PSC

-Chronic cholestatic liver disase >6 months
-Male > female
-Associated with IBD both (UC > crohns in colon)
-All PSC patient > colonoscopy done for them
-Leading to biliary cirhosis and portal HTN

Presentation :
Usually Asymptomatic
If symptomatic ; fatigue and pruritis , jaundice
recurrent cholingitis > think of PSC

Diagnosis :
Hx , elevation of Alk phos by 2-3 fold for more than 6 months
Mild elevated AST , ALT
MRCP : beading appearance
ERCP
Biopsy ; onion skin fibrosis
+Antibodies : P-ANCA , ANA (but not diagnostic)

Small PSC
Normal MRCP but abnormal histology , only 5% of cases

Complications :
ADEK diff (40% A) , steatorhea , celiec , chronic pancreatitis, osteoporosis

Treatment :
No medical treatment
Dilatation or stenting of the strictures
Treat complications
Liver transplant if (portal HTN complications, or recurrent cholingitis)

No response to steroids

Risk of malignancy :
Cholngiocarcinoma , colorectal , liver and pancreatic cancer

PBC

-Chronic cholestatic liver disases > 6months
-Middle aged Female
-Associated with autoimmune diseases :thyroid, rayanoid , sicca syndrome , scleroderma
-Has genetic predisposition
-Associated with smoking

Presentation :
Asymptomatic - 40-50%
Fatigue (most common) , pruritus , xanthoma , xhantholesma (without risk of CAD) , sicca syndrome (dry eyes , dry mouth) , portal HTN , metabolic bone diseases (osteopenia 50% , osteoporosis)

Diagnosis :
Elevated Alk phos 3-4 times
Mild elevated ALT ,AST ,
Elevated biliribin : late
High cholestrol
High IgM
+ AMA , ANA (diagnostic)
Biopsy : florid duct sign
MRCP : normal

Treatment :
Medical treatment : **UDCA** (first line) , OCA , fibrates

No response to steroids

-Low risk of cholngiocarcinoma

Barone Rocks	Primary Biliary Cirrhosis PBC	Primary Sclerosing Cholangitis PSC
Clinical	• Females > Males • Middle age • Fatigue & pruritis • Cholestatic Labs	• Males > Females • 20-40's • Progressive obstructive jaundice • Cholestatic Labs
Site of Involvement	Intrahepatic	Intrahepatic & Extrahepatic
Cause of Obstruction	Granulomatous inflammation destroying bile ducts	Fibrosis destroying bile ducts
Key Microscopic Feature	Florid duct lesion (granulomas)	Concentric "onion-skin" fibrosis around bile ducts
Diagnostic clue	Anti-mitochondrial antibodies (AMA) - Antibodies against the subunit of pyruvate dehydrogenase complex	Beaded appearance of bile ducts on cholangiogram/ERCP/MRCP Baronerocks.com
Association	Other autoimmune disorders Sjögrens, RA, etc.	Ulcerative colitis
Long-term Complication	Cirrhosis	Cirrhosis Cholangiocarcinoma

	Primary Biliary Cirrhosis/Cholangitis:	Primary Sclerosing Cholangitis:
Definition	Ongoing immunologic attack on the intralobular bile ducts that eventually leads to cirrhosis and liver failure	a progressive disease characterized by inflammatory strictures involving the intrahepatic and extrahepatic biliary tree.
MC Symptoms	Pruritis, jaundice Sicca Syndrome	Fever, chills Fatigue and pruritis
Association with IBD	No association with IBD	Inflammatory bowel disease, such as Crohn disease or ulcerative colitis, is identified in about 75% of patients with PSC.
Autoantibodies	Anti-mitochondrial antibodies are the hallmark of PBC	p-ANCA
Male vs Female	Female predominance	Male predominance
Histopathology	Scarring of intrahepatic bile ducts; nonsuppurative cholangitis and destruction of small- or medium-sized bile duct Florid duct lesion	Fibrosis around bile ducts within portal tracts (onion-skin pattern)
Symptoms on diagnosis	Most patients are asymptomatic when diagnosed	Most patients are symptomatic when diagnosed
MRCP findings	No beads on a string appearance on MRCP	MRCP revealing multiple dilatations and strictures (beads-on-a-string) of the intra- and extrahepatic biliary tree
Risk of Cholangiocarcinoma	low risk of cholangiocarcinoma	High risk of cholangiocarcinoma
Response to UDCA	Good response to ursodeoxycholic acid	Poor response to ursodeoxycholic acid

Autoimmune hepatitis

-Intermittently progressive inflammatory liver disease
-With absence of viral hepatitis or alcohol consumption 🍷
-mainly in Middle aged female
-Associated with other autoimmune diseases

Presentation : very similar to viral hepatitis
Fatigue , Artharligia , myalgia, oligomenorrhea , jaundice , hepatosplenomegaly

Diagnosis :
Elevated ALT , AST (in thounsands)
High IgG
High SMA , ANA , p ANCA in type 1 // LKM1 in type 2
Liver biposy :

- Interface hepatitis with lymphoplasmacytic infiltrate
- Rosetting of liver cells
- Bridging necrosis
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Scoring system : Autoantibodies, histology , IgG , abssence of viral hepatitis

Treatment :
-Monotherapy : prednisone (60 mg initial dose)
In : Cytopnea , TMPT diff , malignancy , pregnancy

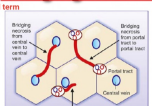
-prednisone is taken as a maintenance therapy

-Combined therapy : prednisone 30 mg w/Azathioprine 50 mg (initial dose)
In : Osteoporosis , HTN , DM , obesity, ance

-Liver transplantation : but there is high recurrence rate

Indications for Treatment

Absolute	Relative	None
AST ≥ 10x normal	Symptoms	No symptoms
AST ≥ 5x normal and γ-globulin ≥ 2x normal	AST < 5x normal γ-globulin < 2x normal	Inactive cirrhosis (No gain from Tx)
Bridging necrosis (histological term)	Interface hepatitis	Portal hepatitis



Sub-Types of Autoimmune Hepatitis

	Type 1 (More common (80%))	Type 2
Age at Presentation	Any age (more with adults)	Predominantly children
Female:Male (both are female-predominant)	4:1	8:1 (more associated with females)
Ig G Levels	Elevated IgG	Variable Ig G
Ig A Levels	Normal	+/- Low IgA
Auto-antibodies	ANA, SMA, PANCA	LKM-1, SLA/LP
Cirrhosis at 3 yrs	~ 40%	~ 80% (worse prognosis)