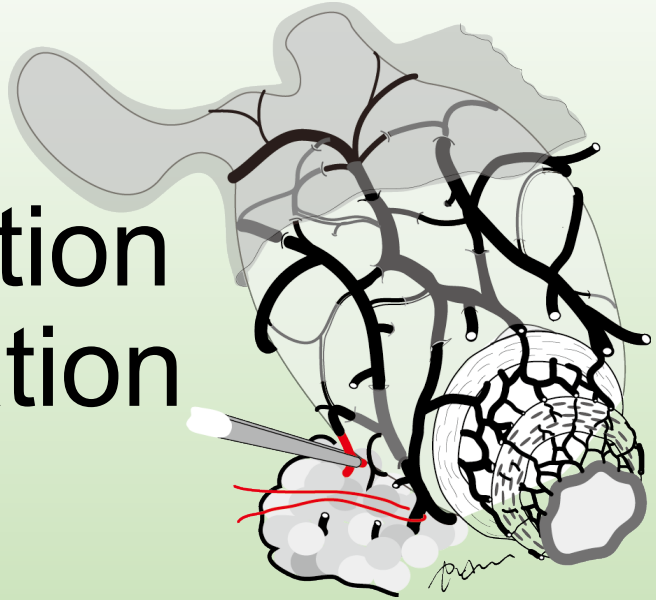
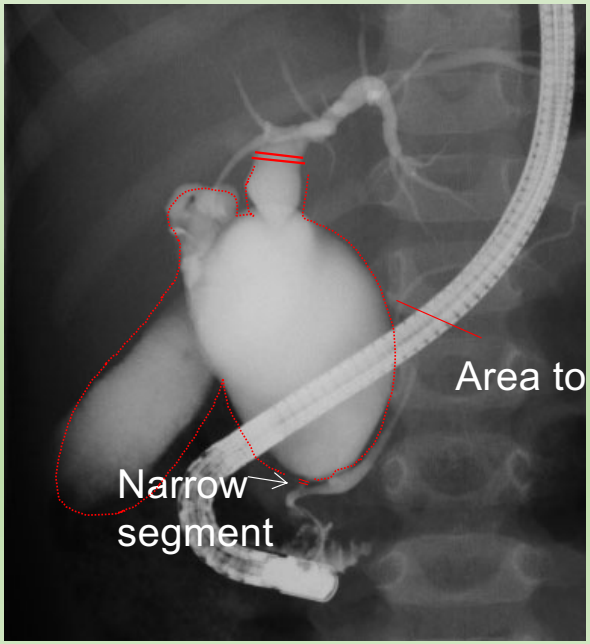
Atlas of Pancreaticobiliary Maljunction & Congenital Biliary Dilatation





Japanese Study Group on Pancreaticobiliary Maljunction (JSGPBM)
Scientific Committee

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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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Introduction

First version, September 2022; Revised, September 2026

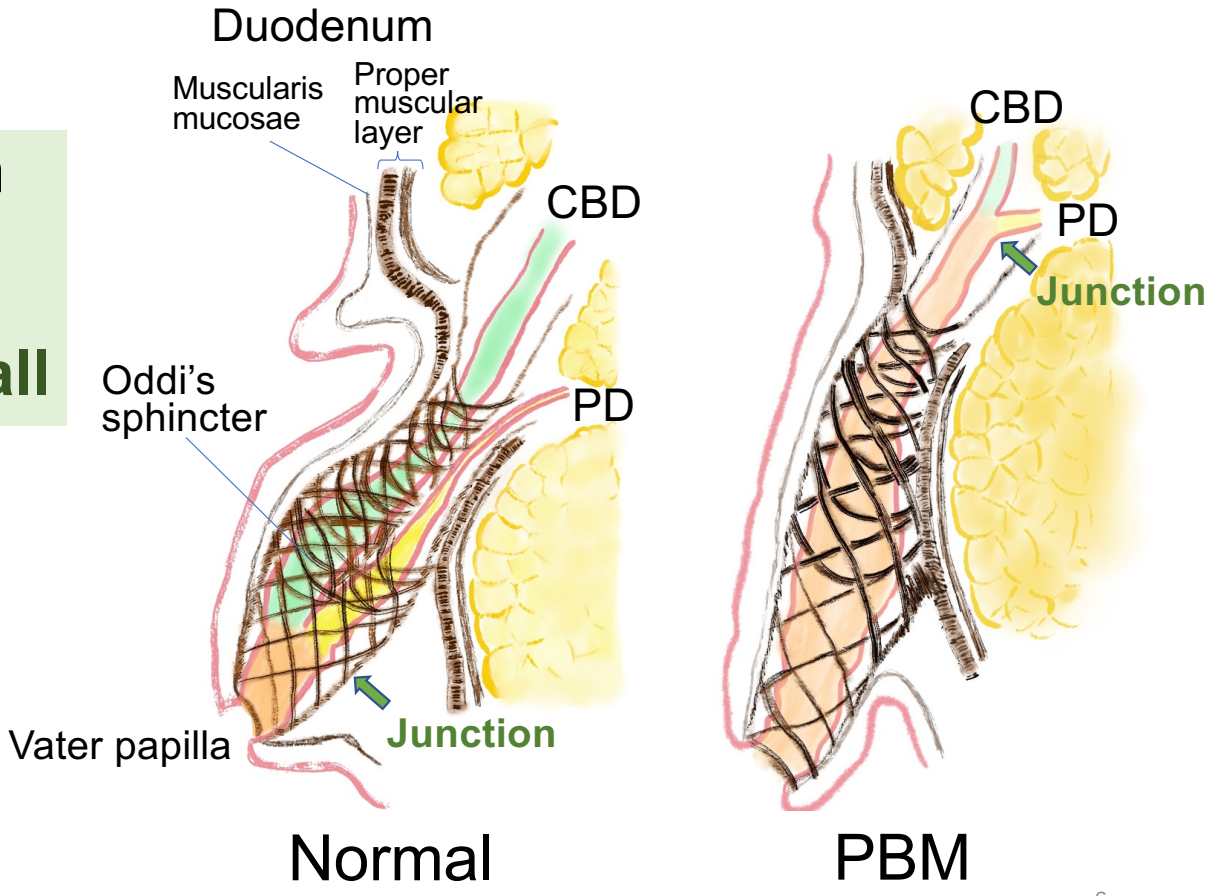
What is Pancreaticobiliary Maljunction (PBM)?

Definition

A congenital malformation in which the **pancreatic duct** and **bile duct join outside the duodenal wall**

The junction outside of the sphincter action is a functional phenomenon of PBM but is not the definition of it. However, when the confluence outside the duodenal wall cannot be shown, identification of the junction outside the sphincter action is enough for a diagnosis of PBM.

See *Basic Section*.

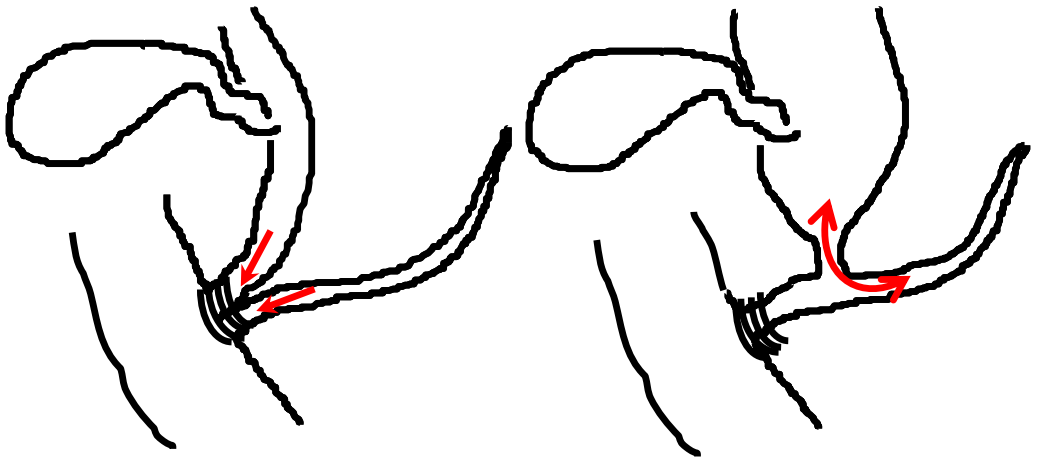


What is Pancreaticobiliary Maljunction (PBM)?

Pathophysiology

In PBM, the sphincter action does not regulate the pancreaticobiliary junction and **two-way regurgitation** of pancreatic juice and bile occurs, resulting in various pathological conditions of the biliary tract and pancreas.

During **childhood**, intermittent **symptoms** including abdominal pain, vomiting, and jaundice occur. During **adulthood**, **biliary cancers** develop.

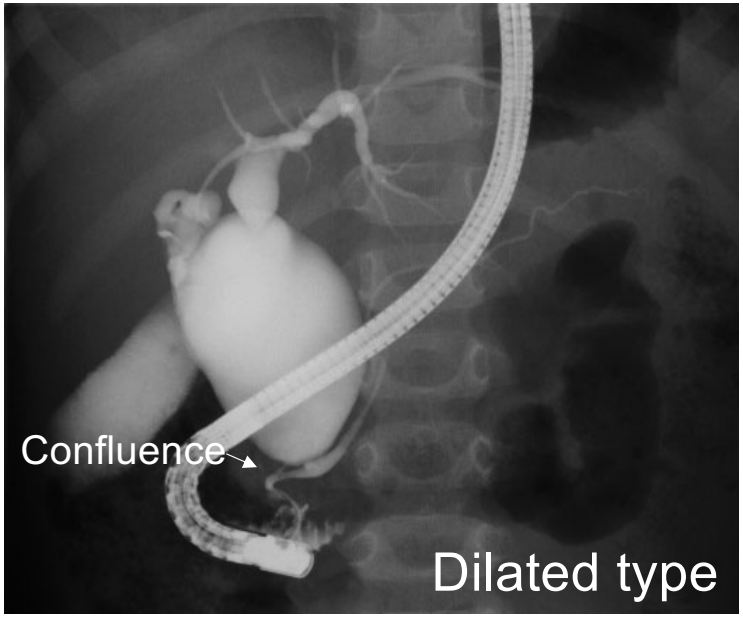


Normal PBM

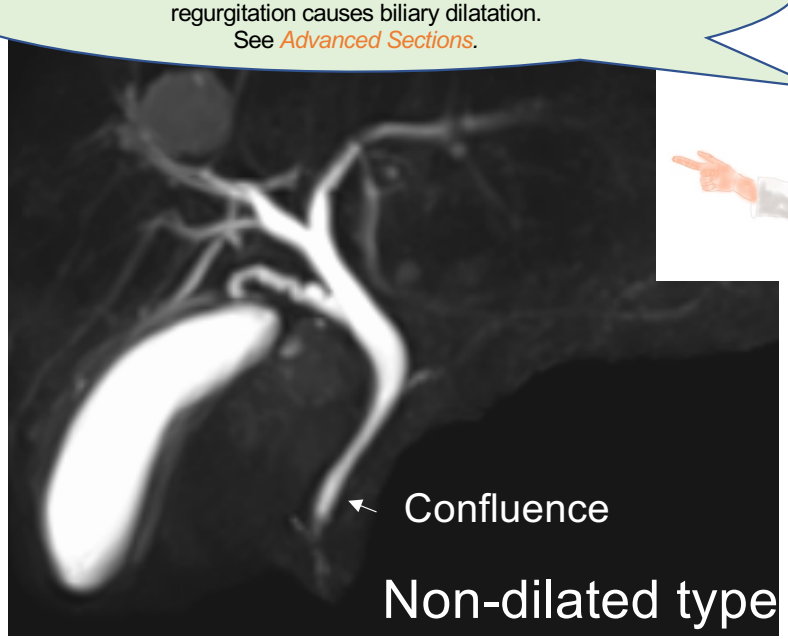
What is Pancreaticobiliary Maljunction?

Two major categories according to whether the bile duct is dilated or not:

- Dilated type = congenital biliary dilatation (CBD)
- Non-dilated type



The definition of non-dilated type remains problematic.
See *Basic and Advanced Sections*.
Existence of non-dilated type denies that pancreatobiliary regurgitation causes biliary dilatation.
See *Advanced Sections*.



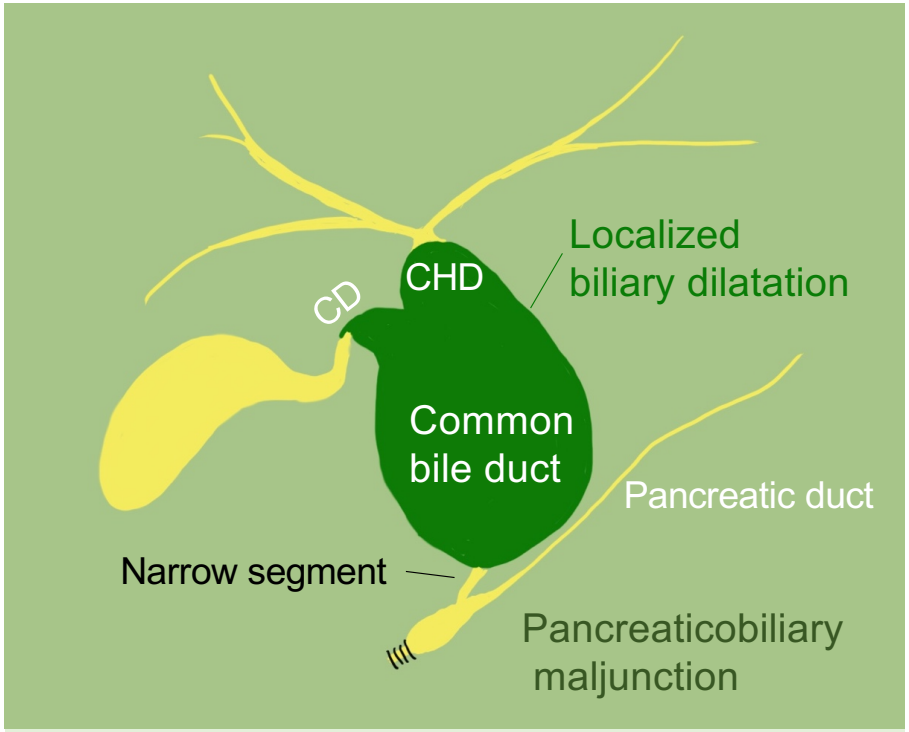


What is Congenital Biliary Dilatation (CBD)?

Definition

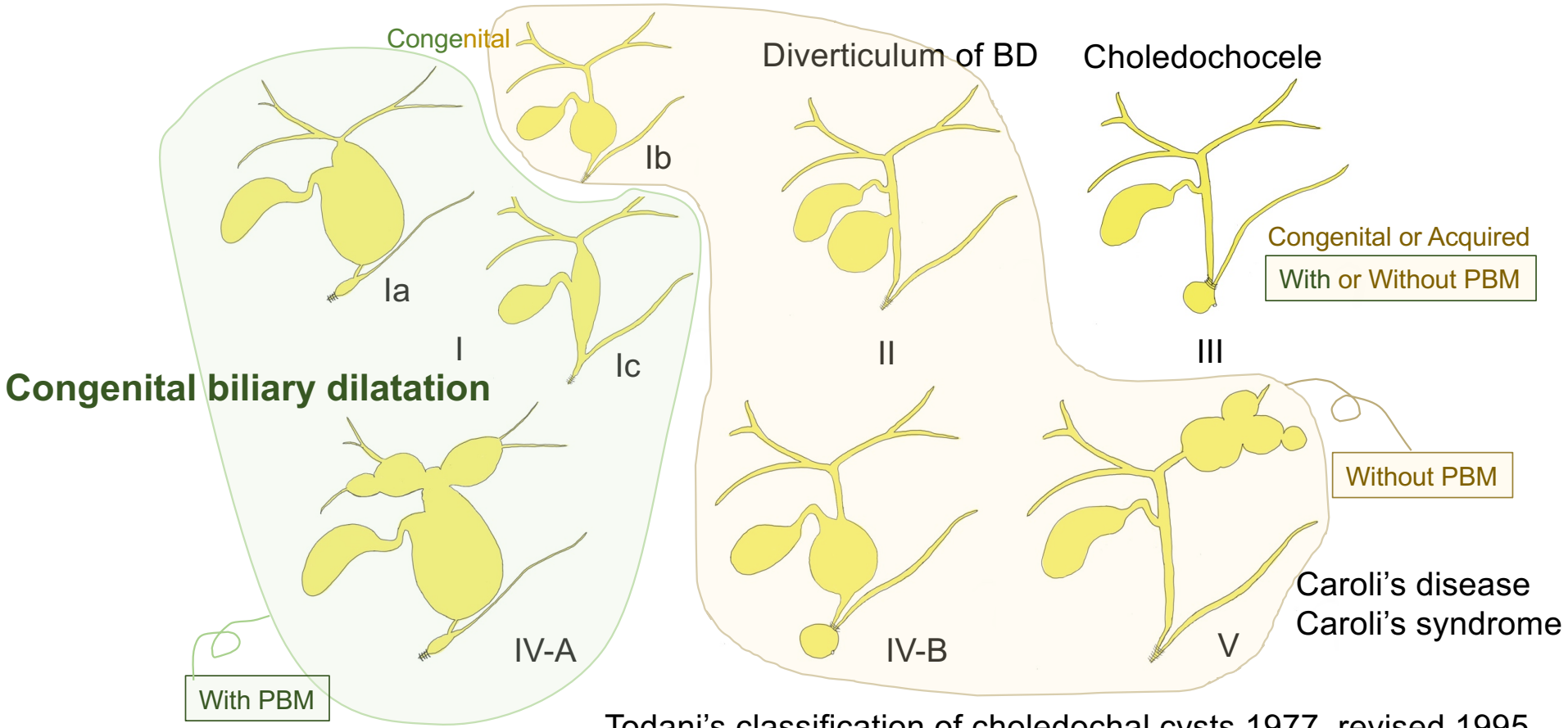
Congenital malformation involving **PBM** and **localized dilatation of the bile duct (BD)** including the common bile duct

Definition of CBD proposed by the Japanese Study Group on Pancreaticobiliary Maljunction (JSGPBM) in 2015



In this case, the common hepatic duct (CHD) and the base of the cystic duct (CD) are dilated, but bile ducts upstream to the CHD and the gallbladder are not dilated (localized dilatation).

The term choledochal cyst involves different entities

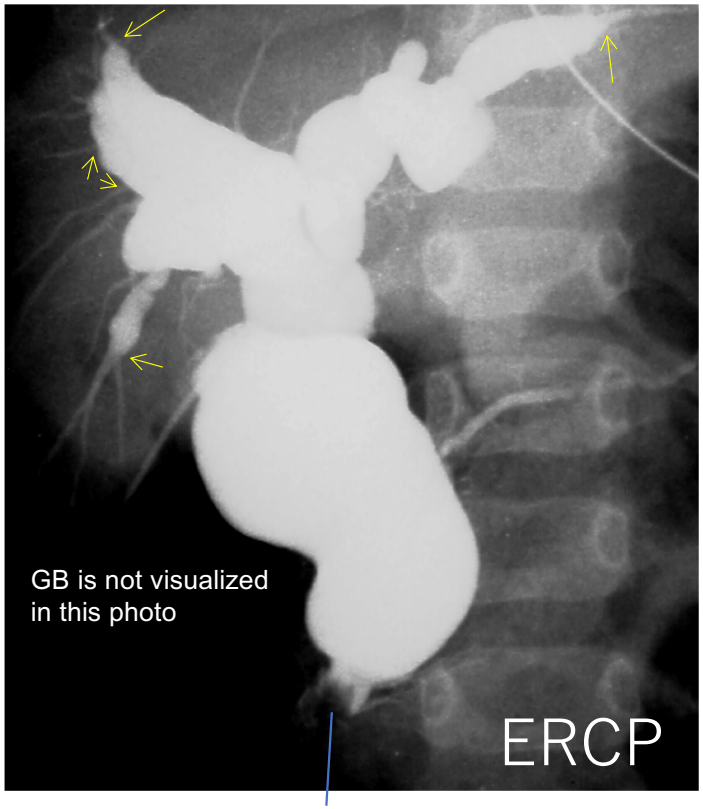


Todani's classification of choledochal cysts 1977, revised 1995

“Localized dilatation of BD” indicates “congenital” disease

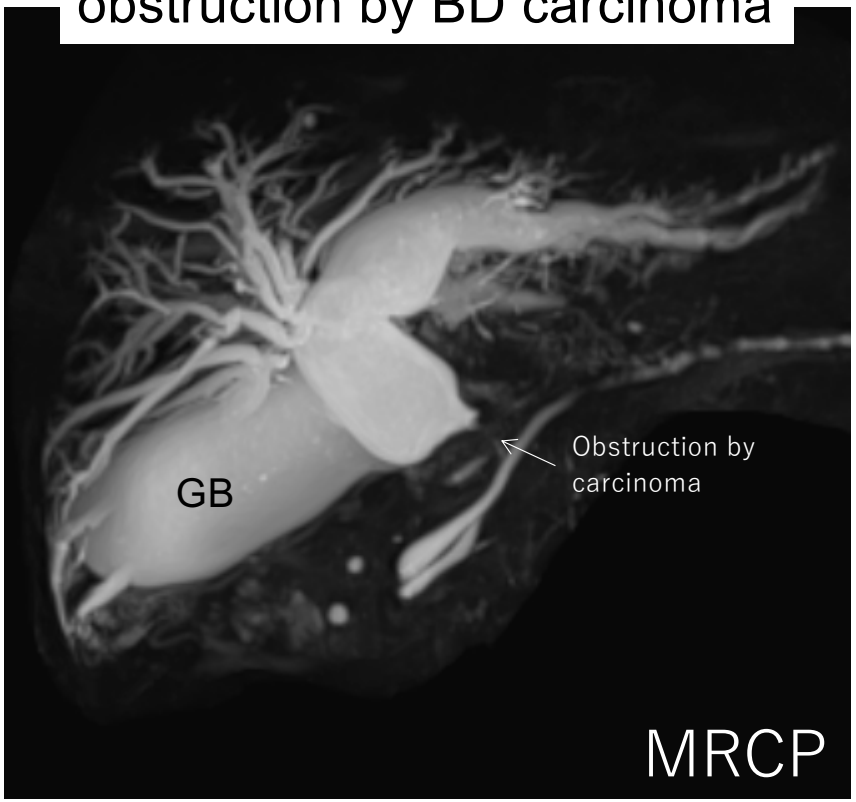
Even when dilatation extends to intrahepatic BDs, dilated areas are localized. **Dilated BDs abruptly change to those of normal caliber** (→).
The GB is never enlarged, although the cystic duct is dilated at its base.
However, in secondary dilatation, dilatation extends smoothly upstream to the peripheral intrahepatic BD.
GB is also enlarged (Courvoisier sign).

Congenital biliary dilatation



Protein plug
compacted in the common channel

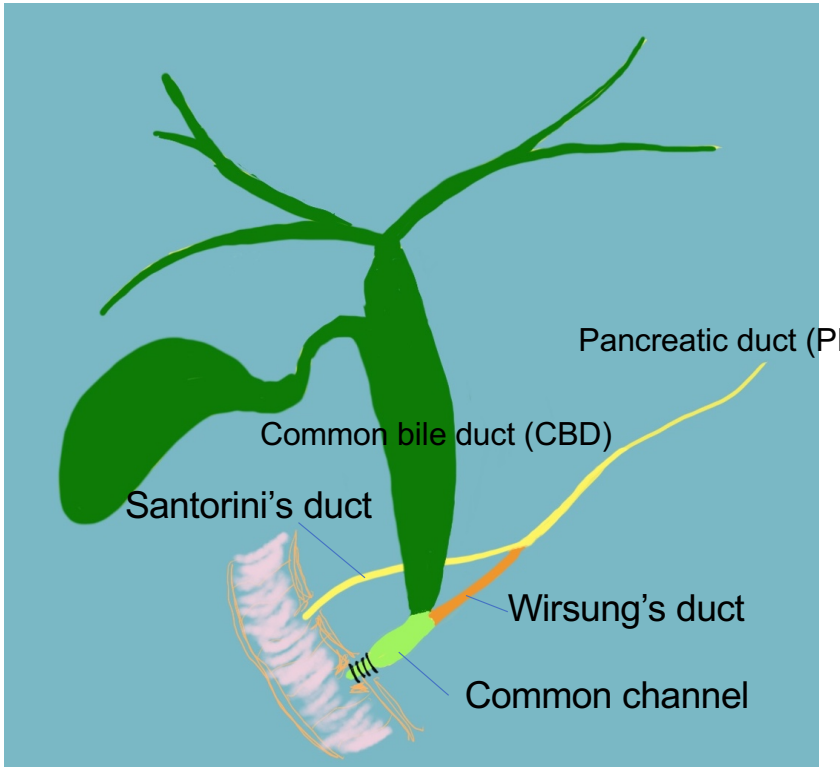
Secondary dilatation due to obstruction by BD carcinoma



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

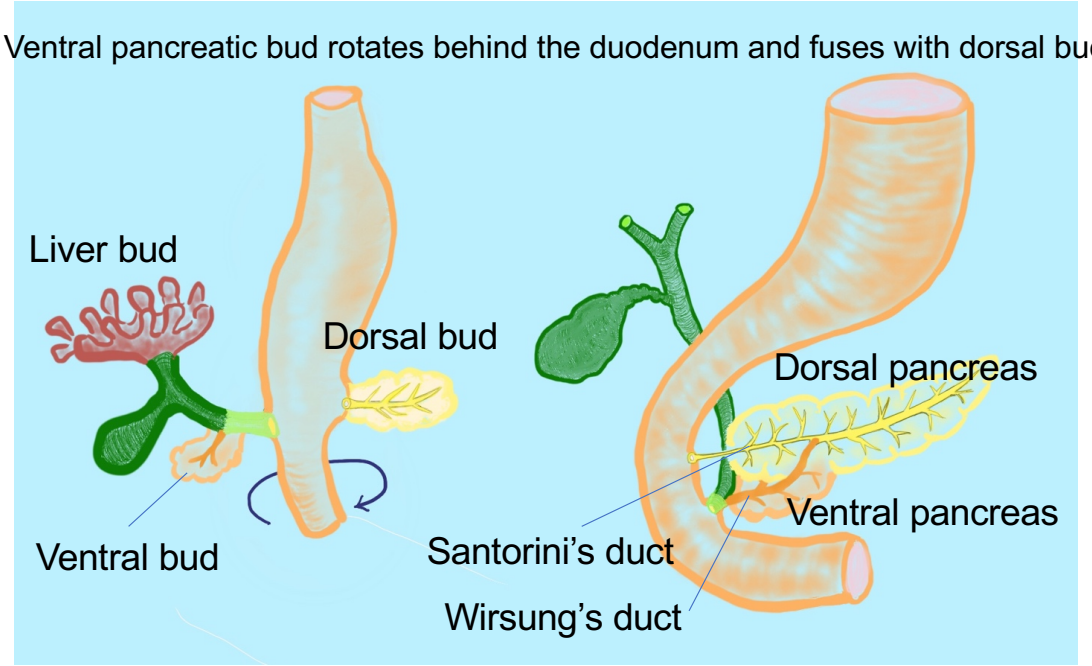
PBM arises from anomalous genesis of ventral pancreatic duct and bile duct

What is wrong in this figure?



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

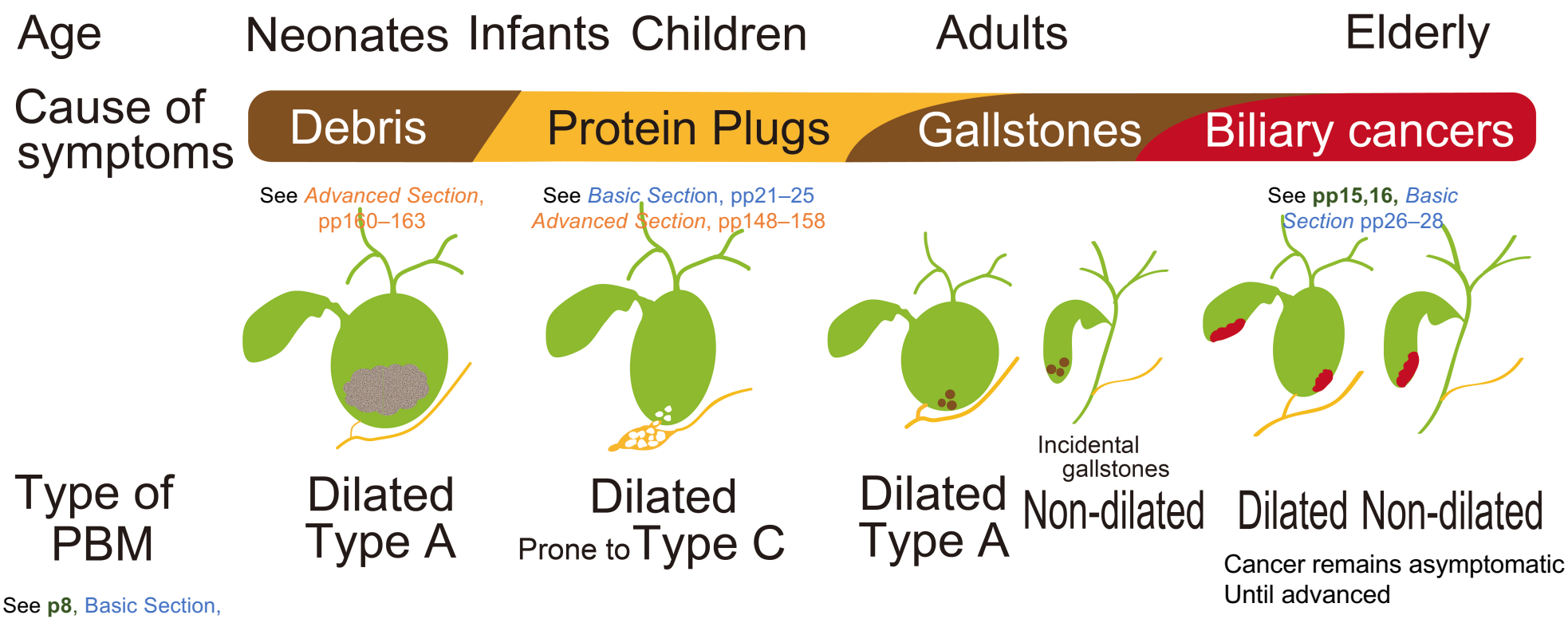
Pancreas development



Connection anomaly never happens between the BD and dorsal pancreatic duct in PBM.

Fusion failure between the ventral and dorsal pancreatic ducts (i.e., pancreas divisum) can occur together with PBM. See *Advanced Section*.

PBM causes symptoms only with secondary factors



See **p8**, Basic Section, pp35–40

Pathogenesis varies by life stage

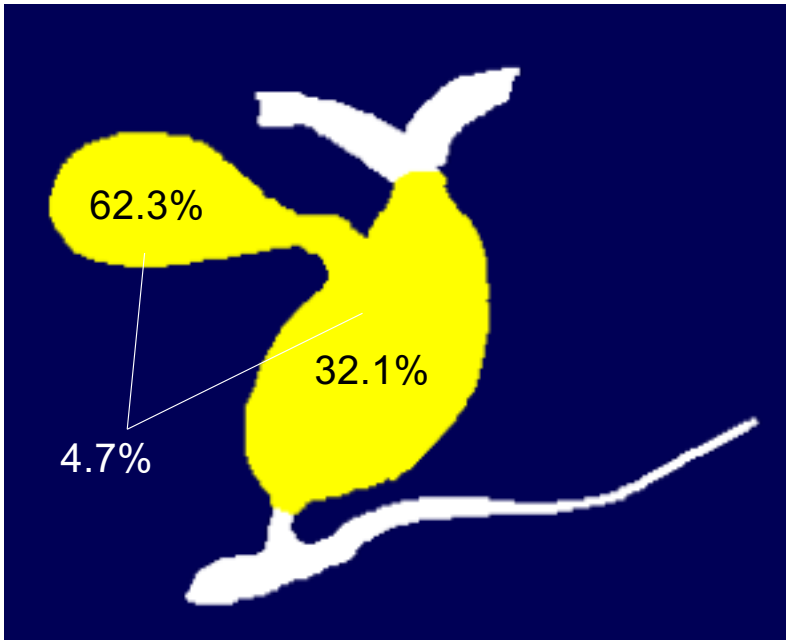
Treatment for pancreaticobiliary maljunction

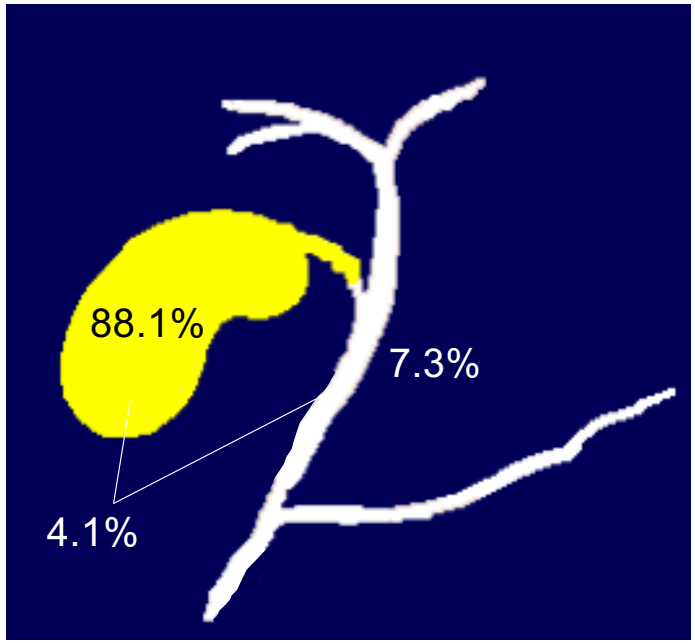
- Rationale for treatment:
  - ✓Not merely to correct malformation!
  - ✓Treatment is indicated to prevent complications.
- Complications of PBM:
  - ✓**Pediatric cases**: Recurrent **symptoms** including abdominal pain, vomiting, and jaundice.
  - ✓**Adult cases**: High incidence of **biliary tract 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.

Biliary carcinogenesis

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





Cancer rate	CBD 21.6%	Non-dilated PBM 42.4%
	215/997 cases	218/514 cases

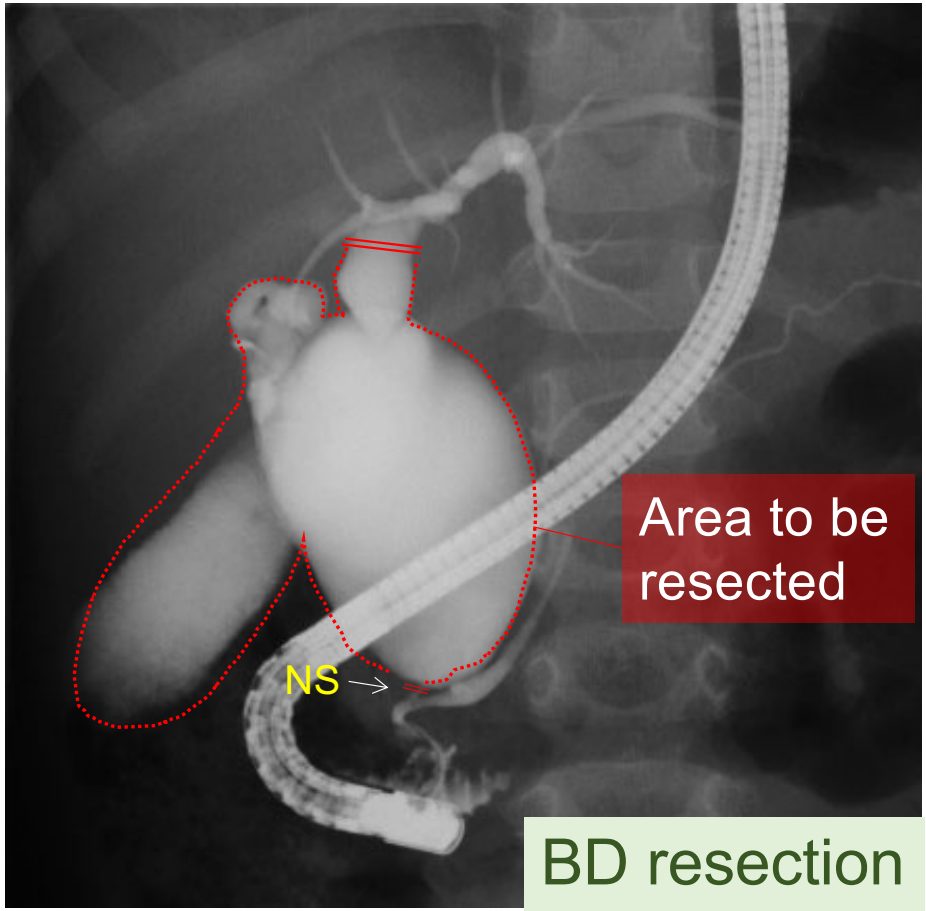
Biliary cancers arise 10–20 years earlier in PBM

	Gallbladder cancer		*P*
PBM	Yes	No	
	36 cases	232 cases	
Age (years)	**58.9** ± 8.8	**69.4** ± 8.7	< 0.01
M:F	1:2.1	1:2.9	NS

	Bile duct cancer		P
PBM	Yes	No	
	6 cases	159 cases	
Age (years)	**54.8** ± 14.3	**68.8** ± 9.1	< 0.01
M:F	1:2	1:0.7	NS

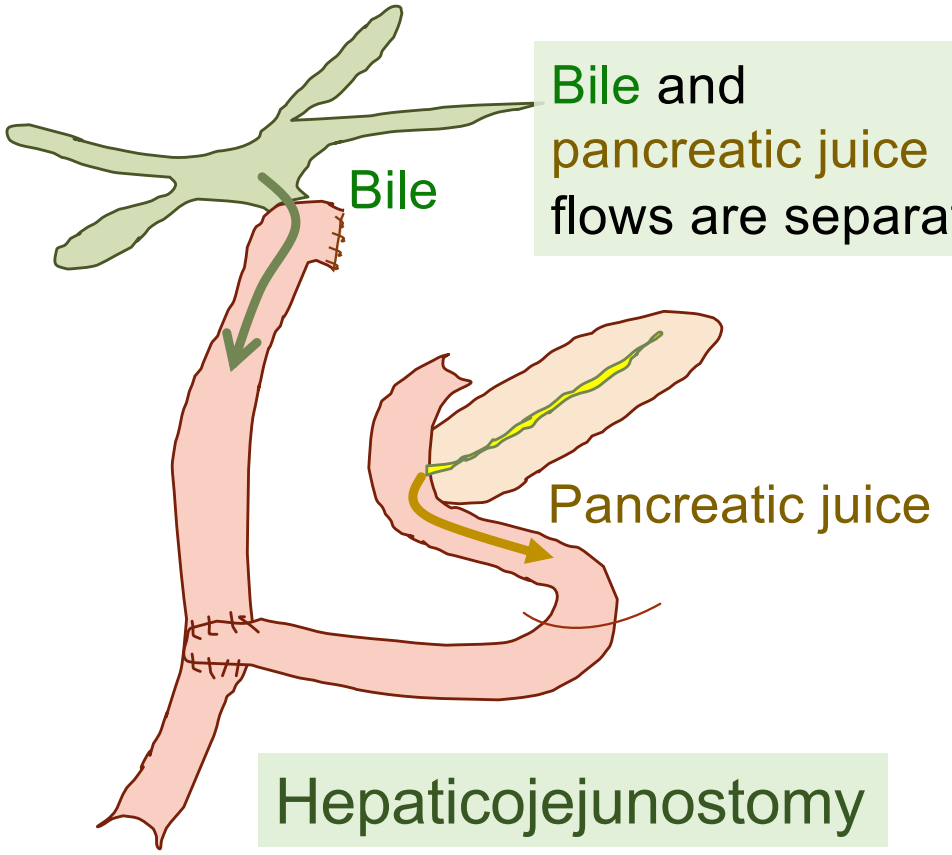
Kamisawa T, Tu Y, Kuwata G, Egawa N, Nakajima H, Tsuruta K, Okamoto A, Matsukawa M.
Biliary carcinoma risk in patients with pancreaticobiliary maljunction and the degree of extrahepatic bile duct dilatation.
Hepatogastroenterology. 2006;53(72):816-8.

Surgery for CBD



NS, narrow segment

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



Recommended terms

- **Pancreaticobiliary maljunction (PBM)** (Official MeSH term since 2020)
- **Congenital biliary dilatation (CBD)**
- **Bile duct resection** as a procedure for CBD

Non-recommended terms

- Pancreaticobiliary malunion or pancreatobiliary malunion
- Choledochal cyst (when indicating CBD [Todani types Ia, c, and IV-A])

The term "choledochal cyst" encompasses various entities, such as CBD, choledochocele, and Caroli's disease.