CHOLEDOCHAL CYST

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2ND YR POST GRADUATE
GENERAL SURGERY

INTRODUCTION

- Congenital focal or diffuse dilations of the biliary tree
- most common congenital abnormality of the biliary tree.
- considered a premalignant condition requiring surgical intervention.
- commonly diagnosed in infancy, but may present in adulthood.

INCIDENCE

- More common in Asia.
- 1 in 1000 live births in Japan.
- 1 in 13000 live births in India.
- rare in Western countries; incidence is between 1:100,000 and 1:150,000.
- Female preponderence- 4:1

CLASSIFICATION

- Original Classification by Alonso-Lej and associates (exclusively involved the extrahepatic duct).
- Revised by Dr. Todani and colleagues in 1977 (included intrahepatic cystic anomalies).

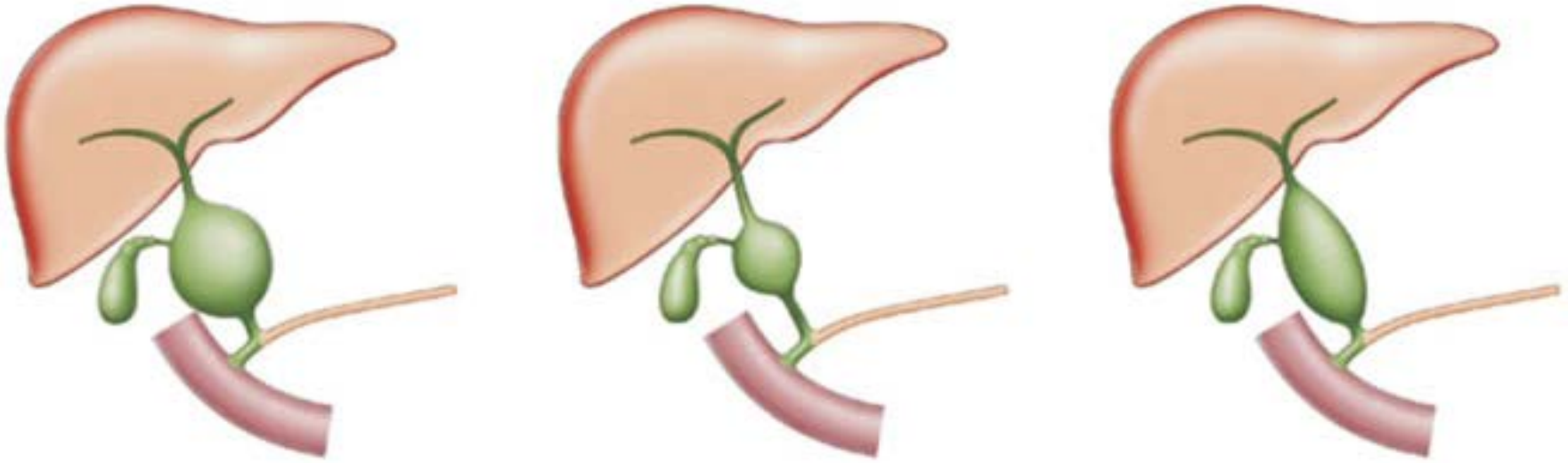
Todani's classification of choledochal cyst

I a- cystic dilation of extrahepatic bile duct (entire) Ib- focal segmental dilation of of extrahepatic bile duct Ic- fusiform dilation of CBD portion of extrahepatic bile duct
II- diverticulum
III- dilation of the distal intraduodenal CBD (choledochocele)
IVa-multiple cysts intrahepatically & extrahepatically IVb- multiple cysts extrahepatically
V- single or multiple intrahepatic cysts without extrahepatic duct dilatation (caroli's disease)

I a- **cystic** dilation of extrahepatic bile duct (entire)

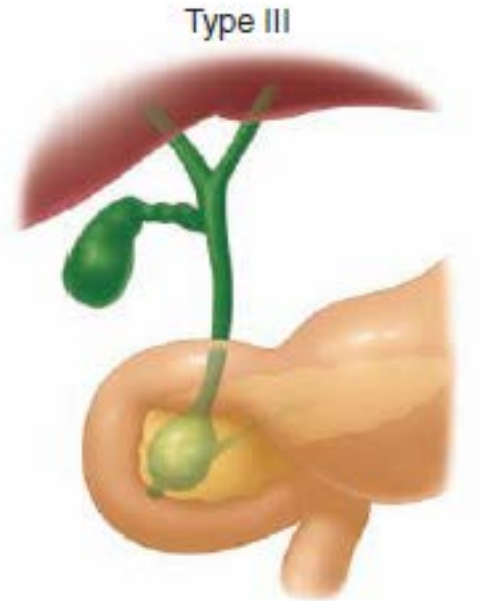
Ib- **focal** segmental dilation of of extrahepatic bile duct

Ic- **fusiform** dilation of CBD portion of extrahepatic bile duct



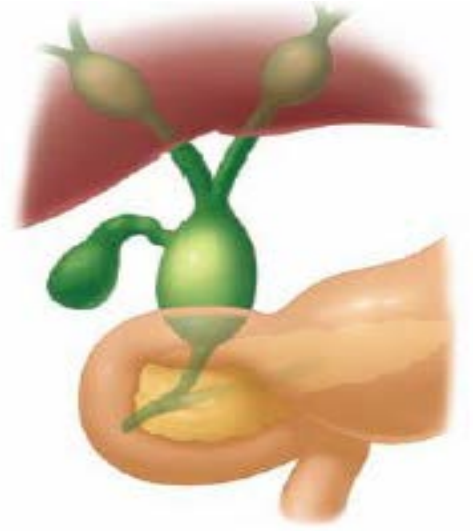
- II- diverticulum

- III- dilation of the distal intraduodenal CBD (choledochocele)



IVa-multiple cysts intrahepatically & extrahepatically

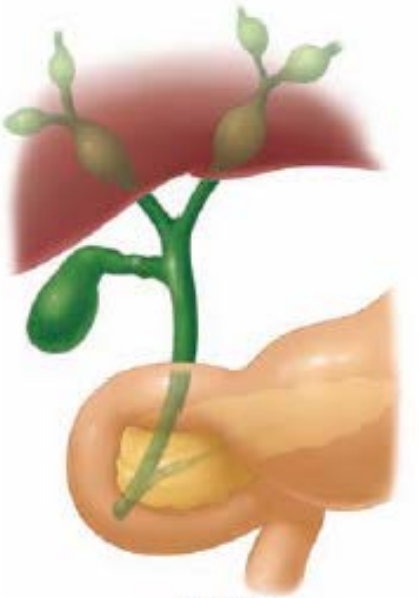
IVb- multiple cysts extrahepatically



Type IVa

Type IVb

- V- (caroli's disease)

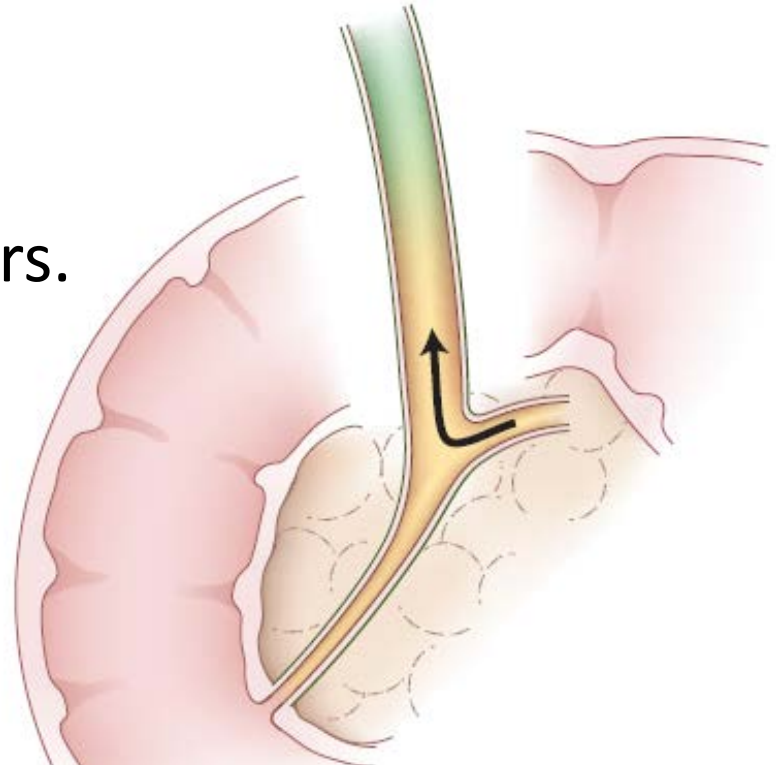


Type V

TYPE		prevalence
I	cystic	79.4%
II	diverticulum	2.7%
III	choledochocele	3.7%
IV	Intra & extra hepatic	13.6%
V	caroli’s disease	0.6%

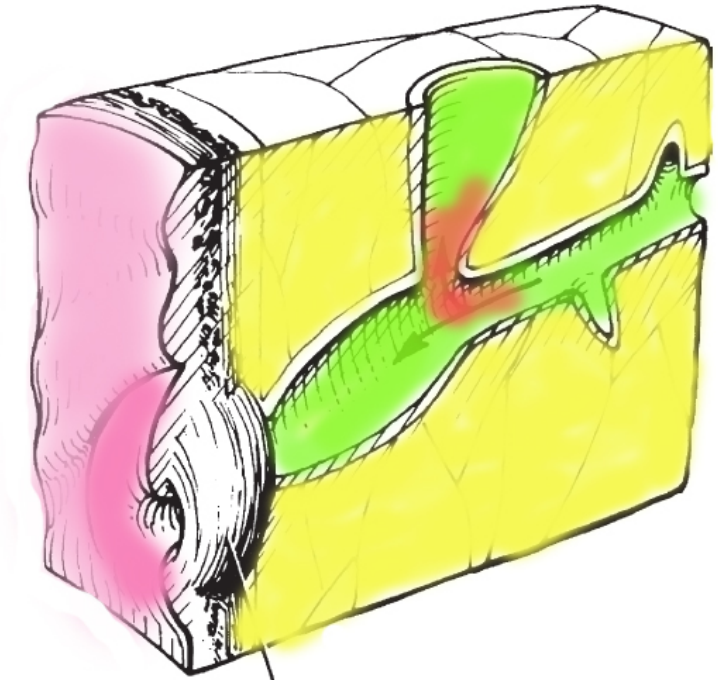
PATHOGENESIS

- Abnormal Pancreatico biliary junction (APBJ)
- Abnormal biliary epithelial proliferation before bile duct cannulation is complete
- Congenital weakness in the bile duct wall
- Bile duct obstruction or distension in the prenatal or neonatal periods
- Fetal viral infection
- genetic or environmental factors.



Abnormal Pancreatico biliary junction

- extramural pancreatico biliary junction
- characterized by a long common channel (over 2 cm)
- Increased reflux of pancreatic juice into the biliary tree



ASSOCIATED ANOMALIES

- Atresia: Biliary , Duodenal, Colonic ,
- septate gallbladder
- Congenital absence of the portal vein
- Pancreatic arteriovenous malformation
- Familial adenomatous polyposis
- Imperforate anus
- **Cardiac:** VSD
- **Renal:** polycystic kidney disease

PRESENTATION

Triad : –

- Abdominal pain
- Jaundice
- Right upper quadrant palpable mass

Infantile form

- Present before 12 months of age
- Present with obstructive jaundice, acholic stools & hepatomegaly
- Signs of hepatic fibrosis & cirrhosis

Adult form

- Anytime after 12 months of age
- May present with fever, nausea, vomiting & jaundice.
- Undiagnosed cases may present with cholelithiasis.

D/D

- Pancreatitis
- Acute cholangitis
- Biliary calculi
- Biliary Obstruction
- Bile Duct Tumors
- Cholangiocarcinoma

DIAGNOSIS

- U/S abdomen.
- CT scan .
- ERCP
- MRCP
- Intraop Cholangiography.
- Tc99 m IDA Scan.
- Liver function tests

OBJECTIVES OF IMAGING

- To classify
- To Mention extension
- To visualize pancreaticobiliary junction
- To rule out association with other diseases
- To search for associated complications

- *Prenatal diagnosis*

Ultrasound - Earliest diagnosis at 25 weeks

COMPLICATIONS

- Stones in GB, cyst, pancreatic duct, intrahepatic biliary tree
- Malignant transformation
- Recurrent pancreatitis
- Cholangitis/Cholecystitis
- Cyst rupture with bile peritonitis
- Bleeding
- Biliary cirrhosis + portal hypertension
- Portal vein thrombosis
- Hepatic abscess

EVOLUTION OF SURGERY

1. Aspiration, marsupialization, external drainage

External biliary fistula

2. cholecystostomy, tube drainage of cyst, hepatic duct drainage after cyst excision

High mortality

3. Cysto duodenostomy

Chronic recurrent cholangitis, biliary cirrhosis, portal HTN

4. loop Jejunostomy

Risk of malignancy

5.Cyst excision with Roux-en-Y hepatico Jejunostomy

6. conduits:

   proximal jejunal conduit

7. Sphincterotomy, Sphincteroplasty

8. Partial Hepatic lobeectomy

9.Liver transplantation

10. Laparoscopic Roux-en-Y- hepatico Jejunostomy

PRINCIPLE OF SURGERY

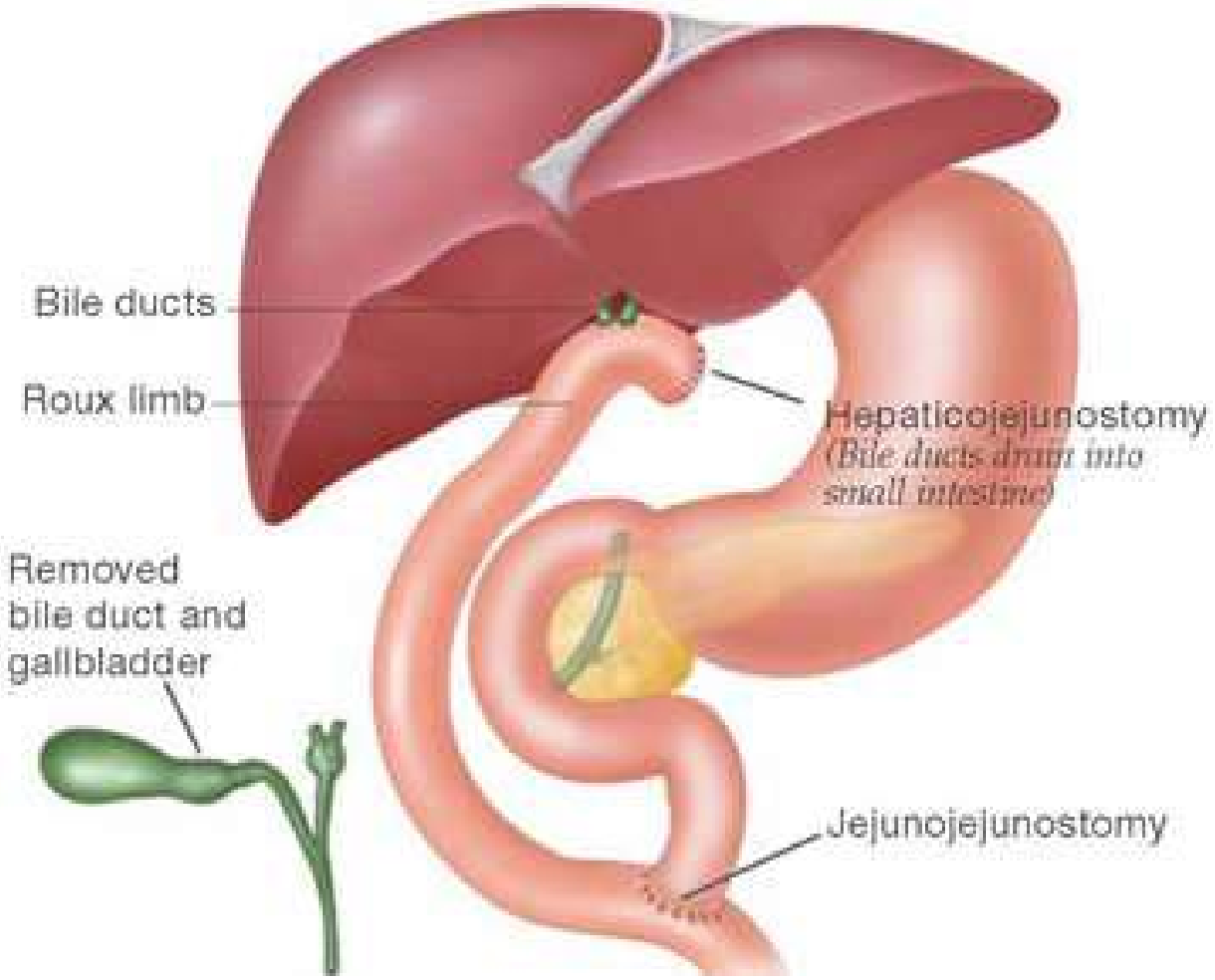
Treatment of choice for choledochal cysts is complete excision of the cyst
+
with construction of a biliary-enteric anastomosis to restore continuity with the GI tract

PROCEDURE

Excision of choledochal Cyst +

Roux-en-Y hepatojejunostomy +

Cholecystectomy



- Roux-en-Y Hepatojejunostomy

(retrocolic isoperistaltic functional end-to-side)

• Occasionally, Cyst adheres densely to the portal vein secondary to long-standing inflammatory reaction
•complete, full-thickness excision of the cyst may not be possible
• serosal surface of the duct is left adhering to the portal vein, while the mucosa of the cyst wall is obliterated by curettage or cautery
• Theoretically, this removes the risk of malignant transformation in that segment of the duct

- Choledochal Cyst dissected away from portal vein and hepatic artery (Lilly Technique?)
- Cyst wall opened till common hepatic duct junction

TREATMENT

- Type I: excision of the cyst,GB and reconstruction by Roux-en-Y hepatico-jejunostomy
- Type II: excision of the diverticulum and suturing of the CBD wall with or without stent
- Type III: endoscopic sphincterotomy/duodenotomy and deroofing.
- Type IV: Extrahepatic biliary resection, cholecystectomy, and biliary reconstruction.
- Type V: Liver transplantation,

POST OP COMPLICATIONS

- **EARLY :** pancreatitis, anastomotic leakage, cholangitis, wound infection.
- **LATE:** formation of intrahepatic strictures and stones, anastomotic stricture, malignancy, cirrhosis, and intrahepatic abscess formation.
- morbidity of 9–41% and mortality 0–3.3%.

Follow up

- Low dose cotrimoxazole for 6 weeks
- Review every 3 months for 1st year
- Anually there after.
- USG- Abdomen, LFT, Sr amylase

PROGNOSIS

- long-term results following resection of a benign choledochal cyst with biliary reconstruction are generally excellent, especially with type I cysts.
- Type IVa cyst patients have the greatest risk for intrahepatic calculi and stricture formation
- Patients with type IVa disease developed strictures at a rate 40%, with virtually all developing cholangitis.

THANK YOU