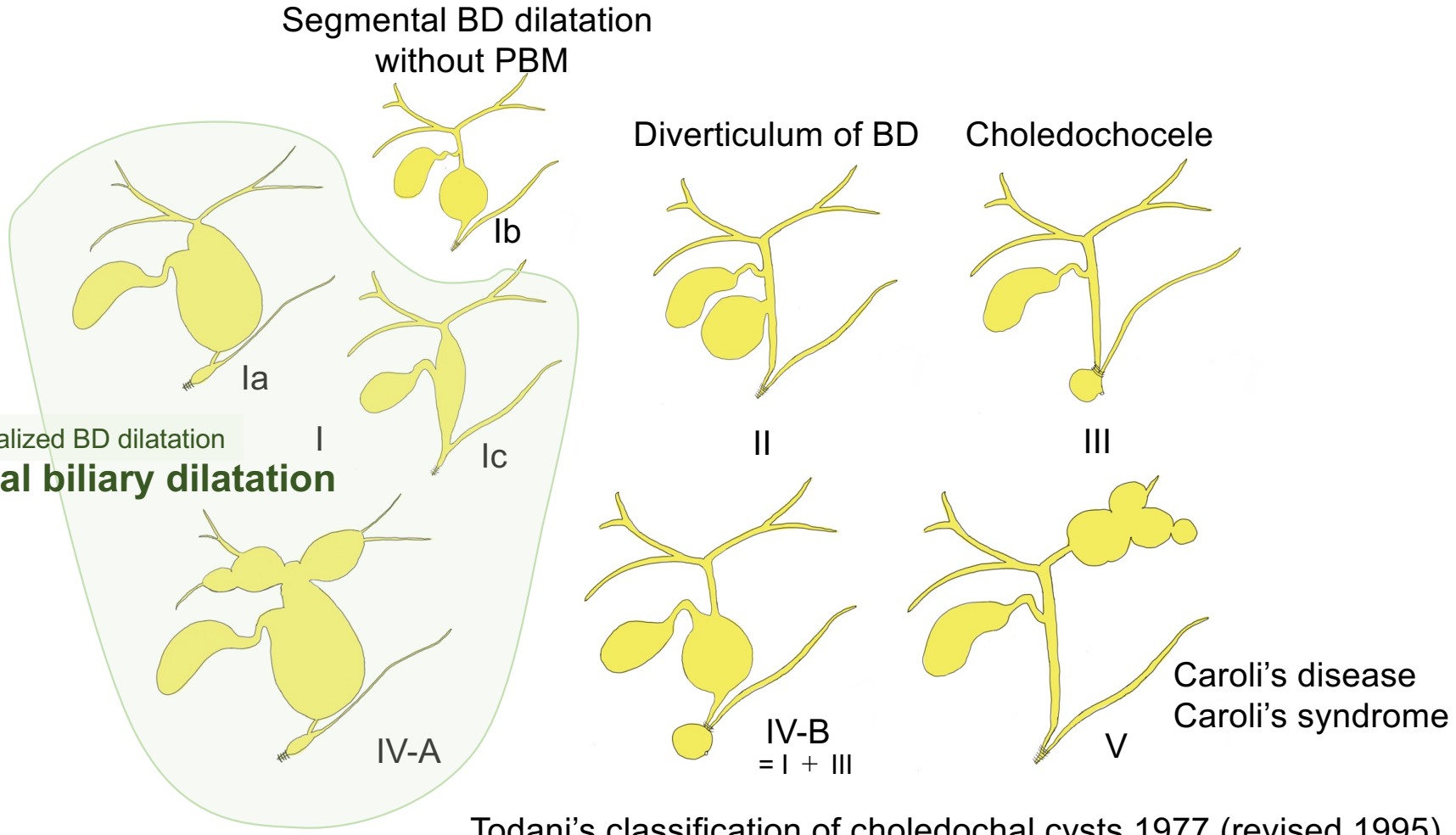
Radiological demonstration

NS is one branch of the PD



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

The term choledochal cyst involves different entities



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

Todani type I

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

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

Extremely rare
See
Expert Section

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

See Advanced Section, pp99–113

Todani type IV

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

- **Type IV** is a multiple cyst
  - **Type IVA** involves multiple cysts in both the intrahepatic and extrahepatic bile ducts. Intrahepatic dilatation affects both lobes, although the left lobe is predominantly involved.
    Based on the shape of the extrahepatic and intrahepatic ducts, type IV is subclassified into three forms, **cystic–cystic**, **cystic–fusiform**, and **fusiform–fusiform**.
  - **Type IVB** refers to multiple dilatations confined only to the extrahepatic bile duct.

Extremely rare

Basics

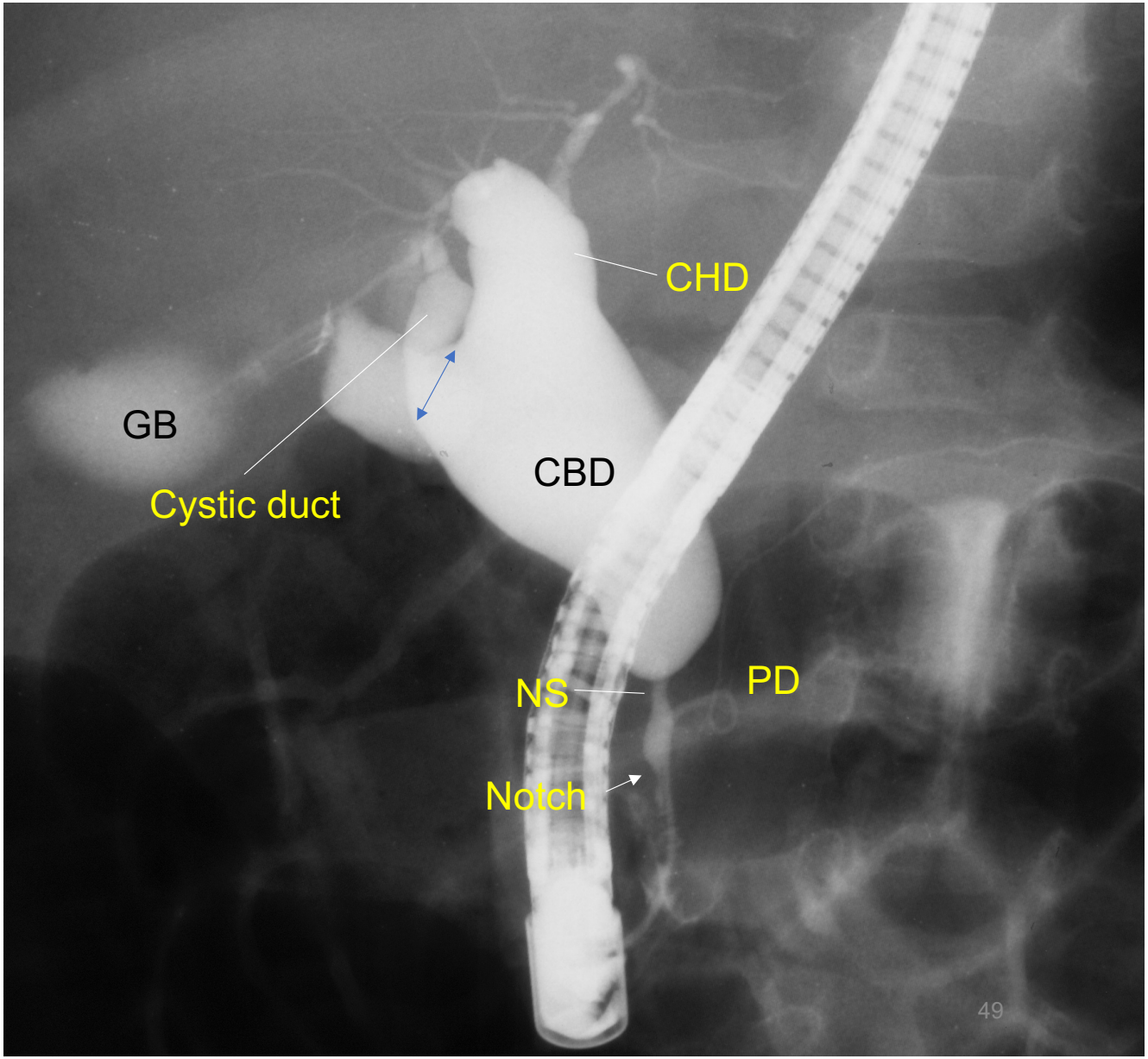
Typical congenital biliary dilatation

First version, September 2022

Case 10 Typical Ia: Pediatric case

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

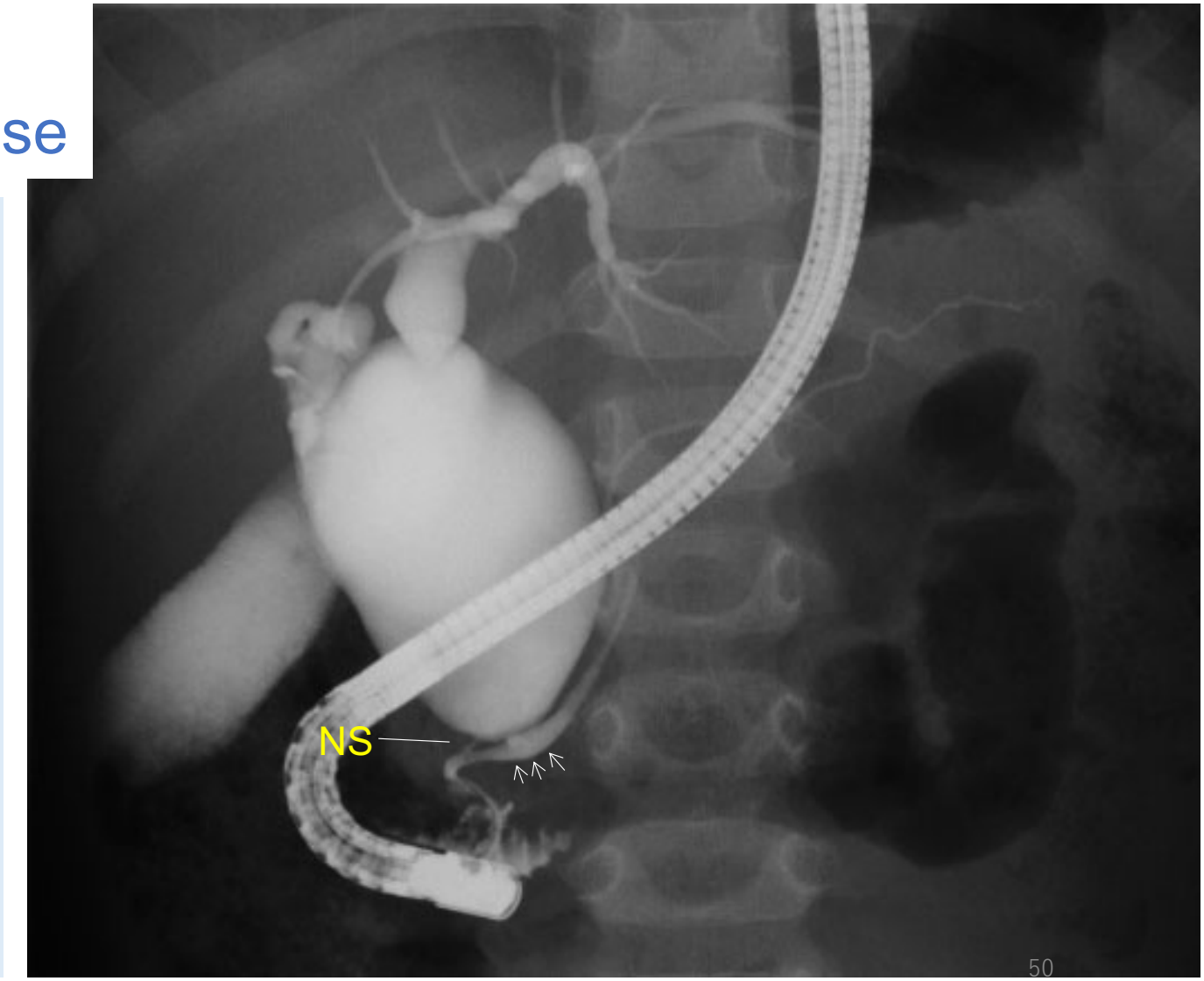
- **Cystically dilated CBD** of 21 mm in diameter
- **Dilatation involves CHD but not its upstream**
- **Cystic duct is dilated at its base** (↔) but GB is not dilated
- **Narrow segment (NS)** exists
- The common channel is not dilated
- JSGPBM classification:
  Type A, stenotic type



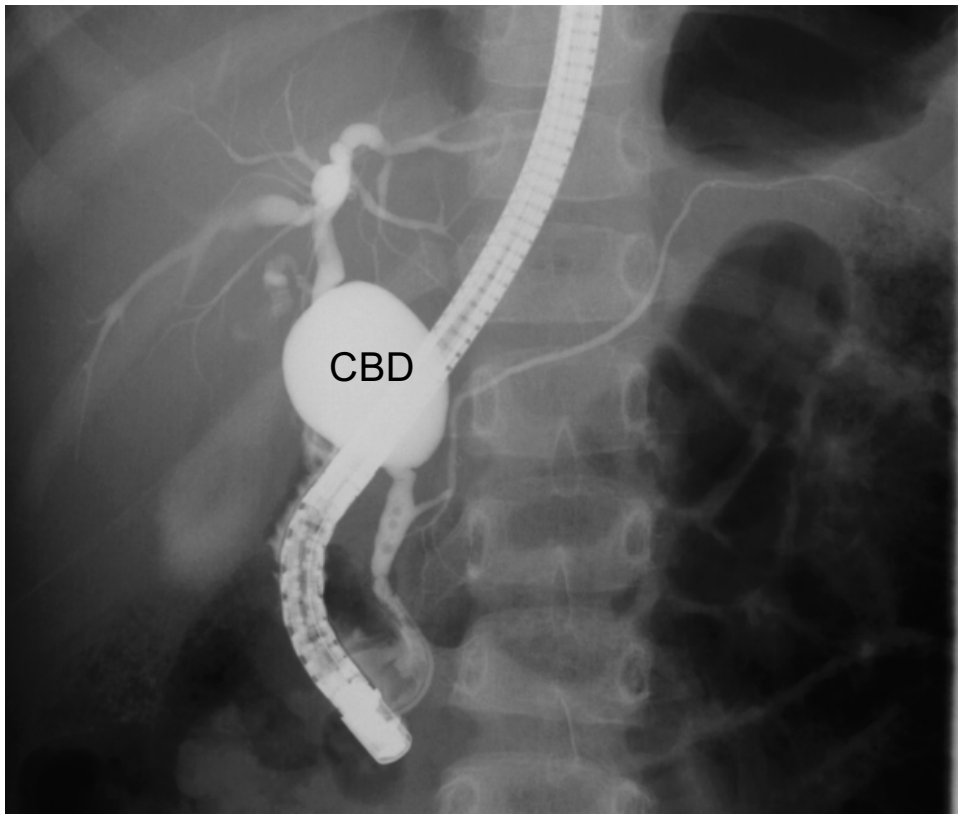
Case 11 Typical Ia: Pediatric case

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

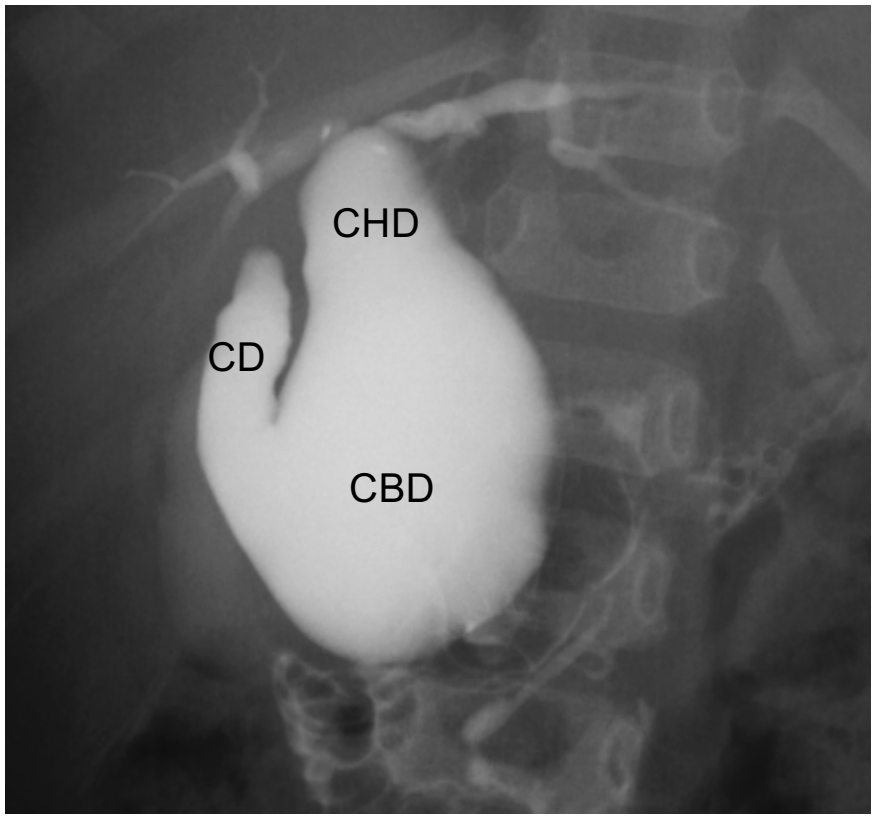
- **Cystically dilated CBD** with a diameter of 40 mm
- **Dilatation involves CHD but not its upstream processes**
- **Cystic duct is dilated at its base**, but GB is not dilated
- **NS** exists
- The common channel is not dilated
- **NS locates not at the cyst bottom but right-ventrally**
  Ventral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



Localized dilatation to various extents even within type Ia pediatric cases



Dilatation limited to the common bile duct (CBD)



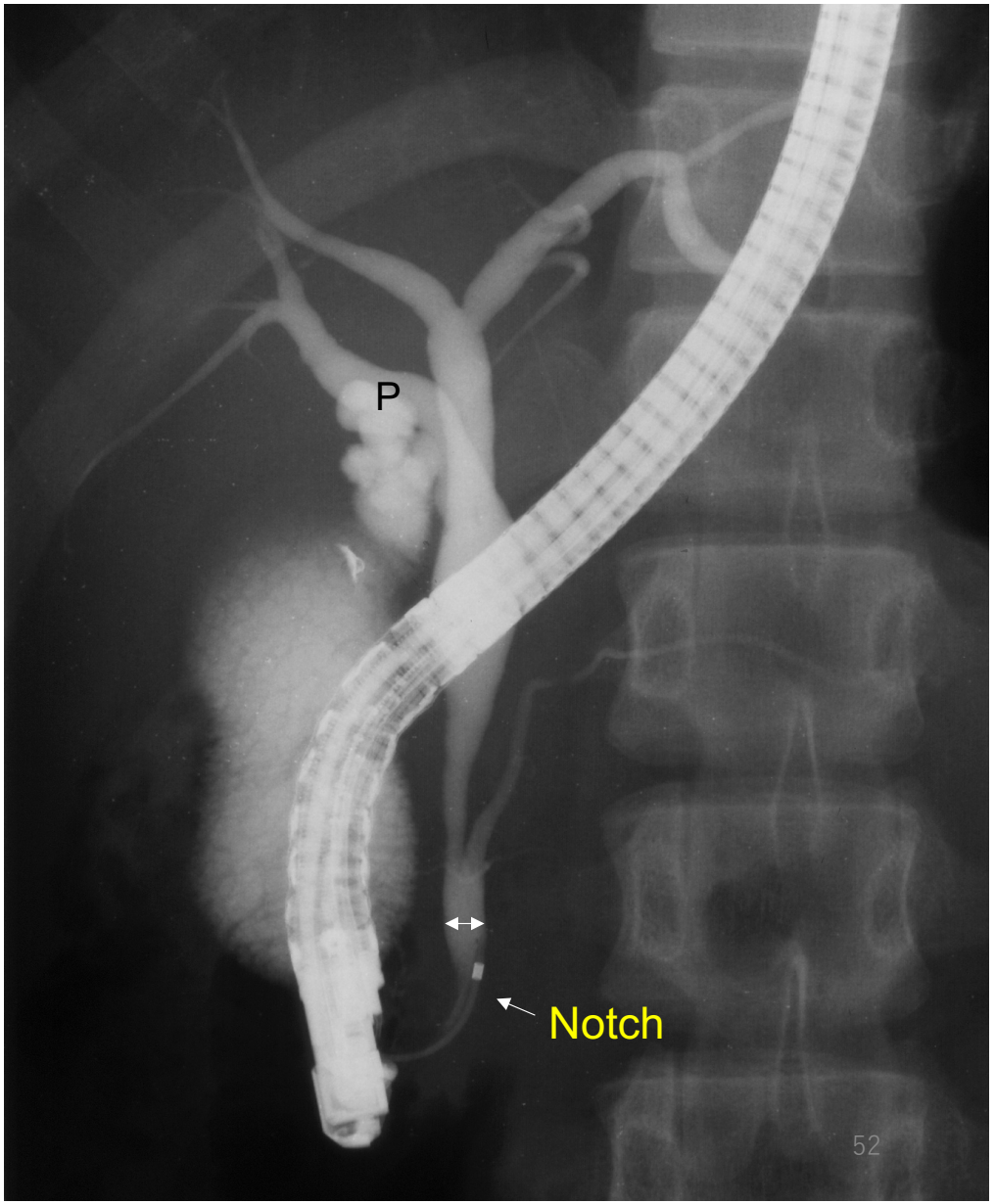
Dilatation involving CBD, CD, and CHD

Case 12

Typical Ic: Fusiform dilatation pediatric case

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

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

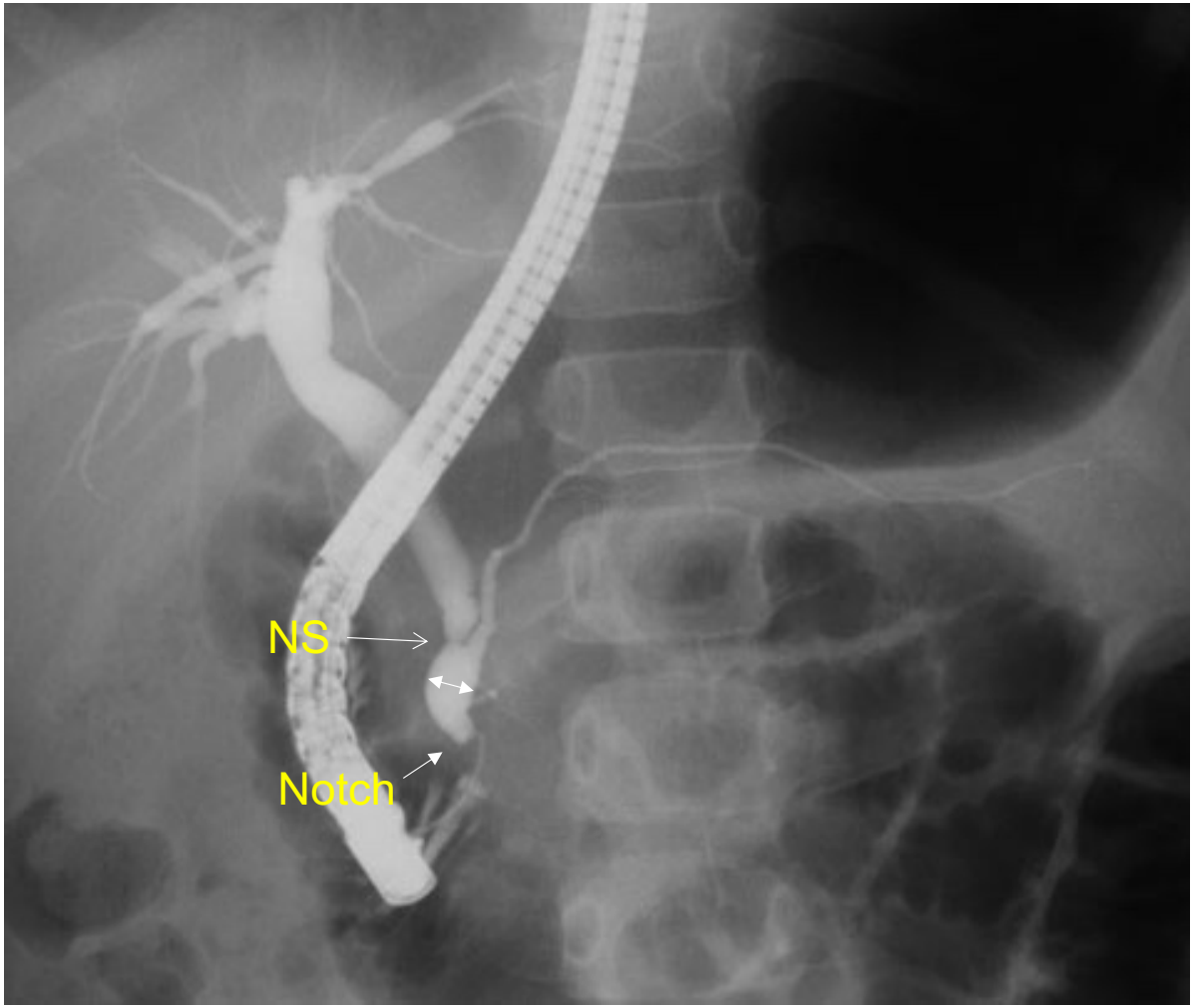


Case 13

Typical Ic: Cylindrical dilatation pediatric case

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

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



Case 14 Typical Ic: Cylindrical dilatation pediatric case

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

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



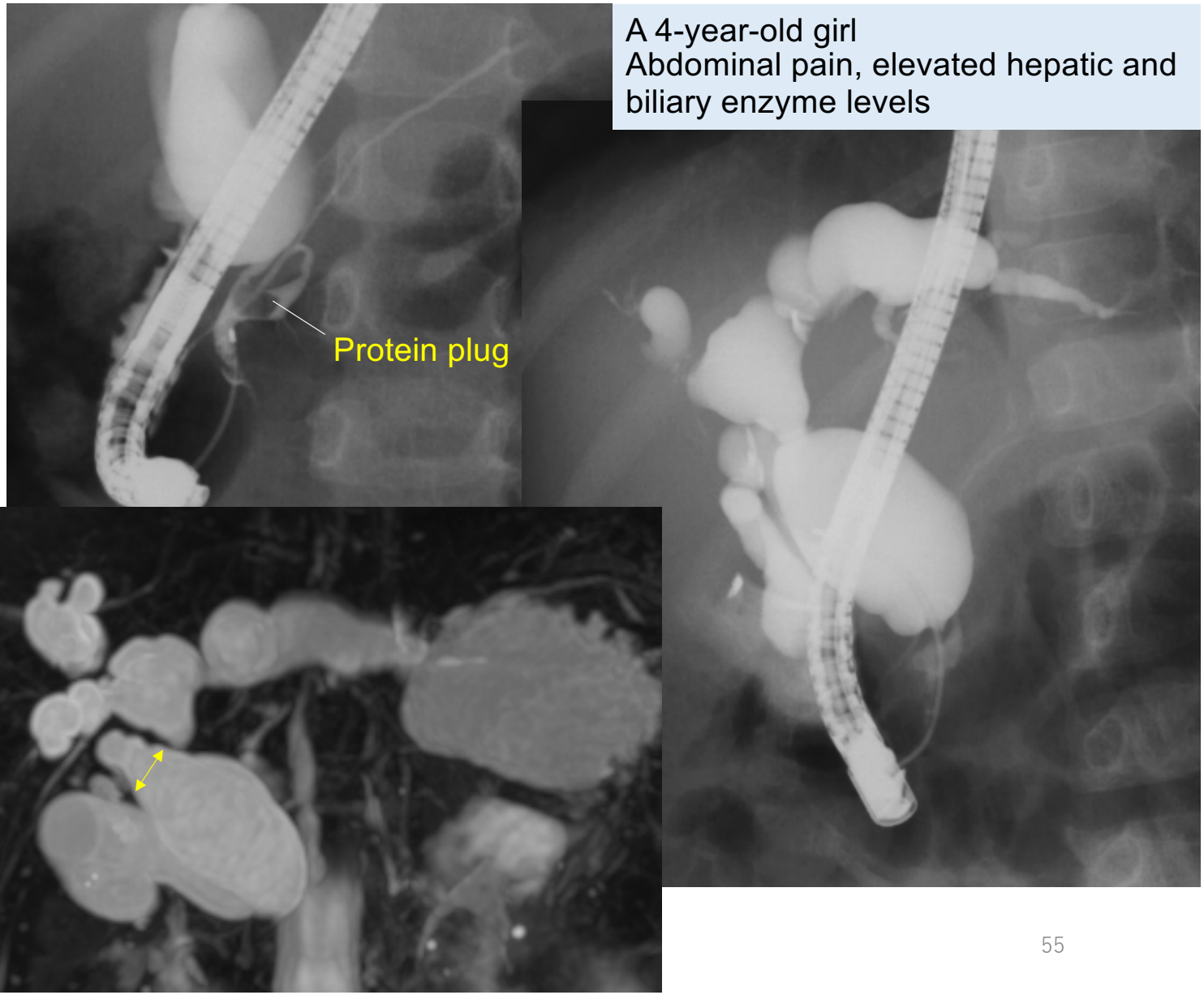
Case 15

Typical VI-A: Cystic–cystic type pediatric case

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

See p85.

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

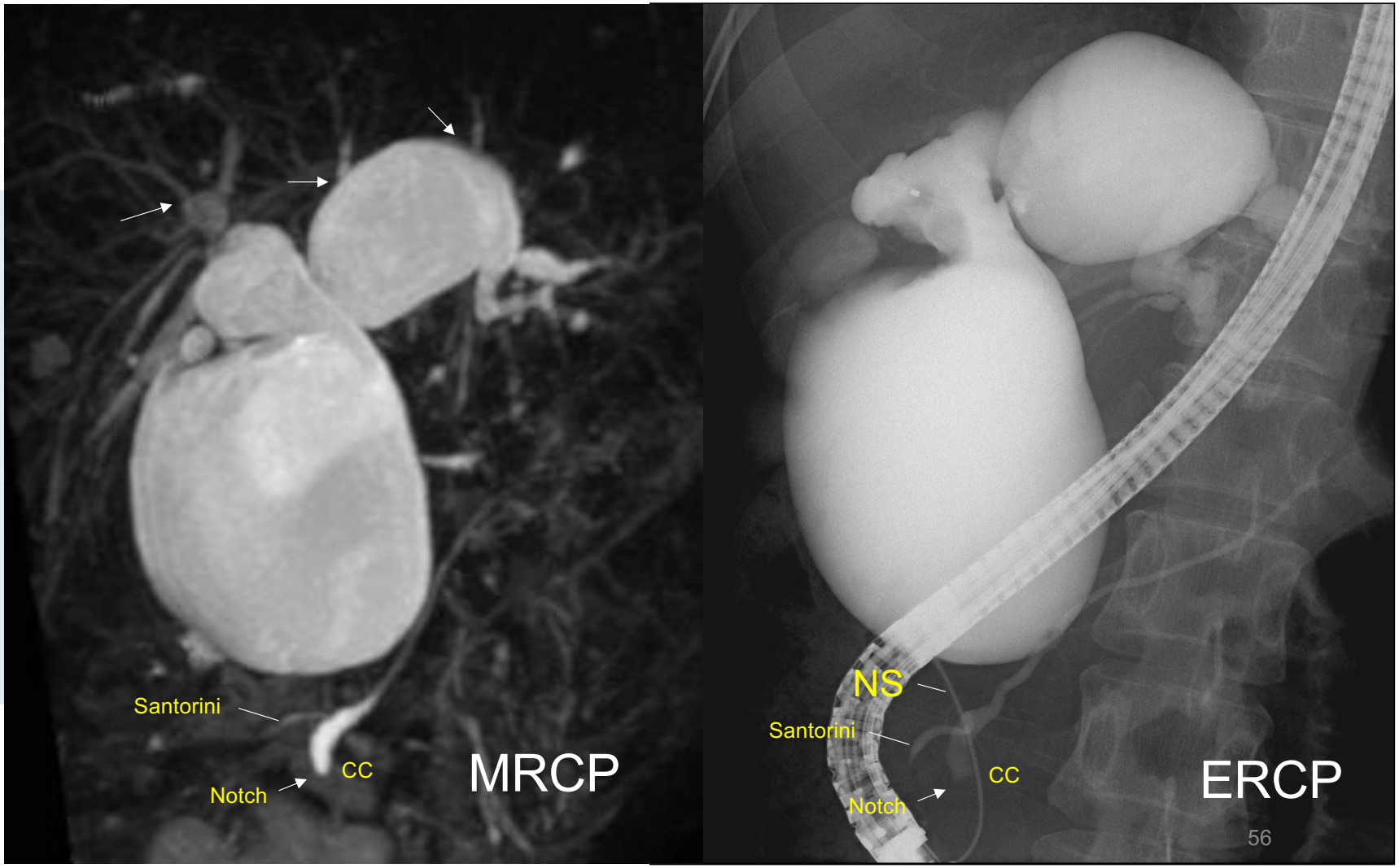


Case 16

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

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

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



CC, common channel

Case 17

Typical IV-A: Fusiform–fusiform type pediatric case

ERCP in a girl

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

