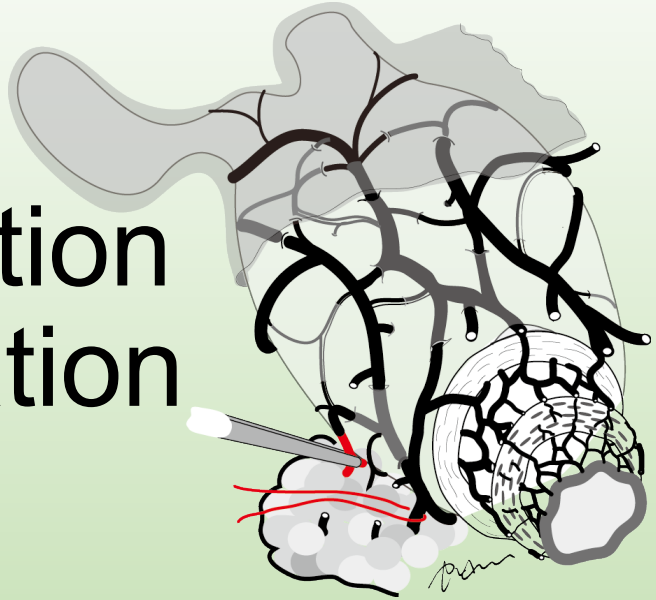
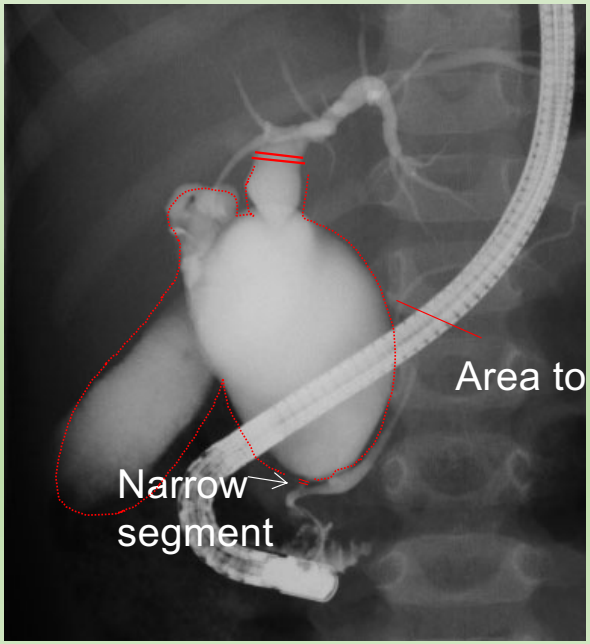
Atlas of Pancreaticobiliary Maljunction & Congenital Biliary Dilatation





Japanese Study Group on Pancreaticobiliary Maljunction (JSGPBM)
Scientific Committee

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Yoshiki Morotomi

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

Contents

Basics

- Pathophysiology, diagnosis & classification 19–39
  - Symptoms in pediatric patients 20
  - Protein plug theory 21–25
  - Biliary carcinogenesis 26–28
  - Diagnosis of PBM 29–34
  - Classification of PBM 35–39
- What is CBD? 40–47
- Typical CBD 48–57
- Nondilated type of PBM 58–68
- Treatment for PBM 69–84
- Postoperative complications 85–96
- High confluence of the pancreaticobiliary ducts 97–100

Part 2

Introduction

- What is PBM? 6–8
- What is CBD? 9–10
- Localized BD dilatation 11
- Embryogenesis 12
- PBM itself is asymptomatic 13
- Treatment of PBM 14
- Biliary carcinogenesis 15–16
- Surgery for CBD 17
- Recommended terms 18

Advanced

- Issues on Todani's classification 101–115
- Nondilated type of PBM 116–144
  - Definition 117–126
  - Pathophysiology and treatment in children 127–134
  - Pathophysiology and treatment in adults 135–144
- PBM seemingly complicated & rare type of PBM 145–149
- Common channel & narrow segment 150–159
- What causes biliary dilatation? 160–167
- Pathophysiology in pediatric patients 168–183
  - Mechanism of protein plug formation 168–178
  - Symptomatology besides protein plug 179–183

Expert

- Prenatal diagnosis, Transitional type to biliary atresia 184–195
- Rare congenital dilatation of biliary tract 196–202
  - Type Ib 198–199
  - Type II, diverticulum 200
  - Type III, choledochocele 201-202

Basics

Non-dilated type of PBM

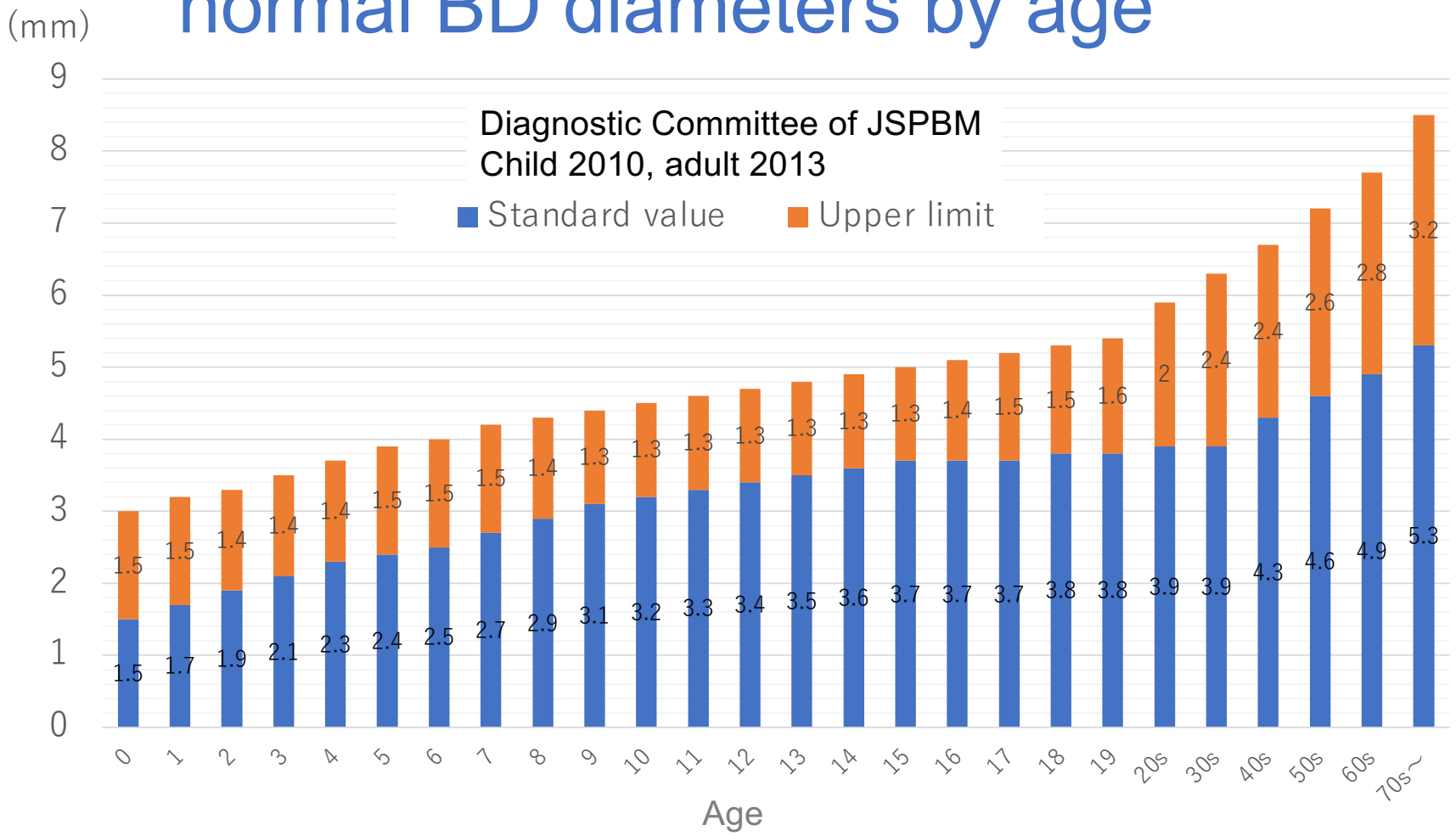
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What is non-dilated type of PBM?

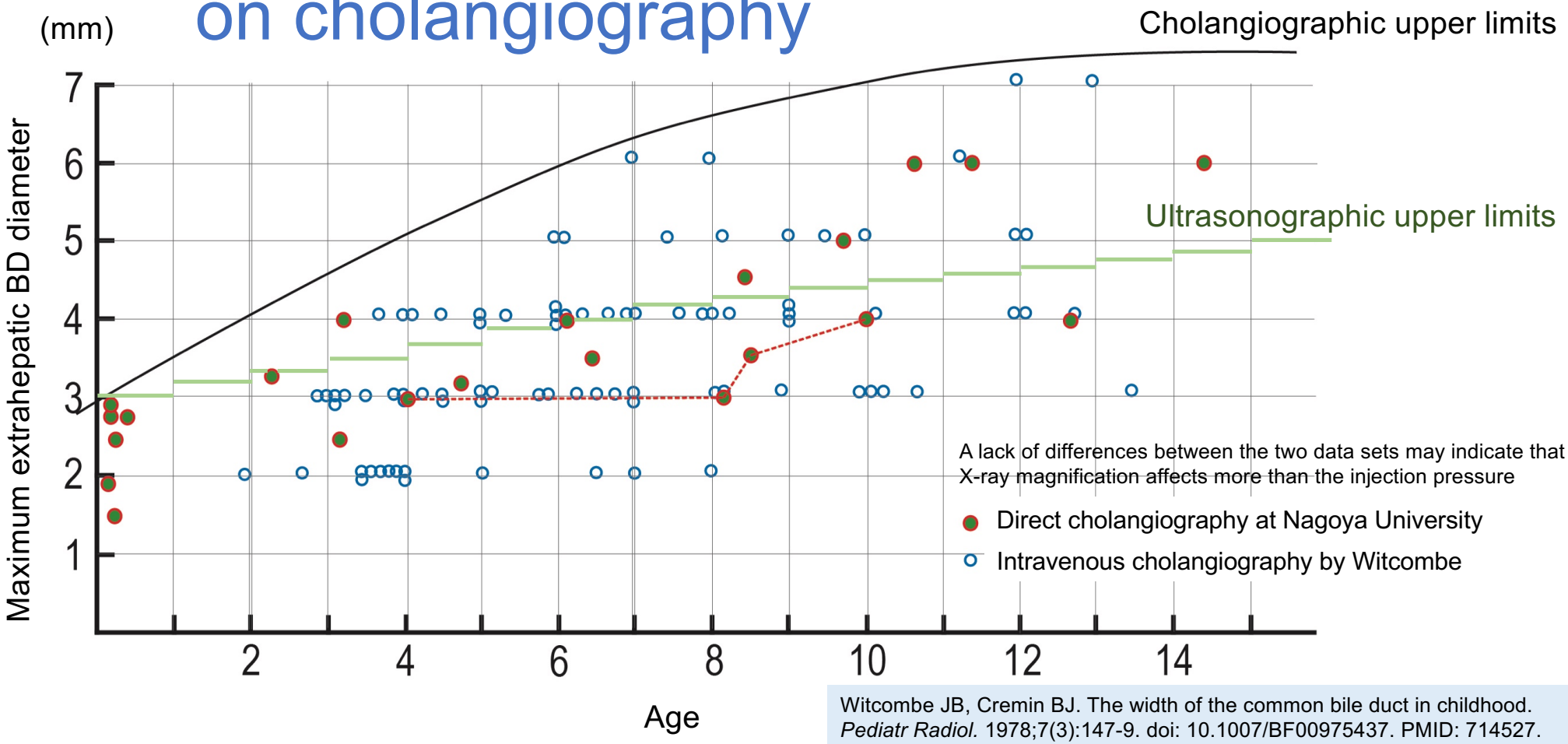
- Definition has not yet been determined.
- BD diameter **increases with age** in both children and adults. Ultrasonographic upper limits of normal BDs by age are determined.
- There is little data of normal BD diameters on direct cholangiography* or MRCP** at present.
- Non-dilated type should have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

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

Ultrasonographic upper limits of normal BD diameters by age



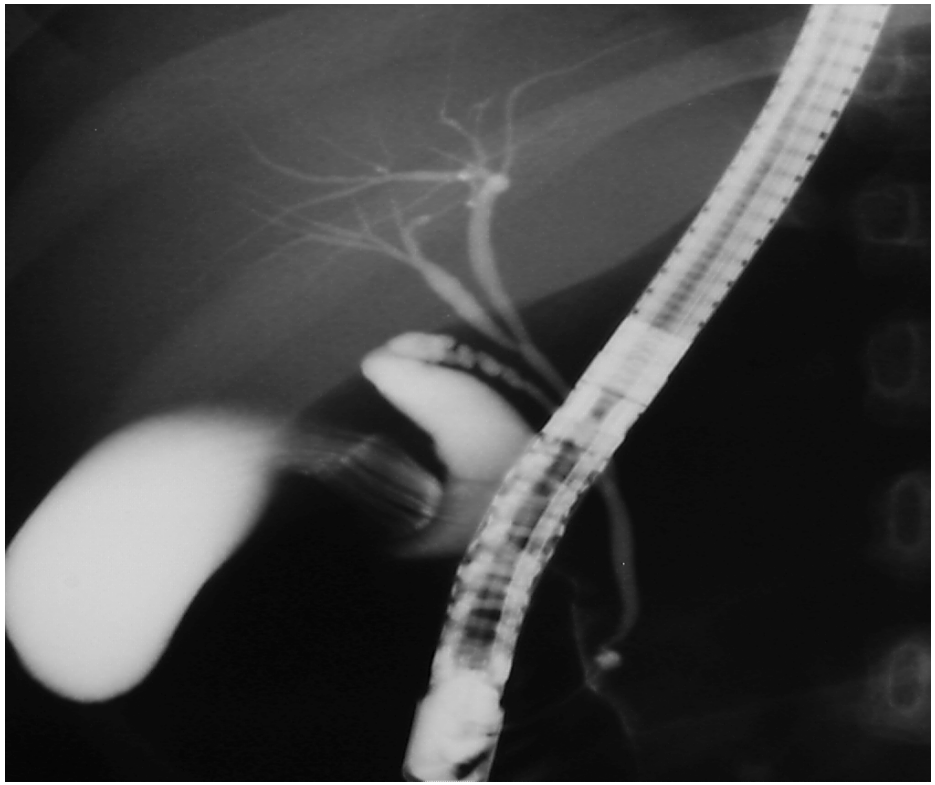
Normal pediatric BD diameters on cholangiography



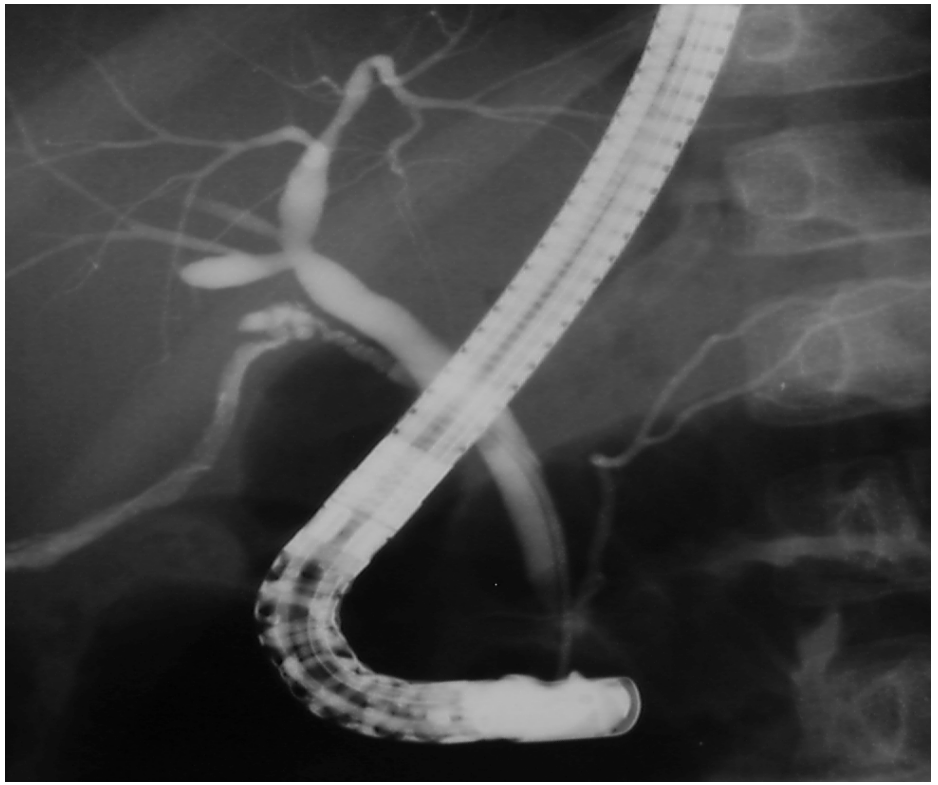
Witcombe JB, Cremin BJ. The width of the common bile duct in childhood. *Pediatr Radiol.* 1978;7(3):147-9. doi: 10.1007/BF00975437. PMID: 714527.

Normal pediatric BD

Appearance varies from thin to thick.



3-year-old girl



6-year-old girl

Case 18

Typical non-dilated type of PBM: Pediatric case

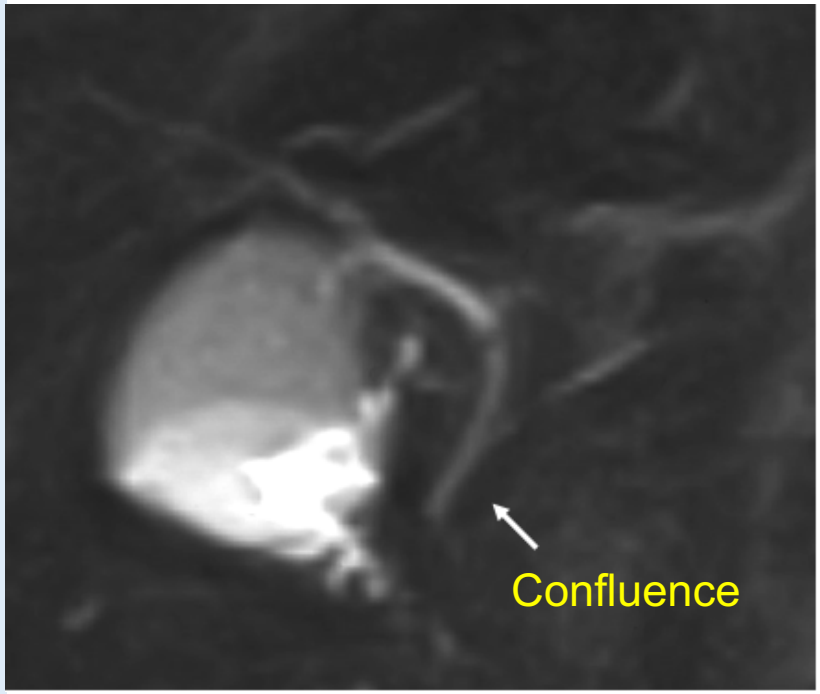
This case had no features of congenital biliary dilatation such as a dilated common channel. True non-dilated type, such as this case, is extremely rare in children.

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

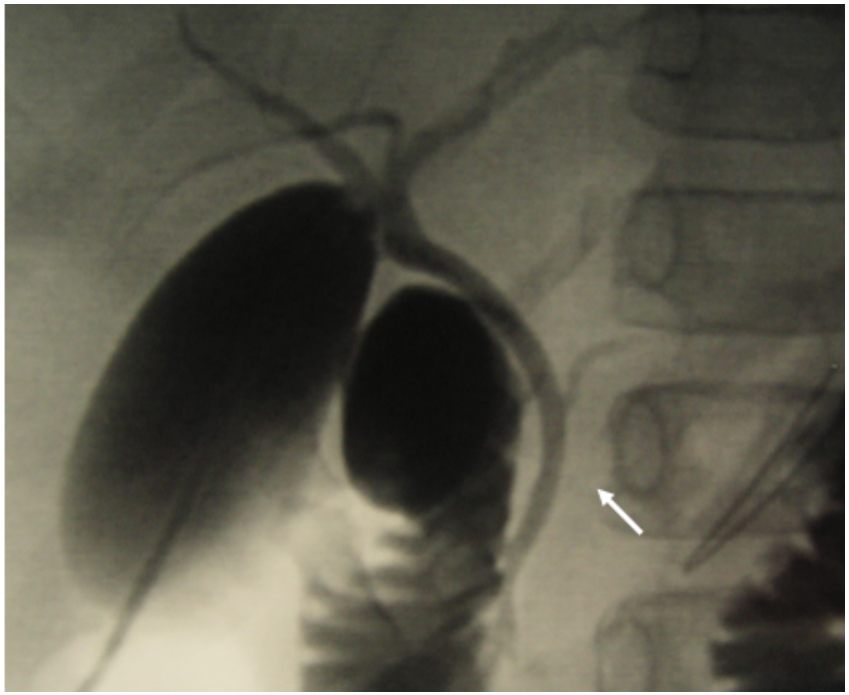
- **Abnormally long common channel = PBM**
  Type B, non-stenotic type
- **Diameter of CBD**
  4 mm on MRCP and cholangiography **within US limits**
- No features of congenital biliary dilatation except PBM

NS, dilated CC, dilated CD

See p41.



MRCP



On-table cholangiography

Case 19

Typical non-dilated type of PBM: Pediatric case

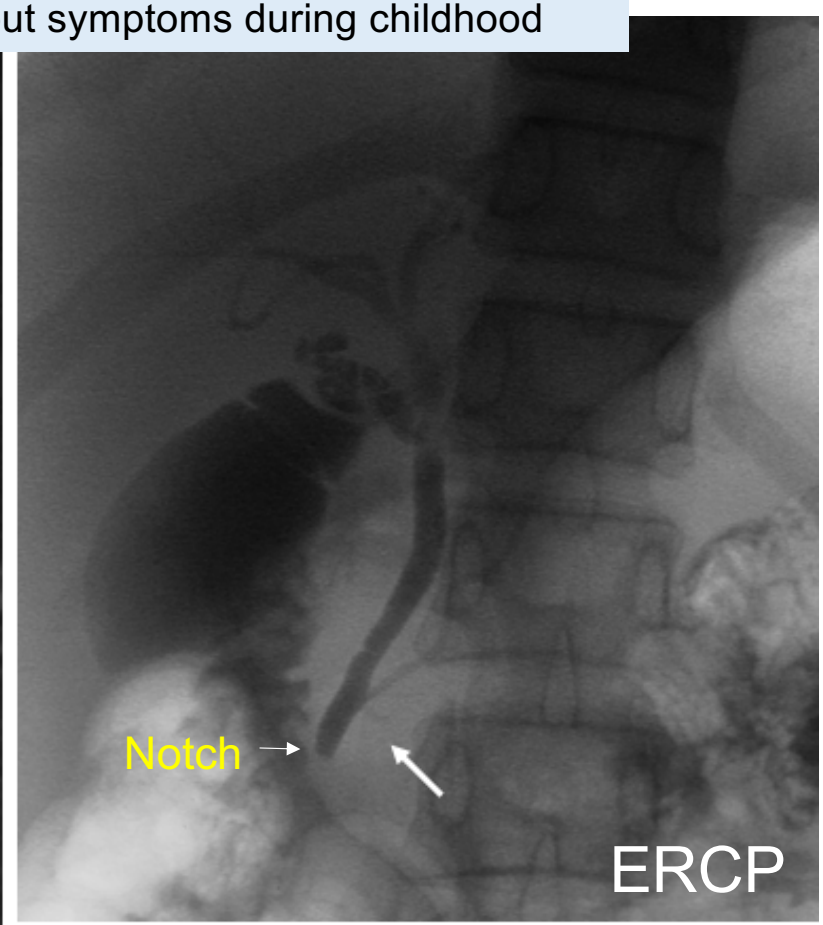
Non-dilated type has not been found without symptoms during childhood

Symptoms are the same as those of congenital biliary dilatation

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

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





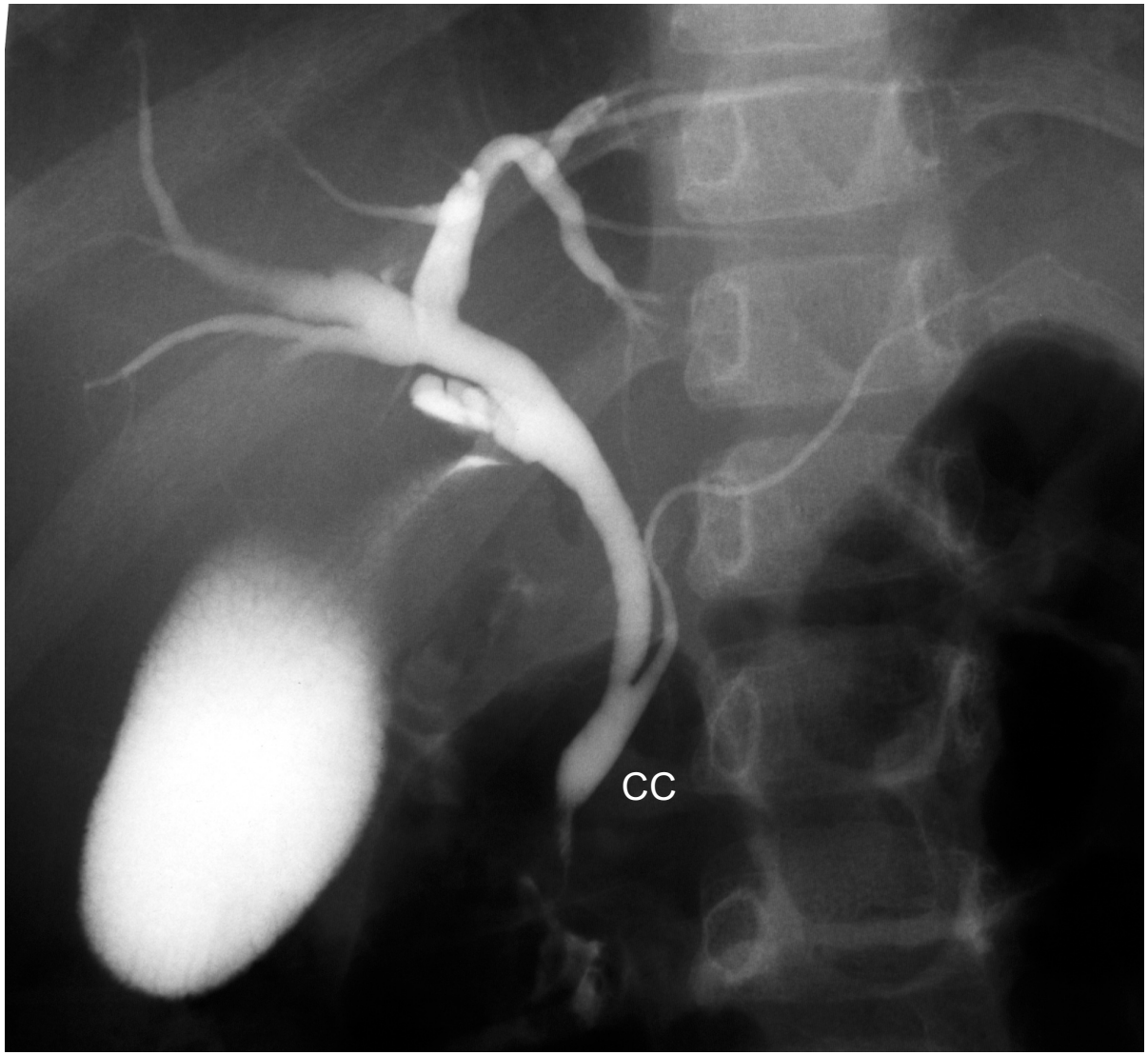
Case 20

Non-dilated type?

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

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

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



Case 21

Typical non-dilated type of PBM: Adult case

Gallbladder cancer often arises in adult non-dilated type

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

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

Non-dilated type is often found without symptoms in adults



From *Introduction Section*, see p31.

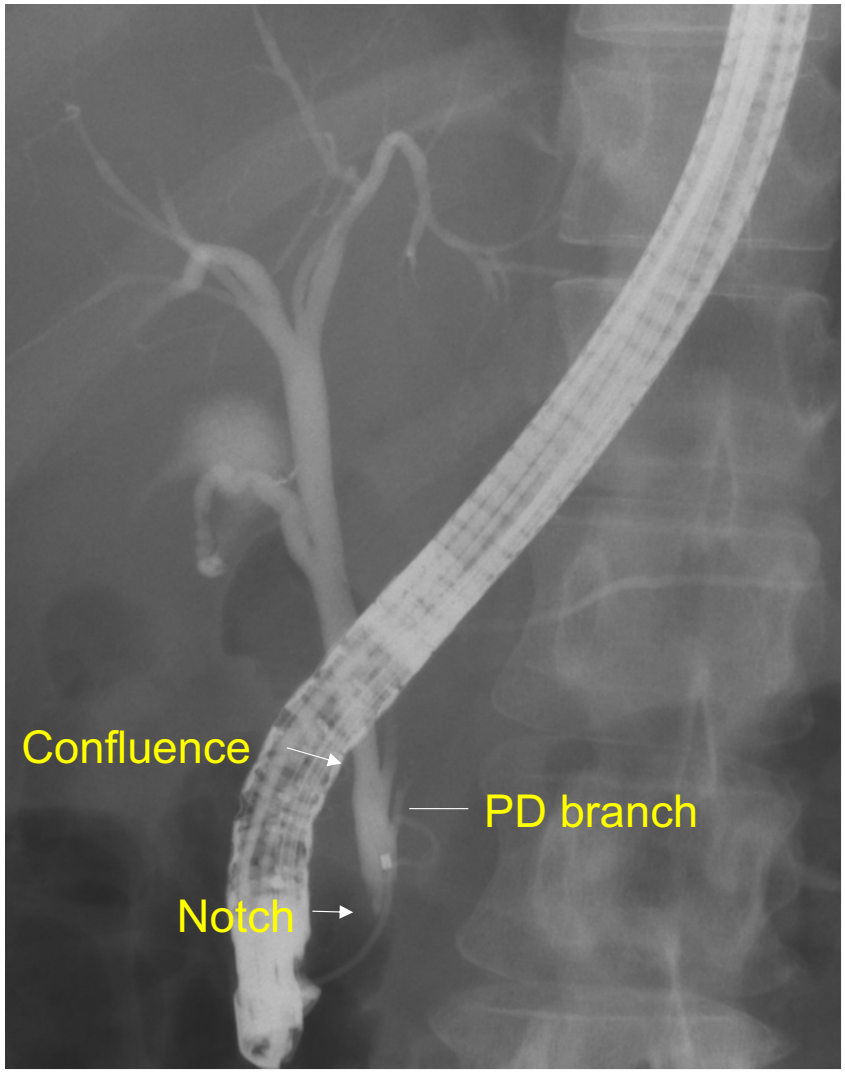
Case 6 Typical non-dilated PBM: Adult case

ERCP in a female in her 40s
Right hypochondrial pain

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

US	4 mm
MRCP	4 mm
ERCP	4 mm

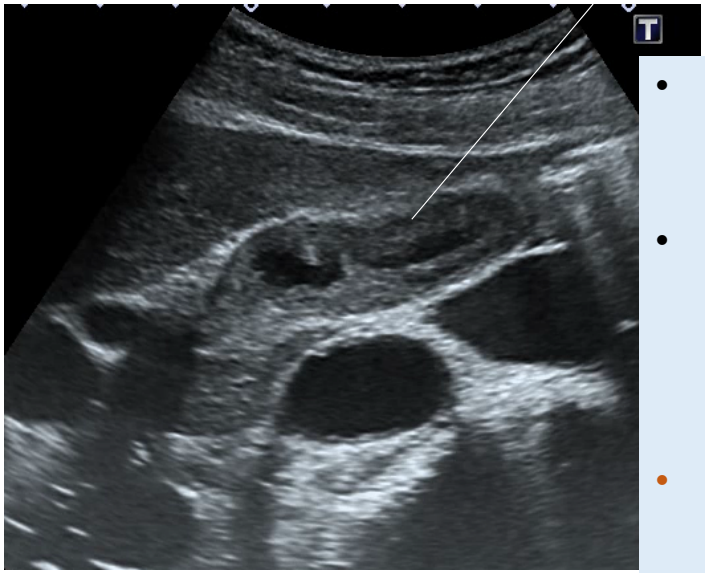
- **No features of congenital biliary dilatation** but for PBM
- A PD branch from the common channel
  The common channel is composed of PD?
  See *Advanced Section*.



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

A female in her 50s

Screening US found GB wall thickness

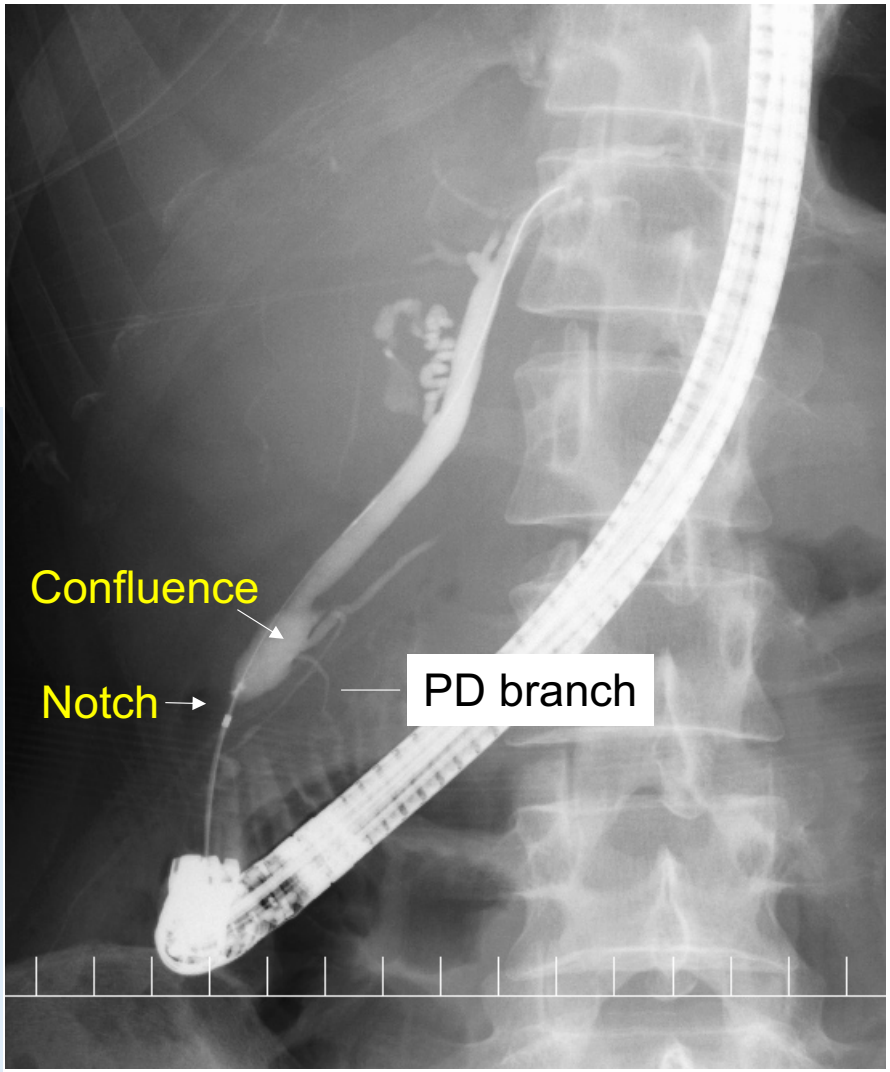


See *Advanced Section*. ←

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

US	4.2 mm
MRCP	6.2 mm
ERCP	5.8 mm

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



Basics

Treatment for PBM

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2nd version, September 2023

Treatment for pancreaticobiliary maljunction

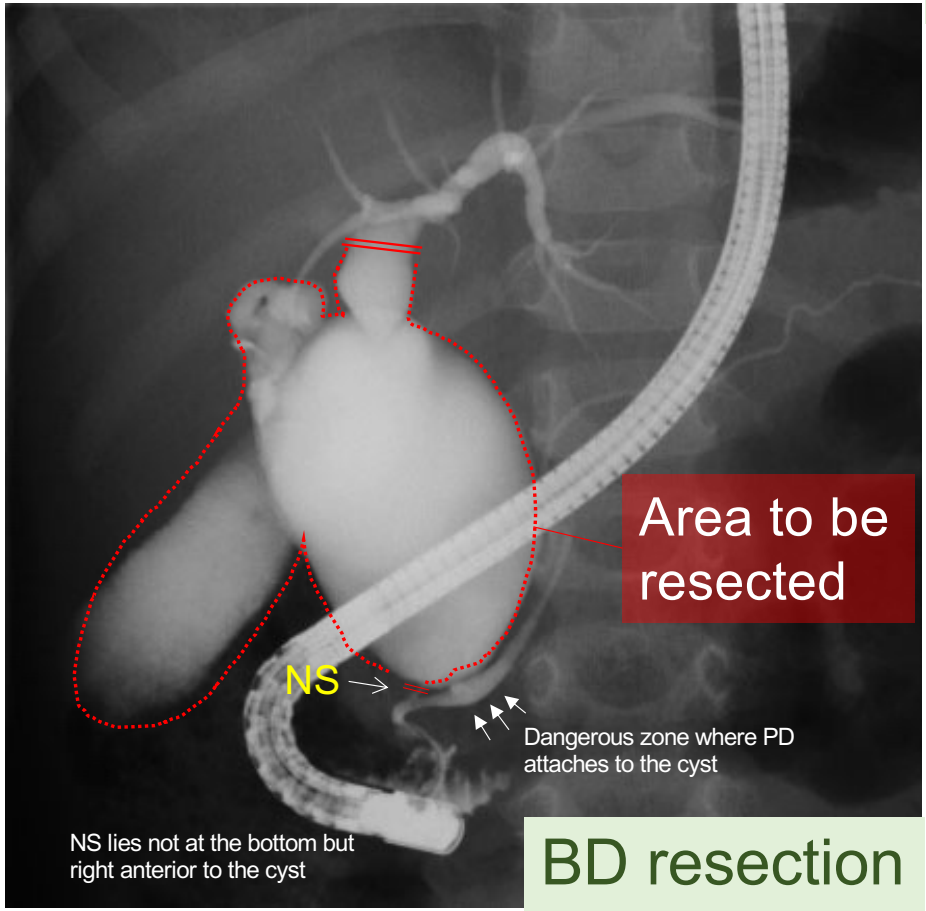
- Rationale for treatment:
  - ✓Not merely to correct malformation!
  - ✓Treatment is indicated to prevent complications.
- Complications of PBM:
  - ✓**Pediatric cases**: Recurrent **symptoms** including abdominal pain, vomiting, and jaundice.
  - ✓**Adult cases**: High incidence of **biliary tract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

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

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

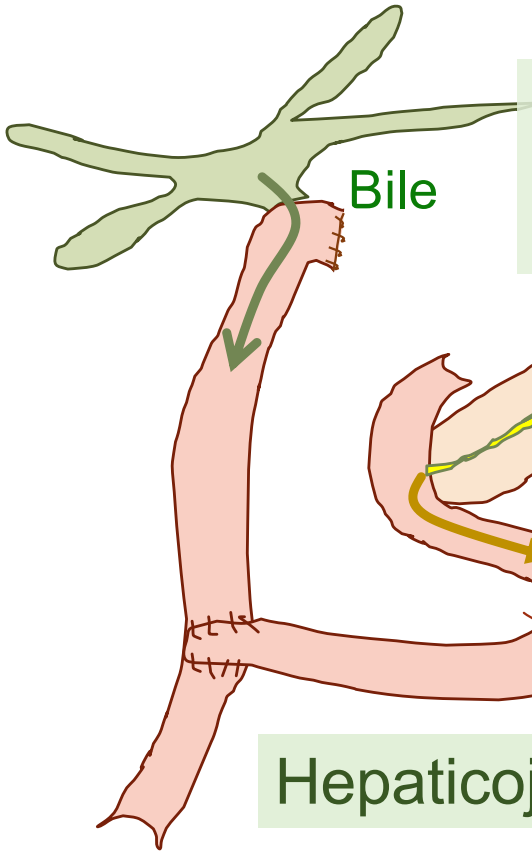
Surgery for CBD

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



BD resection

NS, narrow segment



Bile and pancreatic juice flows are separated

Hepaticojejunostomy

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

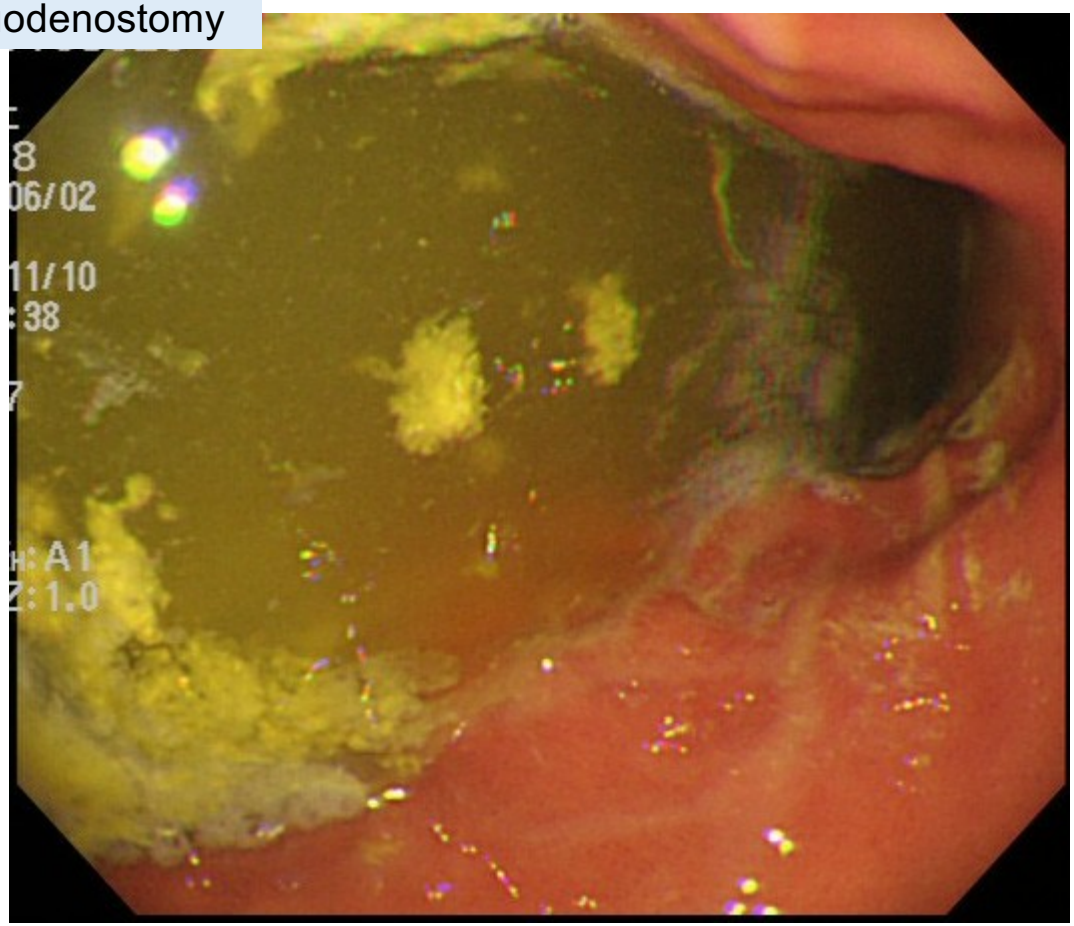
Bile gastritis after hepaticoduodenostomy

One of the reasons not to recommend hepaticoduodenostomy

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

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

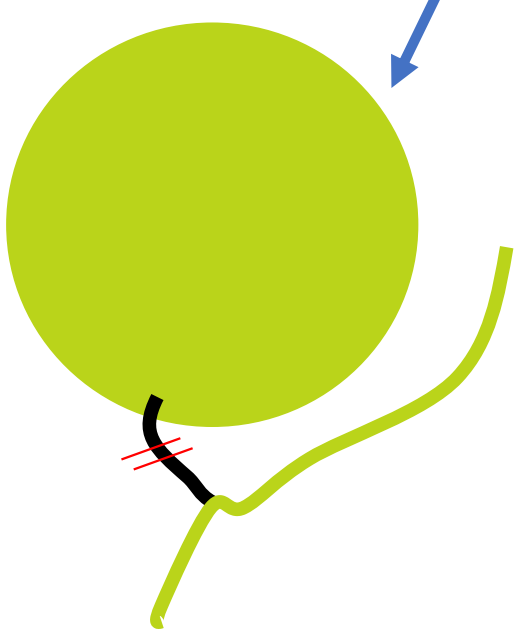
Three months after the examination, she underwent conversion to hepaticojejunostomy, which completely relieved her epigastric pain and endoscopic abnormality.



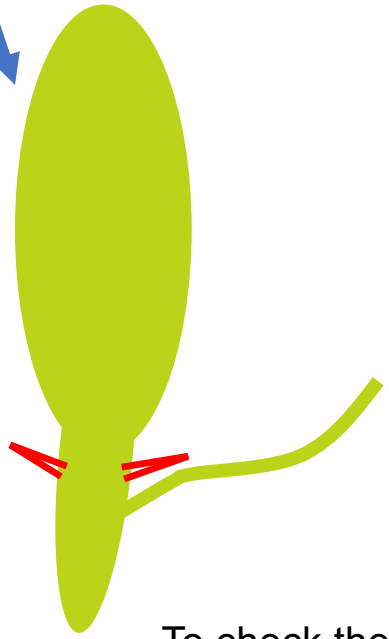
Complete resection of intrapancreatic BD

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

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



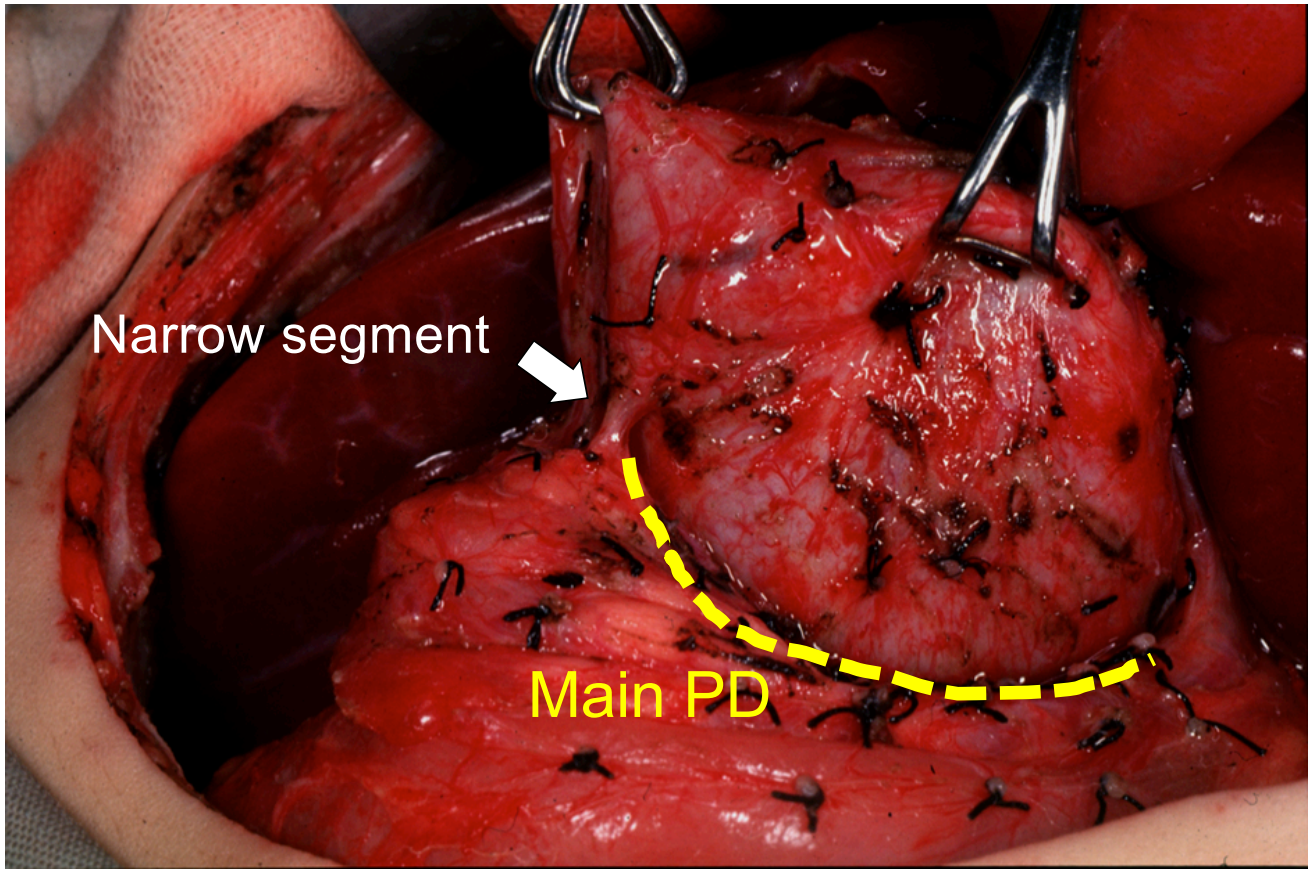
Visual confirmation of narrow segment



To check the distance to PD

Fluoroscopy using clips

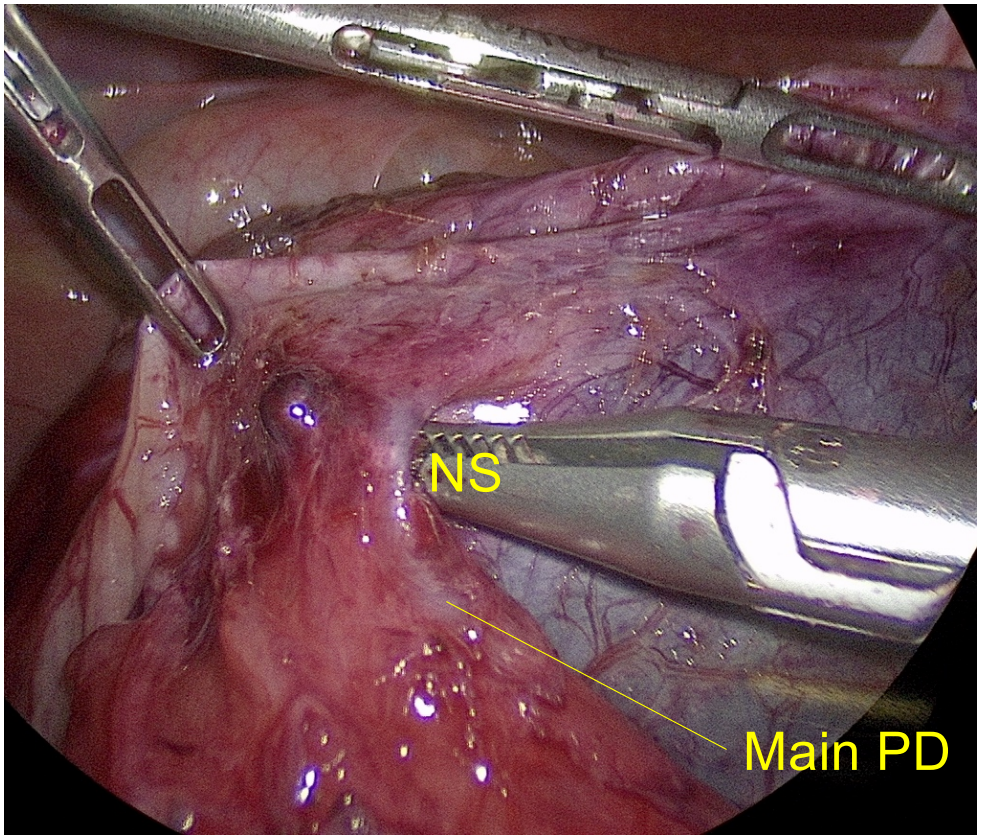
Visual confirmation of narrow segment

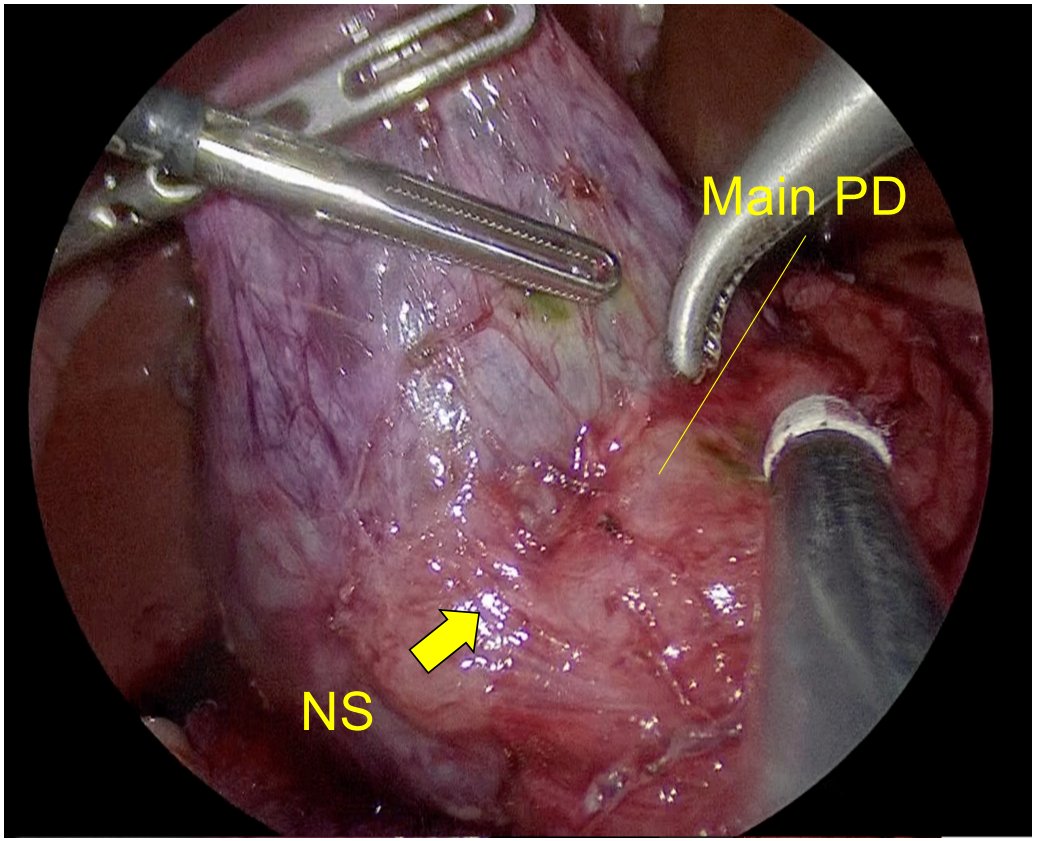


The narrow segment is often located not at the bottom but on the right-ventral side of the cyst.
In this case, care must be taken not to injure the PD because it runs over the cyst surface.

Main PD adheres to the cystically dilated BD

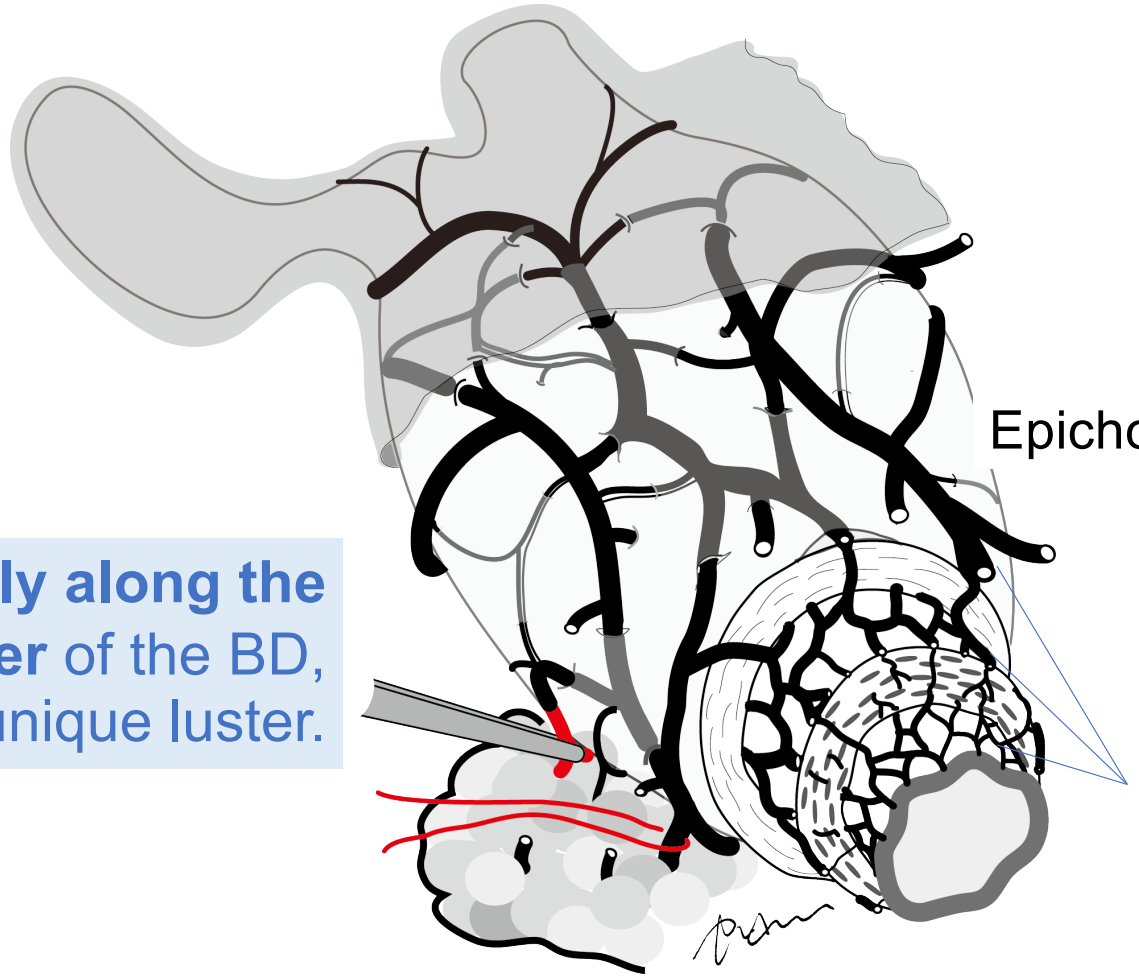
Infant case





How to avoid pancreatic duct injury

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



Epicholedochal plexus

Multilayer vascular plexuses

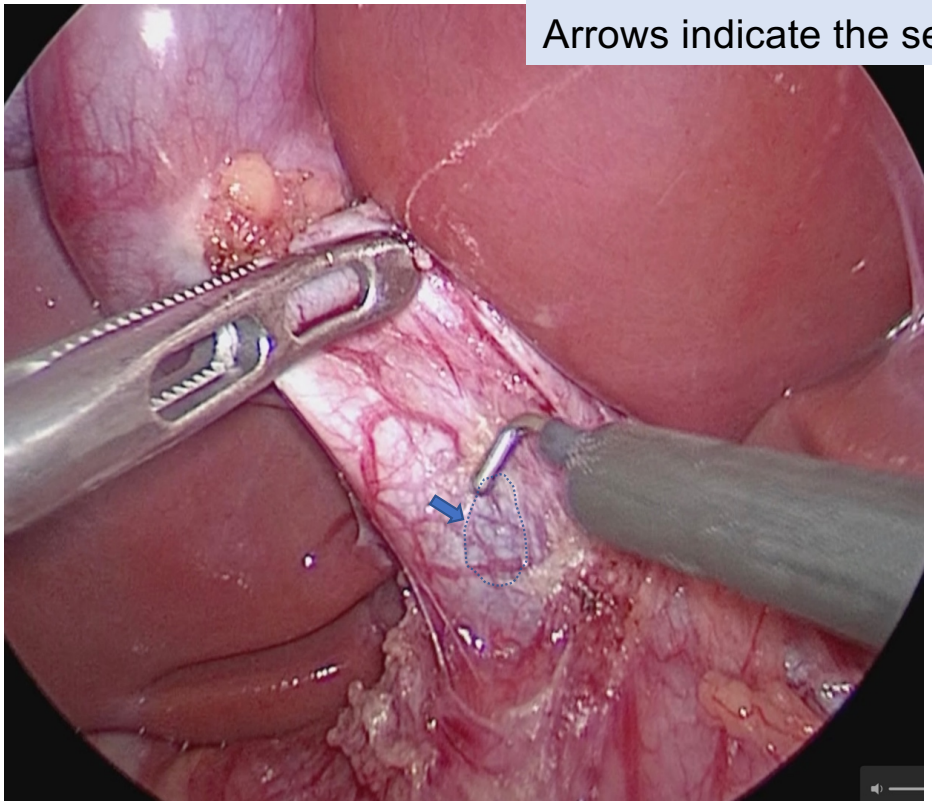
→ Cutting into the BD wall causes bleeding that obscures the layer.

BD serosa has a unique luster

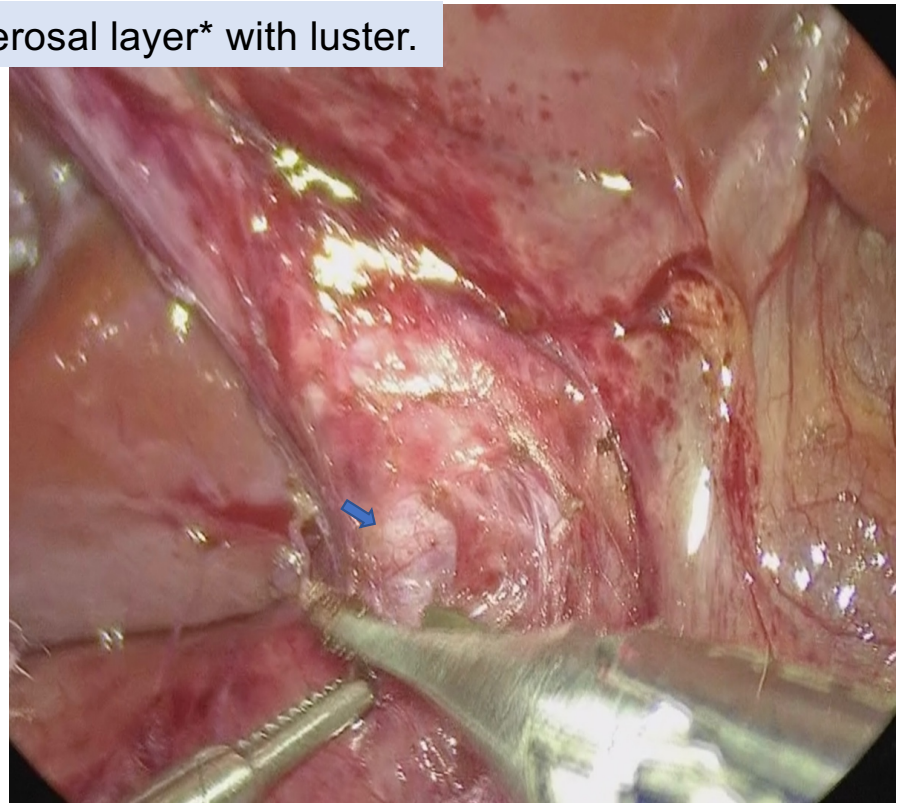
*To be precise, this layer is not the serosa but corresponds to the inner subserosal (ss inner) layer of the gallbladder.

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

Arrows indicate the serosal layer* with luster.



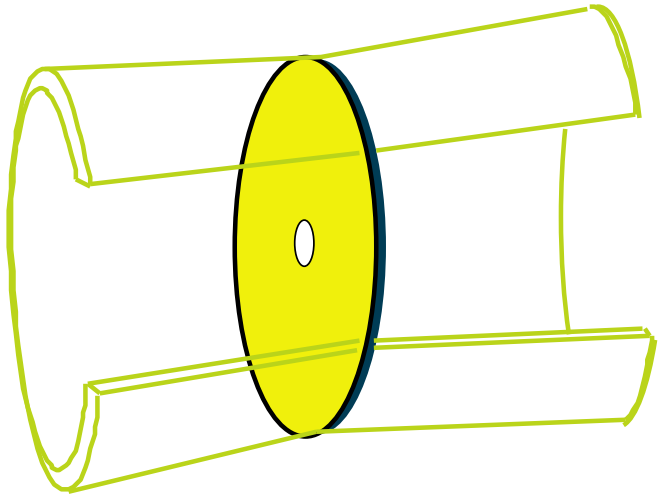
Case without inflammatory adhesion



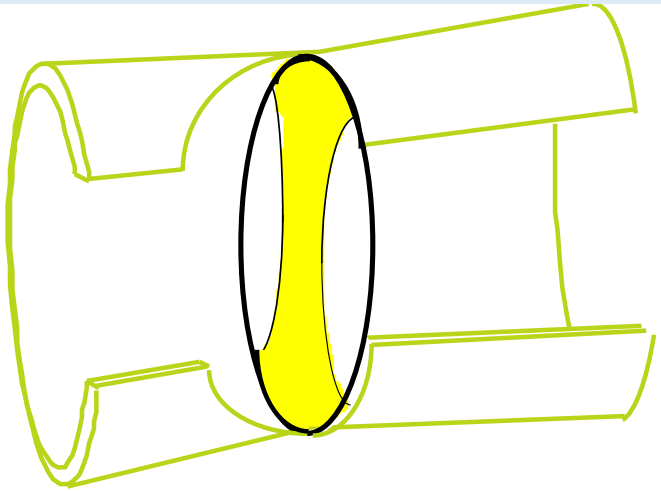
Case with inflammatory adhesion

Two types of congenital BD stenosis around the hepatic hilum

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



Membranous stenosis

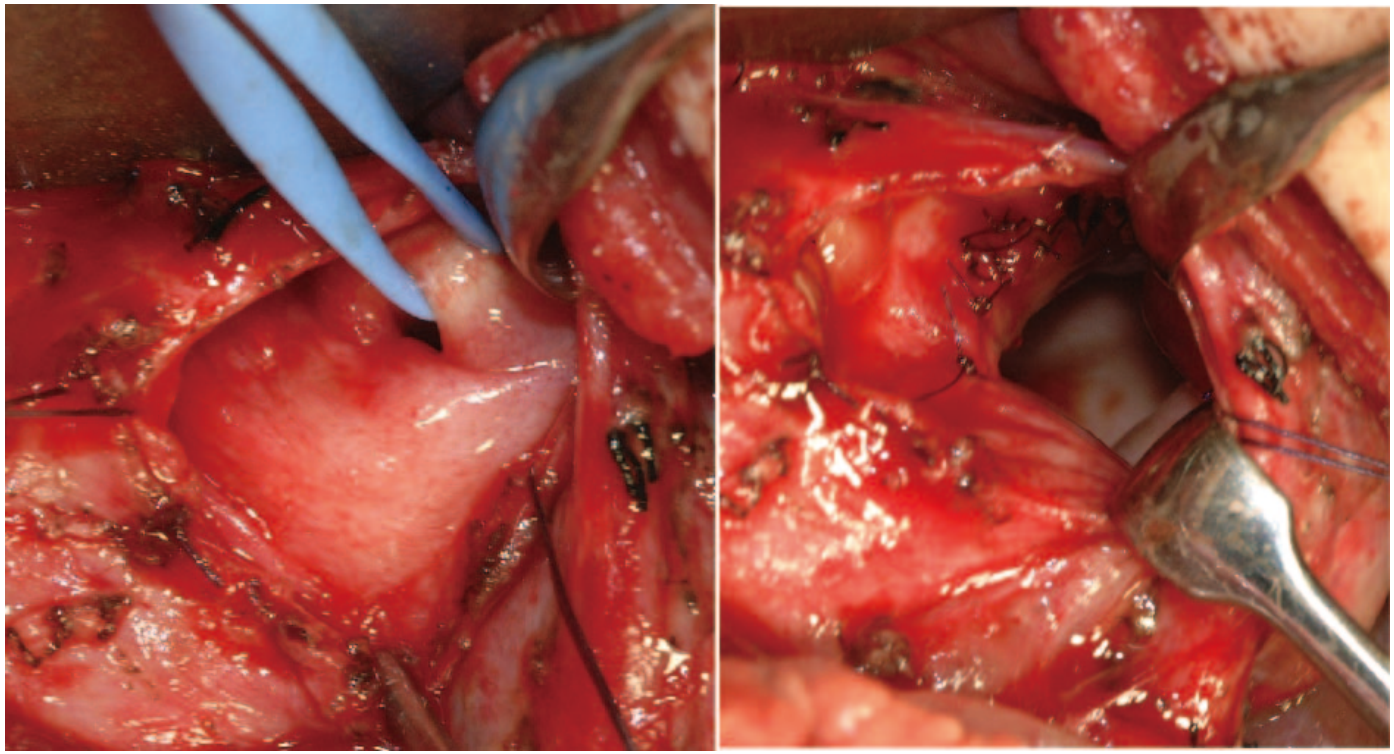


Septal stenosis

Balloon dilatation is ineffective for these stenoses.

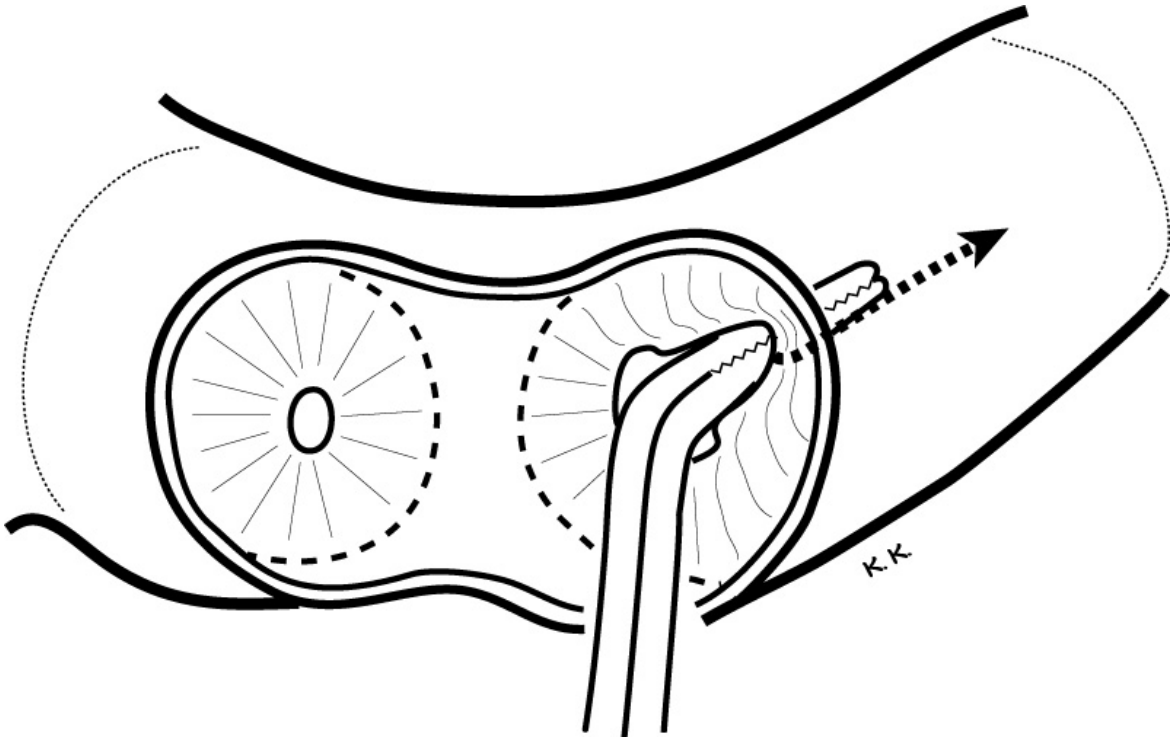
Ando H, Ito T, Kaneko K, Seo T.
Congenital stenosis of the intrahepatic bile duct associated with choledochal cysts. J Am Coll Surg. 1995 Nov;181(5):426-30. PMID: 7582210.

Resection of septal stenosis



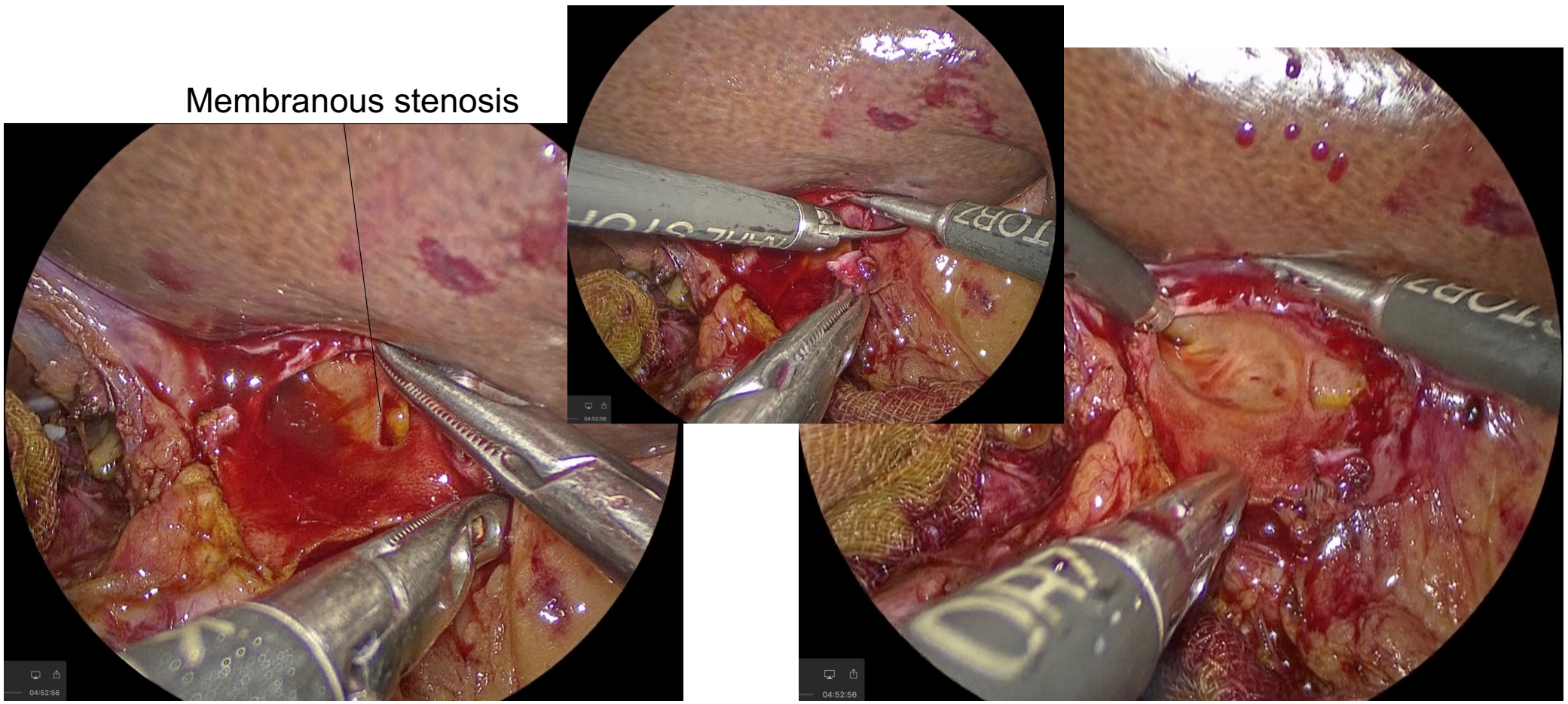
During BD resection

Todani and Lilly's BD plasty for membranous stenosis



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

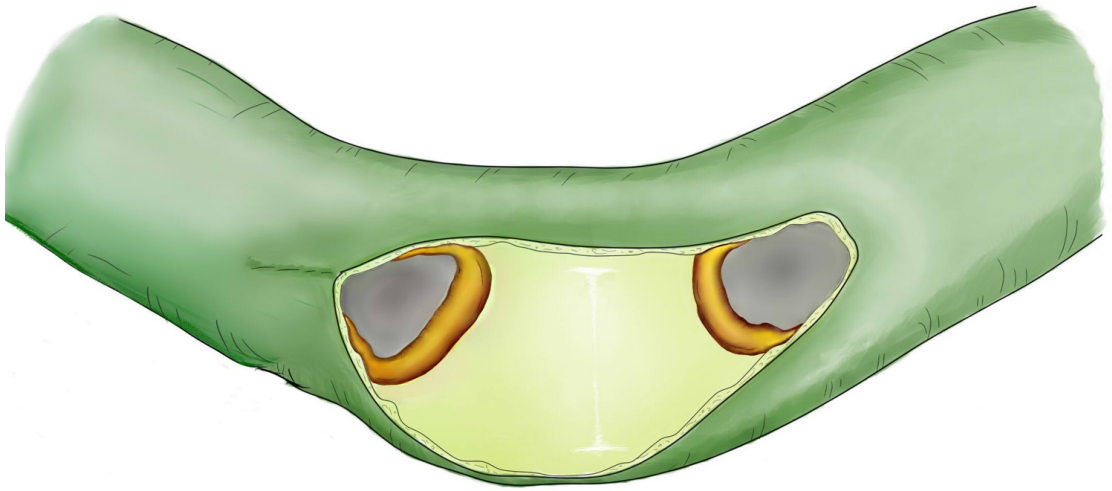
Todani's BD plasty



Hepaticojejunostomy with a wide stoma

Especially important for type IV-A cases





Hepaticojejunostomy in a case with membranous stenoses

This procedure prevents anastomotic stenosis and also relieves congenital stenosis.

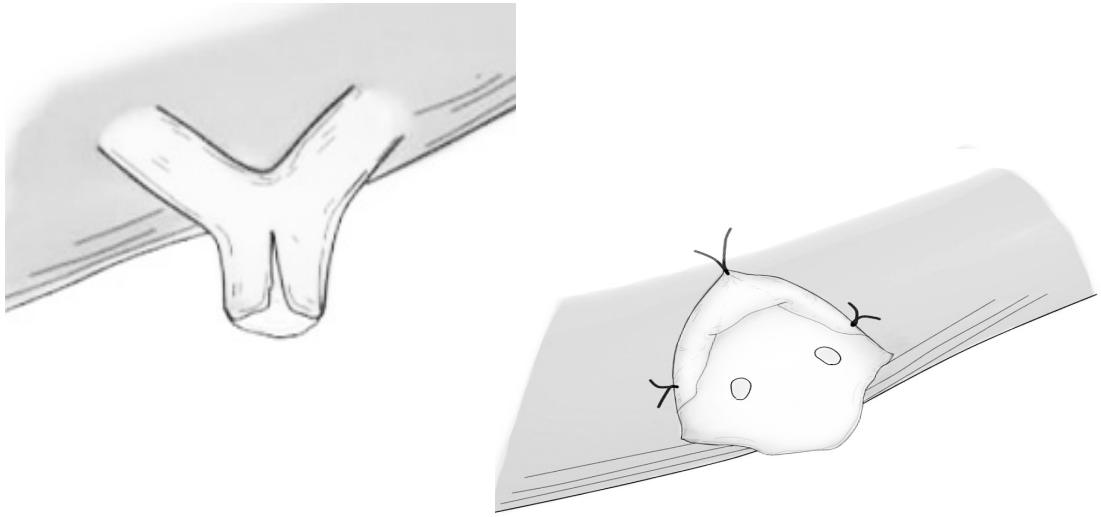
Procedures to prevent anastomotic stenosis in cases with small-caliber hepatic ducts

Open surgery



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

Laparoscopic surgery



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

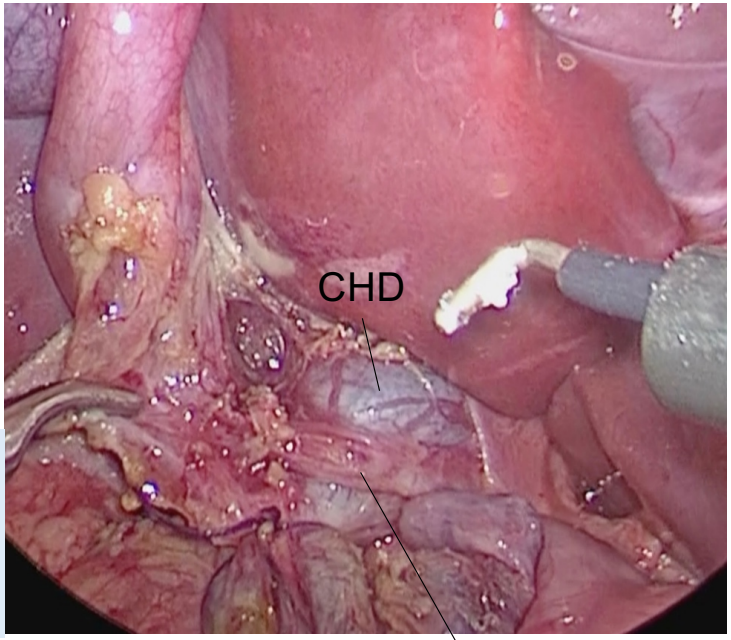
Urushihara et al. 2015

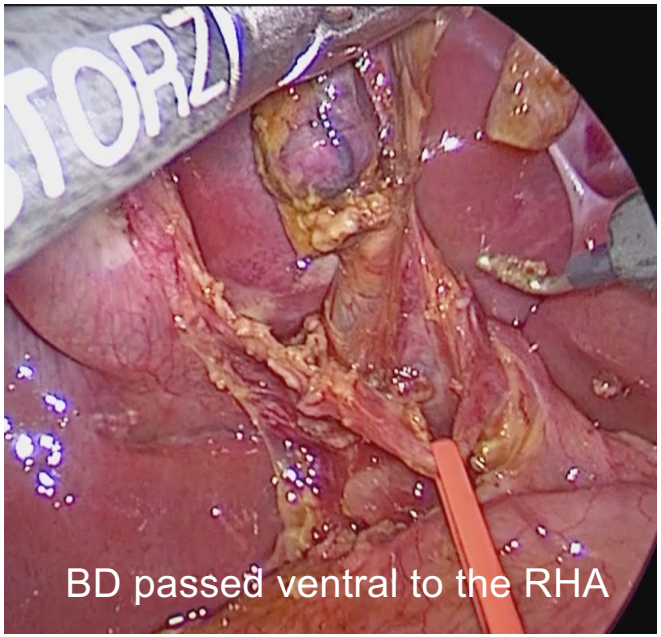
Excessive dissection impairs blood flow, resulting in anastomotic stricture.

Right hepatic artery runs ventral to BD

Case 23

A 4-year-old boy
Type Ia CBD





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

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

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

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

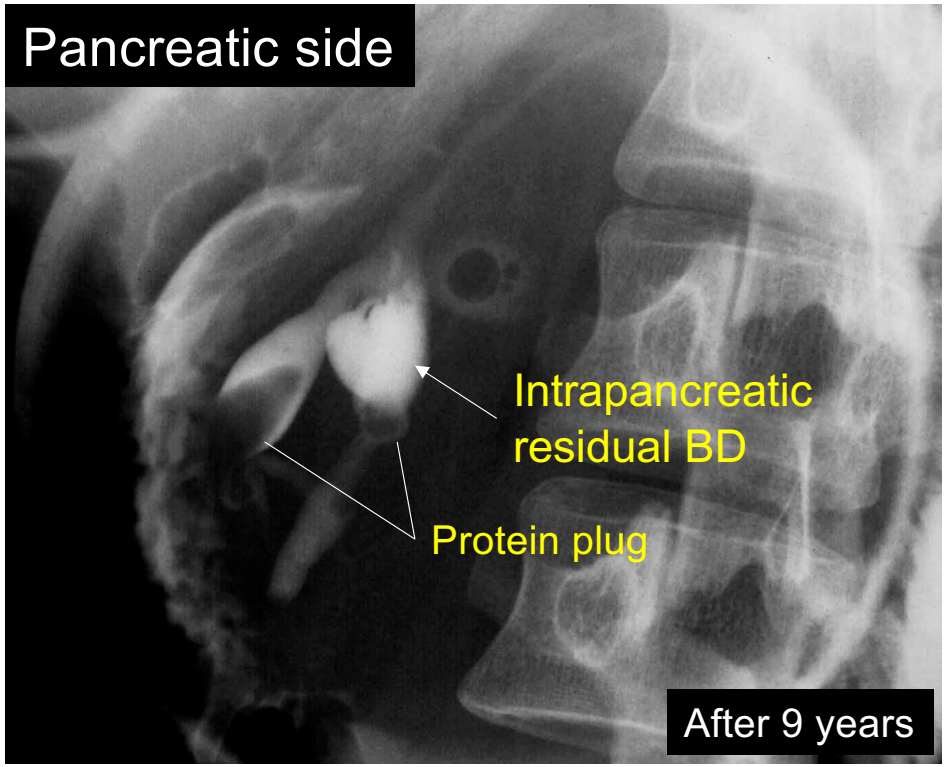
3) Guo WL, Wang J. Preoperative one-stop magnetic resonance imaging evaluation of the pancreaticobiliary junction and hepatic arteries in children with pancreaticobiliary maljunction: a prospective cohort study. *Surg Today*. 2021 Jan;51(1):79-85. doi: 10.1007/s00595-020-02077-5. Epub 2020 Jul 12. PMID: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

Basics

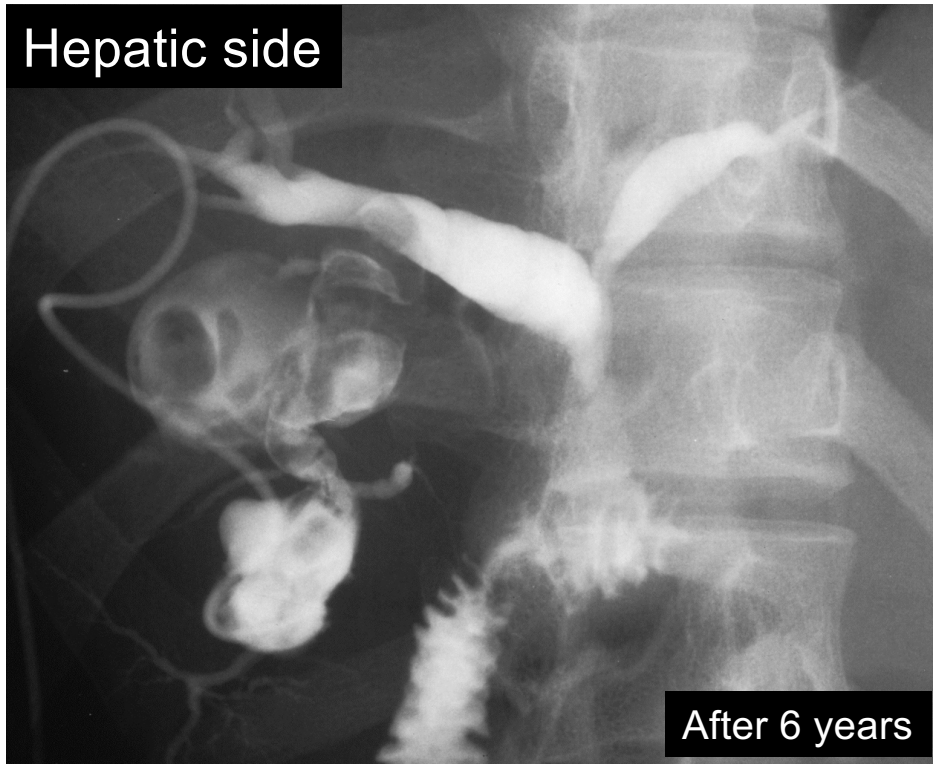
Postoperative complications

First version, September 2022; Revised, September 2026

Long-term complications after BD resection



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



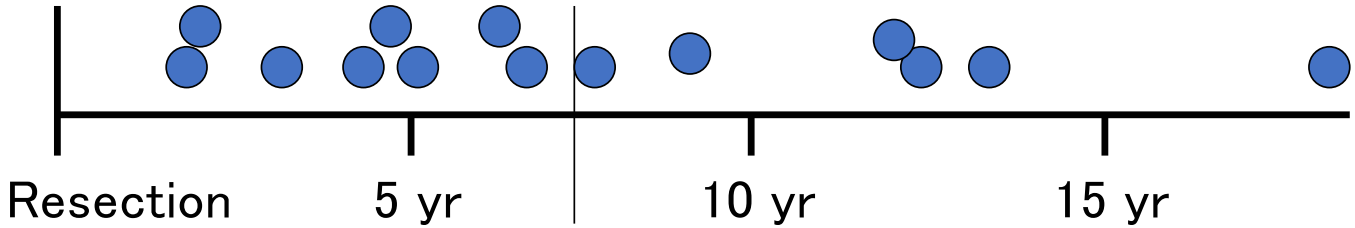
Intrahepatic gallstones and/or cholangitis occur in 7% to 8% of cases several years after BD resection.

Outcomes of residual intrapancreatic bile duct

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

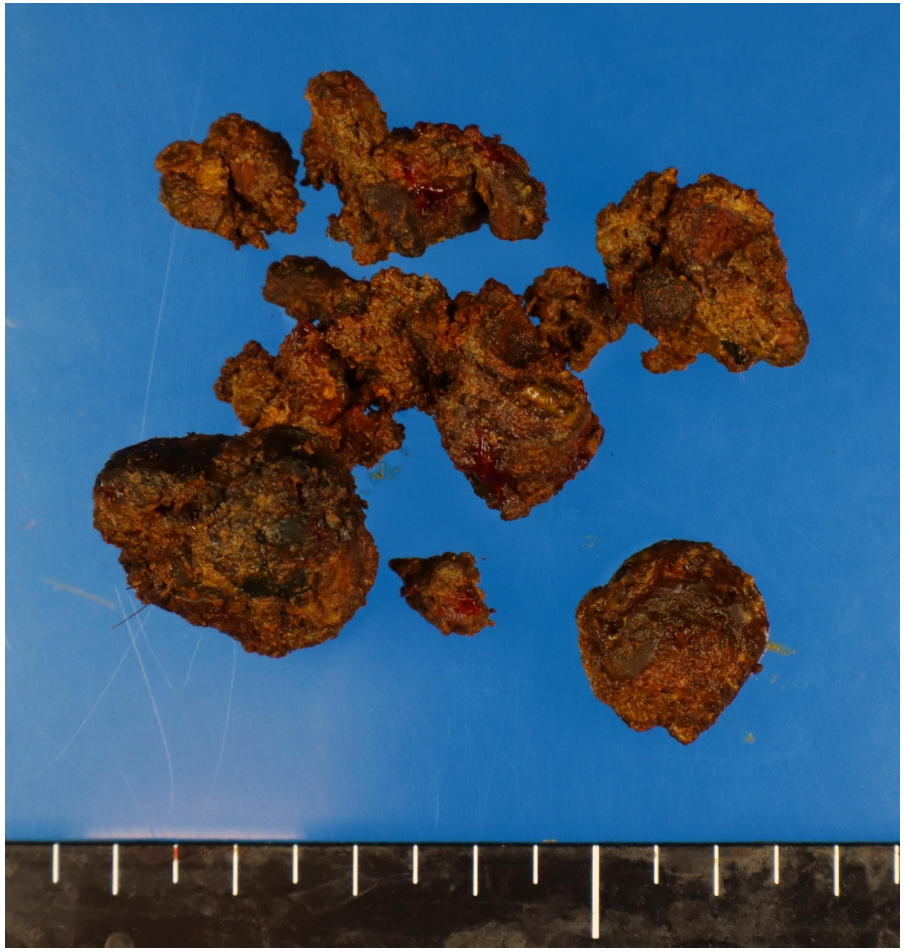
Time from resection to intrahepatic gallstone formation

Pediatric cases (n=14)



Mean: 7.3 yr

Postoperative gallstones are brown pigment stones

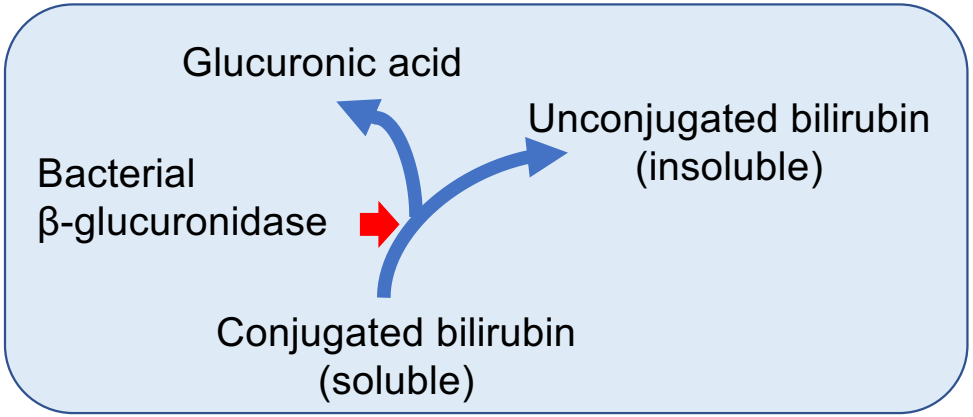


Composition:

Bilirubin calcium + calcium fatty acid

Bacterial β-glucuronidase and phospholipase A1

Bile stasis and bacterial infection



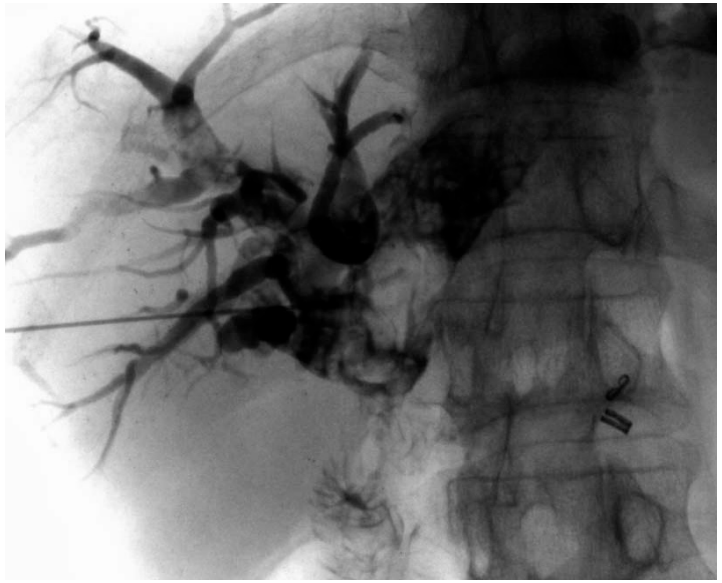
Risk factors for postoperative hepatolithiasis

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

Todani type IV-A



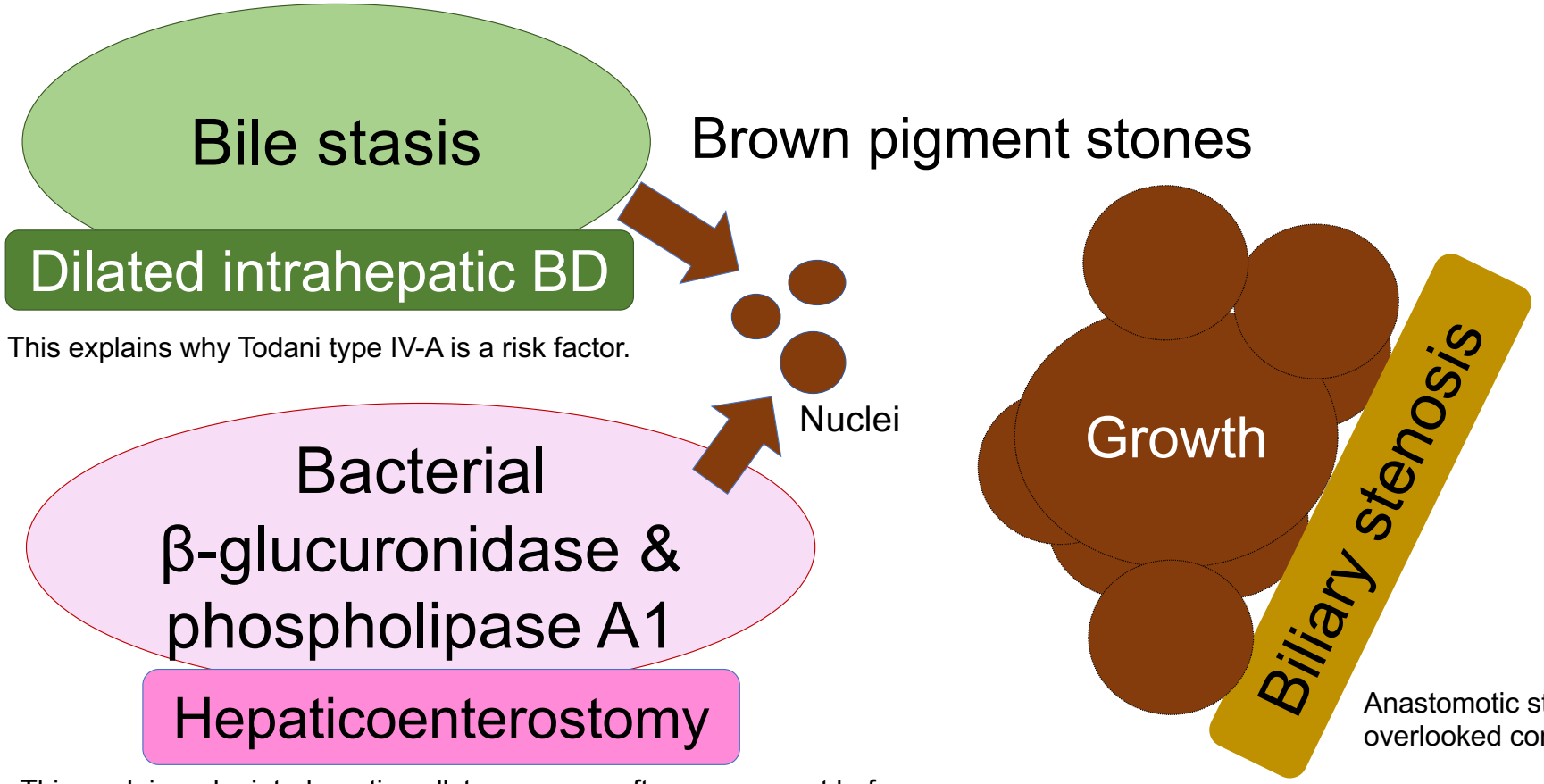
Anastomotic stricture
in non-dilated PBM



Congenital septal stenosis



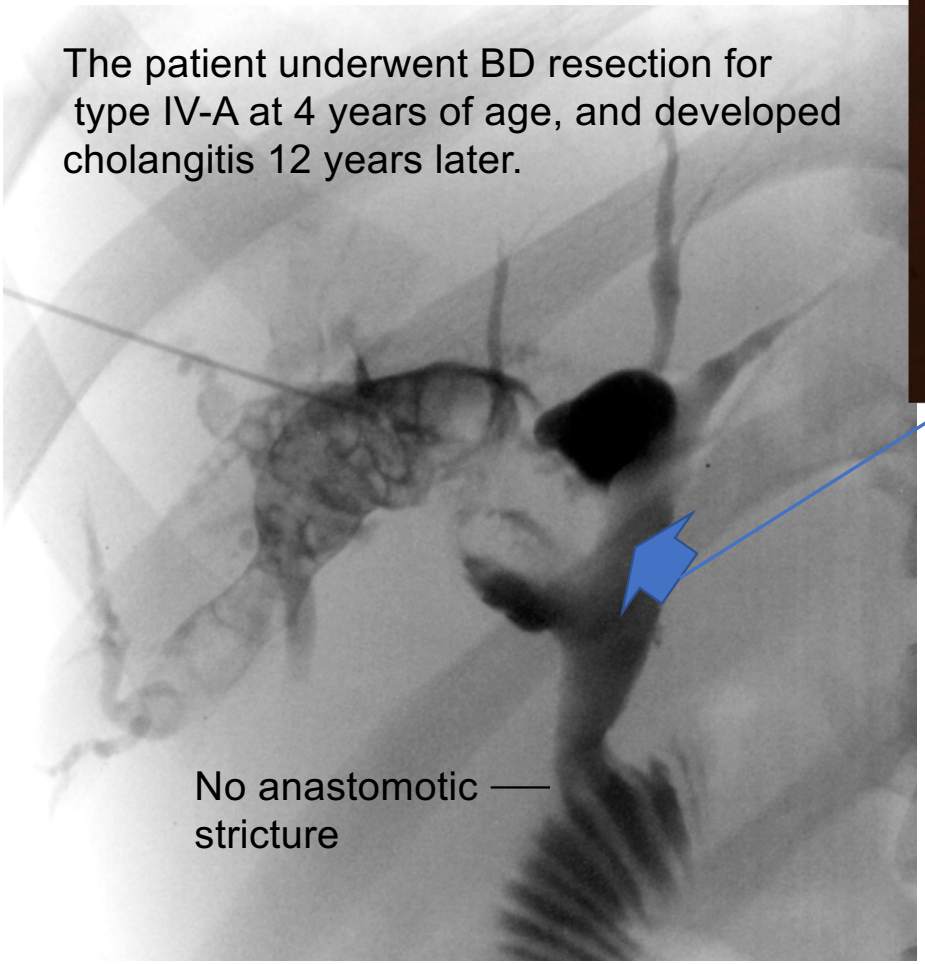
Mechanism of postoperative gallstone formation

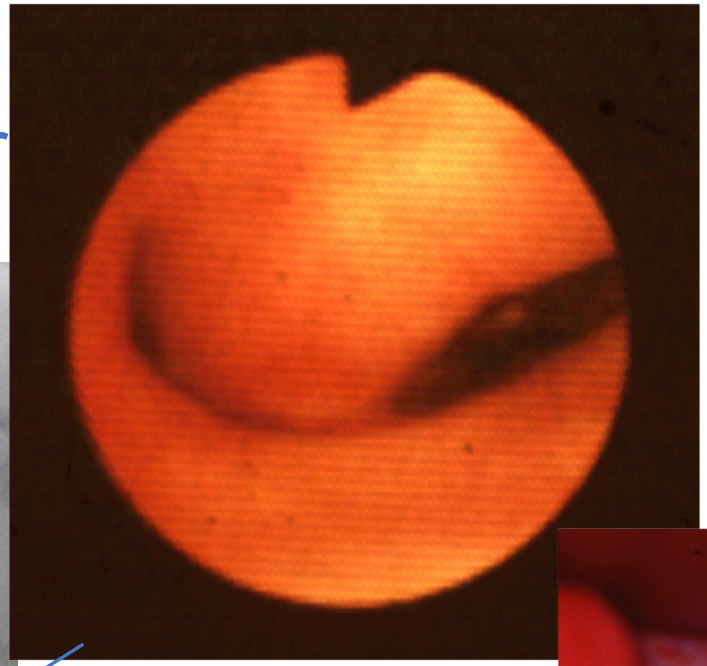


Stone removal alone is insufficient for a permanent cure;
relieving stenosis or resecting the dilated BD (hepatectomy) is required.

Case 24 Intrahepatic gallstones after BD resection

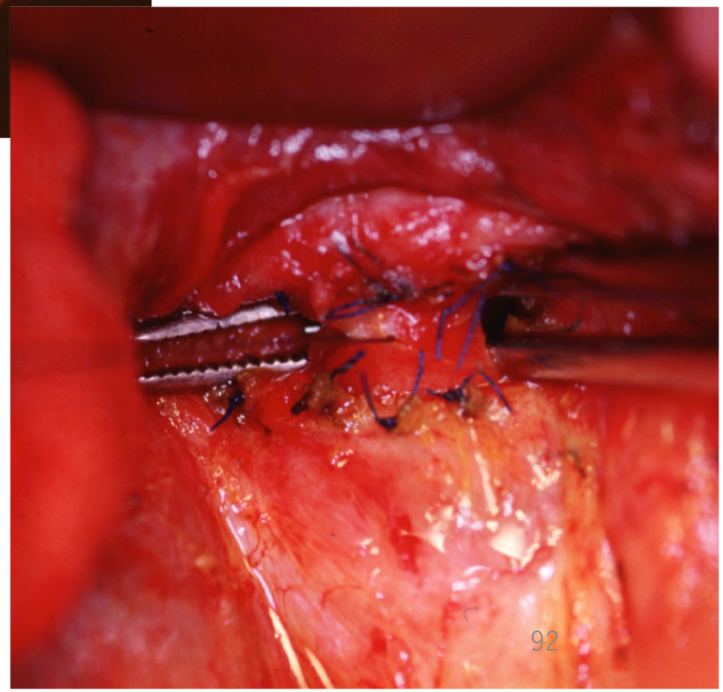
The patient underwent BD resection for type IV-A at 4 years of age, and developed cholangitis 12 years later.





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

Septal stenosis trapping gallstones



Case 25

Refractory cholangitis will occur

BD resection of type IV-A CBD at 1 year of age.

↓

In her 20s, hepatolithiasis (HL) developed, and PTCS* was performed.

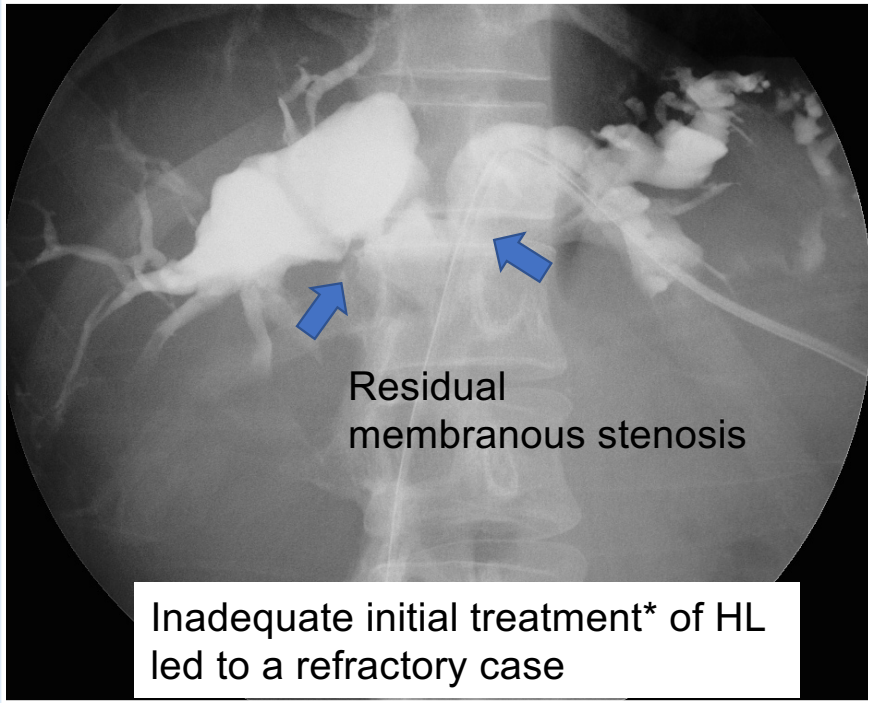
↓

Mild cholangitis recurred repeatedly, but she did not seek medical advice.

↓

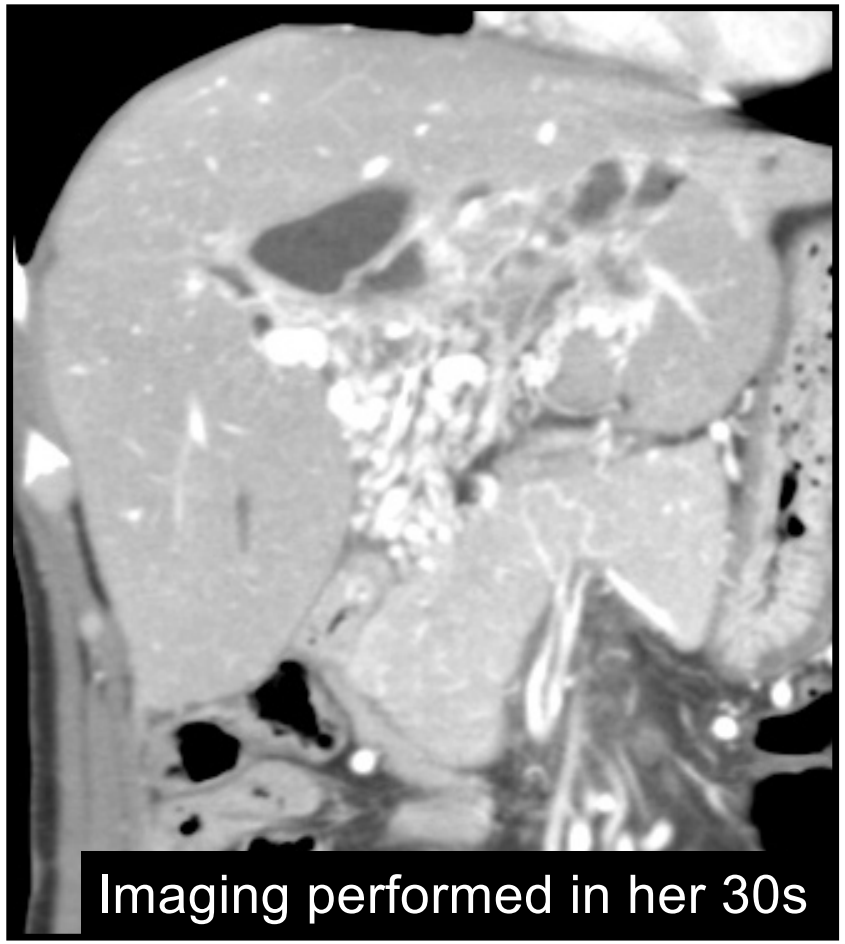
In her 30s, she had severe cholangitis with recurrent HL.

Cavernous transformation of the portal vein



Inadequate initial treatment* of HL led to a refractory case

*PTCS, percutaneous transhepatic cholangioscopic lithotomy



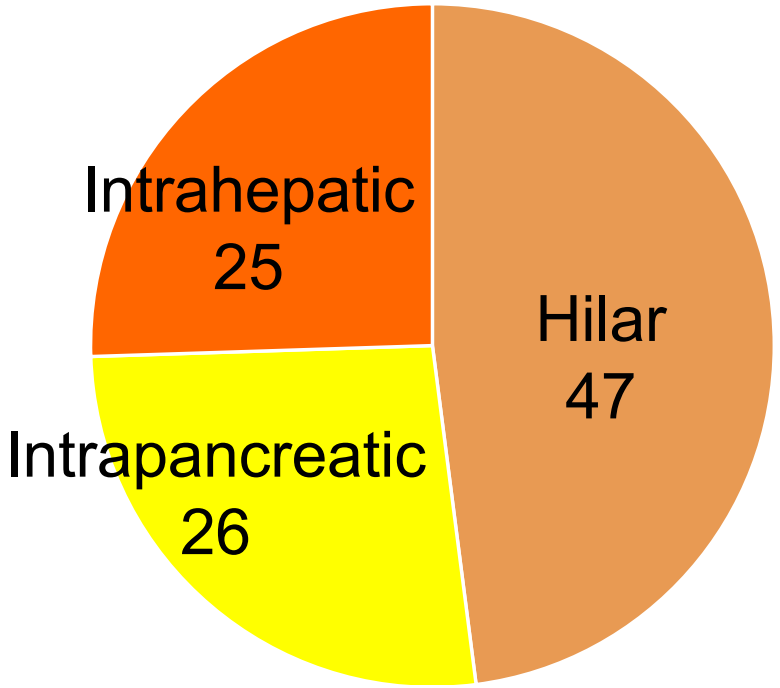
Imaging performed in her 30s

Long-term postoperative complications: Carcinogenesis in the residual BD

Higher incidence even after BD resection

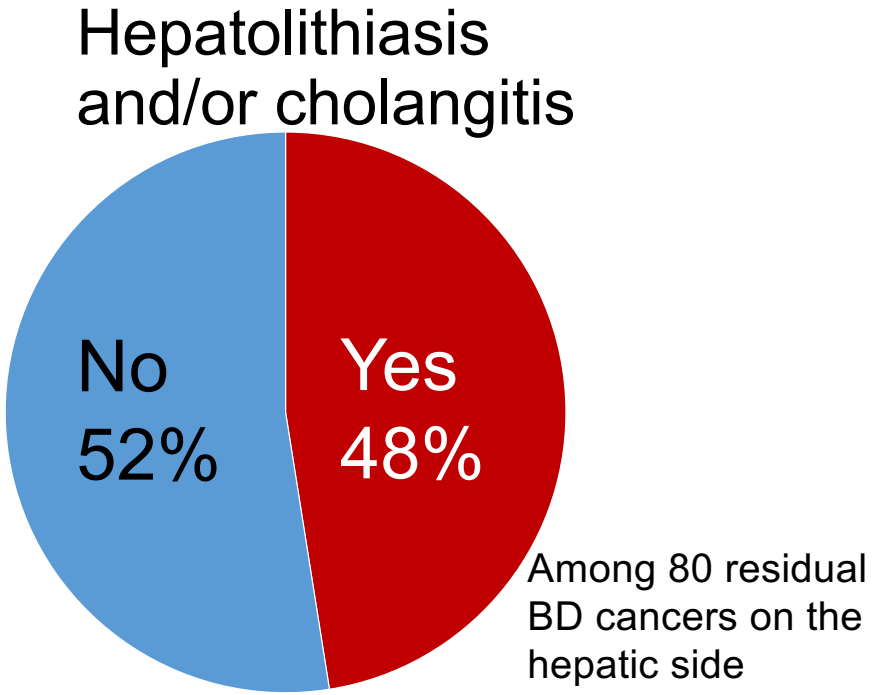
- 107 cases collected from the literature
- Carcinogenesis rate: 2.0%
- Mean interval to carcinogenesis:
  12 postoperative years (mean age: 47 years)

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

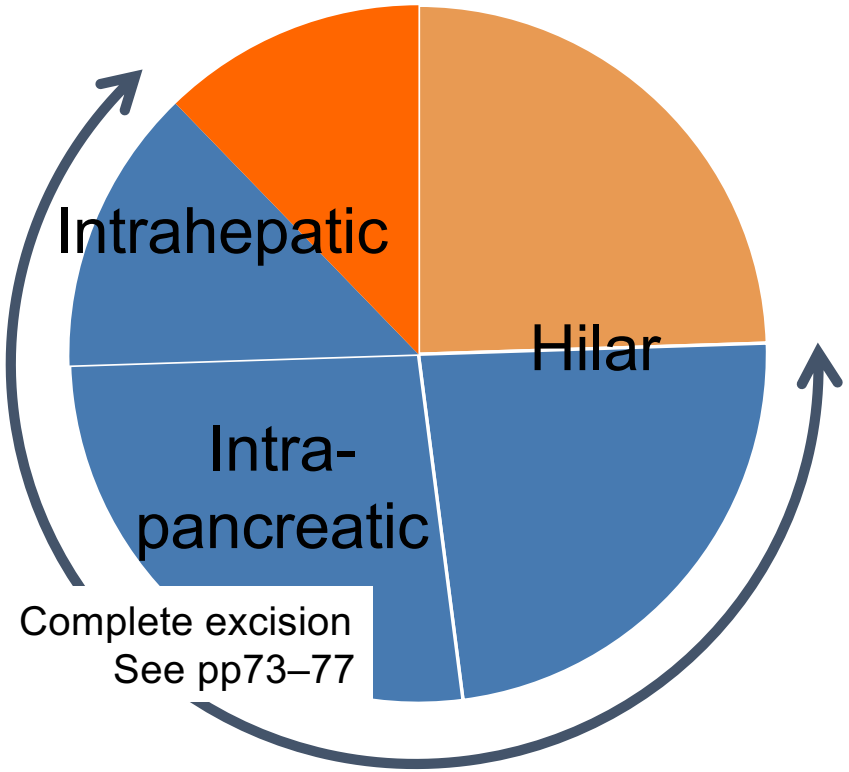


cf. US Survey
Biliary cancer after BD resection: 13/394 (3.3%)
Pediatric cases 4/135 (3.0%) / Adult cases 18/259 (6.9%)
Soares et al. *JAMA Surg* 2015

Many cases of residual BD cancer could have been prevented



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

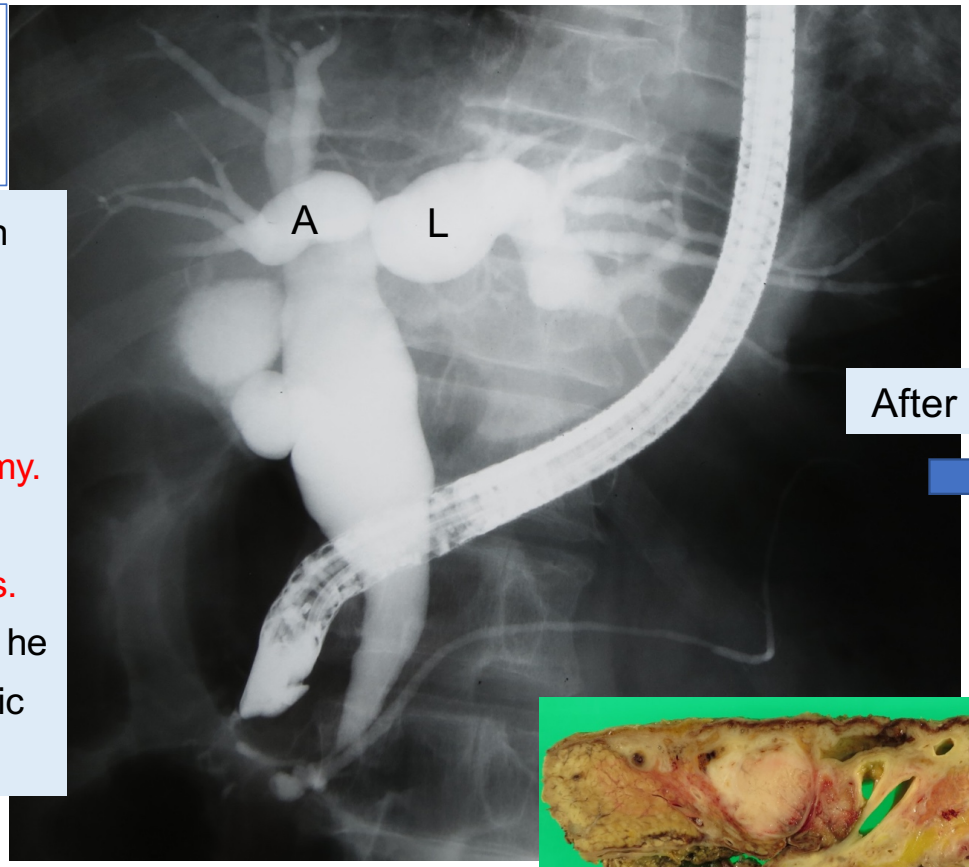


Appropriate procedures could have prevented these cancers.

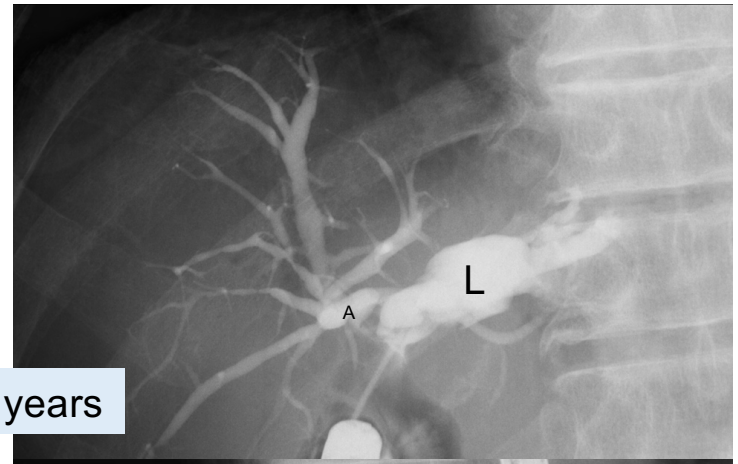
Intrahepatic BD cancer after BD resection

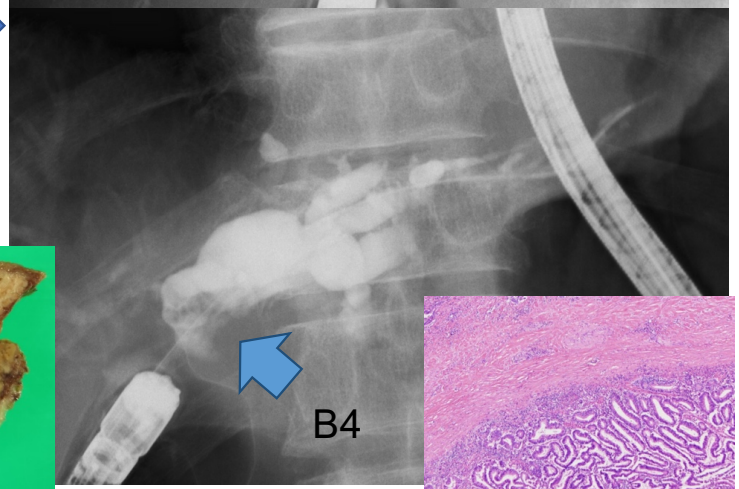
Case 26

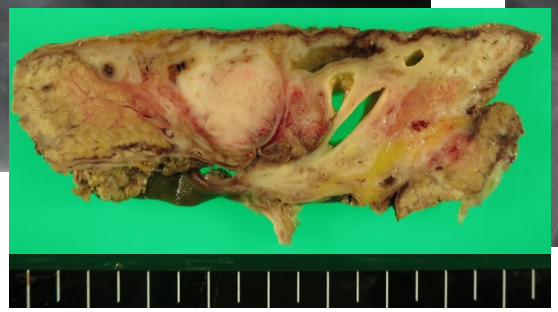
A male in his 60s with type IV-A CBD underwent BD resection with hepaticoduodenostomy. Four years late, he developed cholangitis. Nine years after that, he developed intrahepatic BD cancer in B4.



After 13 years







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

Basics

High confluence of the pancreaticobiliary ducts

First version, September 2024; Revised, September 2026

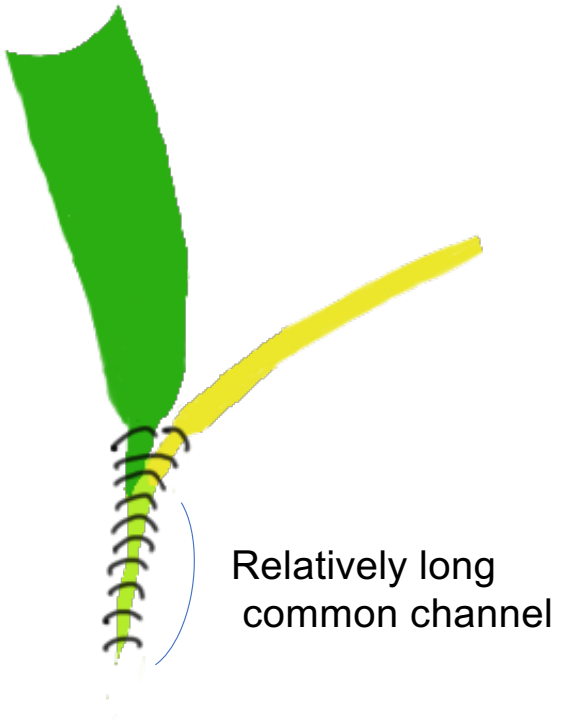
What is High confluence of pancreaticobiliary ducts?

Definition

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

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

Normal — **High confluence** — PBM



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

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

Spectrum from normal to PBM

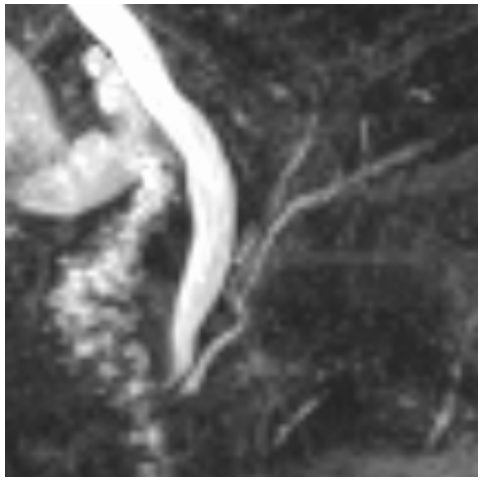
Normal

Occult pancreatobiliary reflux

High confluence

PBM

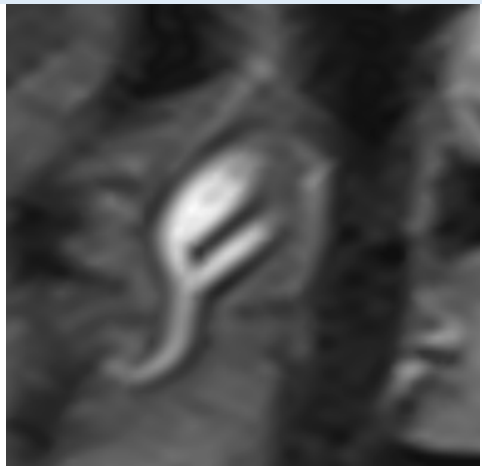
The sphincter function affecting the confluence



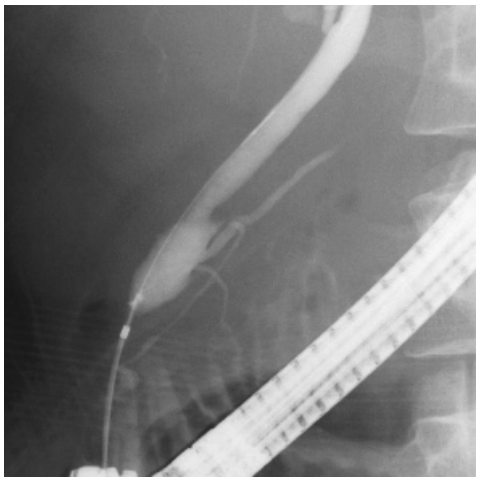
Separate openings



Common channel formation
Cholangiography occasionally visualizes the PD.



Relatively long common channel



Abnormally long common channel

High confluence is a transitional type

High confluence and PBM among patients undergoing ERCP

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

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