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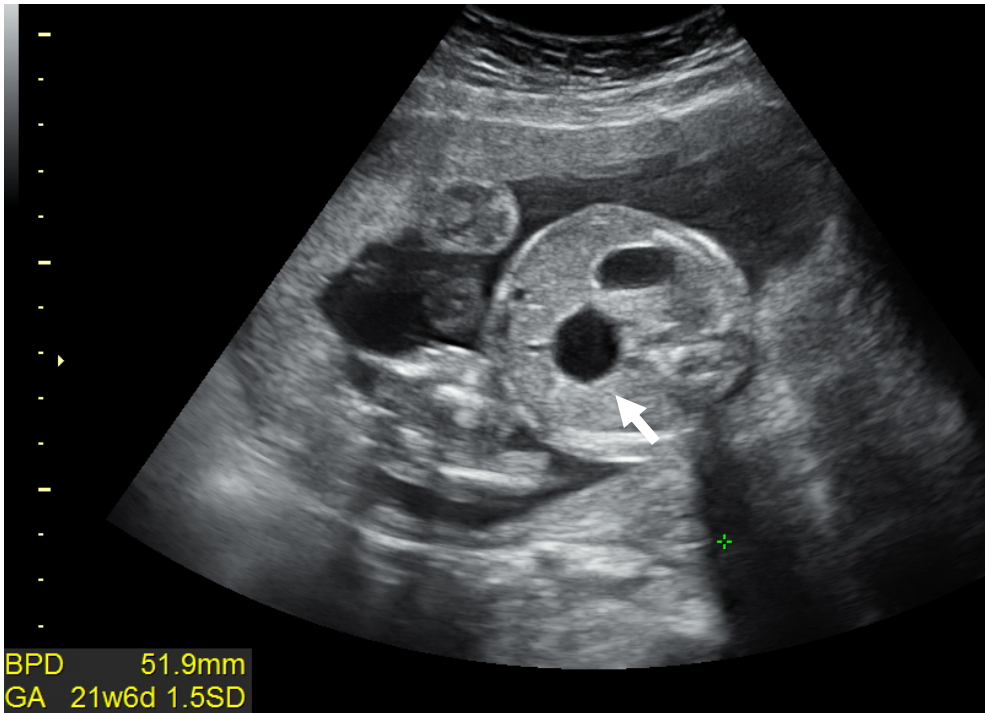
Professional

Prenatal diagnosis

Transition to biliary atresia

First version, September 2024

Both congenital biliary dilatation (CBD) and biliary atresia (BA) are found prenatally as a biliary cyst



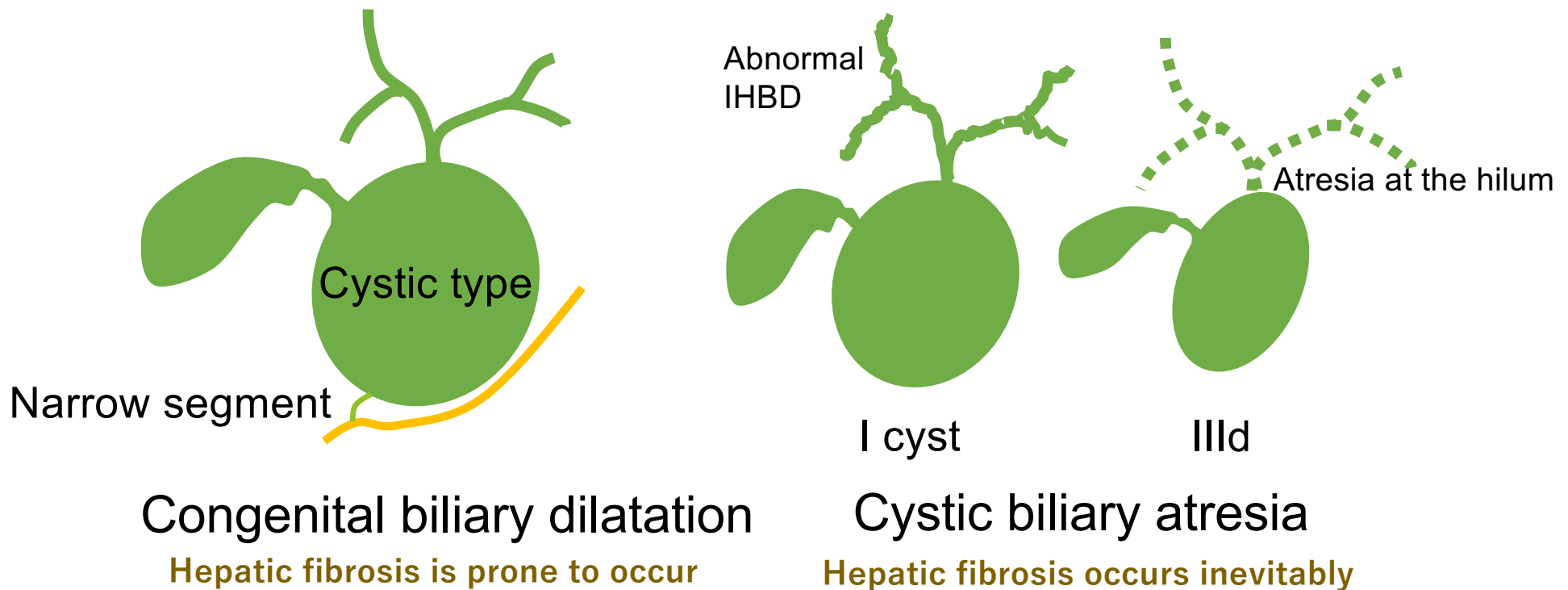
Differential diagnosis is difficult even after birth

→ Congenital biliary dilatation ?
or
Biliary atresia ?

Why?

This fetal ultrasonogram showed connection between the extrahepatic cyst and intrahepatic bile ducts. However, type IIIb biliary atresia was diagnosed after birth.

Fetal biliary cysts present a similar form regardless of final diagnoses



Biliary obstruction causes cholangiopathy specific to neonates and early infants

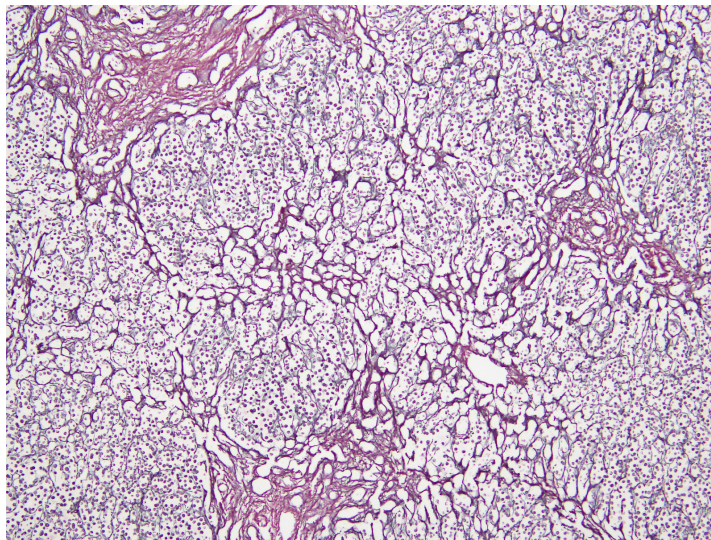
Prenatally diagnosed CBD and hepatic fibrosis

- Prone to hepatic fibrosis
- Fibrosis can occur even in asymptomatic cases.
- The fibrosis improves after bile duct resection suggesting that the fibrosis is reversible.

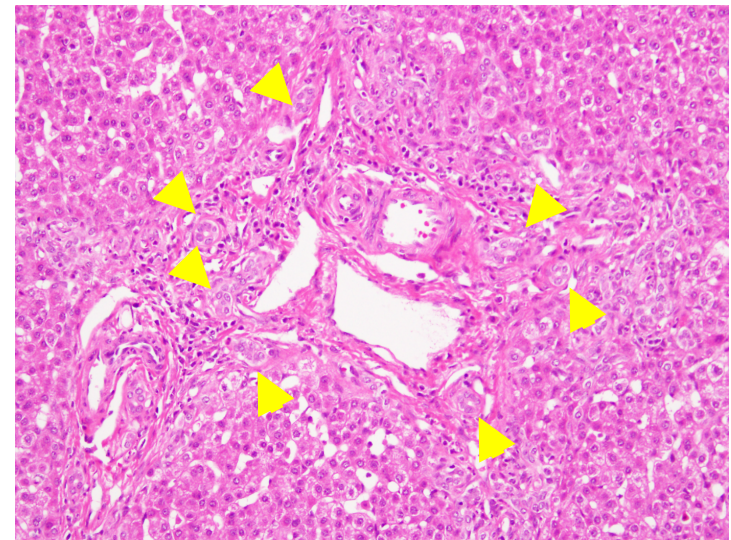
Diagnosis at 29 weeks' gestation

BD resection when acholic stool occurred at 6 months

Liver pathology shows the same cholangiopathy as seen in biliary atresia.

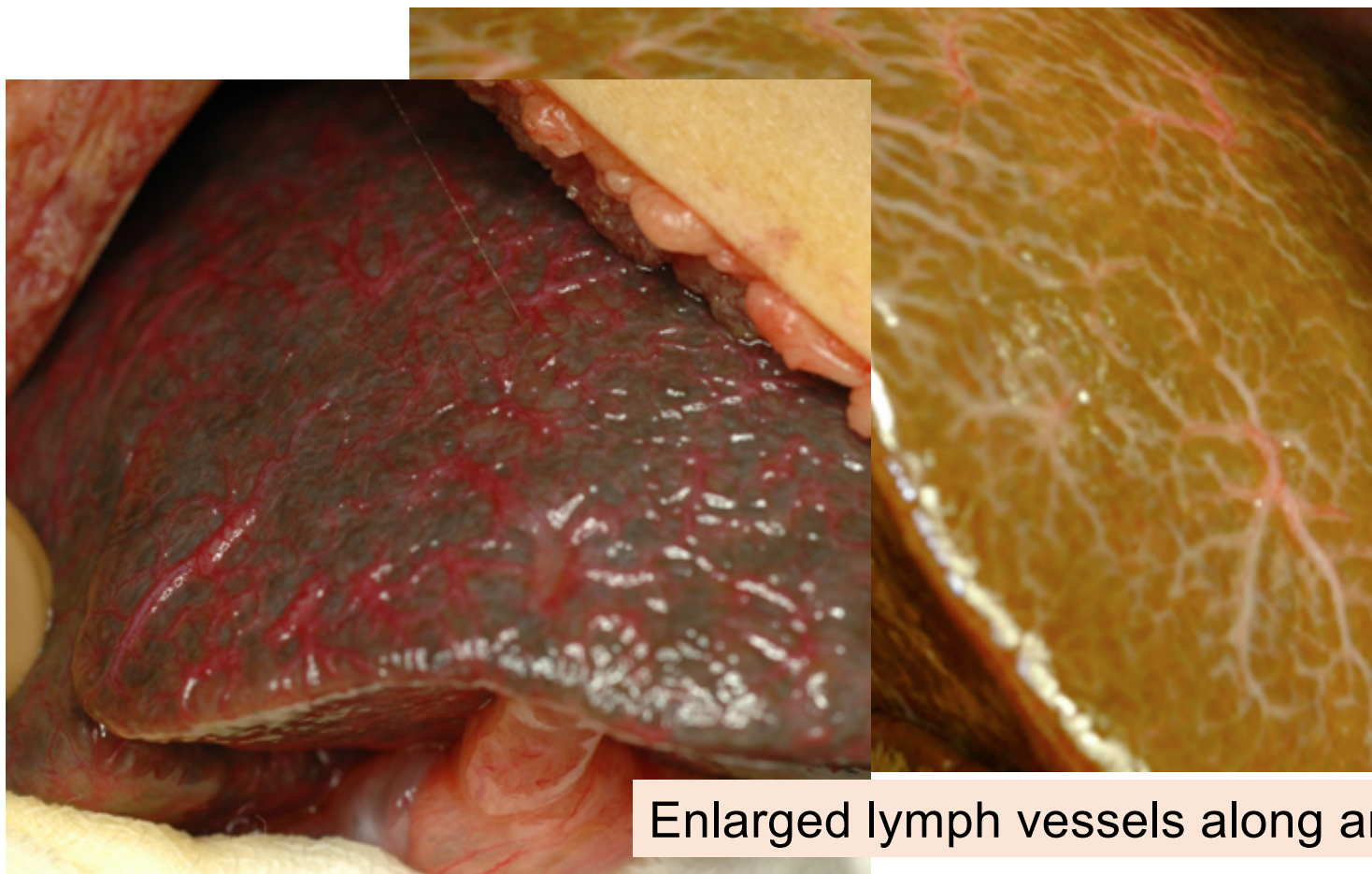


Bridging F1-2



Cholangiocyte proliferation

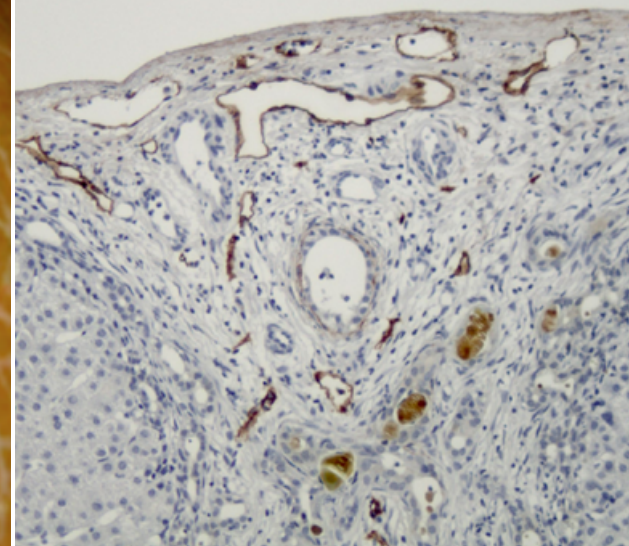
Makibishi sign specific to Biliary Atresia



Enlarged lymph vessels along arteries on the liver surface



Presented by Dr Tainaka, Nagoya Univ.

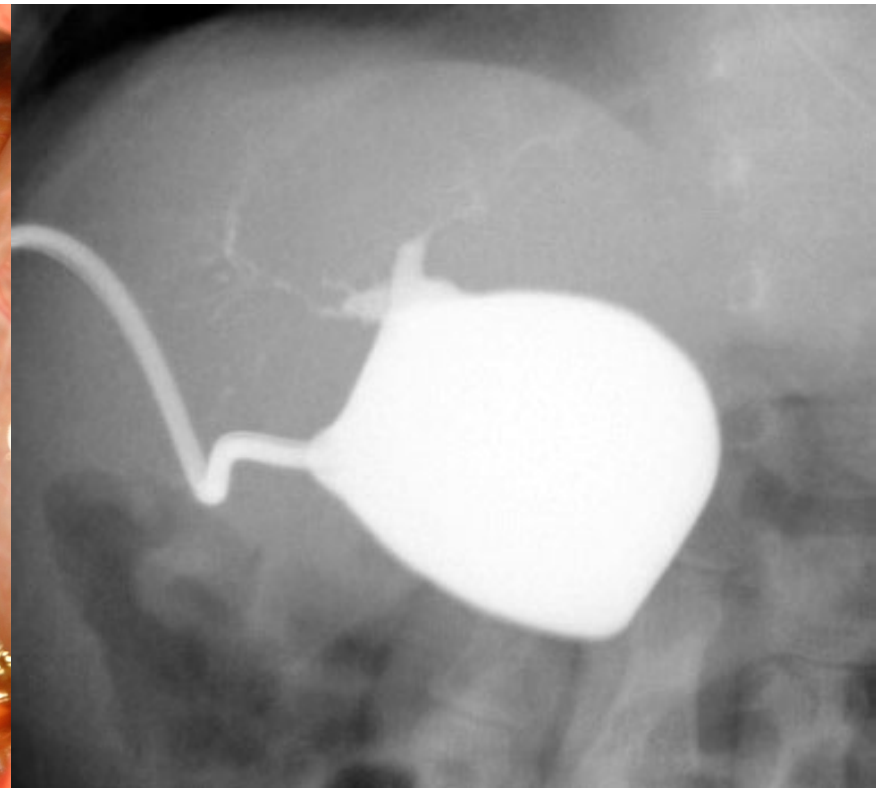
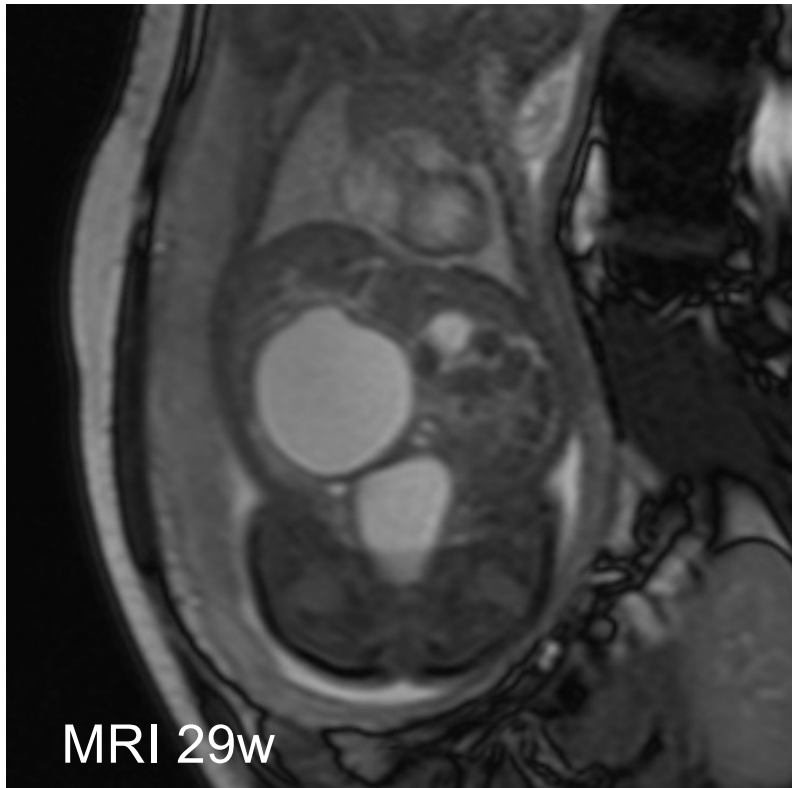


Case **43**

Type I cyst Biliary Atresia

Makibishi sign

Treelike intrahepatic BDs

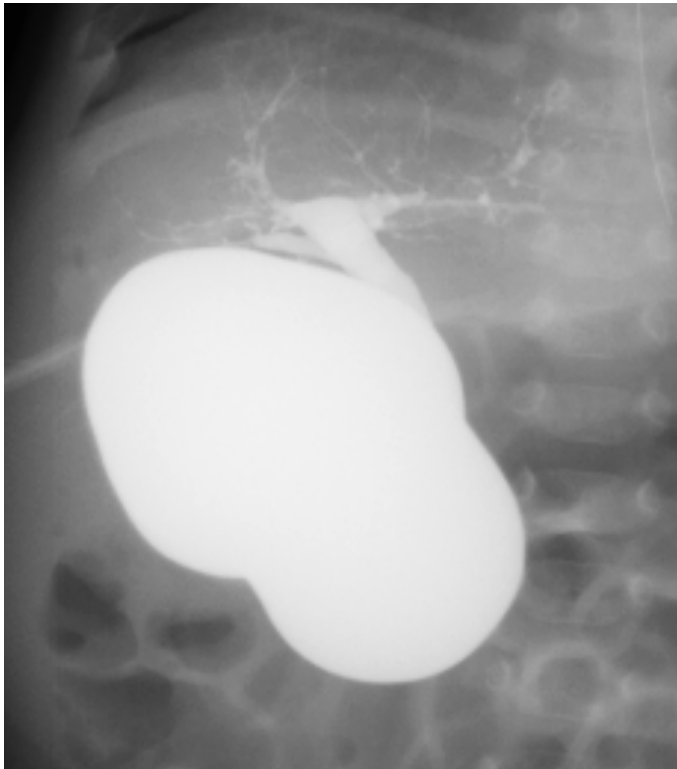


Type I cystic Biliary Atresia

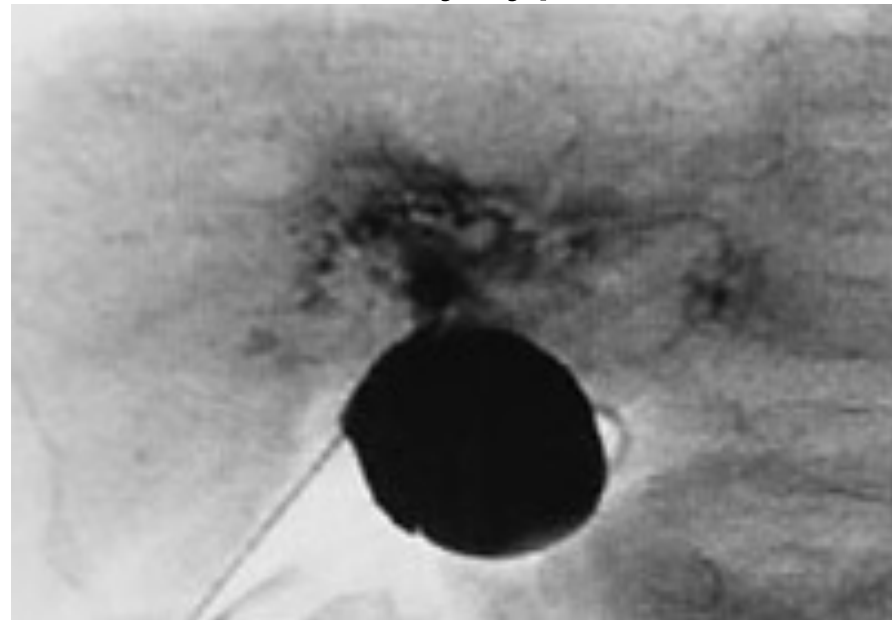
Cholangiography shows

- No flow into the duodenum
- Abnormal IHBD shapes

Treelike type



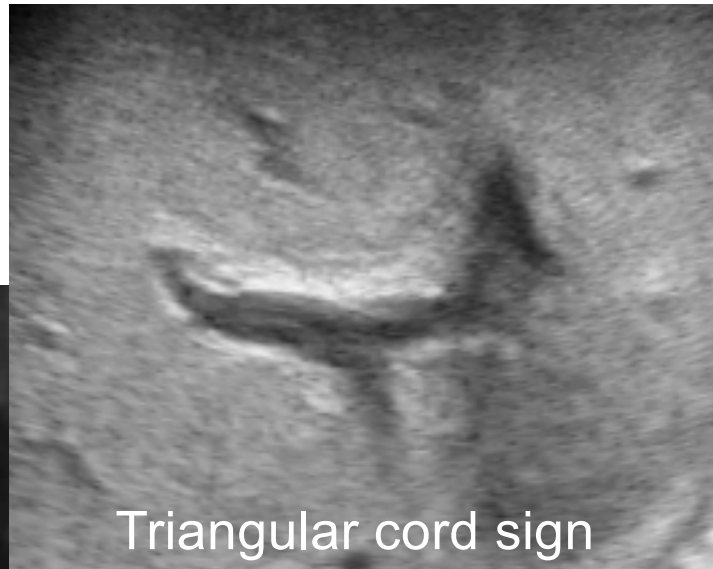
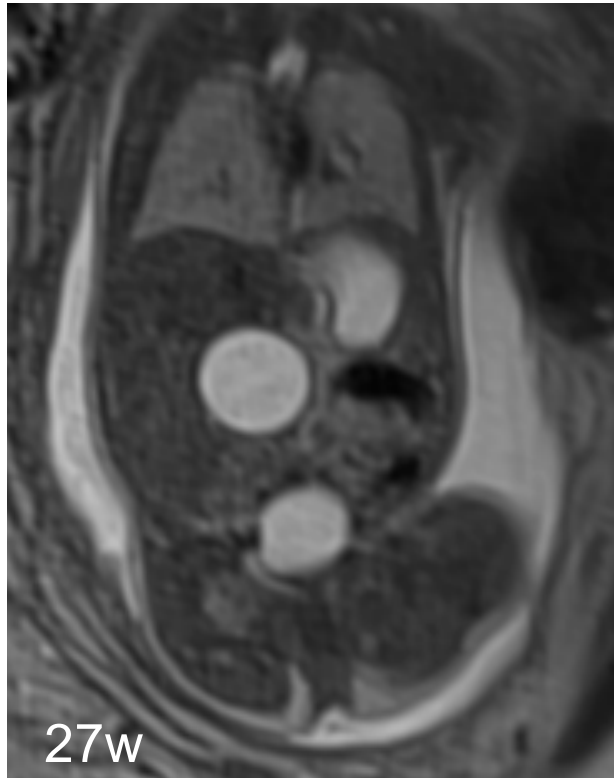
Cloudy type



Nio M, et al. Long-term outcome in type I biliary atresia.
J Pediatr Surg 2006;41:1973-5

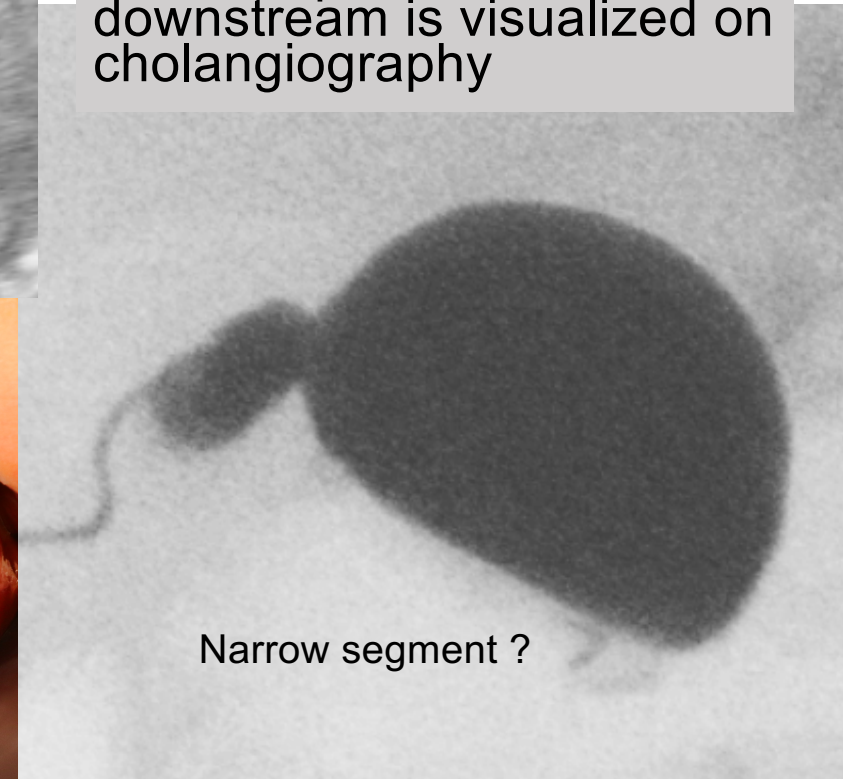
Case **44**

Type IIId BA



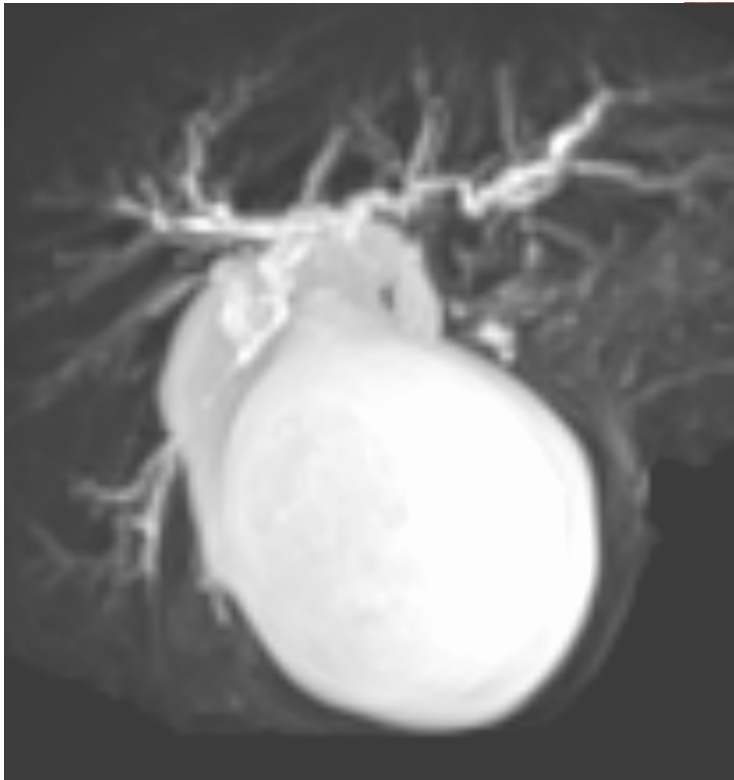
Atresia at the hepatic hilum
requires the Kasai operation

Neither upstream nor
downstream is visualized on
cholangiography



Case 45

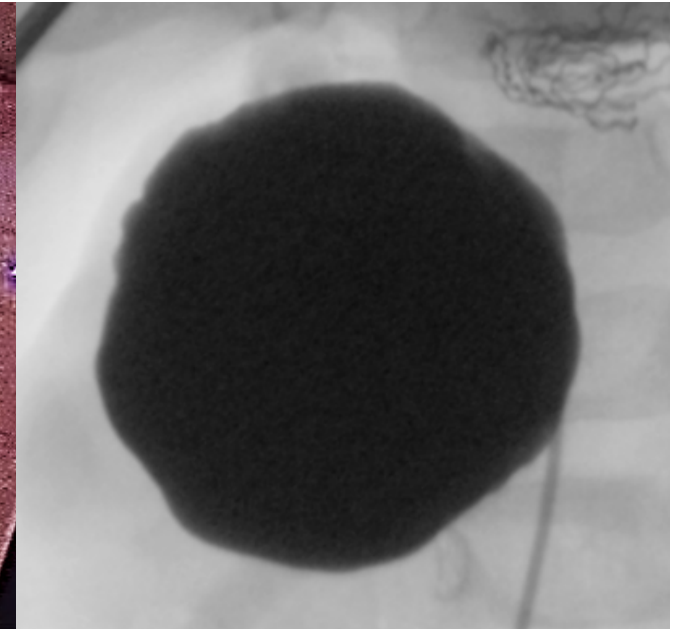
Type Ia CBD > Type I cyst BA



Almost normal IHBD



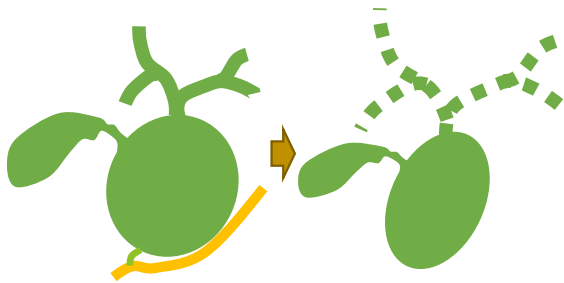
Makibishi sign



Contrast material did not enter
the duodenum

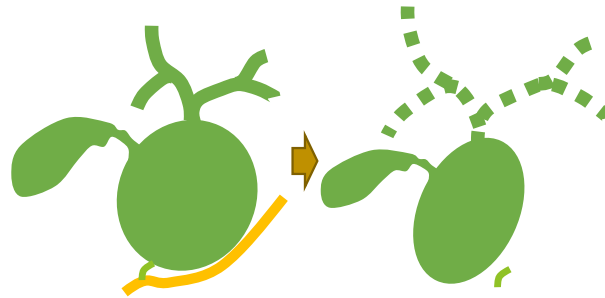
CBD that has features of BA
Intermediate / Transitional Type
Bile duct resection yields good result

There exists a transitional type from CBD to BA



CBD → III d BA

Caponcelli E, et al.
J Pediatr Surg 2008;43:1619



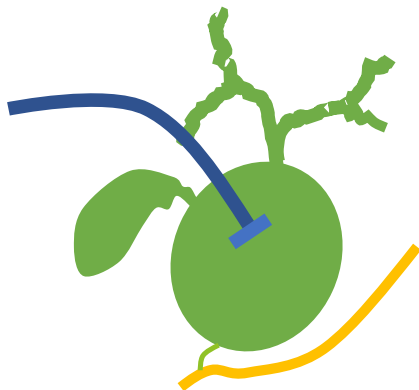
CBD → III d BA (+NS)

Koshinaga T, et al.
BMC Pediatrics 2018;18:158

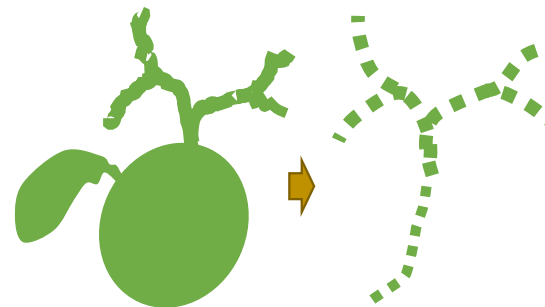


I cyst BA with a dilated LHD
Rapid progress of liver cirrhosis
that results in liver plantation
(The authors describes a type IV-A CBD)

Fumino S, et al.
Pediatr Surg Int 2013;29:1097



I cyst BA
After biliary drainage,
contrast material began to
enter the duodenum,
revealing a NS and PBM.
Suzuki T, et al.
J Hepatobiliary Pancreat Sci
2013;20:465

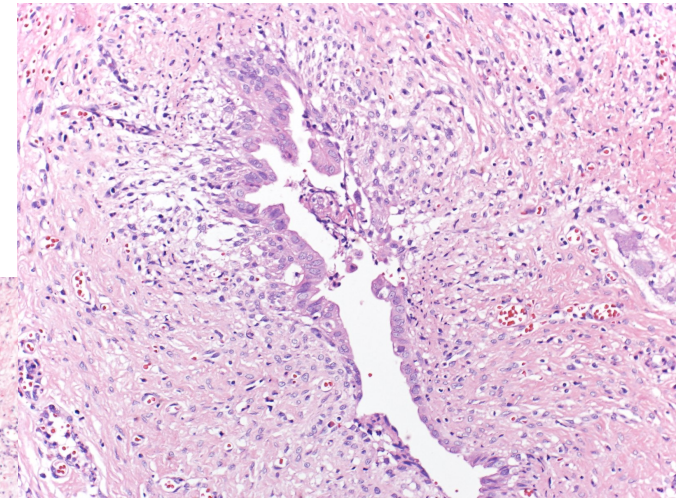


I cyst → III or III d

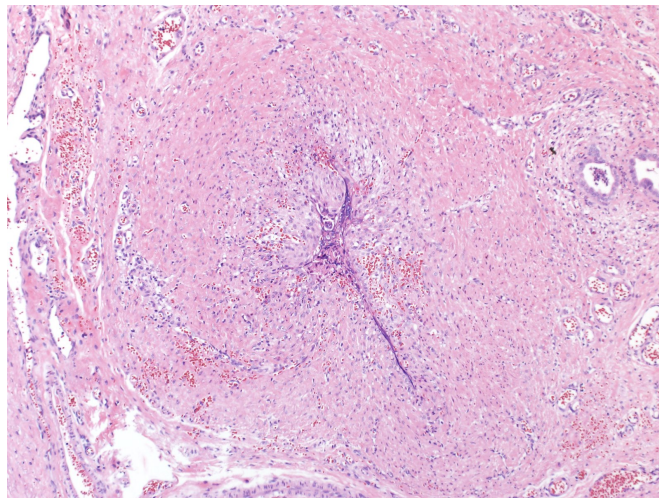
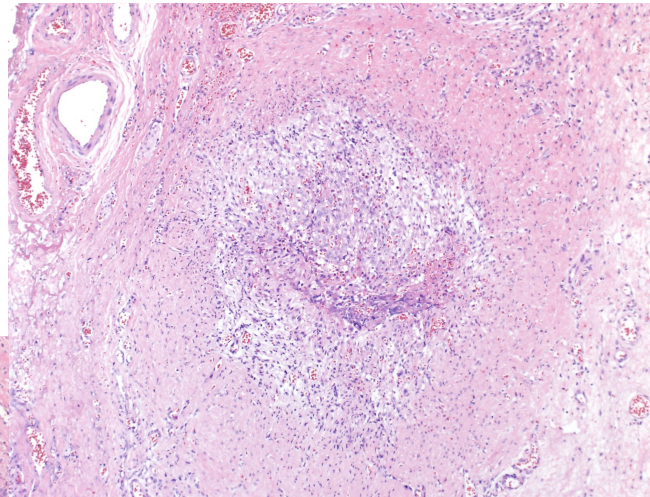
Nakata et al.
J Jpn Pediatr Surg 2000;36:332
Fujishiro J, et al.
Pediatr Sur Int 2005;21:116
Kawasaki et al.
J Jpn Pediatr Surg 2009;45:729
Masumoto K, et al.
Pediatr Sur Int 2011;27:99
Odashima et al.
J Jpn Pediatr Surg 2016;52:1042

Pathology of hilar remnant in type IIIId BA

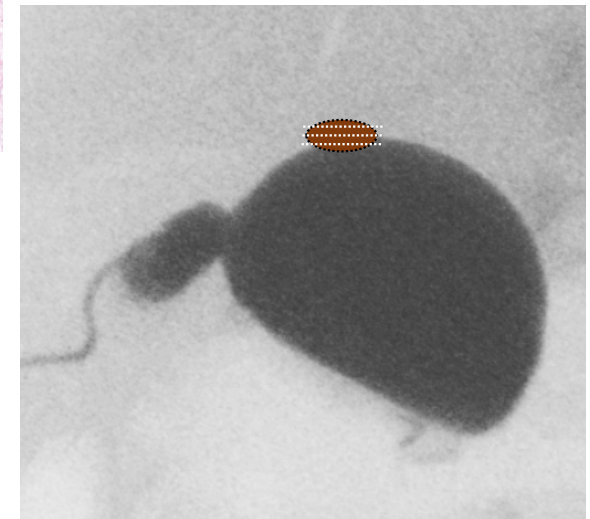
Obstruction progresses from downstream to upstream?



Upstream



Downstream



Hypothesis

Cystic BA originates from CBD with a NS

Infantile obstructive cholangiopathy progresses according to the degree of obstruction at the NS

