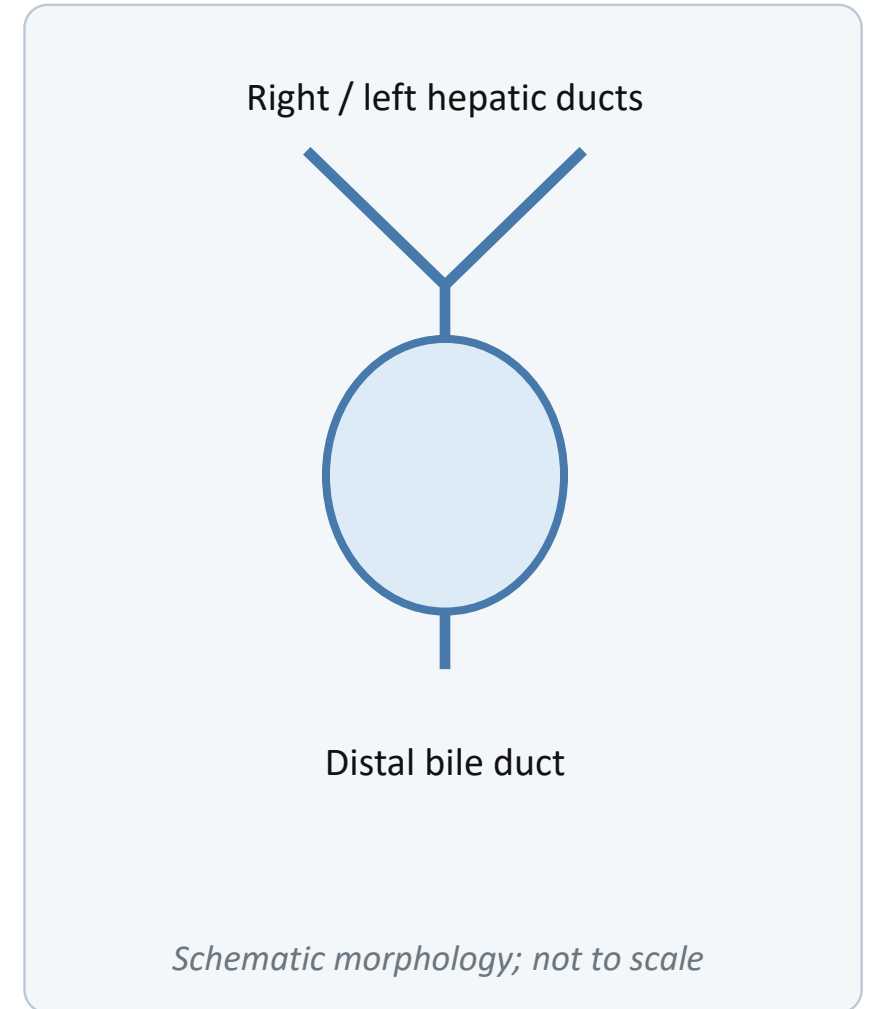


Choledochal Cyst

Congenital dilatation of the biliary tree

Pathogenesis • morphology • clinicopathologic consequences



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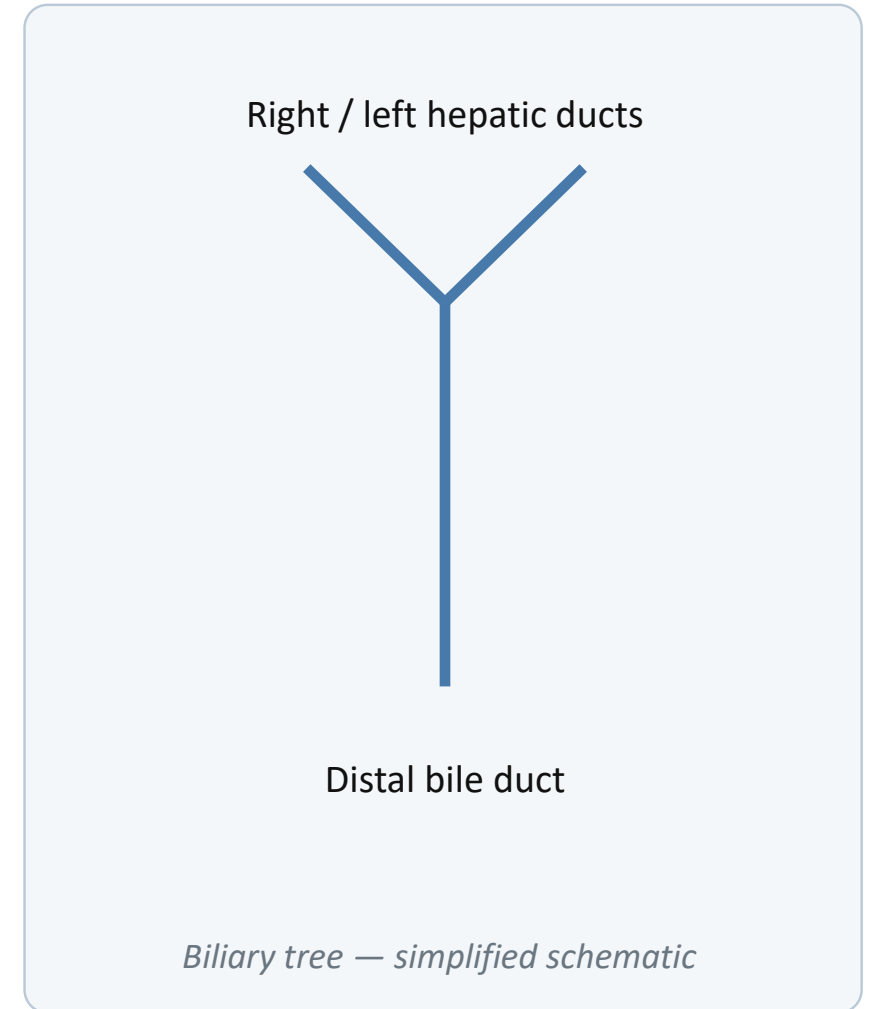
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Learning outcomes

- Define choledochal cyst and distinguish extrahepatic from intrahepatic duct disease.
- Classify major anatomic patterns and interpret the clinicopathologic consequences.
- Relate biliary stasis and epithelial injury to inflammation, stones and cancer risk.
- Outline imaging, pathology examination and principles of definitive treatment.

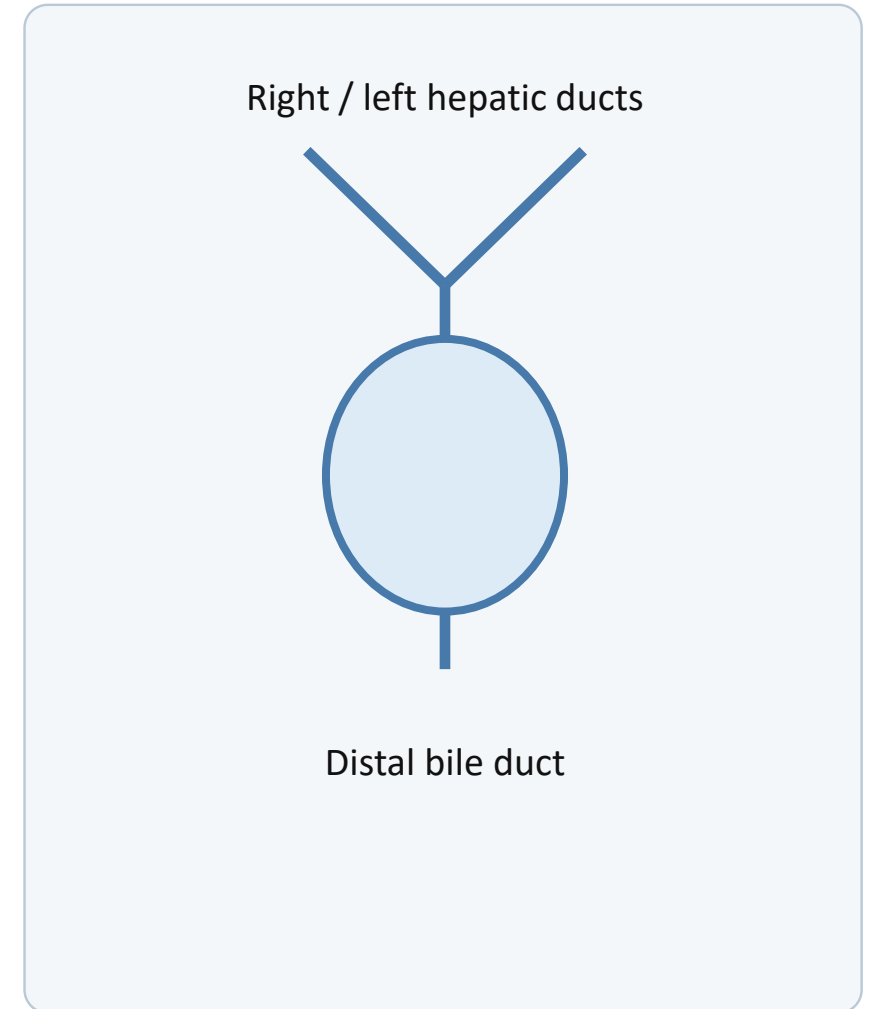
Normal drainage anatomy: An essential baseline

- Right and left hepatic ducts join to form the common hepatic duct.
- The cystic duct joins the common hepatic duct to form the common bile duct.
- The distal duct traverses the pancreatic region before entering the duodenum.
- Cyst location must be described relative to these ductal landmarks.



Definition: Congenital abnormal bile duct dilatation

- Choledochal cysts as congenital dilatations of the common bile duct.
- The term is used clinically for a spectrum of biliary duct cystic malformations.
- An important distinction is true ductal communication versus a separate cystic mass.
- The defining pathology is abnormal biliary duct architecture, not simply a cystic lesion near the liver.



Why anatomic location determines the diagnosis

- Extrahepatic disease involves the common hepatic or common bile duct.
- Intraduodenal dilatation has different morphology and treatment considerations.
- Intrahepatic duct ectasia suggests Caroli disease or combined involvement.
- The extent of involvement predicts risks and influences operative planning.

Duct involvement

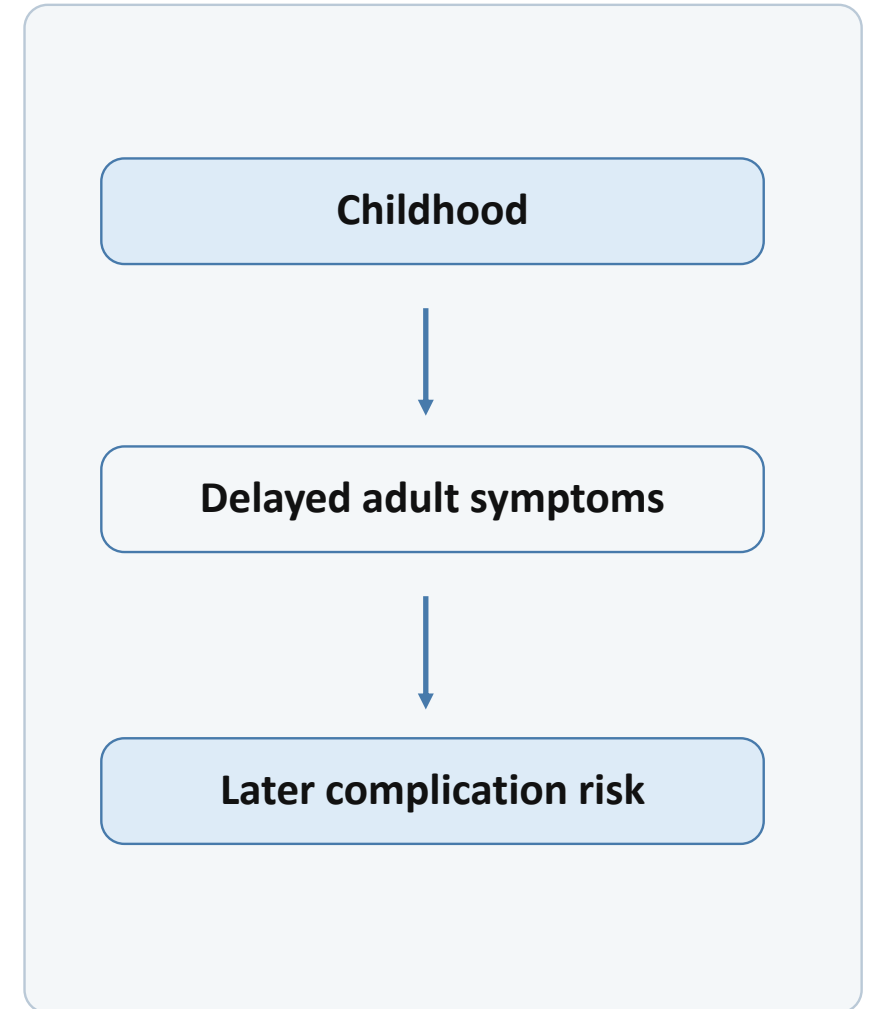


Extrahepatic? Intrahepatic?
Intraduodenal? Diverticulum?

Topology guides the type

Typical age at clinical presentation

- Most symptomatic cases present during childhood, often before age 10.
- Jaundice and recurrent abdominal pain are characteristic initial complaints.
- Approximately 20% become symptomatic only in adulthood (Robbins).
- Congenital origin does not require detection during infancy.



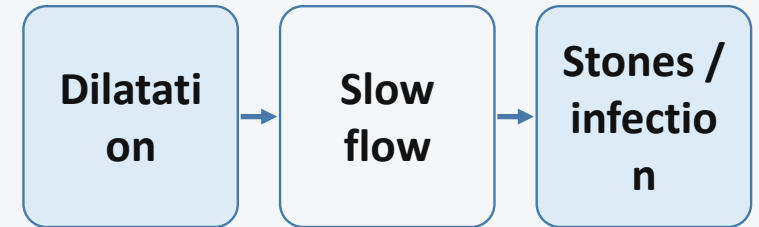
Female predominance is a reproducible feature

- A female-to-male ratio of approximately 3:1 to 4:1.
- The sex distribution helps characterize epidemiology, not establish an individual diagnosis.
- Symptoms and imaging remain essential for case identification.
- A male patient with typical biliary dilatation can still have the disorder.

**Robbins: Female-to-male
ratio $\approx$ 3:1–4:1**

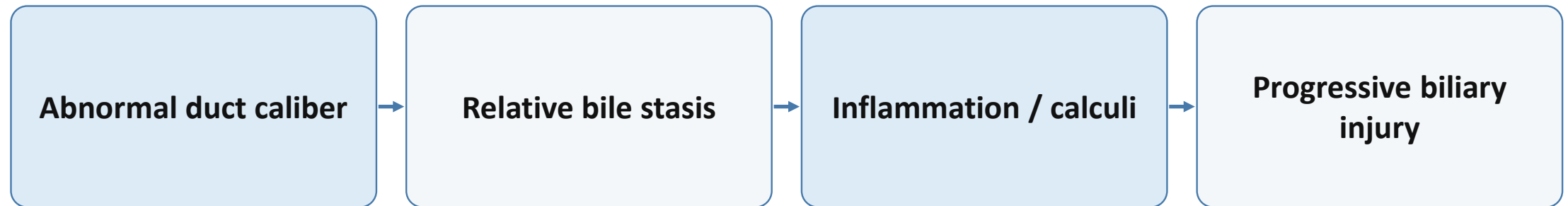
The presenting symptom is often biliary obstruction

- Abdominal pain may resemble recurrent biliary colic.
- Jaundice indicates impaired handling or drainage of biliary pigments.
- Episodes may recur because duct dilatation is a persistent structural abnormality.
- Additional symptoms may reflect stones, infection or pancreatitis.



Biliary stasis links anatomy to complications

A dilated and poorly draining duct changes the local biliary environment.



Dilatation is not merely a radiologic observation; It changes biliary physiology.

Pancreaticobiliary maljunction: Proposed driver

- An anomalous pancreaticobiliary junction may permit reciprocal ductal reflux.
- Pancreatic juice may enter the bile duct outside normal sphincteric regulation.
- Proteolytic pancreatic secretions can injure biliary epithelium.
- This model is important but does not account for every cyst subtype.

Abnormal duct junction

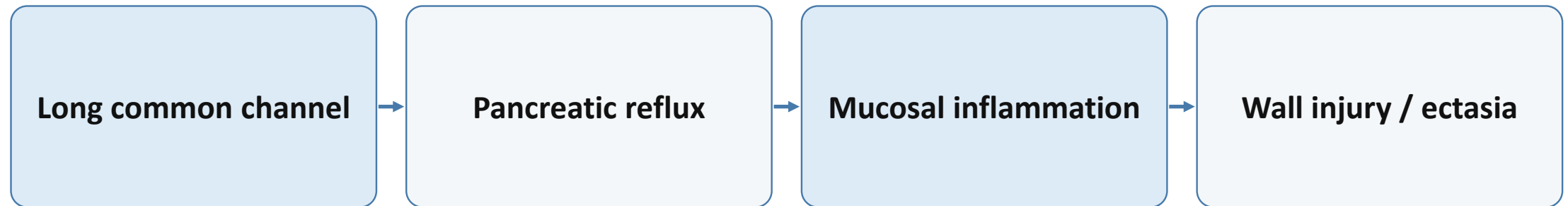


Pancreatic juice reflux



Chronic ductal injury

Proposed pancreatic enzyme injury cascade



Mechanistic framework: Reflux is a commonly accepted hypothesis, not a universal requirement.

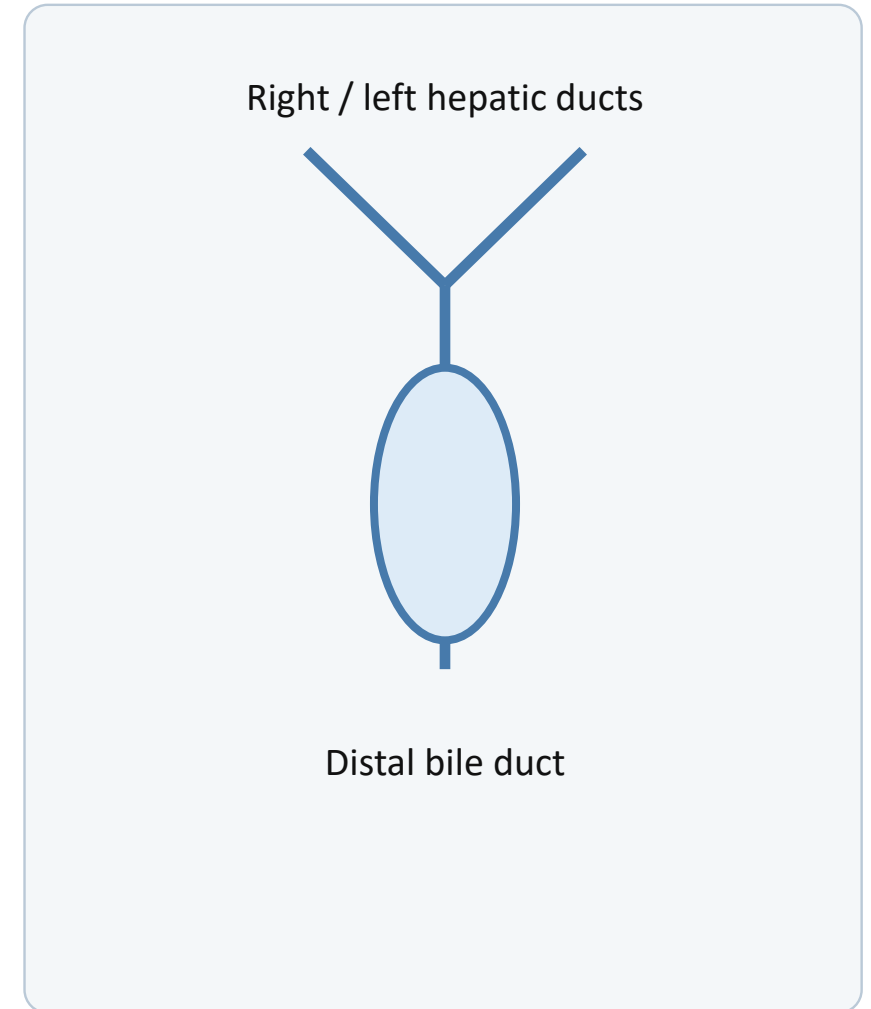
Pathogenesis is heterogeneous across subtypes

- Anomalous duct union is often associated with extrahepatic cystic dilatation.
- Developmental duct abnormalities are important for intrahepatic cystic disease.
- Some lesions, particularly choledochoceles, may differ in pathogenesis.
- An observed cyst does not prove the presence of pancreaticobiliary maljunction.

**No single mechanism
explains every ductal cyst**

Anatomic forms described directly by Robbins

- Segmental or cylindric dilatation of the common bile duct.
- Diverticula arising from extrahepatic bile ducts.
- Choledochoceles protruding into the duodenal lumen.
- Some cases coexist with cystic dilatation of intrahepatic ducts.

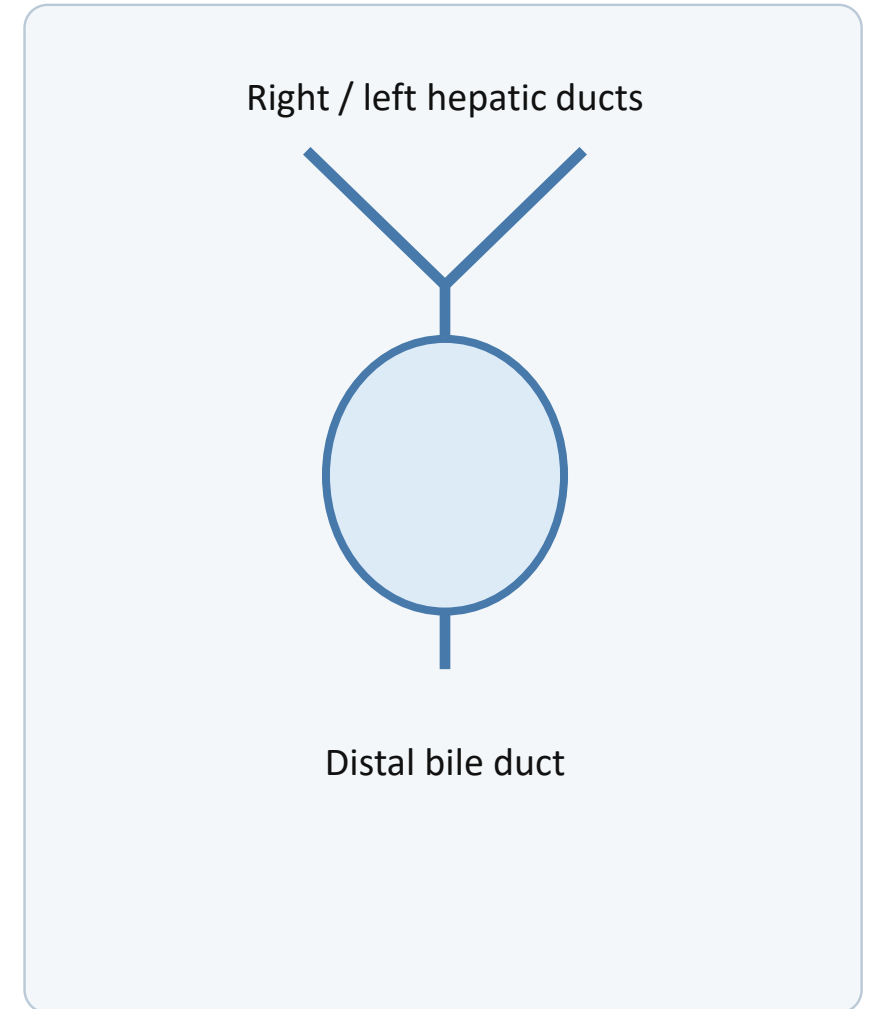


Todani classification: A descriptive anatomic framework

Type	Primary distribution
I	Single segmental, cystic or fusiform extrahepatic duct dilatation
II	True extrahepatic biliary diverticulum
III	Intraduodenal terminal duct dilatation: Choledochocele
IV	Multiple duct cysts: IV-A intra- and extrahepatic; IV-B extrahepatic only
V	Segmental intrahepatic duct ectasia (Caroli disease)

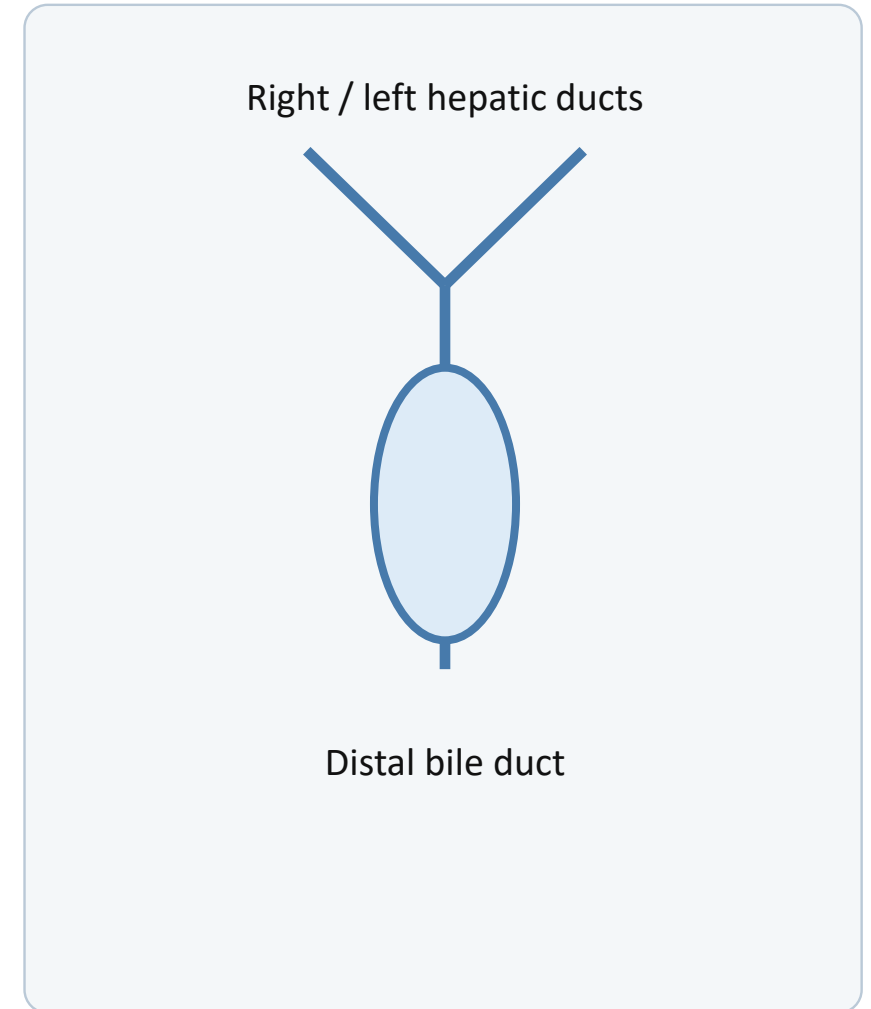
Type I: Isolated extrahepatic duct dilatation

- Abnormal enlargement involves the extrahepatic bile duct.
- Morphology may be cystic, segmental or fusiform.
- The communicating lumen is part of the biliary drainage pathway.
- Type I is the dominant anatomic pattern in many clinical series.



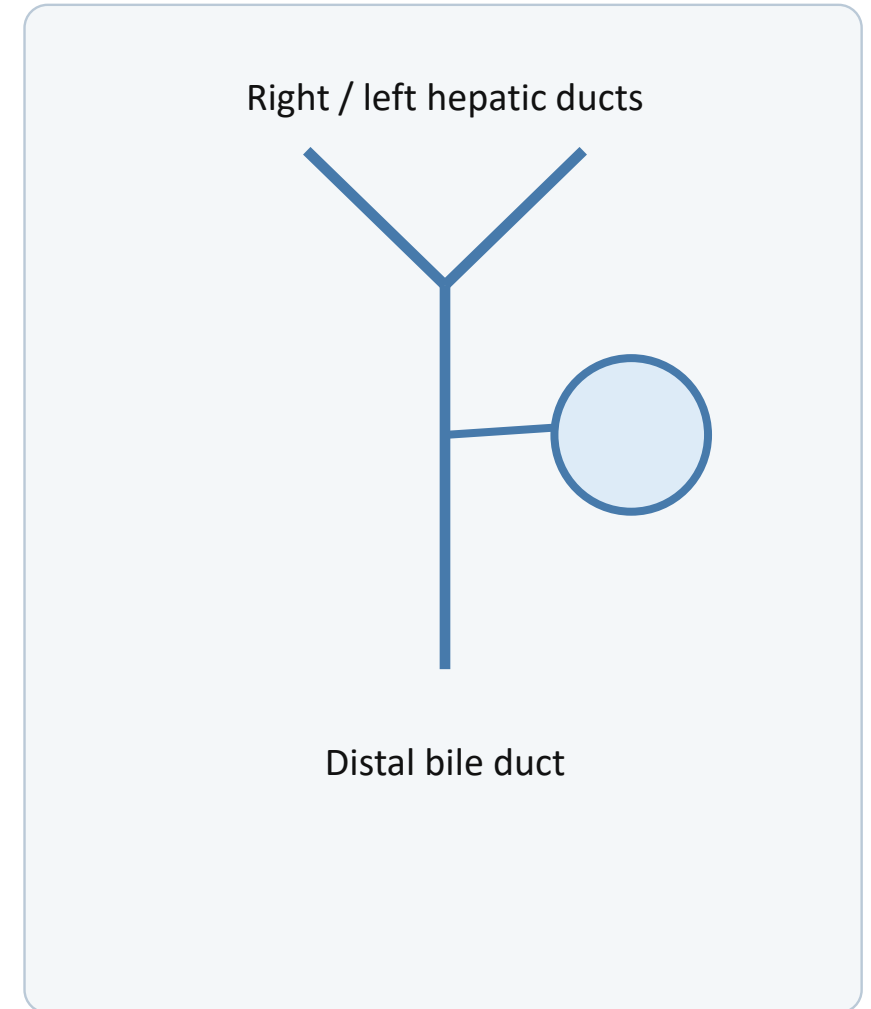
Type I subtypes: Morphology within a single group

- Type IA: Diffuse cystic dilatation of extrahepatic bile duct.
- Type IB: Focal segmental dilatation, often saccular.
- Type IC: Diffuse fusiform extrahepatic dilatation.
- These are morphology-based subdivisions; Naming conventions should be checked against the classification used locally.



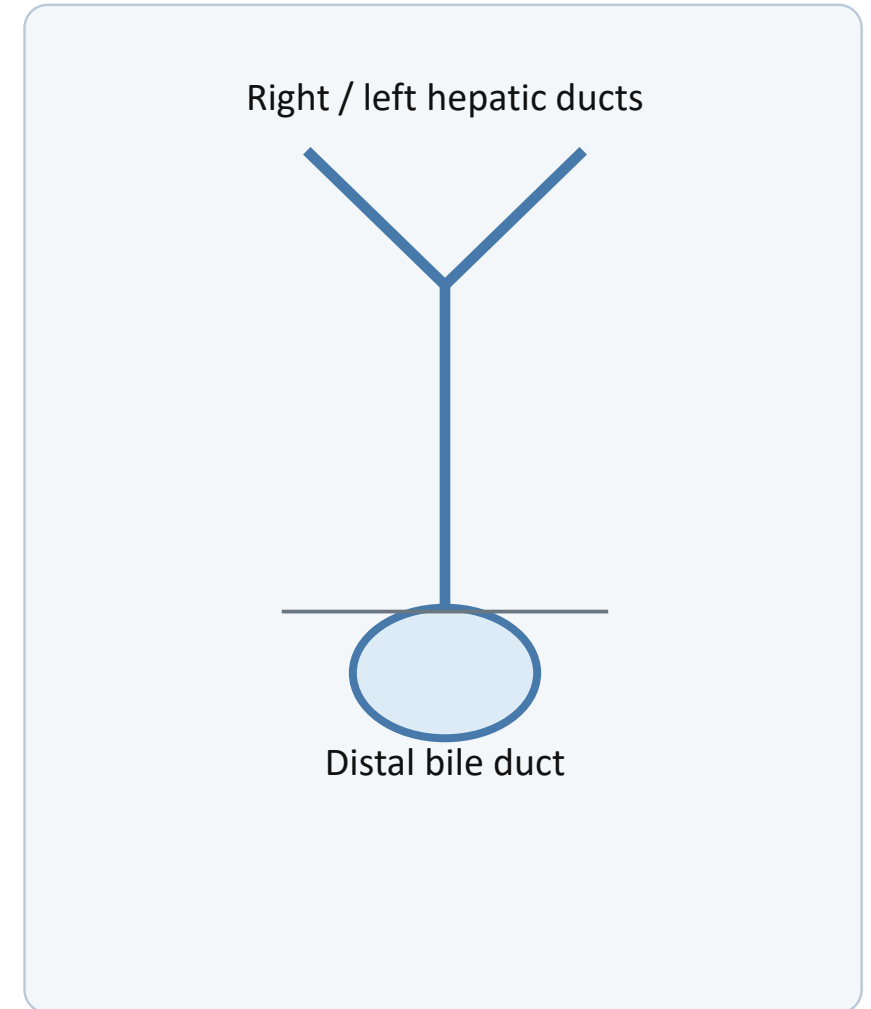
Type II: An extrahepatic bile duct diverticulum

- A focal sac protrudes from the wall of an extrahepatic bile duct.
- The main duct can remain comparatively normal in caliber.
- The diverticular neck may contribute to local stasis.
- Distinguish from a noncommunicating adjacent cyst.



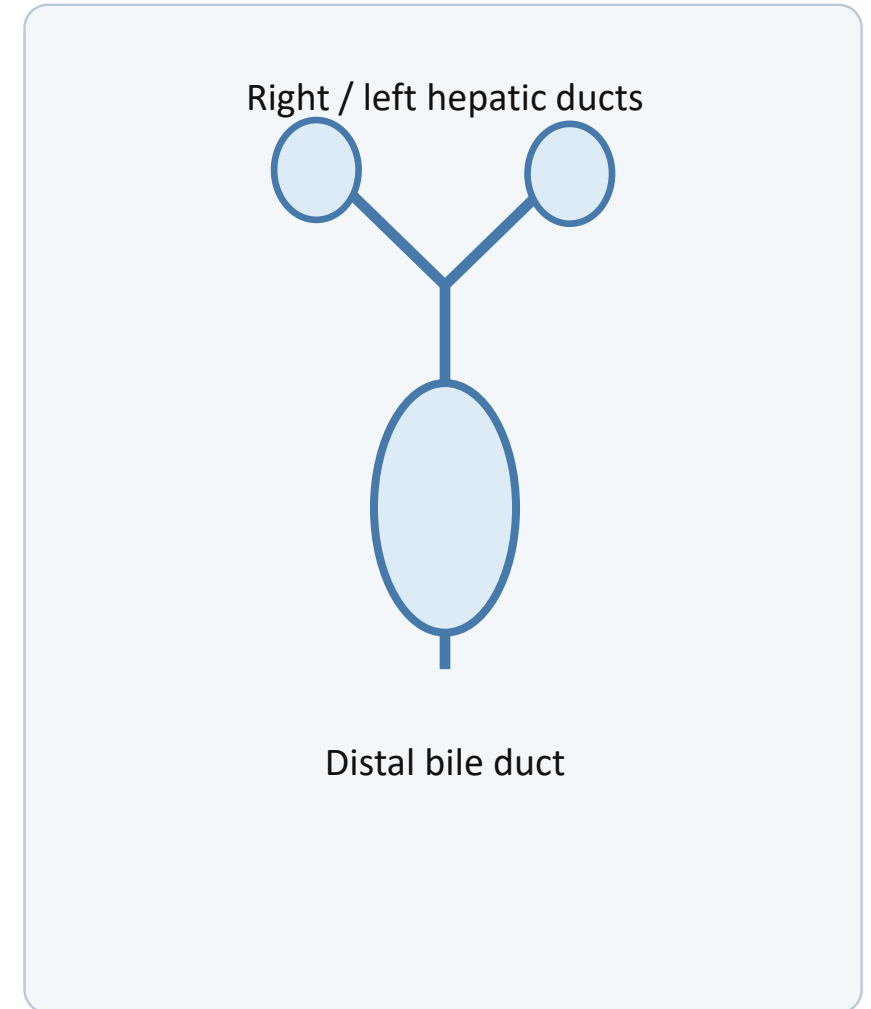
Type III: Choledochocoele

- Cystic enlargement occurs in the distal, intraduodenal duct segment.
- The lesion may bulge into the duodenal lumen.
- Its relationship to the ampulla and pancreatic duct must be mapped.
- Its epithelial lining and malignant potential differ from many large extrahepatic cysts.



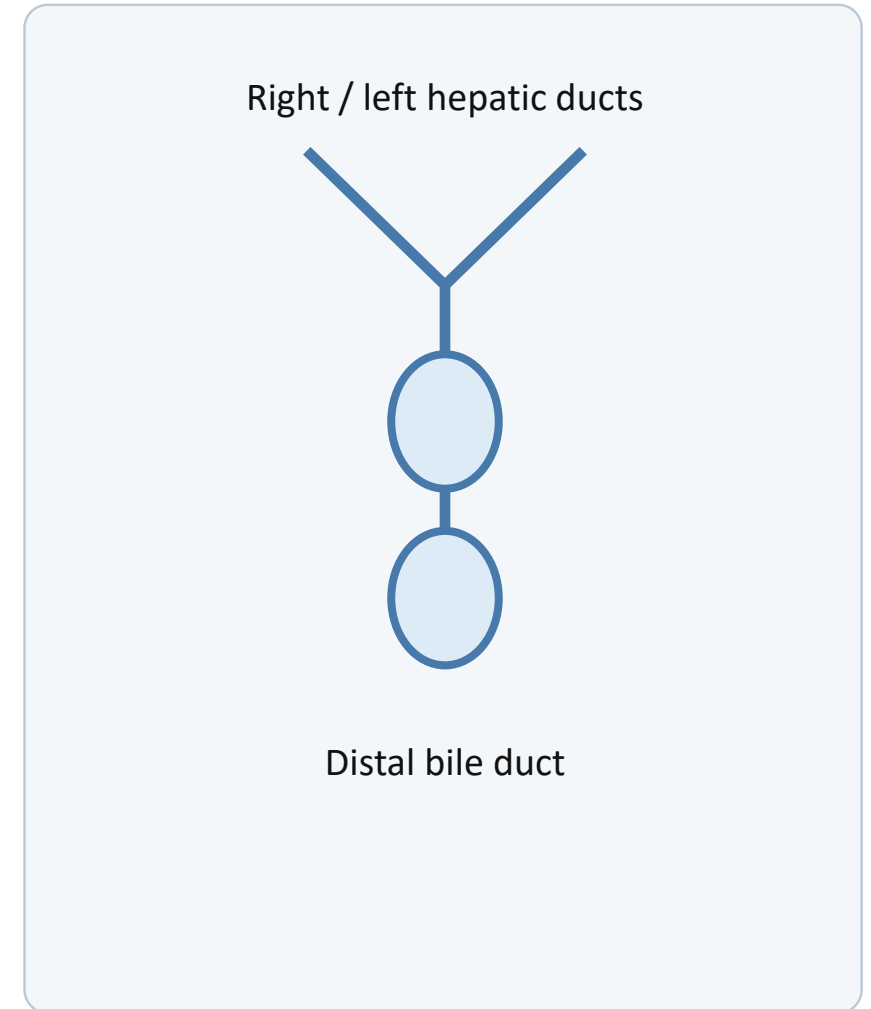
Type IV-A: Intrahepatic and extrahepatic involvement

- Extrahepatic ductal cystic dilatation coexists with intrahepatic duct ectasia.
- Intrahepatic strictures or stasis may persist after extrahepatic reconstruction.
- Mapping the distribution of intrahepatic lesions is essential.
- Management is individualized to the extent of ductal disease.



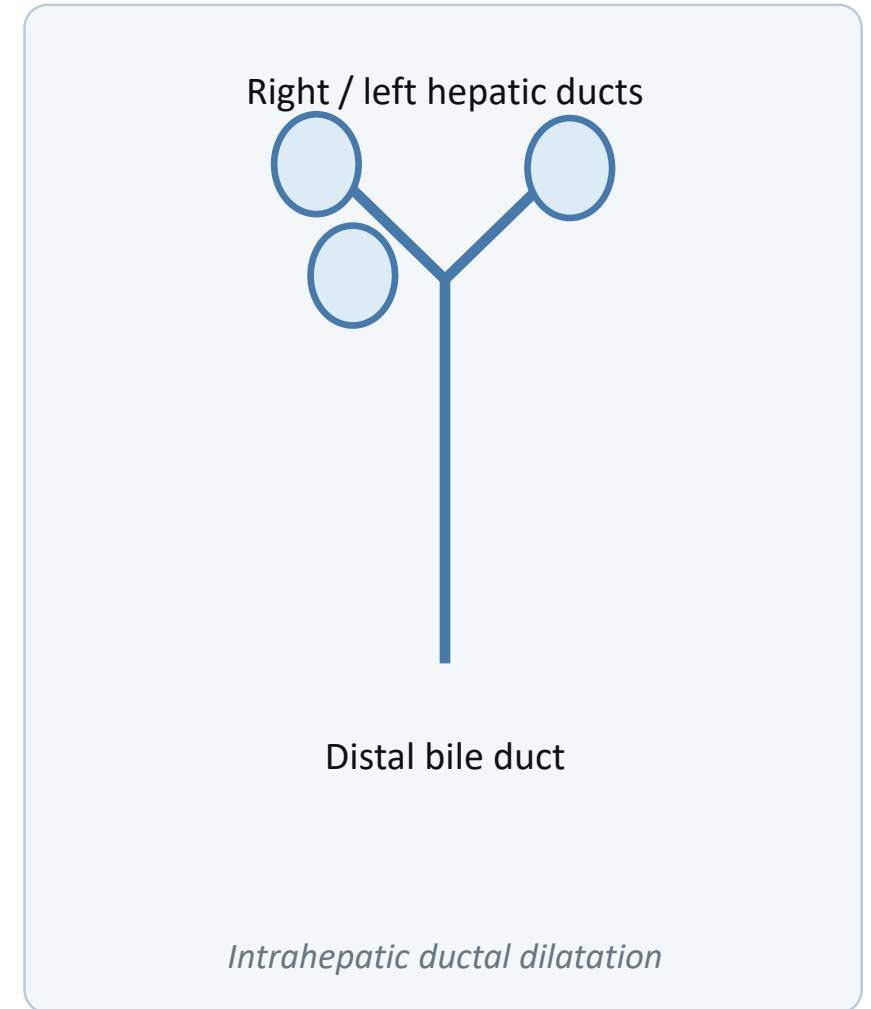
Type IV-B: Multiple extrahepatic cysts

- More than one extrahepatic bile duct segment is dilated.
- Intrahepatic ducts are not part of the defining pattern.
- The distinction from IV-A rests on documenting intrahepatic involvement.
- Complete anatomic mapping matters more than counting visible sacs alone.



Type V: Caroli disease

- Multifocal, segmental cystic dilatation affects larger intrahepatic ducts.
- The duct spaces remain continuous with the biliary tree.
- Caroli disease under fibropolycystic liver disease.
- Its placement in Todani classification does not make all Caroli disease an extrahepatic cyst.



Caroli disease versus Caroli syndrome

Entity	Defining structural association
Caroli disease	Multifocal cystic dilatation of large intrahepatic bile ducts
Caroli syndrome	Caroli-type duct dilatation plus congenital hepatic fibrosis
Congenital hepatic fibrosis	Broad fibrous portal expansions with abnormal bile duct profiles
Choledochal cyst	Typically extrahepatic duct dilatation; May coexist with intrahepatic disease

Developmental duct plate disease is adjacent context

- Ductal plate malformations are a developmental spectrum within fibropolycystic disease.
- Persistence of embryonic periportal ductal plates produces abnormal duct architecture.
- Caroli syndrome combines intrahepatic duct ectasia and congenital hepatic fibrosis.
- Avoid assuming every extrahepatic choledochal cyst is caused by ductal plate malformation.

Prolonged obstruction

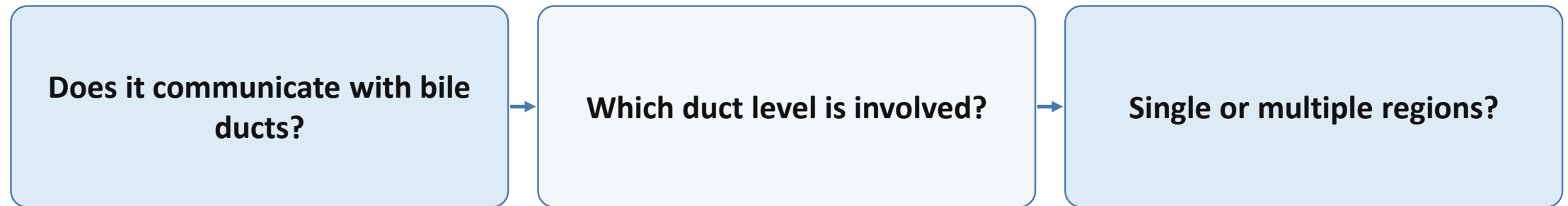


Inflammation / injury



Biliary fibrosis

Three diagnostic questions before assigning a type



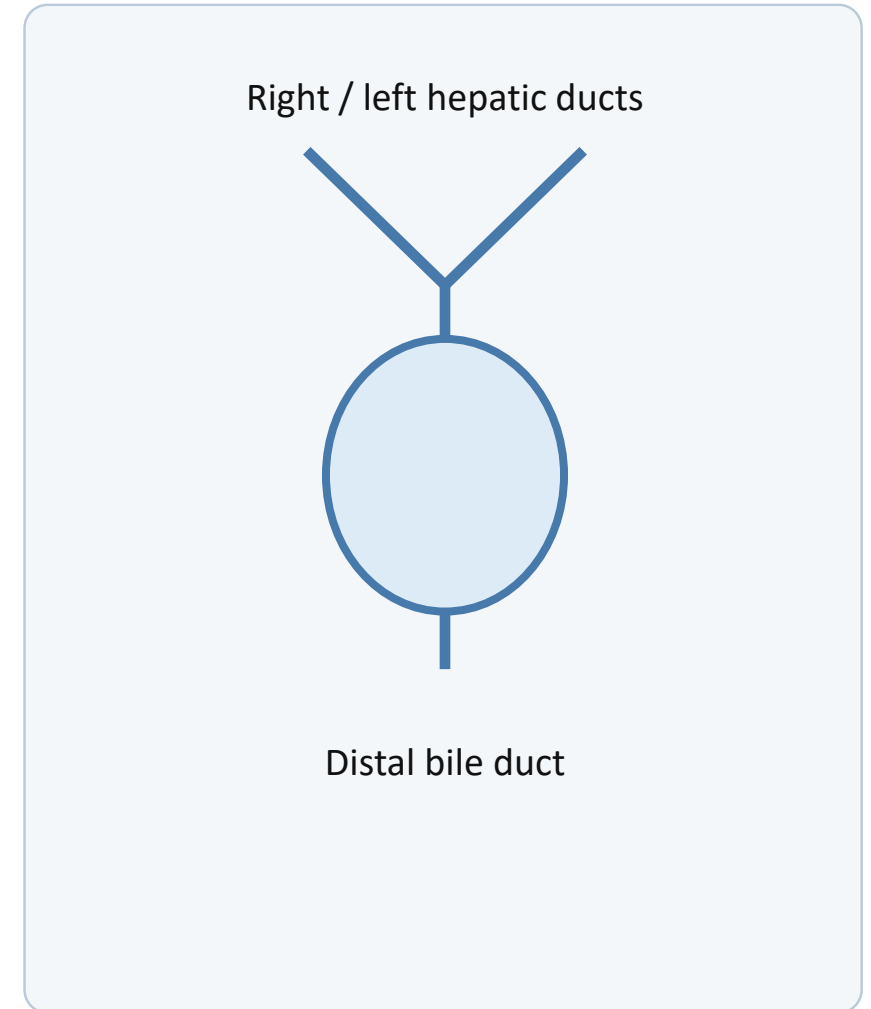
Classify only after demonstrating the complete ductal topology.

Distinguishing congenital cyst from secondary dilatation

Bile duct cyst	Secondary obstructive duct ectasia
Congenital structural morphology	Duct enlarges because an acquired distal obstruction impedes flow
Usually patterned ductal malformation	Dilation may involve ducts upstream of stone or tumor
Classification describes the lesion itself	Search for the obstructing cause
Can develop secondary stones/strictures	Not every enlarged duct is a choledochal cyst

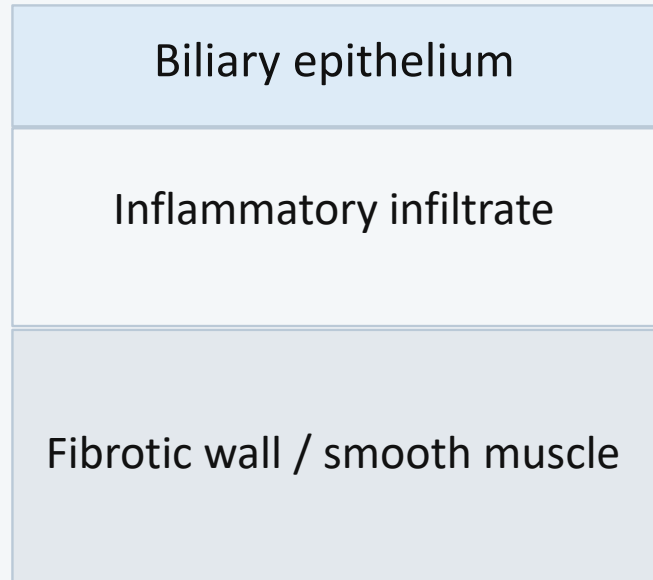
Gross pathology: An expanded bile-filled duct

- The specimen may show localized or elongated enlargement of an extrahepatic duct.
- Wall thickness varies with fibrosis and chronic inflammation.
- The lumen may contain bile, sludge or calculi.
- Mural nodules or focal thickening merit targeted sampling for dysplasia or carcinoma.



The duct wall may become fibrotic

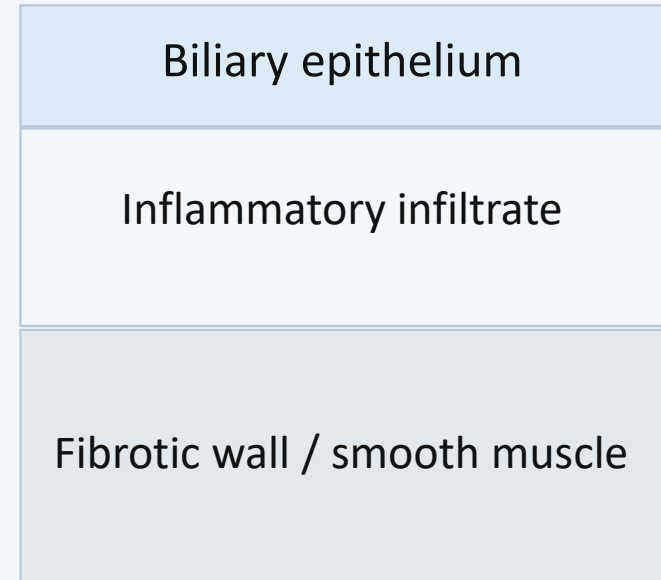
- Expanded ducts can show dense mural collagen with scattered smooth muscle bundles.
- Chronic injury and repair contribute to progressive wall remodeling.
- Inflammatory infiltrates may be present in the wall.
- Severe epithelial injury can leave segments attenuated or denuded.



Schematic wall layers; Not a micrograph

Mucosal lining: Not uniformly preserved

- Biliary-type columnar epithelium can line extrahepatic duct cysts.
- The lining may be flattened, inflamed, hyperplastic or focally lost.
- Choledochoceles may have duodenal-type lining in some cases.
- Sampling and lesion type affect which changes are identified.



Schematic wall layers; Not a micrograph

Histologic chronic inflammation is mechanistically important

- Lymphocytic inflammation may accompany fibrotic remodeling.
- Superimposed neutrophilic inflammation raises concern for cholangitis.
- Reactive atypia can resemble epithelial dysplasia.
- Correlate cytology with architectural disorder, inflammation and clinical context.

Biliary epithelium

Inflammatory infiltrate

Fibrotic wall / smooth muscle

Schematic wall layers; Not a micrograph

Epithelial metaplasia, dysplasia and malignancy

- Recurrent inflammation can be associated with epithelial metaplasia.
- Dysplasia is a premalignant epithelial alteration requiring careful evaluation.
- Invasive adenocarcinoma must be distinguished from reactive atypia.
- Robbins explicitly identifies increased adenocarcinoma risk.



**Potential dysplasia /
neoplasia**

Pathology sampling: The entire duct matters

- Document cyst extent and relationship to resection margins.
- Inspect focally thickened, ulcerated or nodular mucosa.
- Sample suspicious regions separately and include representative cyst wall.
- Report dysplasia, invasive carcinoma and margin involvement if identified.

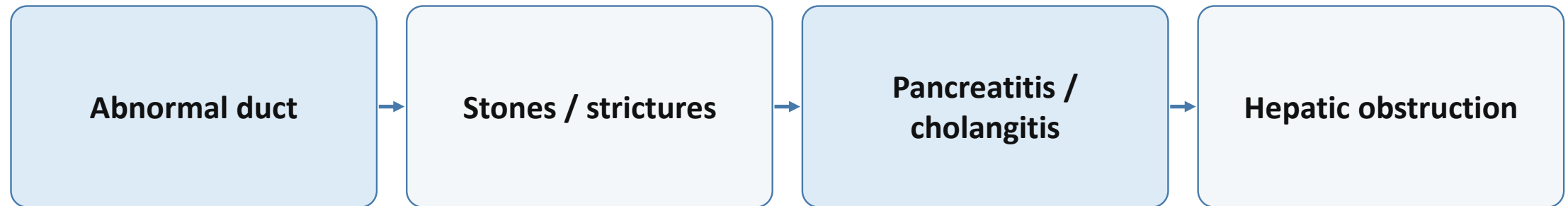
Duct involvement



Extrahepatic? Intrahepatic?
Intraduodenal? Diverticulum?

Topology guides the type

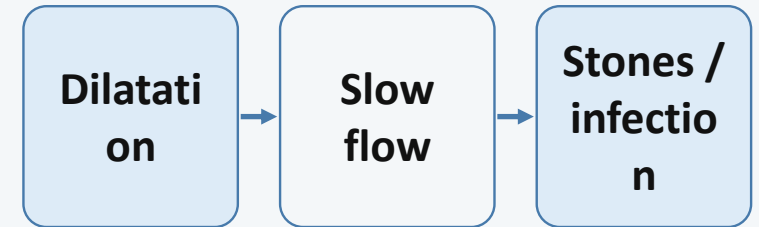
Complication framework: One malformation, several injuries



Robbins also identifies an elevated risk of adenocarcinoma.

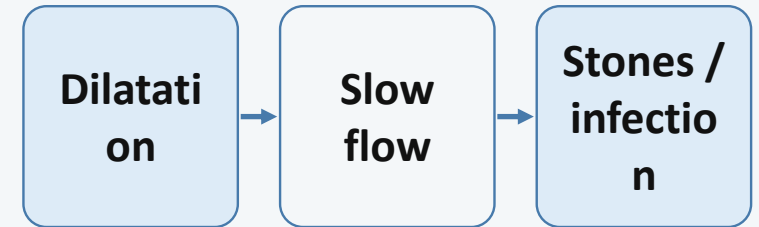
Bile duct stones form in a stagnant system

- Abnormal duct caliber can promote impaired bile clearance.
- Biliary sludge and stones may develop in the dilated segments.
- Calculi can aggravate obstruction and cause intermittent pain.
- Recurrent calculi should trigger investigation for underlying ductal pathology.



Stricture produces a second level of obstruction

- Luminal narrowing may occur at a duct segment or junction.
- A stricture impairs emptying and reinforces bile retention.
- Upstream ductal dilatation may worsen with progressive stenosis.
- Persistent jaundice or cholangitis warrants attention to fixed narrowing.



Ascending cholangitis: An infected obstructed system

- Biliary stasis favors infection when bacteria enter the ductal tree.
- Typical clinical findings can include fever, right upper quadrant pain and jaundice.
- The condition can progress rapidly and requires urgent clinical assessment.
- Definitive prevention depends on resolving the obstructive anatomy.

Prolonged obstruction



Inflammation / injury



Biliary fibrosis

Pancreatitis is a recognized complication

- Abnormal distal ductal anatomy may interfere with pancreatic outflow.
- Reflux or obstruction may provoke pancreatic inflammation.
- Pancreatitis in a child with bile duct dilatation strengthens clinical suspicion.
- Pancreatic duct junction anatomy affects imaging and management.

Abnormal duct junction



Pancreatic juice reflux



Chronic ductal injury

Liver injury from chronic impaired biliary drainage

- Chronic large-duct obstruction can produce cholestatic liver injury.
- Recurrent cholangitis adds inflammatory parenchymal injury.
- Secondary biliary fibrosis may accumulate with prolonged obstruction.
- Portal hypertension can emerge in advanced chronic biliary disease.

Prolonged obstruction



Inflammation / injury



Biliary fibrosis

Adenocarcinoma is the key long-term concern

- Choledochal cysts carry a higher risk of biliary adenocarcinoma.
- Risk differs by lesion type, age and associated abnormalities.
- Malignancy may arise in abnormal ductal epithelium.
- Cancer risk is one reason simple internal drainage is inadequate for many cysts.



**Potential dysplasia /
neoplasia**

Do not present one universal cancer-risk percentage

- Published risk estimates vary across cohorts and age distributions.
- Different studies pool different Todani types and follow-up periods.
- Preoperative cancer and metachronous cancer are distinct outcomes.
- The robust teaching point is increased risk and continued surveillance.



**Potential dysplasia /
neoplasia**

Infancy: Obstructive manifestations may dominate

- An infant may be evaluated for cholestatic jaundice.
- A large cystic duct lesion may become evident on ultrasound.
- Biliary atresia and other neonatal cholestatic causes remain key differential diagnoses.
- Prompt anatomic clarification helps avoid delay in definitive care.

Pattern recognition

Persistent jaundice
requires assessment of the
entire biliary system.

Childhood: Recurrent pain and jaundice

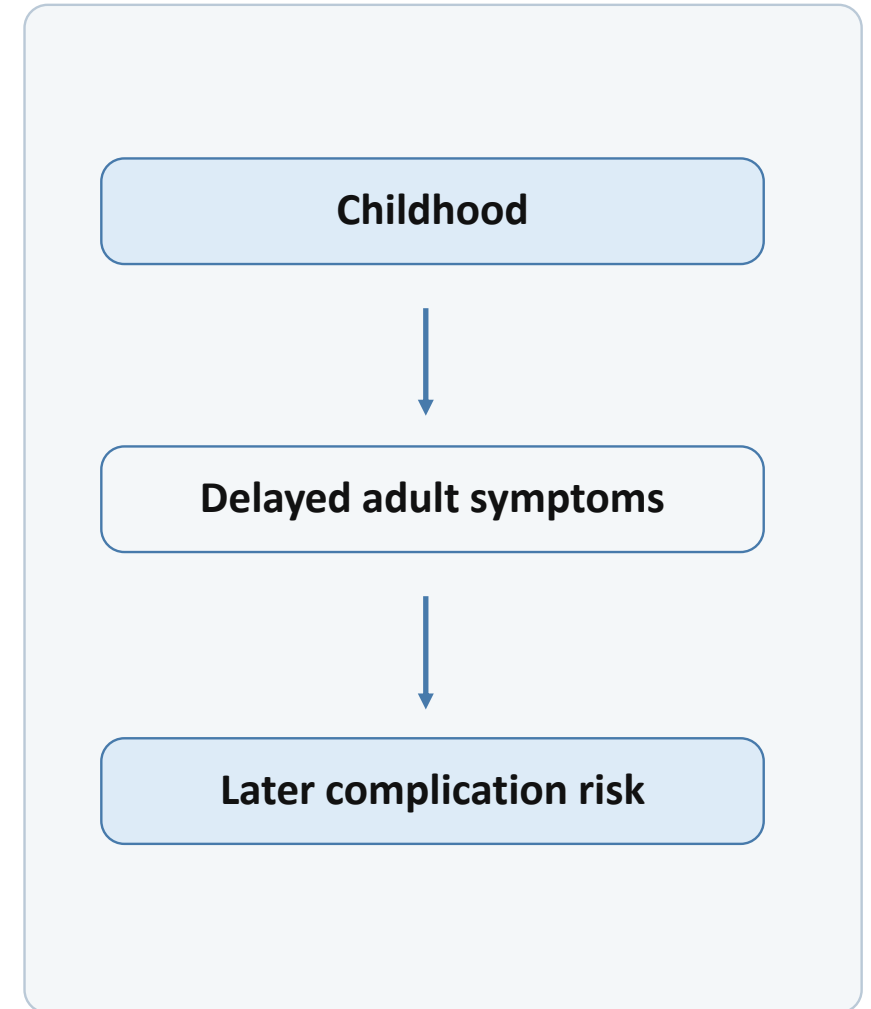
- Recurrent abdominal pain may be interpreted initially as biliary colic.
- Intermittent jaundice may reflect variable obstruction.
- Imaging may detect an enlarged communicating extrahepatic duct.
- Consider associated stone disease, cholangitis and pancreatitis.

Pattern recognition

Recurrent biliary pain is not specific to gallstones.

Adulthood: Symptoms can appear after years

- About one fifth become symptomatic only in adulthood.
- Adults may present with pain, cholangitis, pancreatitis or incidental imaging findings.
- Associated stones or strictures may obscure the congenital anomaly.
- Adult presentations require specific attention to malignant change.



The classic clinical triad is not a screening rule

- The historical triad includes abdominal pain, jaundice and a palpable abdominal mass.
- Many affected patients do not have all three findings.
- Symptoms may be nonspecific and vary with age or complications.
- Absence of a palpable mass does not exclude the diagnosis.

Pattern recognition

Abdominal pain + jaundice
+ palpable mass

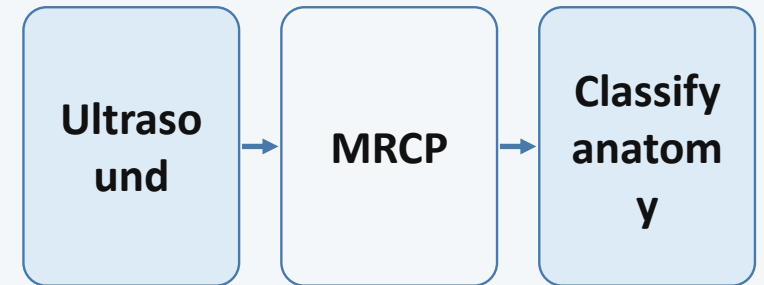
Laboratory tests describe injury, not anatomy

- Bilirubin may rise during clinically significant biliary obstruction.
- Cholestatic biochemical abnormalities may accompany impaired drainage.
- Inflammatory markers can increase during cholangitis.
- No single routine serum assay defines a choledochal cyst type.

Imaging establishes the ductal pattern

Ultrasonography: First-line structural assessment

- Ultrasound can detect a cystic lesion near the hepatic hilum or extrahepatic duct.
- Continuity with the biliary tree is an important observation.
- Assess intrahepatic duct dilatation and associated gallbladder findings.
- Incomplete visualization warrants more comprehensive ductal imaging.



**Clinical correlation + HPB
assessment**

MRCP: Map the entire biliary tree

- Magnetic resonance cholangiopancreatography delineates fluid-filled ducts.
- It helps characterize extrahepatic and intrahepatic disease distribution.
- The distal duct, ampullary region and pancreaticobiliary junction should be assessed.
- Noninvasive anatomic mapping helps guide surgical planning.

Duct involvement

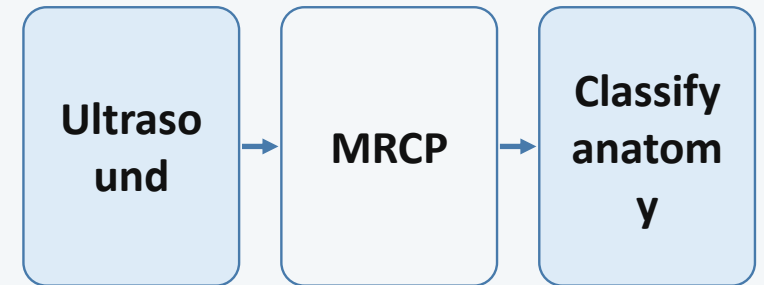


Extrahepatic? Intrahepatic?
Intraduodenal? Diverticulum?

Topology guides the type

CT and ERCP have selective complementary roles

- Cross-sectional imaging may help evaluate mass lesions or complications.
- ERCP can provide ductal detail and therapeutic access when indicated.
- ERCP is invasive and carries complications, including pancreatitis.
- Use each modality for a defined diagnostic or therapeutic question.



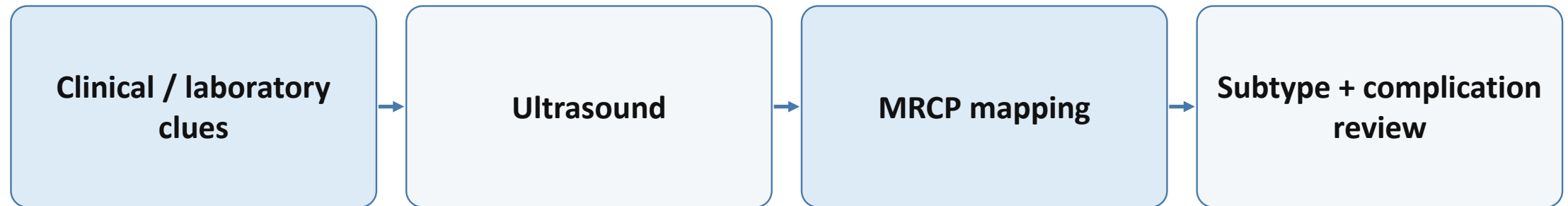
**Clinical correlation + HPB
assessment**

A complete radiology report should describe topology

Report element	Reason it matters
Exact dilated duct segments	Determines whether disease is extrahepatic or intrahepatic
Cystic / fusiform / diverticular	Supports morphological classification
Distal duct and pancreatic junction	Assesses relevant anomalous anatomy
Stones, stenosis, filling defects	Identifies complications or suspicious obstruction
Intrahepatic involvement	Changes operative planning and surveillance

Diagnostic synthesis: Structural recognition first

Symptoms raise suspicion; Duct anatomy establishes the lesion.



Escalate to an experienced hepatobiliary team when complex anatomy is identified.

Differential diagnosis: Pediatric jaundice

Consideration	Discriminating question
Choledochal cyst	Is there congenital cystic dilatation communicating with ducts?
Biliary atresia	Are extrahepatic drainage pathways obliterated or absent?
Obstructing calculus	Is there a focal intraluminal stone and upstream ectasia?
Congenital hepatic fibrosis / Caroli	Are intrahepatic duct and portal fibrosis patterns dominant?

Differential diagnosis: Cyst near the bile duct

Cystic lesion	Distinguishing feature
Choledochal cyst	Cyst communicates with biliary ductal system
Simple hepatic cyst	No ductal communication
Pancreatic pseudocyst	Pancreatic origin or relevant pancreatitis history
Duodenal duplication cyst	Intestinal wall relation and no typical bile-duct origin

Management is dictated by cyst class and extent

- Anatomy, symptoms, complications and cancer risk determine treatment.
- Types I and IV usually require excision of abnormal extrahepatic duct.
- Restoration of physiologic bile drainage is part of definitive reconstruction.
- Complex intrahepatic disease requires individualized hepatobiliary assessment.

Excise abnormal duct



Re-establish bile drainage



Long-term follow-up

Type I: Complete excision and reconstruction

- Excision aims to remove at-risk abnormal extrahepatic biliary mucosa.
- Reconstruction commonly uses Roux-en-Y hepaticojejunostomy.
- Associated gallbladder removal is often part of standard excision.
- Residual strictures or duct abnormalities must be considered in surgical planning.

Excise abnormal duct



Re-establish bile drainage



Long-term follow-up

Type II / III: Treatment differs by anatomy

- Type II diverticula may be treated by excision of the diverticular lesion.
- Type III choledochoceles may be considered for endoscopic therapy in suitable cases.
- The presence of dysplasia, obstruction or complex ductal anatomy alters choices.
- Management recommendations should be individualized by the treating team.

Duct involvement

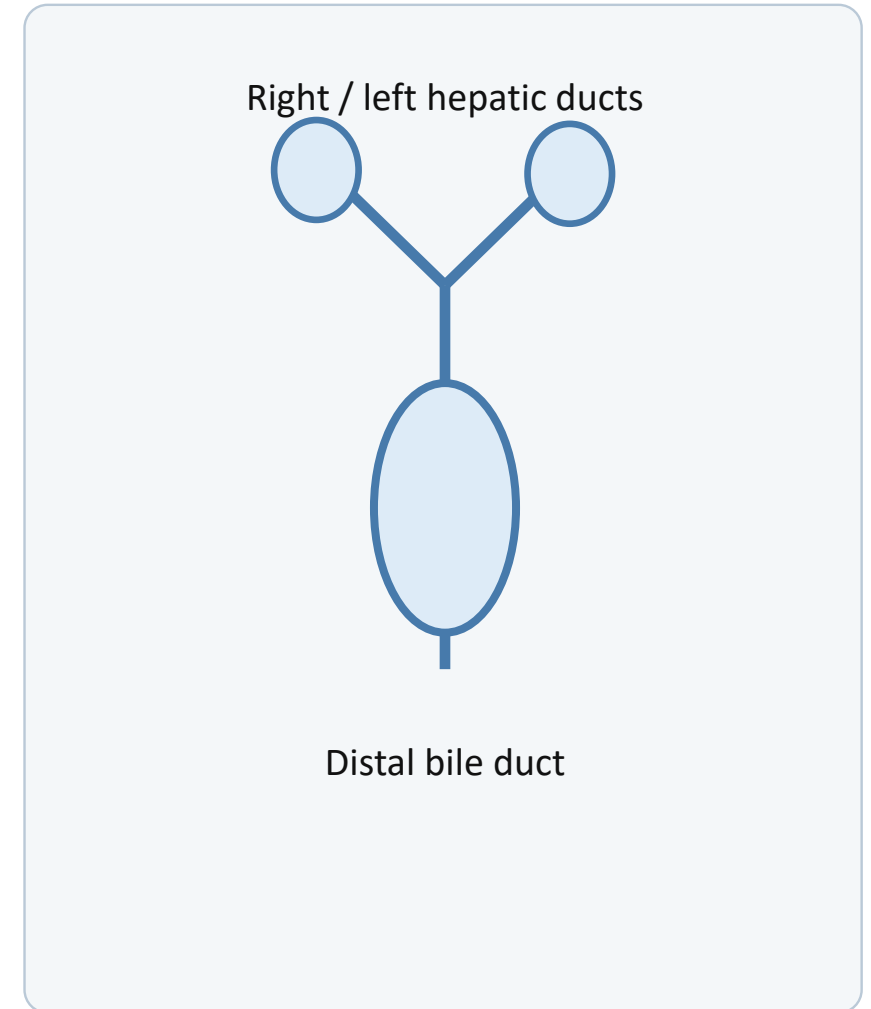


Extrahepatic? Intrahepatic?
Intraduodenal? Diverticulum?

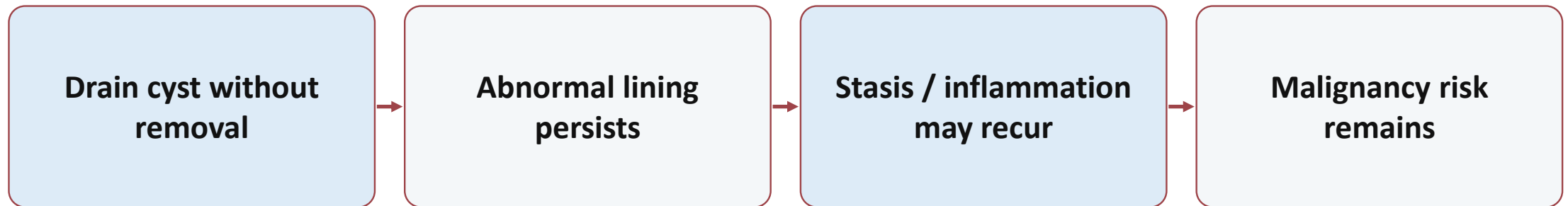
Topology guides the type

Type IV / V: Address intrahepatic disease burden

- Type IV-A disease requires evaluation of both intra- and extrahepatic ducts.
- Persistent intrahepatic strictures and stones may require further intervention.
- Localized Caroli disease may be approached differently from diffuse involvement.
- Selected severe cases may require major hepatic surgery or transplantation.



Why internal drainage alone is inadequate



Complete excision is preferred for lesions with substantial malignancy risk.

After surgery: Early and late complications differ

- Early complications relate to surgery and biliary reconstruction.
- Anastomotic strictures may later impair drainage.
- Recurrent cholangitis or intrahepatic stones can develop.
- Cancer risk may remain elevated despite complete excision.

Excise abnormal duct



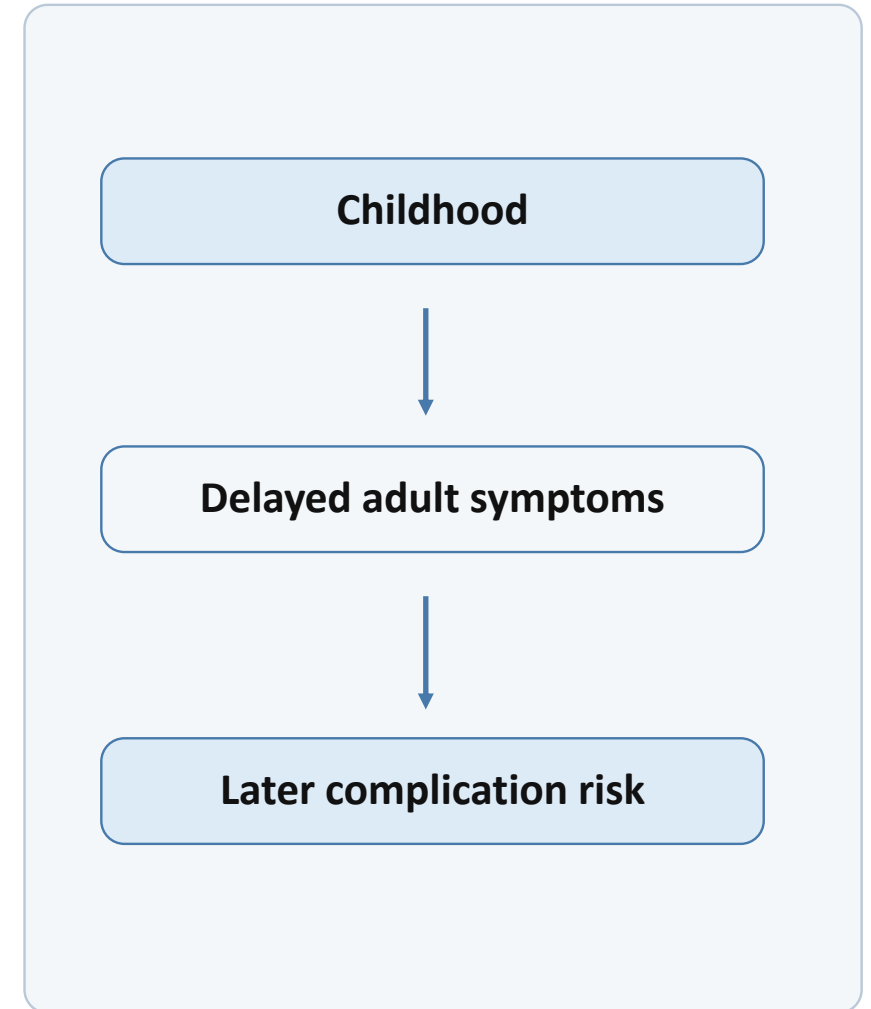
Re-establish bile drainage



Long-term follow-up

Long-term follow-up is an active safety measure

- Assess patients for recurrent pain, jaundice, fever and biochemical cholestasis.
- Follow-up imaging may be required to evaluate strictures or stones.
- Surveillance is particularly important after type I and IV disease.
- Exact intervals should follow specialist guidance and patient-specific risk.



Case 1: Childhood recurrent jaundice

An 8-year-old has intermittent abdominal pain and jaundice.

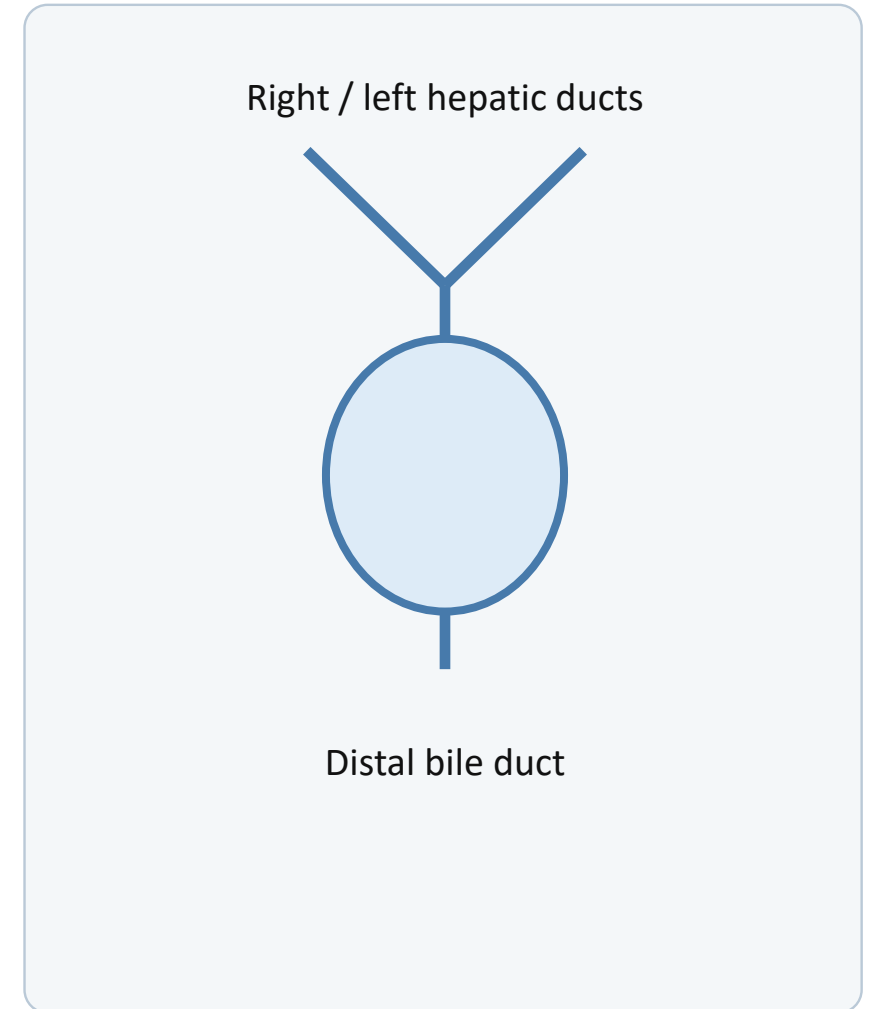
- Ultrasound: Smooth cystic enlargement of the extrahepatic bile duct.
- Intrahepatic ducts appear comparatively normal.
- No obstructing stone is demonstrated.
- Question: Which structural category is most likely?

Pattern recognition

Identify the involved duct
before naming the subtype.

Case 1: Interpretation and next step

- An isolated extrahepatic cystic lesion supports a type I pattern.
- MRCP can define proximal extent and distal duct anatomy.
- Assess for pancreaticobiliary maljunction and associated complications.
- Specialist evaluation is appropriate because definitive excision is usually advised.



Case 2: Pain, infection and intrahepatic duct ectasia

An adult with recurrent cholangitis has extrahepatic and intrahepatic dilated ducts.

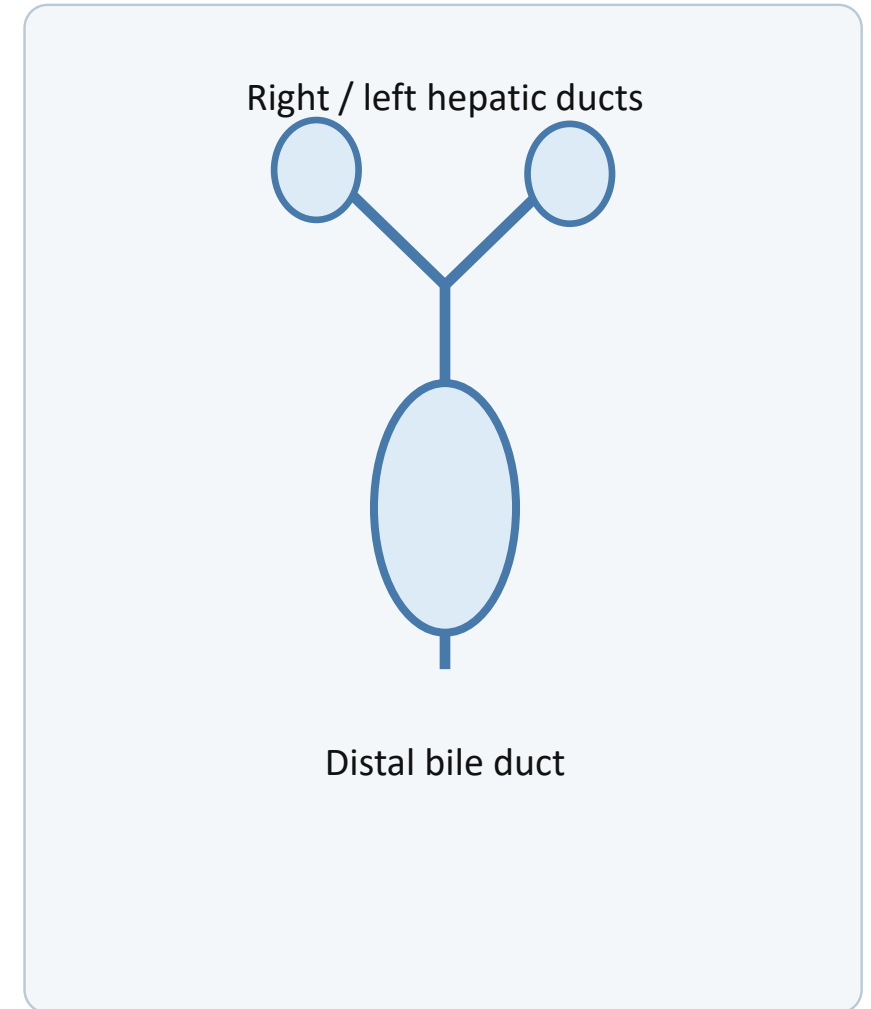
- MRCP: Enlarged extrahepatic duct plus multifocal intrahepatic duct ectasia.
- Several intrahepatic segments contain stones.
- Question: What anatomic category fits best?
- Which residual disease could persist after extrahepatic surgery?

Pattern recognition

Account for both
extrahepatic and
intrahepatic disease.

Case 2: Interpretation and prognostic issue

- The combined distribution supports Todani type IV-A disease.
- Intrahepatic stones and strictures may cause recurrent cholangitis.
- Planning must consider which ducts can be safely resected or drained.
- Surveillance remains necessary after extrahepatic reconstruction.

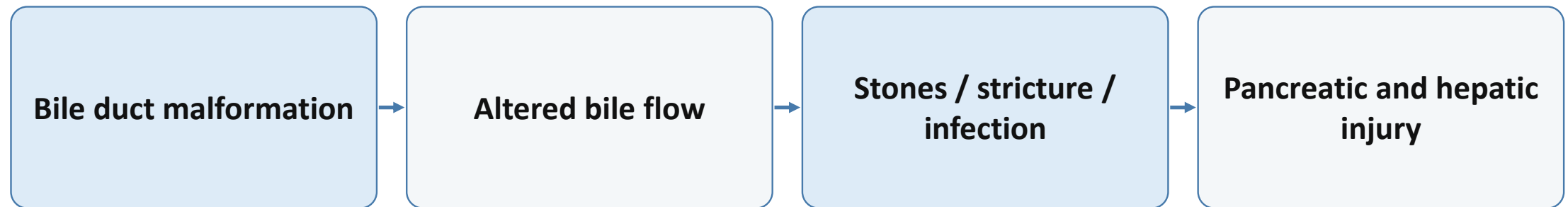


Common errors in clinical-pathology reasoning

Pitfall	Correction
Every cyst near liver = choledochal cyst	Demonstrate communication with biliary ducts
All cysts arise from a single reflux mechanism	Different forms have heterogeneous pathogenesis
Normal adult history excludes congenital disease	Symptoms can first develop in adulthood
Excision removes all future risk	Residual intrahepatic disease and malignancy risk may remain
All types need identical surgery	Type-specific anatomy dictates treatment

Integrated clinicopathologic synthesis

Congenital anatomy is the root condition; Complications arise through time.



Epithelial injury creates an additional, long-term adenocarcinoma concern.

Five facts to retain

- Choledochal cysts are congenital ductal dilatations; Many become symptomatic in childhood.
- The location and number of affected duct segments determine the anatomic type.
- Biliary stasis explains stones, strictures, inflammation and obstruction.
- Risk of adenocarcinoma affects the rationale for complete resection.
- MRCP-based anatomic mapping and long-term clinical follow-up are central.

References and source boundaries

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