Choledochal Cyst

Etiopathogenesis

Clinical Feature

Management

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Introduction

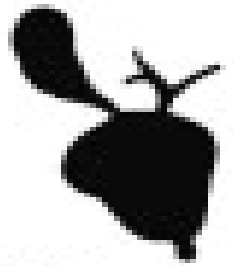
- 1% of all benign biliary disease
- Disease of infancy or childhood
- 20% of patients, the diagnosis is delayed until adulthood
  - associated hepatobiliary pathology
  - complications of previous cyst-related procedures
- Excision is best - concensus

Epidemiology

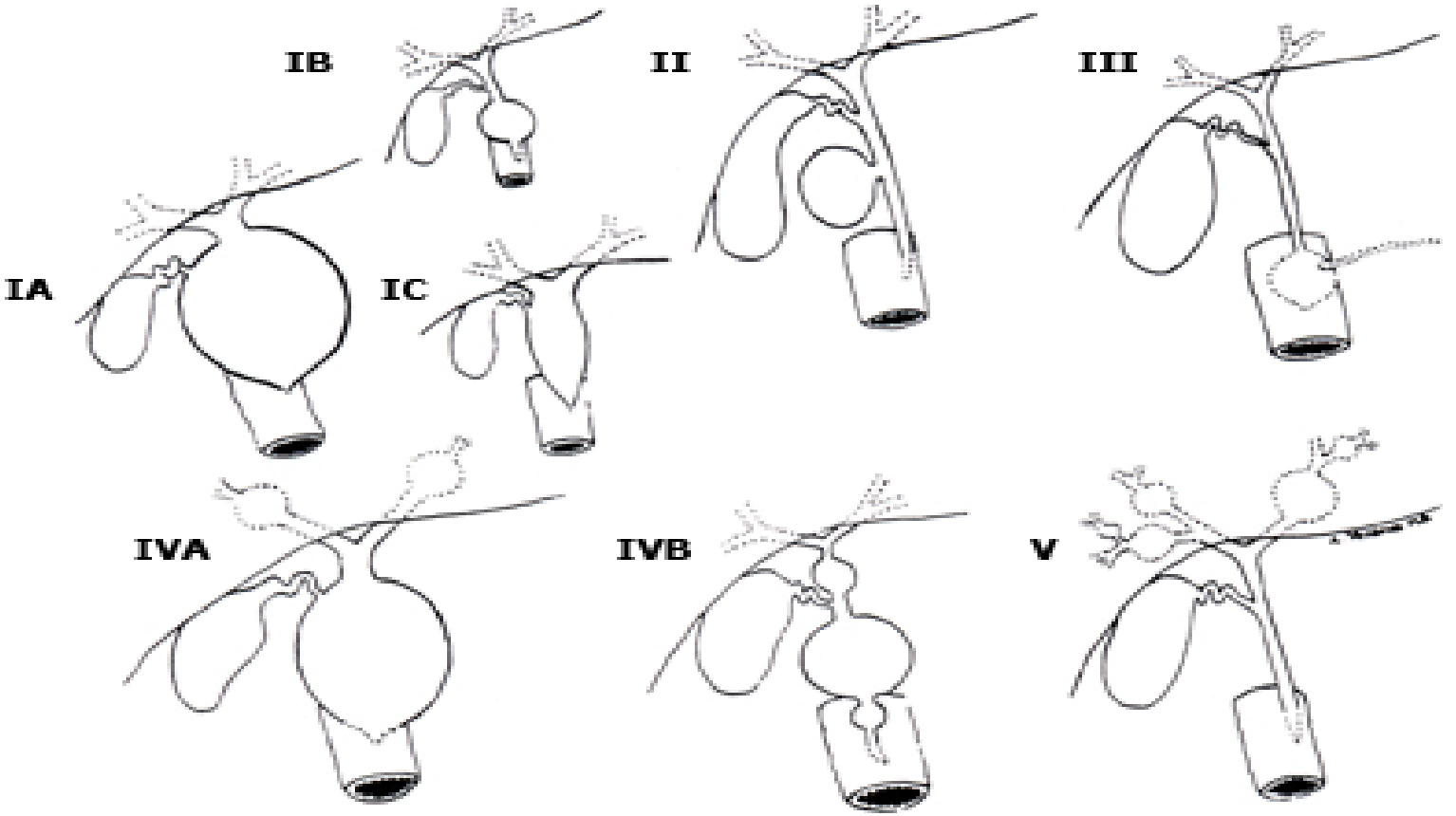
- incidence of biliary cysts - 1:100,000 to 1:150,000
- female to male ratio of 3:1 to 4:1
- one-half and two-thirds of the reported cases occurring in Japan

Todani modification of Alonso-Lej classification (Todani et al, 1977)

Type	Findings	Type	Findings
I	Solitary fusiform extrahepatic cyst	IVA	Fusiform extra- and intrahepatic cysts
II	Extrahepatic supraduodenal diverticulum	IVB	Multiple extrahepatic cysts
III	Intraduodenal diverticulum; choledochocele	V	Multiple intrahepatic cysts; Caroli disease





- Type IA is defined by cystic dilation of the entire extrahepatic biliary tree and is associated with an APBJ. There is no dilation of the intrahepatic ducts. The cystic duct and gallbladder arise from the dilated common bile duct.

- Type IB is defined by focal, segmental (often distal) dilation of the extrahepatic bile duct. Type IB cysts are not associated with an APBJ.
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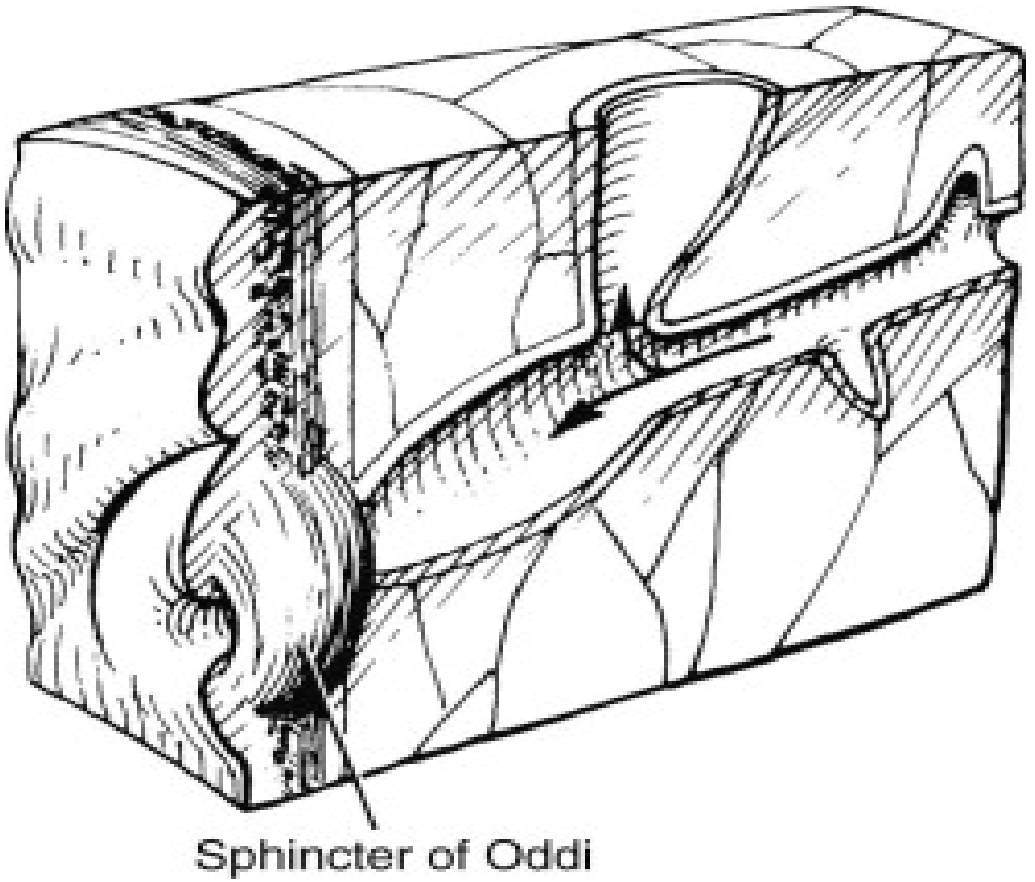
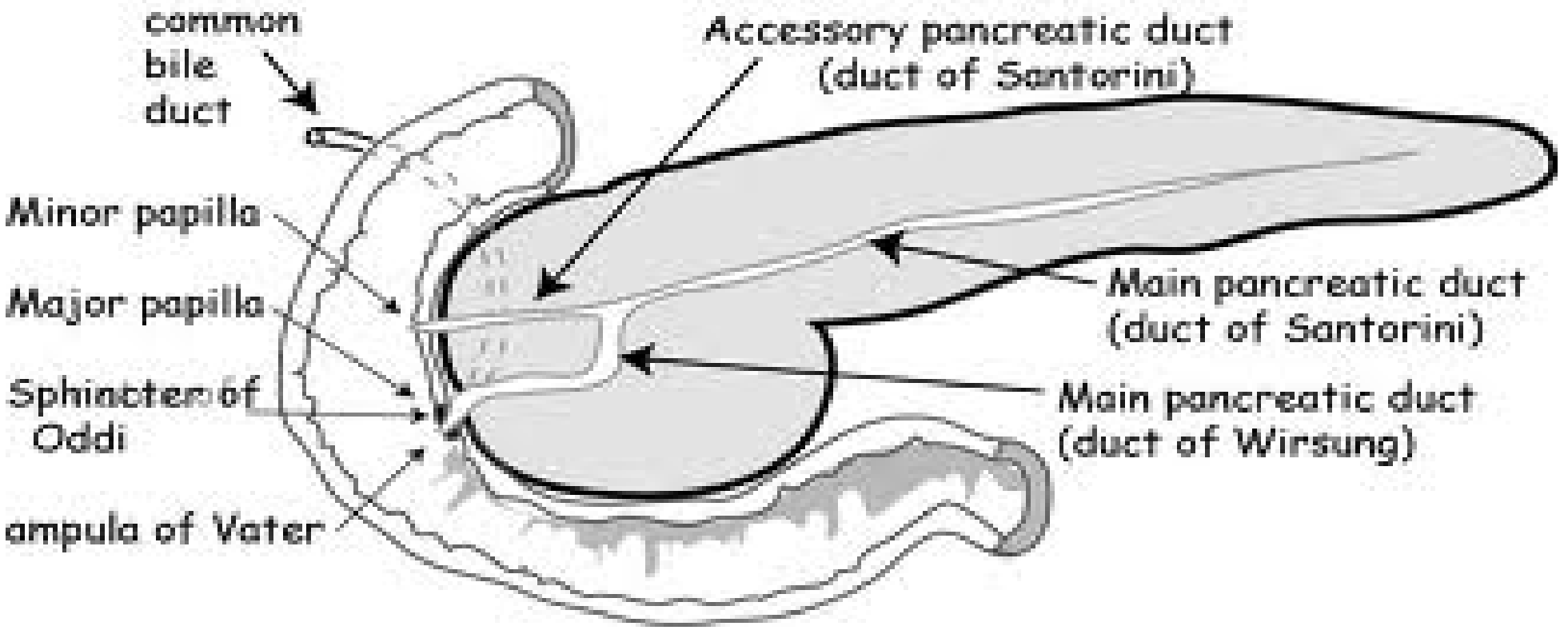
- Type IC is defined by smooth, fusiform (as opposed to cystic) dilation of the entire extrahepatic bile duct. Typically, the dilation extends from the pancreatobiliary junction to the intrahepatic biliary tree. Type IC cysts are associated with an APBJ.

long common channel pancreaticobiliary ductal confluence





APBJ

- rare congenital anomaly, with a prevalence of 0.03 percent
- present in about 70 percent of patients with biliary cysts
- significant risk factor for the development of malignancy with the cyst
- long common duct channel (at least 8 mm, and often over 20 mm, in length)

- presence of an acute or right angle at the bile duct/pancreatic duct junction
- presence or absence of a dilated common channel
- presence or absence of a dilated common channel
- Surgical sphincteroplasty or endoscopic sphincterotomy may be required in the treatment

Other etiology

- hereditary factor
- oligoganglionosis in the distal neck of the cyst
- Cysts may be congenital or acquired
- genetic or environmental predisposition

- Type I choledochal cyst - 79%
- Type II - 2.6%
- Type III or choledochocele - 4%
- Type IV - - 13%

- Caroli's Disease - < 1 % cases

Clinical Feature

- Asymptomatic
- Incidental finding on imaging studies
- Symptoms mimicking calculous biliary tract disease
- Most common initial findings
  - Pain - intermittent, recurrent epigastric or right hypochondrial pain
  - Abdominal tenderness
  - Fever
  - Mild jaundice—the most common initial findings

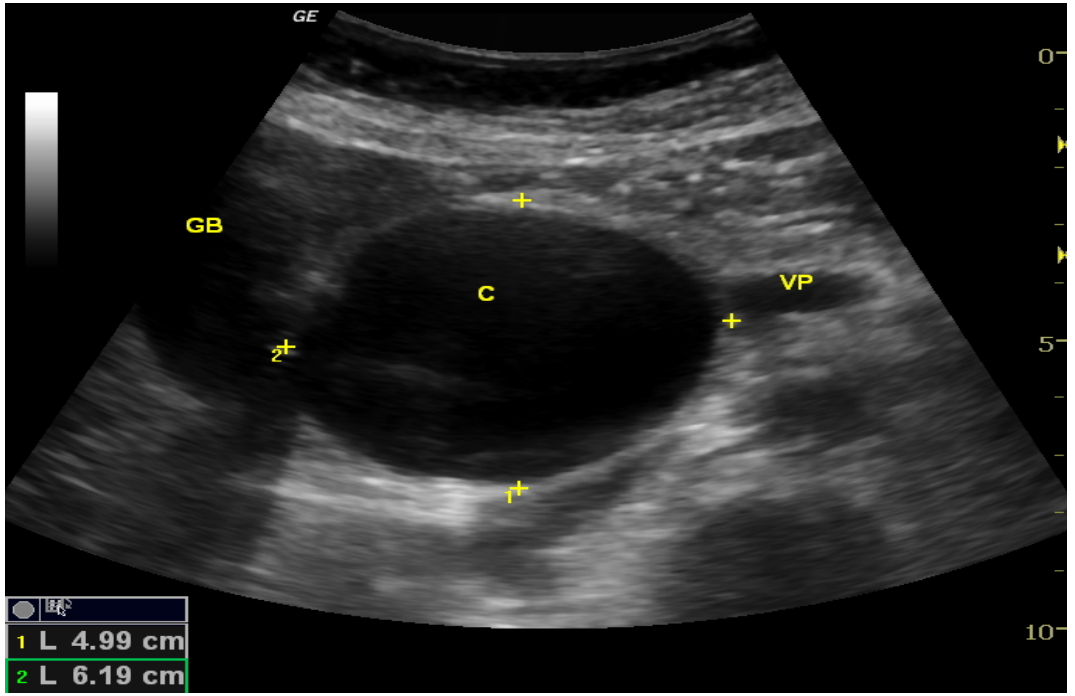
- Fever and rigors – cholangitis
- abdominal mass - uncommon in adults - ? Malignancy
- Nausea, vomiting, and anorexia
- Signs of sepsis
- cirrhosis or hepatic fibrosis from chronic biliary obstruction – 15 % of Adults
  - Hepatomegaly and splenomegaly
  - Hematemesis, melena, and ascites
  - Liver failure - late feature
- Pancreatitis - 30% of patients
  - associated bile duct malignancy in 70 % cases

USG

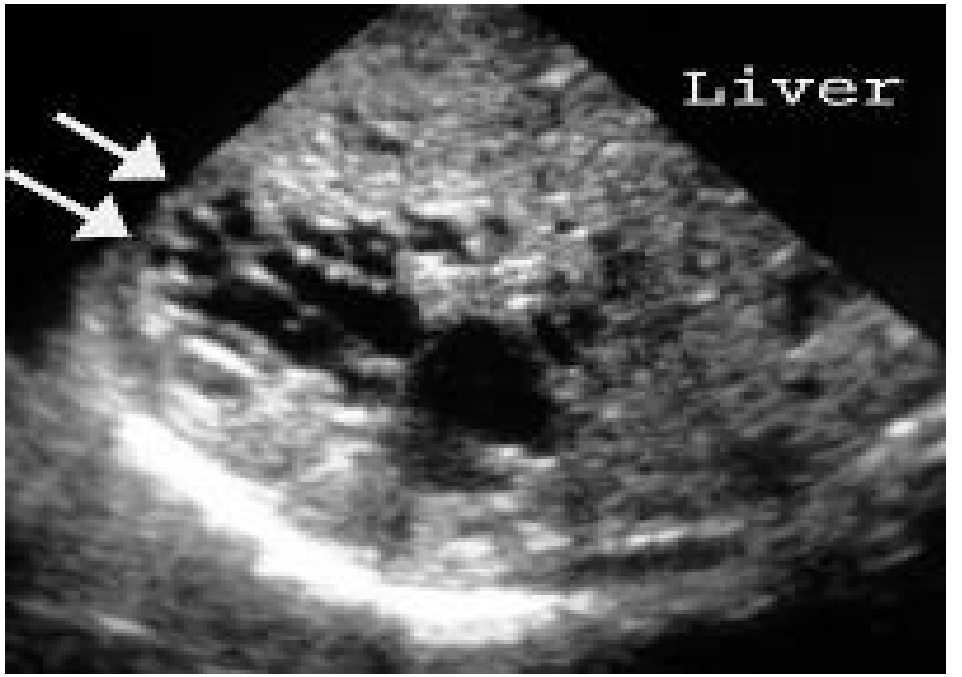
- Initial investigative procedure
- Irregular hypoechoic segmental dilation of the extrahepatic bile duct
- Scan multiple planes
- Define the extent of the dilation
- Focal duct wall thickening or nodularity – malignancy
- Stones within the cyst - echogenic features and acoustic shadowing
- absence of septations - distinguish choledochal cysts from extrahepatic biliary tumors
- Caroli disease - multiple cysts adjacent to the major intrahepatic bile ducts

USG

Type I choledochal cyst



Caroli's disease



- CT combined with intravenous cholangiography
- Direct cholangiography - accurate definition of the type of choledochal cyst
  - Configuration and extent of the cyst
  - Ductal strictures
  - Stones within the biliary and pancreatic ducts
  - Polypoid filling defects that suggest ductal malignancy
  - Relationship of cyst to pancreatic duct

Angle of fusion

- angle between the distal bile duct and pancreatic duct
- avoid intraoperative damage of the pancreatic duct during cyst excision
- recognize stones impacted within the common channel or junction
- exclude distal tumors

ERCP

- Better visualization of an abnormal pancreaticobiliary ductal junction
- Carcinoma can be excluded by biopsy or brush cytology
- intracystic stones can be extracted after papillotomy
- relieve severe cholangitis before surgery
- visualization of the esophagus and stomach to exclude signs of portal hypertension
- biopsy of intracystic masses
- Type III cysts or choledochoceles - endoscopic papillotomy is potentially therapeutic

PTC

- Previous Roux-en-Y cystenterostomy
- Patients with type IV bile duct cysts, in whom ductal strictures or tumors prevent complete visualization of intrahepatic cysts by ERCP
- occasional failure to clearly define the pancreaticobiliary junction
- Percutaneous biliary drainage (PBD) - control of biliary sepsis or to aid surgical reconstruction

MRC

- Noninvasive method of imaging the bile ducts and its anomalies
- accurate anatomic definition of bile duct cysts in neonates, infants, and young children
- limitation of MRC - inability to clearly define the pancreaticobiliary junction (diagnostic accuracy, 69%)
- lack of therapeutic capability.

Hepatobiliary scintigraphy

- technetium-99m-labeled hepatic iminodiacetic acid (HIDA)
- cyst will initially be photopenic, followed by filling of the cyst and delayed emptying of the contrast into the bowel
- demonstrate continuity of cysts with the bile ducts
- useful for extrahepatic cysts
- in cases of cyst rupture

Cystolithiasis

- most frequent accompanying condition in adults
- ranged from 2% to 72%
- bile stasis - primary etiologic factor
- complicates anastomotic strictures after previous cystoenterostomies

Hepatolithiasis

- type IV bile duct cysts
- Stenosis of the major ducts should be assessed
- subsequent intrahepatic abscess formation

Gallbladder disease

- Both acute and chronic cholecystitis, with or without stones, has been recognized in these patients

Pancreatitis

- typically is mild pancreatitis
- ranged from 2% to 70%
- common channel syndrome/pseudopancreatitis
- pattern of pancreatitis is acute and often relapsing
- Chronic pancreatitis associated with bile duct cysts is rare.
- anomalous pancreaticobiliary ductal anatomy

Hepatobiliary malignancy

- between 2.5% and 28%
- Cholangiocarcinoma is the most common malignancy
- adenocarcinoma, adenocanthoma, squamous cell carcinoma, anaplastic carcinoma, bile duct sarcoma, hepatocellular carcinoma, pancreatic carcinoma, and gallbladder carcinoma
- incidence of cyst-associated malignancy is age related, increasing from 0.7% in the first decade of life to more than 14% after age 20 years

- The mean age of patients with cancer associated with bile duct cysts is 32 years
- only 57% of tumors are intracystic
- rest arise elsewhere within the liver or pancreaticobiliary tract.
- prevalence of cancer is significantly greater in type I and IV cysts

- Long-term survival of patients with bile duct cysts and malignancy is rare
- Delayed diagnosis, advanced stage of disease, intraabdominal seeding from previous operations, and tumor multicentricity generally preclude curative resection

Intrahepatic abscess

- Result from recurrent cholangitis
- completely obstructed, pus-filled intrahepatic cysts
- intrahepatic abscess predominant in the left intrahepatic duct
- angulation of the left main duct

Portal hypertension

- Secondary biliary cirrhosis or fibrosis
- Portal vein thrombosis
- Caroli disease with congenital hepatic fibrosis
- hepatosplenomegaly, hematemesis, melena, or ascites

Treatment

- complete cholangiographic definition of the extent of the cystic process
- associated ductal pathology
- control of biliary infections
- Broad-spectrum antibiotics
- percutaneous or endoscopic drainage of infected bile duct cysts

- all bile duct cysts should be excised and bile flow reestablished by mucosa-to-mucosa bilioenteric anastomosis
- If complete excision is not feasible, partial cyst excision and Roux-en-Y cystojejunostomy
- external drainage alone has no role
- long-term follow-up

Surgical options for treatment of choledochal cysts

Type I

EXCISION, ROUX-EN-Y HEPATICOJEJUNOSTOMY
EXCISION, HEPATICO-DUODENOSTOMY
Roux-EN-Y choledocho-cystojejunostomy
Choledochocystoduodenostomy

Type II

EXCISION

Type III

TRANSDUODENAL EXCISION
Transduodenal sphincteroplasty
Endoscopic sphincterotomy

Type IVA

Extrahepatic component
EXCISION, ROUX-EN-Y HEPATICOJEJUNOSTOMY
EXCISION, HEPATICO-DUODENOSTOMY
Intrahepatic component
Hepatic resection ± Roux-en-Y hepatico-jejunostomy
Transhepatic intubation

Type IVB

EXCISION, ROUX-EN-Y HEPATICOJEJUNOSTOMY OR HEPATICO-DUODENOSTOMY
± transduodenal sphincteroplasty

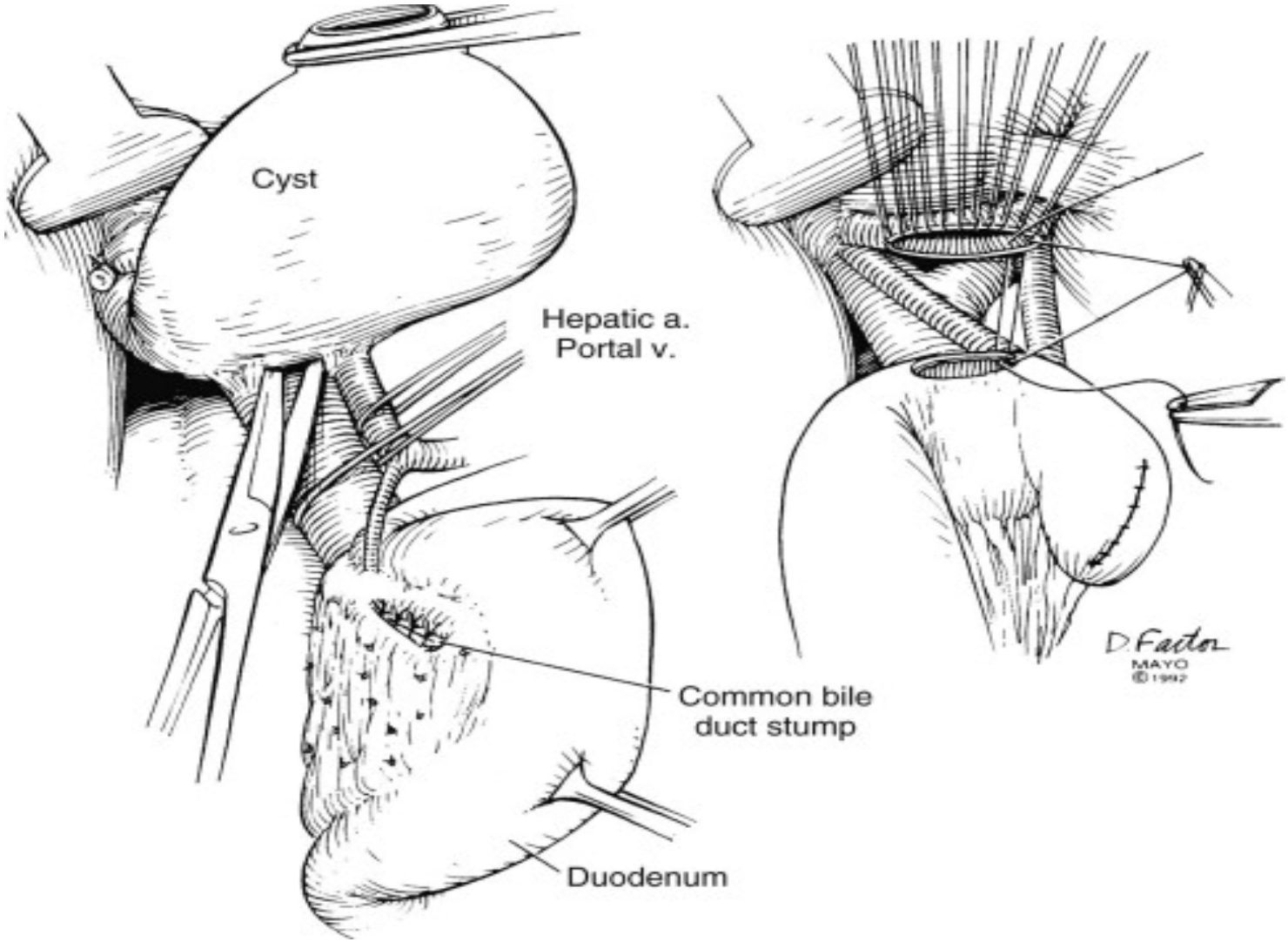
Type V (Caroli disease)

HEPATIC RESECTION
Roux-en-Y intrahepatic cholangiojejunostomy
Transhepatic intubation
Orthotopic liver transplantation

Type I cyst

- Treatment of choice - total cystectomy and Roux-en-Y hepaticojejunostomy
- Reduced incidence of anastomotic stricture, stone formation, cholangitis, and intracystic malignancy
- Morbidity and mortality – same as cyst-enteric anastomosis
- recurrent cholangitis from anastomotic strictures occurs in 10% to 25% of patients

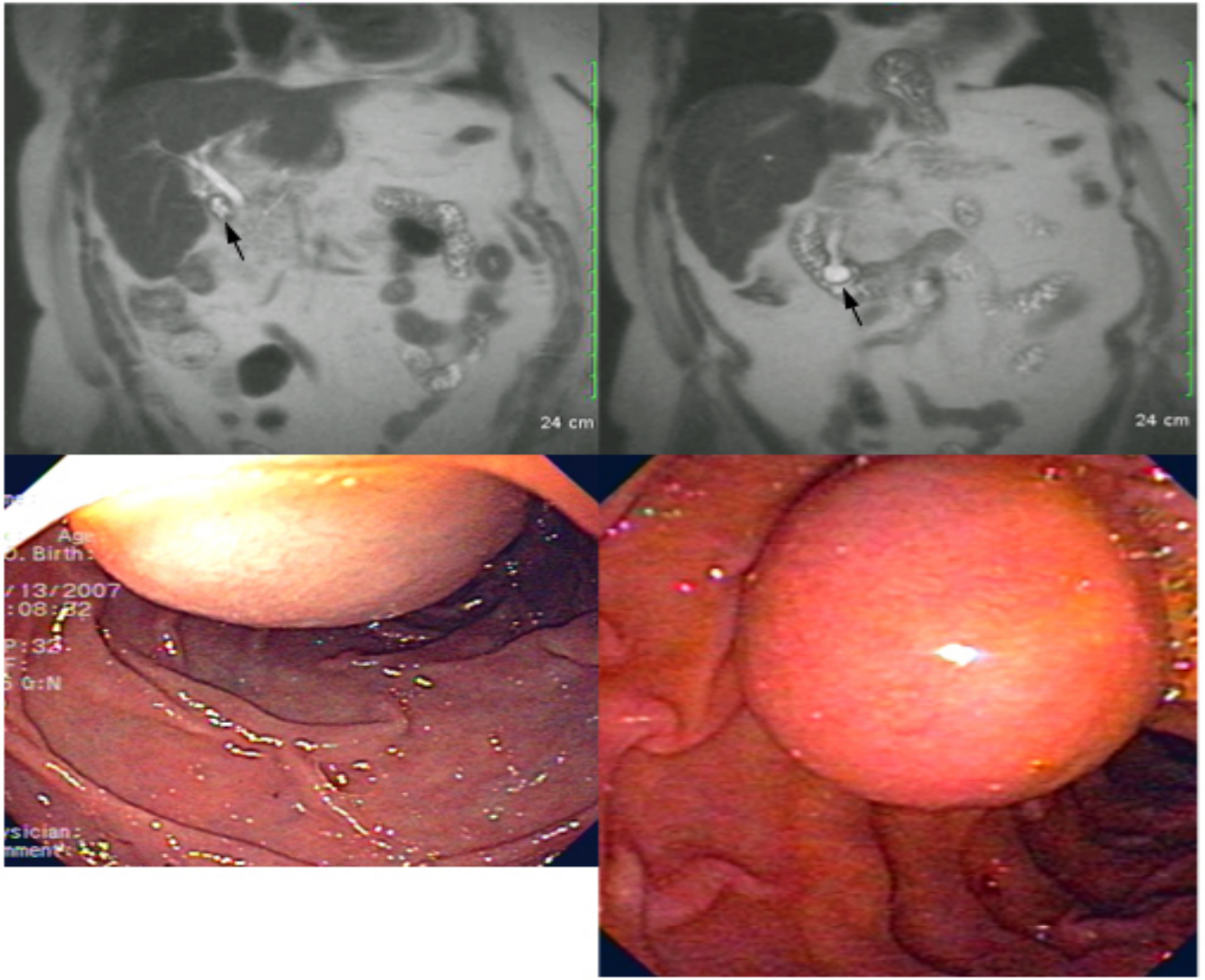
- Reoperation and cyst excision – previous cystenterostomy
- hepaticoduodenostomy - residual biliary epithelium is partially accessible to direct visualization by endoscopy



- Roux-en-Y choledochocystojejunostomy
  - Portal hypertension
  - severe pancreatitis
  - past drainage procedures
- Portal decompression - before biliary reconstruction - central splenorenal shunts / TIPSS
- drainage is undertaken 6 to 12 weeks after portosystemic shunting.

Type II Cyst

- Treatment of choice - excision
- similar to that of cholecystectomy
- neck can either be closed primarily
- T-tube decompression of the common duct



Type III Cyst (Choledochocele)

- endoscopic sphincterotomy and cyst unroofing – tratment of choice
- risk of cancer alone is sufficiently low
- transduodenal cyst excision with or without sphincterotomy
- Damage to the major pancreatic duct – major morbidity and mortality
- Identification of the duct of Wirsung is very important

Type IV Cyst

- Treatment of choice - excision of extrahepatic cyst and Roux-en-Y hepaticojejunostomy
- Transduodenal sphincteroplasty and Roux-en-Y hepaticojejunostomy complete the treatment of type IVB cysts
- Type IVA cysts require more selective management
- lobar hepatic resection is usually required to eliminate the complicated unilobar intrahepatic cystic component

Type IVA cysts

- 90% of patients who had excision of the extrahepatic component of the type IVA cyst had good results, whether or not the intrahepatic component was resected
- cyst drainage alone – satisfactory in < 50% cases
- importance of addressing the presence of variant ductal anatomy and stenoses at the hilum
- Both membranous or bridge like stenoses should be excised circumferentially to their base

Caroli Disease

- Treatment depends on :
  - extent of the intrahepatic bile duct cysts
  - the presence of congenital hepatic fibrosis
  - secondary biliary cirrhosis
  - Carcinoma
- most commonly involving the left ductal system
- Hepatic resection, with or without Roux-en-Y cholangiojejunostomy – treatment of choice

- Resection is always preferable to drainage if the liver parenchyma surrounding the cyst is atrophic
- If resection in patients with localized Caroli disease is not feasible, Roux-en-Y intrahepatic cholangiojejunostomy to the intrahepatic cyst is preferable

- Poor results
  - both lobes of the liver involved
  - portal hypertension from congenital hepatic fibrosis
  - secondary biliary cirrhosis
- Orthotopic liver transplantation has been successfully used for Caroli disease

Laparoscopic Advances

- Palanivelu and colleagues (2008) recently reported the largest series to date of laparoscopic treatment of bile duct cysts.
- In this series of 35 patients, 16 were adults. Conversion to laparotomy was performed in three patients for suspicion of malignancy, and complete cyst excision was performed in 28; in five patients, an incomplete resection was performed as a result of extension of the cyst proximal to the hepatic duct confluence.
- Postoperative complications occurred in 14%, with no perioperative mortality.

Advantages

- superior visualization of the structures around the cyst
- assess neovascularity around the cyst
- separation of the posterior cyst wall from the portal vein
- dissection and anastomosis in the hilar area

	Palanivelu et al, 2008	Hong et al, 2008	Li et al, 2004	Lee et al, 2009	Jang et al, 2006
Patients (n)	35	31	35	37	13
Pediatric (%)*	54	100	100	100	0
Cyst type					
I†	27 (77)	31 (100)	33 (94)	35	9
II	0	0	0	0	0
III	0	0	0	0	0
IV	8 (23)	0	0	2	4
Conversion to open	3 (9)	4 (13)	0	4 (10.8)	1 (8)
Mean cyst size (cm^2)	51	3	4‡	2.8	ND
Complete cyst excision, n (%)	28 (80)	ND	35 (100)	ND	ND
Mean operating time (min)	295	312	258	439	228
Perioperative morbidity, n (%)	5 (14)	4 (13)	1 (3)	15 (40.5)	2 (15)
Mean hospital stay (days)	6.5	8.6	4.5		5.8

Thank You