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Basics

Pathophysiology, Diagnosis, and Classification

First version, September 2022

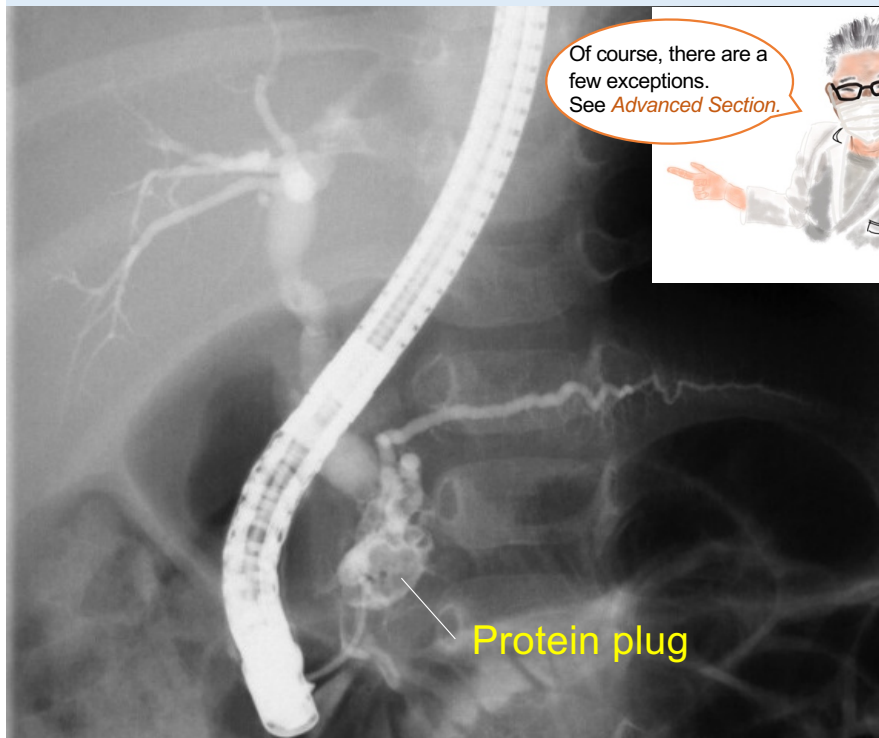
Symptoms in 128 pediatric patients

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

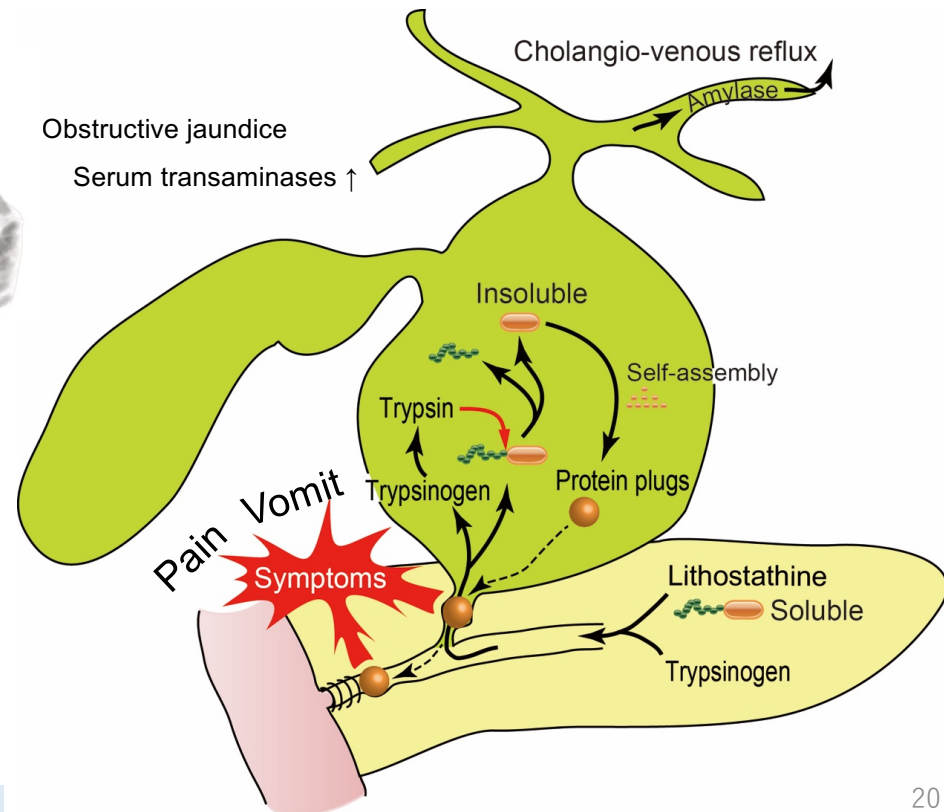
Protein plugs cause symptoms in children.

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

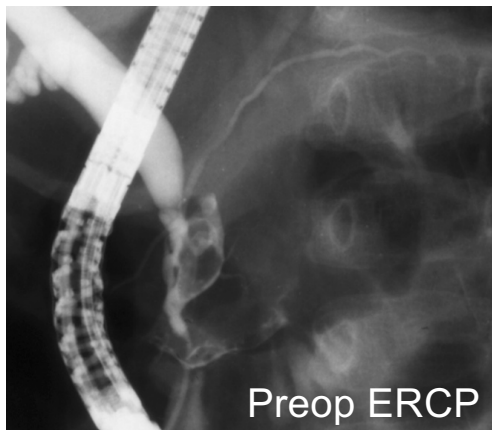


Self-limited

Repeating production



Recurrence



Spontaneous dissolution

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



Case 1

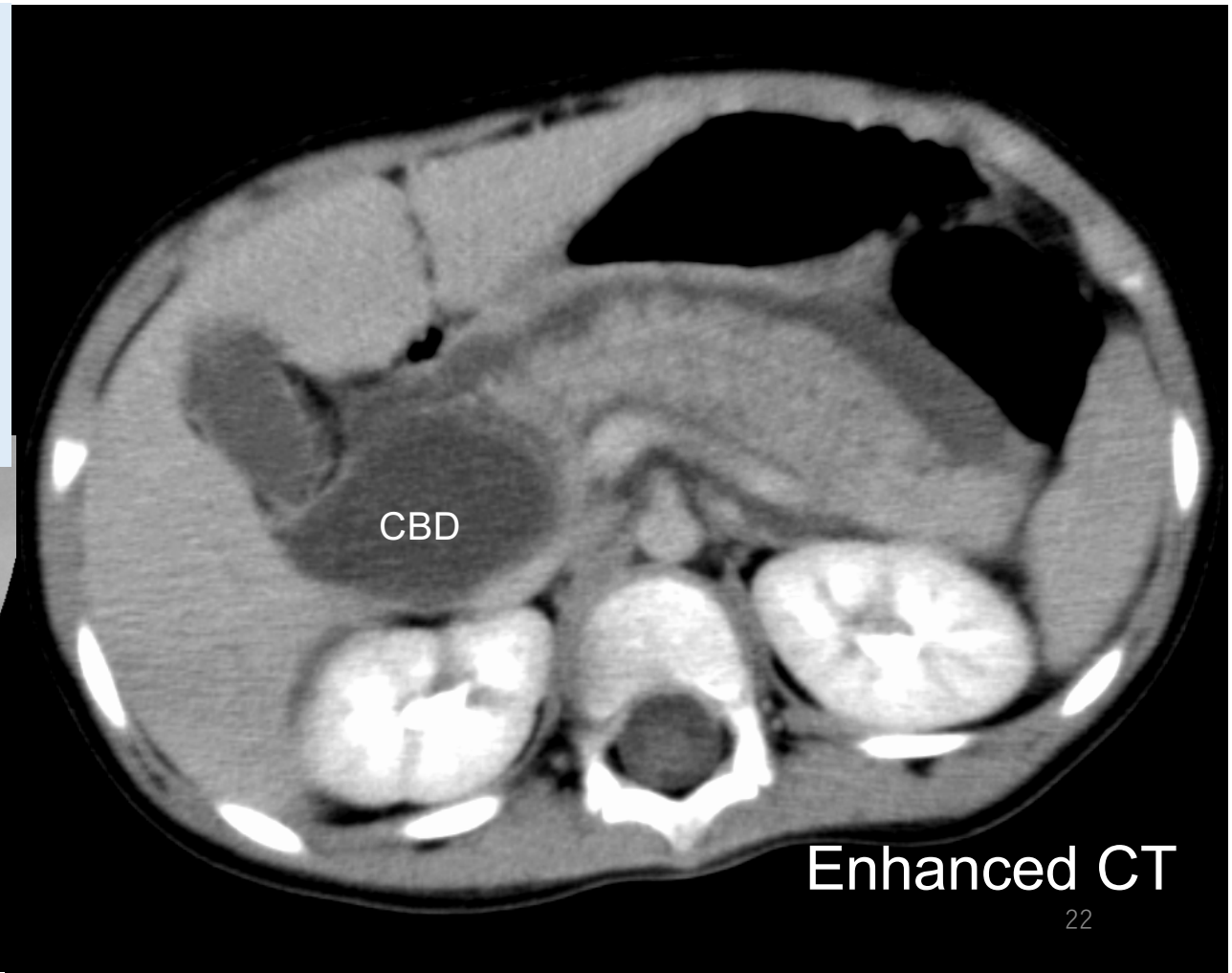
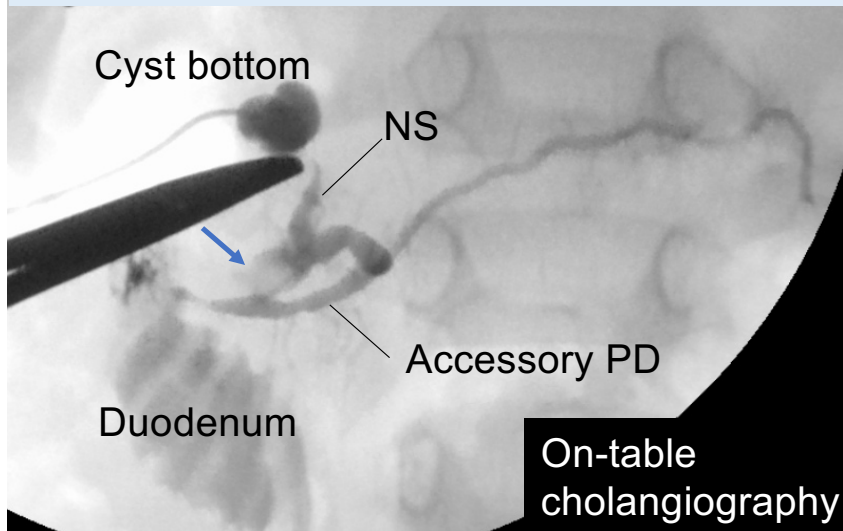
True pancreatitis is observed only in 7% of pediatric patients

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.



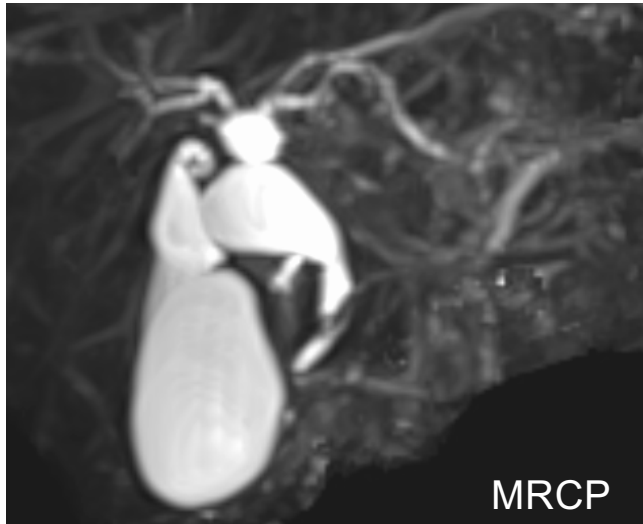
Case 2

Pseudopancreatitis Hyperamylasemia without evidence of pancreatitis

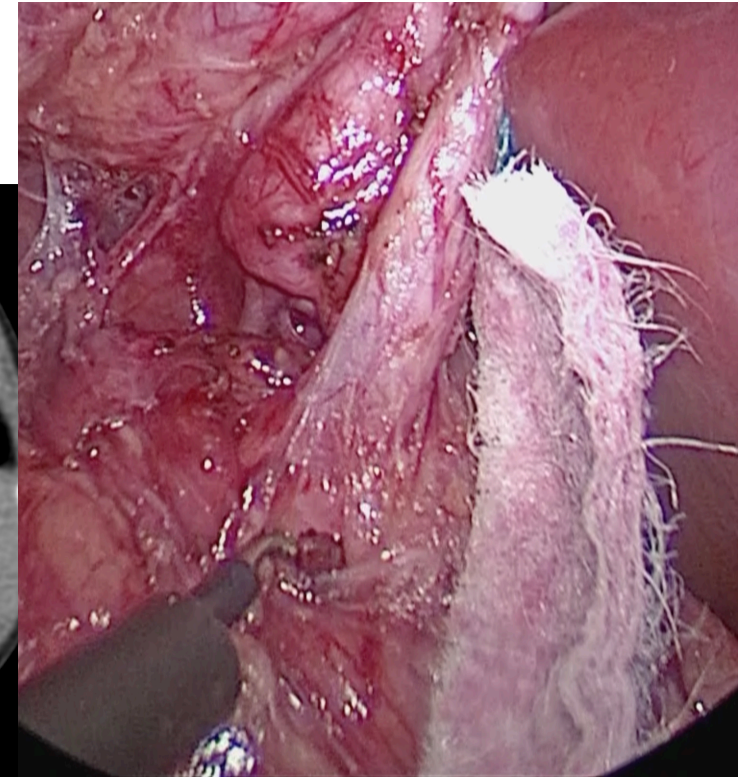
Three-year-old
Repeating **abdominal pain**
and vomiting

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

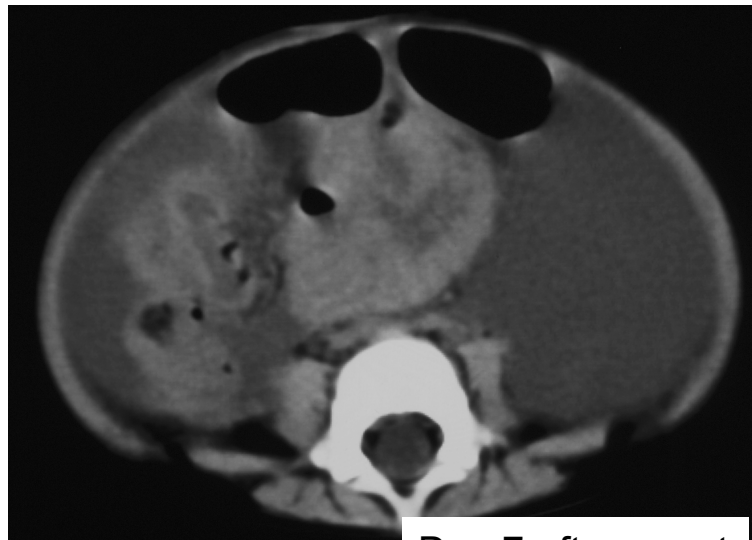
Case 3 Biliary perforation is rare: 3%-6% of children

One-year-old girl

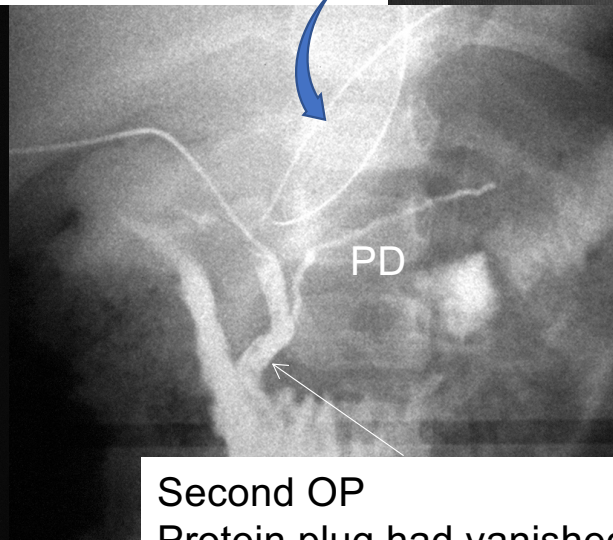
Sudden onset of vomiting and abdominal pain on day 4, diarrhea and abdominal distention on day 5, fever on day 6, and pleural effusion on chest Xp on day 7.

Bile peritonitis causes relatively mild irritation and is often overlooked.

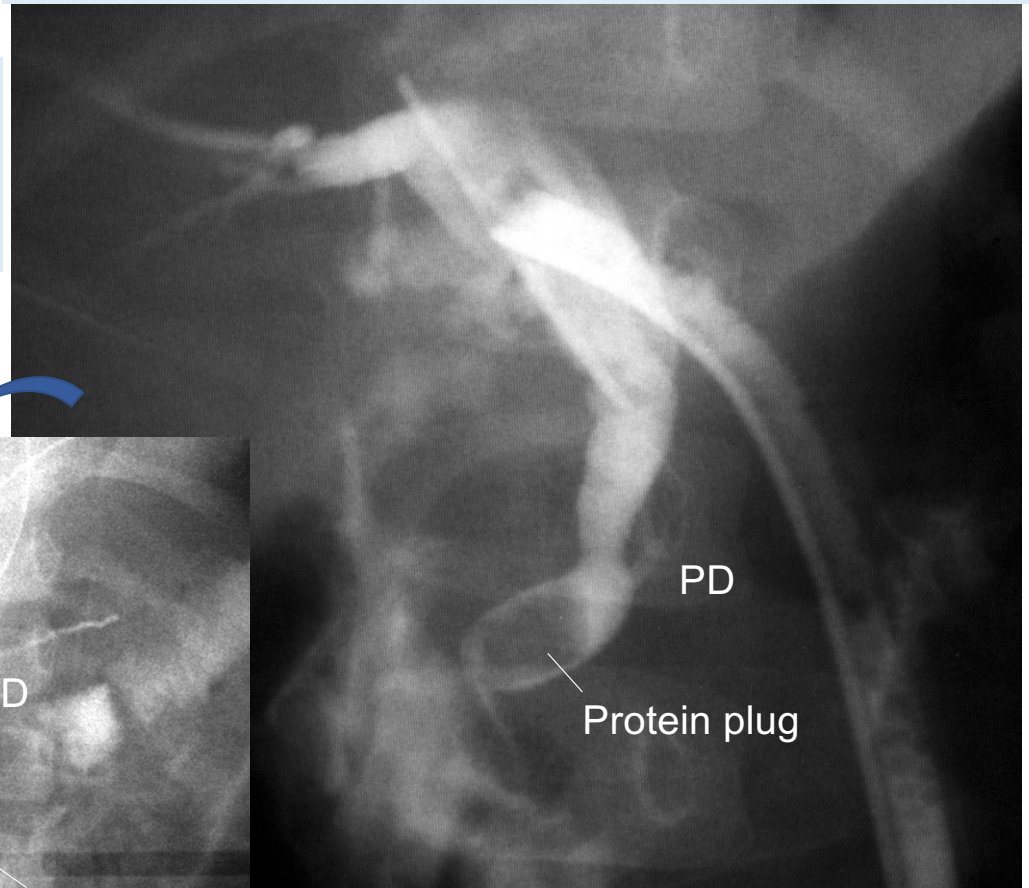
Most protein plugs are fragile and dissolve spontaneously (transient symptoms). However, exceptionally hard protein plugs produce abrupt increases in intraductal pressure, causing a rupture.



Day 7 after onset



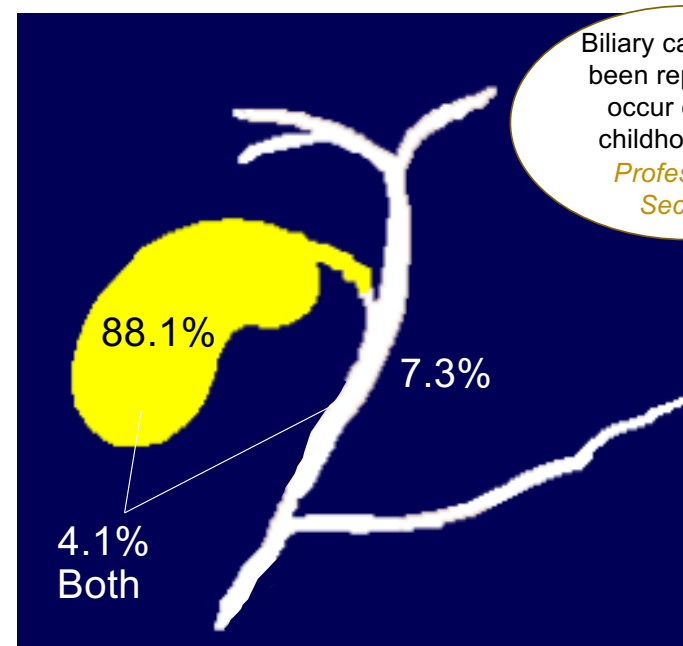
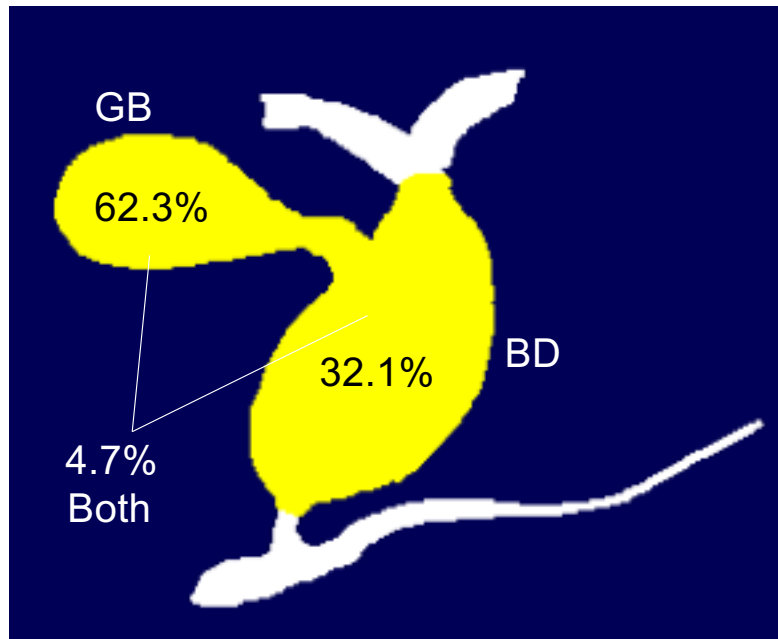
Second OP
Protein plug had vanished



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*.

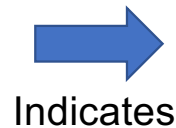


Cancer rate

CBD 21.6%

Non-dilated PBM 42.4%

Carcinogenesis in region where bile stagnates

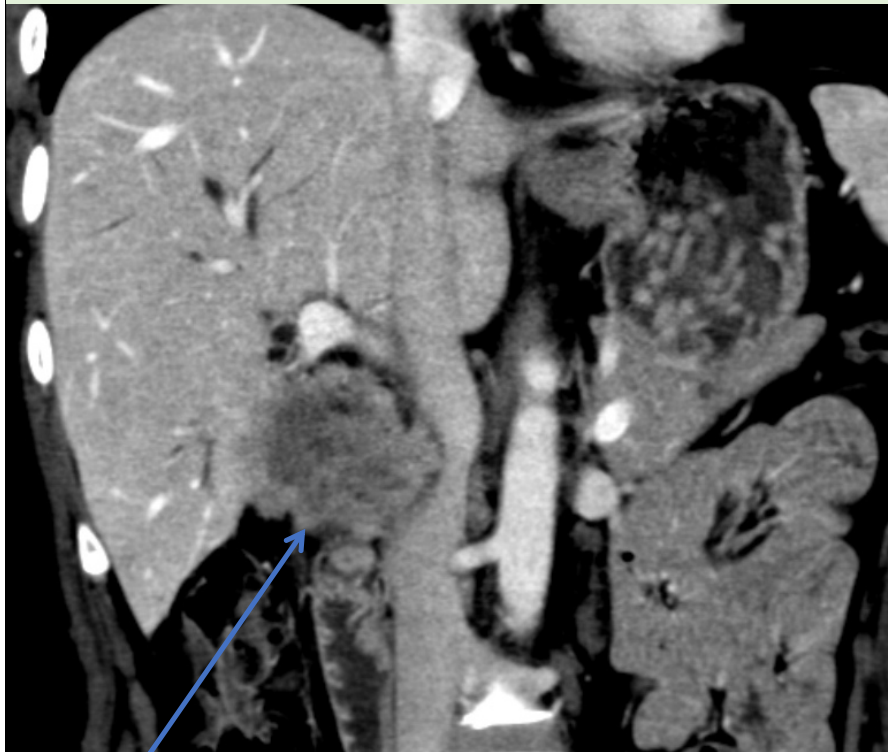


Indicates

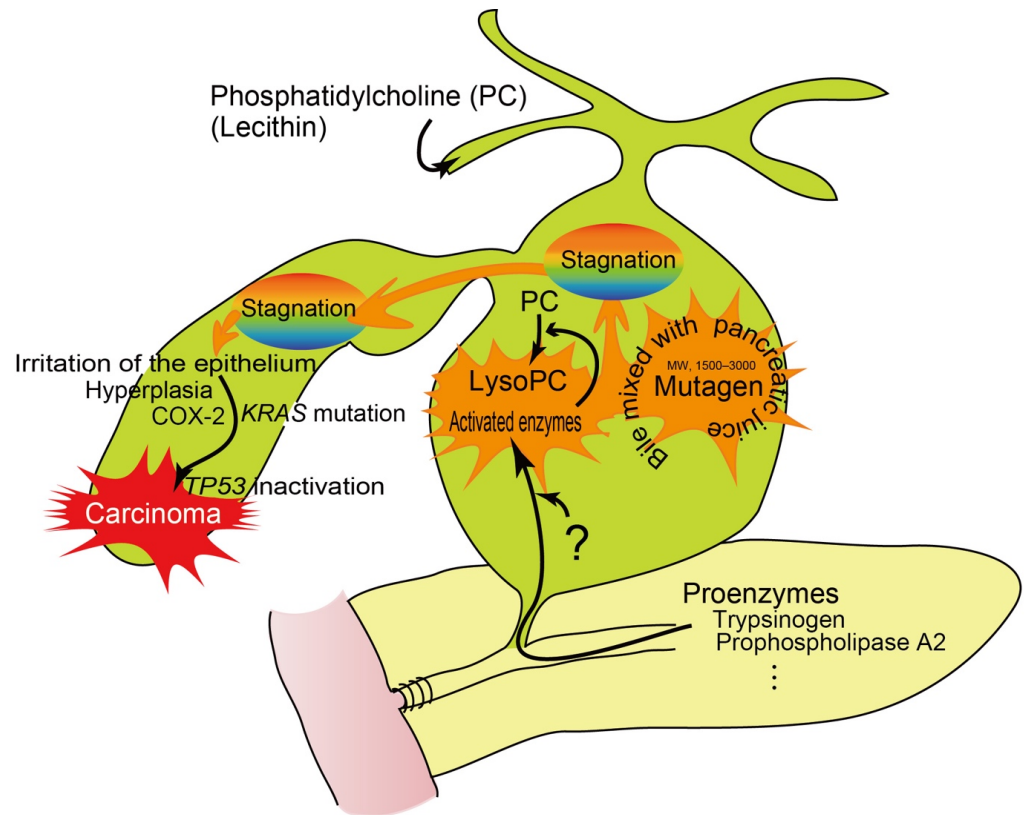
Hazardous substances, produced by a mixture of bile and pancreatic juice, stagnate and cause chronic inflammation, which induces carcinogenesis

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

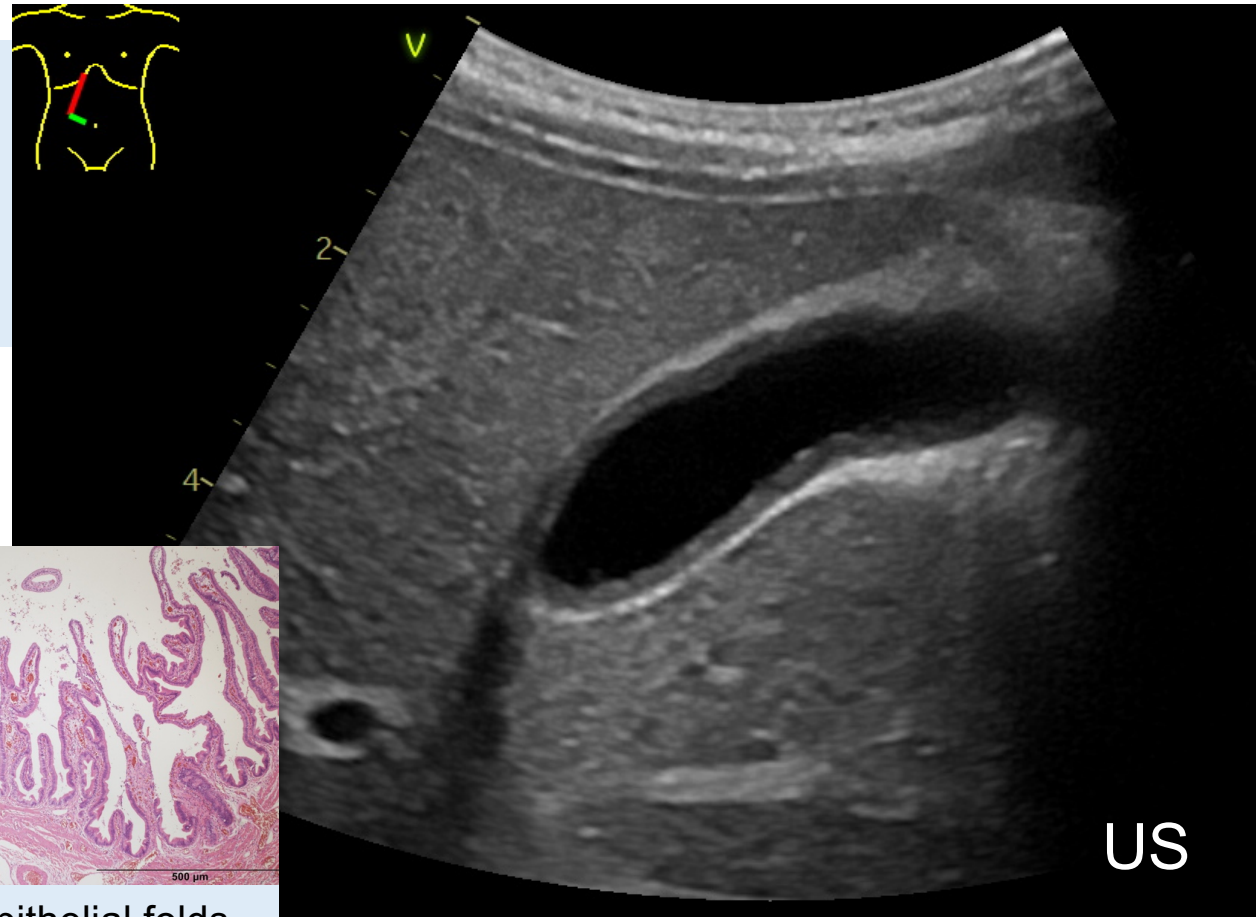
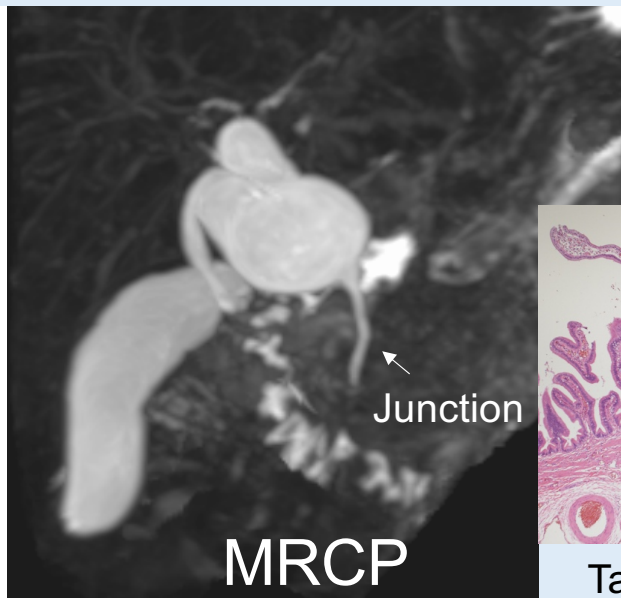
Case 4

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.

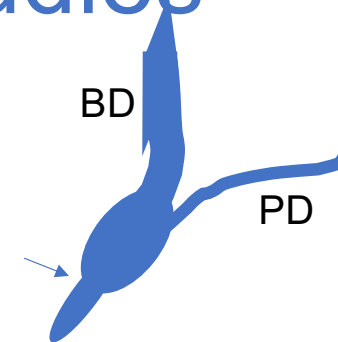
Thick wall of GB on ultrasonography (US)

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.



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).

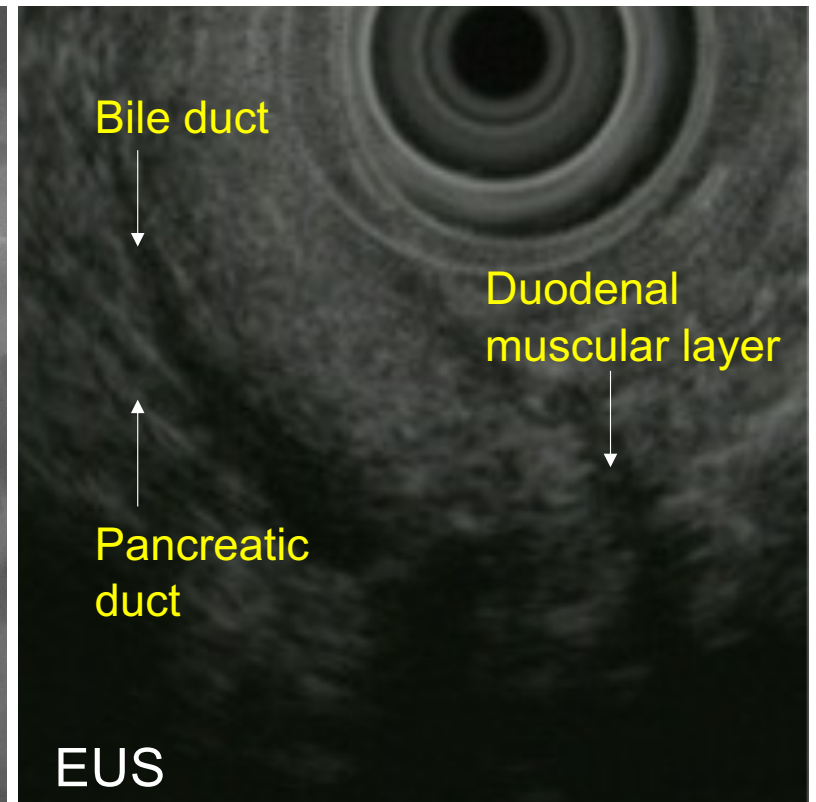
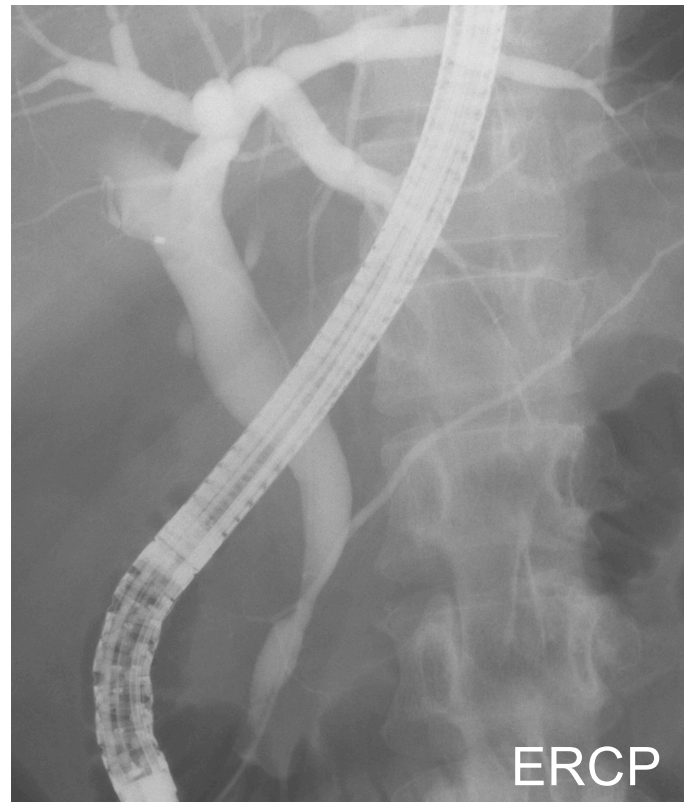
Diagnostic criteria for pancreaticobiliary maljunction 2013
J Hepatobiliary Pancreat Sci 2014;21:159–161

Case 5

EUS directly demonstrates the junction outside the duodenal wall

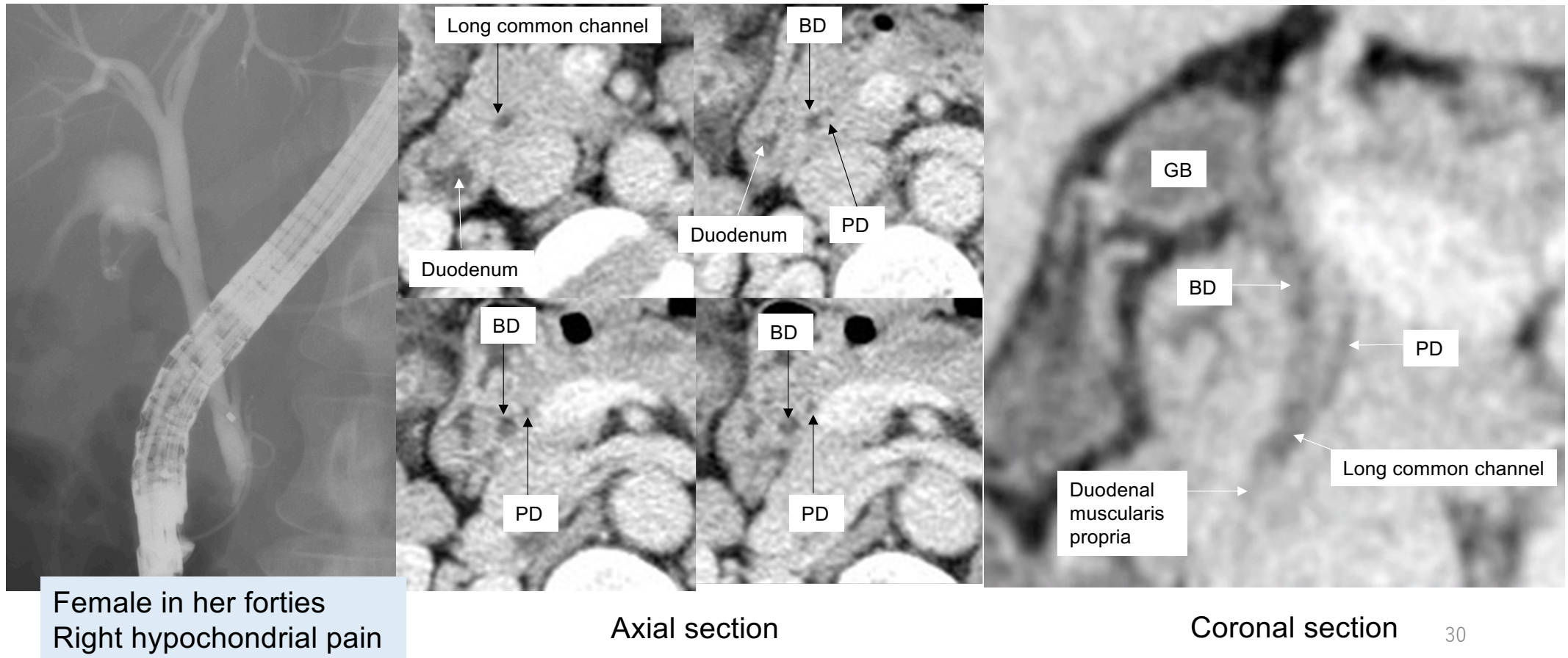
- Female in her forties
Abdominal pain
- EUS shows **union outside the duodenal wall = PBM**

EUS, endoscopic ultrasonography

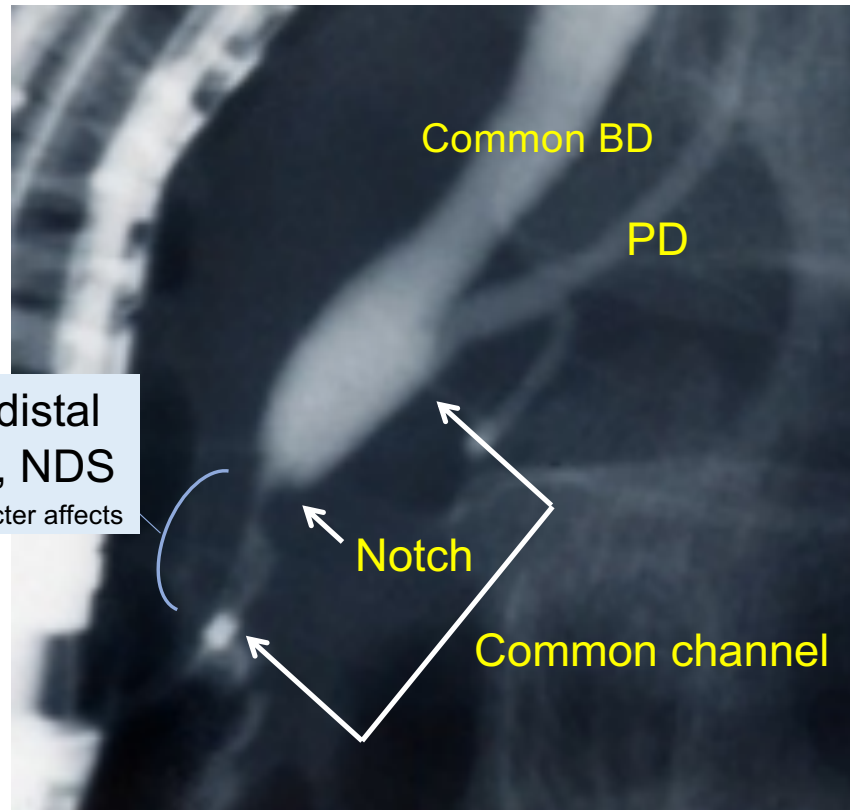


Case 6

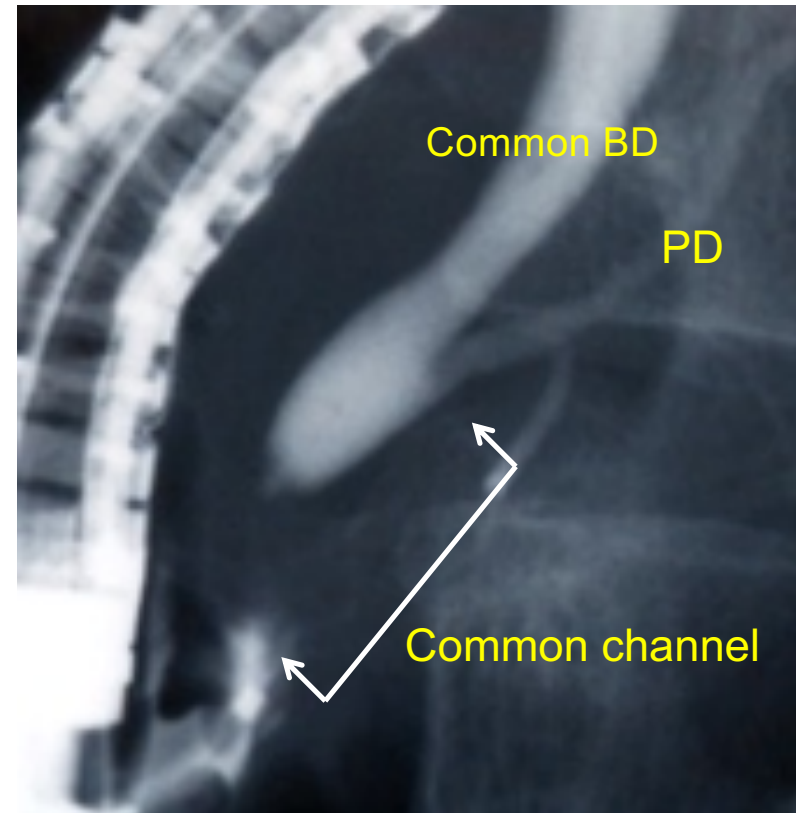
CT directly demonstrates
the junction outside the duodenal wall



Notch on the common channel = PBM



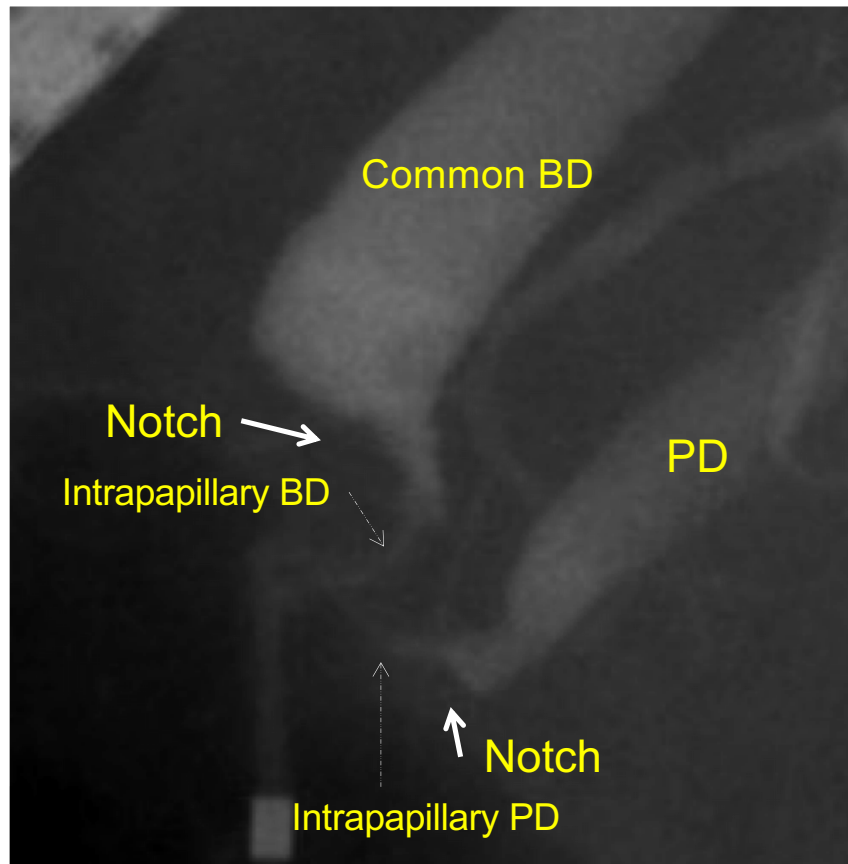
Relaxation phase
of Oddi's sphincter



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

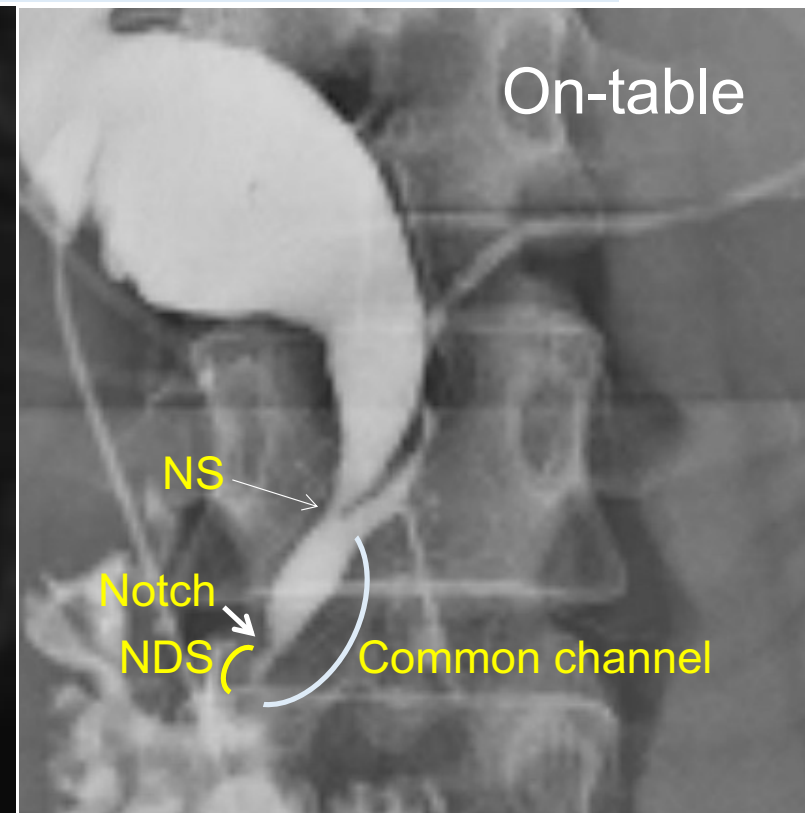
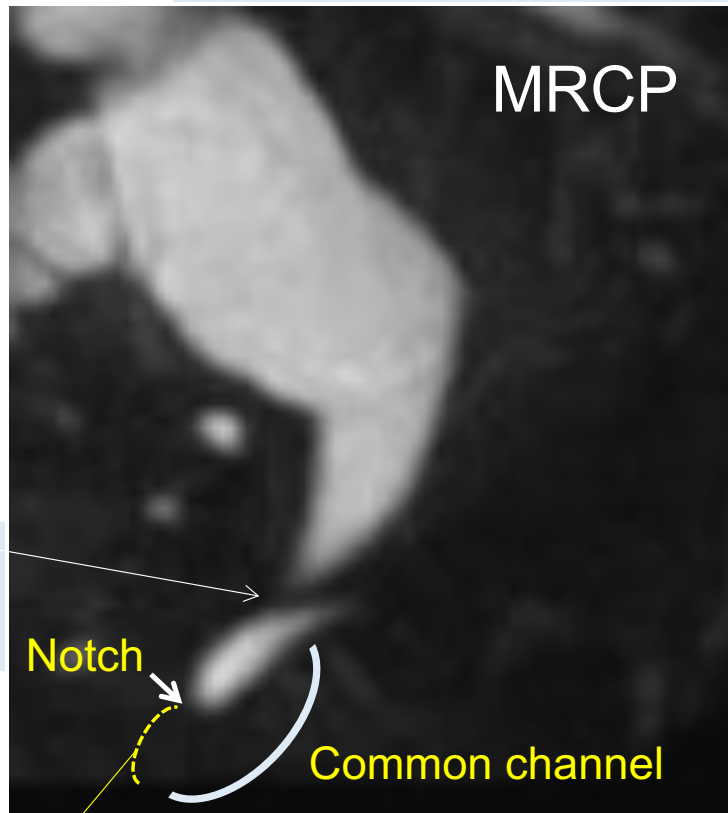


Contraction phase
of Oddi's sphincter

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

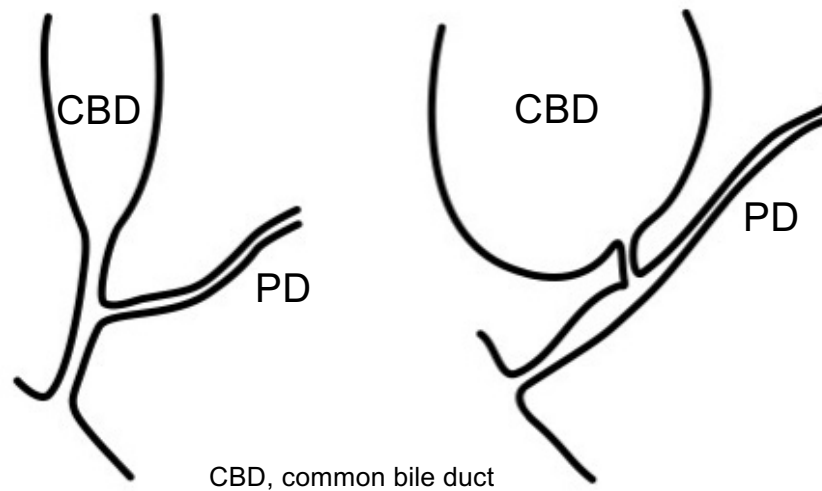


NDS not depicted → MRCP may not elucidate precise length of the common channel

Old Classification of PBM

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

Kimura's classification 1976

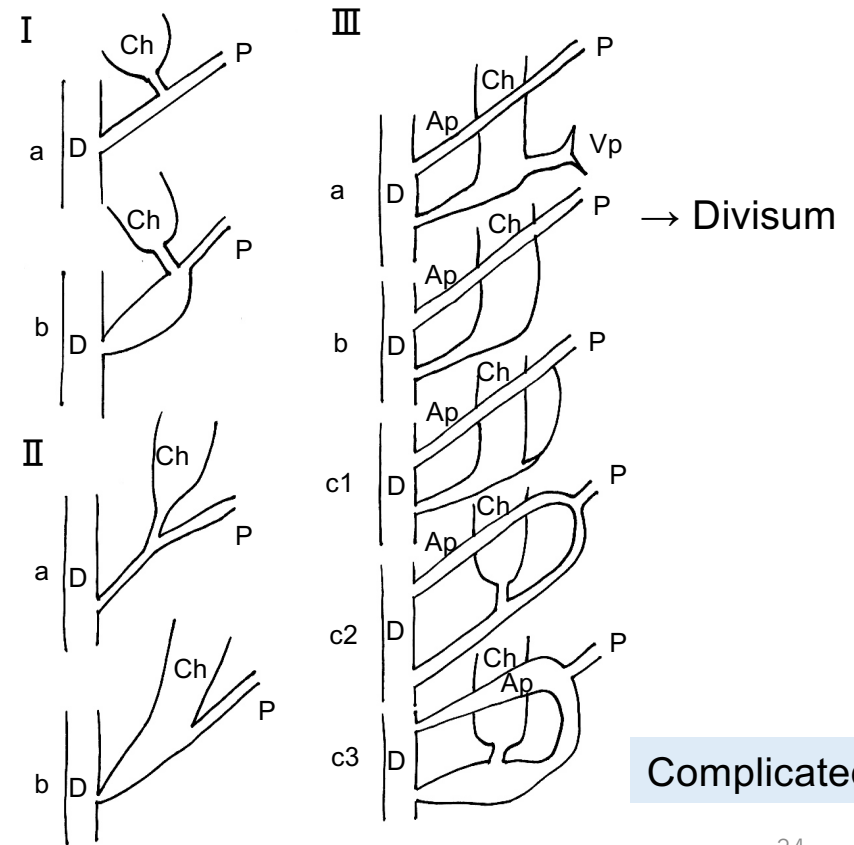


PC type
PD enters CBD

CP type
CBD enters PD

Remains basic even today

Komi's revised classification 1991



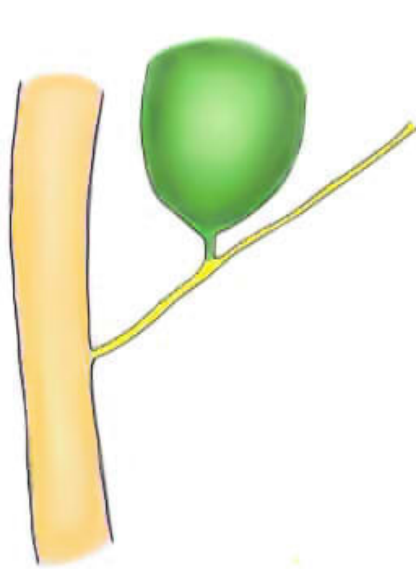
Ch, choledochus; Ap, accessory PD

Complicated

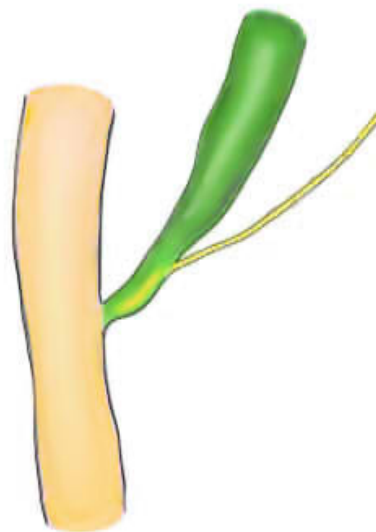
Classification by JSGBPBM

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

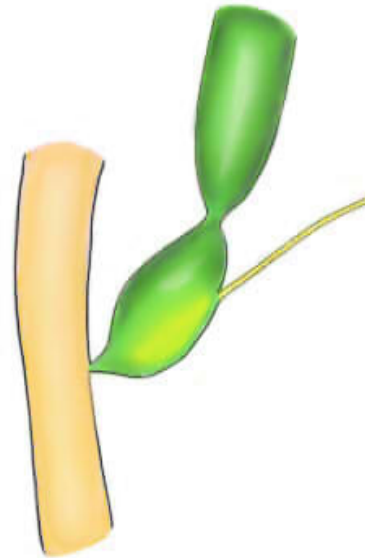
Based on the existence of narrow segment and dilatation of common channel



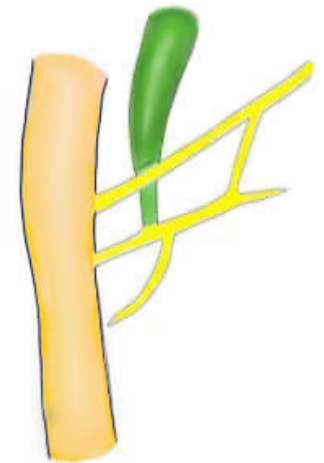
Type A
Stenotic type



Type B
Non-stenotic type



Type C
Dilated channel type

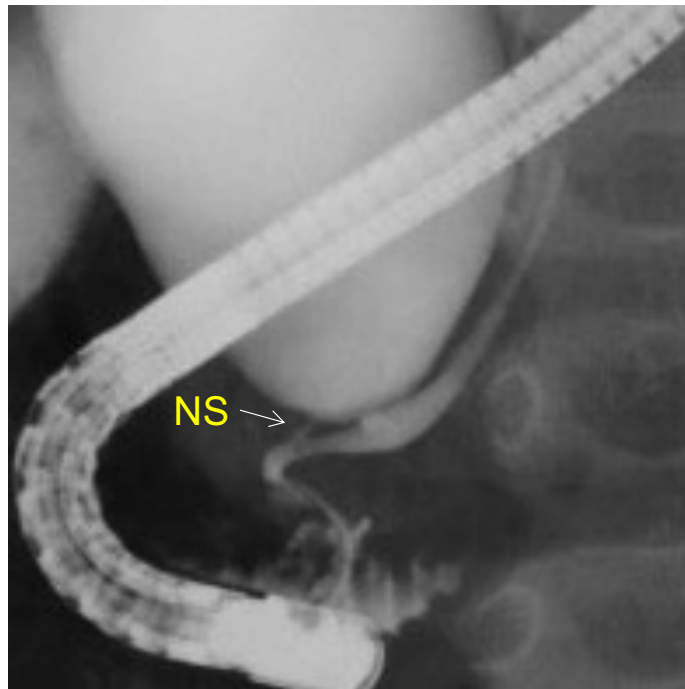


Type D
Complex type

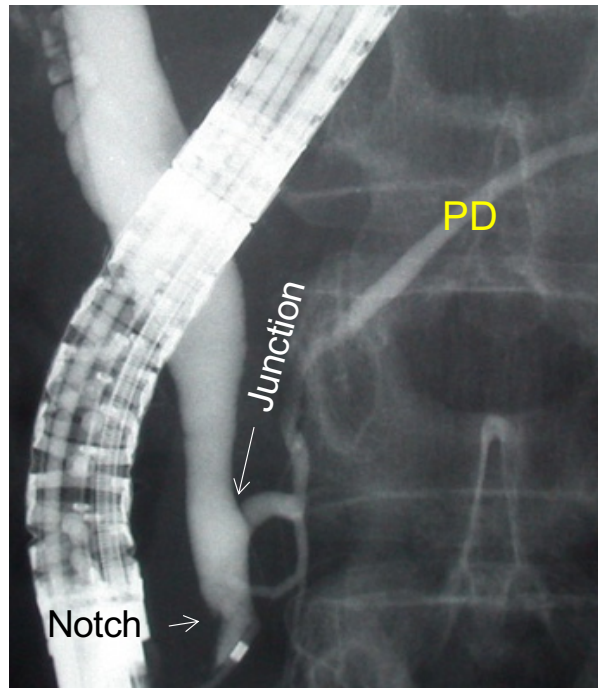
This classification is indifferent to existence of accessory PD (Santorini's duct)

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

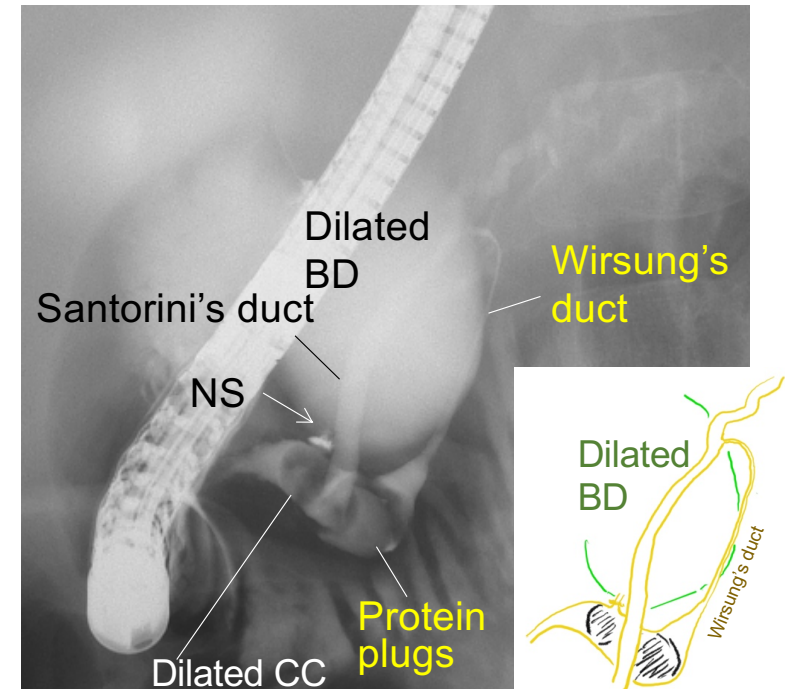
Classification by JSGBPBM



Type A
Stenotic type



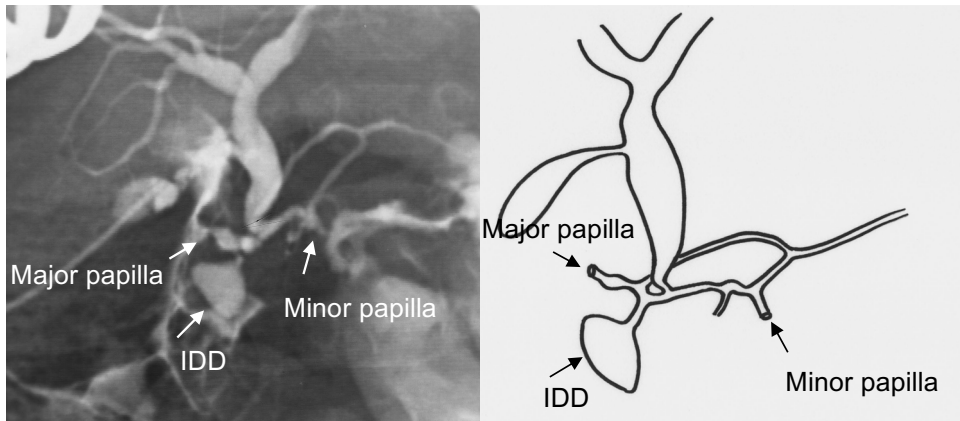
Type B
Non-stenotic type



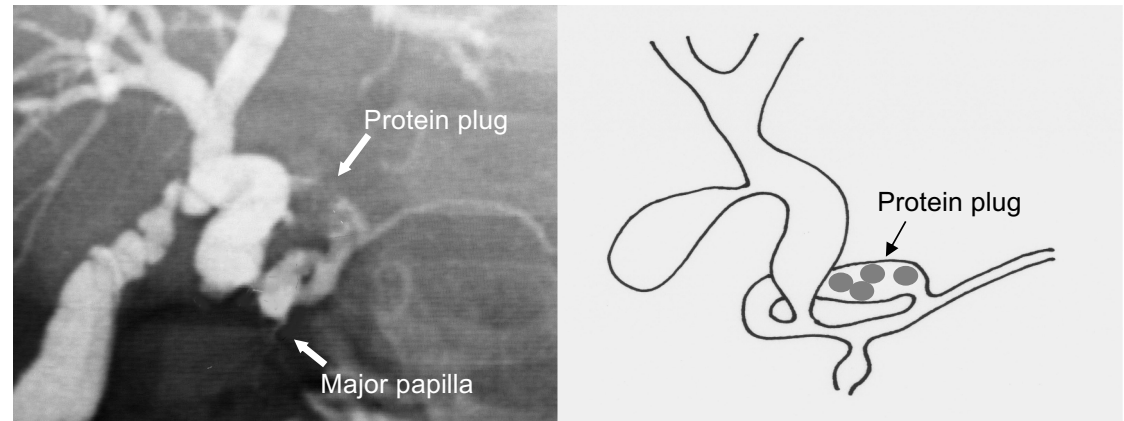
Type C
Dilated channel type

Type D: Complex type

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



IDD, intraluminal duodenal diverticulum

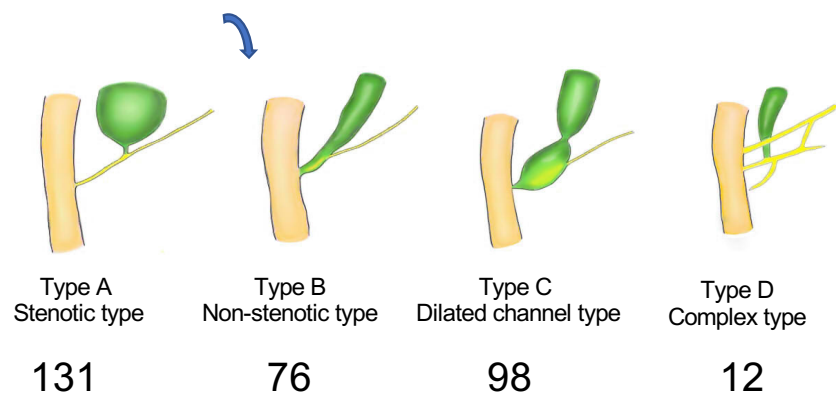


One-year-old girl:
Annular pancreas and
intraluminal duodenal diverticulum

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

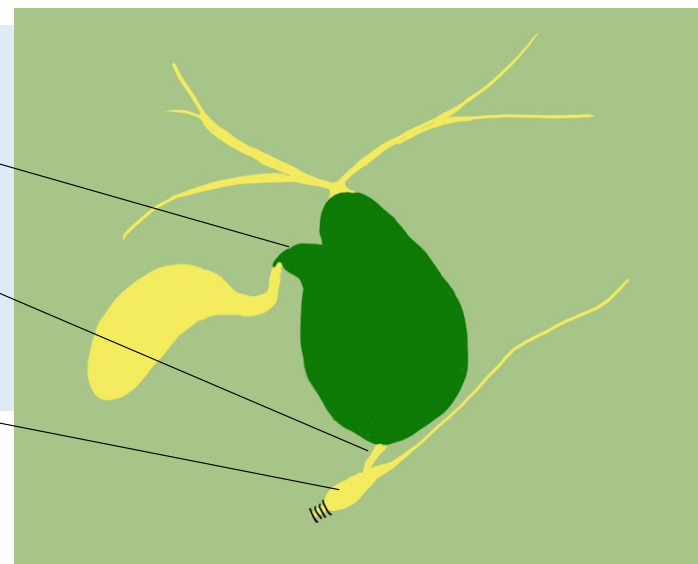
Features that define CBD

- 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



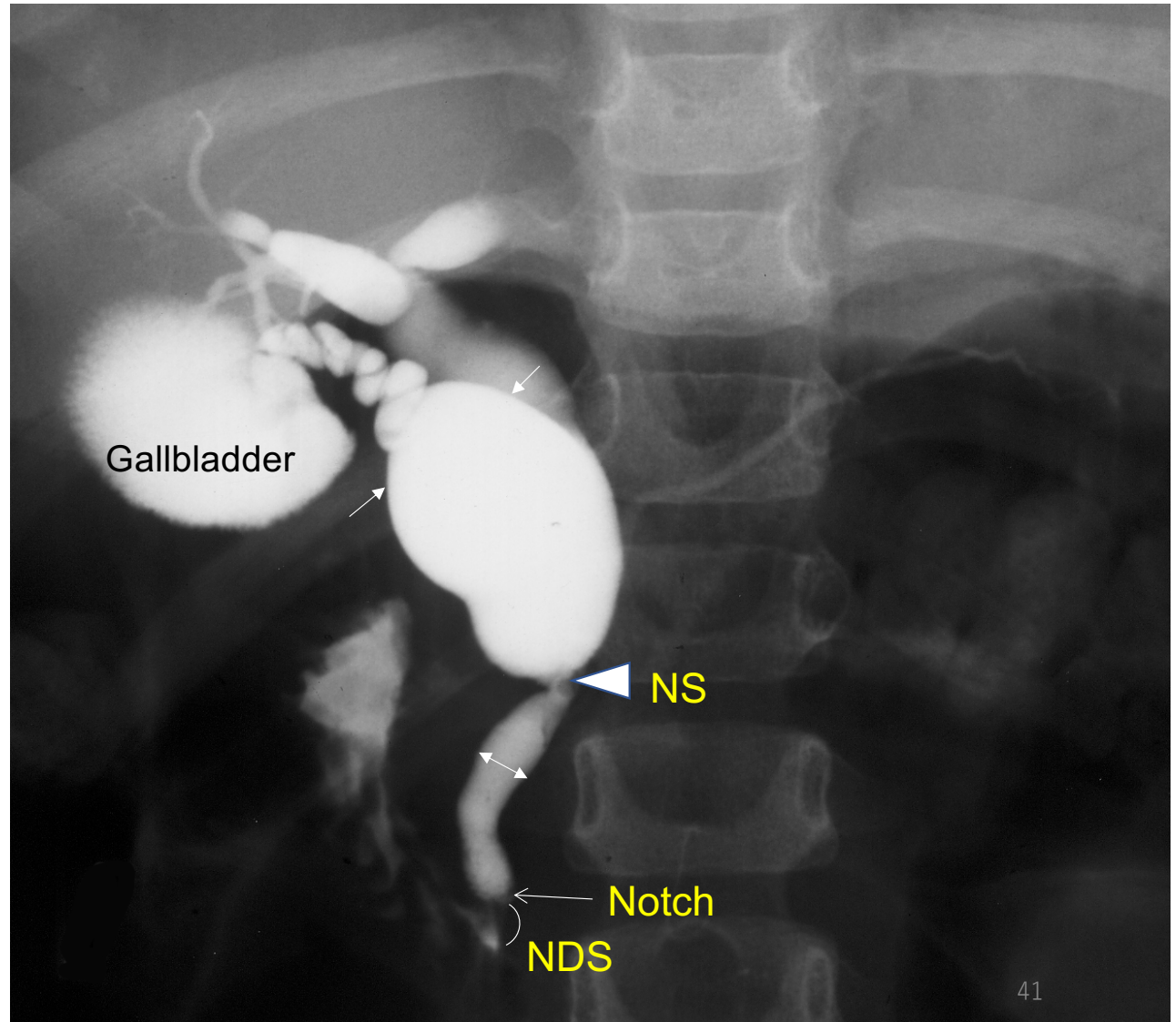
Case 7

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

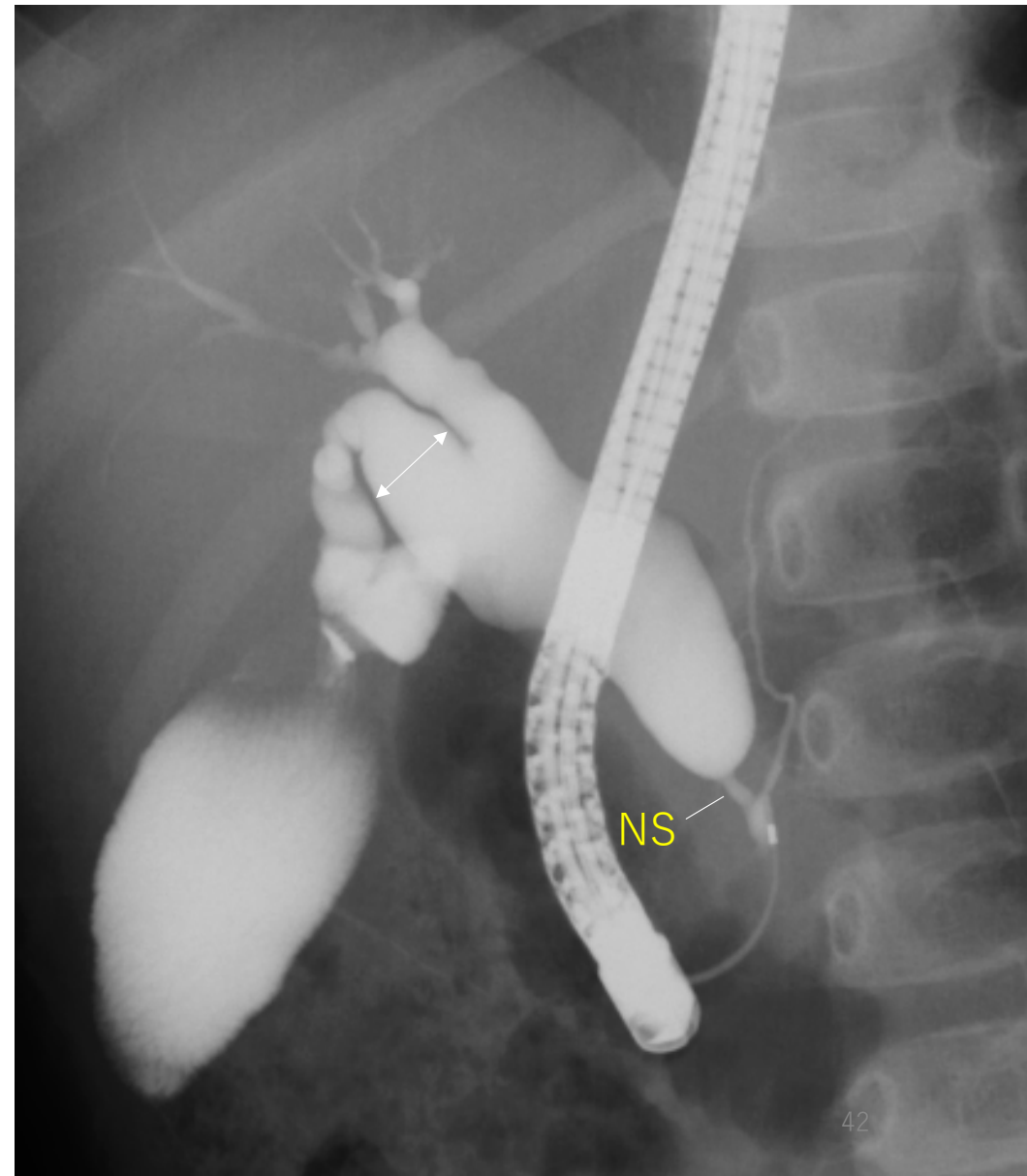


Case 8

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

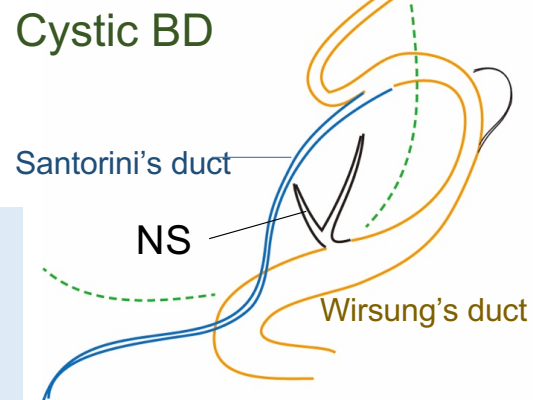
Histopathological demonstration

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

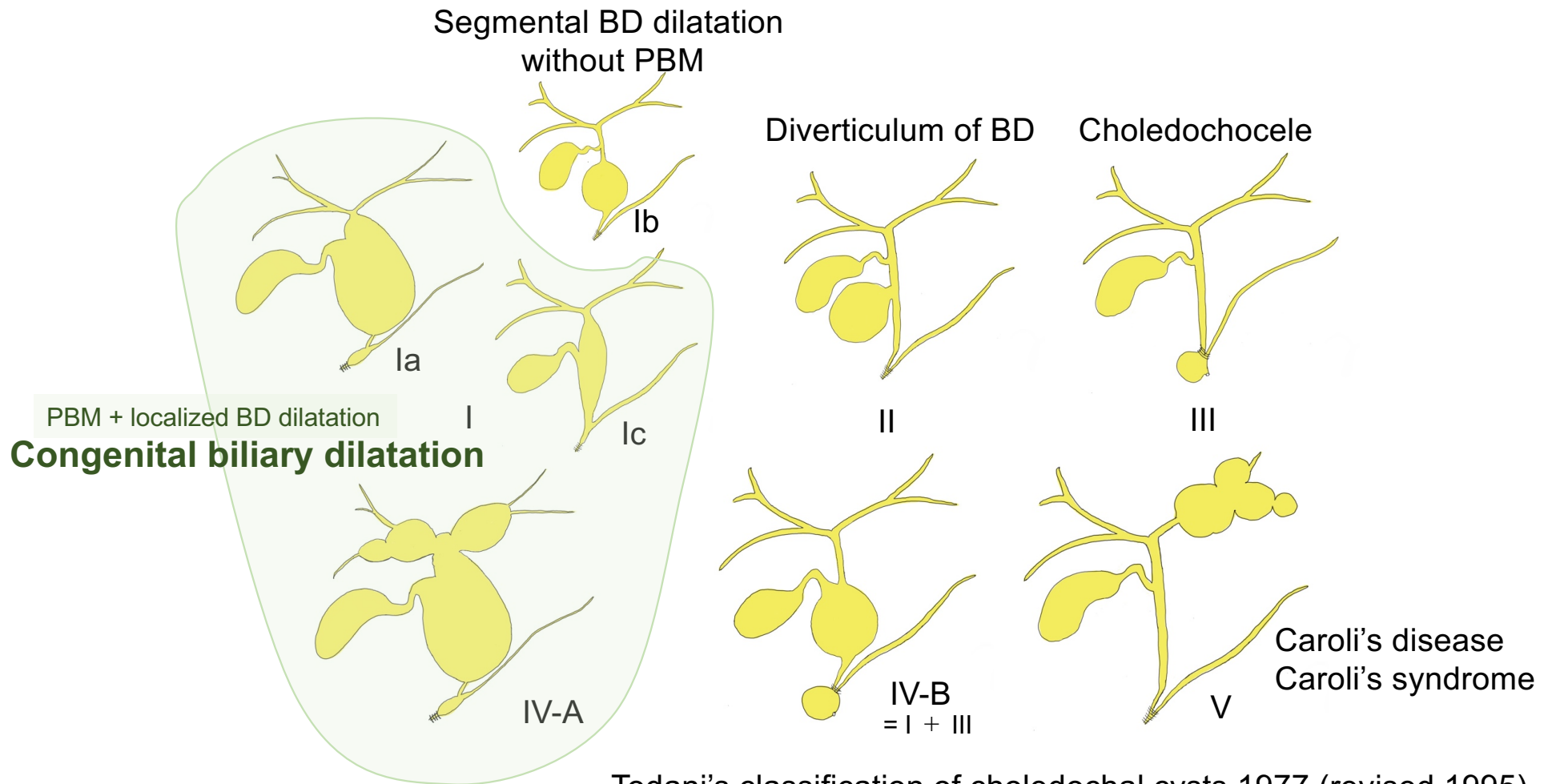


NS is one branch
of PD



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 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