

Fibropolycystic disease

Congenital malformations of the biliary tree

Ductal plate malformation · biliary cysts · portal fibrosis

An integrated clinicopathologic lecture

Dr Abu Suraih Sakhri

MBBS, MD (Pathology)

www.abusuraihsakhri.com

Learning outcomes

- Explain persistent fetal ductal plates as the developmental substrate.
- Distinguish duct hamartomas, biliary cystic dilatation, and congenital hepatic fibrosis.
- Differentiate Caroli disease from Caroli syndrome.
- Link structural abnormalities to cholangitis, portal hypertension, and clinical risk.

Definition: A biliary developmental spectrum

- A heterogeneous family of lesions—not one uniform cystic disease.
- The primary abnormality is congenital malformation of the biliary tree.
- Expression ranges from incidental findings to clinically important portal disease.
- Morphology depends on which portal-tract / ductal compartment is involved.

Three recurring morphologic patterns

Small-duct pattern

Von Meyenburg complexes

Localized bile duct hamartomas

Diffuse involvement may signal broader disease

Cystic-duct pattern

Biliary cysts or duct dilatation

Caroli disease: Large intrahepatic ducts

Can coexist with portal fibrosis

Fibrotic pattern

Congenital hepatic fibrosis

Broad collagenous portal bands

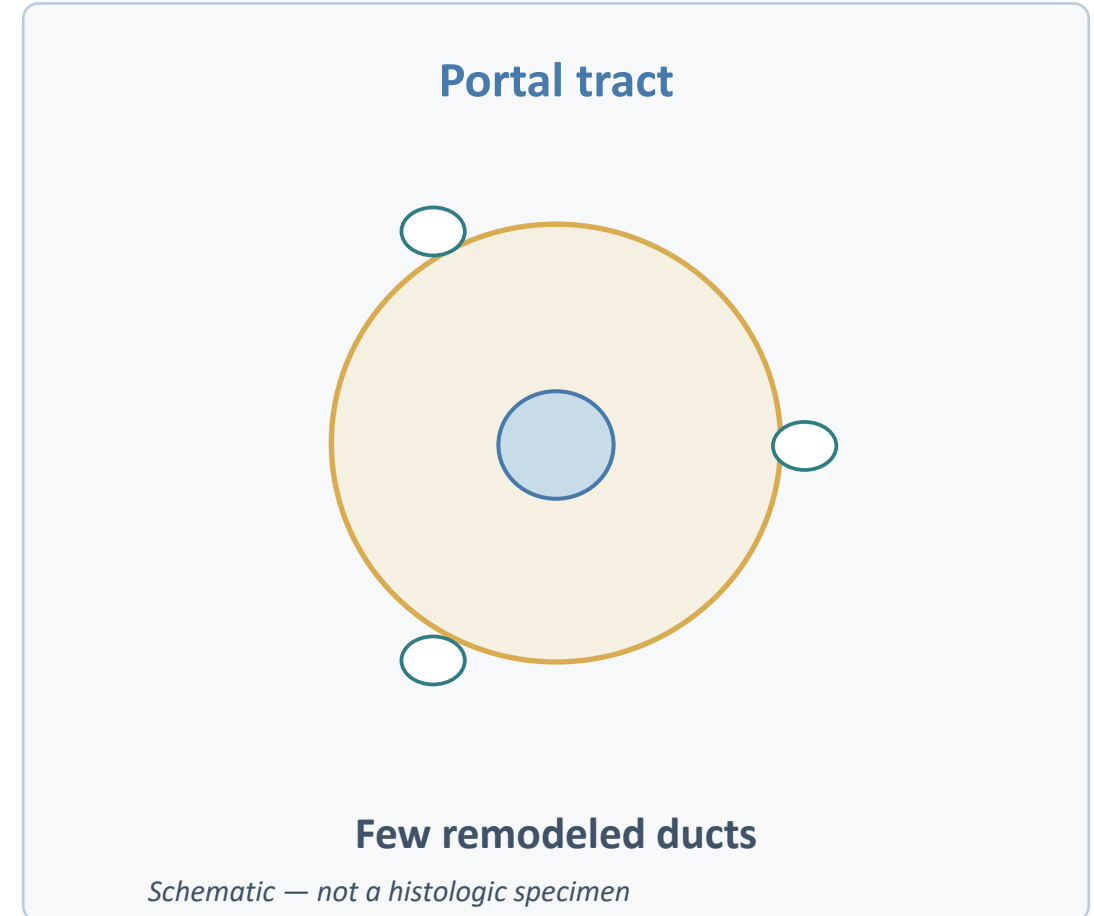
Portal hypertension and varices

The common developmental substrate

- All three patterns belong to the spectrum of ductal plate malformations.
- Persistence of fetal periportal ductal plates produces abnormal ductal structures.
- The resulting lesion may be small-duct, cystic, fibrotic, or combined.
- A shared developmental origin explains the overlap between named conditions.

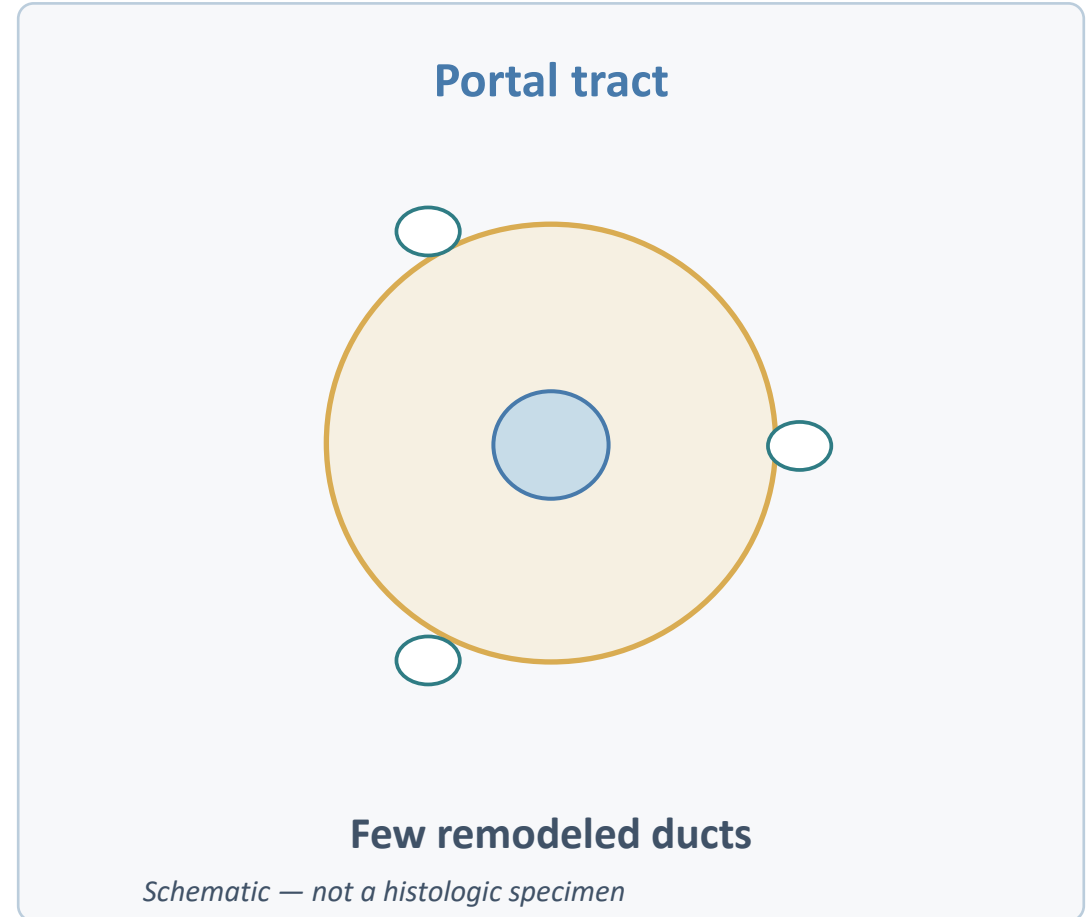
Portal-tract anatomy determines location

- Bile ducts normally accompany the periportal vascular structures.
- The malformation arises in the fetal periportal ductal-plate region.
- Small versus large involved tracts generate different lesion patterns.



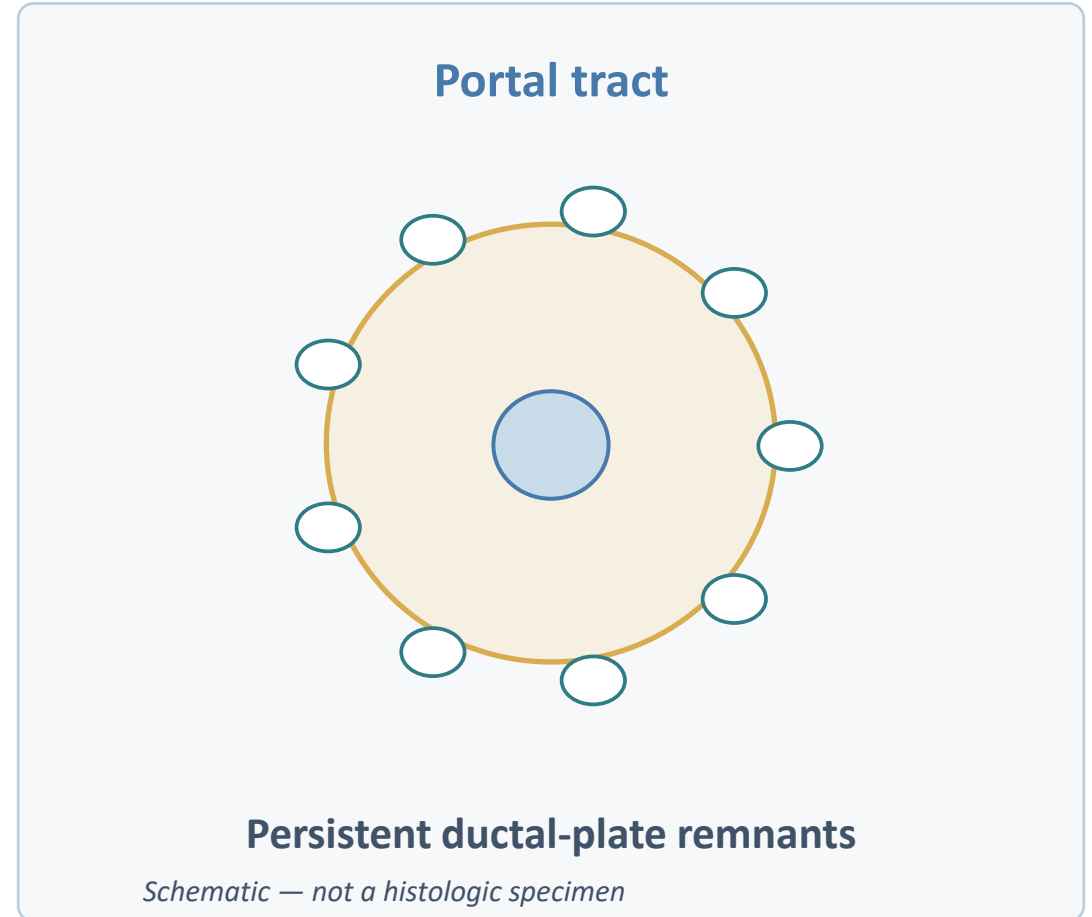
Ductal plate: The fetal precursor

- The fetal ductal plate is a periportal developmental structure.
- Its remodeling ordinarily yields a more restricted duct arrangement.
- Persistent plate remnants underlie the malformation spectrum.



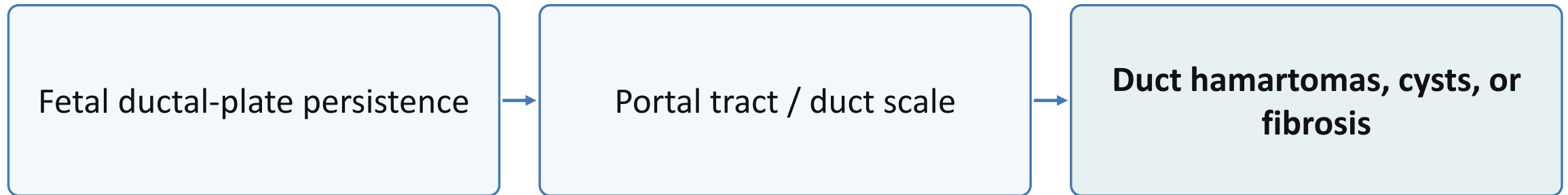
Persistence alters biliary architecture

- Remnants of the fetal ductal plate remain around portal tracts.
- Anomalous duct profiles persist rather than resolving normally.
- The final appearance varies with ductal scale and associated fibrosis.



From portal-tract caliber to phenotype

The caliber of affected portal tracts influences lesion size, shape, and distribution.



Interpretation

A continuum of patterns, rather than unrelated lesions.

Developmental malformation versus acquired obstruction

Fibropolycystic spectrum

Congenital biliary-tree maldevelopment

Abnormal duct profiles from ductal plates

May coexist with broad portal fibrosis

Acquired obstructive disease

Obstruction is not the initiating definition here

Clinical consequences can overlap

Do not infer developmental origin from cholestasis alone

Morphologic components can overlap

Hamartomatous

Von Meyenburg complexes

Small bile duct lesions

May be widespread

Cystic

Dilated ducts / true cysts

Intra- or extrahepatic

Large intrahepatic ducts in Caroli

Fibrotic

Broad portal septa

Abnormal duct profiles

Portal hypertension

Why some cases are incidental

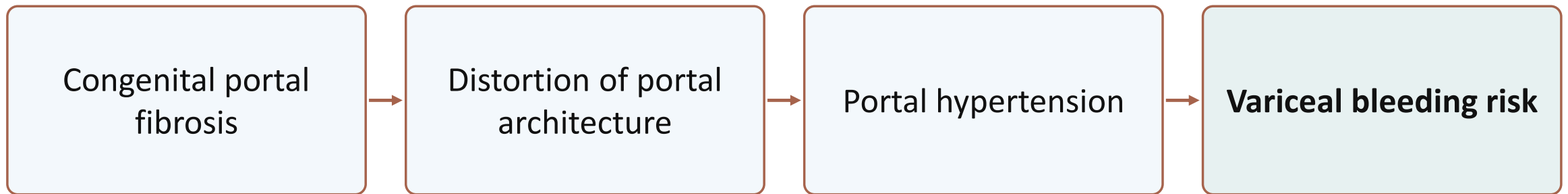
- Lesions can be detected during radiographic assessment for other reasons.
- Surgery and autopsy may reveal otherwise unsuspected abnormalities.
- Occasional von Meyenburg complexes also occur in normal individuals.
- The distribution and associated changes determine the significance.

The severe end of the clinical spectrum

- The most severe forms may become evident in late childhood or adolescence.
- Hepatosplenomegaly and portal hypertension can dominate presentation.
- Clinically important portal disease may occur without overt hepatic dysfunction.
- Structural and vascular consequences need separate assessment.

Portal hypertension may precede dysfunction

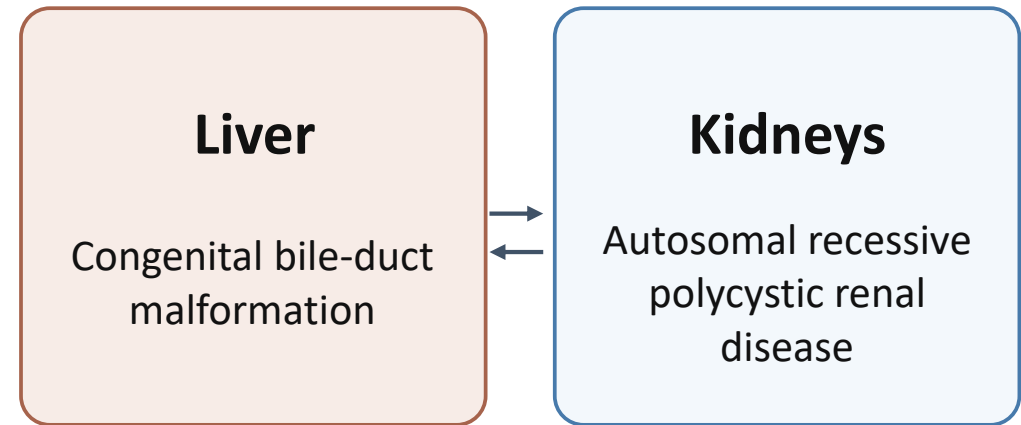
Do not equate preserved hepatocellular function with absence of serious portal disease.



This is especially relevant in congenital hepatic fibrosis.

Associated renal malformation

- Fibropolycystic liver disease often coexists with autosomal recessive polycystic renal disease.
- The association suggests a broader developmental disease spectrum.
- The textbook does not state that every affected patient has renal disease.



Reported association — not an obligatory pairing

A clinically important malignancy association

- An increased risk of cholangiocarcinoma.
- The statement applies to the fibropolycystic disease spectrum.
- No numerical risk estimate is supplied in the assigned excerpt.
- Risk association does not establish that every individual lesion is premalignant.

The disease spectrum in one view

Small-duct pattern

Von Meyenburg complexes

Localized bile duct hamartomas

Diffuse involvement may signal broader disease

Cystic-duct pattern

Biliary cysts or duct dilatation

Caroli disease: Large intrahepatic ducts

Can coexist with portal fibrosis

Fibrotic pattern

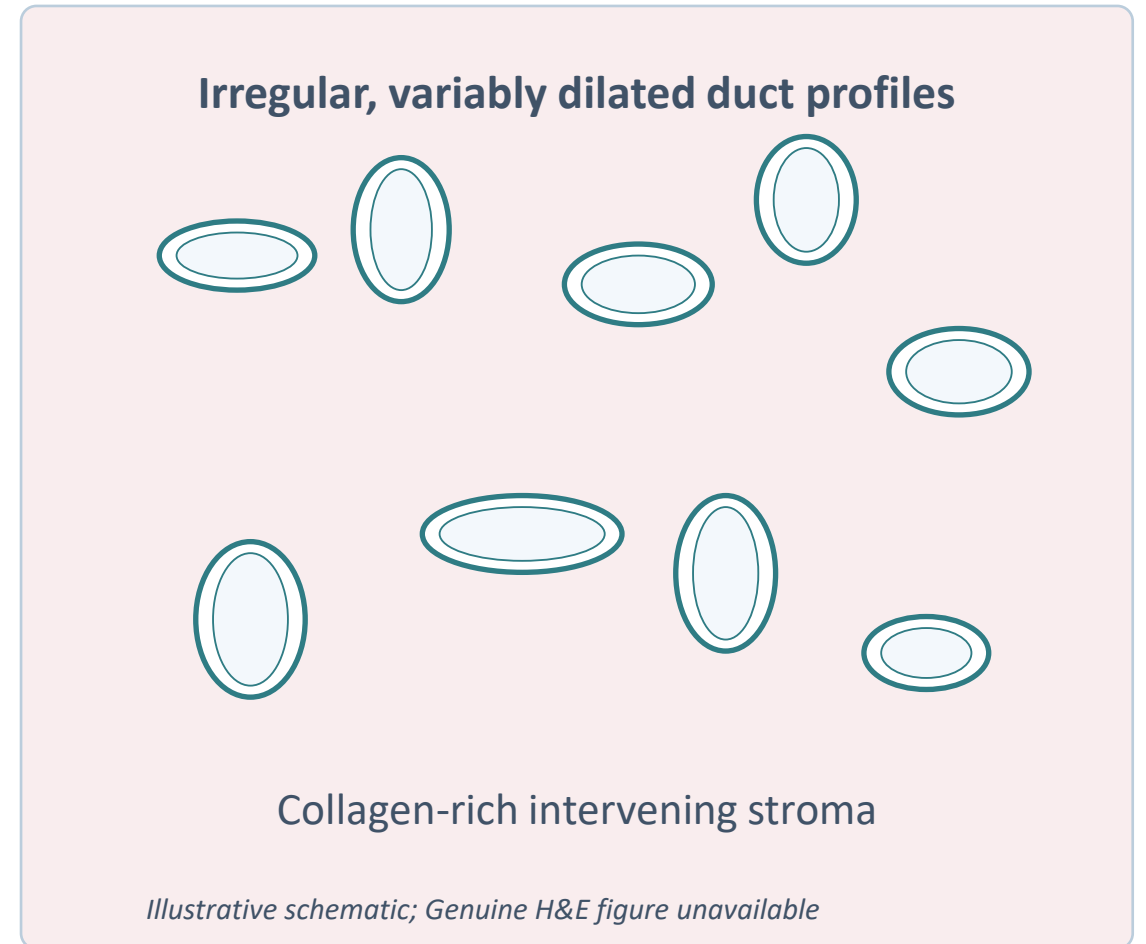
Congenital hepatic fibrosis

Broad collagenous portal bands

Portal hypertension and varices

Von Meyenburg complex: The small-duct lesion

- A von Meyenburg complex is a small bile duct hamartoma.
- It represents a ductal plate–related malformation.
- The finding becomes more informative when widespread rather than isolated.



Occasional versus diffuse bile duct hamartomas

Occasional complexes

Can occur in otherwise normal liver

May be incidental

Alone, not sufficient to infer severe disease

Diffuse complexes

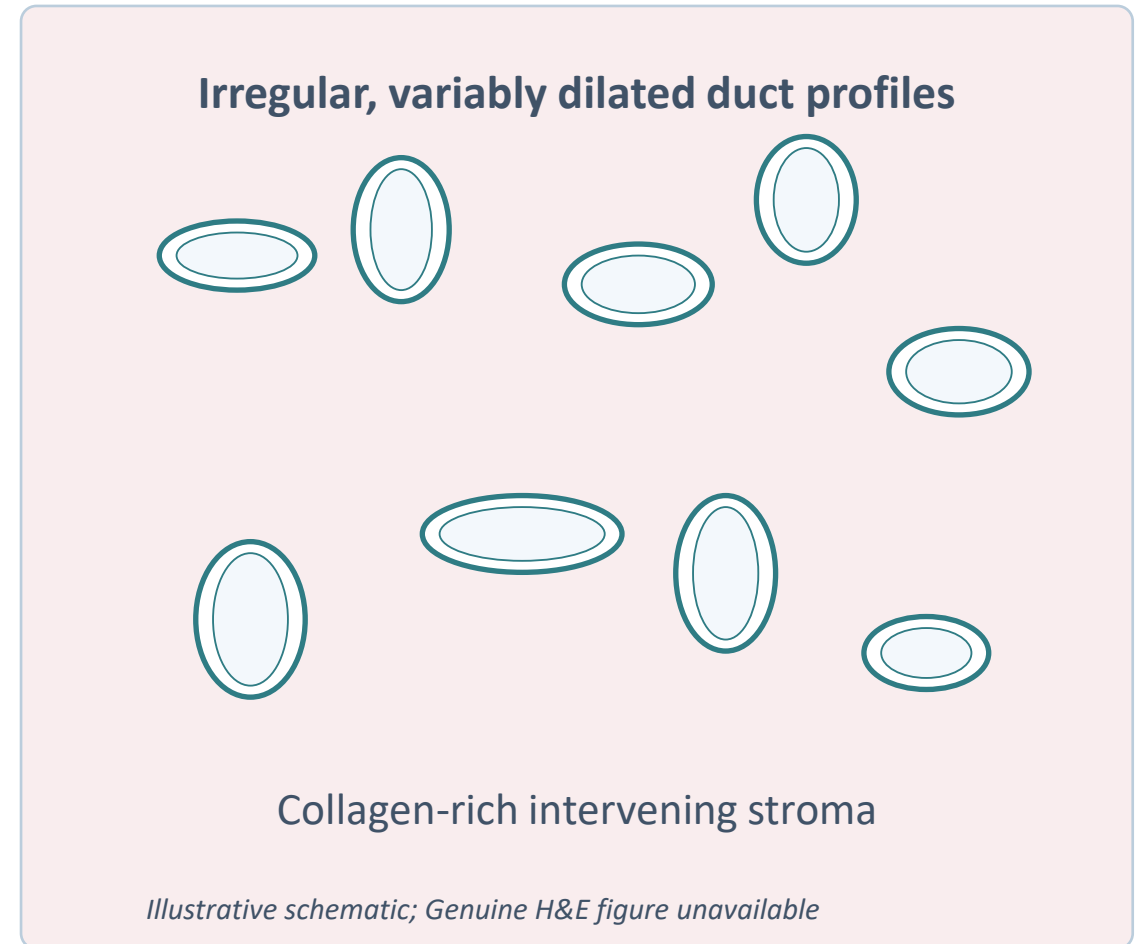
Signal underlying fibropolycystic disease

Prompt recognition of a wider pattern

Look for cystic and fibrotic components

Von Meyenburg morphology: An interpretive schematic

- Small, irregular duct profiles form the hamartomatous lesion.
- Dilated, curvilinear bile duct outlines.
- The schematic is explanatory; It is not a substituted H&E slide.



Why the duct contours matter

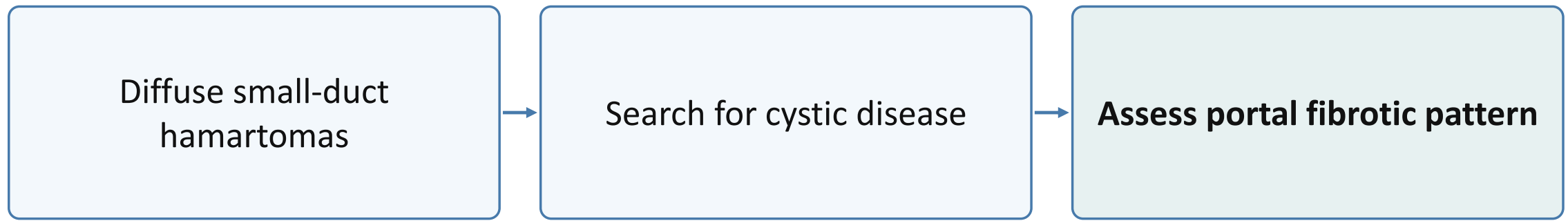
- Irregular curvilinear duct profiles are a structural clue to the lesion.
- The pattern fits persistence of abnormal fetal ductal structures.
- Distribution—occasional versus widespread—changes interpretation.
- Avoid diagnosing broader disease from one isolated duct profile.

Interpret a bile duct hamartoma in context

- Establish whether the abnormality is isolated or multifocal.
- Identify any associated intrahepatic biliary cysts or dilated ducts.
- Assess whether congenital hepatic fibrosis is also present.
- Integrate microscopy with the pattern of presentation.

Diffuse complexes: A marker, not the whole diagnosis

Widespread hamartomas may point to clinically important fibropolycystic disease.



This is a clinicopathologic inference, not a validated diagnostic algorithm.

Morphology exercise: Small duct lesions

Clinical / pathology vignette

Several minute, irregular bile duct hamartomas are described in an otherwise unremarkable liver. No additional cystic or fibrotic pattern is identified.

Reasoning prompts

Name the lesion.

What does isolated occurrence imply?

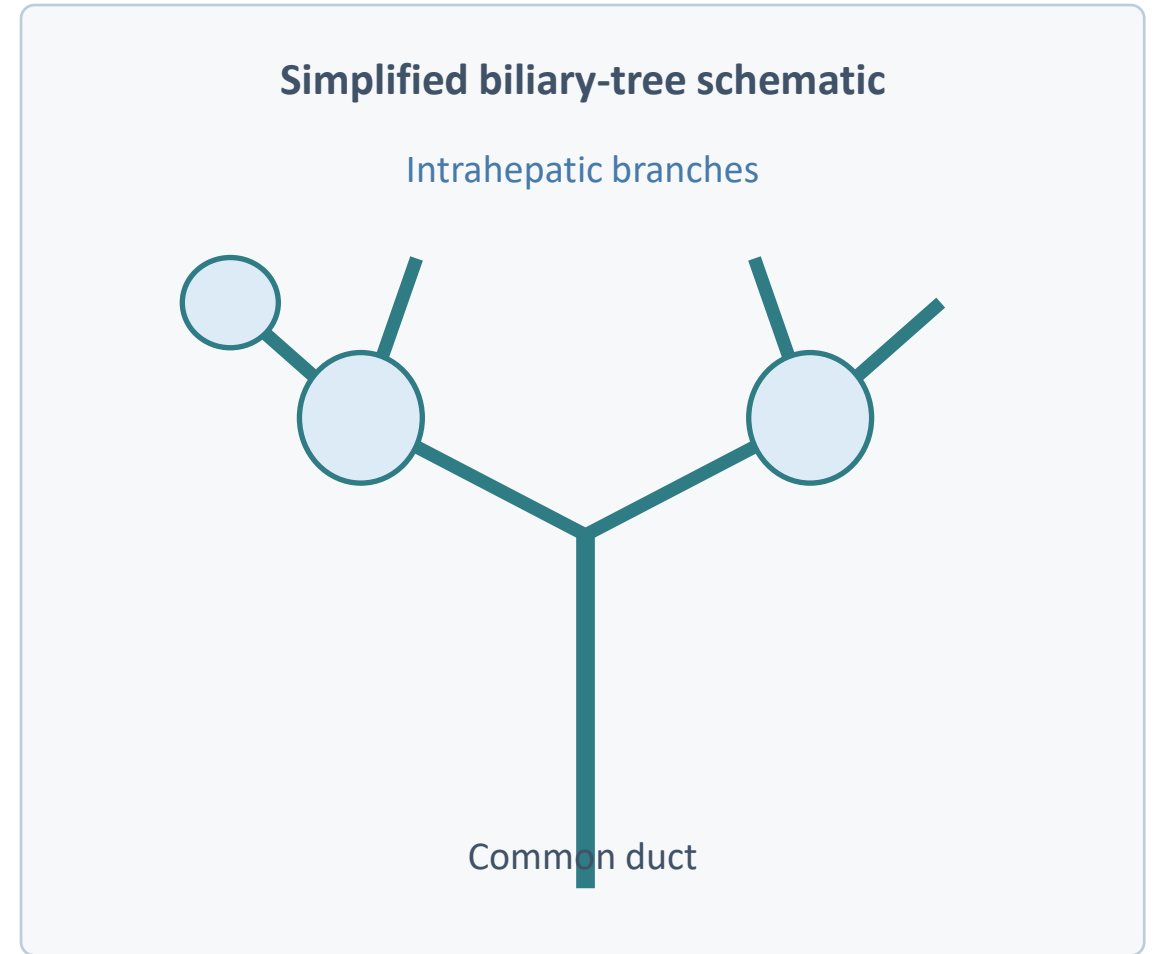
What would diffuse lesions change?

Biliary cysts: The second major component

- Single or multiple biliary cysts may be present.
- Lesions may be intrahepatic or extrahepatic.
- Bile ducts can be cystically dilated; True cysts may also occur.
- Cystic disease and portal fibrosis are not mutually exclusive.

Anatomic location is diagnostically useful

- Intrahepatic dilation involves ducts within the liver.
- Extrahepatic cysts can involve the larger outflow ducts.
- Caroli disease specifically refers to multifocal dilation of large intrahepatic ducts.



Single cyst versus multiple abnormalities

- A solitary lesion and multifocal disease can both lie within the spectrum.
- The location alone is not enough to define a complete syndrome.
- Assess the affected duct caliber and presence of portal fibrosis.
- The larger structural pattern determines the diagnostic term.

Dilated ducts and true cysts are not synonyms

Cystically dilated duct

Part of the existing biliary tree

Duct lumen is enlarged

Underlying duct continuity is conceptualized

True cyst

A distinct cystic lesion

May coexist with dilated ducts

Can be intrahepatic

Teaching distinction

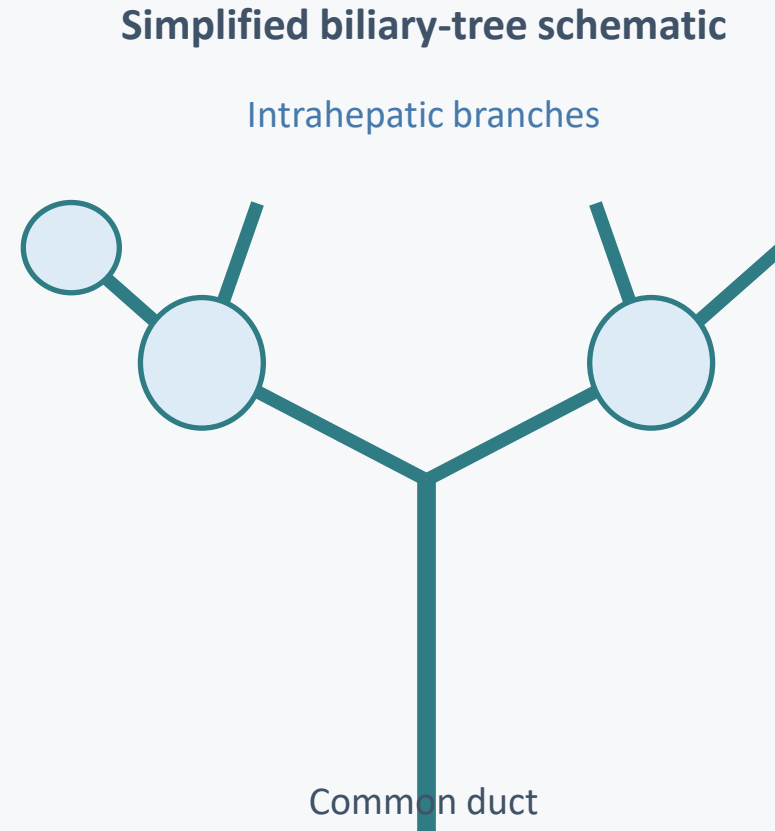
Both forms occur

Report the actual structure described

Do not collapse all cysts into Caroli disease

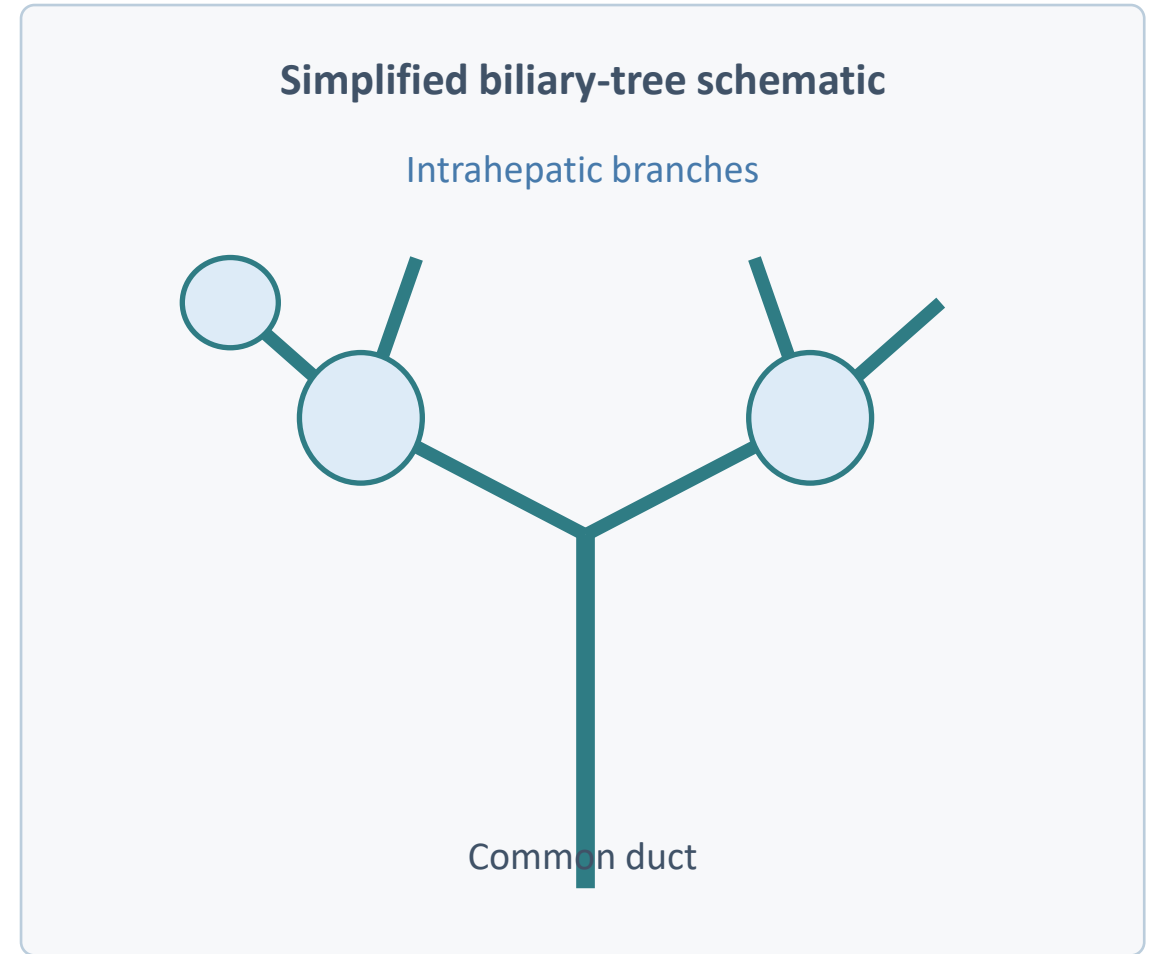
Caroli disease: Definition

- Multifocal cystic dilation affects large intrahepatic bile ducts.
- The named pattern is based on the anatomy of duct dilation.
- Additional congenital hepatic fibrosis changes the designation to Caroli syndrome.

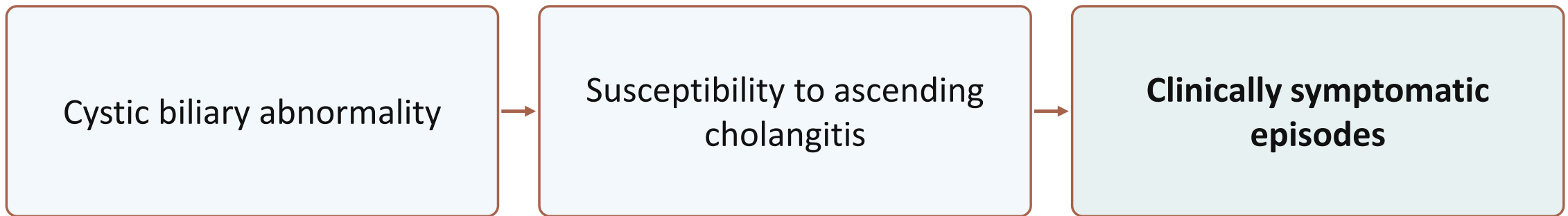


Caroli disease: Which ducts are enlarged?

- The abnormal dilatation is intrahepatic, not merely common-duct dilation.
- Multiple large-duct segments are involved.
- Its cystic pattern remains part of the ductal plate malformation spectrum.



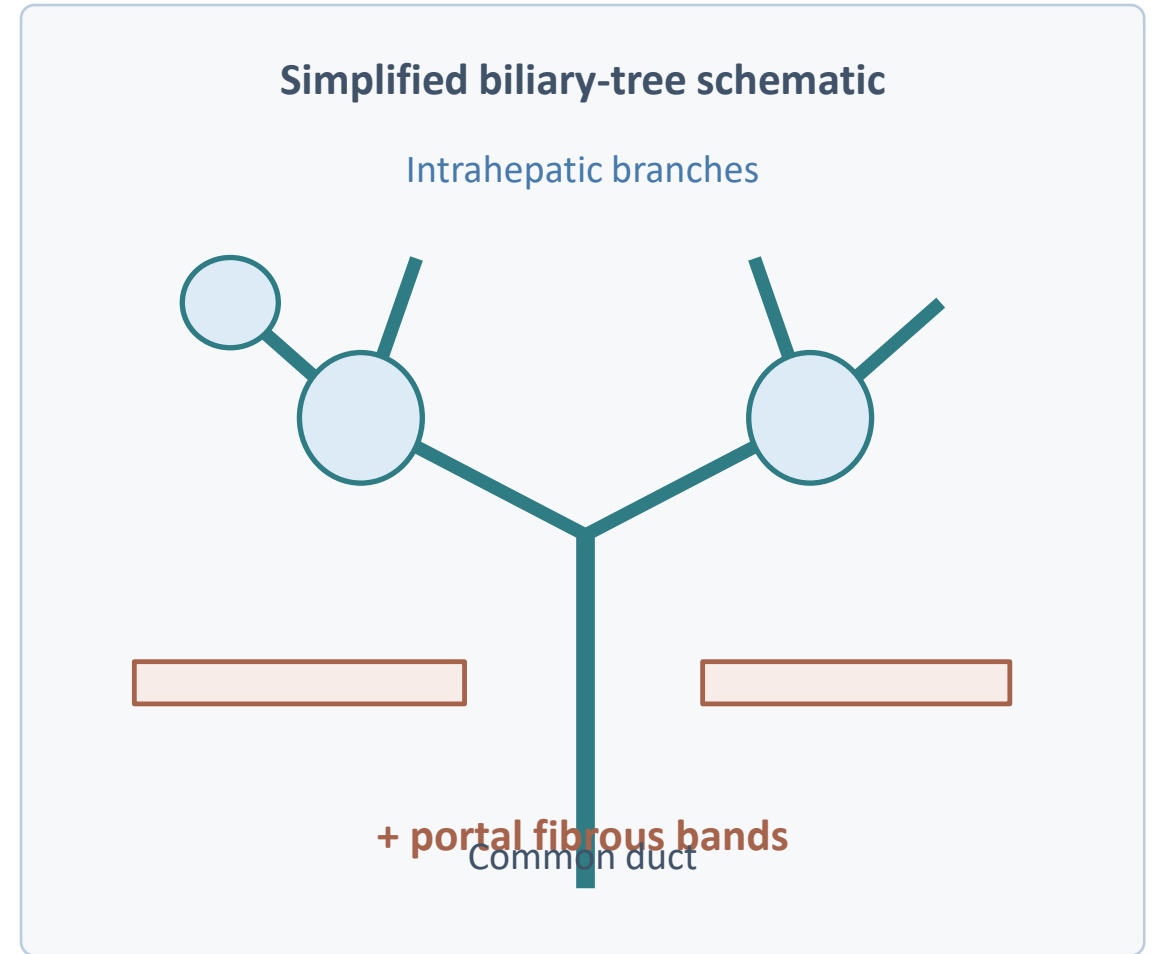
Cystic disease can present with cholangitis



- Robbins particularly notes symptoms from ascending cholangitis in isolated cystic lesions.
- The excerpt does not provide a frequency or clinical prediction rule.

Caroli syndrome: A combined phenotype

- Caroli-type biliary duct dilatation is present.
- Congenital hepatic fibrosis is also present.
- The term “Caroli syndrome” depends on the combination of both patterns.



Caroli disease versus Caroli syndrome

Feature	Caroli disease	Caroli syndrome
Large intrahepatic duct dilation	Present	Present
Congenital hepatic fibrosis	Not required for the name	Present by definition
Position in spectrum	Cystic phenotype	Combined cystic–fibrotic phenotype
Clinical interpretation	Consider biliary manifestations	Consider biliary and portal manifestations

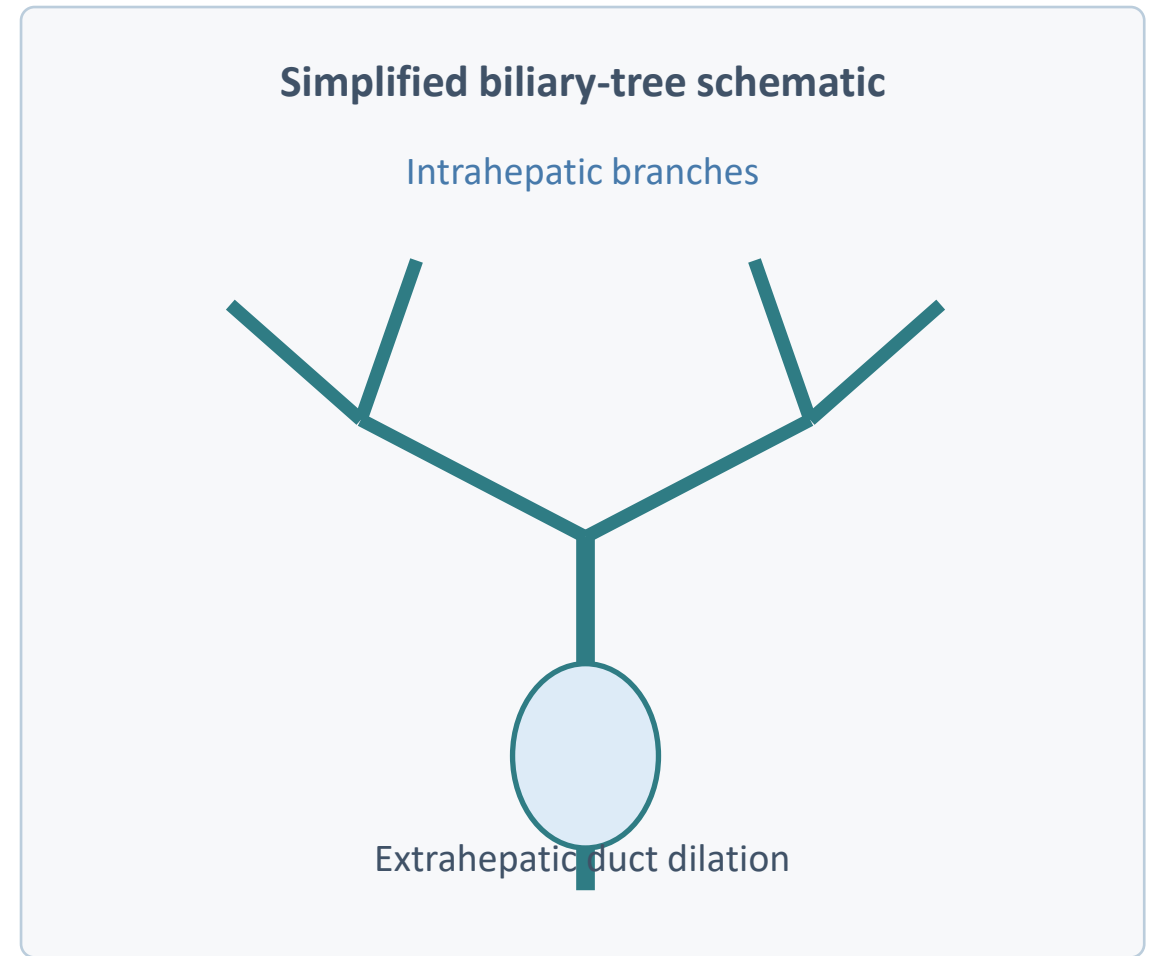
Caroli disease versus choledochal cyst

Dimension	Caroli pattern	Choledochal cyst
Primary named location	Large intrahepatic ducts	Common bile duct
Core anatomic change	Multifocal cystic dilatation	Congenital common-duct dilatation
Can coexist?	Yes—Robbins notes overlap	Yes—may coexist with Caroli
Important complications	Ascending cholangitis may occur	Stones, obstruction and malignancy associations

Choledochal-cyst comparison incorporates the immediately preceding Robbins section (p. 782).

Extrahepatic duct dilation is a different locator

- A choledochal cyst is a congenital dilation of the common bile duct.
- This is distinct from the defining large intrahepatic dilation of Caroli disease.
- Combined intrahepatic and extrahepatic abnormalities can occur.

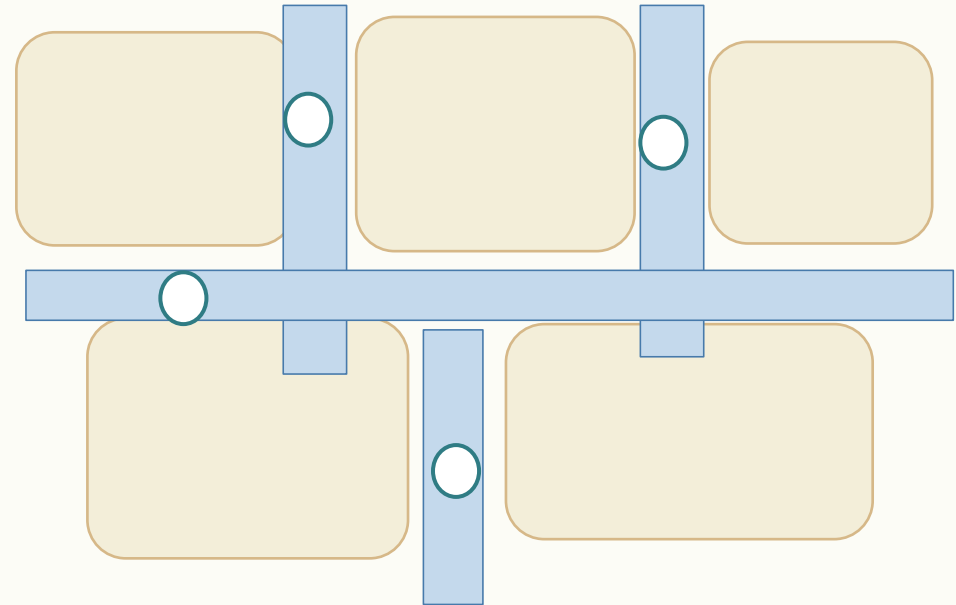


Clinical reading of cystic biliary disease

- Confirm whether the dilated structures are intrahepatic or extrahepatic.
- Determine whether dilation is focal or multifocal.
- Identify coexistence of congenital hepatic fibrosis.
- Relate cholangitic episodes to the structural biliary phenotype.

Congenital hepatic fibrosis: The portal pattern

- Portal tracts are enlarged by broad collagenous tissue.
- Fibrous septa divide liver tissue into irregular islands.
- Abnormally shaped bile ducts are embedded within fibrous areas.



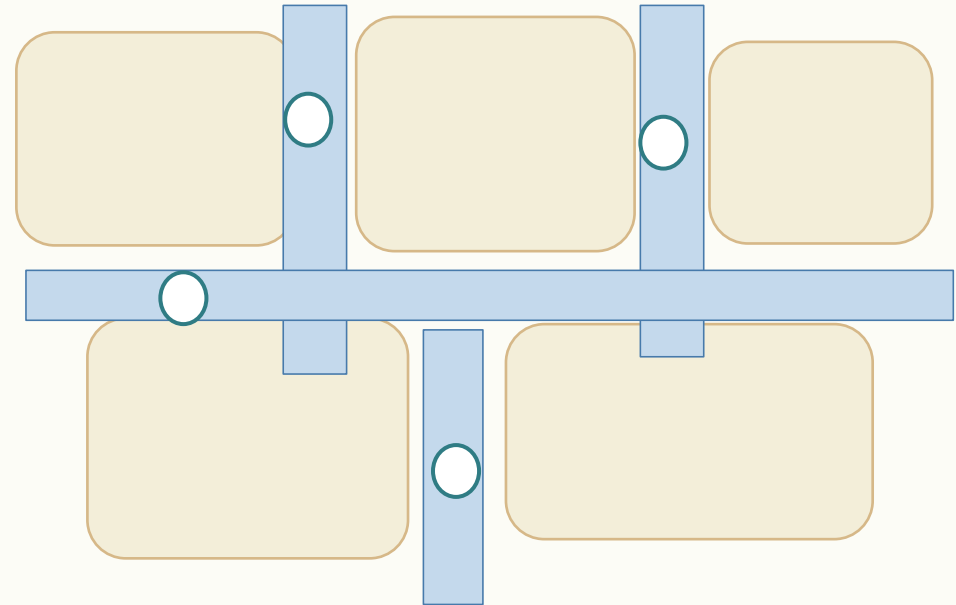
Relatively preserved parenchymal islands

Blue bands = portal collagenous fibrosis

Schematic — not a clinical specimen

Broad portal bands: The central morphologic clue

- The lesion is defined by prominent fibrous expansion of portal regions.
- Collagenous bands form septa between relatively preserved liver islands.
- The fibrosis is spatially related to the abnormal bile ducts.



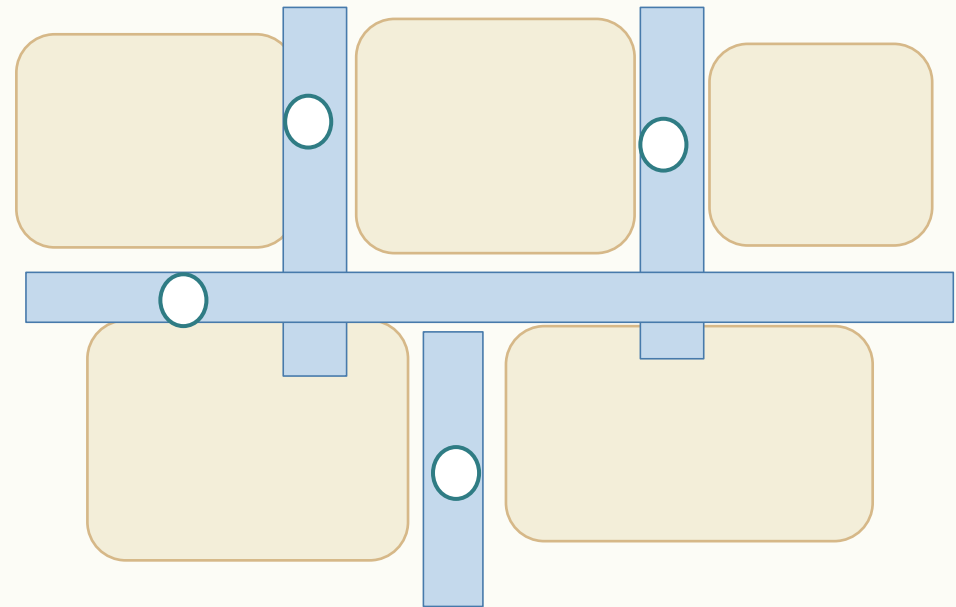
Relatively preserved parenchymal islands

Blue bands = portal collagenous fibrosis

Schematic — not a clinical specimen

Abnormal bile ducts within the fibrous tissue

- Variable numbers of duct profiles occupy the collagenous portal bands.
- Bile ducts have abnormal shapes rather than normal uniform profiles.
- Their presence links the fibrosis to congenital biliary maldevelopment.



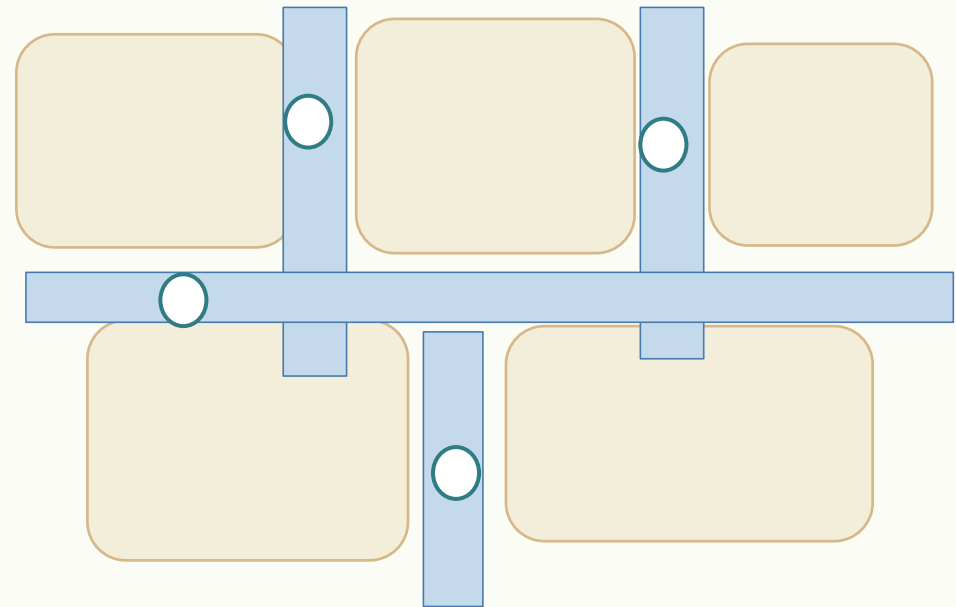
Relatively preserved parenchymal islands

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Schematic — not a clinical specimen

Architectural pattern: Irregular parenchymal islands

- The broad septa subdivide hepatic tissue into irregular islands.
- Relatively normal intervening parenchyma in its figure caption.
- Recognize the portal/biliary malformation, not merely “fibrosis”.



Relatively preserved parenchymal islands

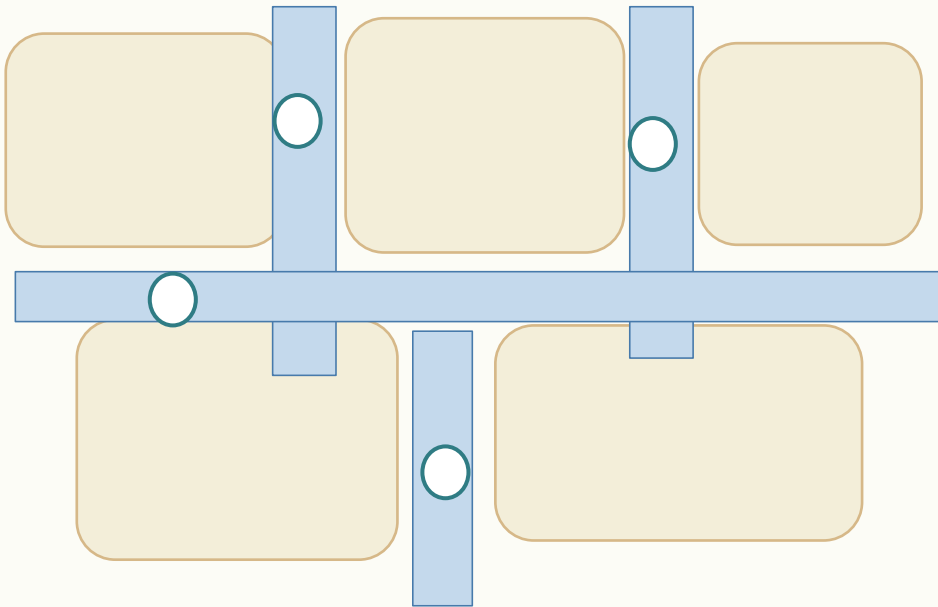
Blue bands = portal collagenous fibrosis

Schematic — not a clinical specimen

Bile-duct continuity is retained

- Abnormally shaped ducts remain in continuity with the biliary tree.
- This feature distinguishes a remodeled ductal system from unrelated cystic spaces.
- Duct continuity is part of the structural interpretation of the disease.
- The assigned excerpt does not specify a required imaging test to prove it.

Congenital hepatic fibrosis: Integrated schematic



Relatively preserved parenchymal islands

Blue bands = portal collagenous fibrosis

Schematic — not a clinical specimen

Read the pattern

Broad portal collagen bands

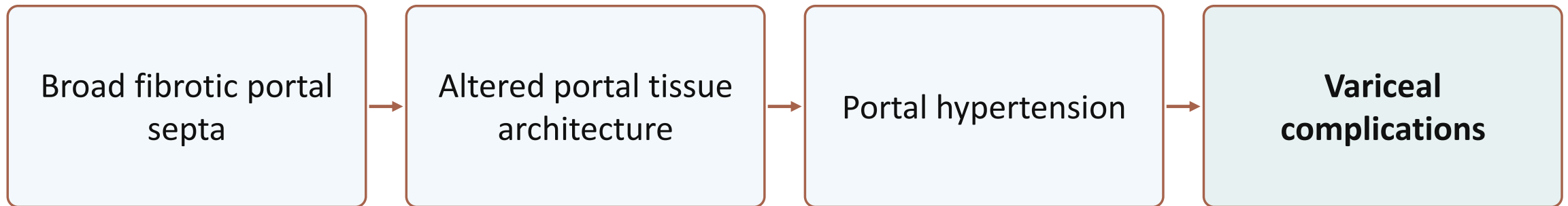
Irregular parenchymal islands

Abnormal ducts in scar

Ducts remain continuous

From portal fibrosis to portal hypertension

Congenital hepatic fibrosis carries the risk of portal hypertension.



The sequence is a teaching interpretation of the morphology–clinical association.

Bleeding varices are an important complication

- Robbins singles out variceal hemorrhage as a complication of portal hypertension.
- The risk follows the portal fibrotic phenotype.
- Recognition of portal hypertension may be clinically urgent.
- The excerpt does not define surveillance intervals or treatment protocols.

Hepatic function and portal effects can diverge

- Some severe presentations show portal hypertension despite limited hepatic dysfunction.
- The distinction is explained by prominent structural portal abnormalities.
- Clinical assessment should not rely solely on hepatocyte function.
- Hepatosplenomegaly may provide an additional clue.

Congenital hepatic fibrosis is not synonymous with cirrhosis

Congenital hepatic fibrosis

Developmental ductal-plate spectrum

Broad portal septa and abnormal ducts

Intervening parenchyma relatively normal in depicted example

Cirrhosis (contextual comparison)

Also associated with portal hypertension

Portal hypertension alone does not determine etiology

Look for the defining biliary developmental pattern

Pathologic comparison of the three components

Component	Central morphologic clue	Interpretive implication
von Meyenburg complex	Small bile duct hamartomas	Diffuse pattern supports broad disease
Biliary cystic pattern	Cysts or dilated ducts	Large intrahepatic multifocal dilation = Caroli
Congenital hepatic fibrosis	Broad portal collagen + abnormal ducts	Portal hypertension and varices
Combined pattern	Caroli-type dilation + fibrosis	Caroli syndrome

Case: An isolated tiny bile duct lesion

Interpretation

A small bile duct hamartoma is found incidentally. The remaining liver is reported as structurally unremarkable.

What the finding means

Most direct designation: Von Meyenburg complex

Incidental isolated lesions can occur

Do not automatically label advanced fibropolycystic disease

Case: Numerous small duct hamartomas

Clinical / pathology vignette

A pathology description identifies widely distributed von Meyenburg complexes in a liver specimen.

Reasoning prompts

Why does distribution matter?

Which two other structural patterns should be considered?

What can and cannot be inferred without more data?

Case: Multifocal intrahepatic dilatation

Clinical / pathology vignette

Imaging identifies multiple cystically dilated large intrahepatic bile ducts, with no congenital hepatic fibrosis reported.

Reasoning prompts

Name the ductal phenotype.

What added finding would justify “syndrome”?

Which complication is relevant to the biliary pattern?

Case: Combined cystic and fibrotic change

Interpretation

Multiple enlarged intrahepatic bile ducts coexist with broad fibrous portal septa and abnormal duct profiles.

What the finding means

Classification: Caroli syndrome

It combines two fibropolycystic patterns

Consider biliary and portal clinical consequences

Case: Portal hypertension in adolescence

Clinical / pathology vignette

An adolescent has hepatosplenomegaly and portal hypertension. A biopsy description mentions broad portal collagen bands with numerous abnormal ducts.

Reasoning prompts

Which fibropolycystic component is dominant?

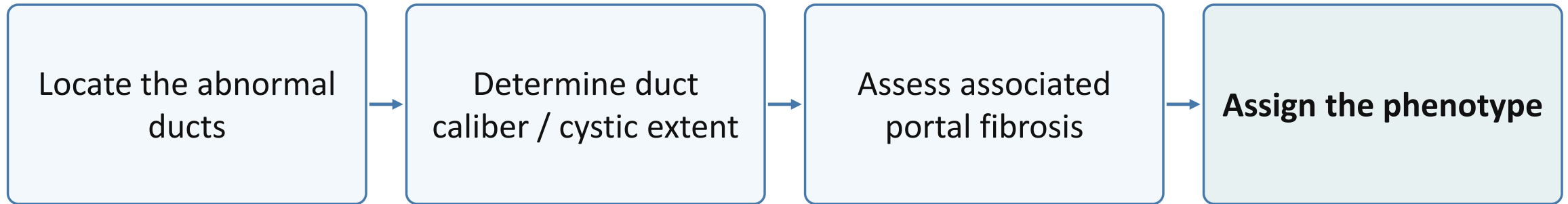
Why might hepatocellular function be relatively preserved?

What vascular complication should be anticipated?

Integrating morphology with clinical consequence

Observed finding	Likely component	Clinical relevance
Isolated small duct hamartoma	von Meyenburg complex	Often incidental
Diffuse hamartomas	Wider fibropolycystic disease	Look for other components
Large intrahepatic duct ectasia	Caroli disease	Ascending cholangitis
Broad portal septal fibrosis	Congenital hepatic fibrosis	Portal hypertension / varices
Dilation plus fibrosis	Caroli syndrome	Mixed biliary + portal phenotype

A practical morphology-first reading sequence



- Consider the distribution of von Meyenburg complexes.
- Review the presence of hepatic or renal developmental associations.
- Relate any portal hypertension or cholangitis to the identified anatomy.

High-yield distinctions

Unifying lesion

Persistence of fetal periportal ductal plates
Different portal-tract calibers yield different patterns
Renal association may be present

Classification

Von Meyenburg: Bile duct hamartoma
Caroli: Large intrahepatic duct ectasia
Caroli syndrome: Ectasia + congenital hepatic fibrosis

Clinical importance

Ascending cholangitis with cystic disease
Portal hypertension and varices with fibrosis
Elevated cholangiocarcinoma risk in the spectrum

References and figure provenance

Primary source

Robbins, Cotran & Kumar. Pathologic Basis of Disease. 11th edition (2026). Chapter 18, Liver and Gallbladder, “Fibropolycystic Disease,” pp. 782–783.

Adjacent parent context

“Structural Anomalies of the Biliary Tree” and “Choledochal Cyst,” p. 782, used for anatomic comparison.

All tissue/duct diagrams are original native PowerPoint schematics; no textbook plate or synthetic micrograph is reproduced.