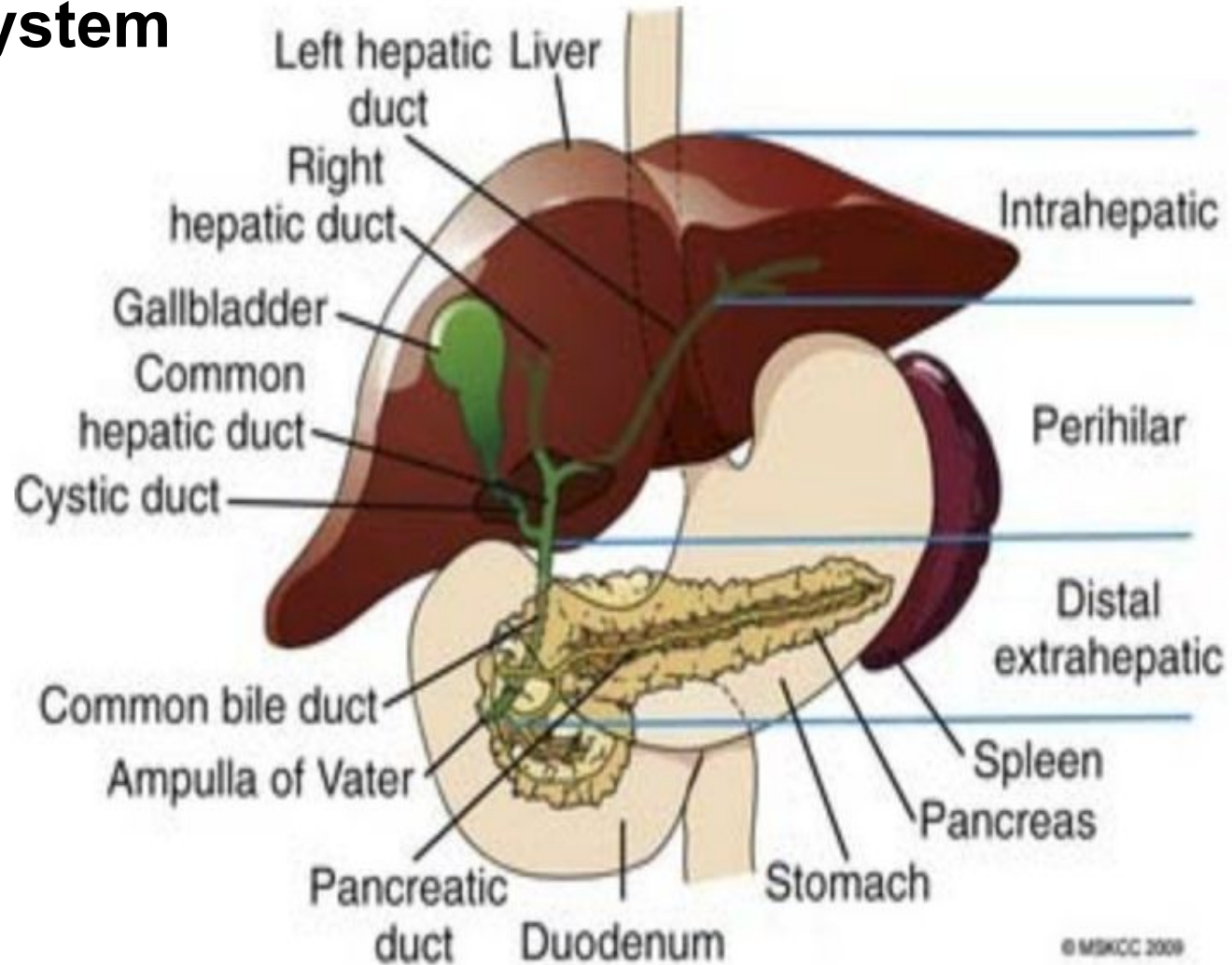
An anatomical illustration of the liver and biliary system. The liver is shown in a reddish-brown color, with a network of blue and red blood vessels branching across its surface. A yellow gallbladder is visible at the bottom left, connected to the biliary system. The background is a soft, out-of-focus purple and blue gradient. A semi-transparent purple rectangular box is overlaid on the right side of the image, containing the title and author information.

Primary Sclerosing Cholangitis in the IBD Population

**By, Sandra Scolari
R.N., B.S.N, C.G.R.N**

Beth Israel Lahey Health Hospital

The Biliary System



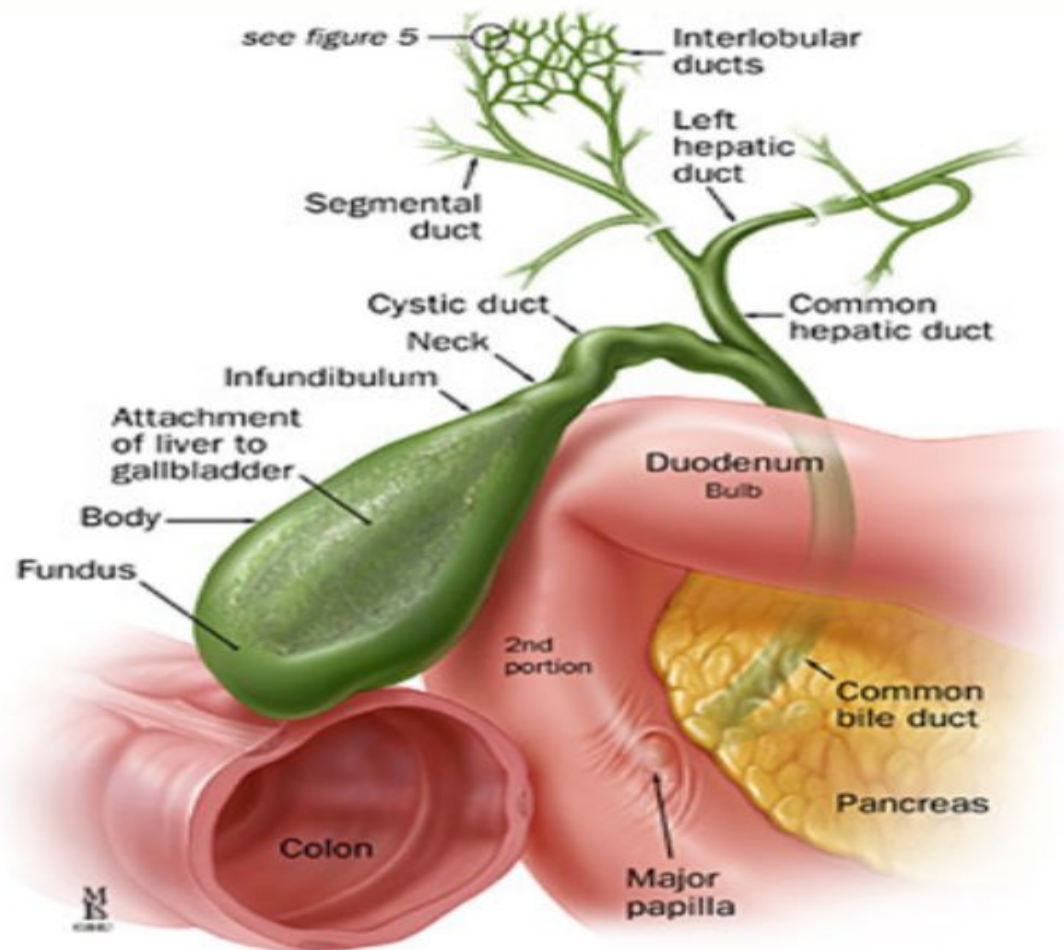
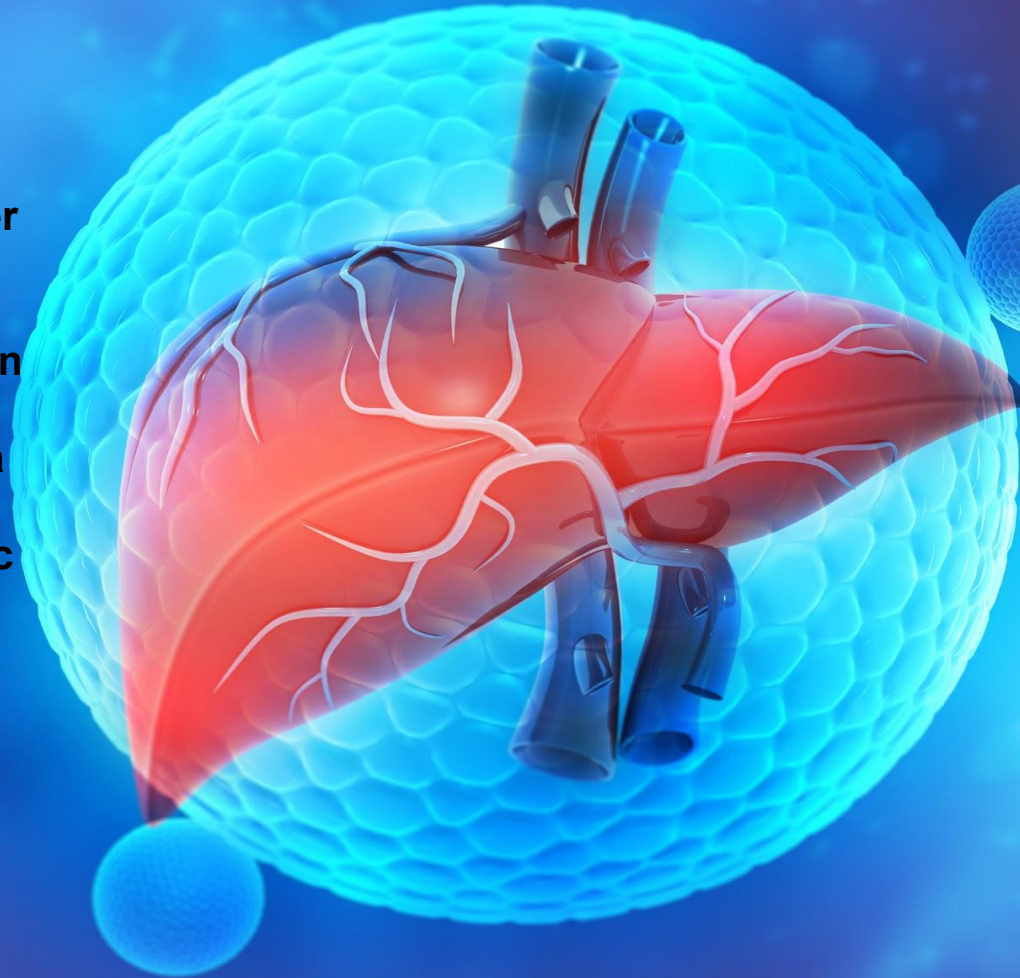


Figure 4. Anatomy of the biliary tree and adjacent structures.

www.johnhopkinsmedicine.org picture credit

What is Primary Sclerosing Cholangitis?

- Chronic liver disease.
- Cholestasis
- Inflammation
- Fibrosis
- Effects Intra and Extrahepatic ducts.



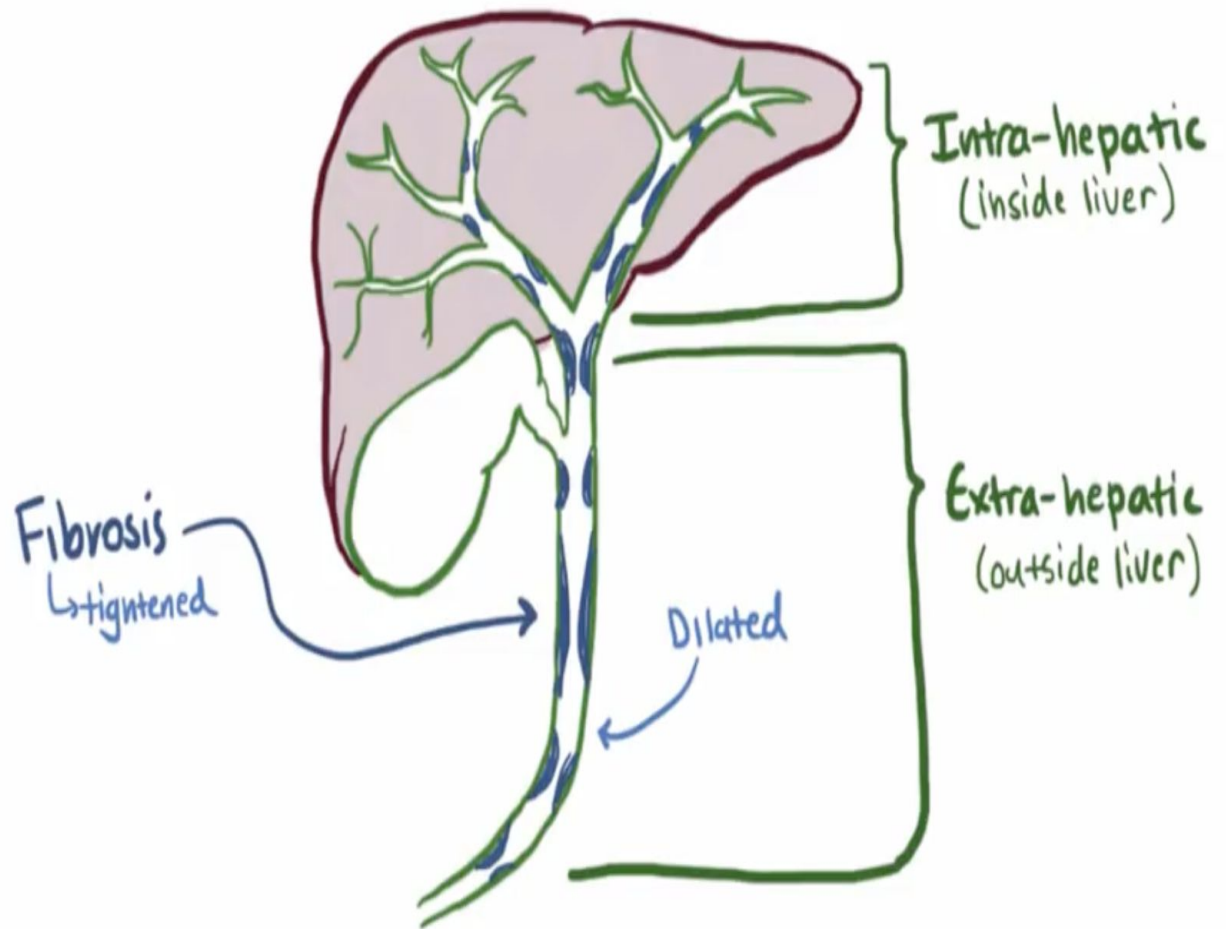
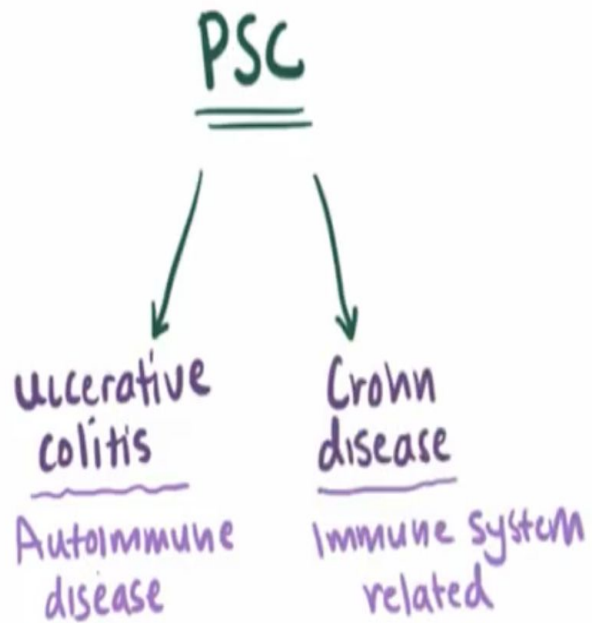
Primary Sclerosing Cholangitis

- Rare disease < 200,000
- 30-40 male(70%)
- UC(87%)
- Crohn's(13%)
- Genetic and Environmental.
- Therapy aimed at symptom management.

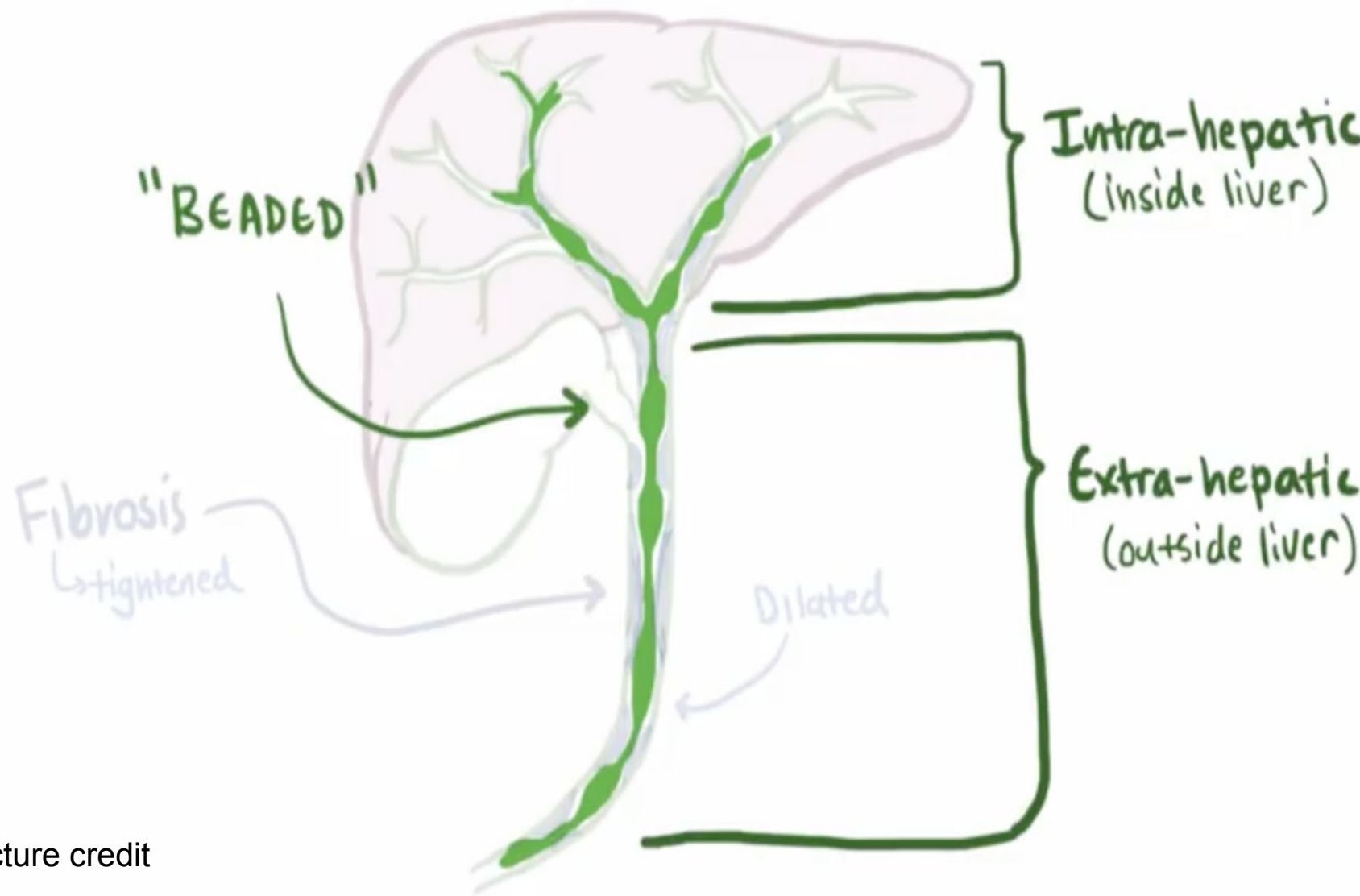
Journal of Hepatology Vol.67 2017 1298-1323

else

Tissue



www.osmosis.org picture credit



PRIMARY

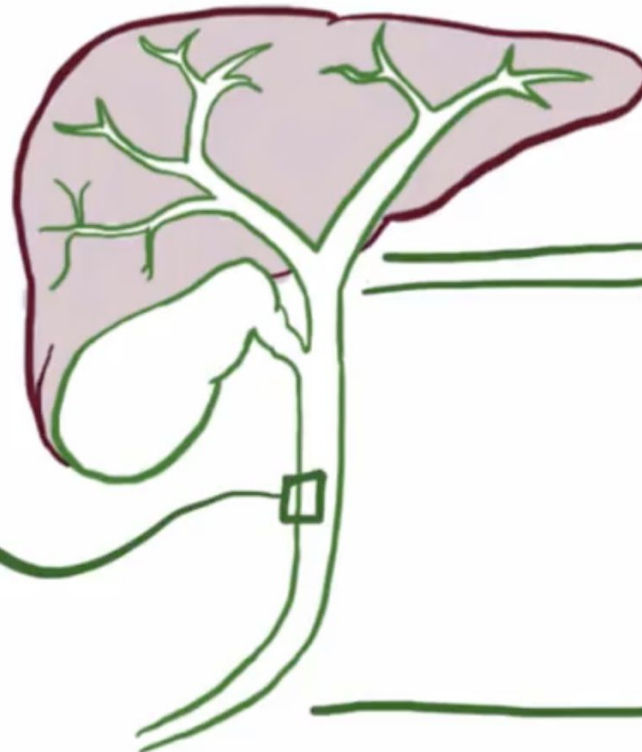
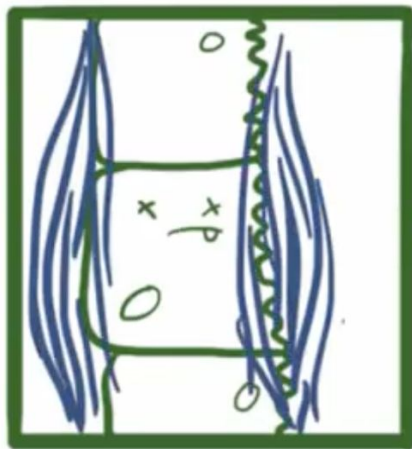
not caused
by something
else

SCLEROSING CHOLANGITIS (PSC)

Hardening
of
tissue

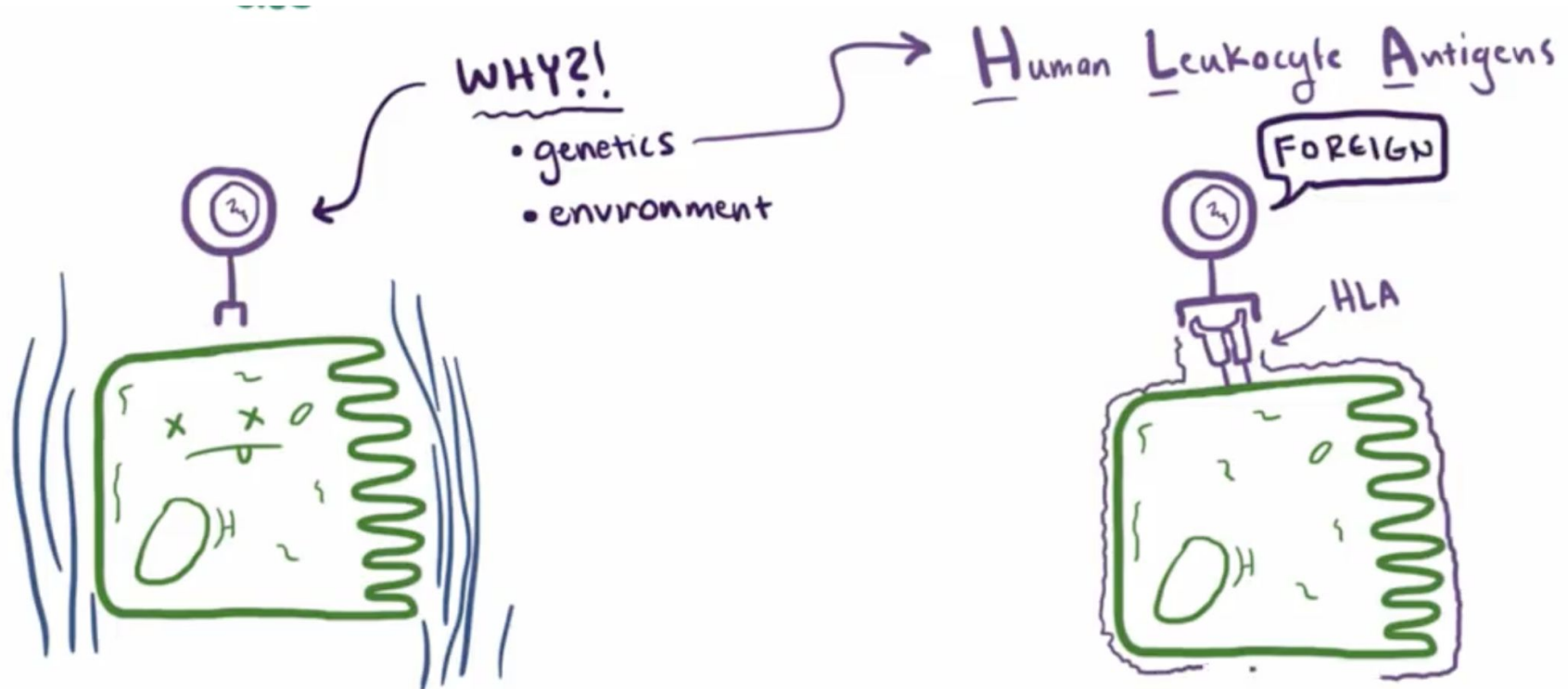
Bile ducts

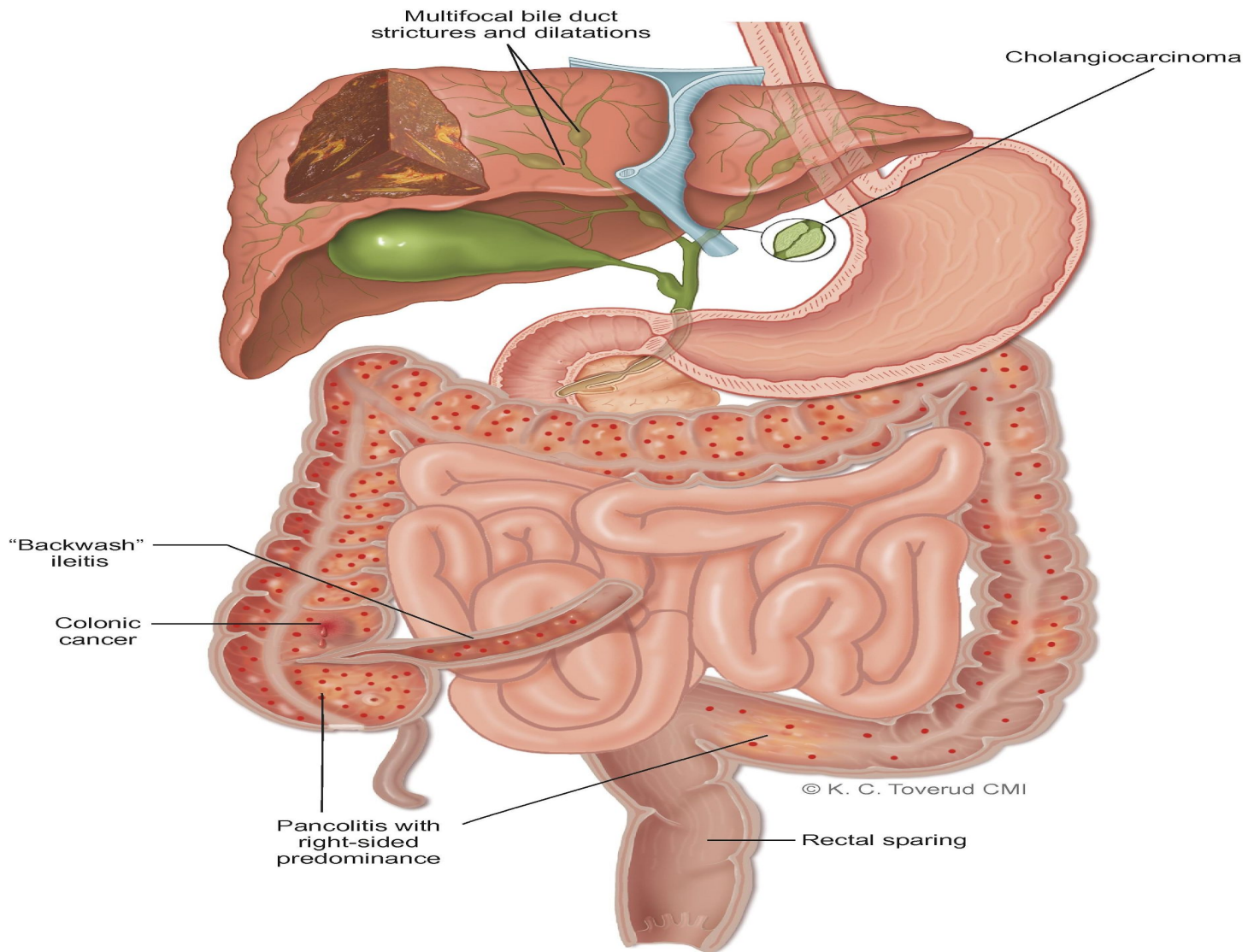
Inflammation



Intra-hepatic
(inside liver)

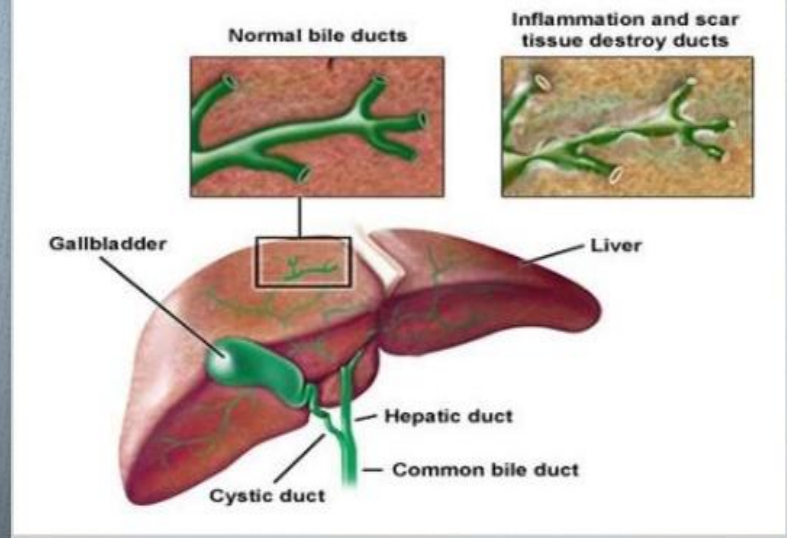
Extra-hepatic
(outside liver)





Pathophysiology AND Histopathology findings

Pathophysiology



Histopathology



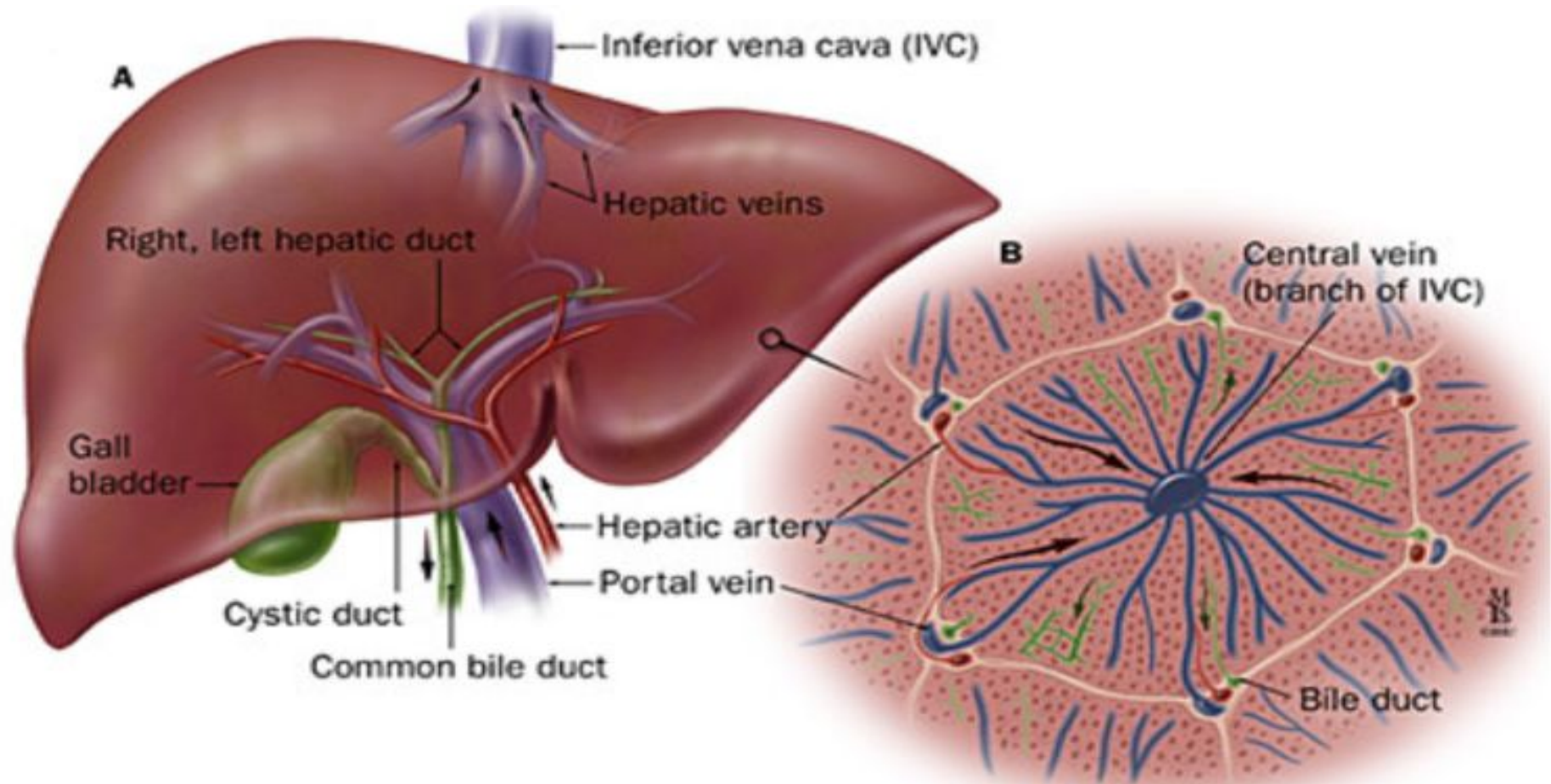


Figure 7. A, Normal gross anatomy of the liver; B, Histological (microscopic) view of a liver lobule.

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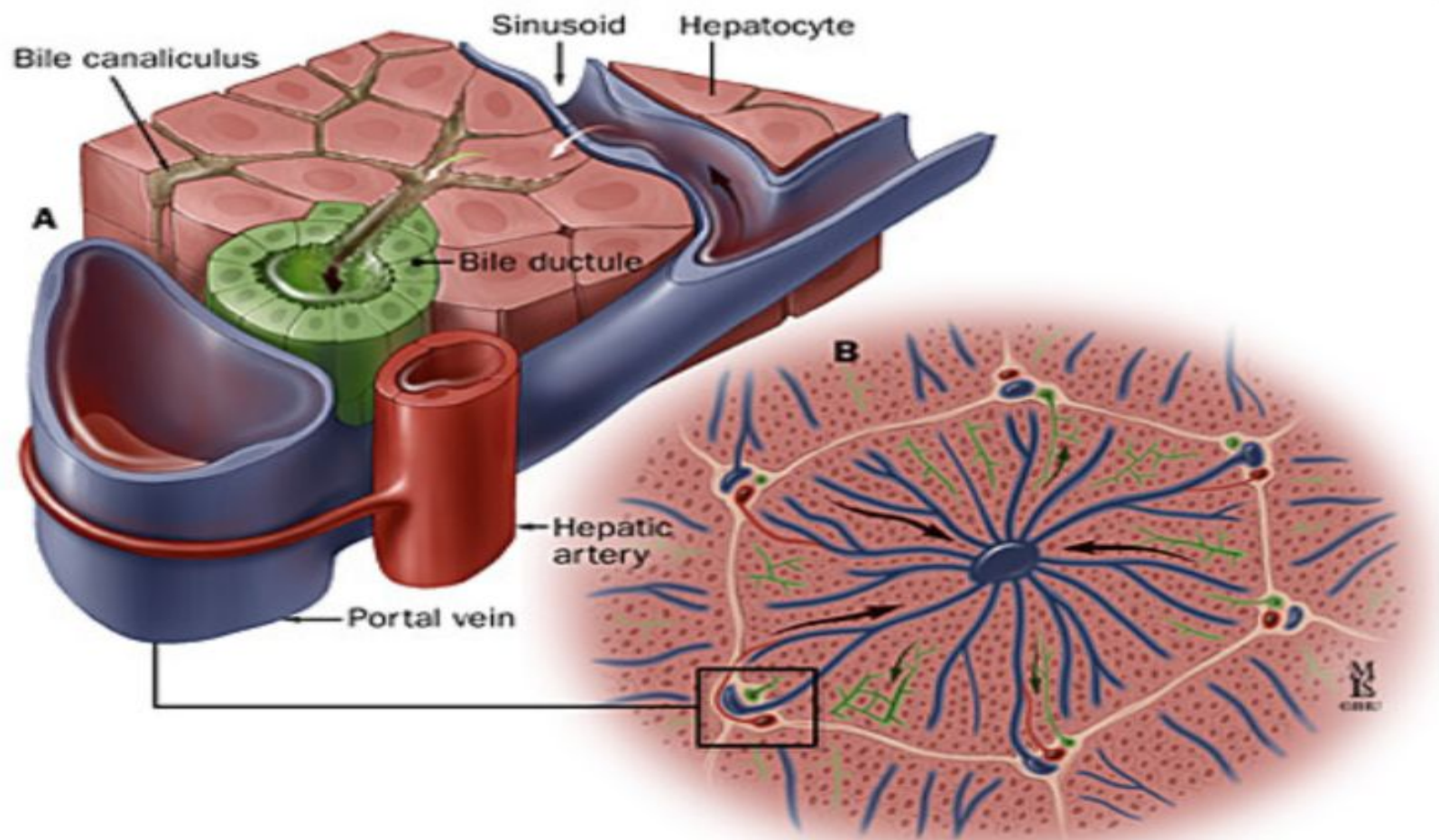
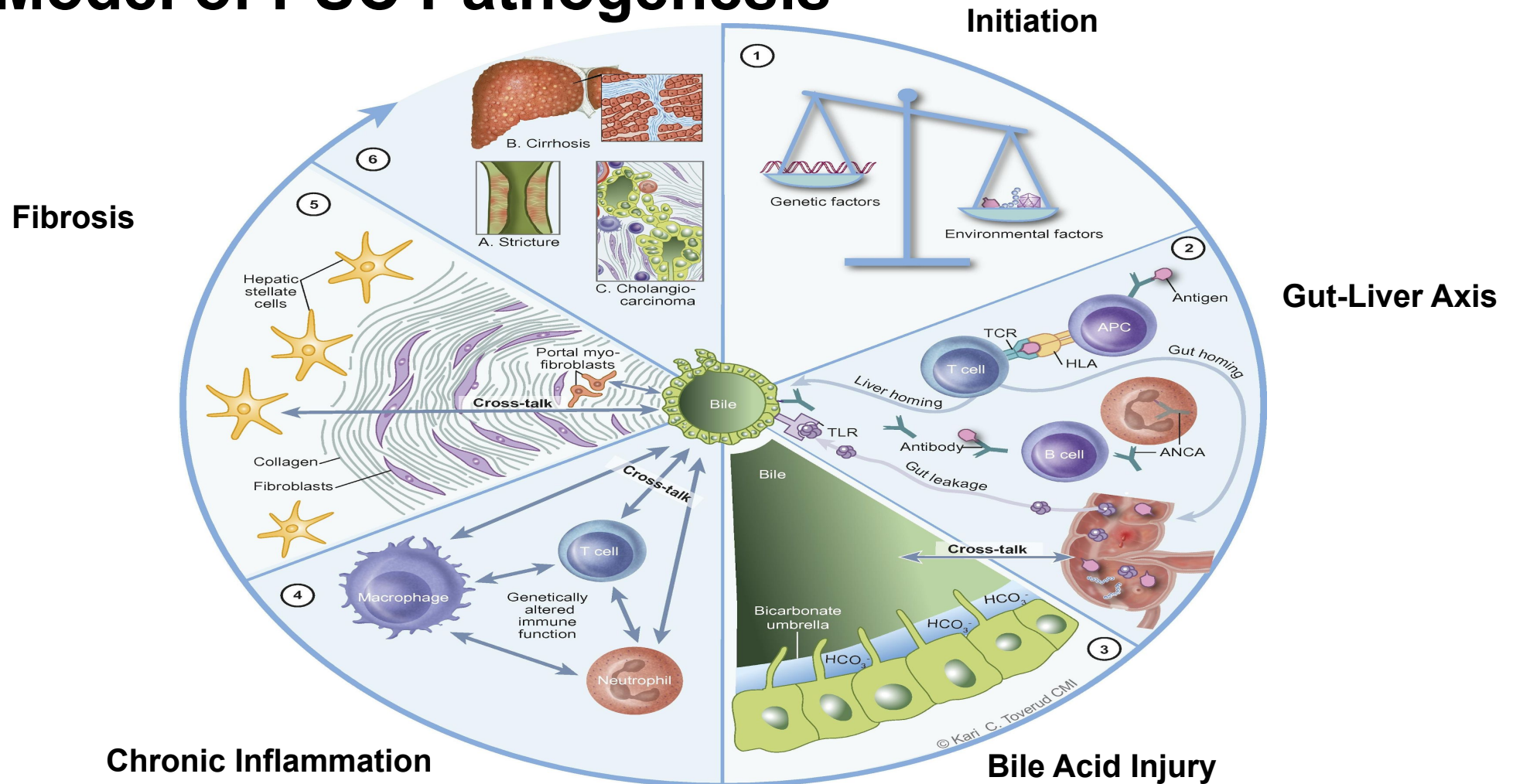


Figure 6. A, Liver lobule; B, histological image with magnified view of the portal tract.

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Model of PSC Pathogenesis



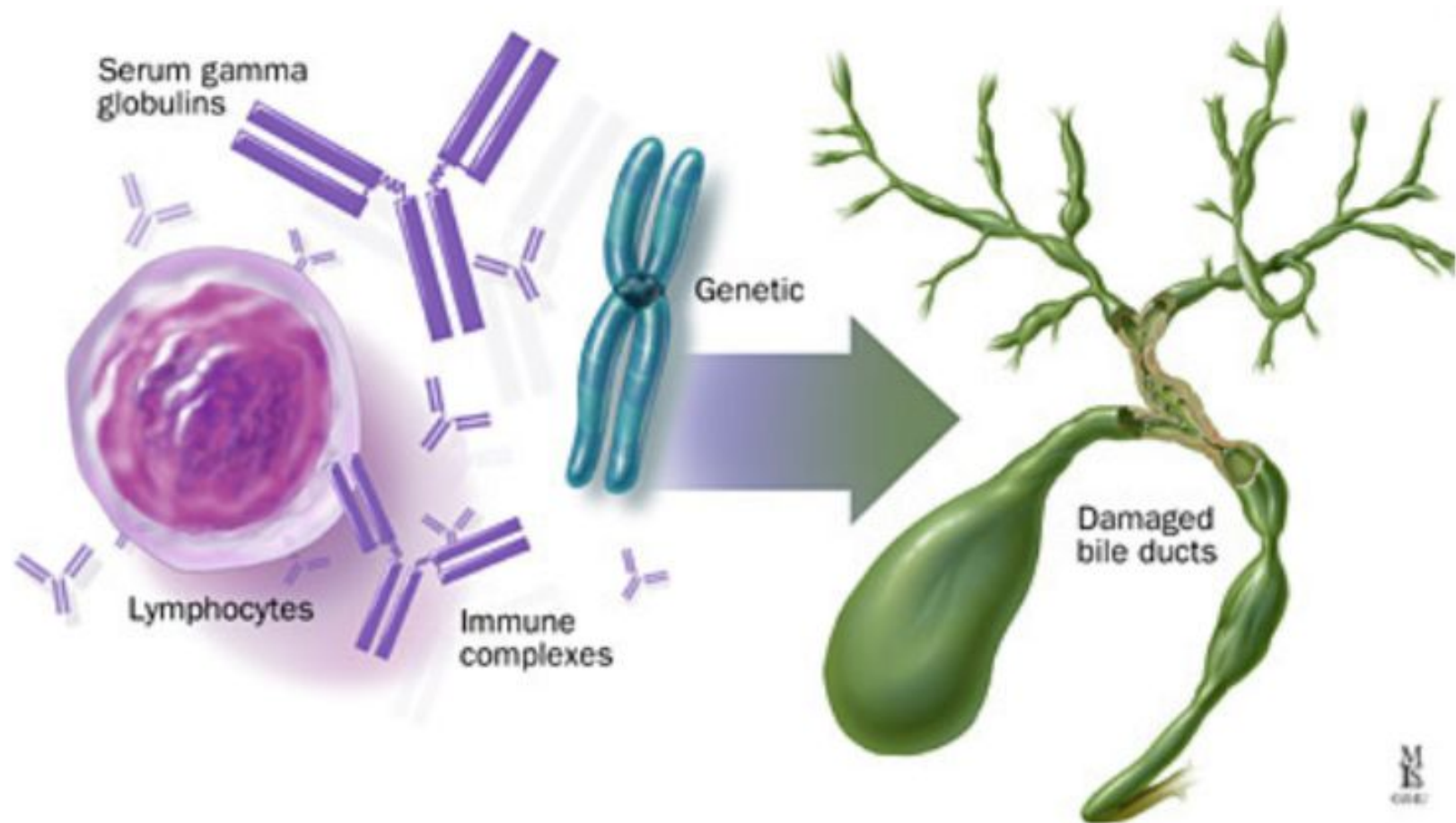


Figure 8. Summary of potential causative mechanisms of primary sclerosing cholangitis.

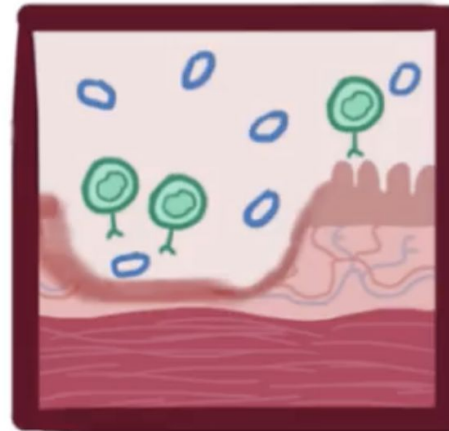
www.johnhopkinsmedicine.org picture credit

ULCERATIVE COLITIS

Add to playlist

affects mucosa
& submucosa
of large intestine
only

Different from
CROHN DISEASE



mucosa

submucosa

muscle layers

AUTOIMMUNE

* T cell's target unclear

ULLER

P-ANCA

(perinuclear antineutrophil)
cytoplasmic antibody



Neutrophil

Cross-react?



Gut
bacteria

Structural
Similarity?

www.osmosis.org picture credit

Signs and Symptoms of PSC



- **Fatigue/Malaise**
- **Pruritus**
- **Jaundice**
- **Weight loss**
- **Dark urine**
- **Light-colored stools**
- **Right Upper quadrant pain**
- **Recurrent febrile episodes with bacterial cholangitis**
- **Night Sweats**
- **Hepatomegaly and Splenomegaly**

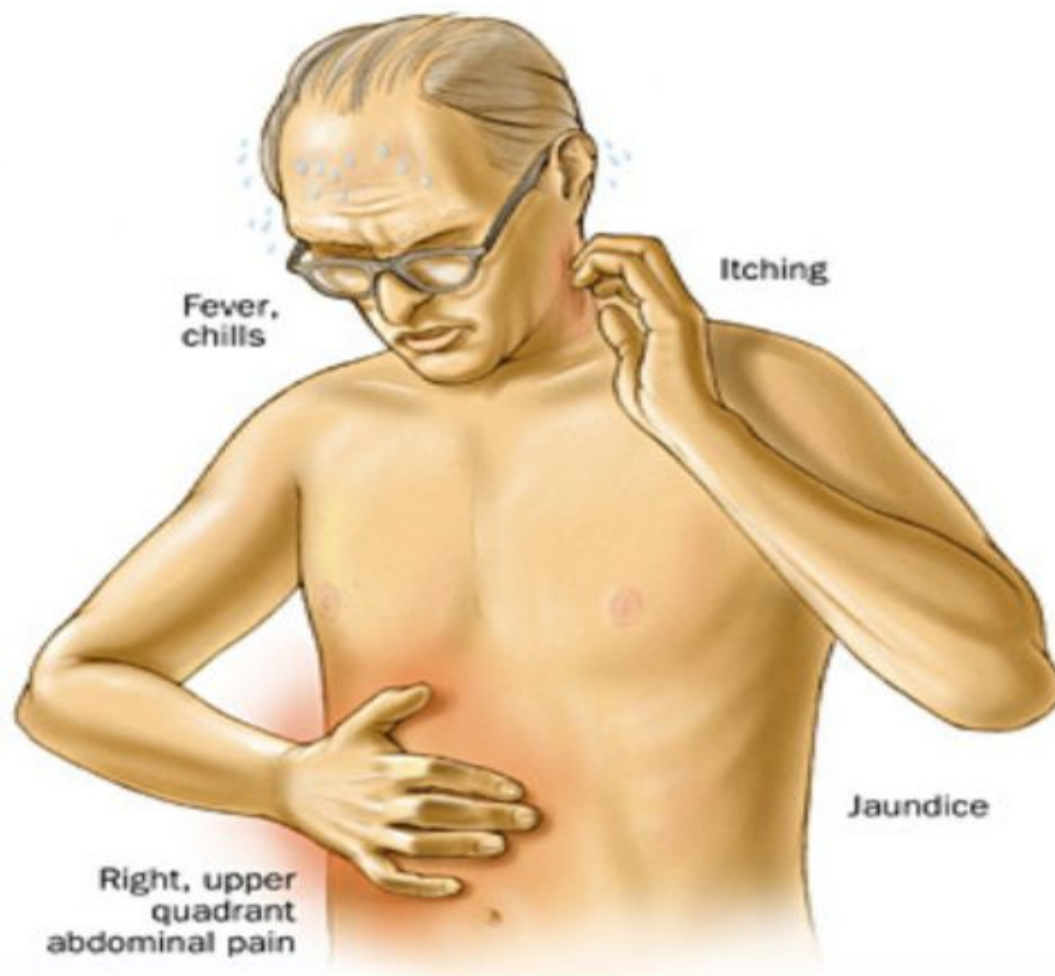
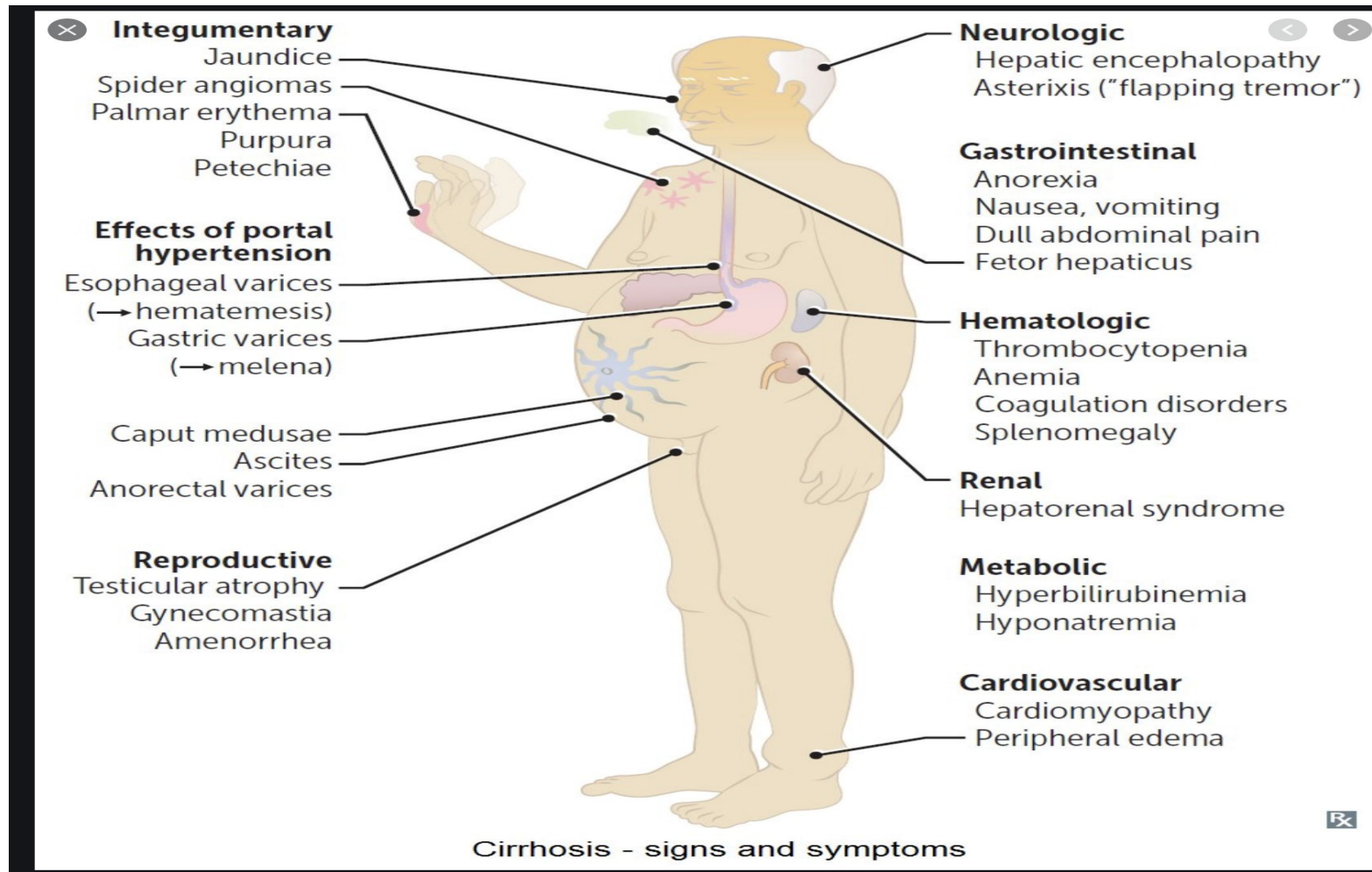


Figure 3. Symptoms of advanced disease.

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With Progression of Disease



Manual of Medicine: picture credit

Hepatocellular carcinoma

0.3%-2.8% lifetime incidence
Unknown risk over general population

Cholangiocarcinoma

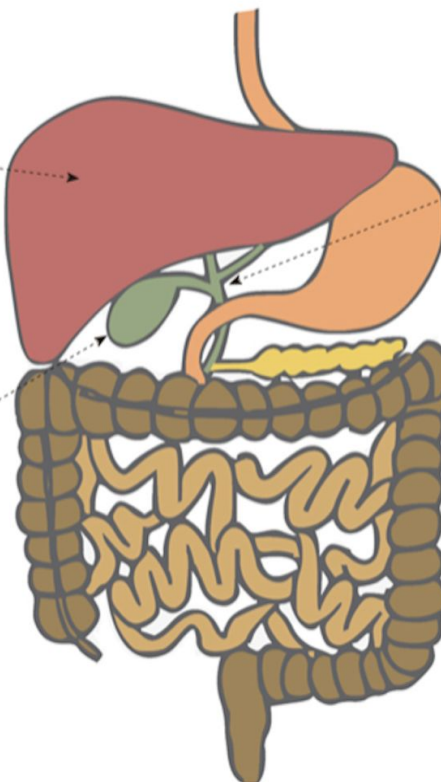
Up to 20% lifetime incidence
400x-1500x risk over general population

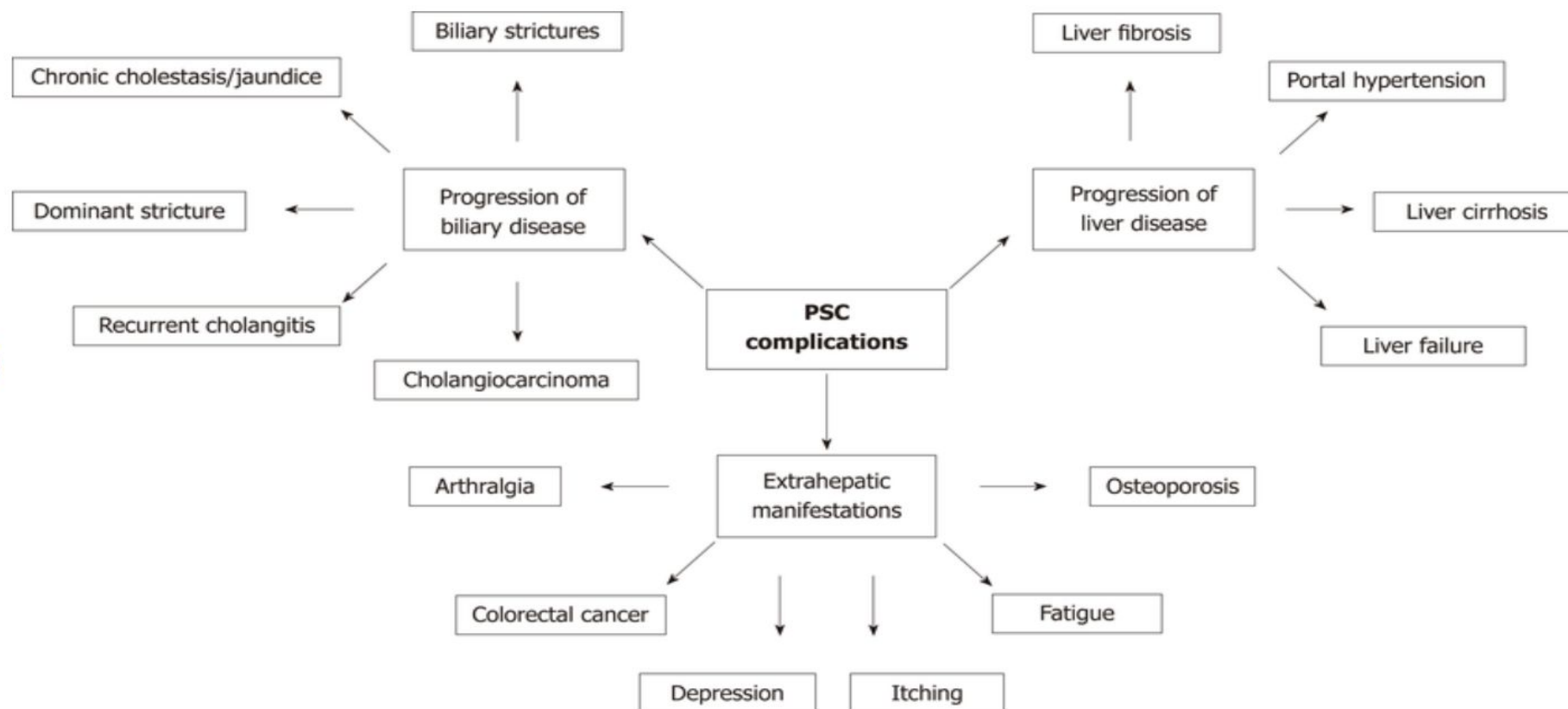
Gallbladder carcinoma

1%-3.5% lifetime incidence
10x risk over general population

Colorectal cancer

20%-30% lifetime incidence in PSC-IBD
10x risk¹ over general population
4x risk¹ over patients with UC





Summary of complications resulting from disease progression in primary sclerosing cholangitis. PSC: Primary sclerosing cholangitis.

| [World Journal of Gastroenterology](#) 25(6):644-658 · February 2019

BILE in the BLOOD

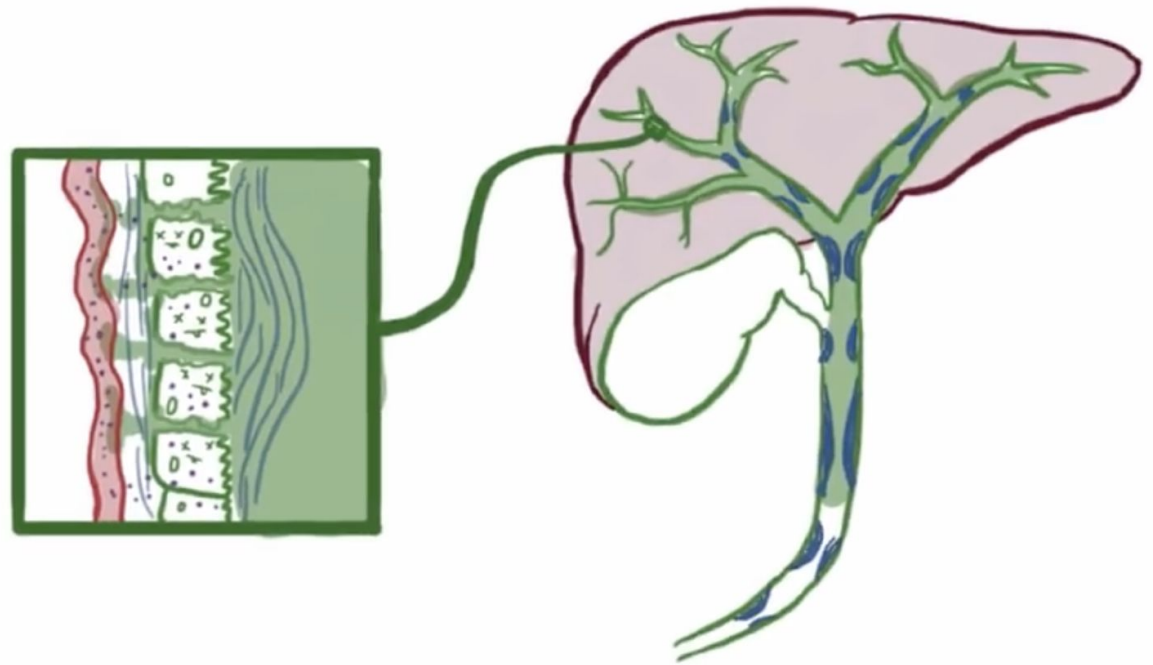
Pruritus (itchiness)
Bile salts



↑ conjugated bilirubin

↑ ALP + GGT

↳ enzymes from liver



BILE in the BLOOD

Pruritus (itchiness)
Bile Salts



↑ conjugated bilirubin

→ KIDNEYS

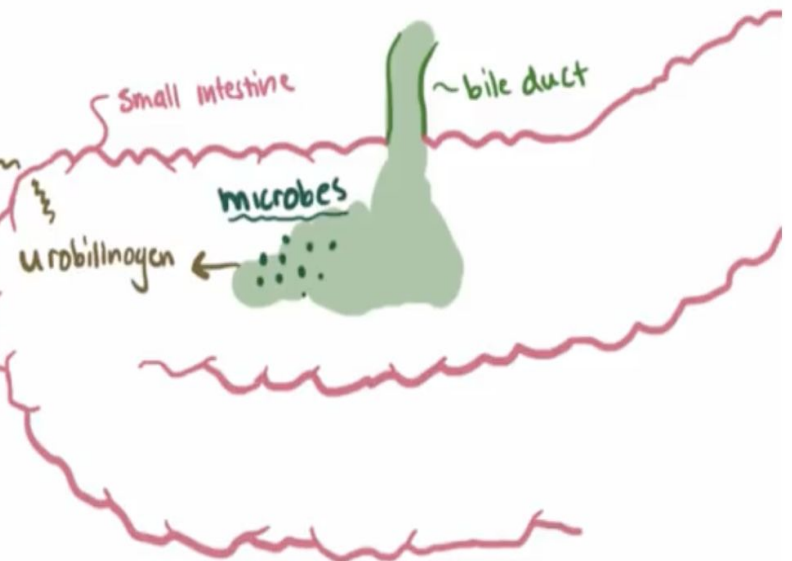
↑ ALP + GGT

↳ enzymes from liver

↑ conjugated bilirubin } bilirubinuria

Normally

Small amount UROBILINOGEN



*** BILE ***
in the
BLOOD

Pruritus (itchiness)
Bile salts



↑ conjugated bilirubin → KIDNEYS

↑ ALP + GGT

↳ enzymes from liver

Urobilinogen
NOT in the
BLOOD

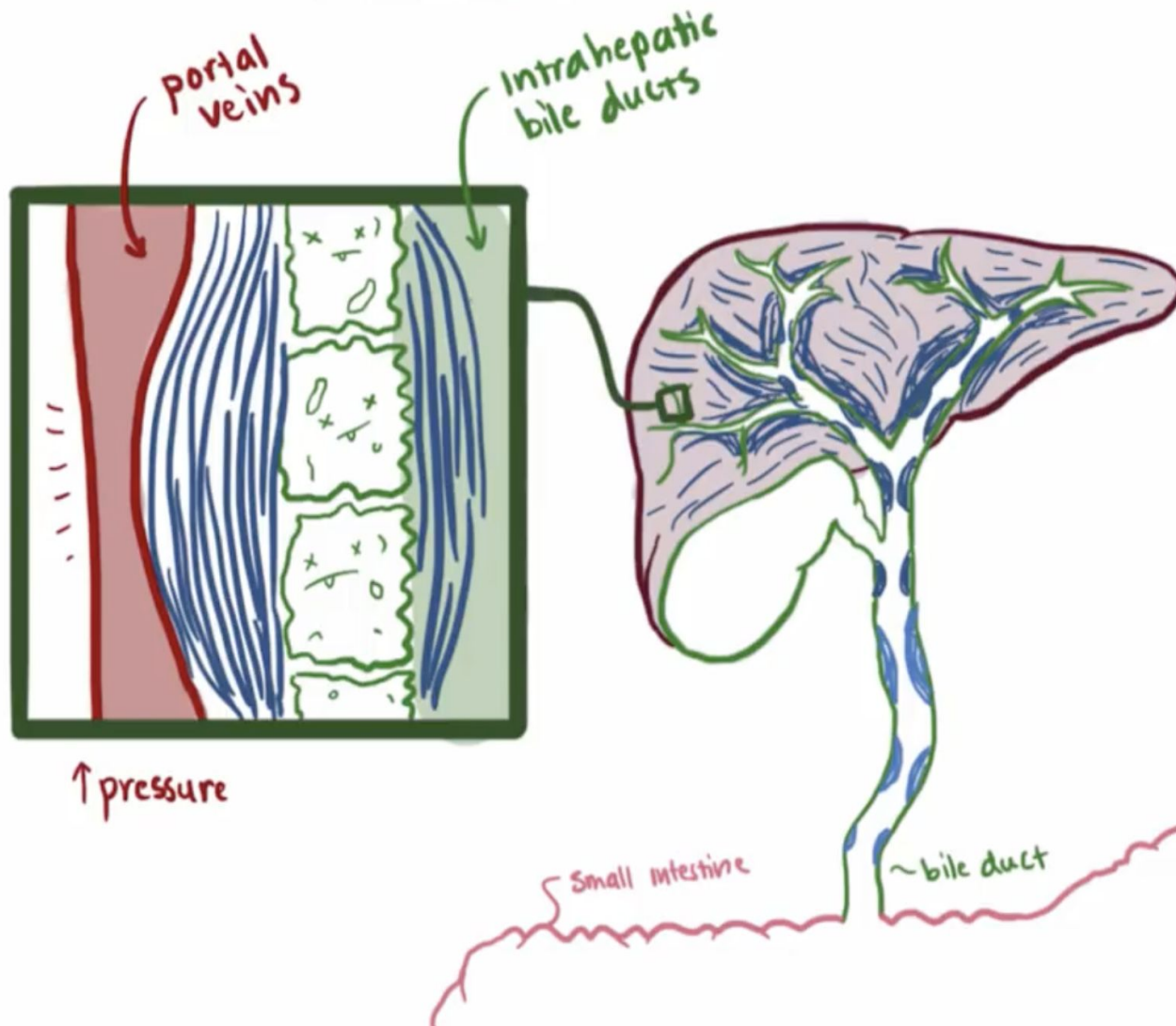
* bile has to get to
gut for urobilinogen
production!



PSC
NO UROBILINOGEN



↑ conjugated bilirubin } bilirubinuria
↓ urobilinogen



LIVER COMPLICATIONS

* Portal hypertension

↓
Fluid in Spleen + liver
(hepatosplenomegaly)

* CIRRHOSIS

↳ Stops functioning
from damage

~ esophagus

~ stomach

TREATMENT

- * immunosuppressant & anti-inflammatory medications
 - ↳ NONE proven to slow disease
- * LIVER transplant
 - ↳ for advanced liver disease

LIVER COMPLICATIONS

- * Portal hypertension
 - ↓
 - Fluid in Spleen + liver (hepatosplenomegaly)
- * CIRRHOSIS
 - ↳ Stops functioning from damage

BILE DUCT COMPLICATIONS

- * ↑ cholangiocarcinoma

Labs

LIVER FUNCTION TESTS

Test		Actual Measurement Value
Total Protein	g/L	79
Albumin	g/L	41
Globulin	g/L	28
Total Bilirubin	umol/L	8
Conjugated Bilirubin	umol/L	2
Alk.Phosphatase	IU/L	112
ALT	IU/L	22
AST	IU/L	21
Gamma GT	IU/L	

Imaging



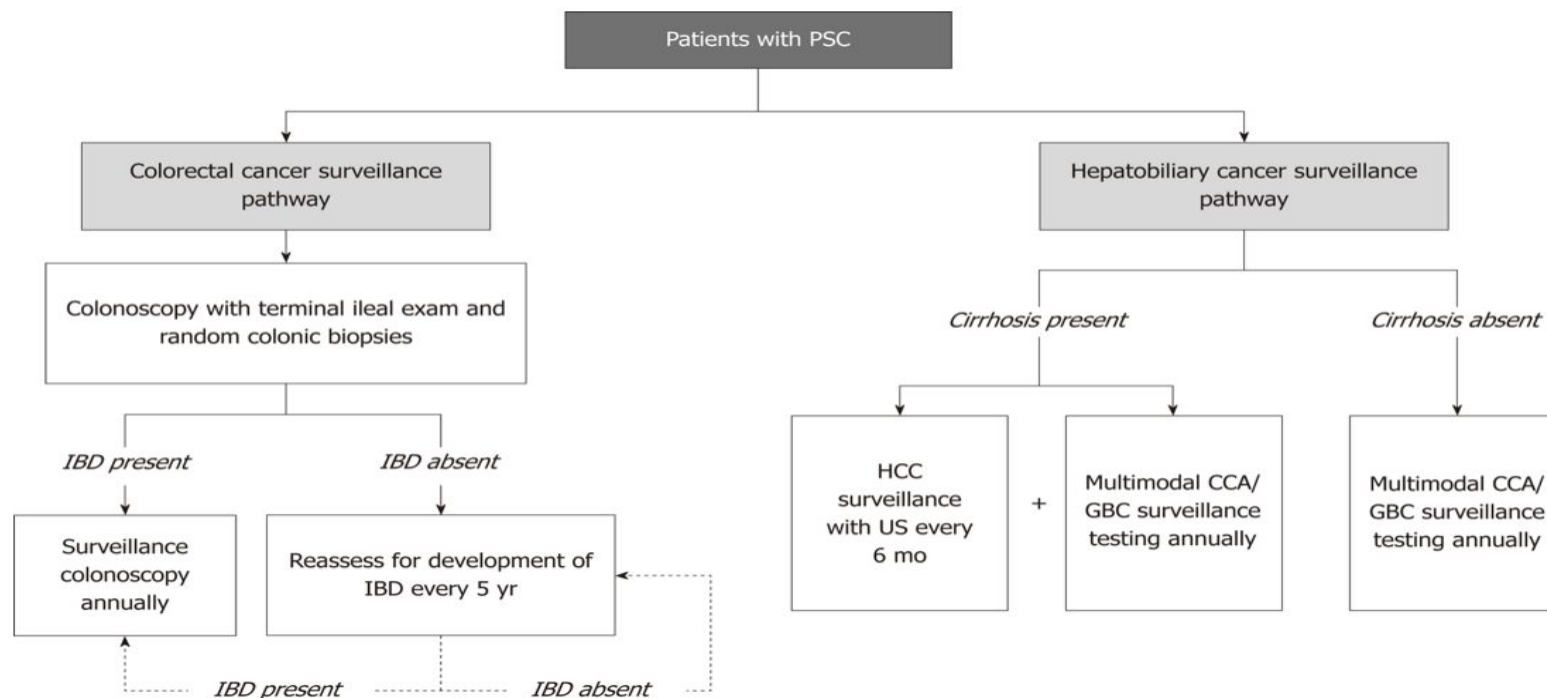


Figure 2 Overview of cancer surveillance in patients with primary sclerosing cholangitis, beginning at time of primary sclerosing cholangitis diagnosis. This overview is based on recommendations from the American Association for the Study of Liver Disease practice guidelines[44]. CCA: Cholangiocarcinoma; GBC: Gallbladder carcinoma; HCC: Hepatocellular carcinoma; IBD: Inflammatory bowel disease; US: Ultrasound; PSC: Primary sclerosing cholangitis.

- LFT's
- Immunoglobulin
- Autoantibodies
- MRCP
- ERCP
- Liver Biopsy
- Colonoscopy
- CT or MRI

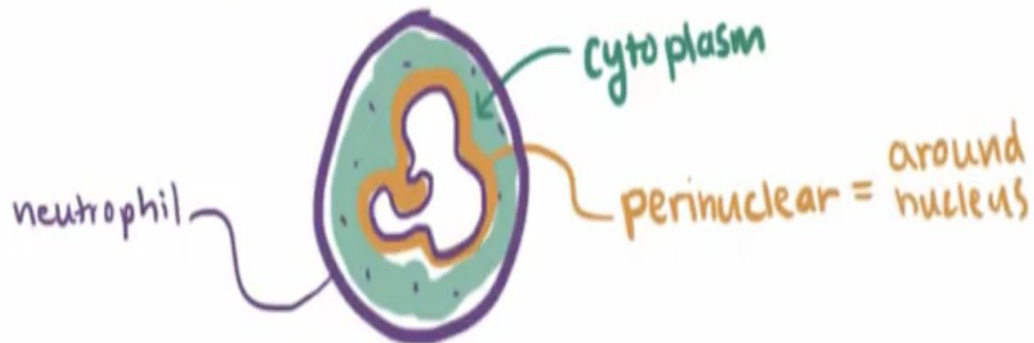
CASE

MORE AUTOIMMUNE EVIDENCE

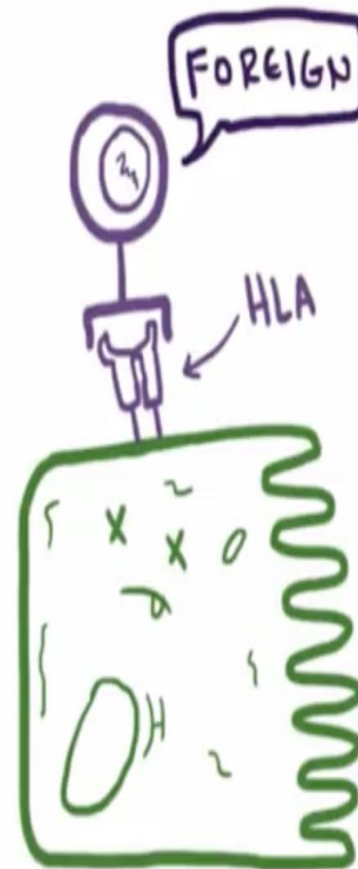
↑ IgM antibodies

↑ p-ANCA

perinuclear anti-neutrophil cytoplasmic antibody



Human Leukocyte Antigens

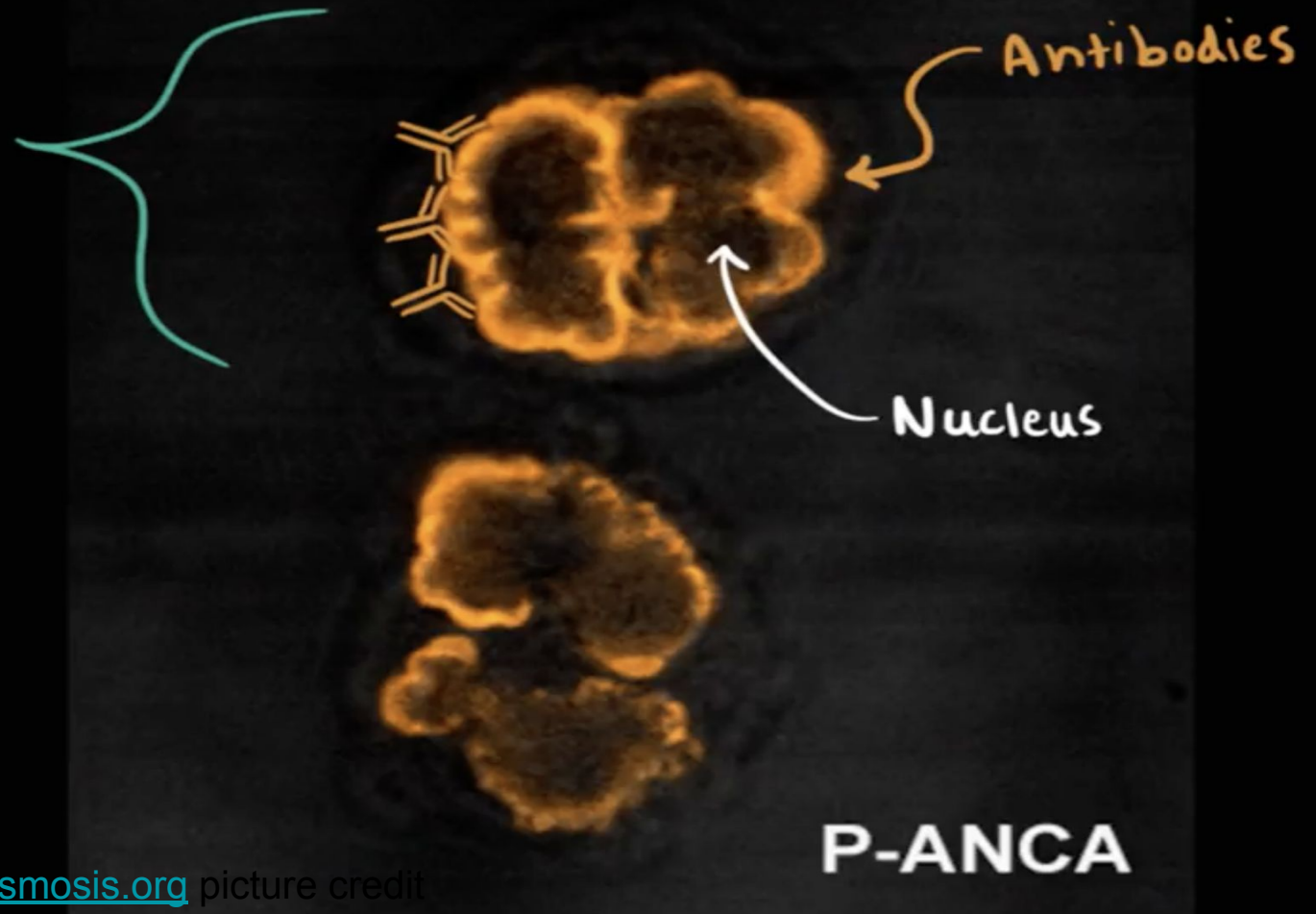


PSC patients:

- HLA-B8
- HLA-DR3
- HLA-DRw52a

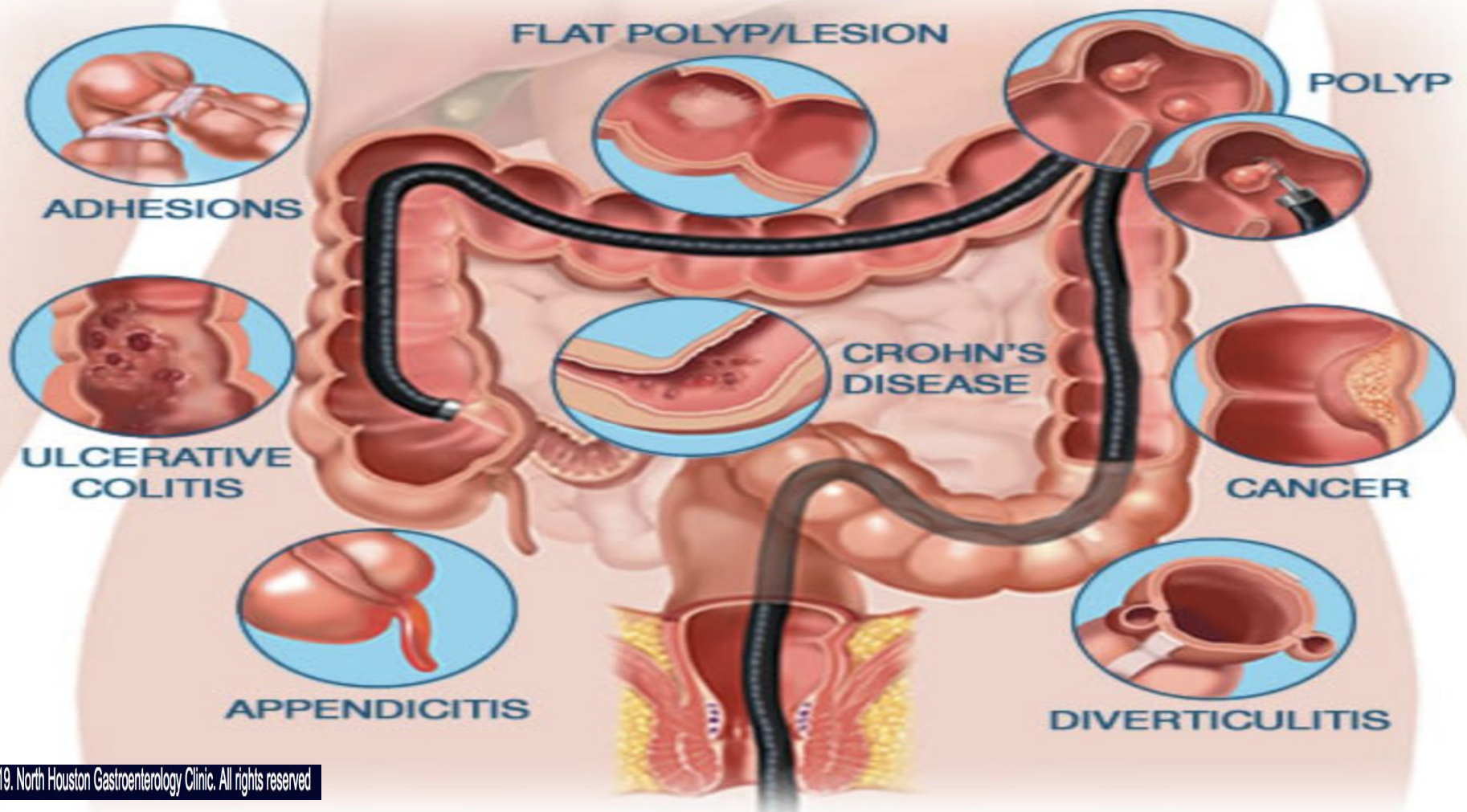
www.osmosis.org picture credit

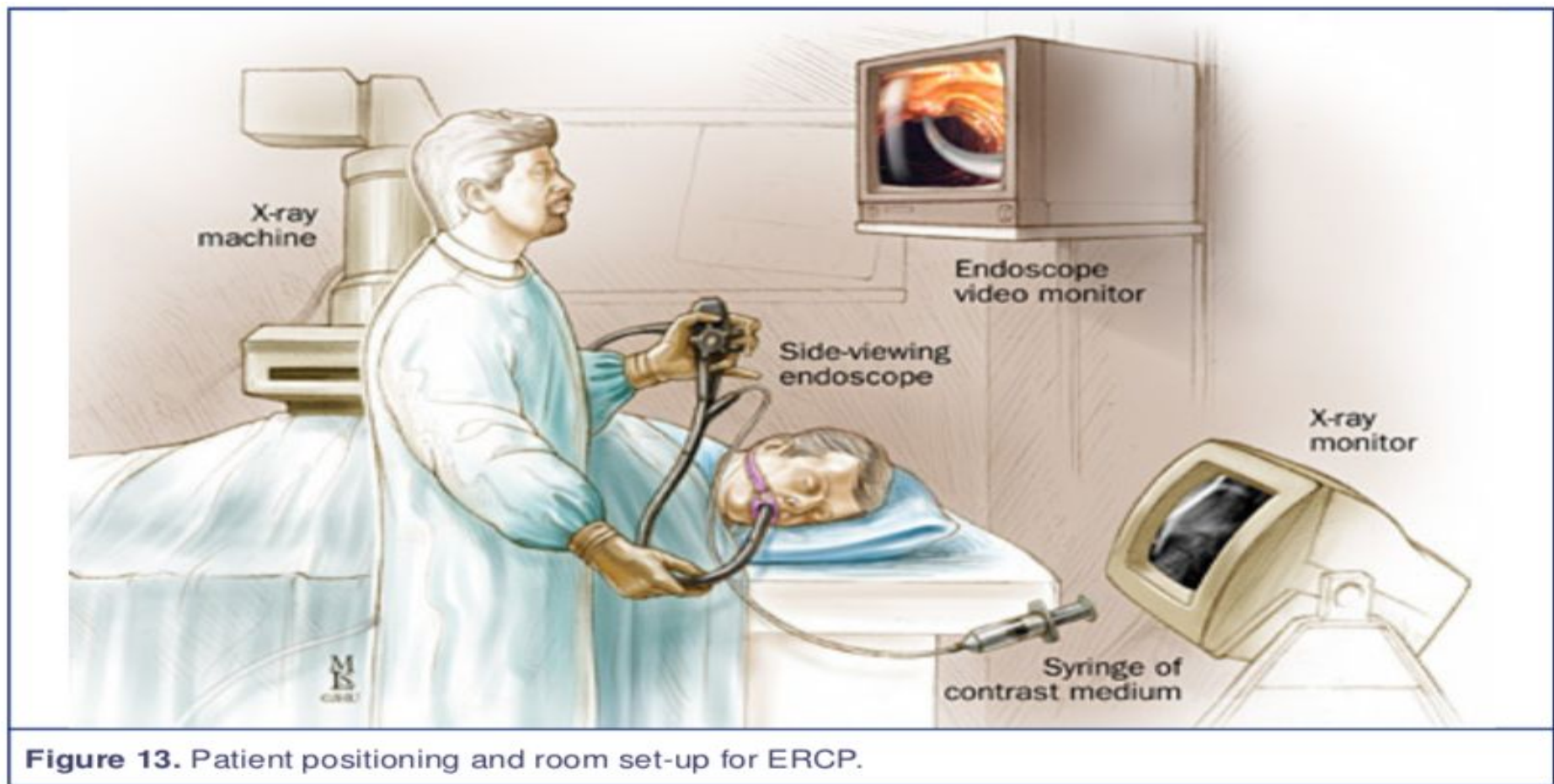
~80% Patients
with PSC



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The COLONOSCOPY



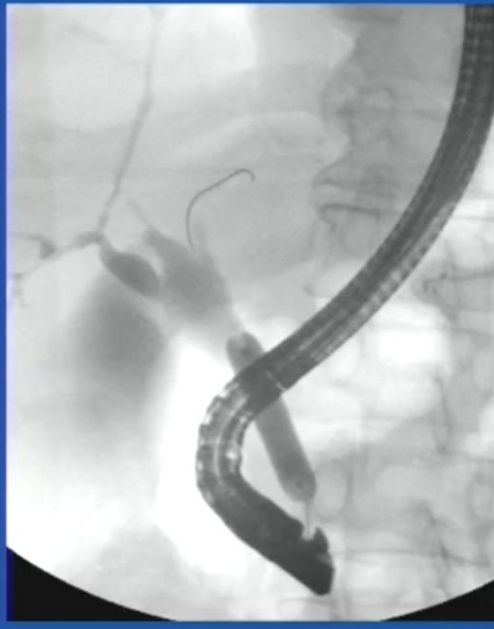


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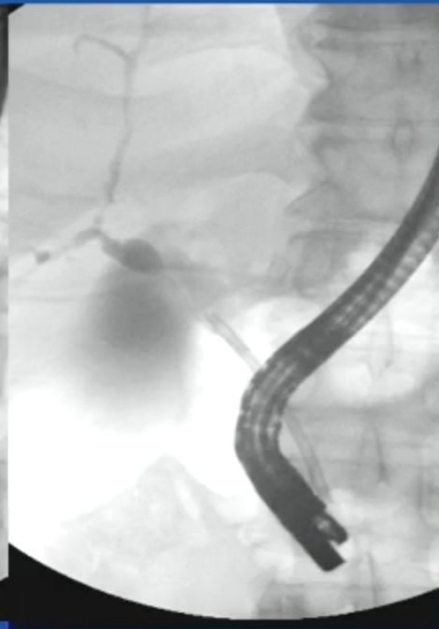
Balloon Dilation and Stenting in PSC



stricture



dilation

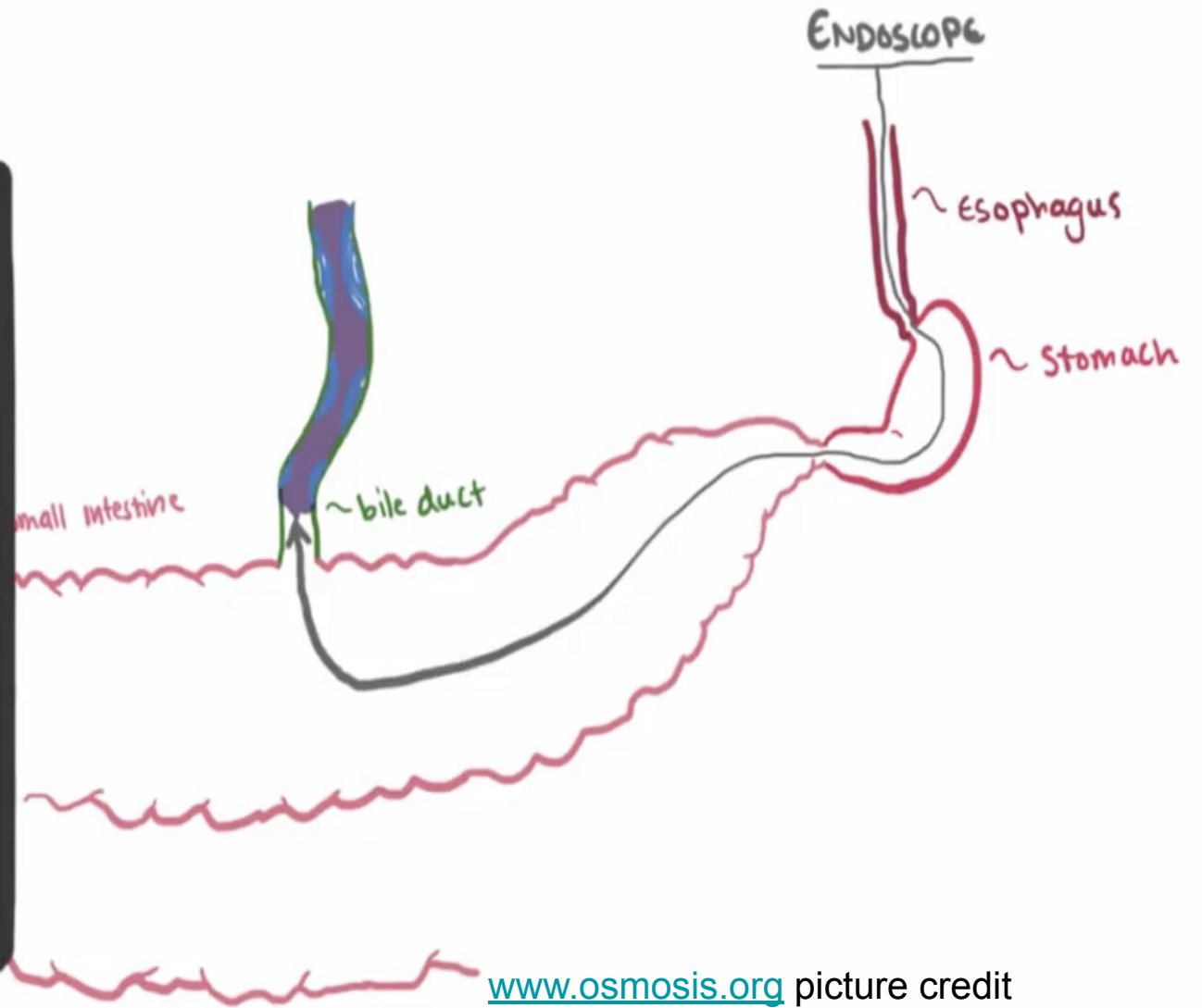
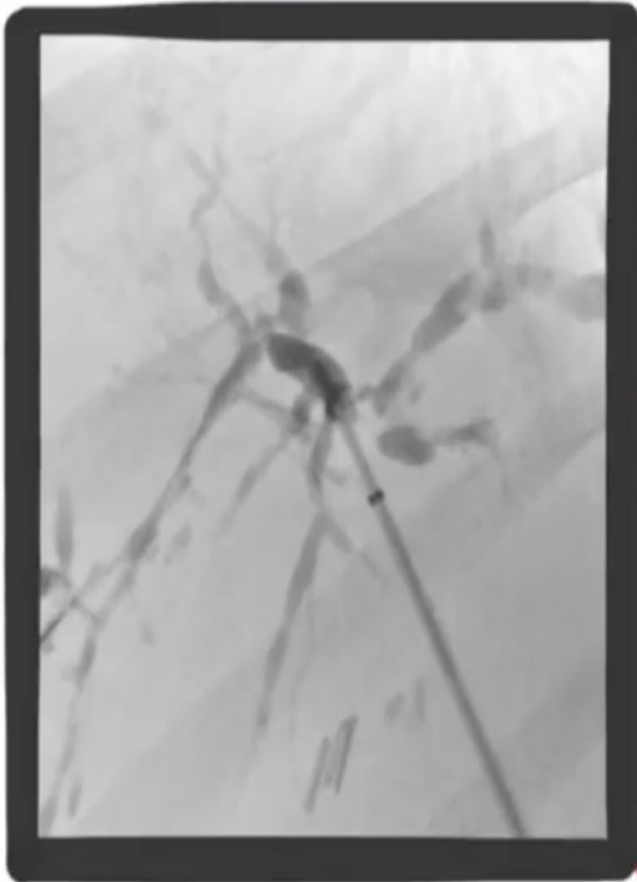


stent



Post stent

ERCP



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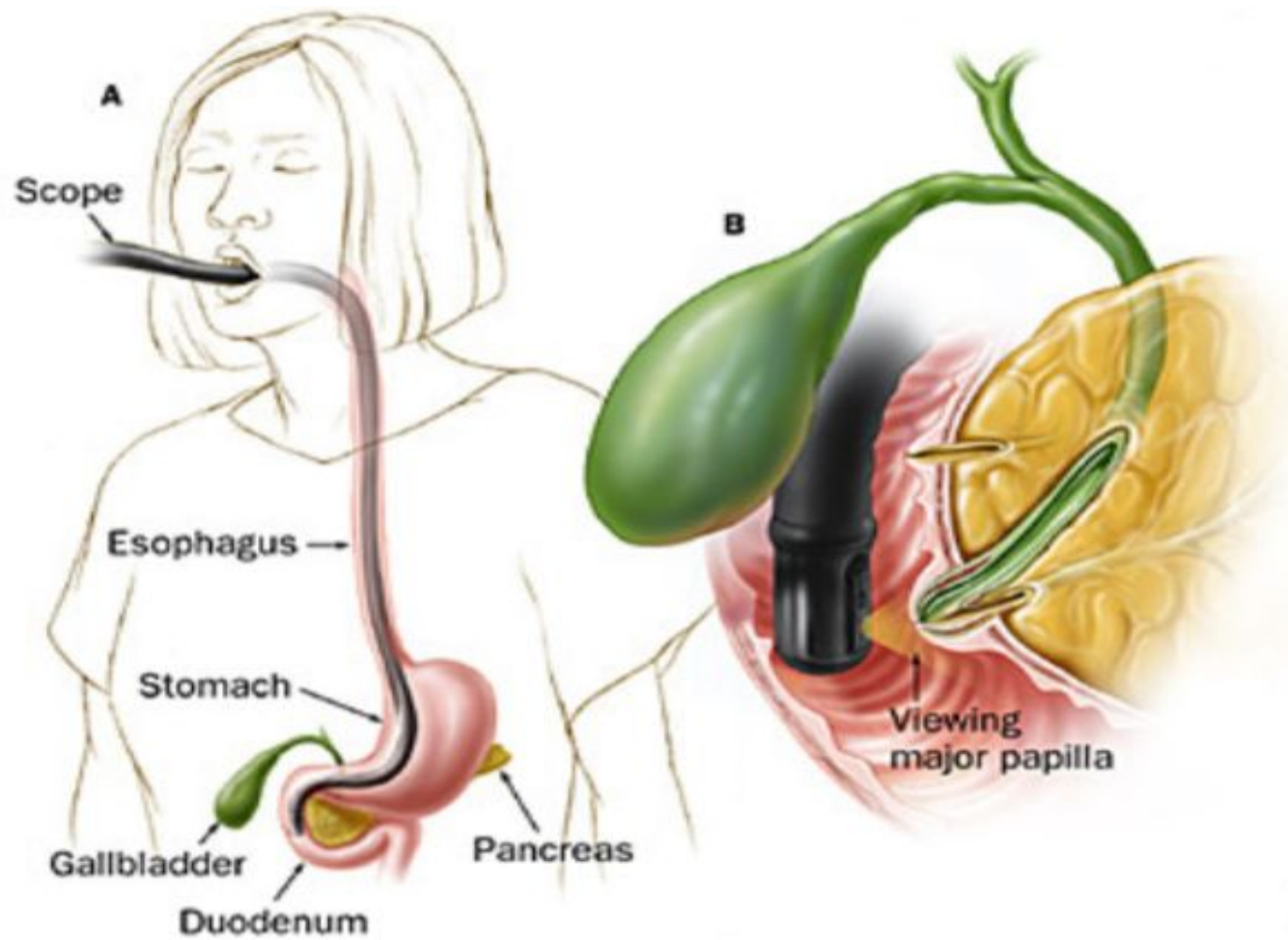
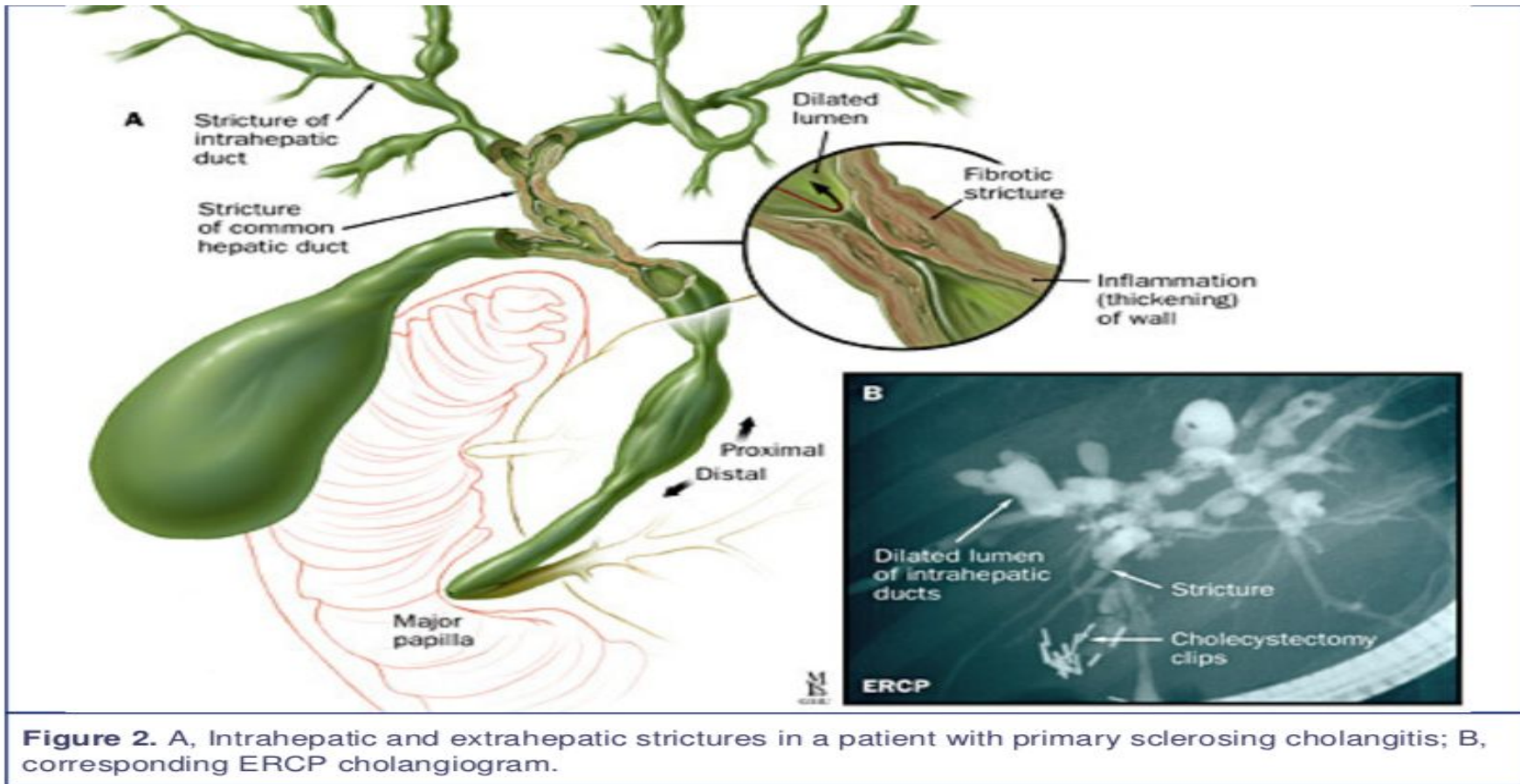


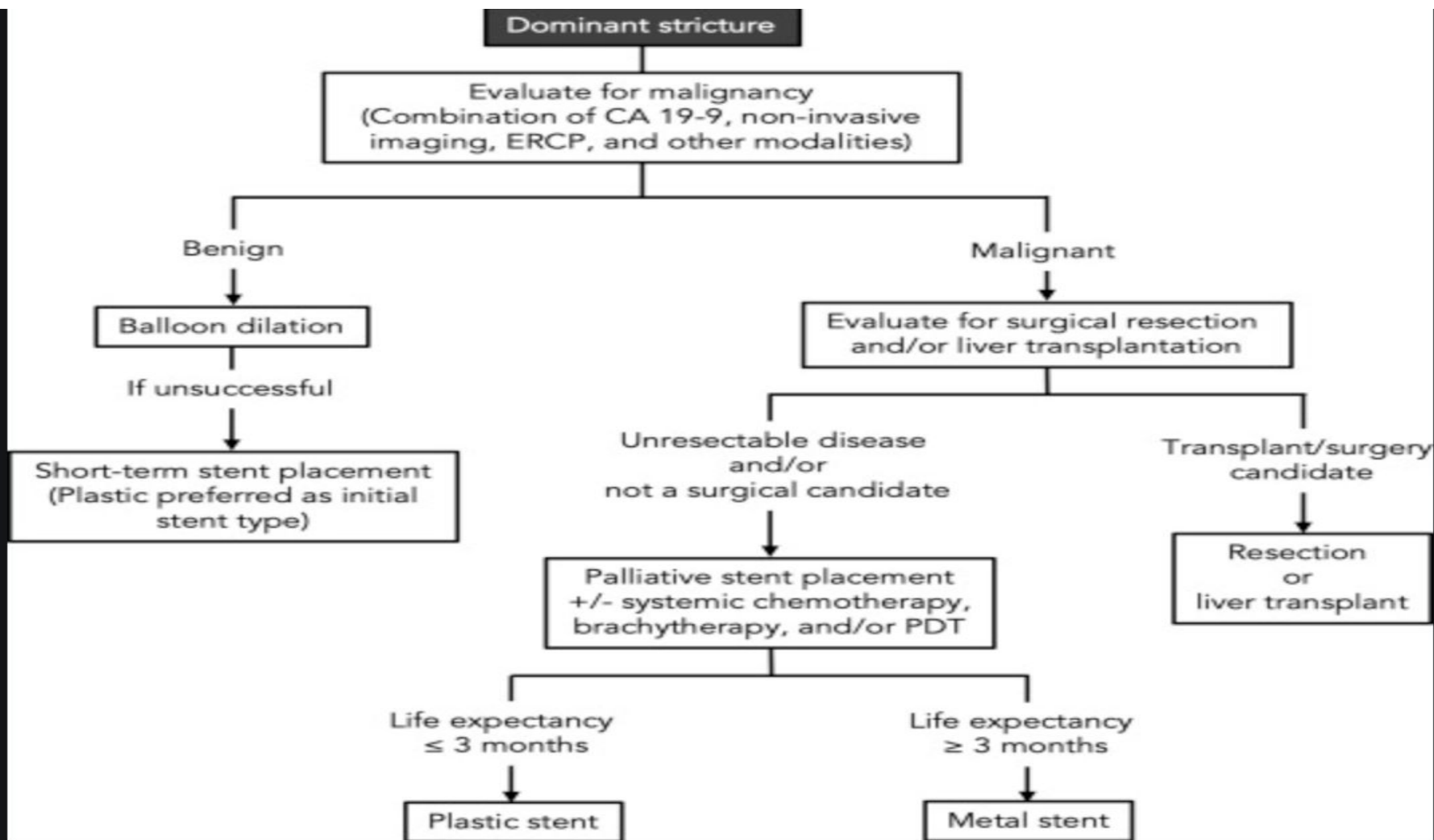
Figure 11. A, B, Position of the side viewing endoscope facing the major papilla.

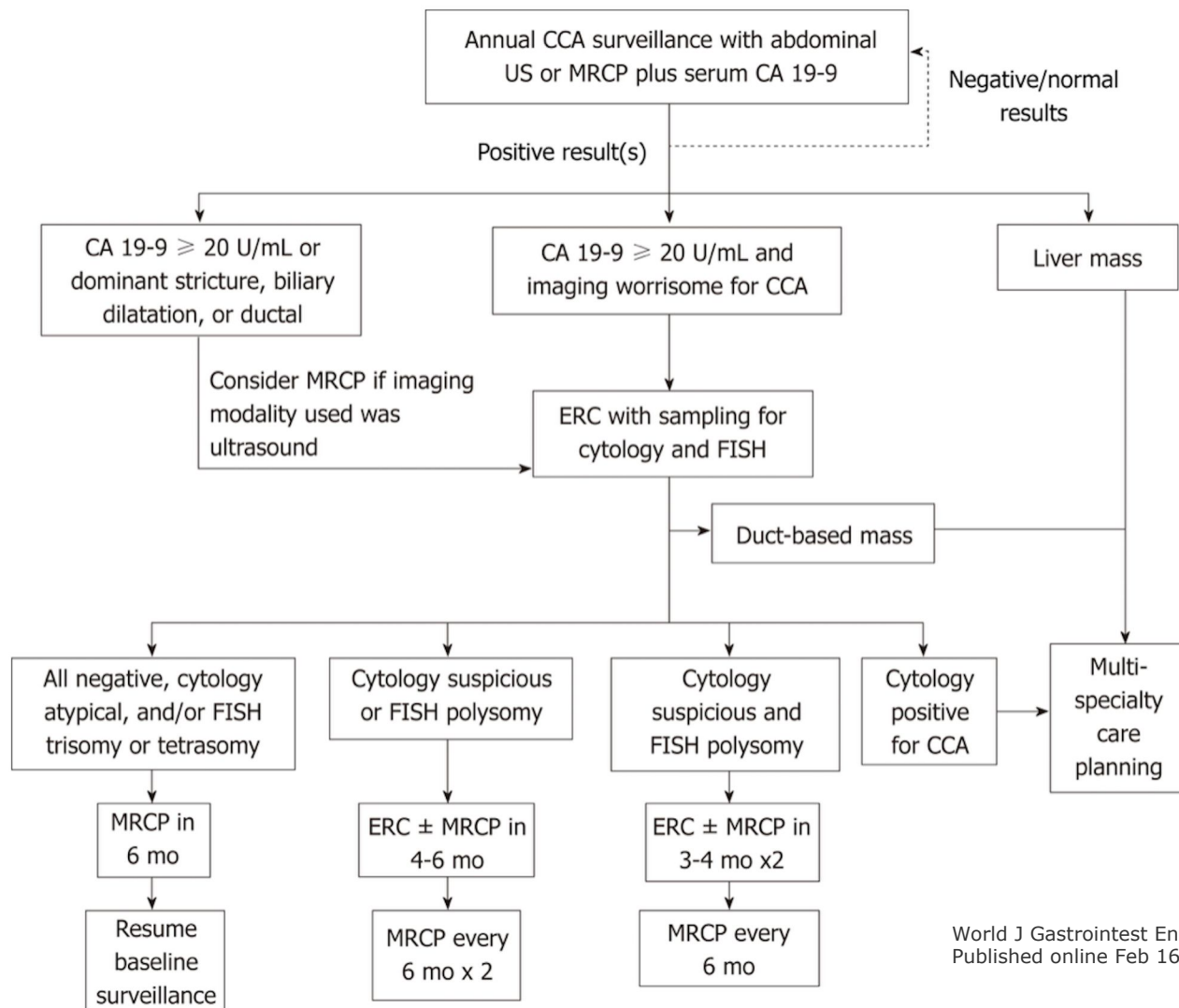
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ERCP: to help remove blockages caused by strictures by placing temporary plastic stents and enlarge narrowed ducts by performing balloon dilatations. Brushings of the bile ducts also help to diagnose bile ducts cancers.

www.johnhopkinsmedicine.org picture credit





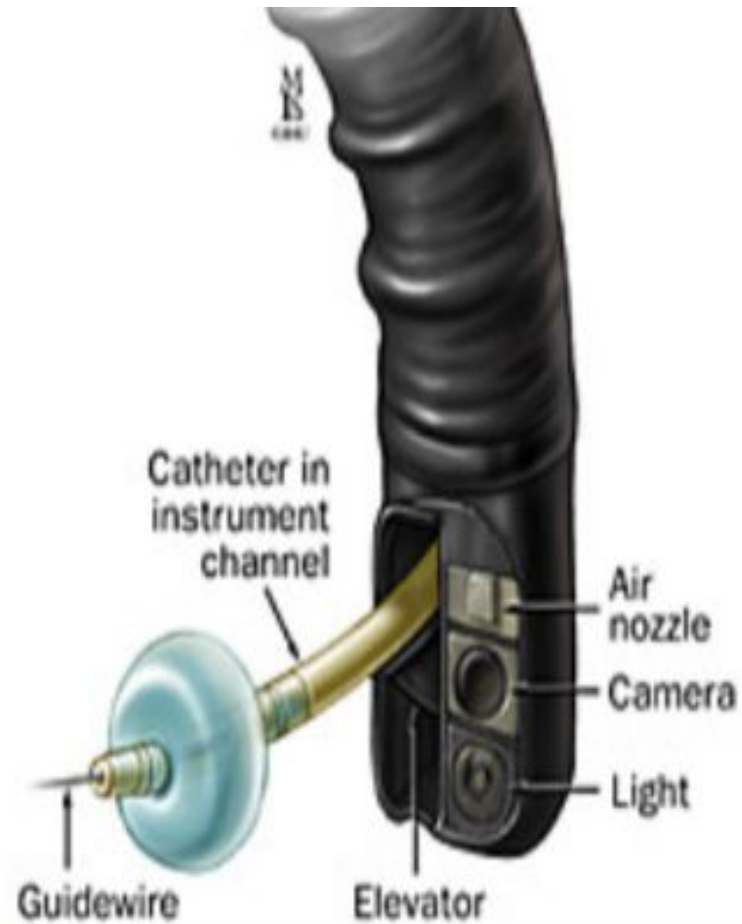


Figure 12. Tip of the side viewing endoscope.

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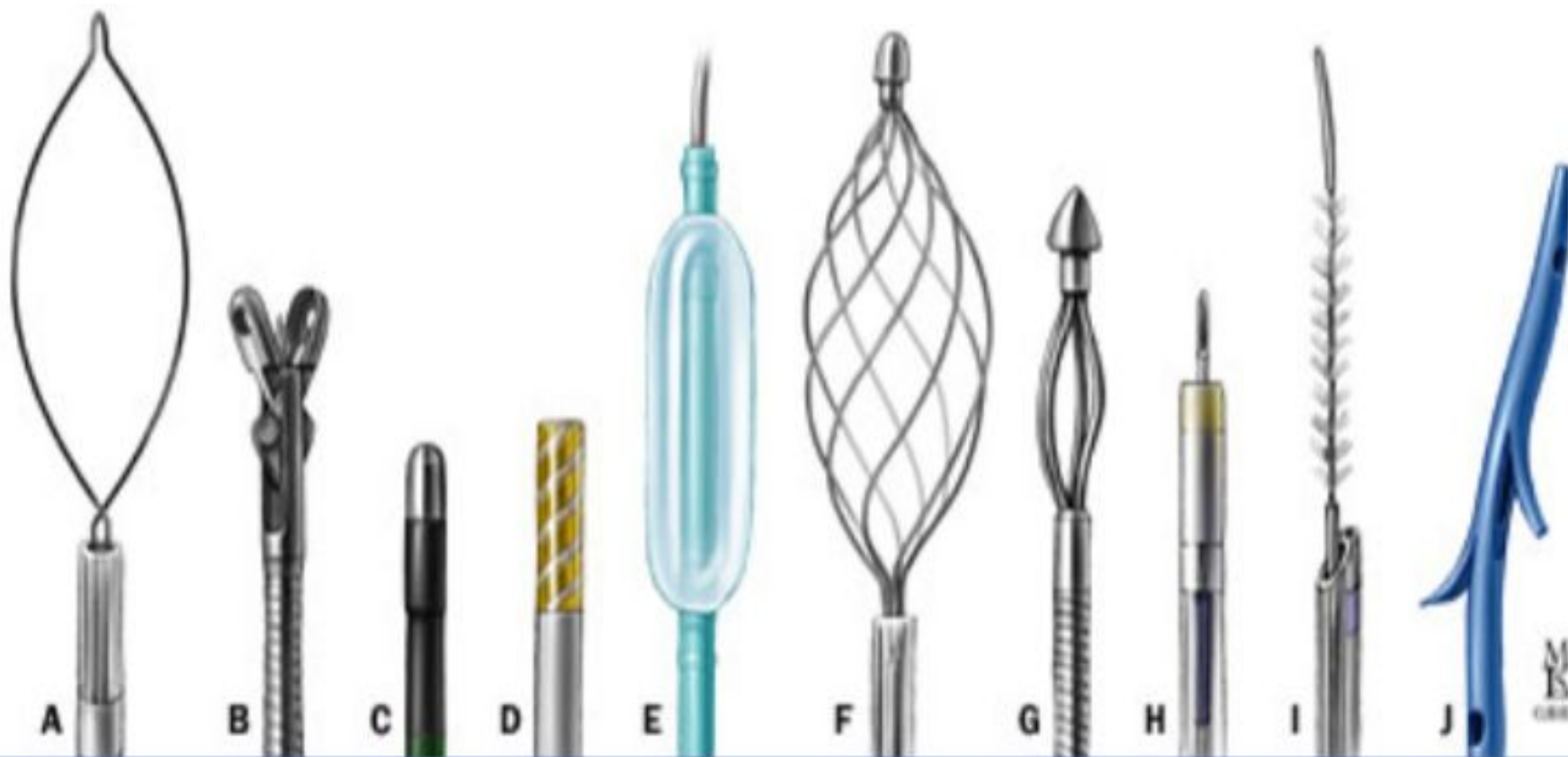


Figure 14. Accessory instruments used through the endoscope; A, snare; B, biopsy forceps; C, heater probe; D, BICAP probe; E, dilation balloon with guide wire; F, stone retrieval basket; G, mechanical lithotripter; H, sclerotherapy needle; I, cytology brush; J, plastic stent.

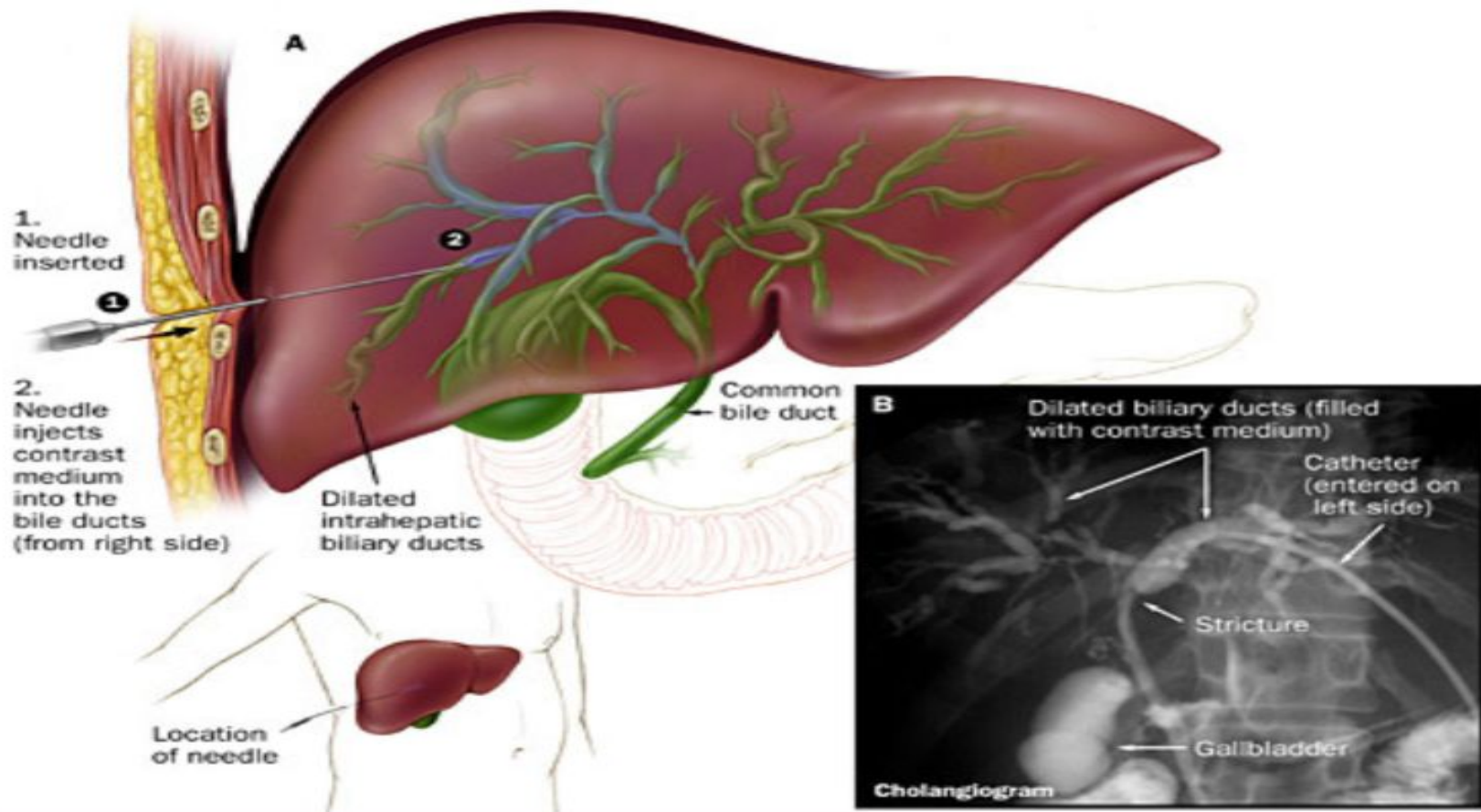


Figure 15. A, Technique of percutaneous transhepatic cholangiography; B, corresponding cholangiogram.

PTC: If there is a failure to cross the stricture by ERCP then a percutaneous transhepatic drain is placed.

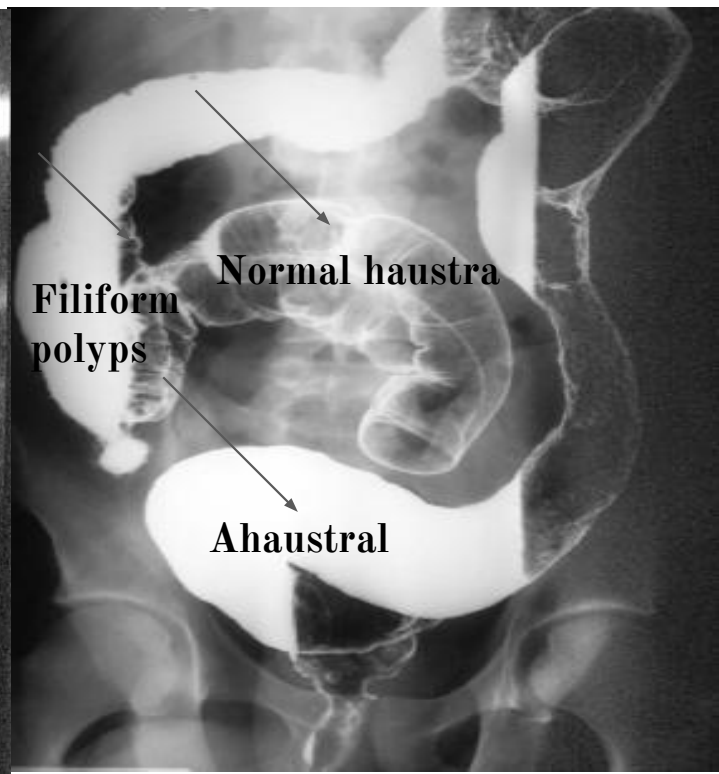
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Ercep intrahepatic bile strictures and beading.

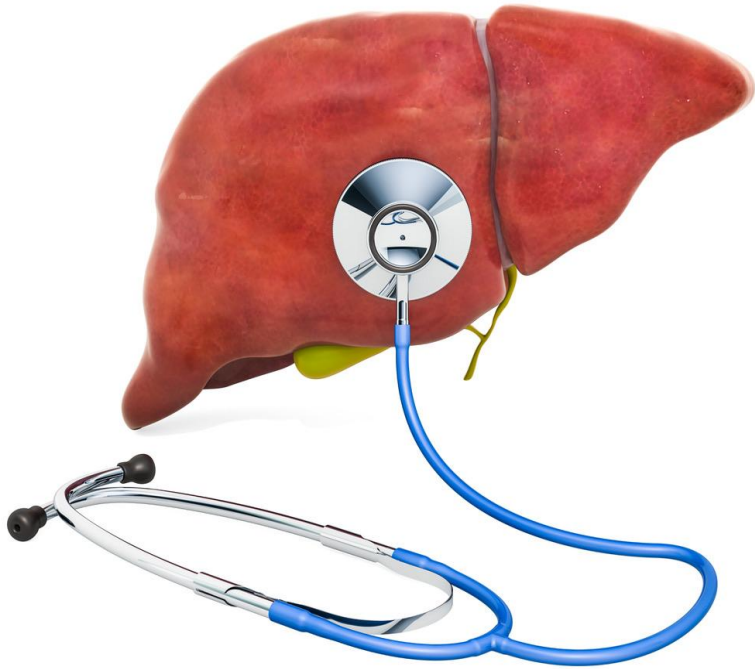


MRCP shows R And L hepatic duct strictures and dilated proximal left duct.



Filiform polyps(finger-like projections) and an ahaustral("lead pipe appearance") colon secondary to UC.

Treatment of PSC



- **Symptomatic Treatment**
- **Pruritus**
- **Cholangitis**
- **Fatigue**
- **Bone disease**
- **Autoimmune Hepatitis**
- **High IG4 levels**
- **Ursodiol**
- **Biologics**
- **ERCP**
- **PTC**
- **Liver Transplantation**

Liver Biopsy: (Is invasive procedure involving a biopsy of liver tissue to assess how far the PSC has progressed.)

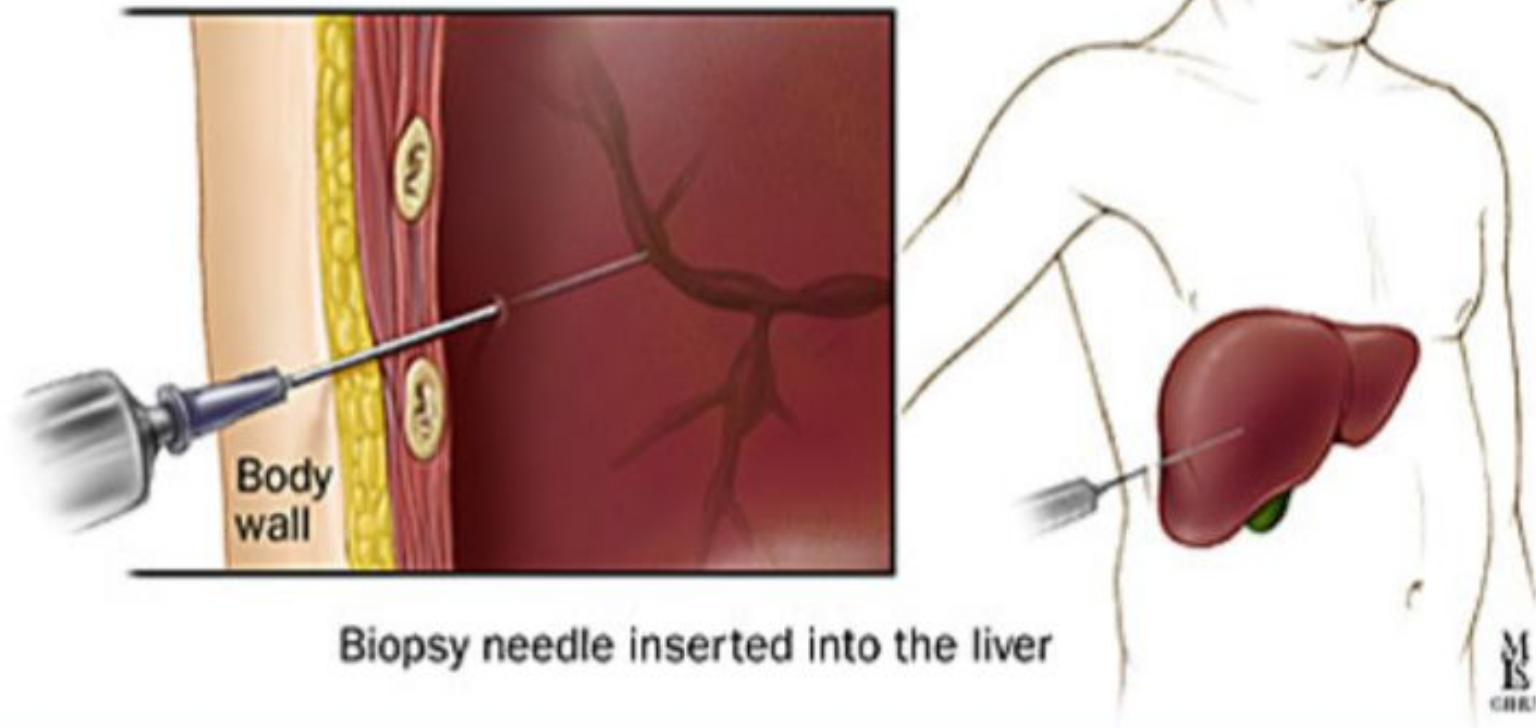
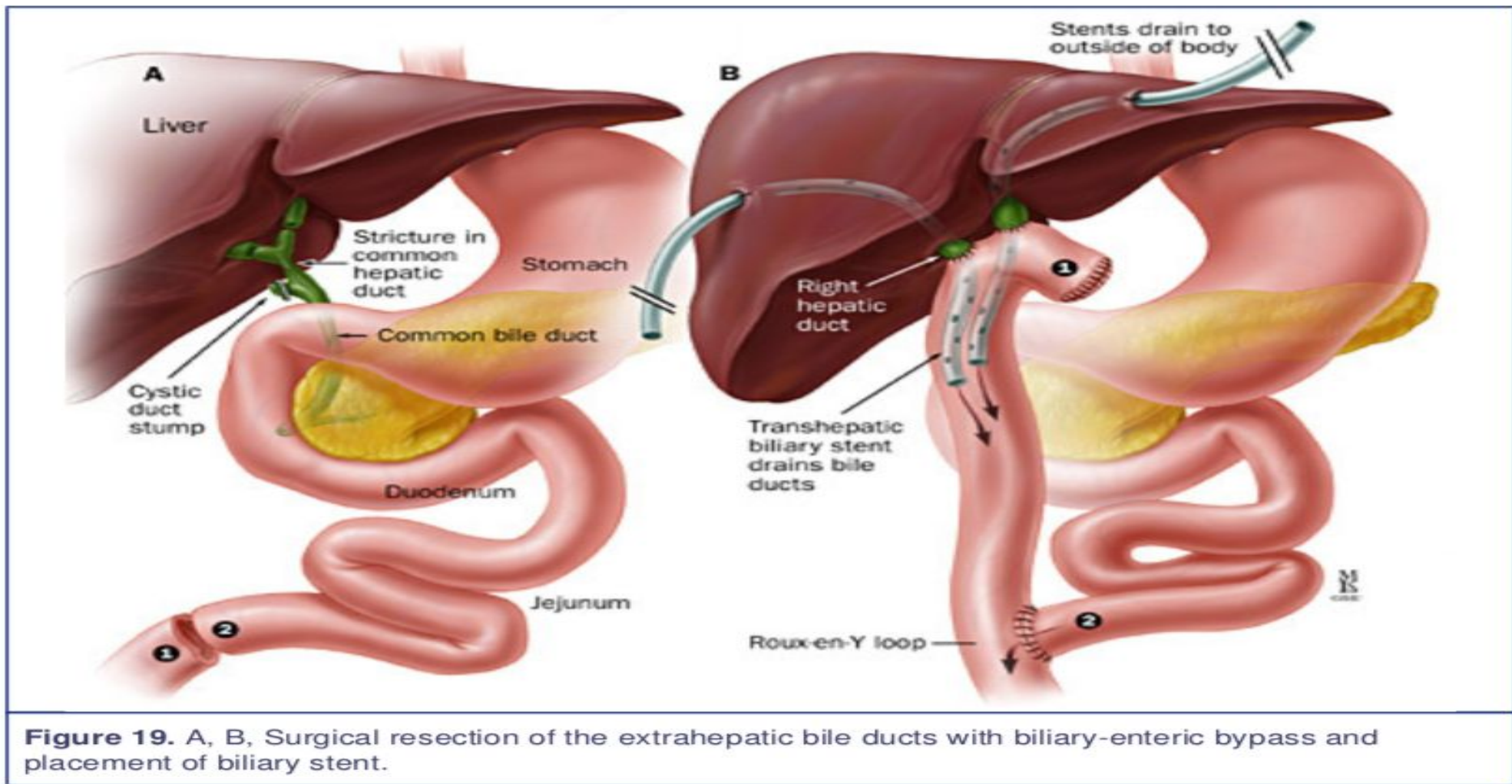


Figure 9. Technique for liver biopsy.

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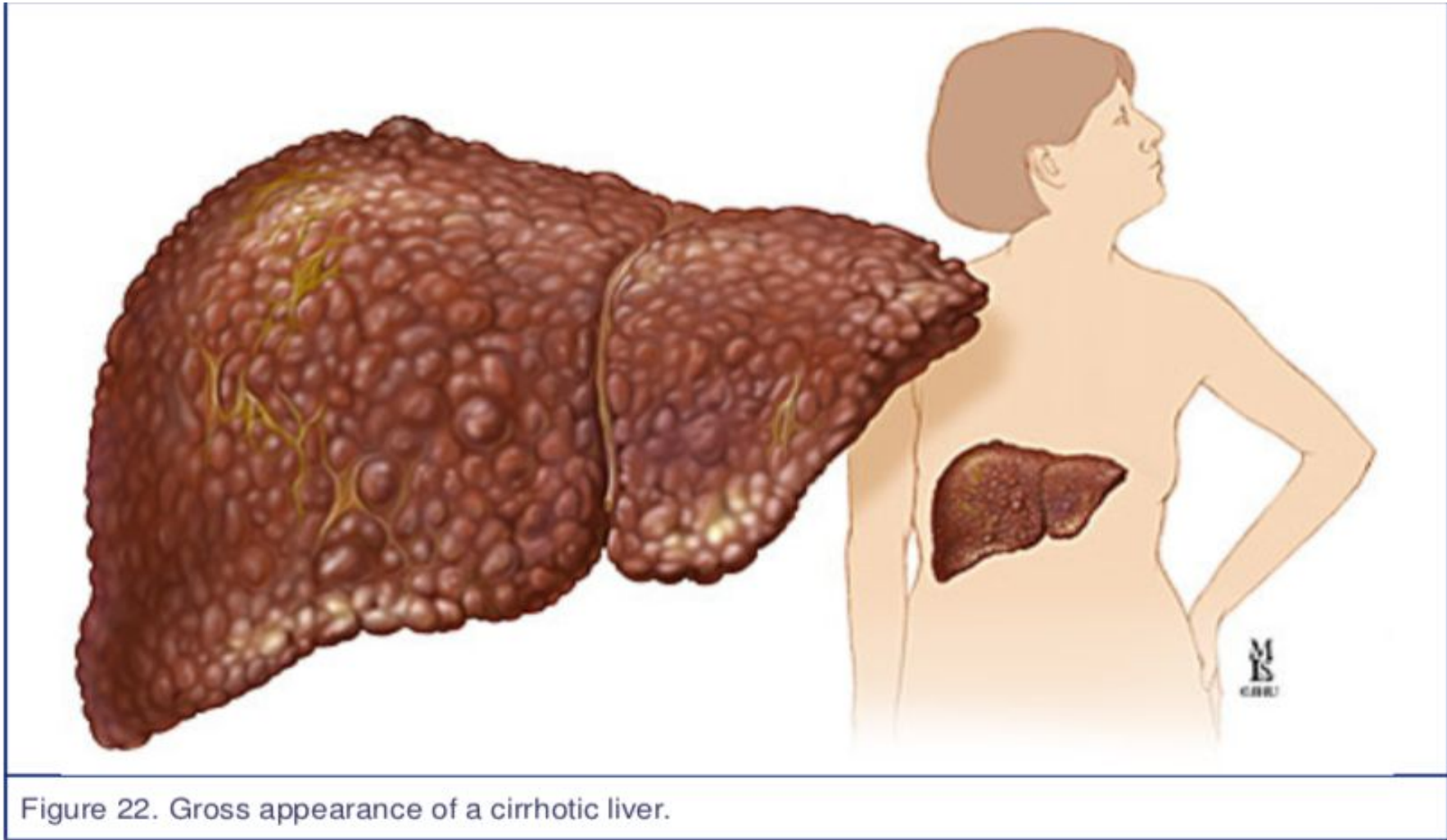


Figure 22. Gross appearance of a cirrhotic liver.

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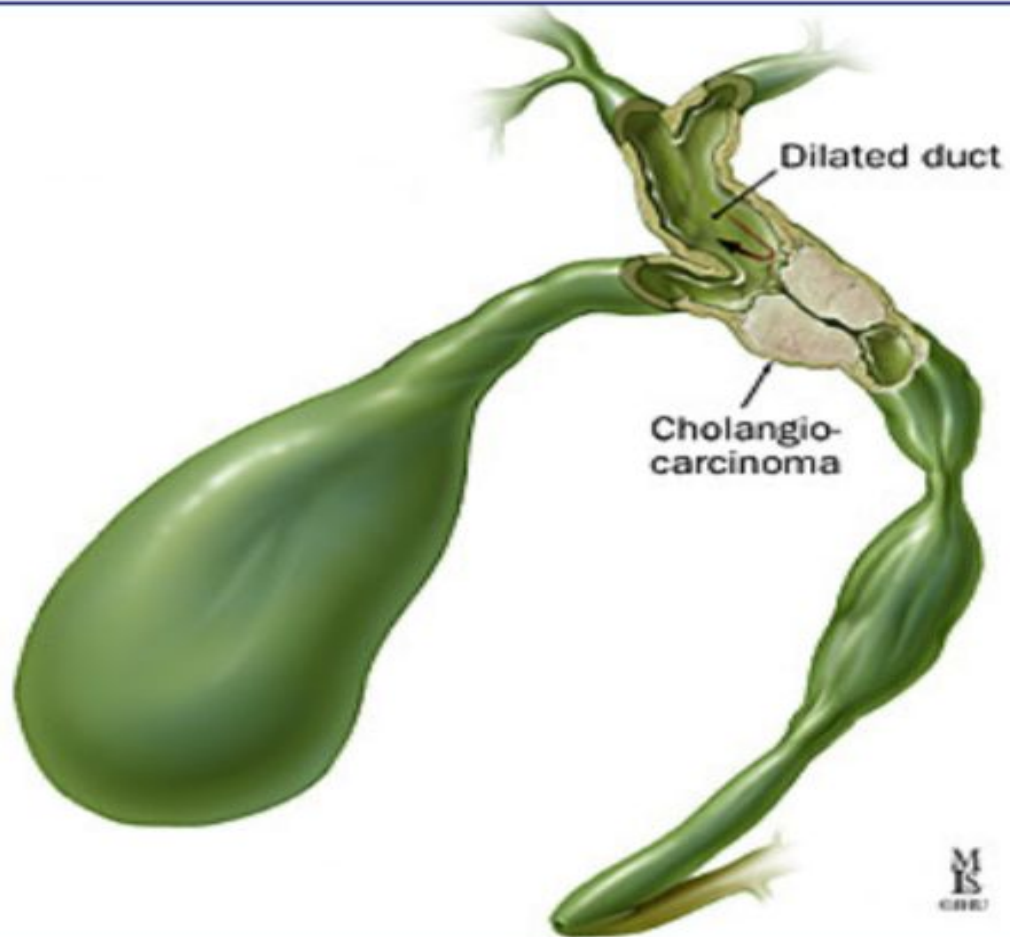


Figure 21. Cholangiocarcinoma of the common bile duct with resultant ductal dilation.

www.johnhopkinsmedicine.org picture credit

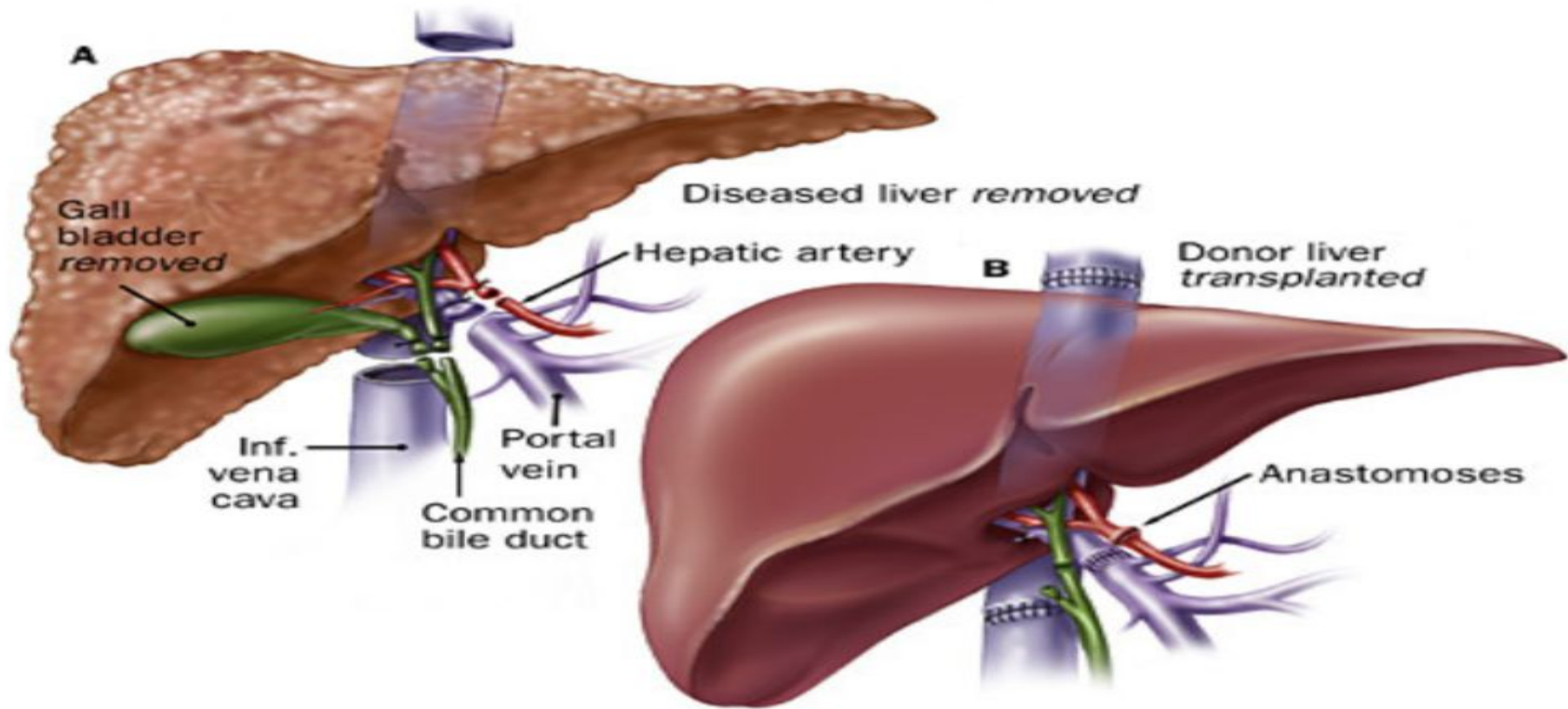
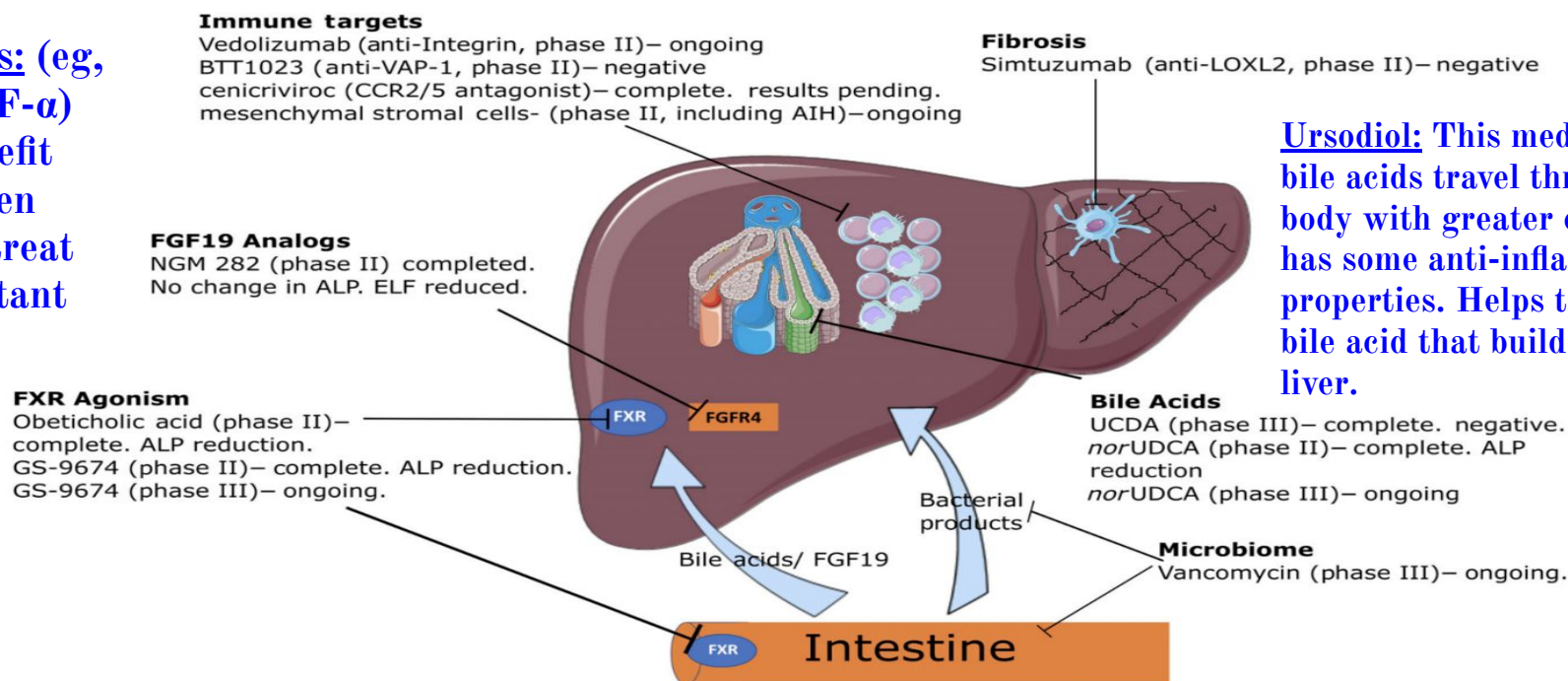


Figure 20. Liver transplantation.

Biologics: (eg, anti-TNF- α) may benefit PSC when used to treat concomitant IBD



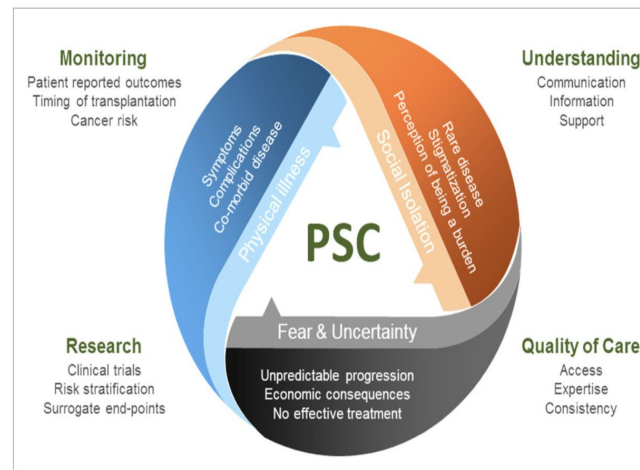
Ursodiol: This medicine helps bile acids travel through the body with greater ease. It also has some anti-inflammatory properties. Helps to fight toxic bile acid that build up in the liver.

FIGURE 3 Phase II and III clinical trials in primary sclerosing cholangitis. Abbreviations: CCR2/5, c-c chemokine receptor type 2/5; AIH, autoimmune hepatitis; ALP, alkaline phosphatase; ELF, enhanced liver fibrosis score; FGFR4, fibroblast growth factor receptor 4; LOXL2, lysyl oxidase homolog 2; norUDCA, norursodeoxycholic acid; UDCA, ursodeoxycholic acid; VAP, vascular adhesion protein. Figure adapted using graphics from ServierMedical Art.

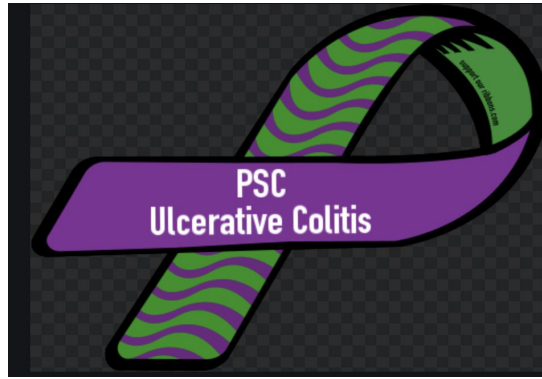
This a graph showing Phase 2 and Phase 3 clinical trials of certain drugs to treat PSC.

Nursing Implications and Considerations for patients with PSC

- Stress Management
- Diet/Nutrition
- Medication for symptom management
- Treatment: ERCP and Surgical Management
- Financial Burden of Disease



Conclusion:



- Psc is a rare, chronic, slowly progressive autoimmune liver disease that affects mainly young and middle-aged men, commonly associated with inflammatory bowel disease.
- No known medical treatment to halt the progression of disease is available; patients may benefit from endoscopic therapy and liver transplantation when appropriate.
- PSC is associated with a higher lifetime risk of hepatobiliary and colorectal malignancies.