Pediatric Hepatobiliary Surgery

Part 1

Biliary Atresia

Part 2

Choledochal cyst

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Biliary Atresia

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1.1. Basics

1.2. Clinical Features

1.3. Management

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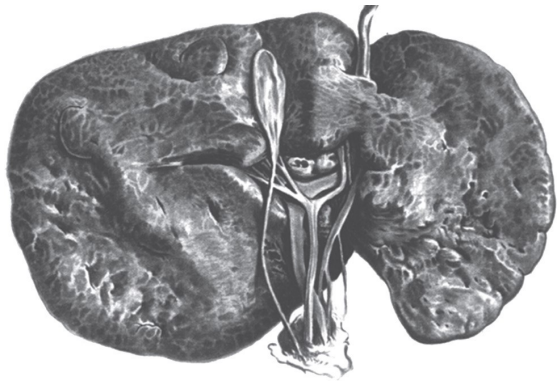
Definition

Congenital obliteration of bile ducts (panductular cholangiopathy)

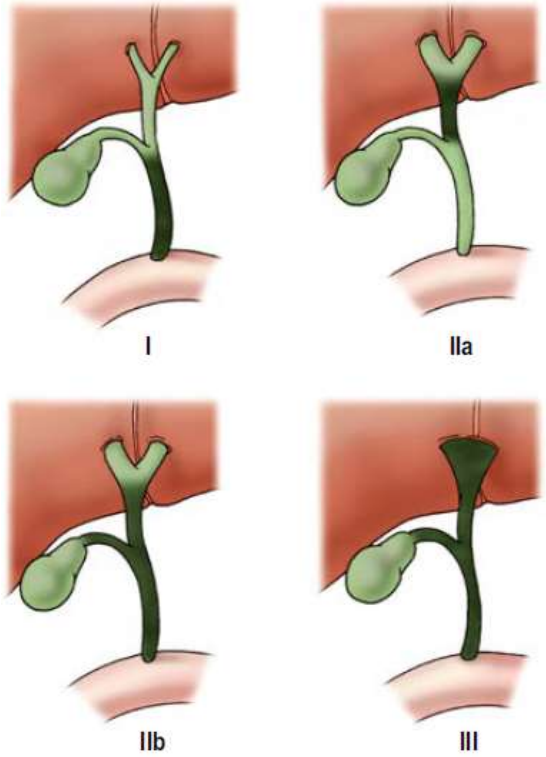
- **Extrahepatic ducts** are **atretic** (cone shaped fibrous mass ant to portal vein bifurcation)
- **Intrahepatic ducts** are **patent in early infancy** (progressive fibrosis)

***It is assumed that minute ducts are present in fibrous cone of tissue (basis of Kasi's procedure)*



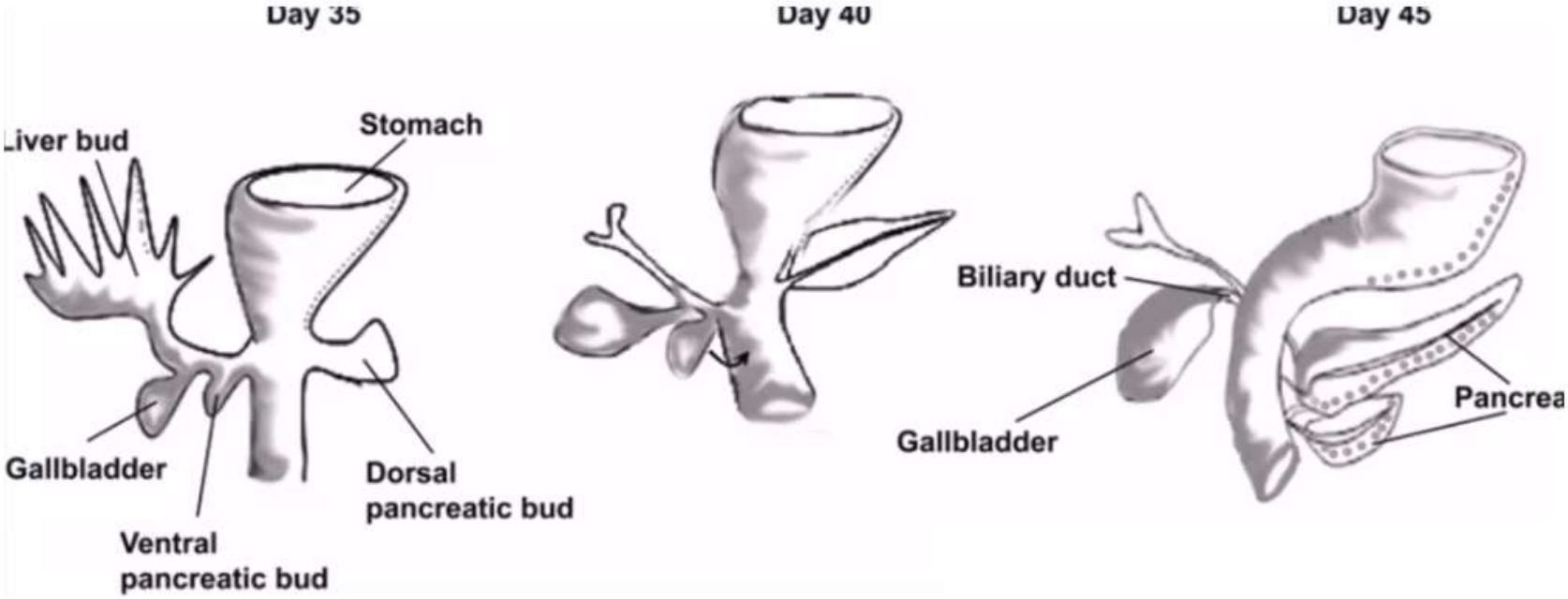
Classification

- 25% **"correctable"** (patent extrahepatic ducts)
  - Type I – atresia of CBD
  - Type II – atresia CHD (IIA- CBD spared, IIB- CBD involved)
- 75% **"non-correctable"** (atresia of all extrahepatic)
  - Type III – atresia upto porta hepatis
- 10% **Cystic variant –** *all types can have cystic component*
  - *causes confusion with CC*
  - May coexist with BASM
  - Detected earlier & better response to kasi

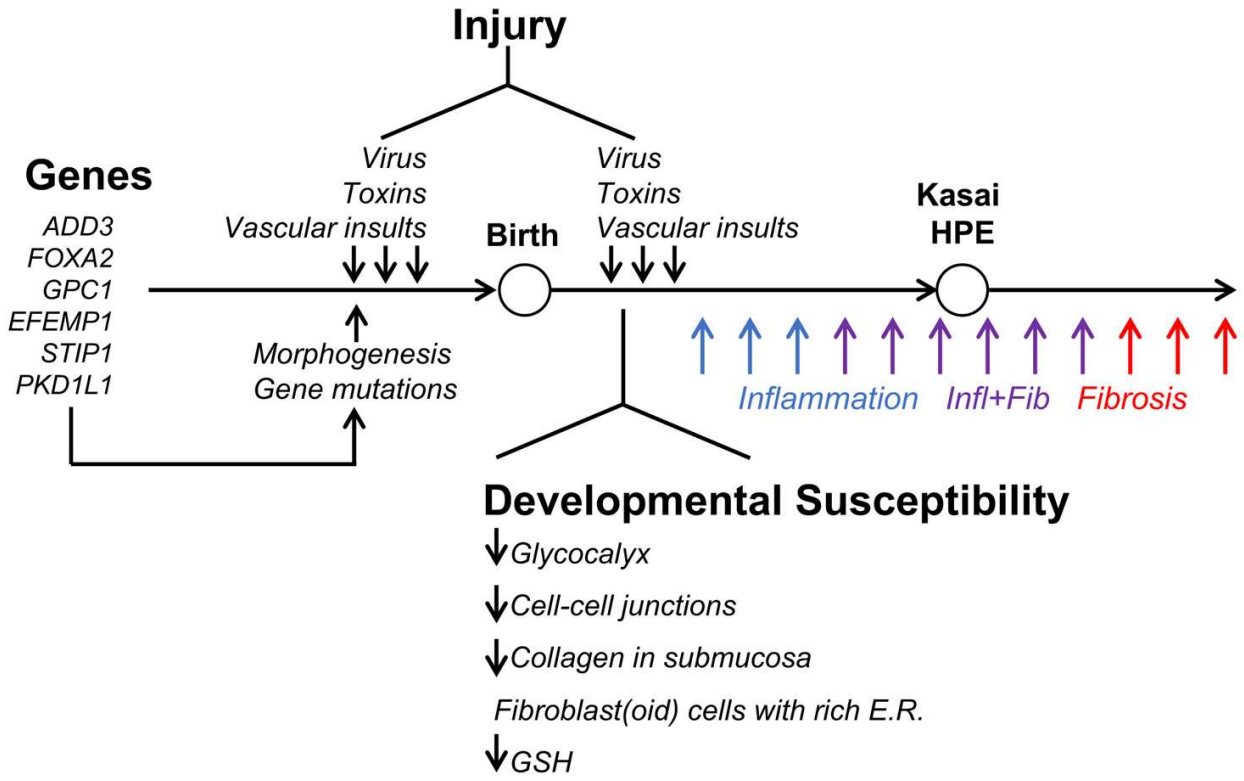


Etiology

- **Non syndromic (85%)** - jaundice-free period after birth
  - **Isolated BA**
  - **CMV variant** (10%) **-** Variable incidence based on geography, poor response to kasi
- **Syndromic (15%)** - Early onset, poor outcome after kasi
  - **BASM** (10%) – Polysplenia, situs inversus, preduodenal portal vein, absent IVC, CHD
  - **BA with non-laterality defects** (5%) - Cardiovascular, GI and GU malformations



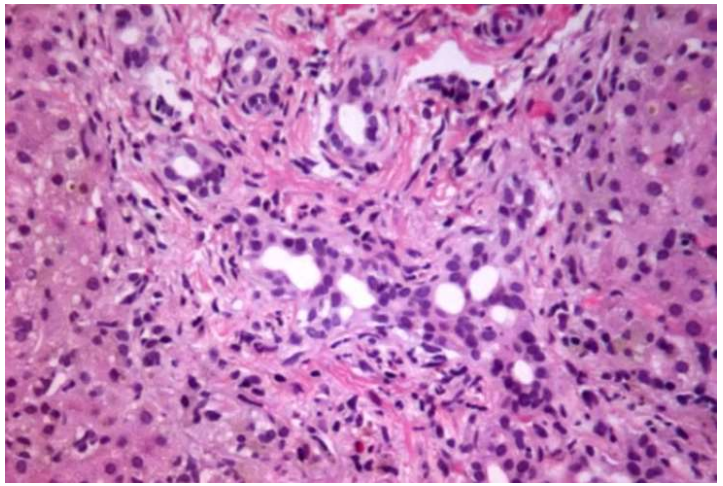
- **Embryologic theories**: theory of abnormal hepatic diverticulum differentiation is a possible explanation for syndromic BA. However, no mechanism is accepted for largest group of BA (isolated BA). A theory of developmental arrest has been proposed which assumes that intra & extrahepatic ducts develop from d/t sources and BA occurs if linkage doesn't occur at 10-12wk. This is supported by finding of porta hepatis in normal fetus at 12wk is similar to BA.



- **Factors implicated in development of BA**: Complex interplay between **genetic predisposition, virus triggers and autoimmunity.** Finally culminate in bile duct injury triggering inflammation and fibrosis typical of BA. Experimental studies have induced BA in mice by administering rotavirus.

Pathology

- Bile duct changes
  - Bile Duct proliferation (paucity in Alagile syndrome)
  - Bile plugs
- Portal tract changes
  - Portal Inflammation
  - Portal Edema
  - Portal fibrosis
- Parenchymal changes
  - Steatosis
  - Multinucleated giant hepatocytes
  - Extramedullary hematopoiesis



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Epidemiology

- Leading indication for liver transplant in children
- 1 in 10,000-20,000
  - Asian (1:5,000 in Taiwan)
  - Females (slight predominance)

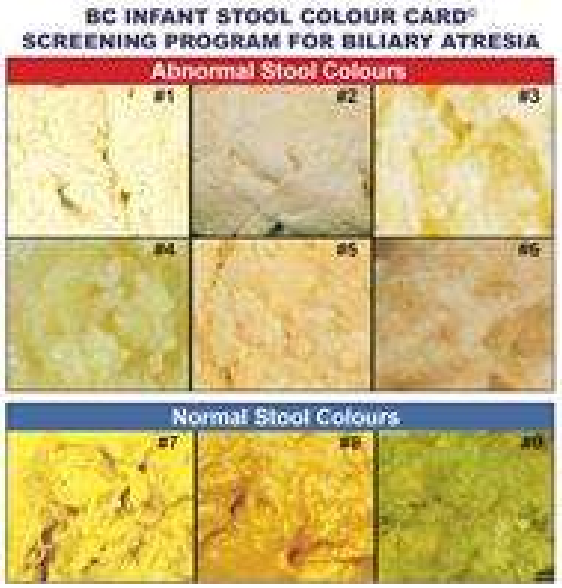
Presentation

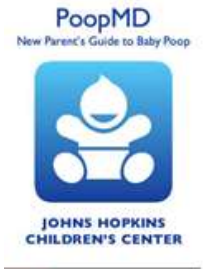
Classically term, NBW, Presents in first few weeks of life

- **Jaundice** (progressive)
- **acholic stool** (pale, grey, clay color)
  - Screening in Asia with Stool card (now has an app- PoopMD)
- **Dark urine**
- **Examination**

  *Palpate liver **(**hardness)

  ** Duodenal aspiration (bilious aspirate r/o BA)*





DDx: Obstructive Jaundice

Medical

- TORCH infections
- Cystic fibrosis
- A1 antitrypsin deficiency
- Alagille syndrome
- Familial cholestasis syndromes
- TPN ass. cholestasis

Surgical

- Biliary atresia (80%)
- Obstructive CC (10%)
- Inspissated bile syndrome (5%)
- Spontaneous perforation of bile duct (<2%)

Labs

- LFT
  - Bilirubin (Direct >2, total may be normal)
  - Inc ALP & GGT (if low can r/o BA)
- Rule out other causes
  - A1 antitrypsin
  - TORCH titer
  - Other metabolic (CF, hypothyroidism, galactosemia)

SCIENCE TRANSLATIONAL MEDICINE | RESEARCH ARTICLE

BILIARY ATRESIA

Large-scale proteomics identifies MMP-7 as a sentinel of epithelial injury and of biliary atresia

Chatmanee Lertudomphonwanit,[1,2] Reena Mourya,[1] Lin Fei,[3] Yue Zhang,[3]* Sridevi Gutta,[1] Li Yang,[1,4] Kevin E. Bove,[5] Pranavkumar Shivakumar,[1] Jorge A. Bezerra[1]†

HEPATOLOGY

AASLD

RAPID COMMUNICATION | HEPATOLOGY, VOL. 68, NO. 6, 2018

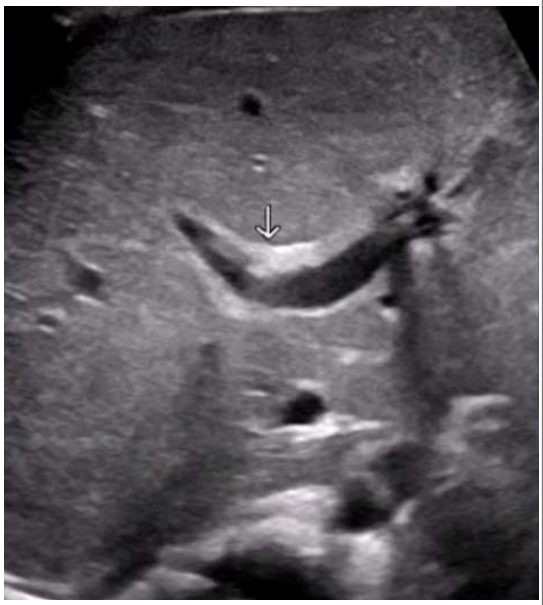
Diagnostic Accuracy of Serum Matrix Metalloproteinase-7 for Biliary Atresia

Li Yang,[1,2]* Ying Zhou,[1]* Pei-pei Xu,[1] Reena Mourya,[2] Hai-yan Lei,[3] Guo-qing Cao,[1] Xiao-li Xiong,[4] Hui Xu,[1] Xu-fei Duan,[3] Na Wang,[4] Lin Fei,[5,7] Xiao-pan Chang,[1] Xi Zhang,[1] Meng Jiang,[1] Jorge A. Bezerra,[2] and Shao-tao Tang[1]

- **MMP7 (matrix metalloproteinase 7):** A large scale proteomic analysis of around 30 BA patients in USA (2017) identified serum MMP7 level as a marker of epithelial injury in BA. The finding was validated in China (2018) with 135 patients being worked up for cholestatic jaundice. The cut off value of 52.85ng/dl had sensitivity of 99% and specificity of 95% (?better than GGT)

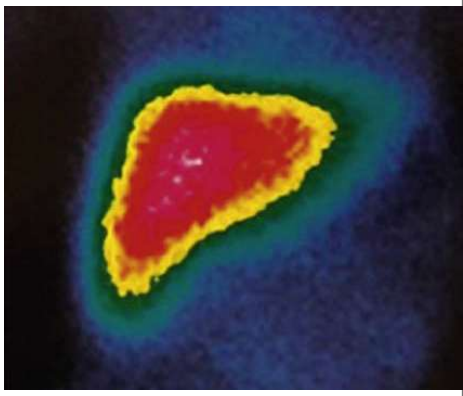
Imaging

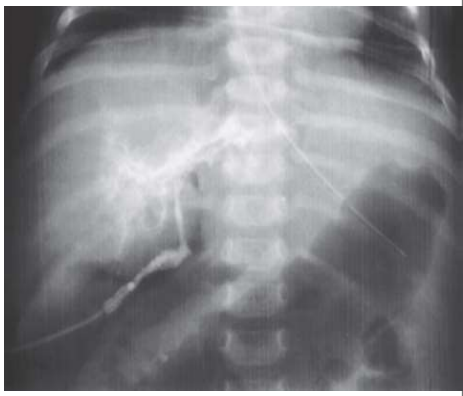
- **Ultrasound**
  - **No Biliary dilatation** unlike other obst (intrahepatic also affected by inflmn)
  - **GB abnormal (irregular) or absent**
  - **Triangular cord sign** (inc echogenicity at location of CHD at porta hepatis)
  - Other ass. Anomalies (polysplenia syndrome)
- **?MRCP** role is less in BA (used to see dilatation or cyst. not obliteration)



Additional workup

- **HIDA Scan** (For questionable diagnosis like in prolonged TPN)
  - presence of excretion r/o BA (But absence can't reliably diagnose)
  - needs pre-medication with phenobarb for 5 days (delays surgery)
- **Percutaneous Biopsy** (UK group, diagnostic in 85%)
  - False positive (TPN, A1 antitrypsin), equivocal (hepaitis),
  - false negative (early age (4-6wk), inadequate sample)
- **ERCP** (UK group, if biopsy suspicious but not diagnostic)
  - Invasive (requires GA)
- **Laparoscopy assisted cholangiography** (Japanese group, avoids biopsy)
  - Gold standard is direct observation of porta hepatis & cholangiography
  - Cholangiography can also be done percutaneous to avoid laparotomy





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Timing of surgery

- **Urgent -** Before 2mo (cirrhosis will occur at 3-4mo)

Preop preparation

- **Vitamin K** 1-2mg/kg for several days
- **Bowel preparation** dec intestinal gas
  - PO antibiotic (kanamycin)
  - Enema (glycerine)
  - early NPO (24hr)

Surgical options

Hepatico-enterostomy: if large enough CHD (>5mm)- 20%

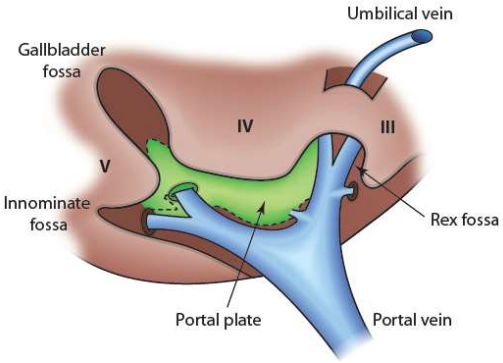
Porto-enterostomy (Kasi procedure): required in 80-90%

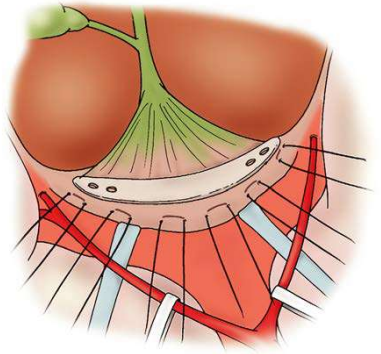
***cyst-enterostomy (**in cystic BA) is being abandoned b/c it has been shown cyst doesn't have epithelium (stricture, cholangitis)

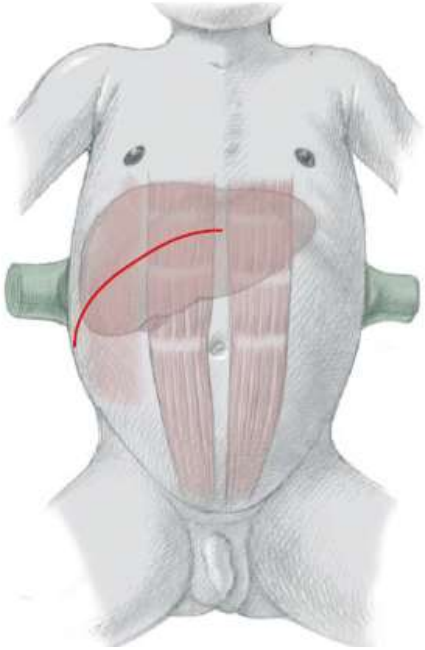
***GB–porta hepatis anastomosis** can be done inn type IIA (5%)

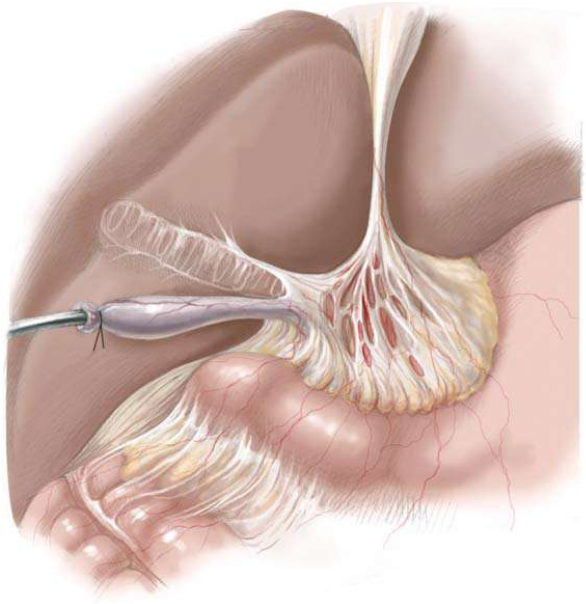
Surgical principles

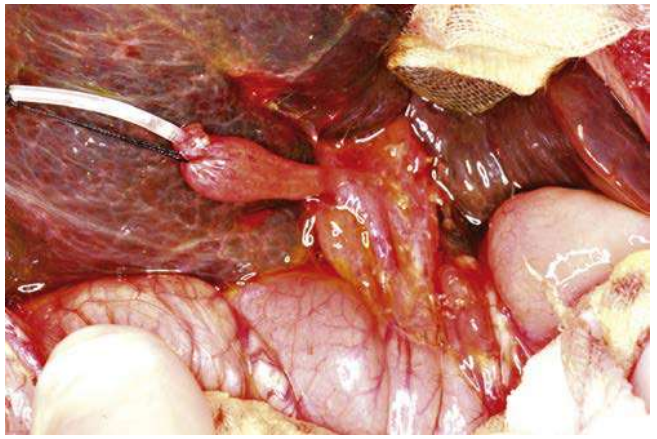
- Remove all extrahepatic bile ducts (+fibrous remnants)
  - **dissection should be deep and wide** (deep until liver capsule, laterally up to where arteries divide into segmental branches)
- intestine anastomosed to portal plate
  - **Sutures should be superficial, avoid transected surface, avoid 2 & 10 O'clock** (micro ducts may be present)



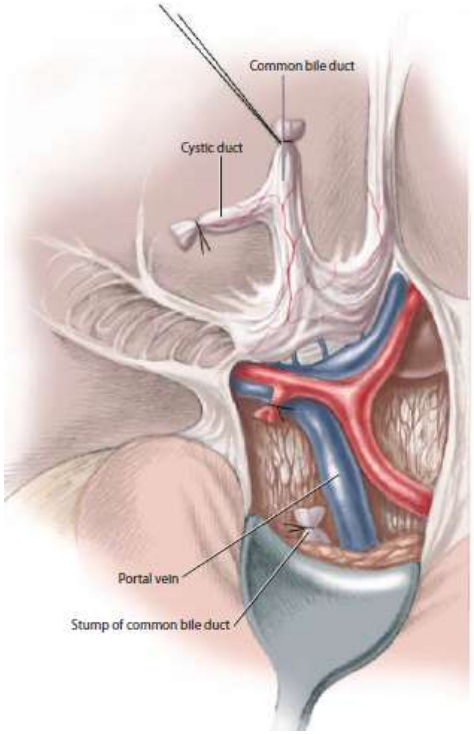


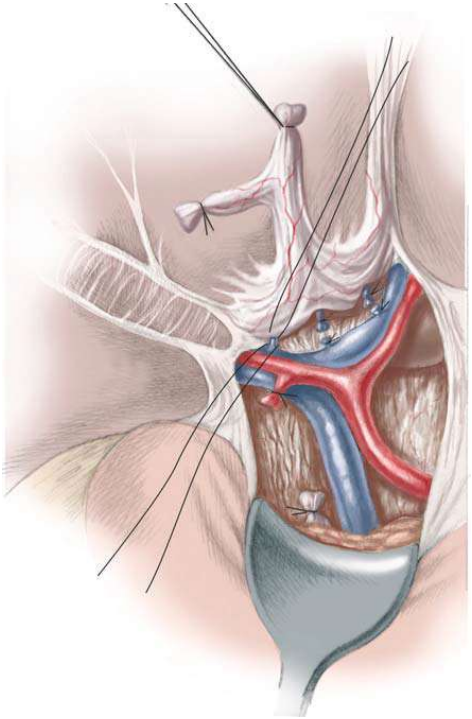


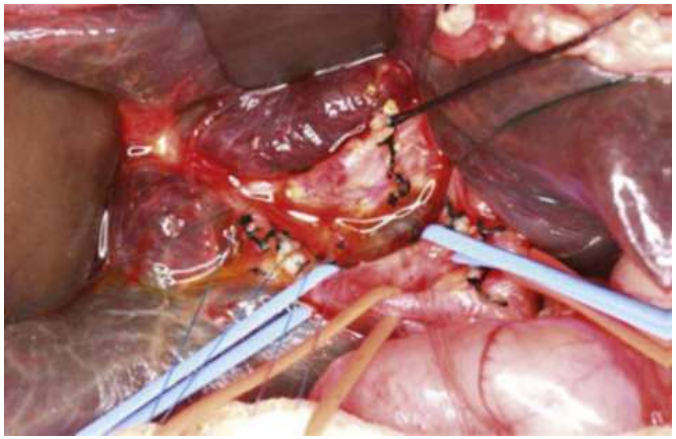




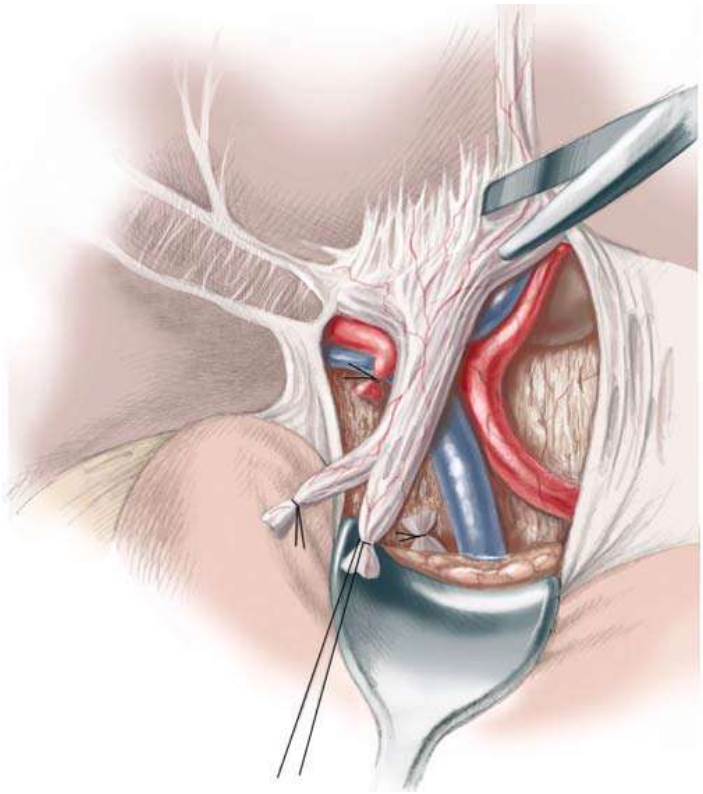
- **Exposure**: Supine, insert pillow, right subcostal incision (flank to midline). May mobilize & **exteriorize liver** to allow adequate visualization. Check for cirrhosis & portal HTN. Liver is enlarged, firm & green. **Liver biopsy** is taken. GB may be atretic or hypoplastic & filled with white mucus. **Cholangiogram** performed via 4Fr NGT if there is bile in GB (to confirm occlusion of extrahepatic ducts). GB is freed from liver and dissection advanced toward CHD.

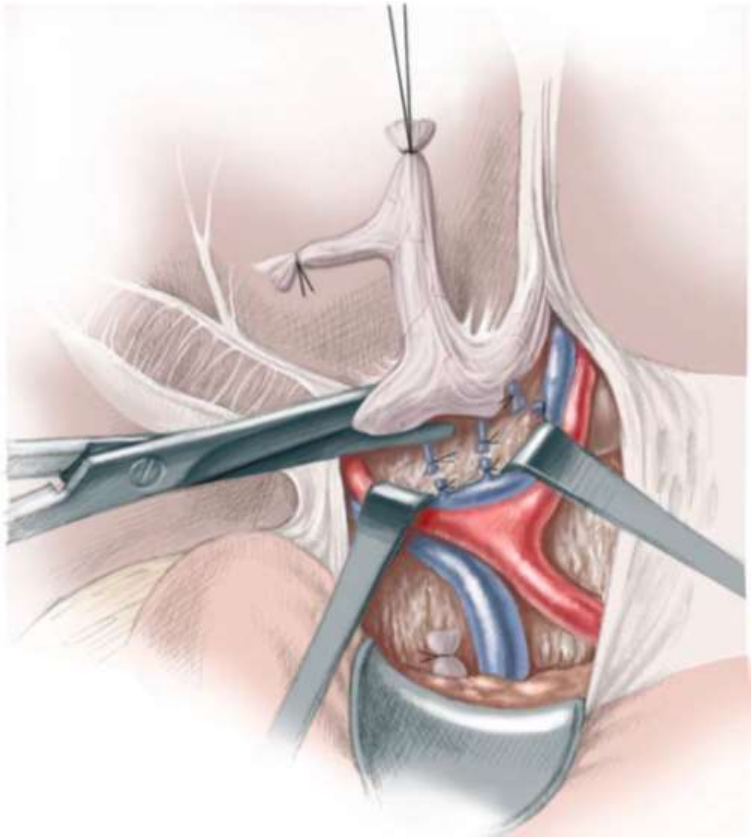






- **Porta hepatis dissection (posterior) :** CBD remnant is carefully dissected and divided adjacent to duodenum. CHD remnant (cone shaped fibrous tissue cranial to bifurcation of portal vein) is freed from portal vein and hepatic arteries (vessel loop). The posterior aspect of the fibrous remnants is exposed deep and wide enough behind the bifurcation of the portal vein. Small branches bridging from the portal vein to the fibrous remnants are identified and divided.





- **Porta hepatis dissection (anterior) & Transection :** sharp dissection continued b/n fibrous remnant and quadrate lobe of liver. Dissection should be as wide as possible (right - ant branch of rt hepatic artery upto left – where obliterated umblical vein joins Lt portal vein). Finally fibrous remnants are transected carefully at level of posterior surface of portal vein. If hemorrhage occurs use only irrigation with warm saline for 10 min. (**no ligation/cautery**)

Pediatr Surg Int (2015) 31:1177–1182
DOI 10.1007/s00383-015-3799-4

ORIGINAL ARTICLE

Near-infrared fluorescence cholangiography with indocyanine green for biliary atresia. Real-time imaging during the Kasai procedure: a pilot study

Yutaka Hirayama[1] · Yasushi Iinuma[1] · Naoyuki Yokoyama[2] · Tetsuya Otani[2] · Daisuke Masui[3] · Naoko Komatsuzaki[3] · Naruki Higashidate[3] · Shiori Tsuruhisa[3] · Hisataka Iida[1] · Kengo Nakaya[1] · Shinichi Naito[1] · Koju Nitta[1] · Minoru Yagi[3]

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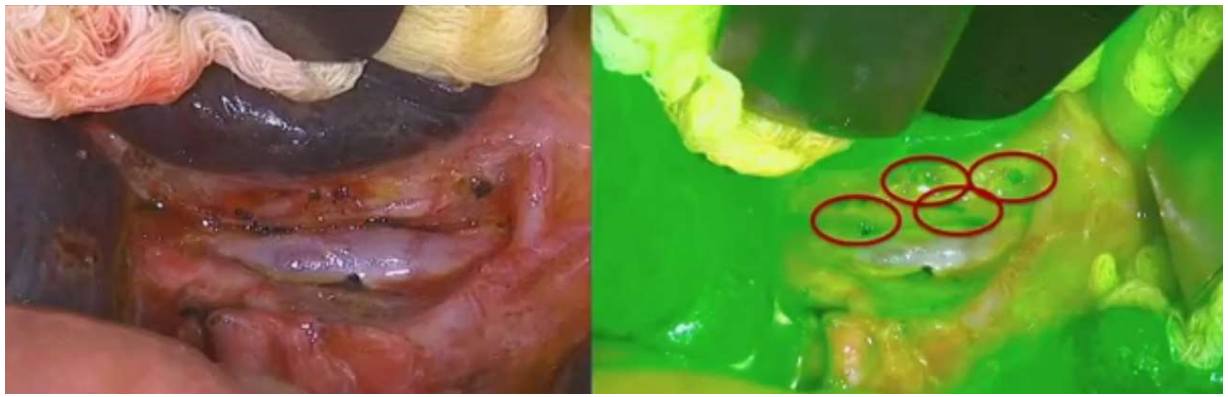
Journal of Pediatric Surgery

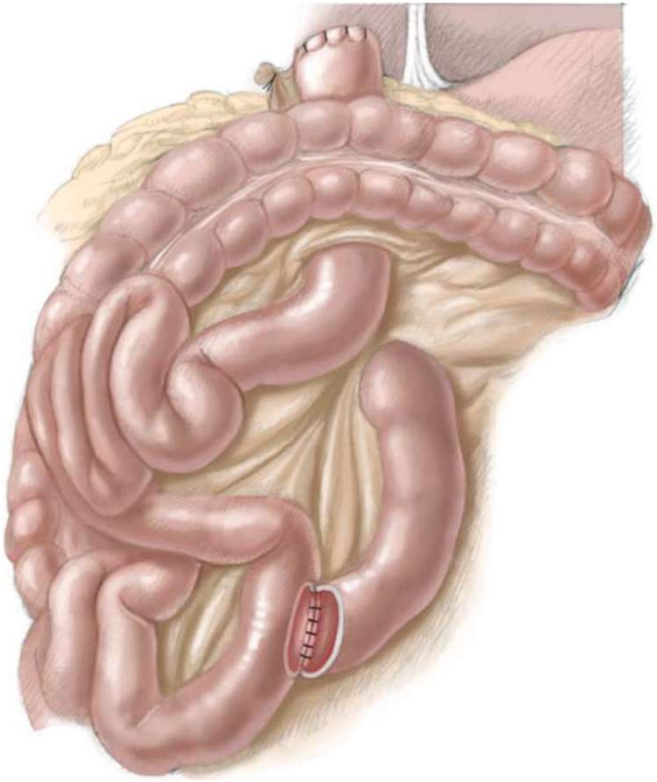
journal homepage: www.elsevier.com/locate/jpedsurg

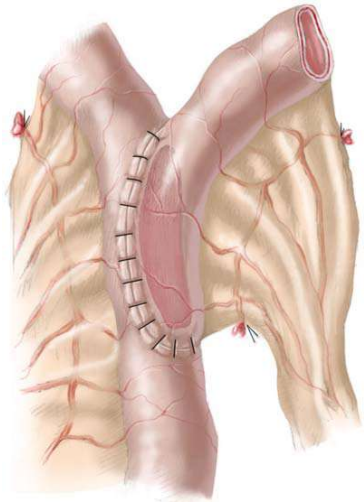
The outcome of real-time evaluation of biliary flow using near-infrared fluorescence cholangiography with Indocyanine green in biliary atresia surgery

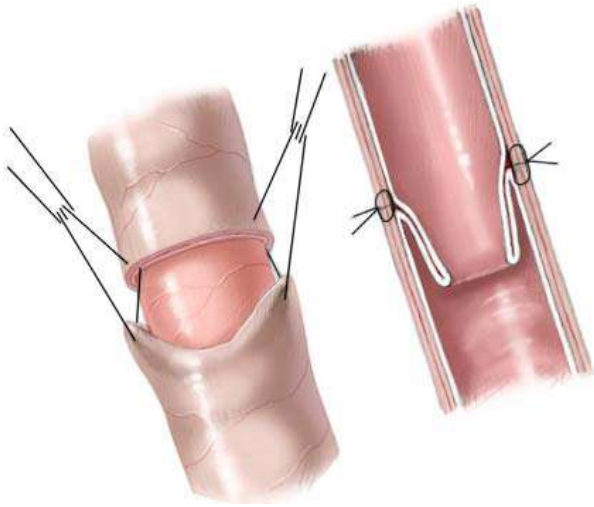
Yusuke Yanagi [a,*], Koichiro Yoshimaru [a], Toshiharu Matsuura [a], Yuichi Shibui [b], Kenichi Kohashi [b], Yoshiaki Takahashi [a], Satoshi Obata [a], Ryota Sozaki [a], Tomoko Izaki [a], Tomoaki Taguchi [a]



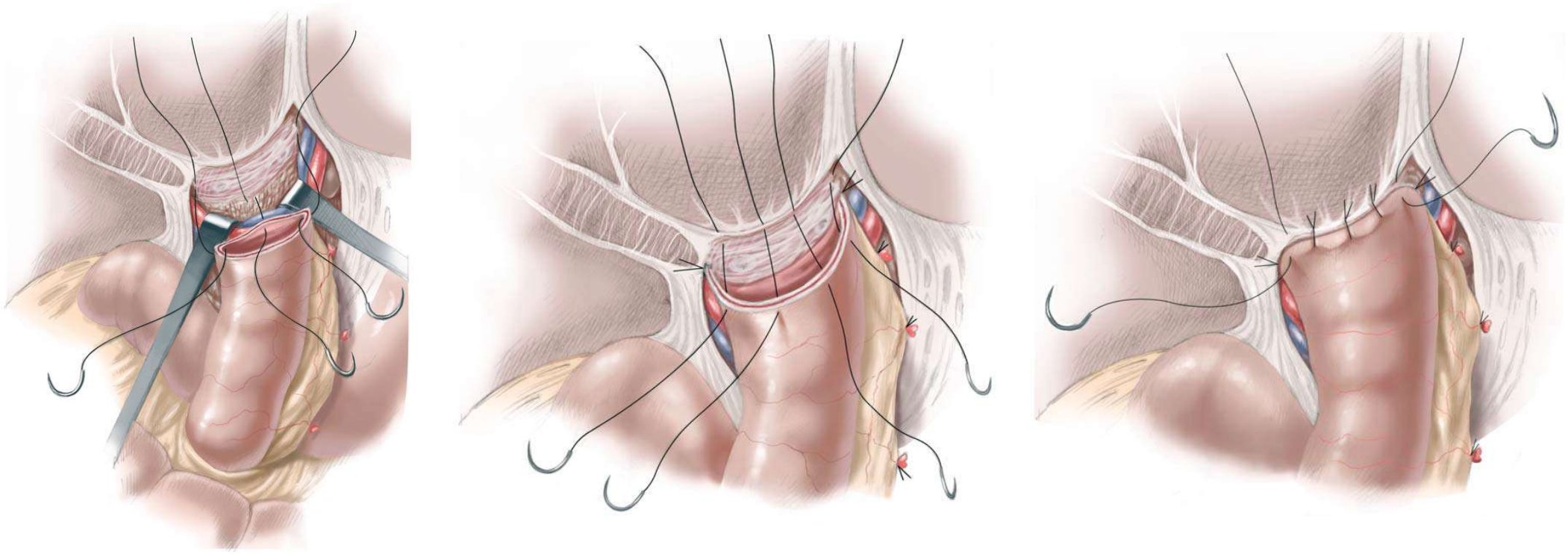
- **Indocyanine green as an adjunct during dissection:** to demonstrate weather or not hilar plate is adequately transected (weather there are patent biliary radicles at fibrous cone). If there is no fluorescence, cut into the plate further until there is fluorescence. The whole liver may be green during fluorescence b/c of biliary stasis but the more intense areas correlate with patency.







- **Preparation of Roux jejunal limb:** ***Details are discussed in next section (choledochal cyst).***
- Long jejunal roux limb (50-70 cm) may decrease chance of cholangitis. *The* Japanese group also perform a double-valve (spur valve and intussuscepted valve) hepatic porto-jejunostomy. However, some say despite modifications rate of cholangitis is similar to the original kasai.



- **Porto-jejunostomy**: full thickness 5-0/6-0 absorbable sutures used to anastomose intestine **to the parenchyma around transected end** of fibrous remnants at porta hepatis (Careful not to place stiches on transected surface). After all posterior sutures are placed, they are tied. Anterior row of sutures placed in same manner. Additional interrupted 5-0 silk seromuscular sutures are used to fix the jejunum at porta hepatis. (from anterior wall to quadrate lobe and from posterior wall to hepatoduodenal ligament). Penrose drain is placed in foramen of winslow.

Postop care

- **Nutritional optimization -** Maintain good weight gain
  - Supplement Vit ADEK
- **Antibiotic** - used by most but protocol is different (prevent inflammation)
  - **Type** (Amikacin/ Amoxacilin/ cephalosporin/ Bactrim)
  - **Duration** (3mo/6mo/ until bilirubin <2/ until CRP <0.3/ until leukocytosis resolve)
- **Steroids (**prednisolone) – use is controversial. (cohleretic + prevent inflammation),
  - Some start empirically (some immediately postop, some start with PO feeding ~ 4d)
  - some start only if no response a few days after kasai
  - some use for cholangitis not responding for antibiotics
- **Cholertics (**ursodiol) – adjunct (to improve drainage)
- **Antiviral** (valgangciclovir)–for IgM positive CMV infection

Research

Original Investigation

Use of Corticosteroids After Hepatoportoenterostomy for Bile Drainage in Infants With Biliary Atresia
The START Randomized Clinical Trial

Jorge A. Bezerra, MD; Cathie Spino, DSc; John C. Magee, MD; Benjamin L. Shneider, MD; Philip Rosenthal, MD; Kasper S. Wang, MD; Jessi Erlichman, MPH; Barbara Haber, MD; Paula M. Hertel, MD; Saul J. Karpen, MD; Nanda Kerkar, MD; Kathleen M. Loomes, MD; Jean P. Molleston, MD; Karen F. Murray, MD; Rene Romero, MD; Kathleen B. Schwarz, MD; Ross Shepherd, MD; Frederick J. Suchy, MD; Yumirle P. Turmelle, MD; Peter F. Whitington, MD; Jeffrey Moore, MS; Averell H. Sherker, MD, FRCP(C); Patricia R. Robuck, PhD, MPH; Ronald J. Sokol, MD; for the Childhood Liver Disease Research and Education Network (ChiLDREN)

Corresponding Author: Jorge A. Bezerra, MD, Cincinnati Children's Hospital Medical Center, 3333 Burnet Ave, Cincinnati, OH 4522 (jorge.bezerra@cchmc.org).

Supplemental content at jama.com

Pediatr Surg Int (2016) 32:193–200
DOI 10.1007/s00383-015-3836-3

CrossMark

REVIEW ARTICLE

Steroids after the Kasai procedure for biliary atresia: the effect of age at Kasai portoenterostomy

Athanasios Tyraskis[1] · Mark Davenport[1]

✉ Mark Davenport
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[1] Department of Paediatric Surgery, King's College Hospital, Denmark Hill, London SE5 9RS, UK

- **Steroids in BA**: ** USA study didn't find difference in bile drainage with high dose steroid. The study from UK showed benefit of steroids affecting liver damage (AST, APRi) and restoring bile flow. However, efficacy of adjuvant steroid was associated with younger age at KPE.

Contents lists available at ScienceDirect

Journal of Pediatric Surgery

journal homepage: www.elsevier.com/locate/jpedsurg

Adjuvant therapy of cytomegalovirus IgM + ve associated biliary atresia: Prima facie evidence of effect☆,☆☆,★

Filippo Parolini, Nedim Hadzic, Mark Davenport *

Department of Paediatric Surgery, Denmark Hill, London, UK SE5 9RS
Department of Hepatology Kings College Hospital, Denmark Hill, London, UK SE5 9RS

- **Adjuvant therapy for CMV**: UK study (2019) found anti-CMV therapy improved success after Kasi in CMV IgM+ve BA patients. Improved jaundice clearance (21% vs 75%) and transplant (57% vs 25%)

Follow-up

2nd wk post op, then every 6 wk for 6 mo

- Weight check, Vitamin D level
- LFT (Bilirubin, liver enzyme, albumin, Coagulation profile)
- AFP
- Ultrasound (check for cirrhosis and portal htn)

***Prevent inflammation first 4 wk** (until biloenteric fistula develop)- antibiotic, steroid, choleretic

- Bilirubin can worsen 1st week
- pigmented stools and a definite fall in bilirubin should occur by 4th wk (? total <1.5)

Outcome

Survival used to be 0% >> 50% with Kasi only and 90% with transplant

- **Success depends on**
  - Age at kasi (most morbidity after kasai is due to the damage that already occurred preop)
  - Degree of liver fibrosis (and presence of microscopic ducts at porta hepatis)
  - presence of cholangitis
- **Failure to clear jaundice**
  - Never cleared jaundice (3mo post kasi)- require Liver transplant by end of 1st year of life.
  - Raising bilirubin after initial normalization - try medical first, revise Kasai if persist
- **Cholangitis** (40-50%): can result in fatal septicemia or re-obliteration of hepatic portoenterostomy.
  - ***Consider roux loop obstruction if cholangitis occurs years later after successful kasi*
- **Cirrhosis, Portal HTN, hypersplenism:** fibrosis might continue despite successful Kasi.
- **Other** – intrahepatic bile lake cysts, hepatic malignancy,

	Period	*n*	Age at Kasai (D) (median/mean)	Clearance jaundice (%)	4- to 5-year native liver survival	4- to 5-year true survival
Centralised Series						
England and Wales (45)	1999–2009	443	54	56%	46	90
Decentralised National Series						
France (48)	2003–2009	329	59	33%–39%[a]	33–39[a]	85–92[a]
Swiss (49)	1994–2004	48	59	n/a	37	91
Netherlands (51)	1987–2008	214	214	n/a	46	73
Canada (52)	1996–2002	150	55	n/a	39%	82
Multicentre, Not National						
United States (54)	1997–2000	104	61[a]	~40%[b]	n/a	n/a
North America (18)	2004–2010	159	62	46	n/a	n/a
National Asian						
Japan (53)	89–98	1381	65	57–62[c]	52–62[d]	70–78[d]

- **Experience affecting outcome** : highest jaundice clearance and native liver survival in Japan (case volume). UK also has comparable outcome because cases are centralized (only 3 centers manage BA)

Difficult scenarios

Management of late presenters (after 3mo)

- Determination of liver function is important (high morbidity & low success of kasai)
  - **no signs of cirrhosis on U/S** – laparotomy and see the liver to decide
  - **signs of cirrhosis on U/S** (heterogenous surface, ascites) – plan for transplant if available

  If transplant not available* - *go ahead with Kasi (accepting the risk)

- Determination of age at onset should also be considered (may be acquired BA)
  - **If pale stool started at a later age** - go ahead with Kasi if time from onset is <2mo

Choledochal Cyst

2

2.1. Basics

2.2. Clinical Features

2.3. Management

Choledochal Cyst

2

2.1. Basics

2.2. Clinical Features

2.3. Management

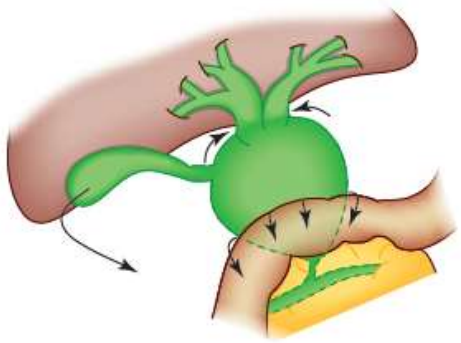
Nomenclature

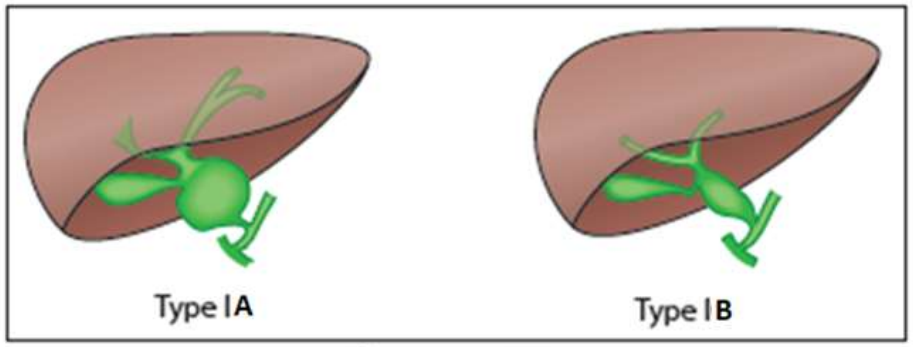
- **Choledochal cyst:** original terminology
- **Choledochal malformation:** b/c not all are cystic/round/globular (can be fusiform, spherical, cylindrical)
- **Biliary cysts :** b/c include intrahepatic cyst

Definition

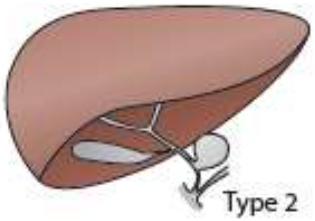
- Congenital dilatation of biliary tract
- Can occur anywhere but most common site is the CBD
- Consider when CBD diameter is greater than upper limit for age

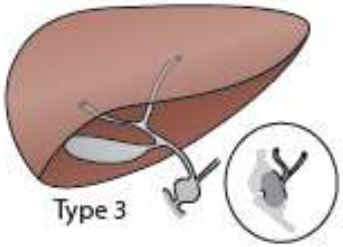
Age	Max. CBD diameter
<6 yr	4 mm
6-12 yr	6 mm
>12 yr	7 mm

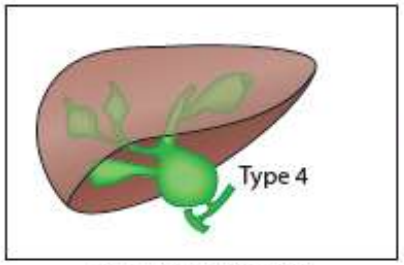




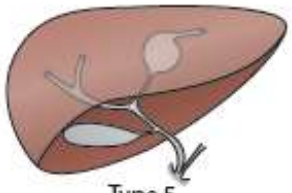
Common ~ 80%







Common 10%–20%



- **Todani Classification**: type I (80-90%) CBD dilatation (cystic/fusiform), type II CBD diverticulum
  type III Choledochocele, type IV multiple cysts, type V intrahepatic cysts (Caroli's disease)

*Forme fruste (common channel syndrome): pancreaticobiliary malunion with little/no dilatation (similar presentation)

Contents lists available at ScienceDirect

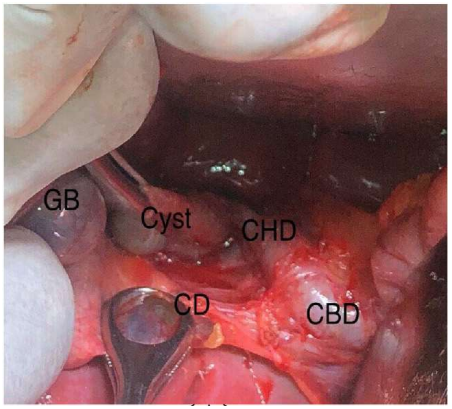
International Journal of Surgery Case Reports

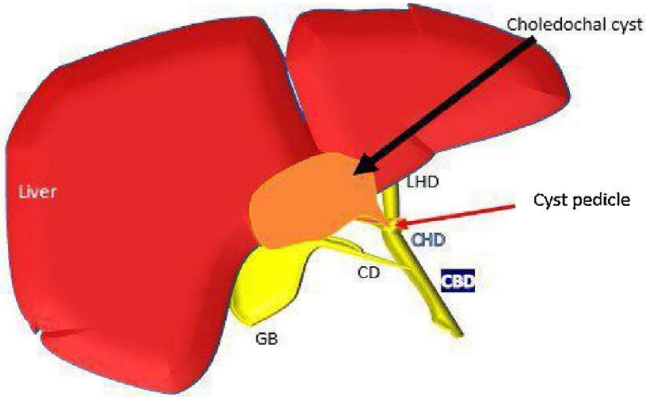
journal homepage: www.casereports.com

Unusual variant of choledochal cyst in a child: A case report, in Tertiary Specialized Hospital, Ethiopia

Workye Tigabie, Hadush Tesfay, Dagnachew Tamrat, Kassahun Raya, Tihitena Negussie*

Addis Ababa University, College of Health Sciences, Ethiopia



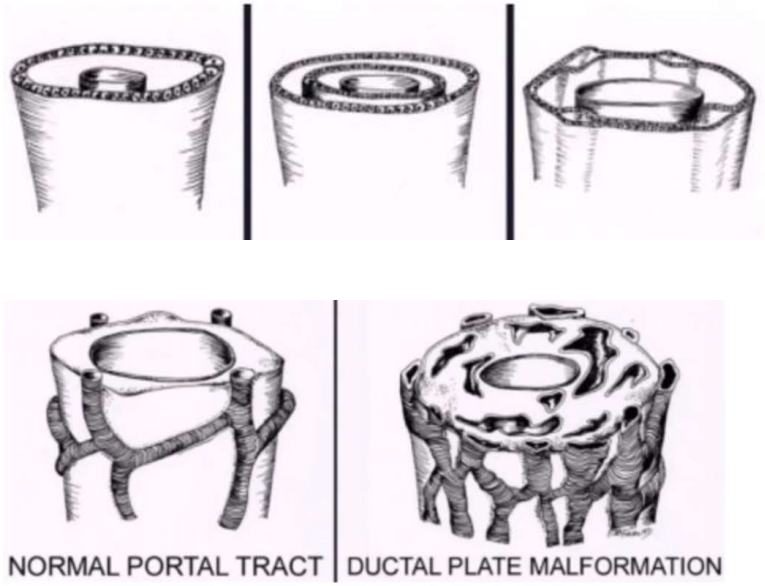


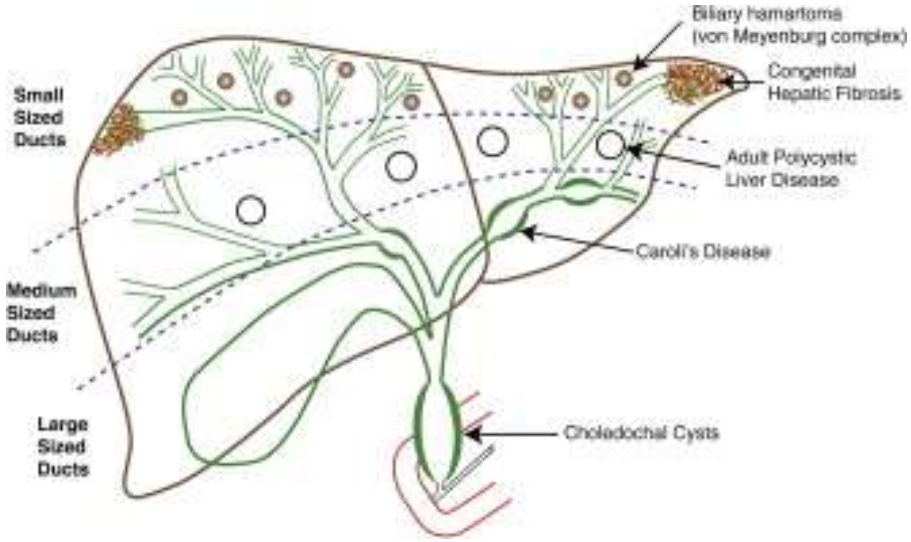
- **Report from Ethiopia**: variant of CC with diverticula arising from CHD instead of CBD. Cyst excision and primary closure done.

Etiology

No theory explains all five types (may be multiple etiology like biliary atresia)

- **Type I, IV**
  - ***"The obstructing segment hypothesis"*** (Distal CBD stenosis)
    - Animal models – CBD obstruction in fetal lamb led to CC
    - Intraop measurement - High choledochal pressure in CC, especially type 1A (cystic)
  - ***"The pancreatic Reflux hypothesis"*** (long common pancreaticobiliary channel)
    - Animal models – pancreatic reflux in puppies led to high pressure but no mural weakening/diltation
    - Intraop measurement - bile amylase inversely related to pressure. Higher amylase in type 1B (fusiform)
- **Type II:** ? acquired (Healed perforation)
- **Type III**: obstruction of the ampulla or a congenital duodenal duplication.
- **Type V**: ductal plate malformation





- **Ductal plate malformations**: ductal plate (single layer surrounding portal vein) duplicates and partly fuses to form bile ducts. Normally each portal vein is surrounded by bile duct**. Arrest in ductal plate remodeling results in numerous, ectatic bile ducts.** Arrest at 12th wk (level of large duct) causes calori disease. Small duct malformation leads to caroli syndrome (multiple small cysts and hepatic fibrosis, genetic, ass. with PCKD)

Pathology

Depends on patients age and degree of cholangitis

- Bile ducts
  - pancreatico-biliary malunion (90%)
  - dilated intrahepatic ducts
  - stricture/stenosis
  - inflammatory reaction (mucosal ulcerations, pericystic inflamation)
  - epithelial metaplasia (adenocarcinoma)
  - sludge/Stones
- Liver
  - Normal
  - periportal fibrosis
  - Cirrhosis

Choledochal Cyst

2

2.1. Basics

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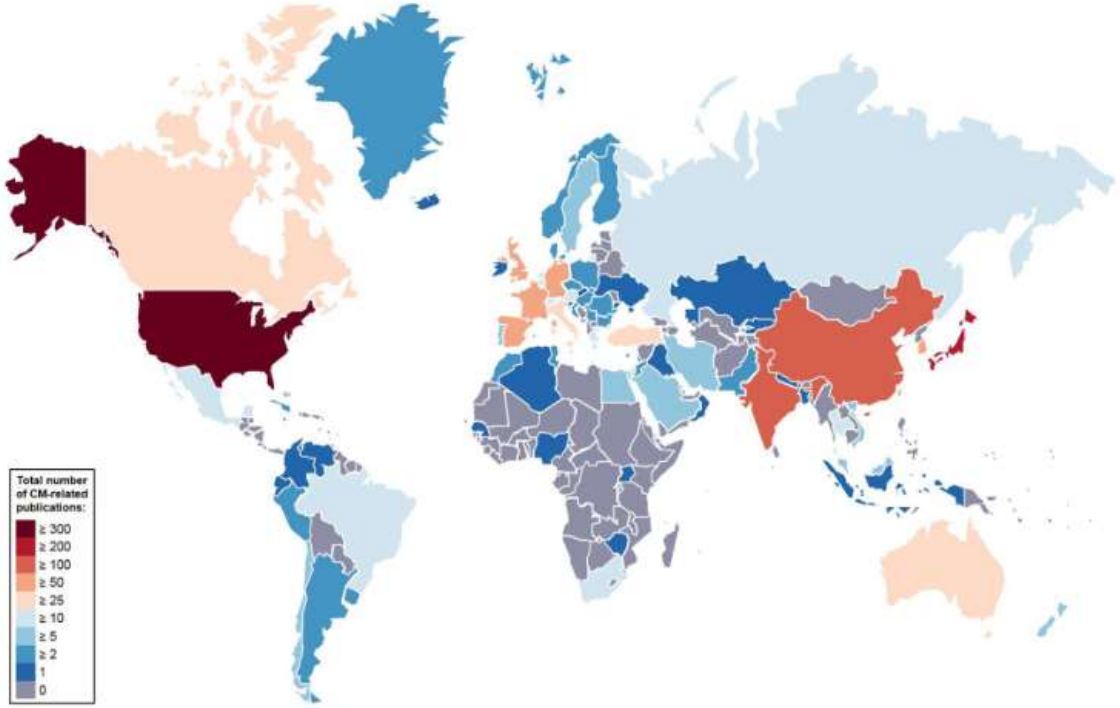
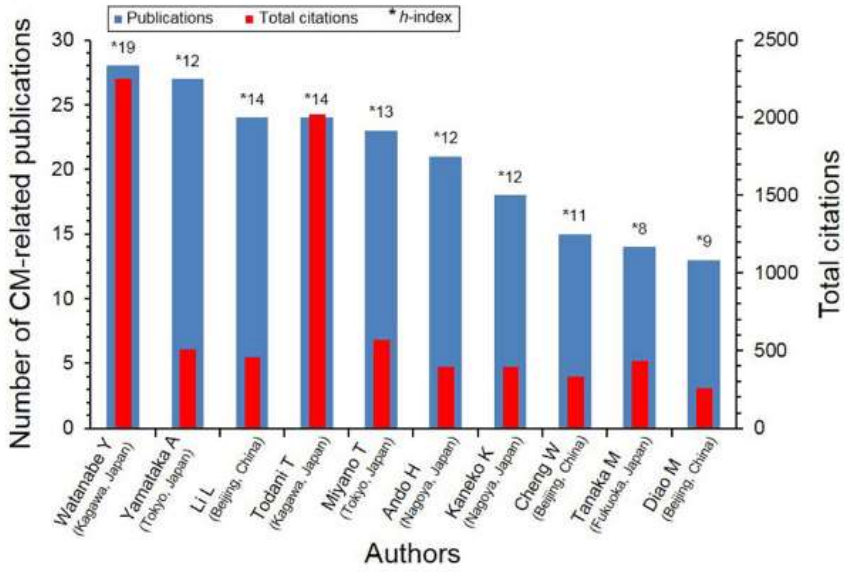
Epidemiology

- **Incidence** = 1:100,000
- **Female : Male** = 3 : 1 to 4:1
- **Age at presentation** = 90% by 10 yr



Choropleth mapping visualizing global publication volume in the field of CM research



- **Geographic distribution**: CC is more common in Asia (1:1,000). Most of the reported cases are from Japan (33-50%). The ten most productive authors in the field of choledochal cyst are either from Japan or China.

Presentation

Depends on age of onset.

- **Infantile form** (more severe- earlier hepatic fibrosis)
  - Prenatal diagnosis (6%)
  - Jaundice (icterus, discolored stool)
  - Abdominal mass (cystic type)
- **Adult form** (more symptoms, may also be diagnosed prenatally)
  - Abdominal pain (Classic Triad – jaundice, pain, mass in 20%)
- **Complications**
  - Cholangitis & pancreatitis – vomiting, fever
  - Perforation (5%) - hemobilia
  - Malignancy (type 4, adolescent/adult)
  - Cirrhosis (<2%)

Labs

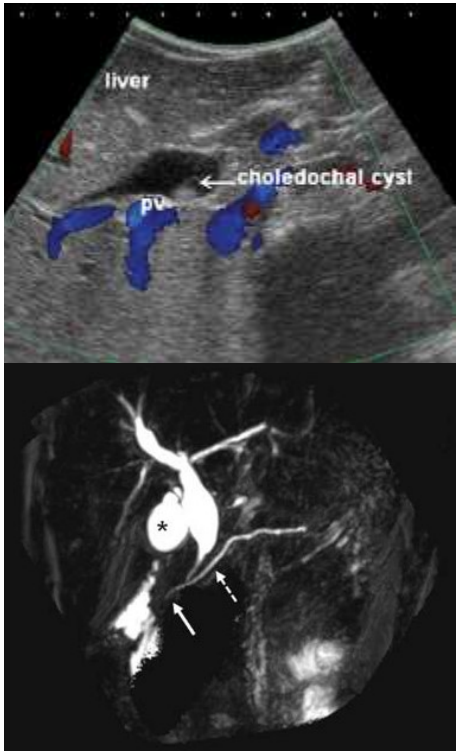
May be normal or show evidence of biliary obstruction

- Bilirubin
- ALP
- Coagulation profile

Imaging

Ultrasound: initial choice

- CC – contour, position
- proximal ducts
- vascular anatomy
- Liver - echotexture



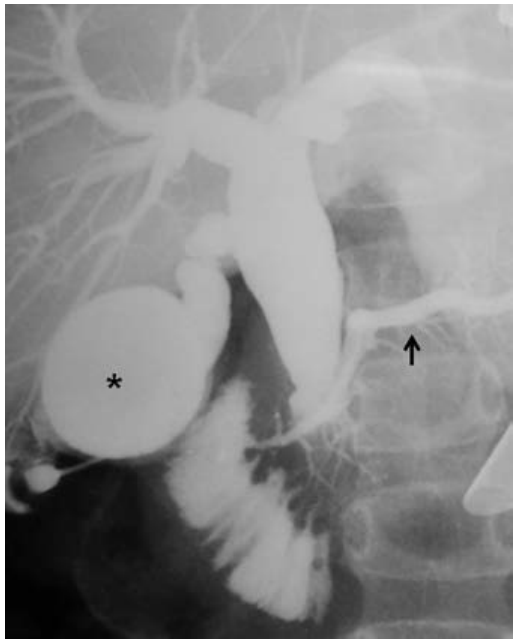
MRCP: gold standard

- highly accurate in detection & classification

Imaging

ERCP

- Common channel syndrome, borderline biliary dilatation (excellent anatomy Pancreatobiliary junction)
- invasive & has comp (panreatitis, perforation...)



Intraop cholangiography

- If anatomic detail can't be demonstrated on MRCP and ERCP

CT

- If pancreatitis or tumor is suspected, Fort type IV & V cyst

Radioisotope scan

- in doubtful fusiform (Liver function, biliary excertion), perforation

Choledochal Cyst

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2.1. Basics

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Timing of Surgery

- Safe in all age groups.
- **Earlier surgery for Infantile type** (to limit liver fibrosis)
    - As early as surgeon feels comfortable (? 1 mo, ? 3-6mo)
- **Urgent surgery** if obstructed

Preop preparation

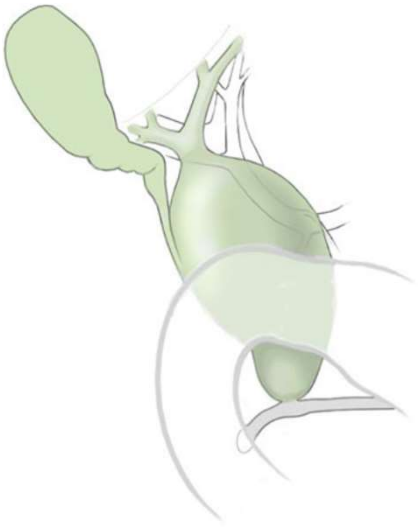
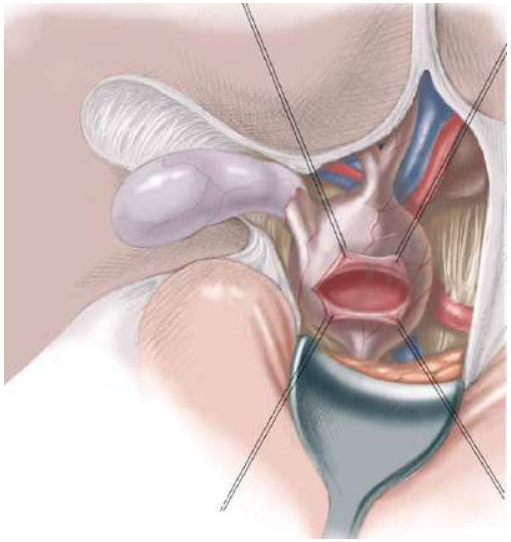
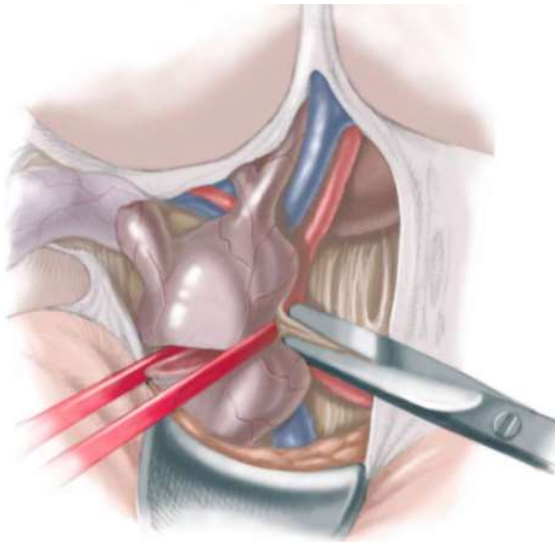
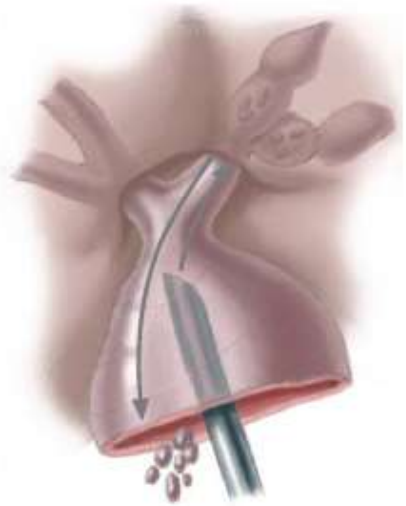
- **Treat cholangitis** (antibiotics)
- **Correct prolonged PT** (vit K)
- Drugs for ascaris (in areas where ascaris is prevalent)

Surgical options

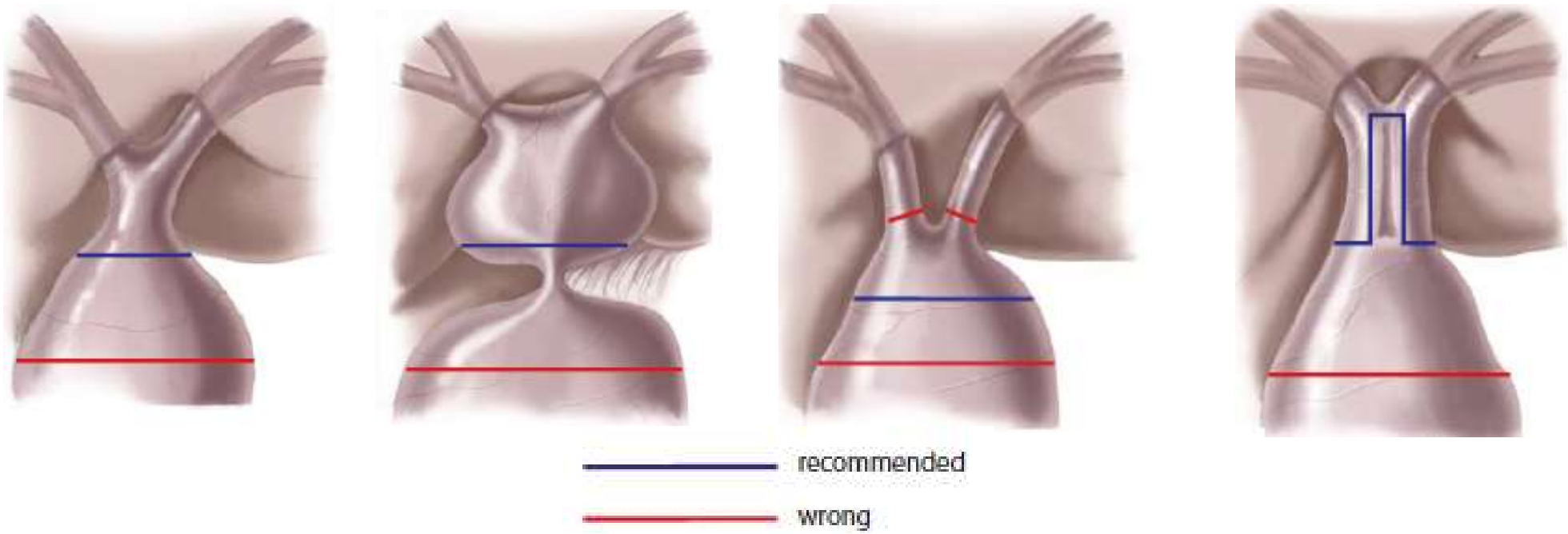
- **Common types of CC (type 1 & 4)**
  - Cystectomy with internal drainage (hepaticojejunostomy)
  - + liver resection = type 4 with hepatolithiasis, intrahepatic duct stricture or hepatic abscess
  - **ERCP & sphincterotomy = done in some centers for mild fusiform diltation
- **CBD Diverticula (type 2)**
  - Small neck = cystectomy with Primary closure (with T tube decompression)
  - Intrapancreatic CBD/large neck = cystectomy with internal drainage
- **Choledochocele (type 3)**
  - Small (<3cm) - Endoscopic sphincterotomy + unroofing
  - Larger – transduodenal excision + pancreatic duct reimplantation
- **Caroli disease (type 5)**
  - Localized = partial hepatectomy
  - Diffuse = liver transplant

Surgical Principles

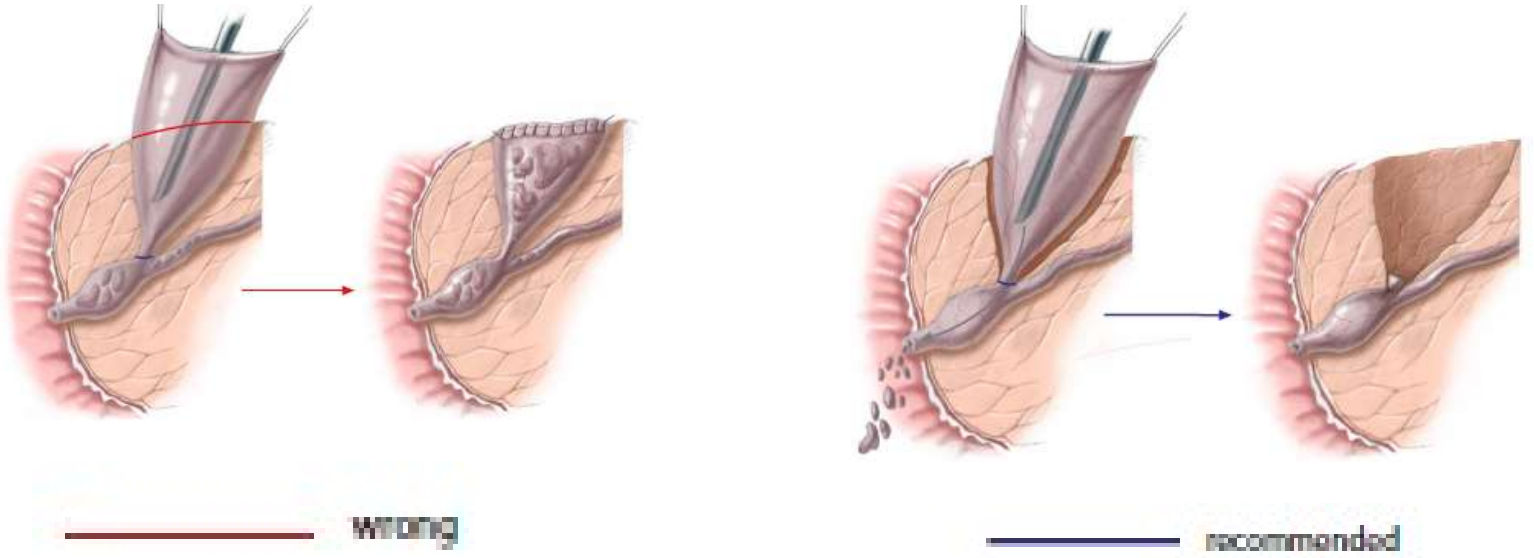
- **Cystectomy** (cyst-enteric anastomosis abandoned due to cholangitis, pancreatiits, stone, malignancy)
  - **acceptable to leave some cysts in type 4 CC (resolve after surgical relief)
- **Bilio-enteric anastomosis**
  - Hepatico–duodenostomy (shorter, more physiologic but no valve-cholangitis & inc reflux- gastritis-? malignancy)
  - hepatico-jejunostomy (longer surgery, duodenal ulcer, fat malabsorption, twist of roux limb, intest obst)
- ***Intraoperative endoscopy** (neonatal cystoscope): recommended by the Japanese group
  - Ensure debris is cleared, detect anatomic variations of CHD and distal CBD (determine site of resection)
  - Decision in type 4. (dilatation or ductal-plasty may be done for ductal stenosis up to 1st branch of intrahepatic)
- ***External drainage:** *for perforated, unstable*

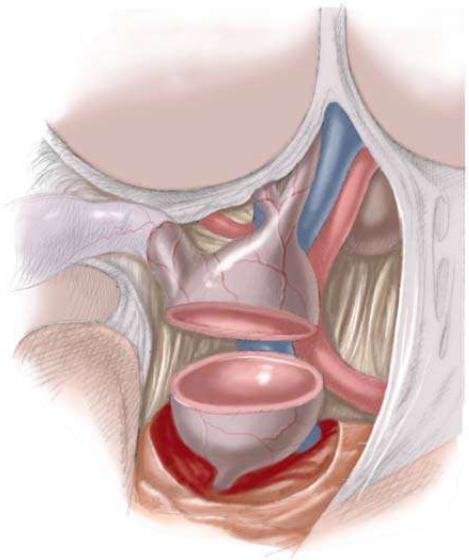
- **Cyst dissection:** extended subcostal incision. Retrograde cholecystectomy but gallbladder continuity is maintained with the choledochal cyst. Cyst is mobilized. The Japanese group prefer to always open the anterior wall transversely to facilitate safe dissection. Catheter/scope is placed through biliopancreatic channel to irrigate protein plugs & calculi.

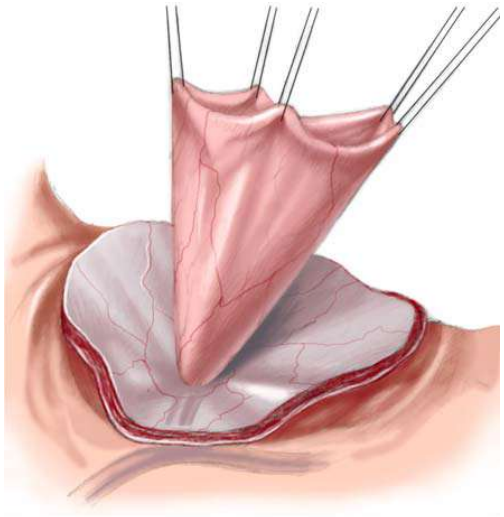


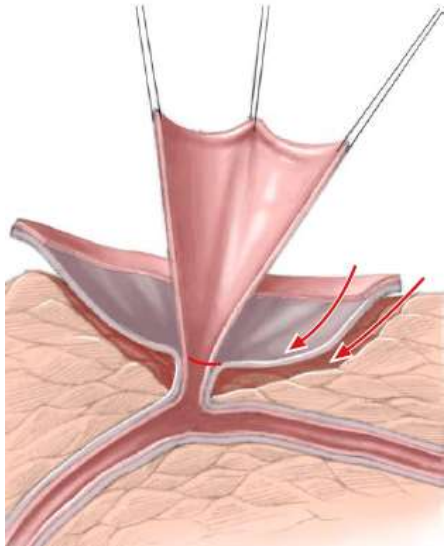
- **Cyst Excision (proximal):** Cyst divided proximally 5mm from hepatic duct confluence and irrigated. CHD length above 10 mm will become kinked. However, lumen of the CHD should be inspected before trimming because there might be anatomic variations (luminal stenosis, separate opening, or septum)



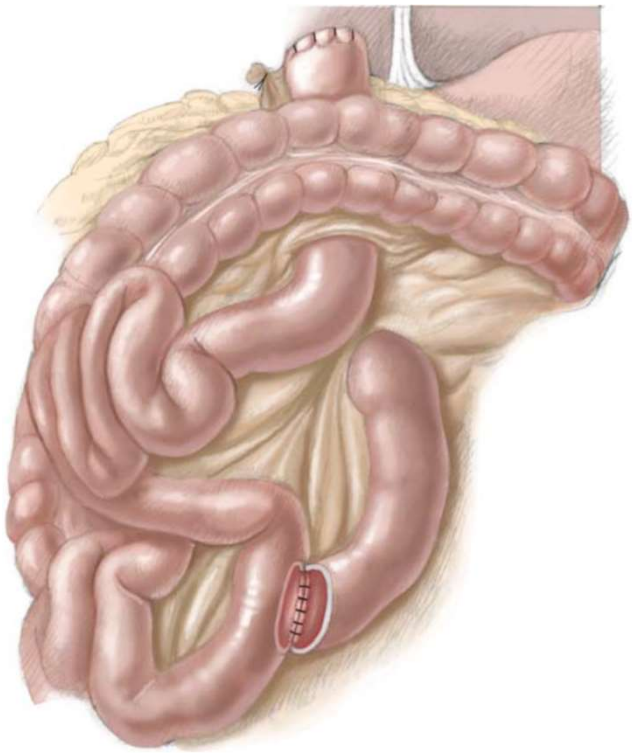
- **Cyst excision (Distal):** Distal end divided at end of CBD making sure not to leave the intra-pancreatic portion of the CBD behind and not to injure the pancreatic duct. If resected along the red line, cyst will reform.





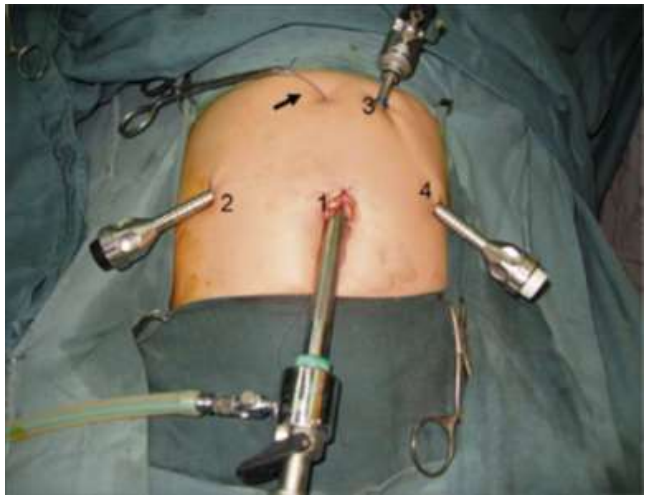


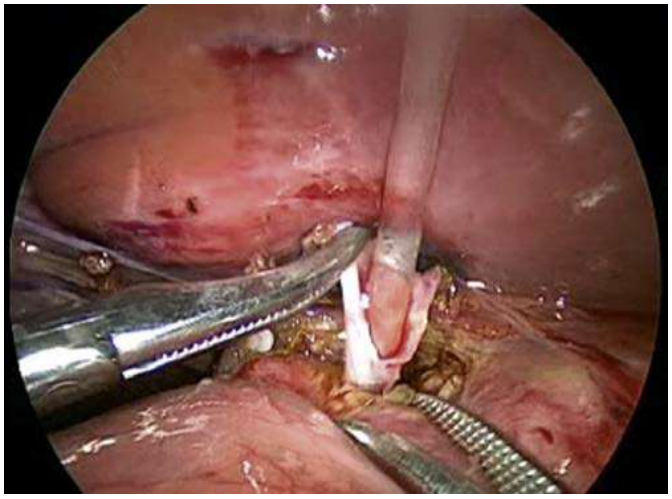
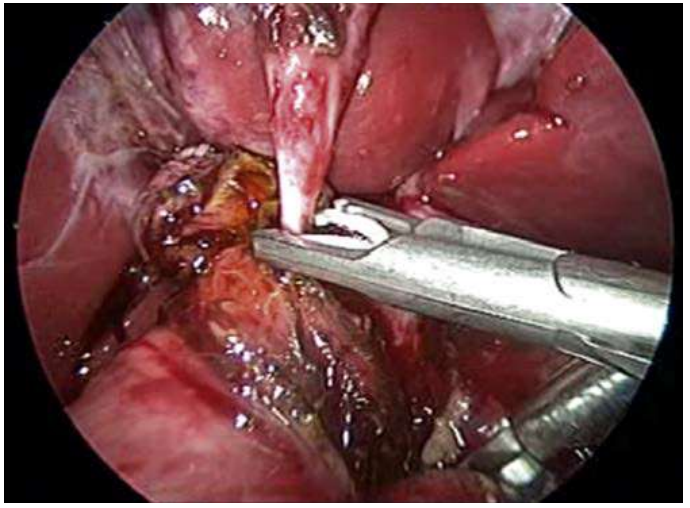
- ***Mucosectomy (Lilly technique):** If the cyst is extremely inflamed and adhesions are very dense (older children) the wall should be opened . Mucosectomy may be performed rather than full-thickness dissection. Following mucosectomy, the distal end of the cyst is transfixed twice using 3/0 or 4/0 absorbable sutures. The distal stump is either left or buried in the muscle wall of the surrounding cyst.



- **Roux loop preparation:** jejunum divided a few cm distal to ligament of treitz (neonate- 10cm, infant-20 cm, older-30cm). Jejuno-jejunostomy (end-side) to create Roux limb which is passed retrocolic (through window in transverse mesocolon to right of MCA). Length of the roux limb is predetermined by some surgeons (30-60 cm) but it should be decided intraop so that the roux limb fits into the splenic flexure (this length doesn't cause redundency) Mesentric defects are closed. **Some recommend fixing the roux limb to the native jejunum for upto 8 cm cranially so that contents flow smoothly down (otherwise anastomosis becomes T shaped leading to bile stasis & reflux of GI content in roux limb)

- **Hepaticojejunostomy**: can be anastomosed end-side or end-end (preferred if no significant size discrepancy). If end-to-side anastomosis is unavoidable, it should be as close as possible to the closed end so there will be no blind pouch. Otherwise elongation of the blind pouch will occur later in life and cause bile stasis. Use 5-0/6-0 PDS. Interrupted sutures are used if CHD diameter is <1cm. If CHD is too small left HD may be incised longitudinally. anastomosis placed above transverse colon mesentry to help prevent intestinal obstruction from adhesions.



- **Laparoscopic approach:** can be used if no perforation or adhesion (no previous hepatobiliary surgery)

Intraoperative accidents

- **Portal vein injury –** keep dissection close to cyst as possible.
- **Hepatic duct injury –** occurs when hepatic bifurcation is low away from hilum. Avoid by internal inspection before excising cyst.
- **Pancreatic duct injury –** prevent by understanding bilopancreatic anatomy preop. Avoid by Internal inspection of distal CBD to identify orifice of common duct.
- **Roux limb twist –** careful inspection before hepaticojejunostomy

Postop care

- **Antibiotics** – 5 days
- **Subhepatic drain** – remove on day 5 if no leak
- **Feeding** – 2-3d (when NGT is clear)

Complications

- 5-9% complication, early mortality ~0%
- **Early**
  - Bleeding
  - Intestinal obstruction (0-5%)
  - **Biliary leak** (0-5%) & **pancreatic fistula** [antibiotic, drainage, NGT & TPN]
- **Late**
  - **Cholangitis** (2-3%) & pancreatitis (2.5%) [antibiotics]
  - **Stricture** (1-2%) & Intrahepatic calculi [ERCP or reoperation]
  - **cholangiocarcinoma** (4%) = residual cyst, stone, recurrent cholangitis, stricture, intrahepatic dilation

Follow up

- Long term follow up (cholangiocarcinoma)
    - **Labs** (CA 19-9, Liver transaminase, Bilirubin)
    - **Ultrasound-** if symptomatic or lab abnormality
    - **MRCP** – depending on ultrasound & labs

Difficult scenarios

- Differentiating **type 4 CC Vs type I CC with dilated intrahepatic ducts**
  - If obstructed, dilatation may be due to type I. But generally both are the same (not important to d/t)
- Differentiating **dilatation due to CBD stone Vs underlying choledochal cyst** in older children
  - Cystic CC are easy to d/t. Difficult to d/t fusiform, but consider presence of common channel.
- Differentiating **cystic BA Vs CC** in prenatally diagnosed
  - If obstructed (jaundiced) difficult to distinguish, but both need urgent surgery
  - If not jaundiced and bilirubin comes down first 2 weeks of life, it is CC

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