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Basics

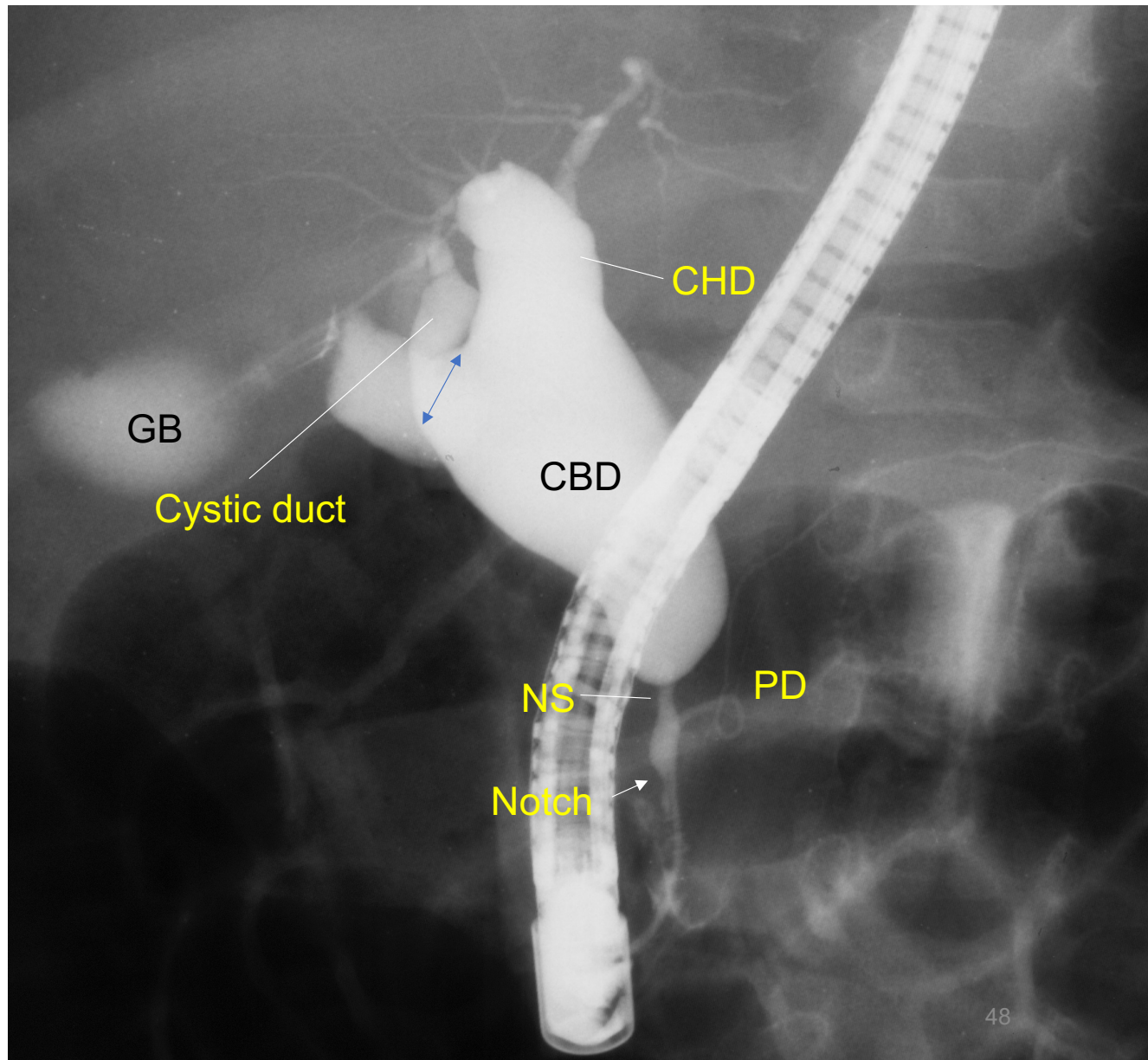
Typical Congenital Biliary Dilatation

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

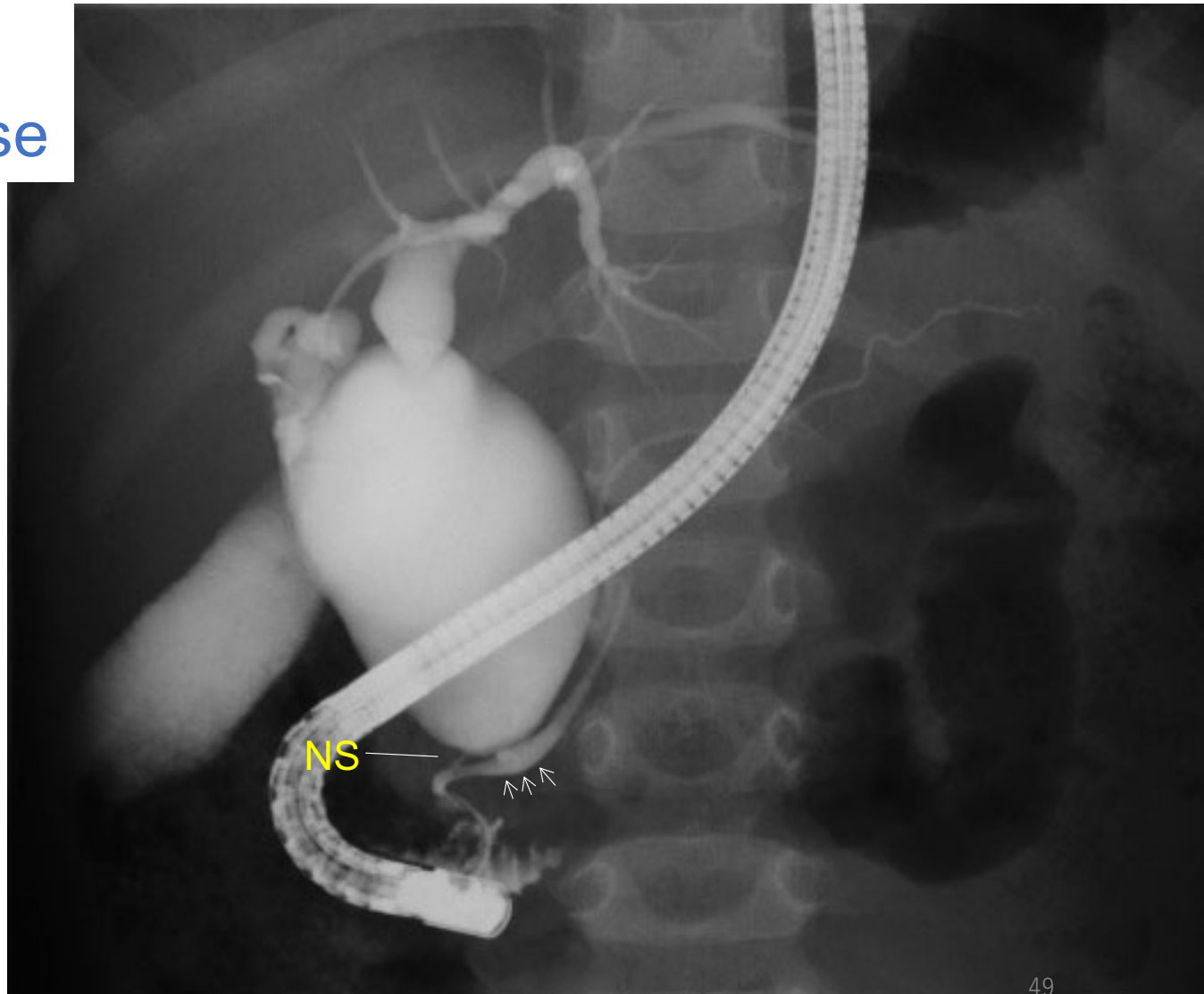


Case 11

Typical Ia: Pediatric case

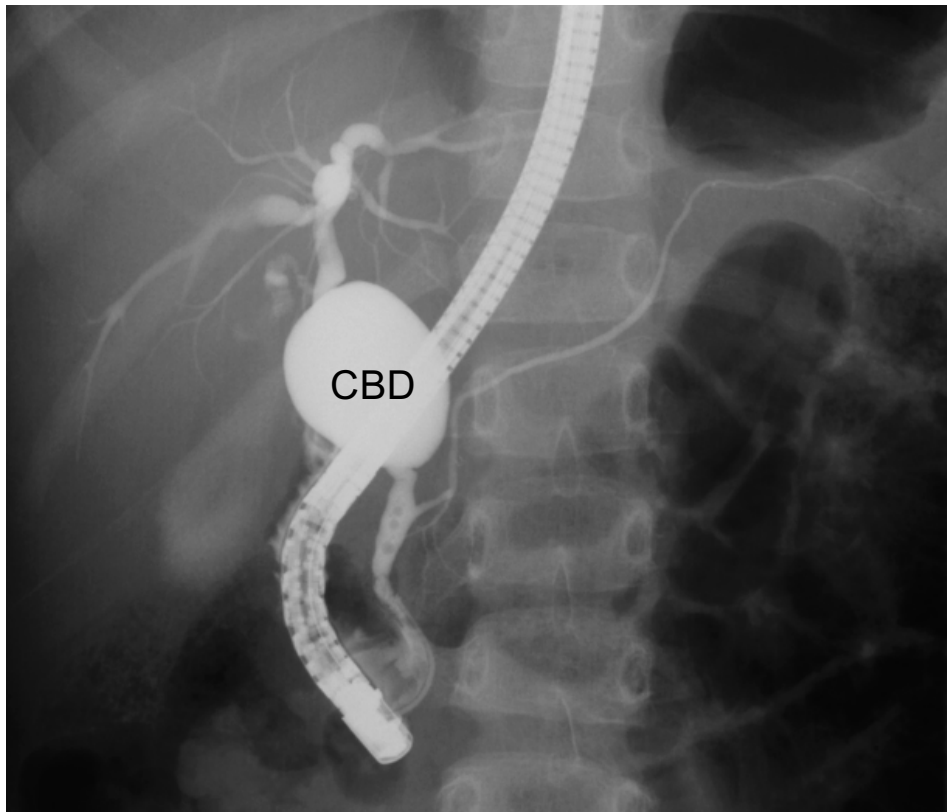
ERCP in a 3-year-old girl
Abdominal pain, vomiting, and elevated serum transaminase levels

- **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

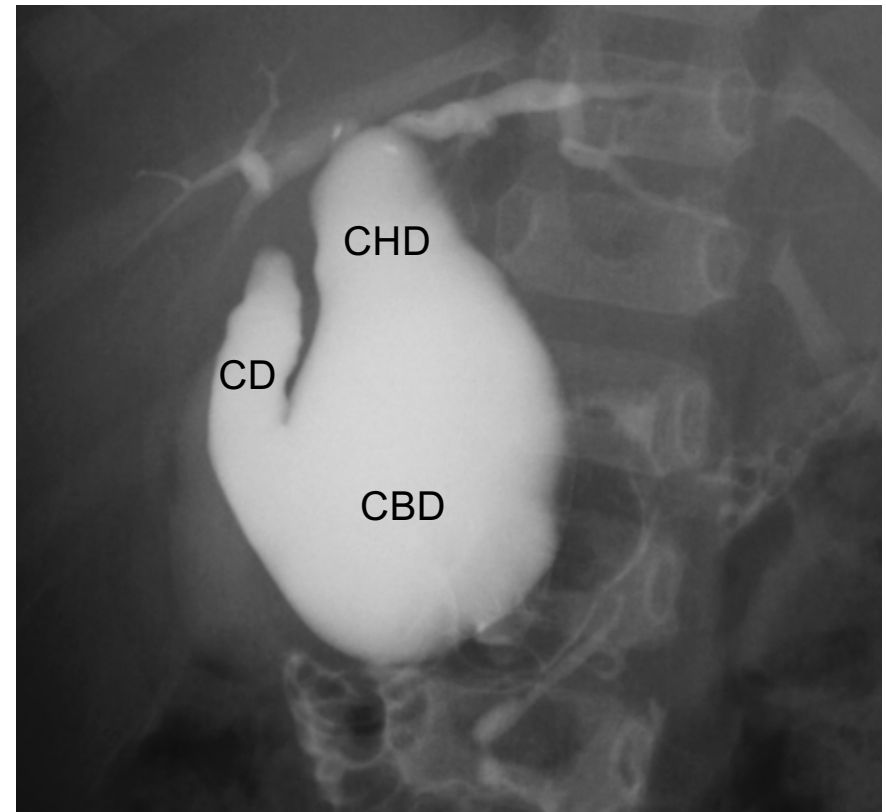


Type Ia: Pediatric case

Localized dilatation to various extents



Dilatation limited to the common bile duct (CBD)



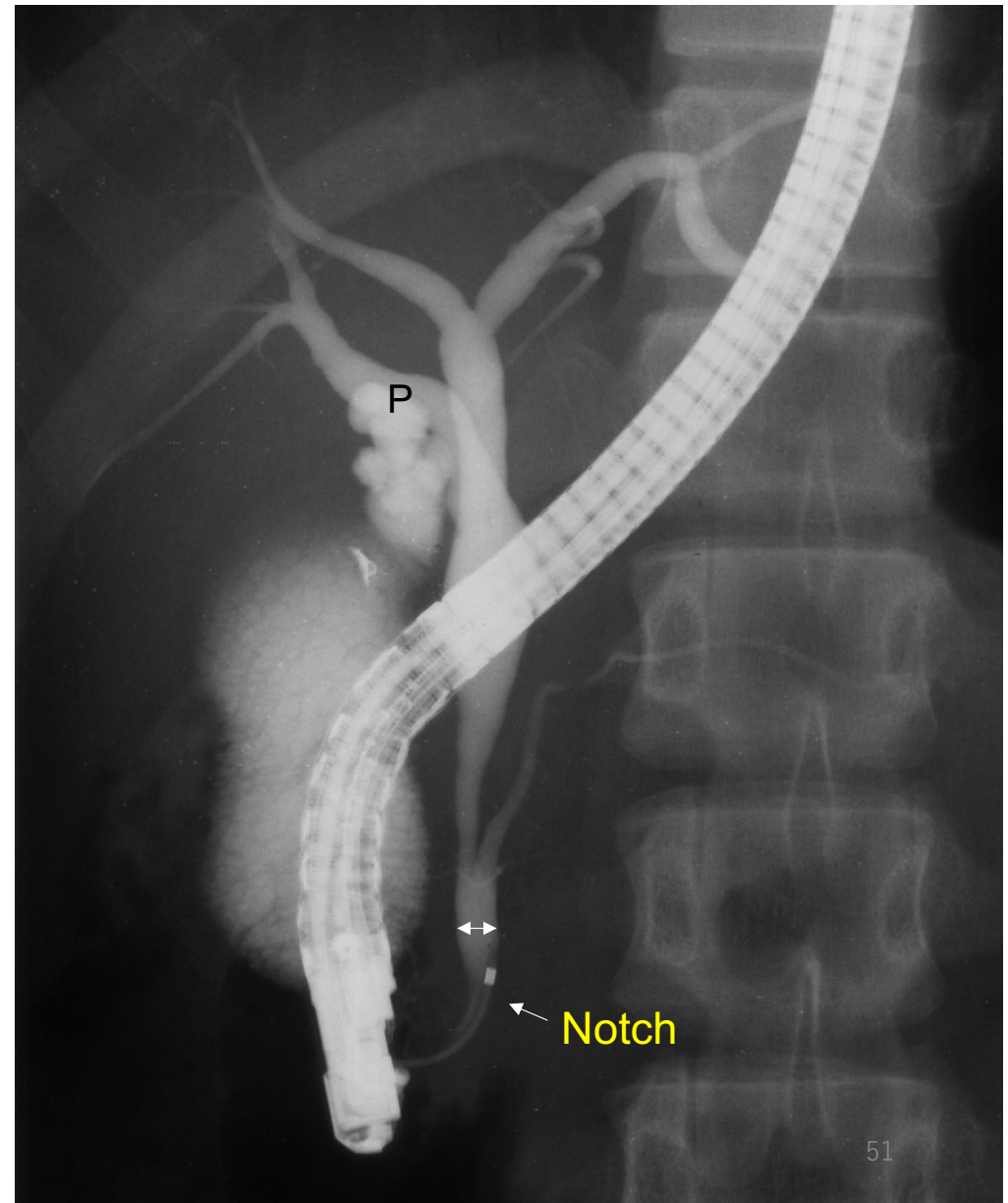
Dilatation involving CBD, CD, and CHD

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

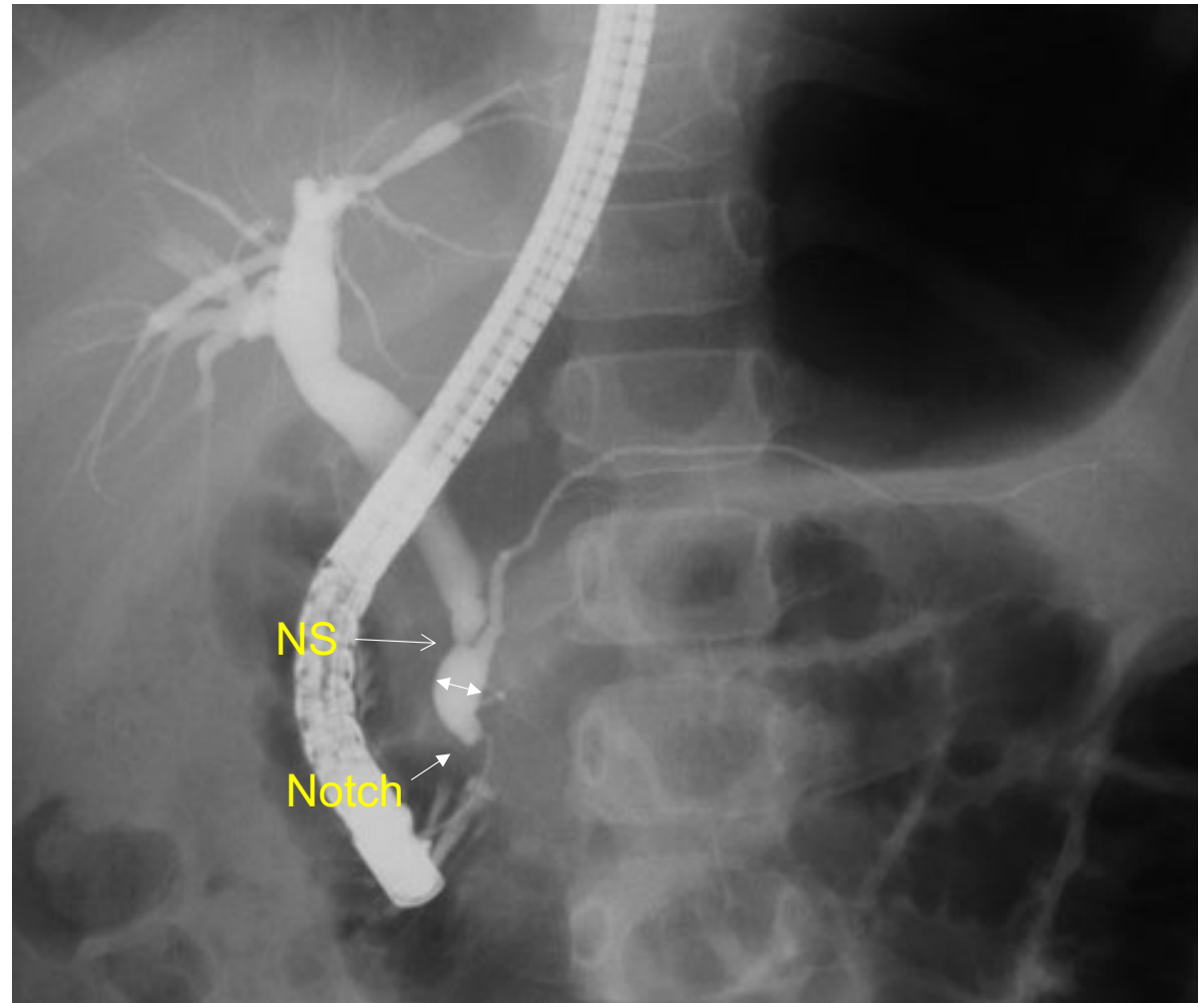


Case 13

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

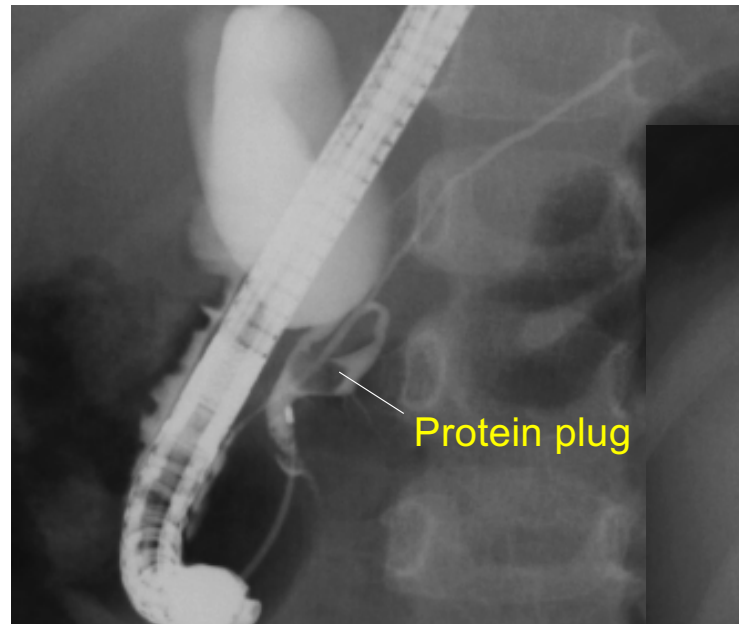


Case 15

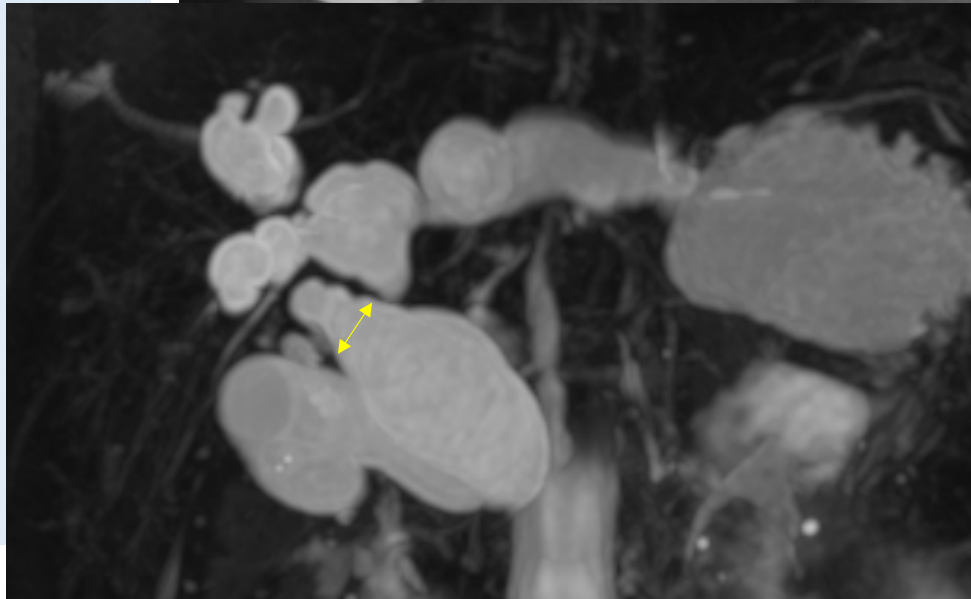
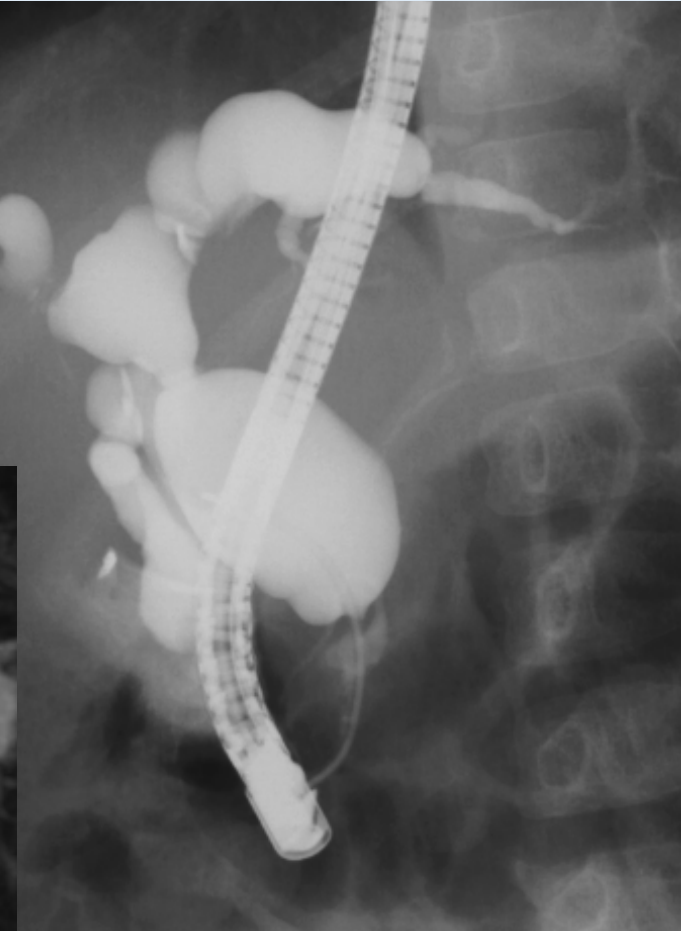
Typical VI-A: Cystic–cystic type pediatric case

- 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



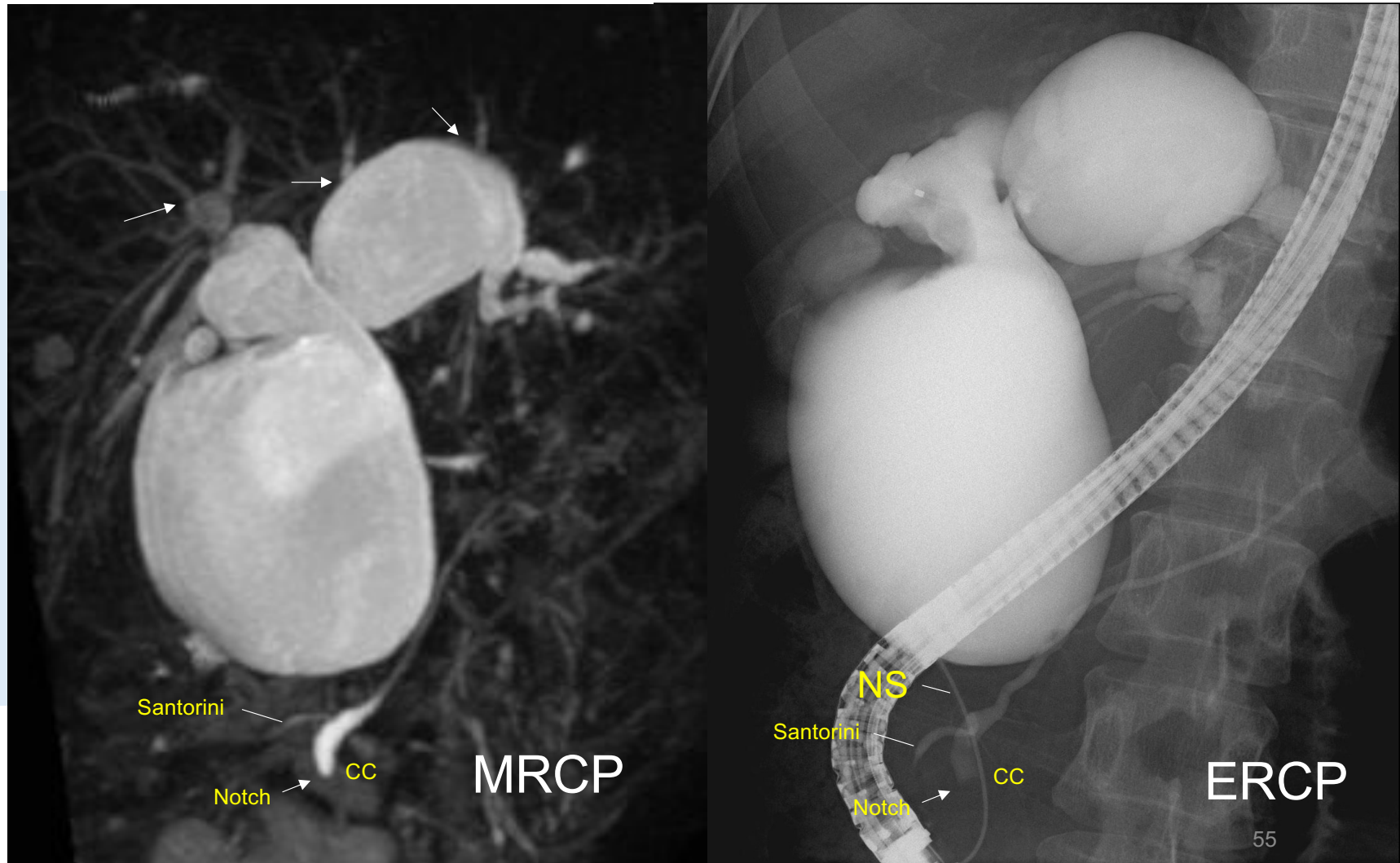
Case 16

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

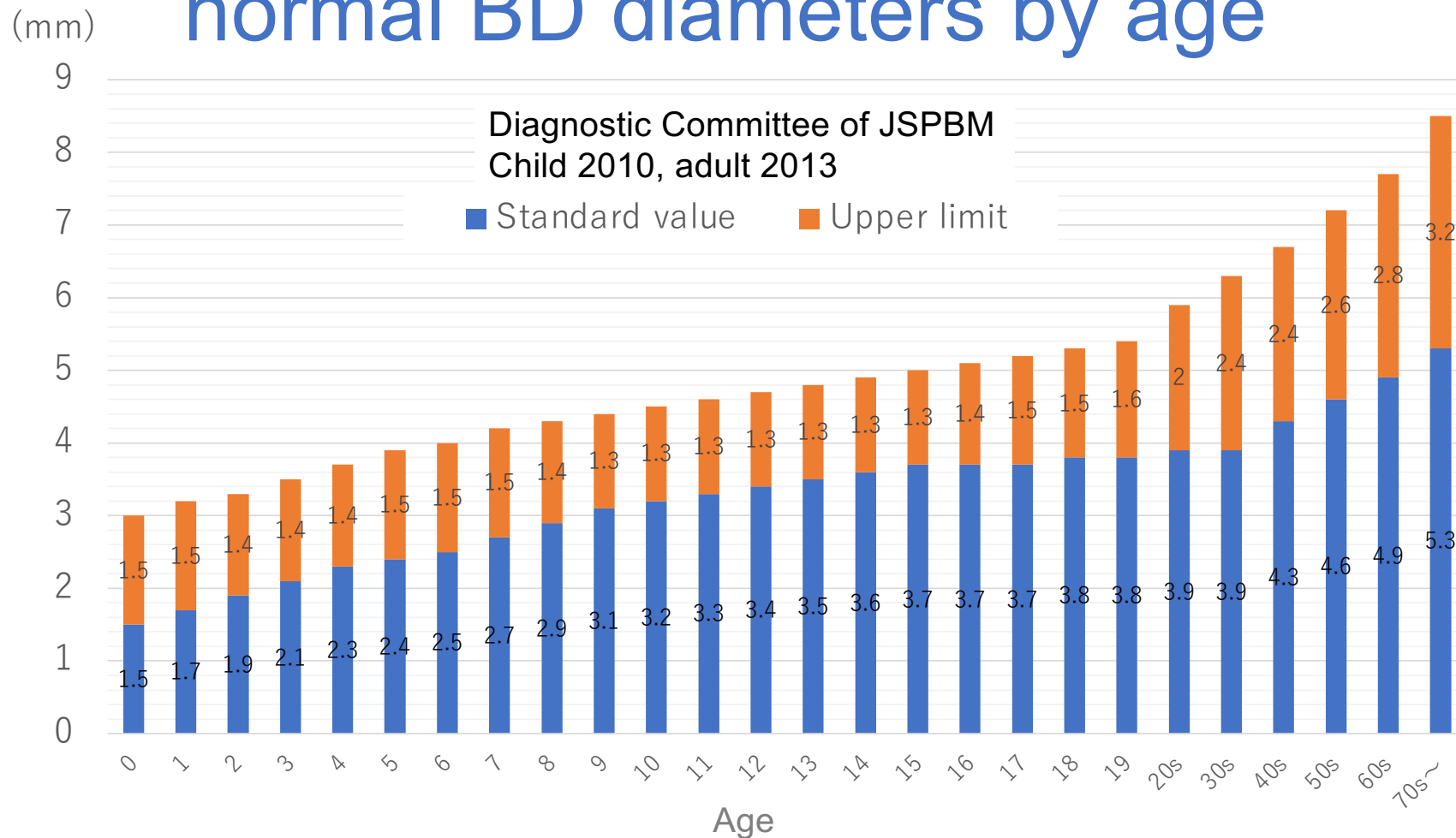
2nd version, September 2024

What is non-dilated type of PBM?

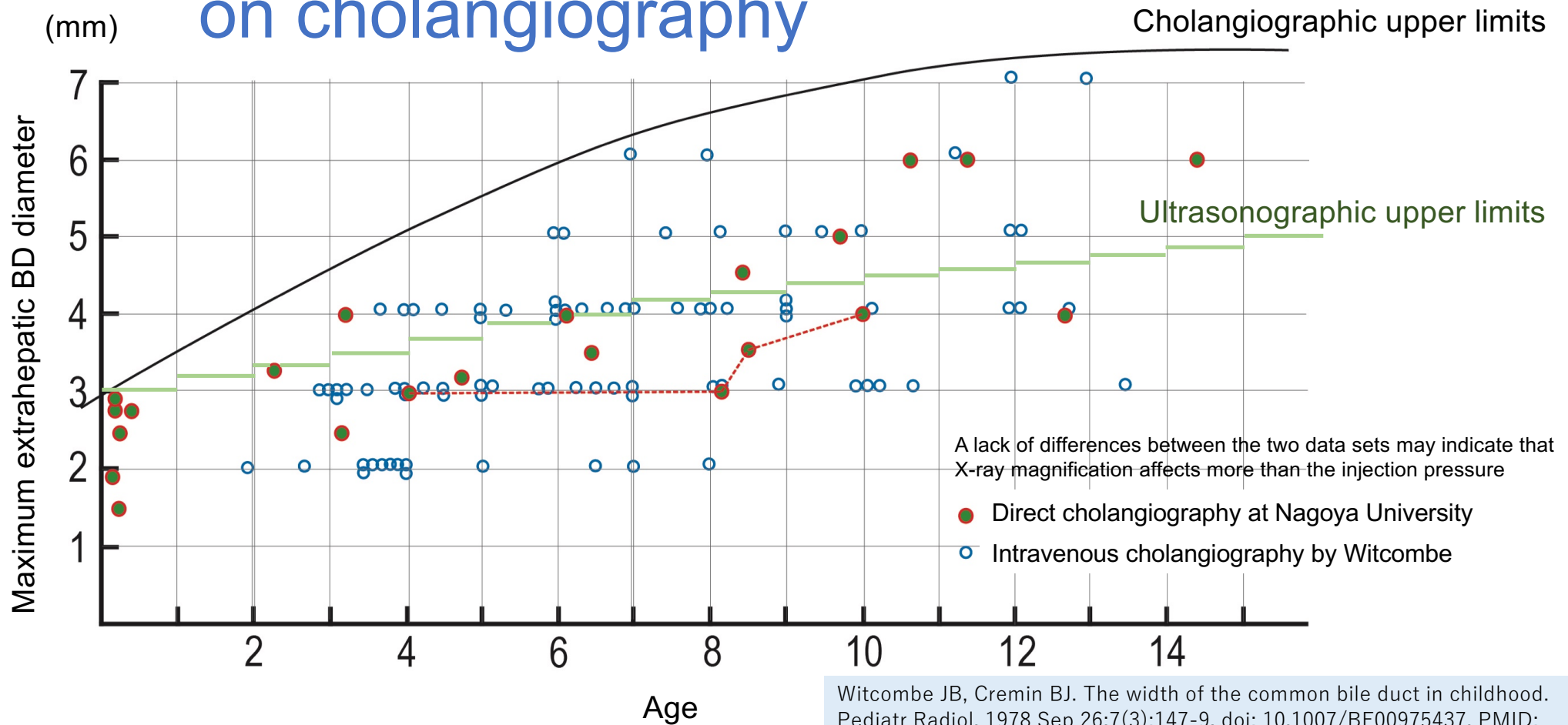
- Definition has not yet been determined.
- BD diameter **increases with age** in both children and adults. Ultrasonographic upper limits of normal BDs by age are determined.
- There is little data of normal BD diameters on direct cholangiography* or MRCP** at present.
- Non-dilated type should have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



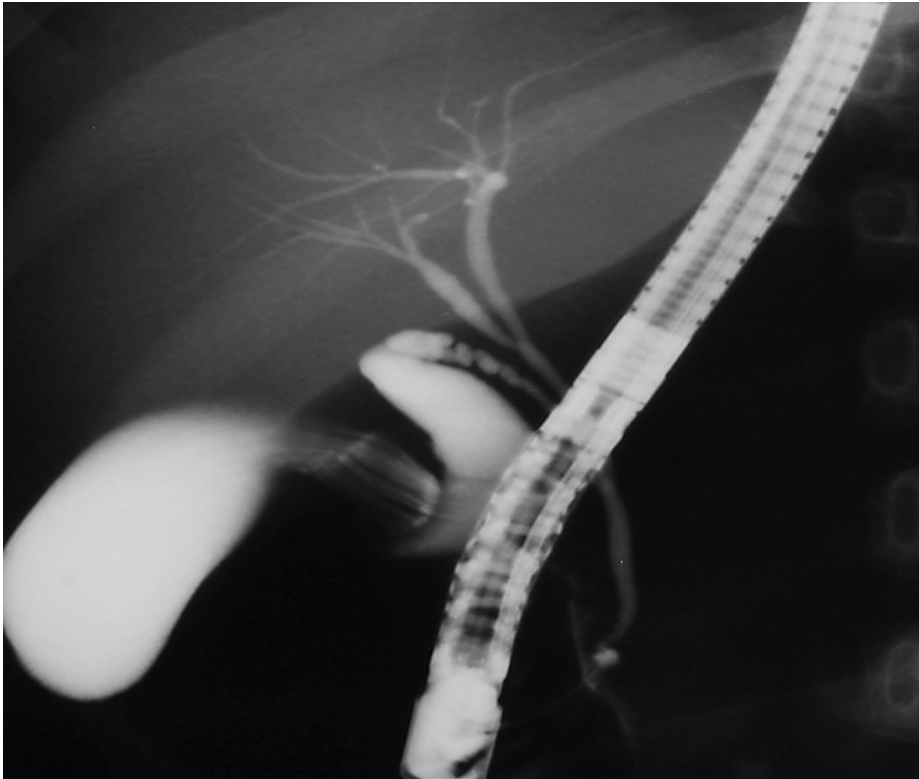
Normal pediatric BD diameters on cholangiography



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

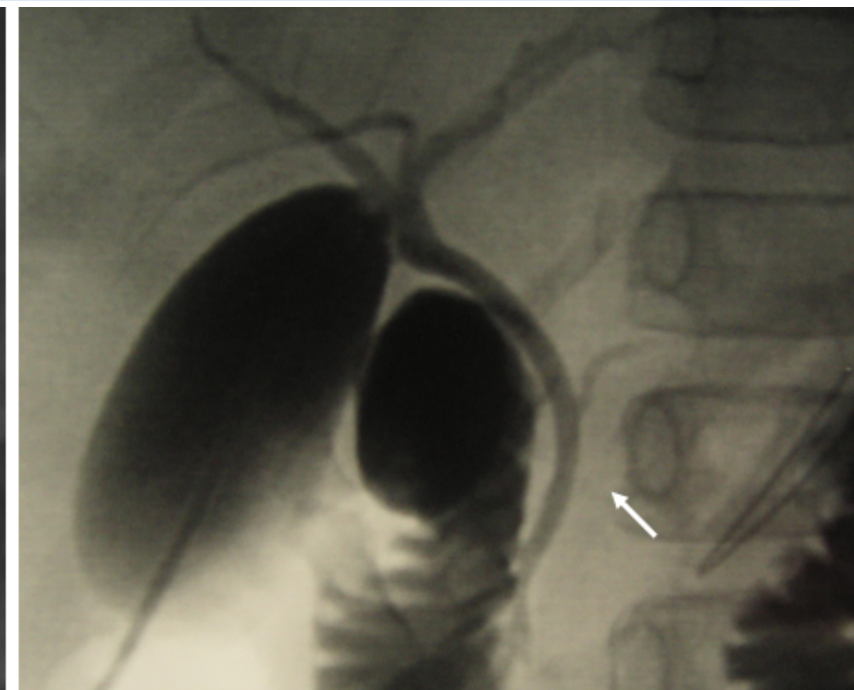
Six-year-old girl
Abdominal pain, vomiting,
hyperamylasemia

- **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

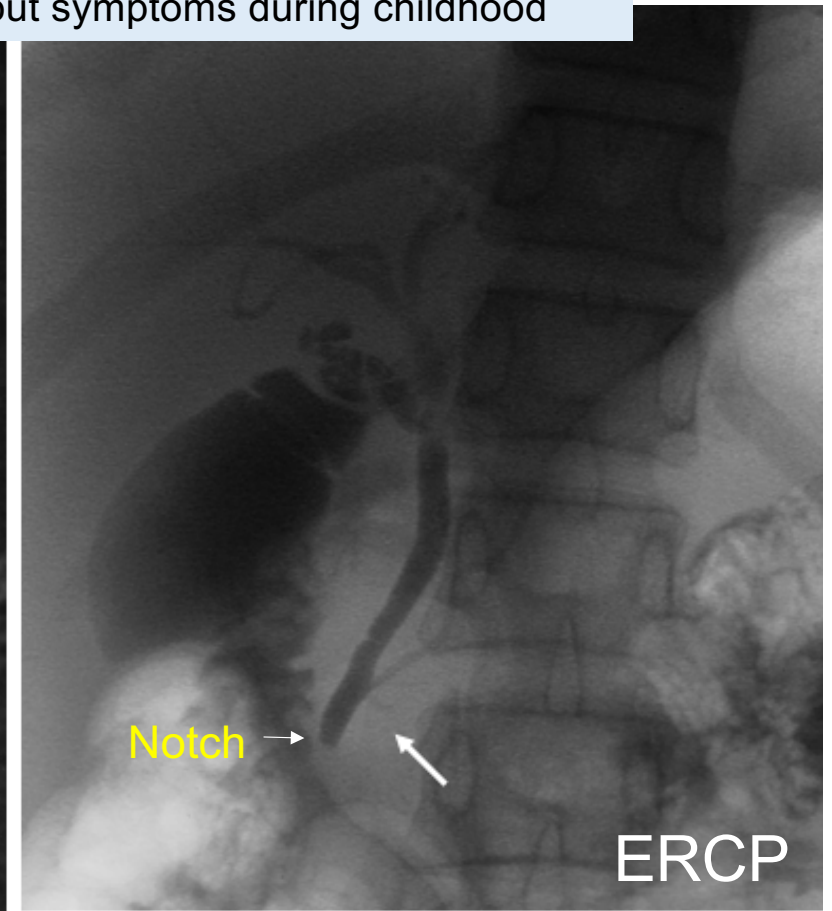
Typical non-dilated type of PBM: Pediatric case

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

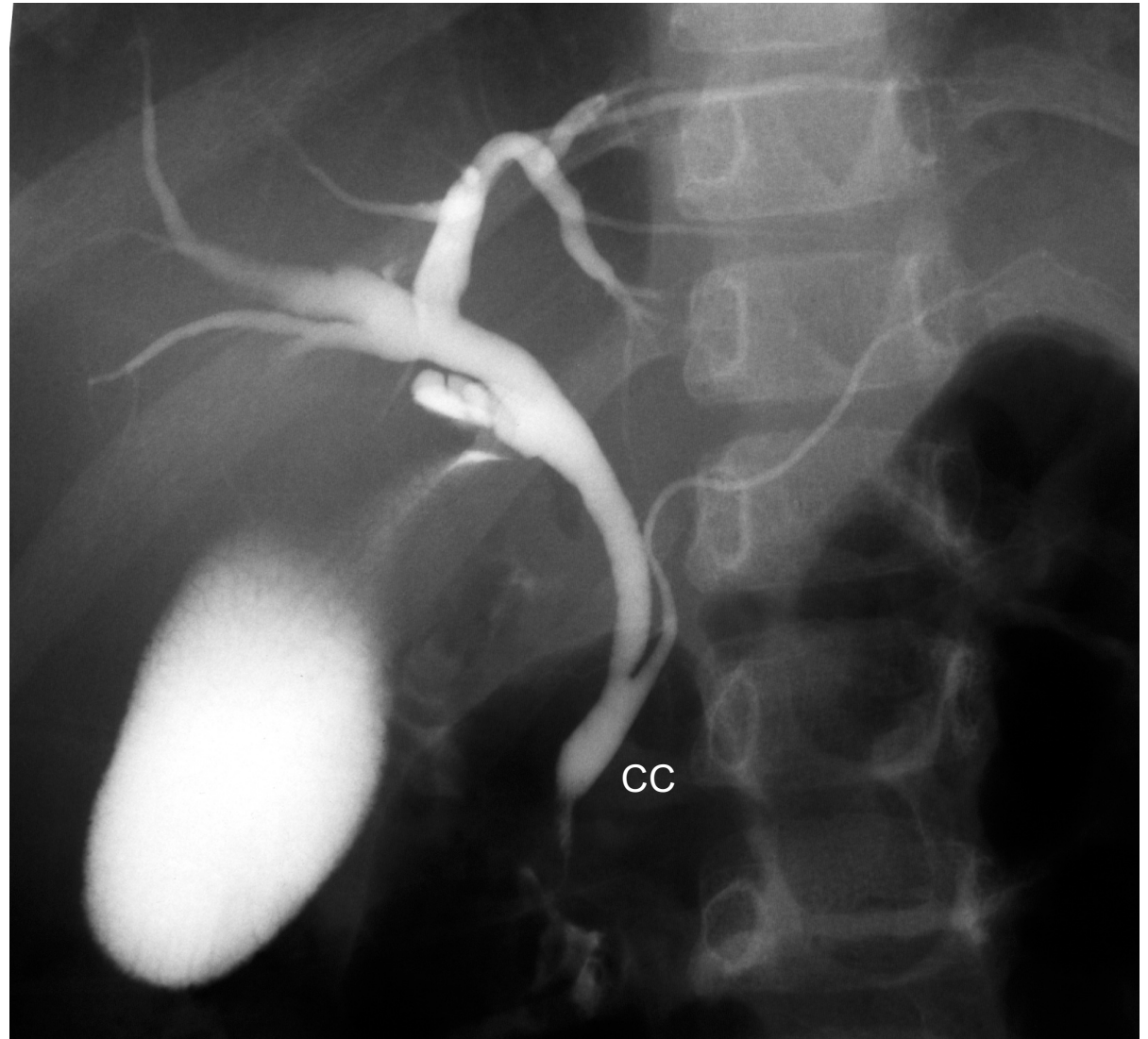
Non-dilated type?

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

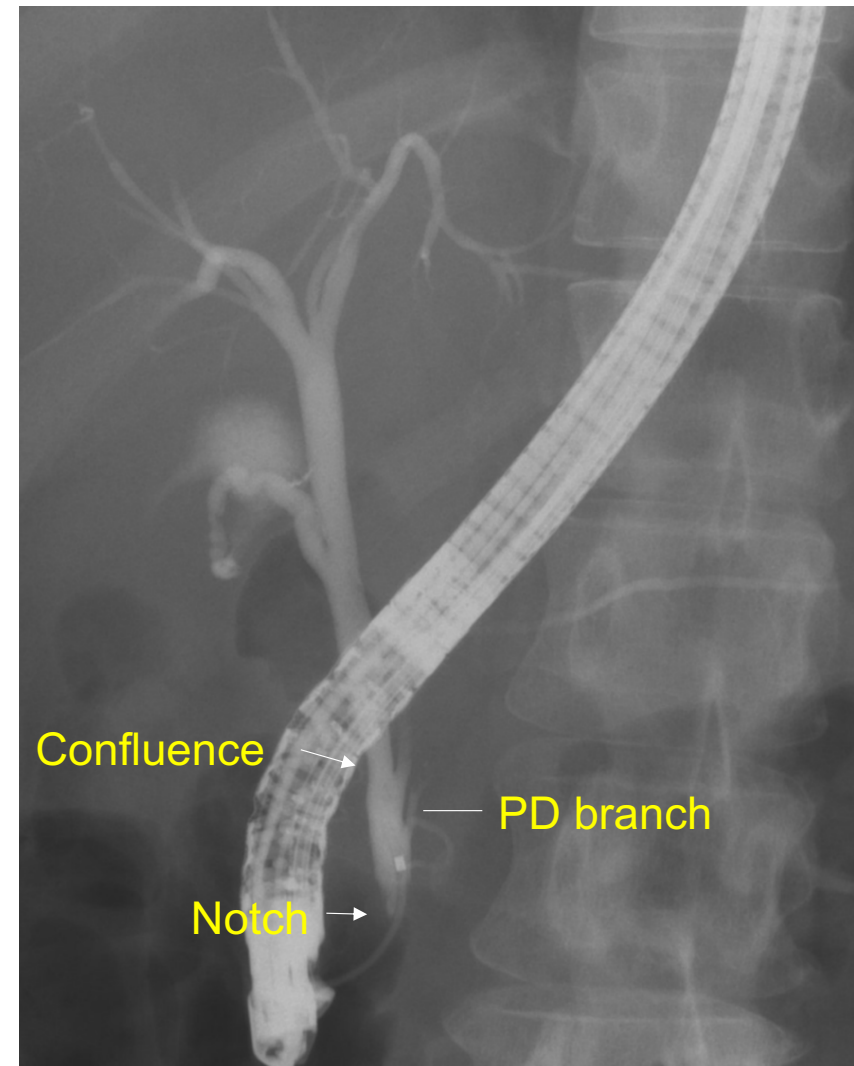
Typical non-dilated PBM: Adult case

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.

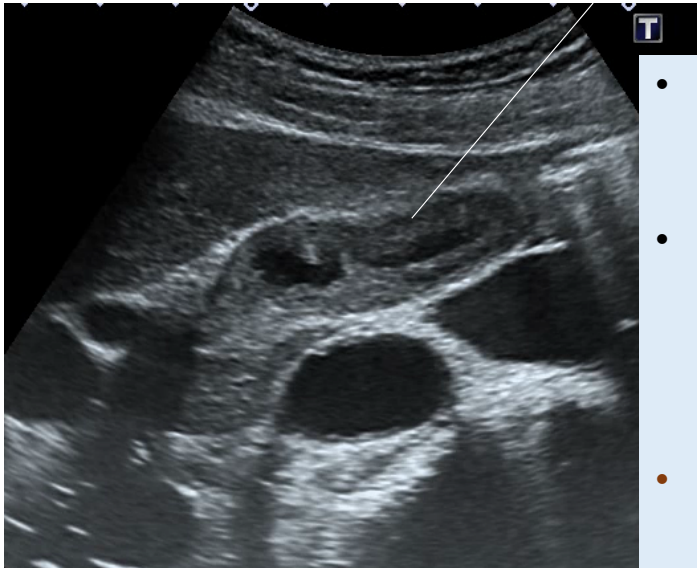


Case 22

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

A female in her fifties

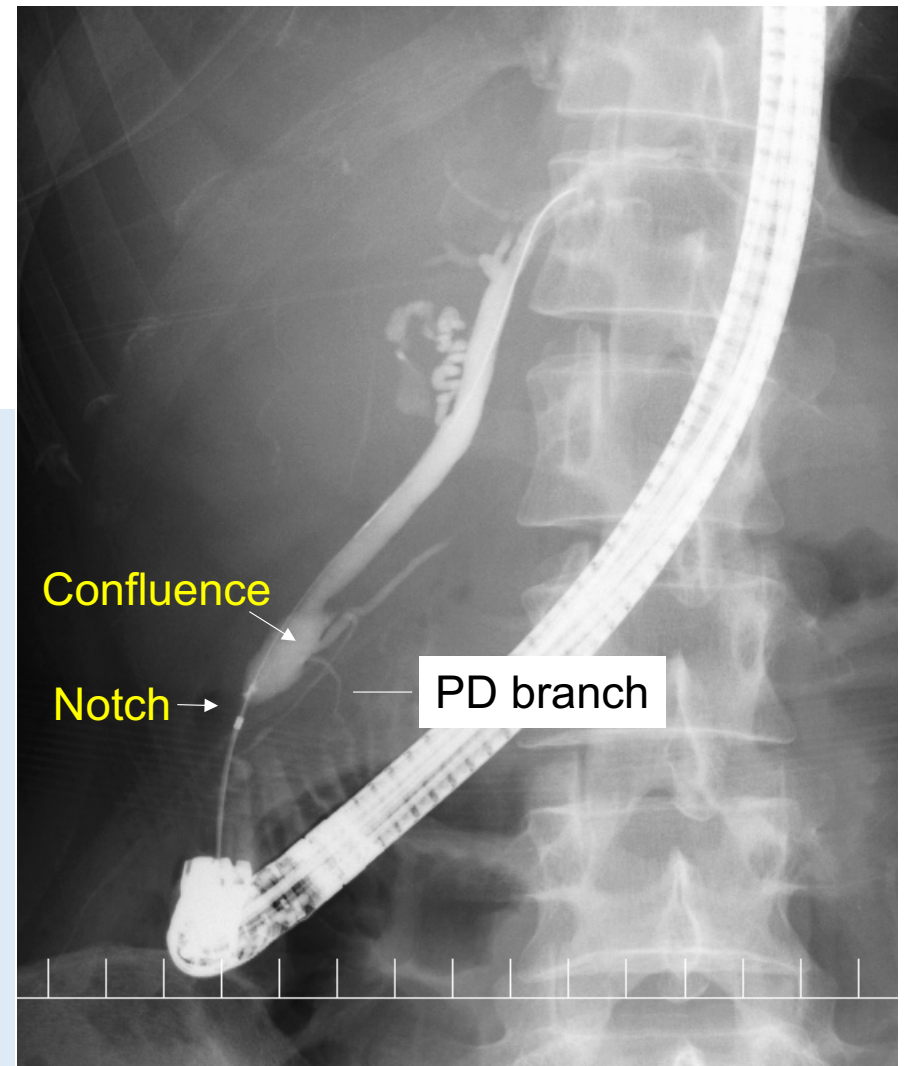
Screening US found GB wall thickness



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

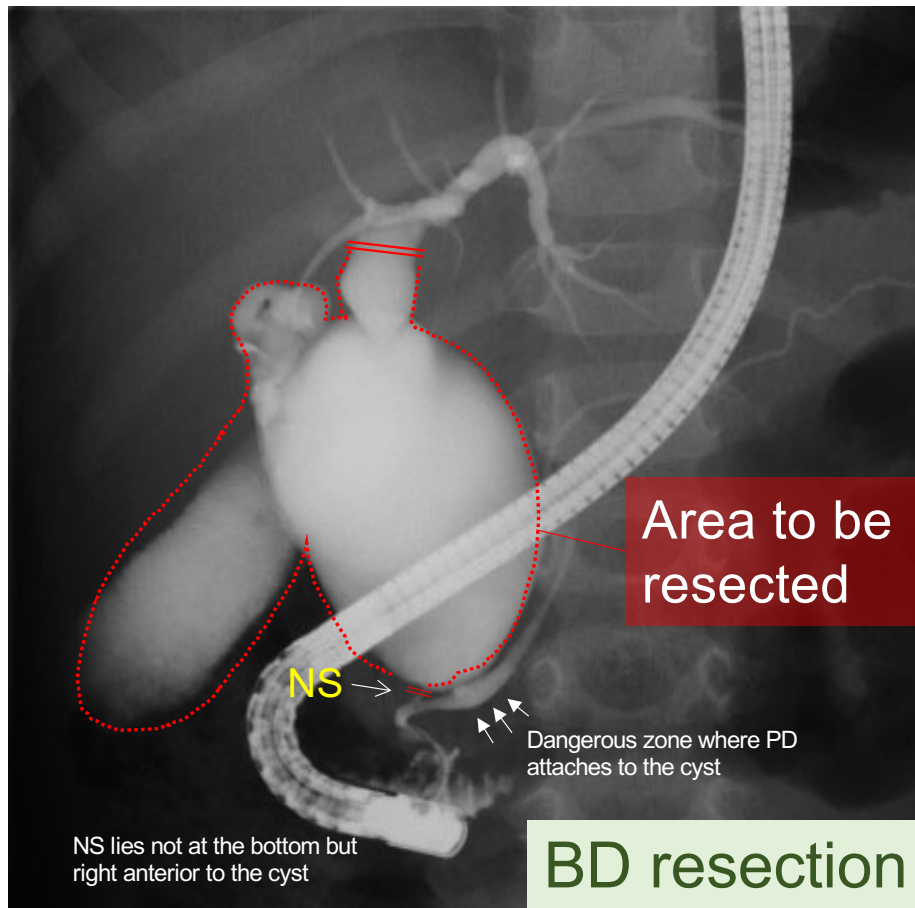
Treatment for pancreaticobiliary maljunction

- 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!

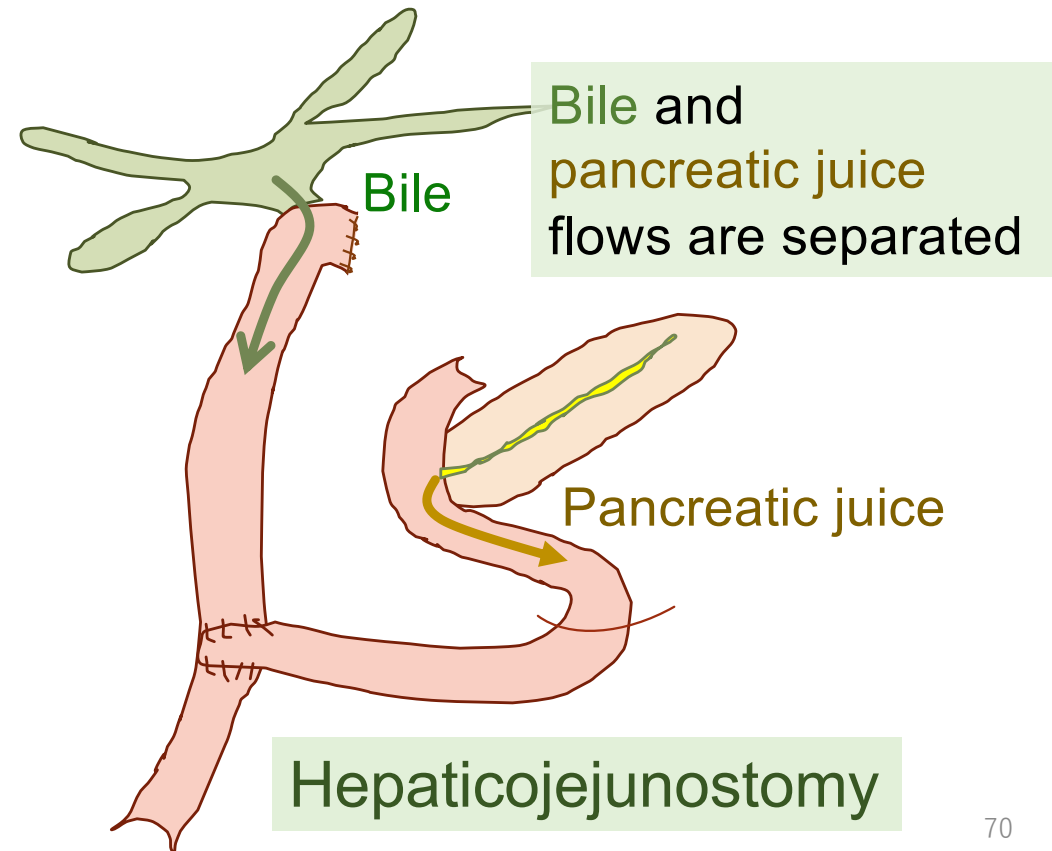
*No consensus on the need for bile duct resection because of little incidence of carcinogenesis at the BD
In pediatric cases, bile duct resection is common because the same symptoms as those of CBD occur due to the same mechanism, i.e. protein plugs.

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

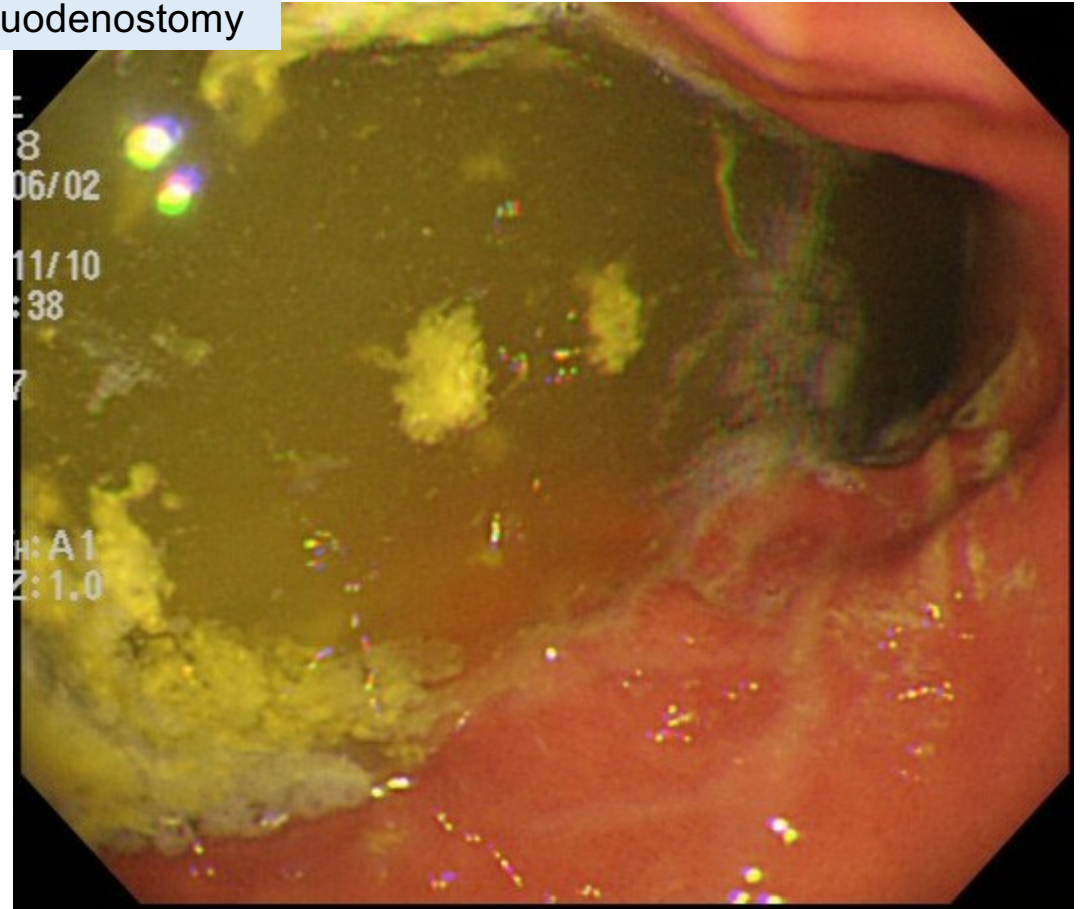
Bile gastritis after hepaticoduodenostomy

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.

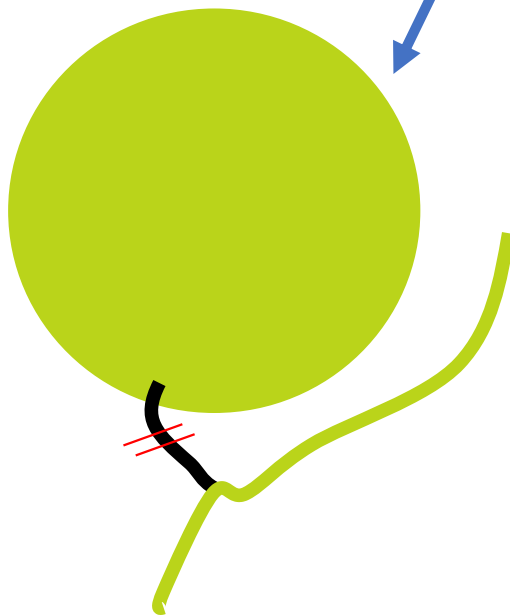
Three months after the examination, she underwent hepaticojejunostomy from duodenostomy, which relieved her epigastric pain and endoscopic abnormality.



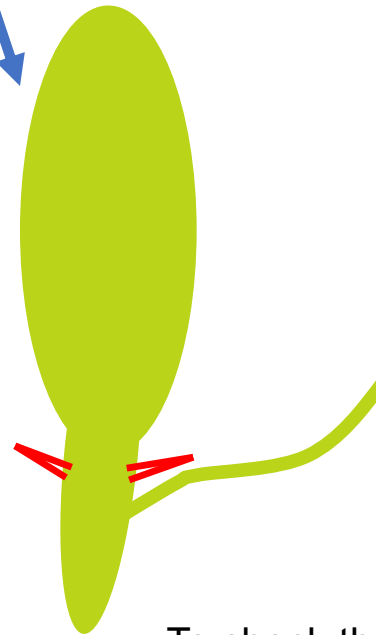
Complete resection of intrapancreatic BD

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



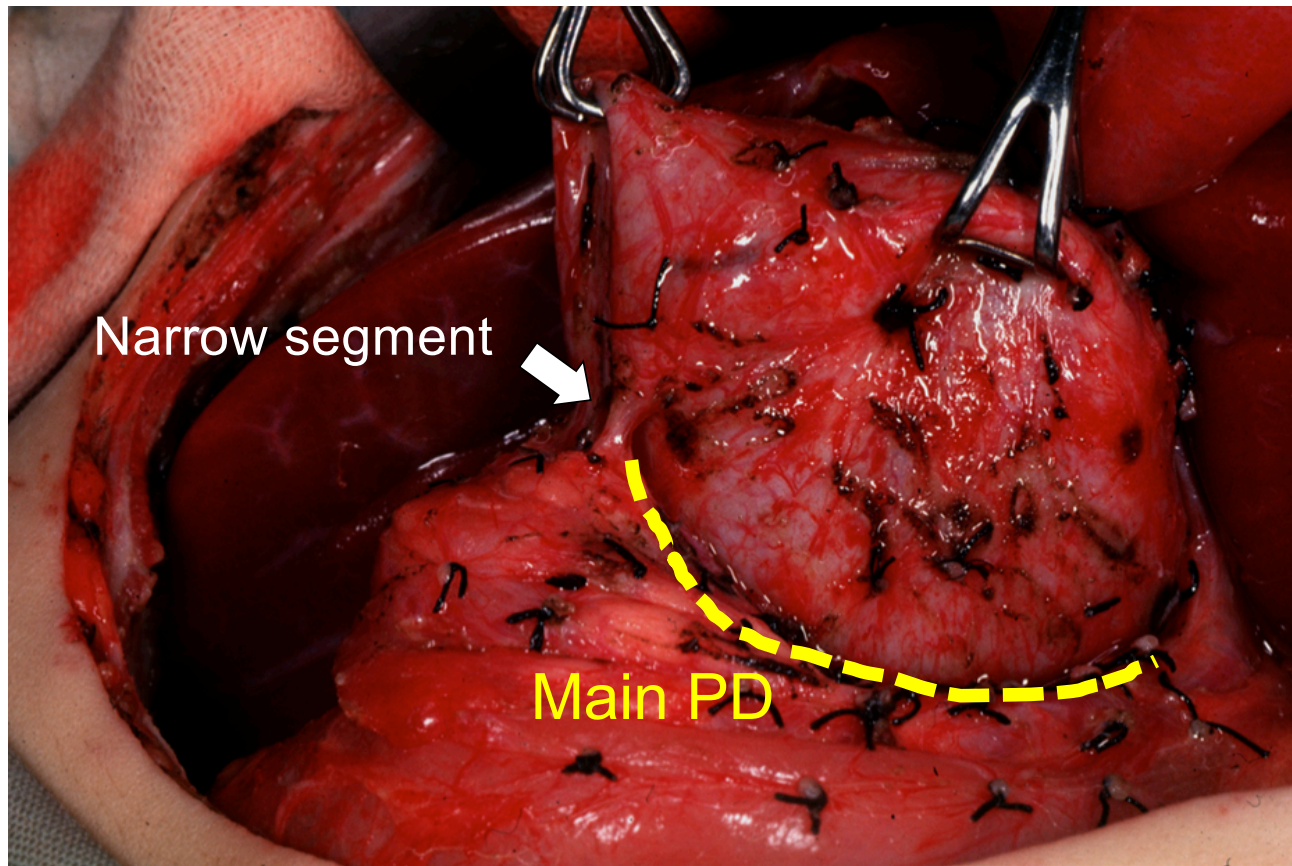
Visual confirmation of narrow segment



To check the distance to PD

Fluoroscopy using clips

Visual confirmation of narrow segment

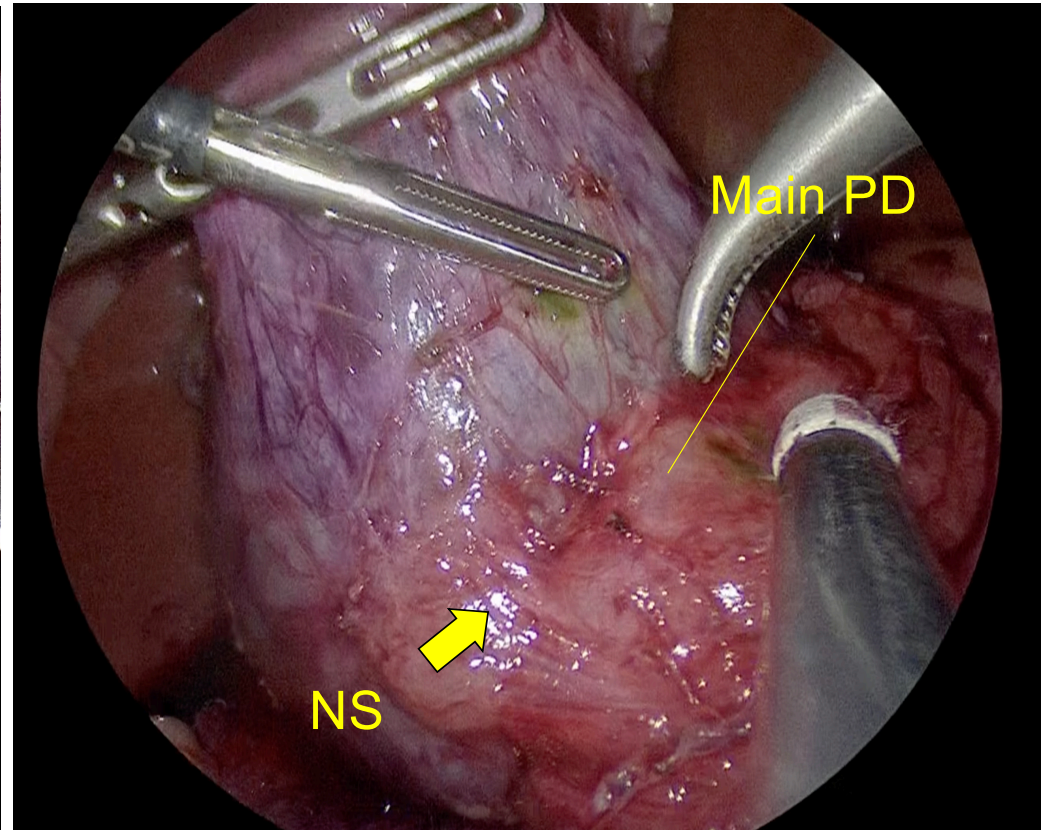
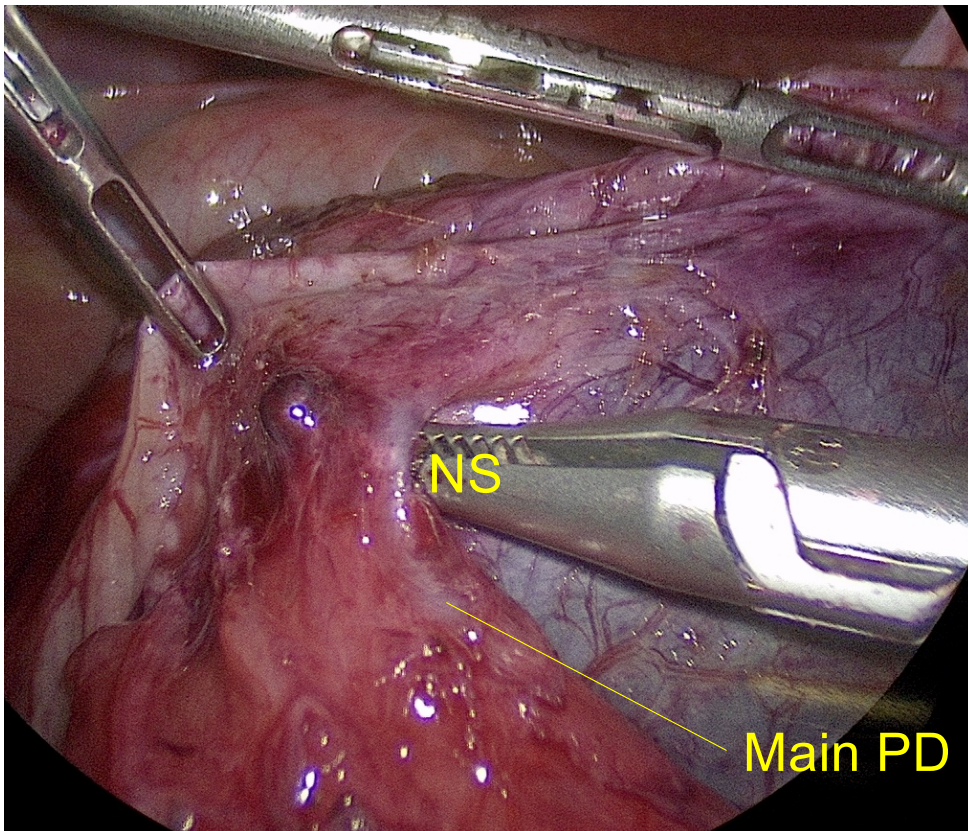


Narrow segment is often located not at the bottom but right-ventral to the cyst.

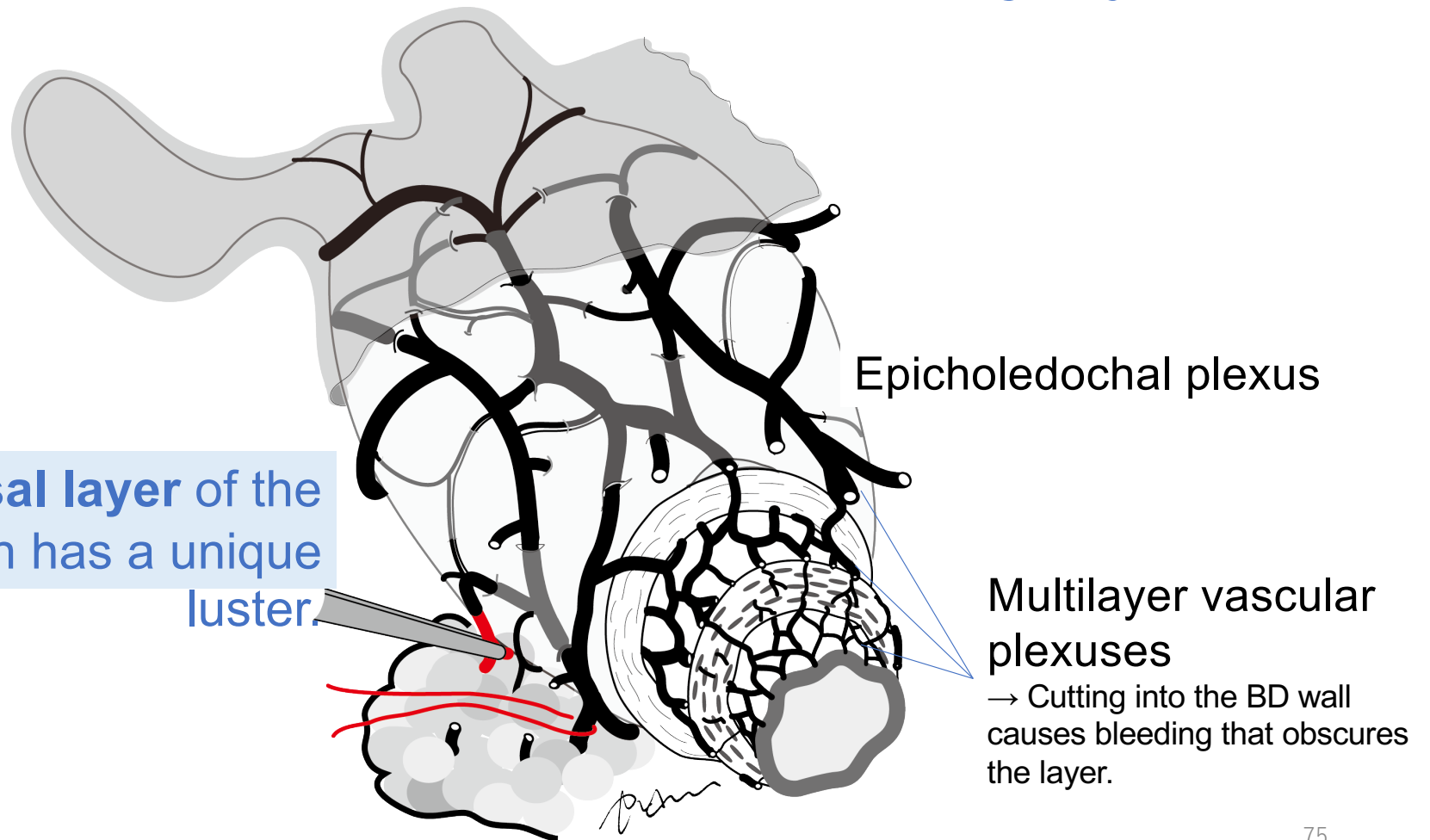
In this case, care must be taken not to injure the PD because the PD runs on the cyst.

PD adheres to the cystically dilated BD

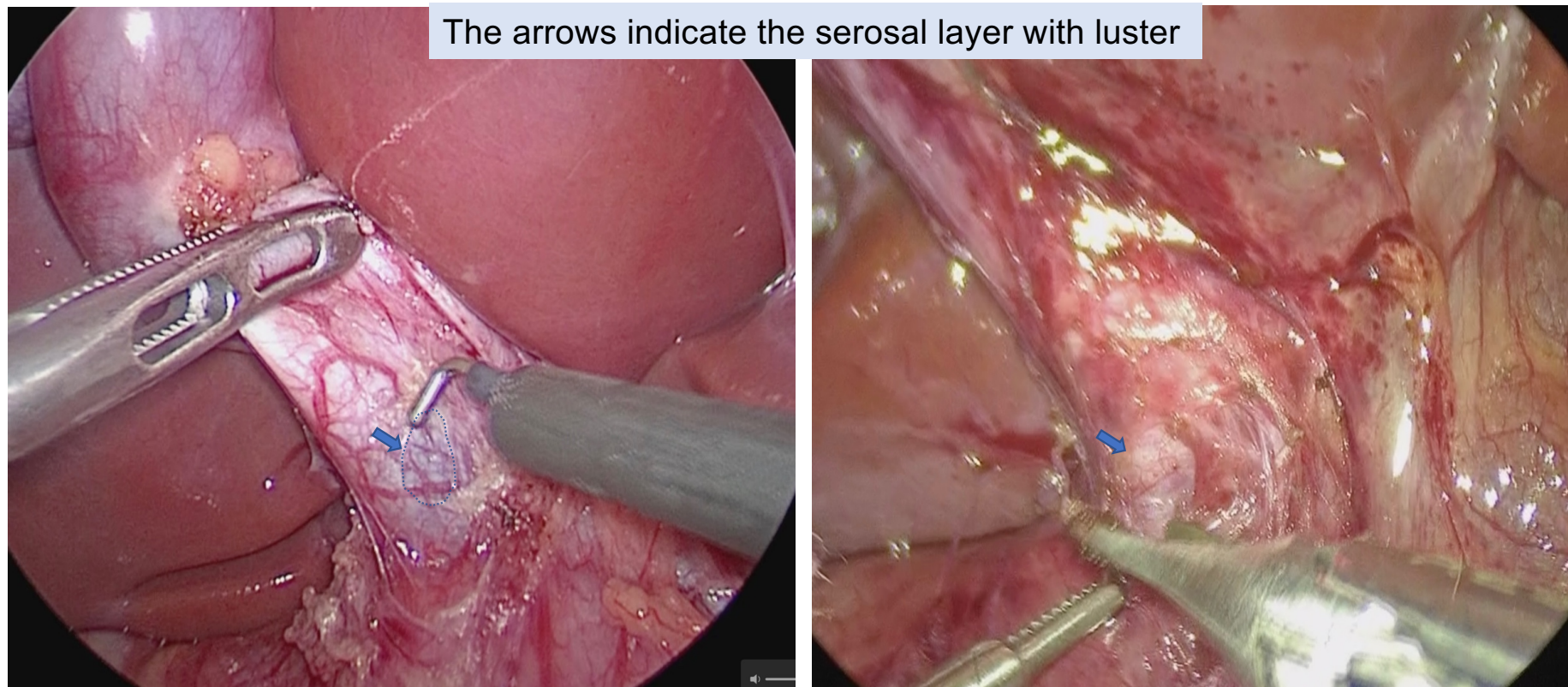
Infant case



How to avoid a pancreatic duct injury



BD serosa has a unique luster

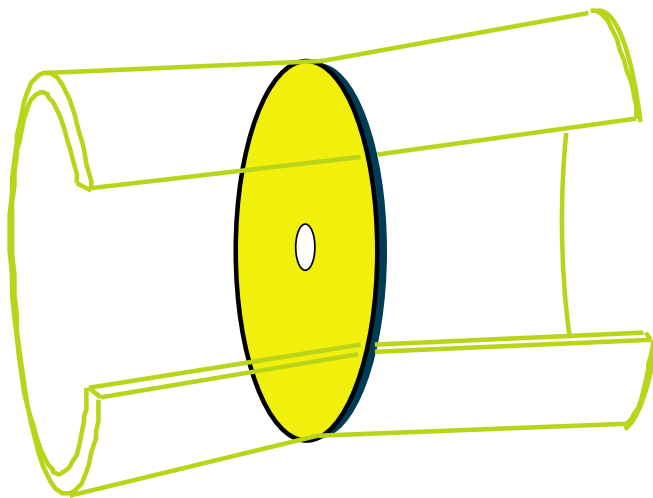


Case without inflammatory adhesion

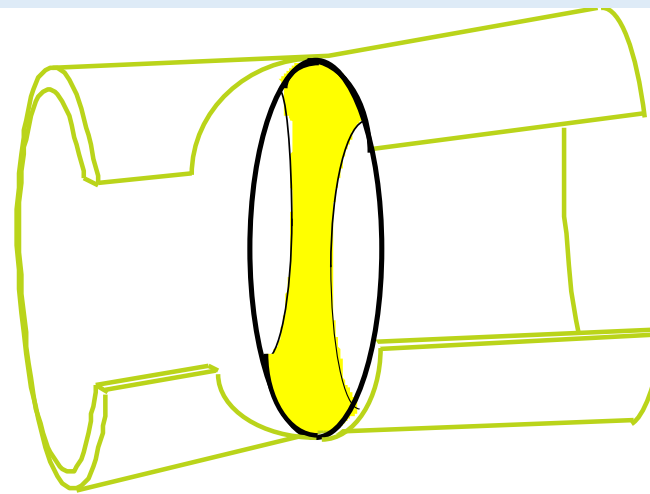
Case with inflammatory adhesion

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

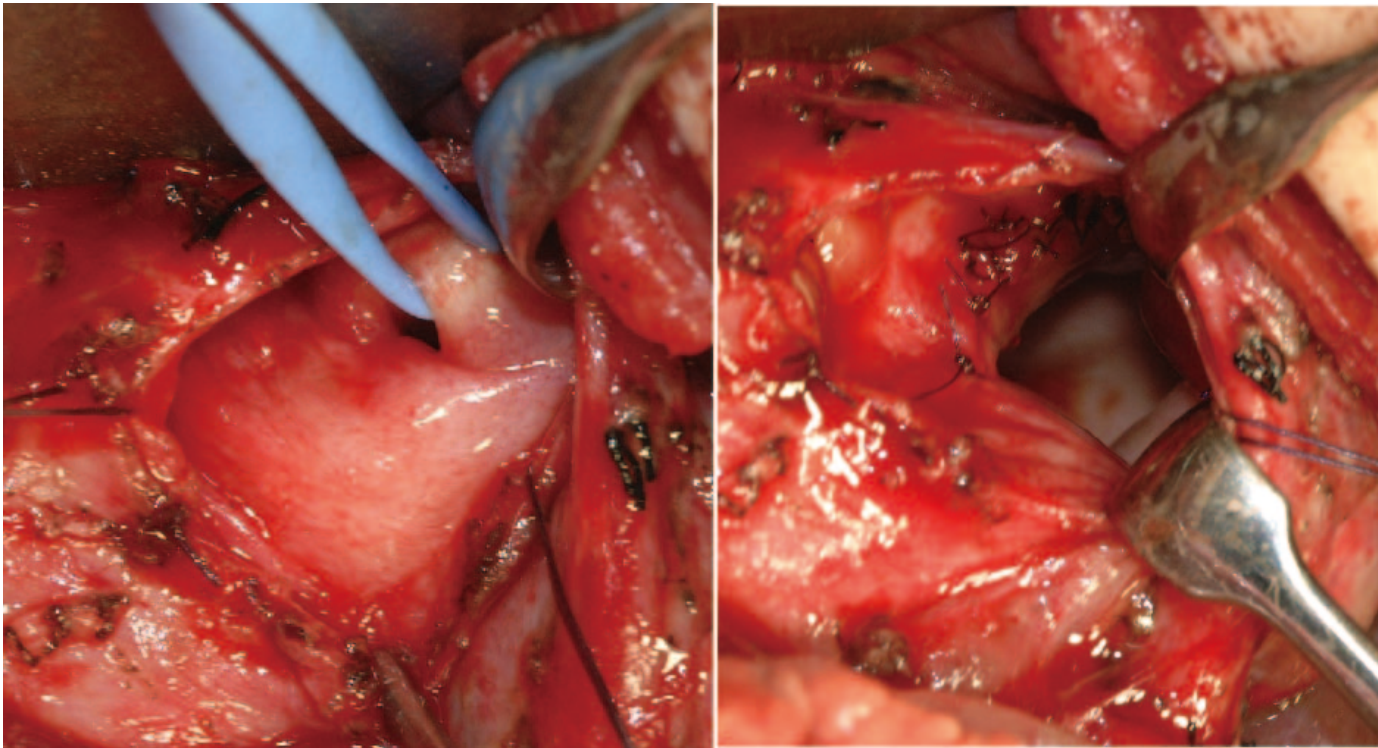


Septal stenosis

Balloon dilatation is not effective for these stenoses.

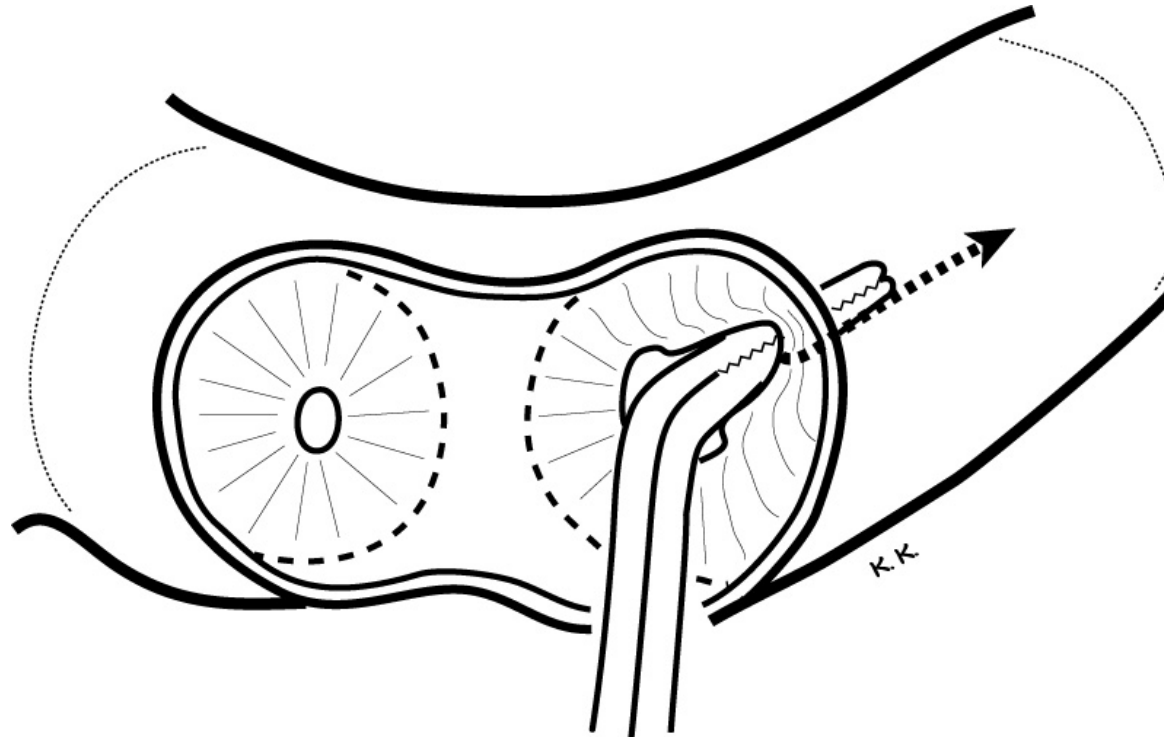
Ando H, Ito T, Kaneko K, Seo T.
Congenital stenosis of the intrahepatic bile duct associated
with choledochal cysts. J Am Coll Surg. 1995
Nov;181(5):426-30. PMID: 7582210.

Resection of septal stenosis



At the time of BD resection

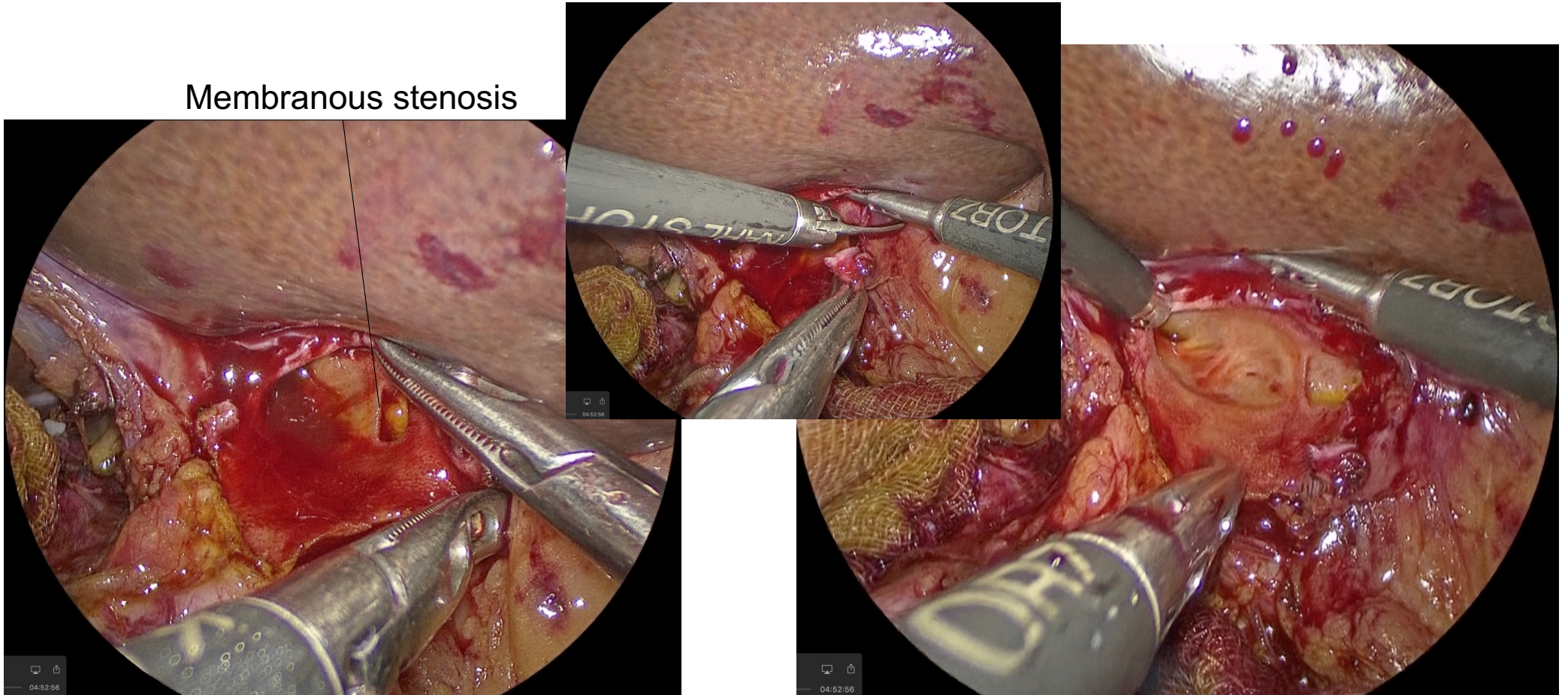
Todani and Lilly's BD plasty for membranous stenosis



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

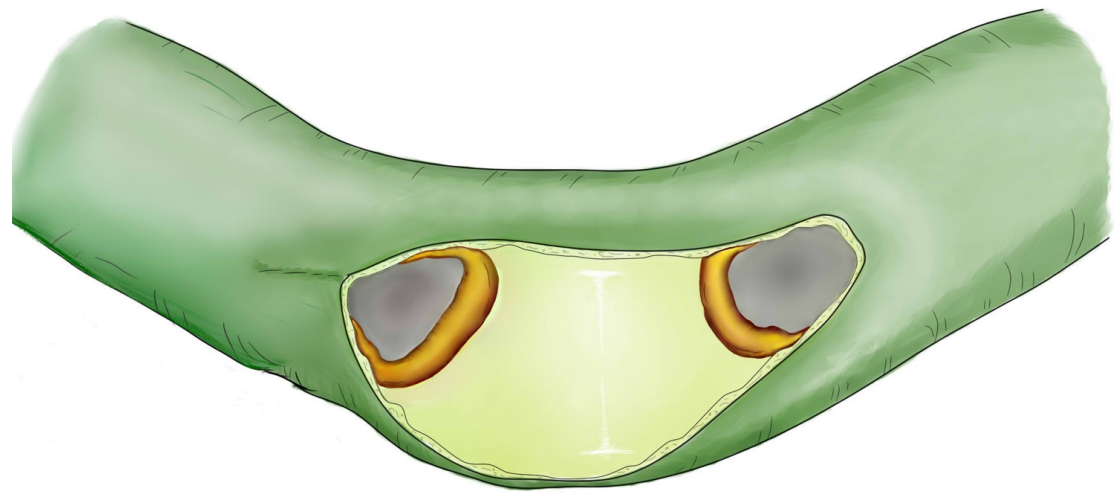
Todani's BD plasty

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

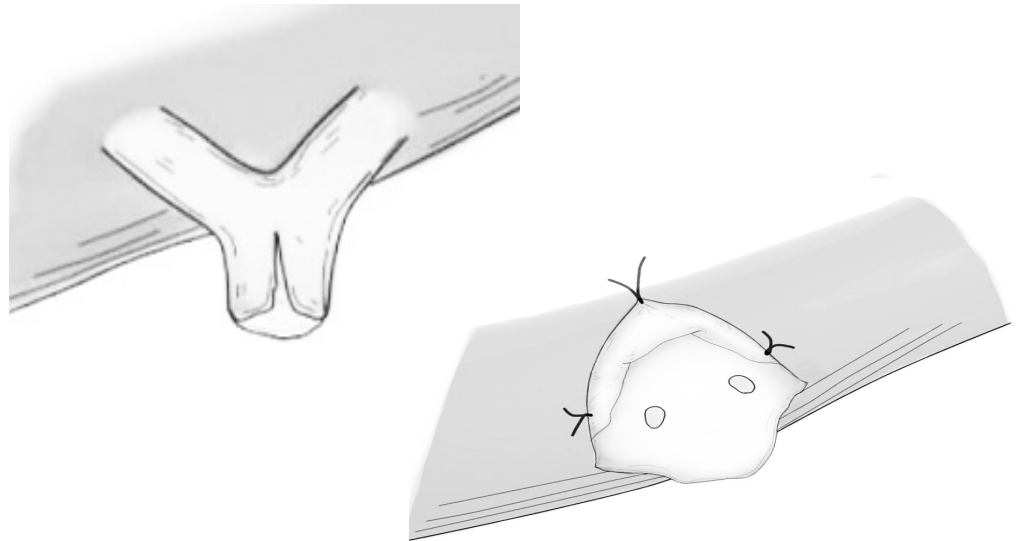
Procedures to prevent anastomotic stenosis in cases with small-caliber hepatic ducts

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

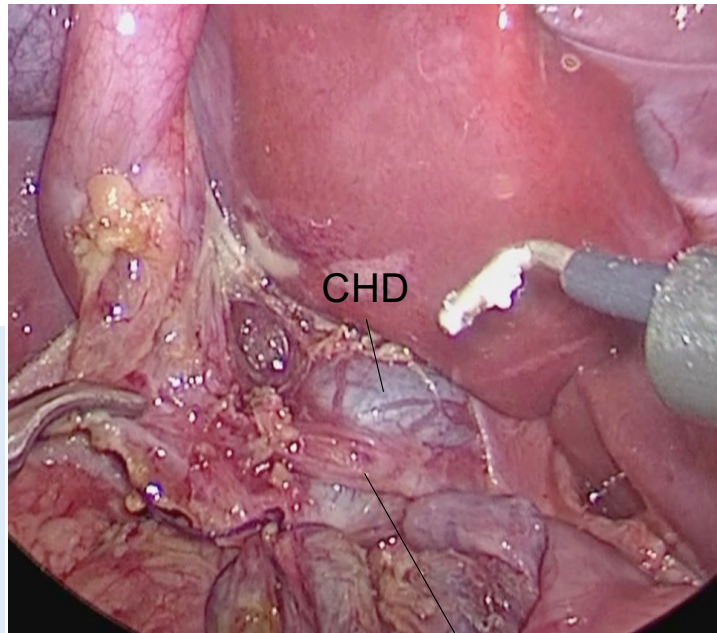
Case 23

Four-year-old boy
Type Ia CBD

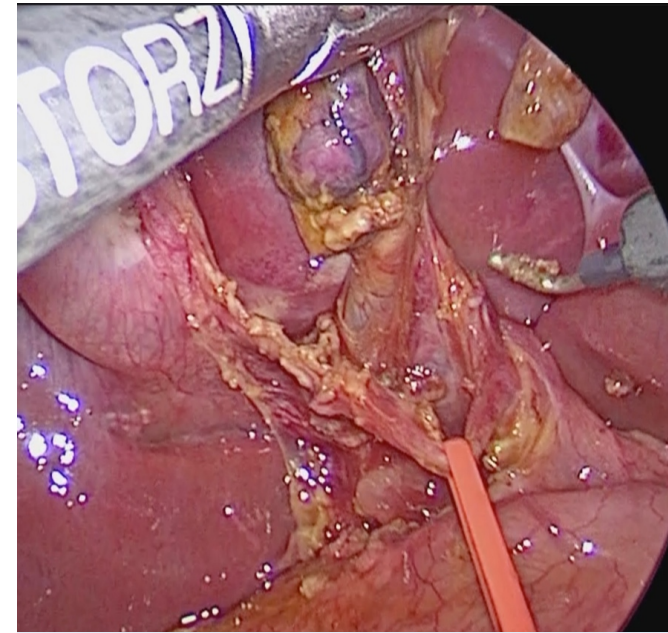
RHA ventral to CHD

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

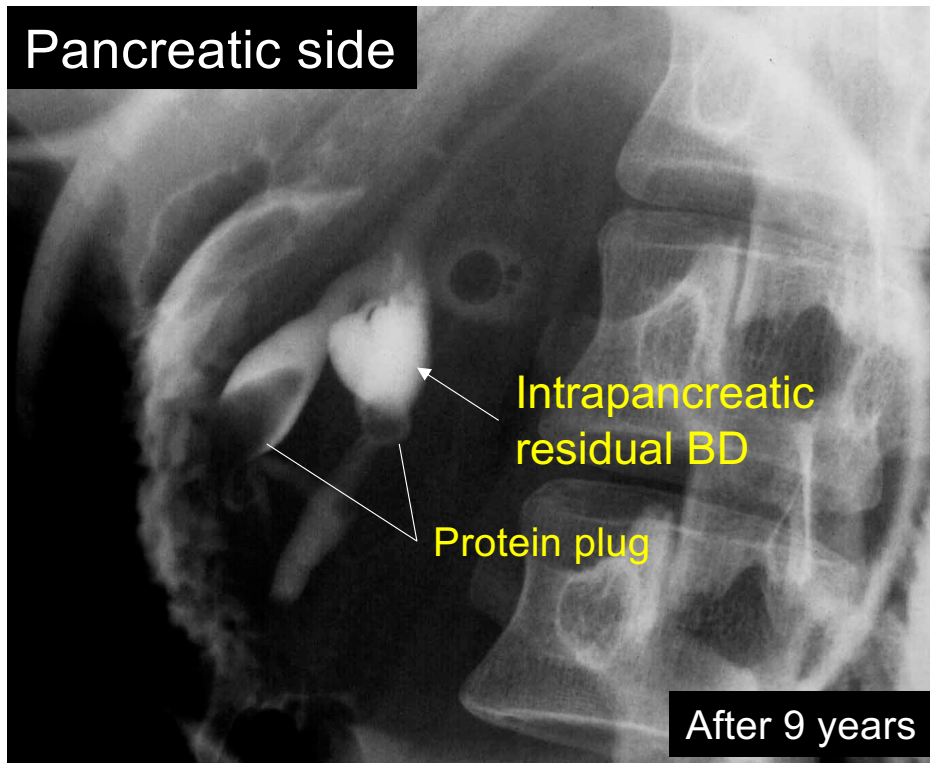
3) Guo WL, Wang J. Preoperative one-stop magnetic resonance imaging evaluation of the pancreaticobiliary junction and hepatic arteries in children with pancreaticobiliary maljunction: a prospective cohort study. Surg Today. 2021 Jan;51(1):79-85. doi: 10.1007/s00595-020-02077-5. Epub 2020 Jul 12. PMID: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

Basics

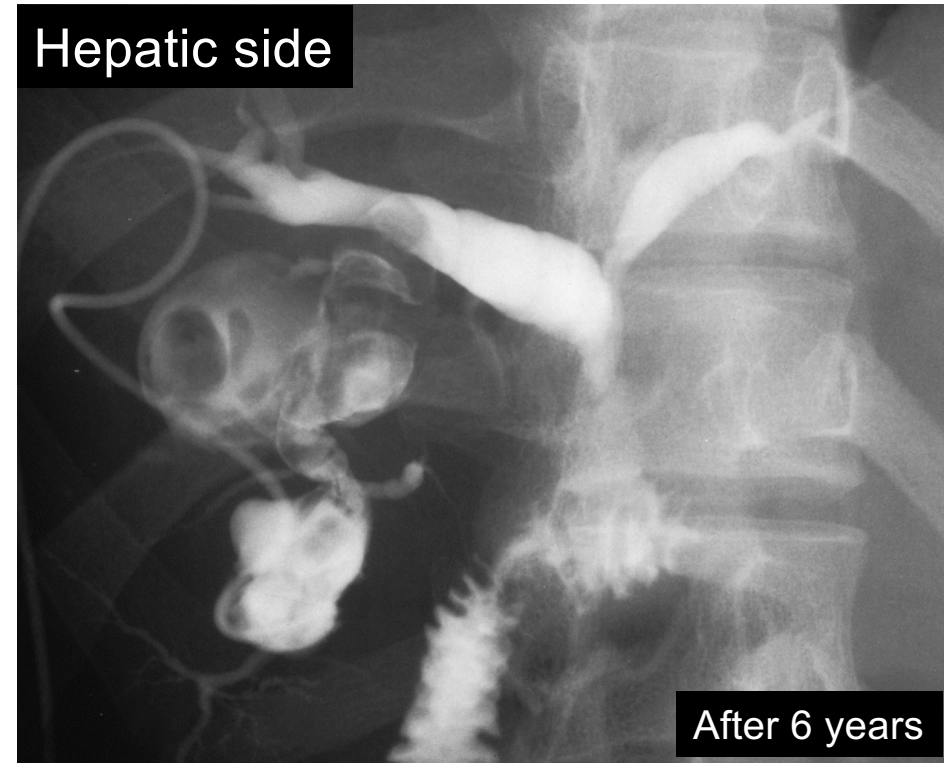
Postoperative Complications

First version, September 2022

Long-term complications after BD resection



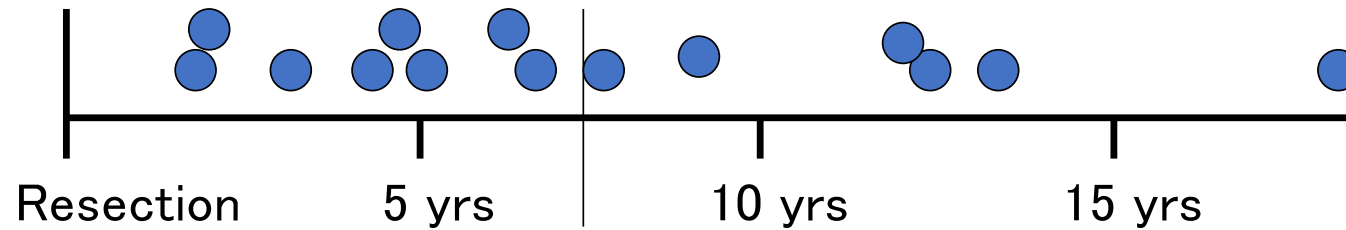
A residual intrapancreatic BD would cause protein plugs or intrapancreatic stones and cancers in the residual BD. Complete excision of intrapancreatic BDs is necessary.



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

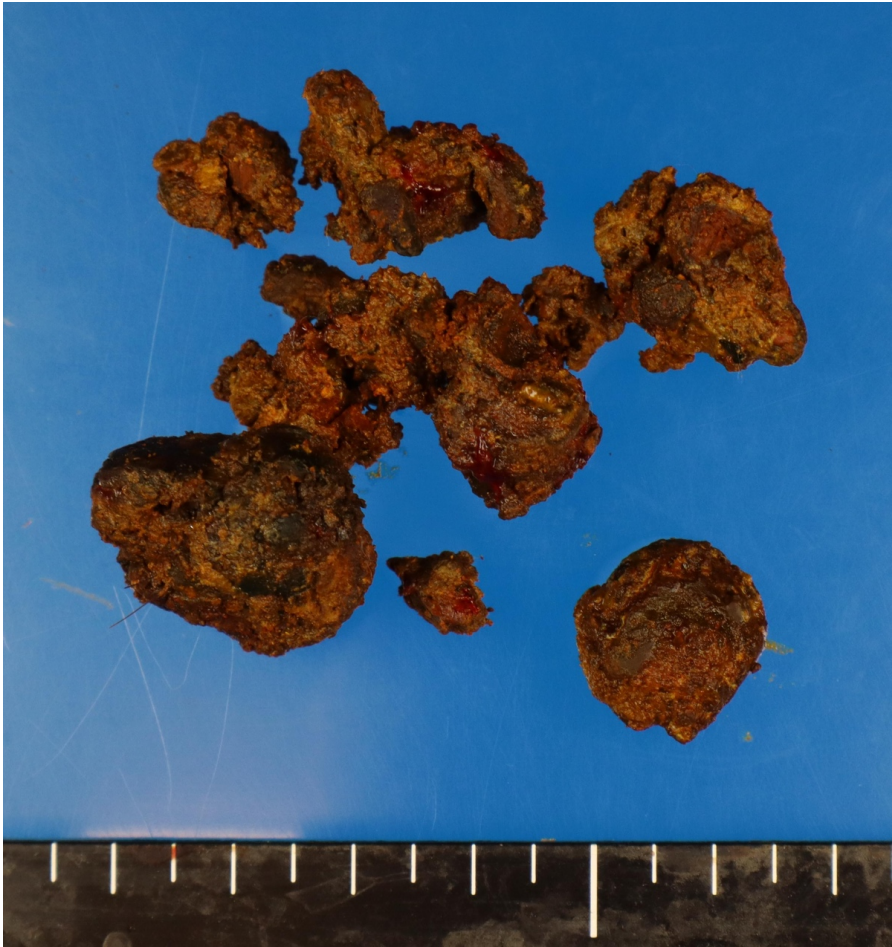
Period from resection to intrahepatic gallstone formation

Pediatric 14 cases



Average 7.3 yrs

Postoperative gallstones are brown pigment stones



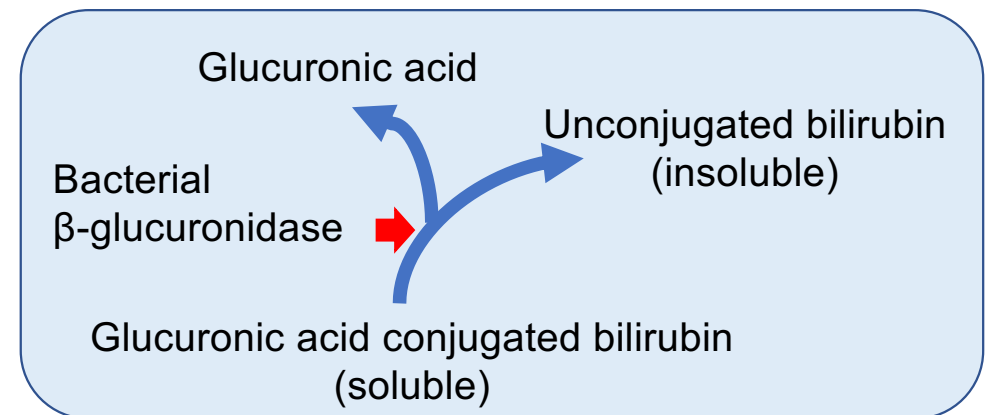
Composition:

Bilirubin calcium + fatty acid calcium



Bacterial β -glucuronidase and phospholipase A1

Bile stasis and bacterial infection



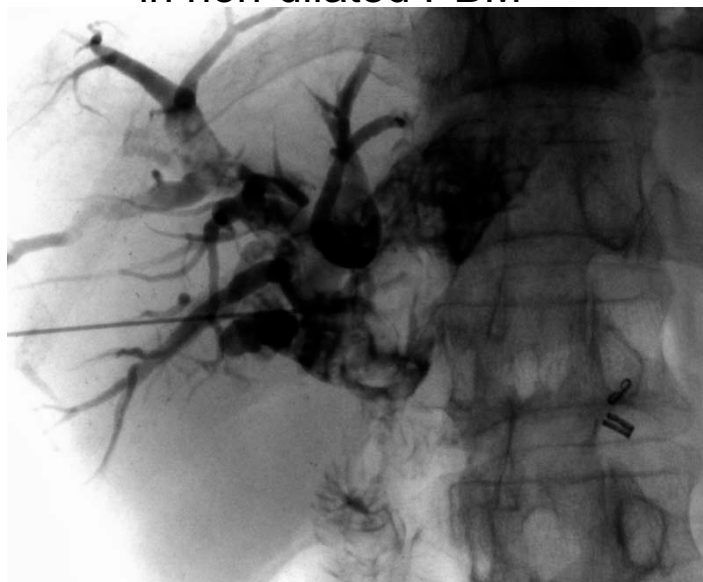
Risk factors of postoperative hepatolithiasis

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

Todani type IV-A



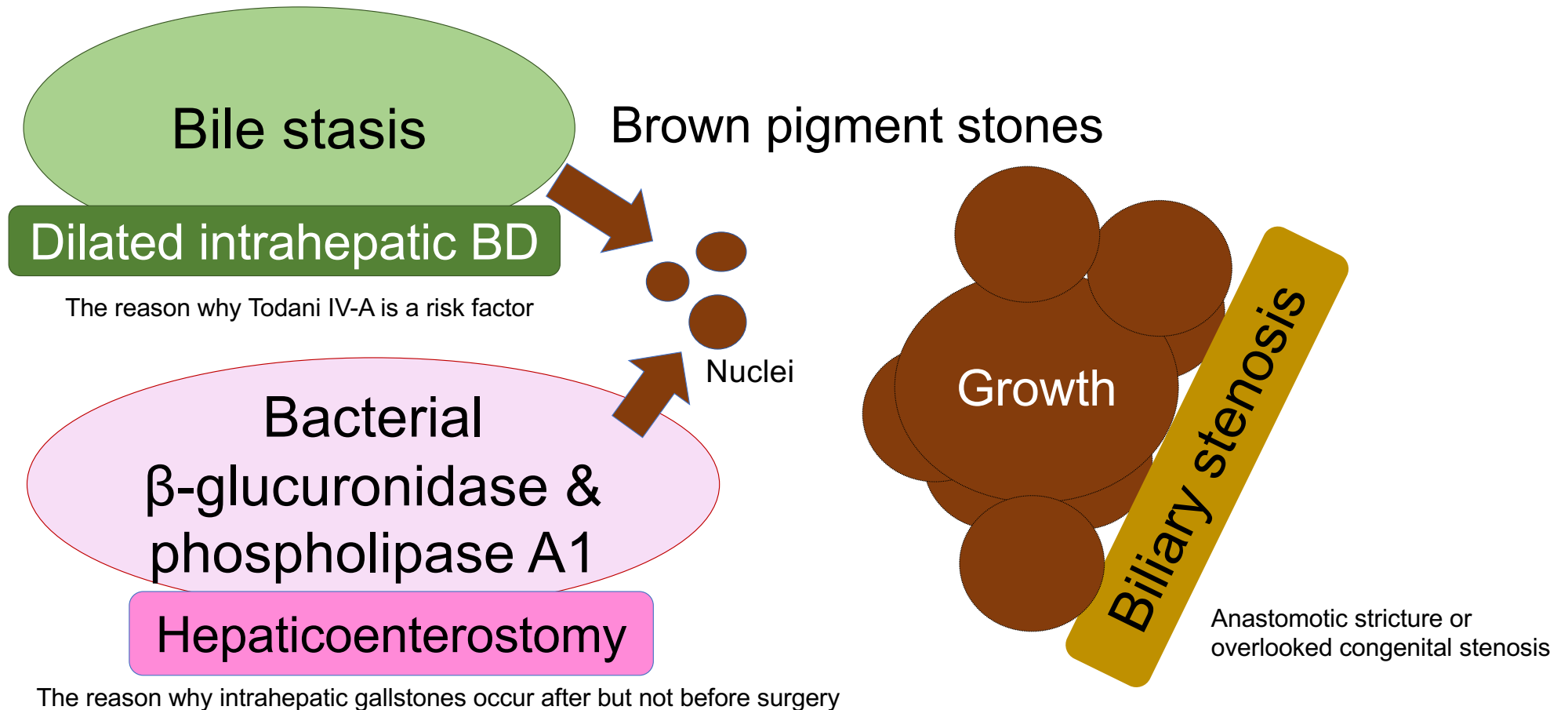
Anastomotic stricture
in non-dilated PBM



Congenital septal stenosis



Mechanism of postoperative gallstone formation

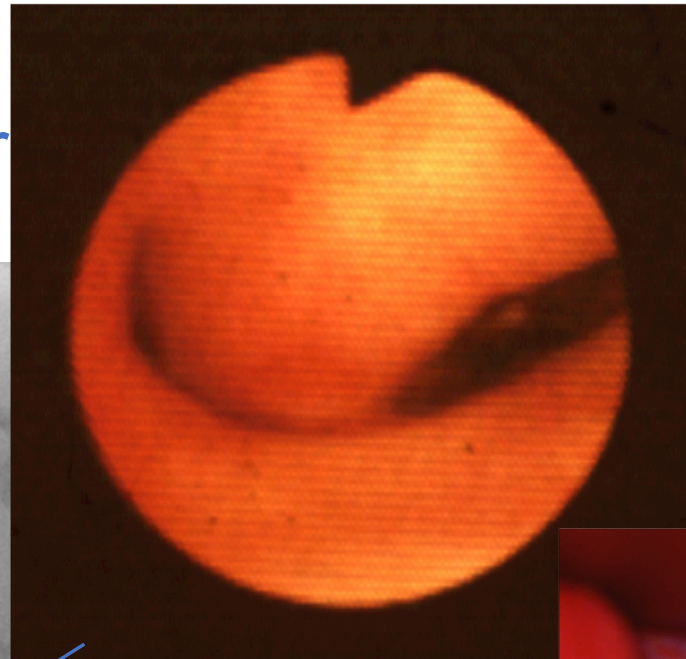
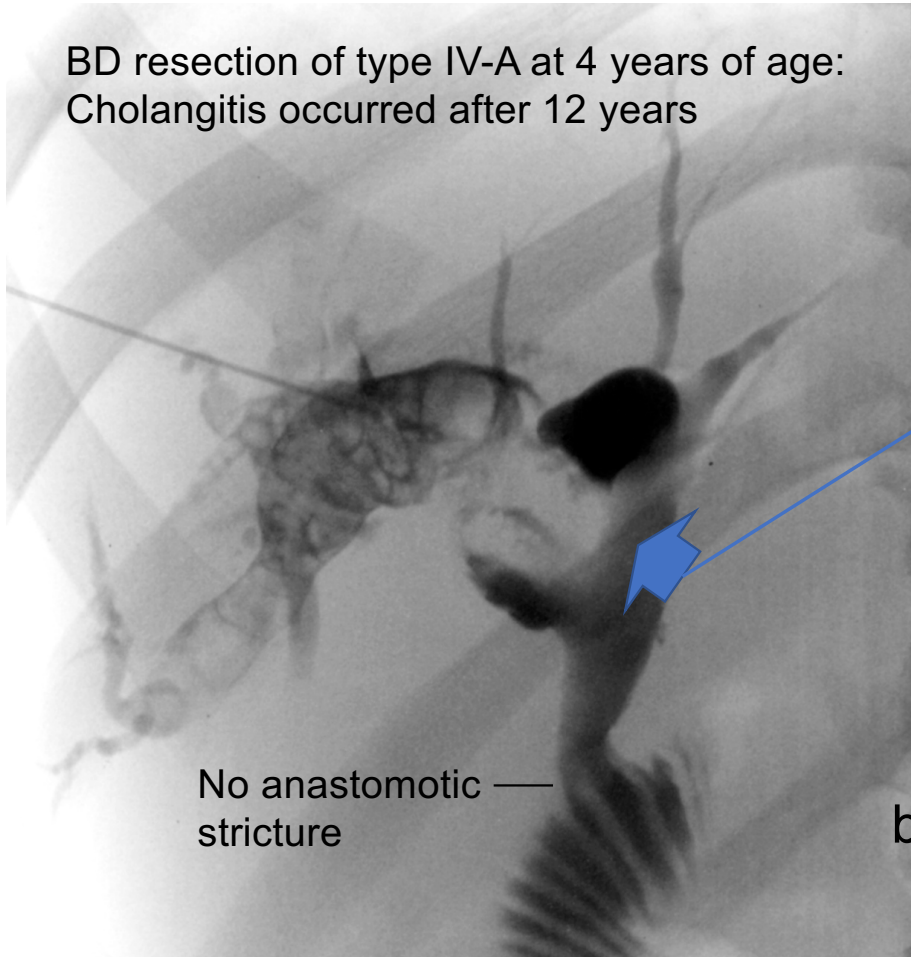


Only removal of stones may be insufficient for a cure; this needs to relieve stenosis or dilated BD (hepatectomy).

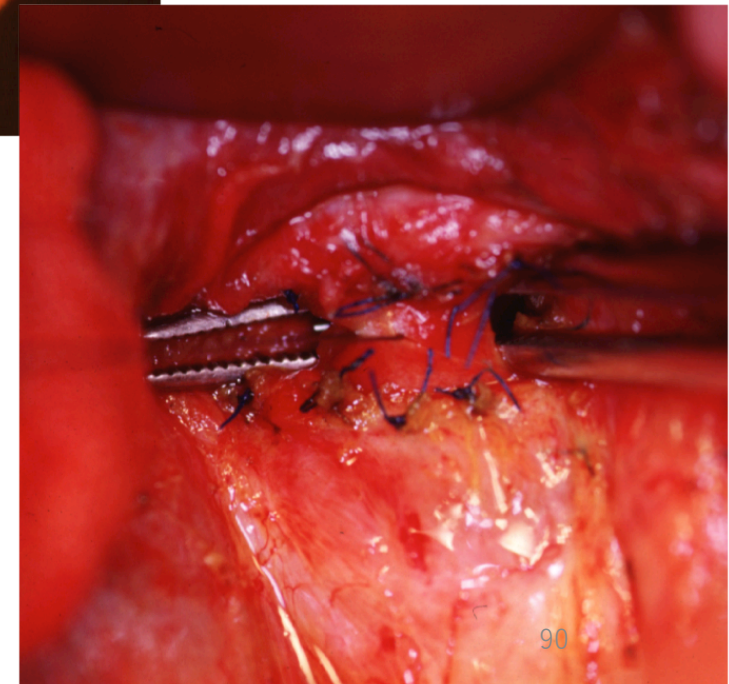
Case 24

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

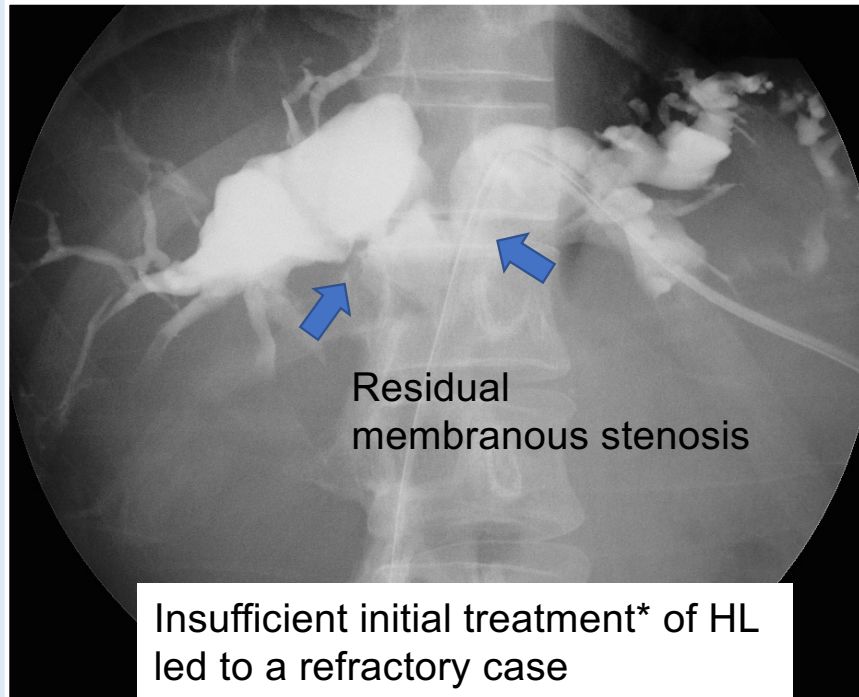


Refractory cholangitis will occur

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

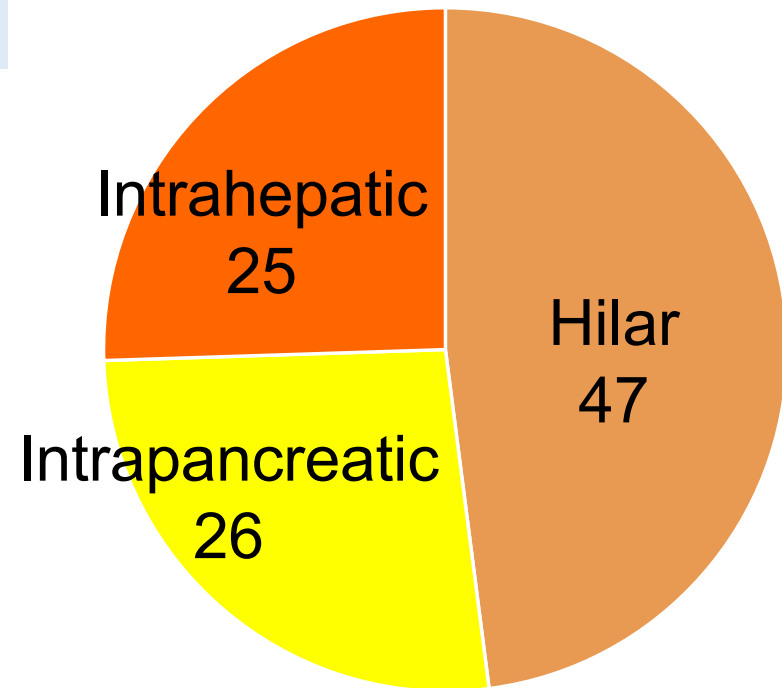
Kamisawa T, Kaneko K, Itoi T, Ando H. Pancreaticobiliary maljunction and congenital biliary dilatation. Lancet Gastroenterol Hepatol. 2017 Aug;2(8):610-618. doi: 10.1016/S2468-1253(17)30002-X. PMID: 28691687.

cf. Survey in USA

Biliary cancer after BD resection 13/394 3.3%

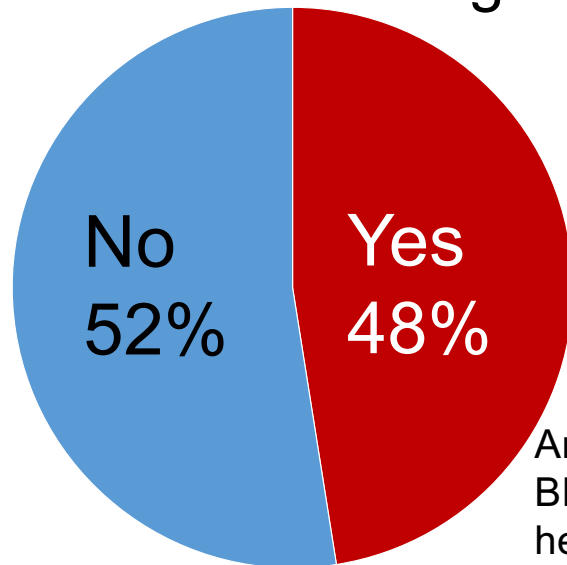
Children 4/135 3.0%, Adults 18/259 6.9%

Soares et al. JAMA Surg 2015



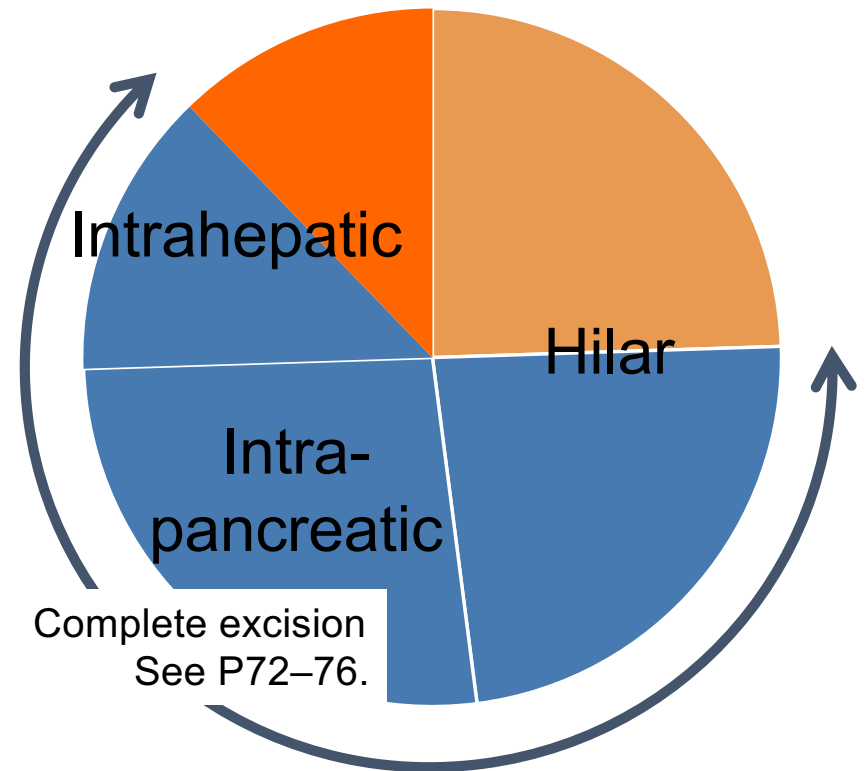
Many residual BD cancers could have been prevented

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

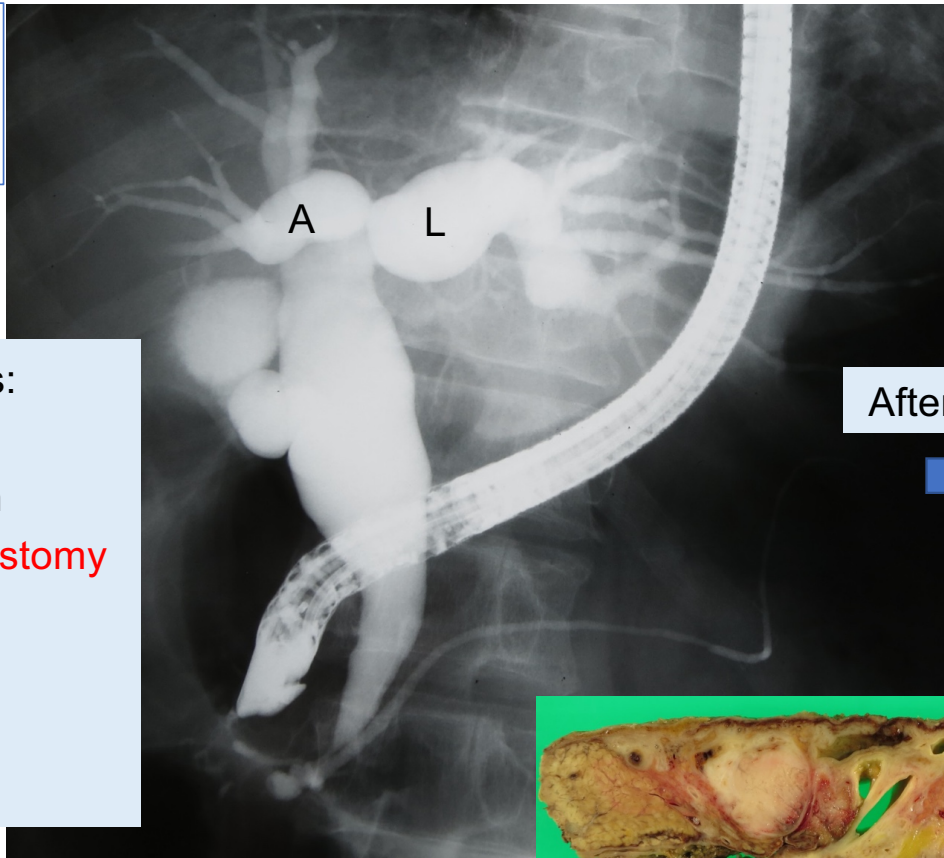
Cholangitis

↓ After 9 years

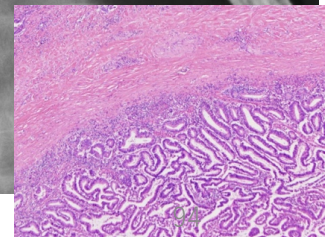
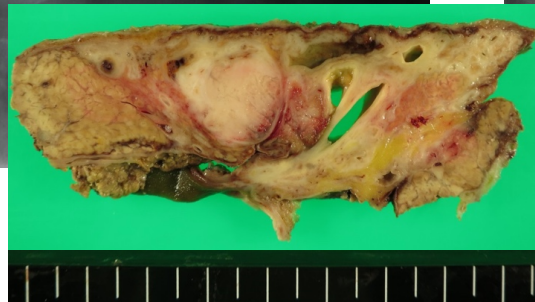
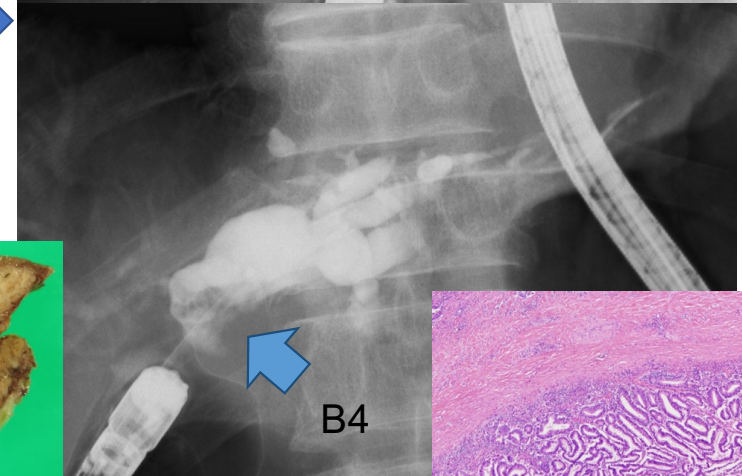
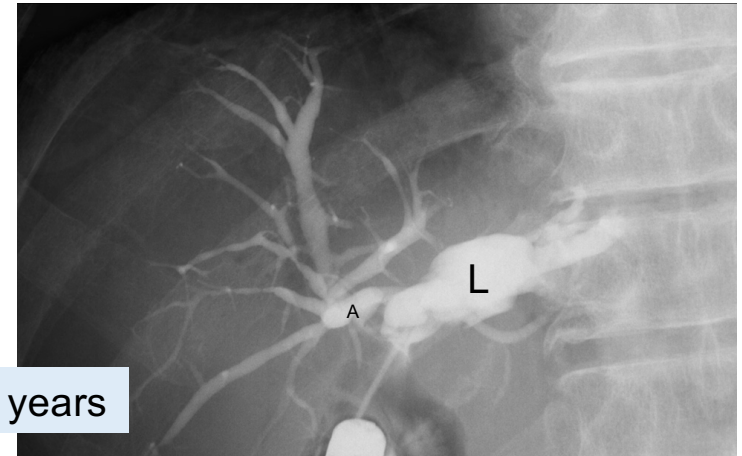
BD cancer (B4)

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

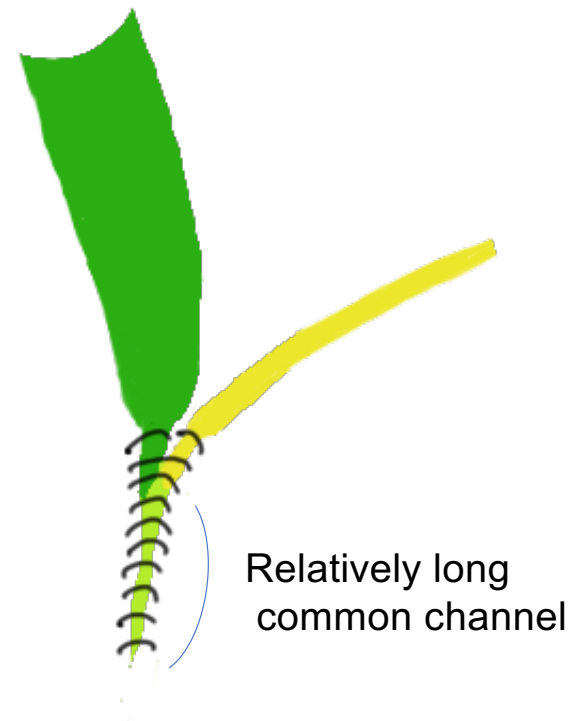
High confluence is the **transitional type between normal and PBM**, and frequently causes **pancreatobiliary reflux**. It is known that patients with high confluence often have gallbladder cancers as is seen in pancreaticobiliary maljunction. Sai et al. proposed a similar concept of **occult pancreatobiliary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

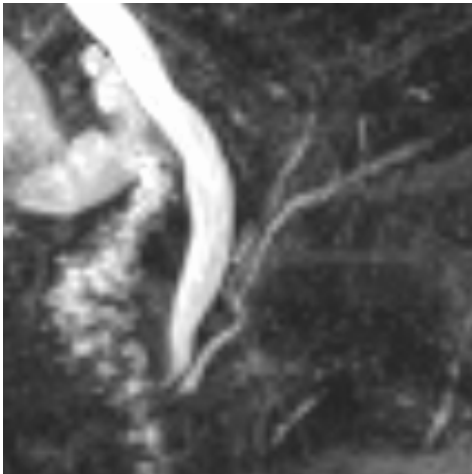
Normal

Occult pancreatobiliary reflux

High confluence

PBM

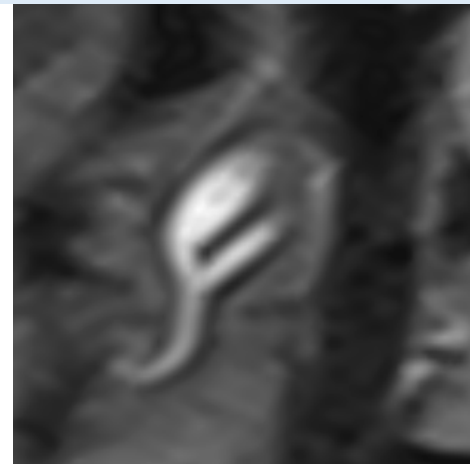
The sphincter function affecting the confluence



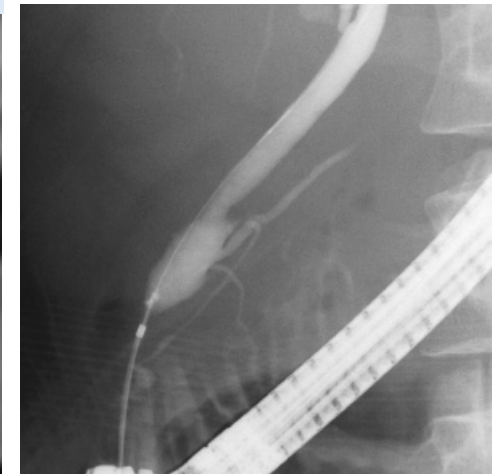
Separate openings



The common channel formed
Cholangiography occasionally
shows the PD.



Relatively long
common channel



Abnormally long
common channel