

► Primary Sclerosing Cholangitis [Young ♂ with UC]

- Cholestatic disease $\Rightarrow \uparrow ALP + GGT$
- Frequently associated with IBD, mostly UC
- Inflammation & fibrosis of biliary tree, leads to biliary cirrhosis & PHTN
- Etiology \gg T-cell abnormal function / infection / portal bacteremia
- Clinical picture \gg ↑ALP, in setting of colitis, may be asymptomatic.

confirm diagnosis \rightarrow ERCP or MRCP = Abnormal cholangiogram

- Clinical presentation \gg

Fever
jaundice \rightarrow Charcot Triad
AVP pain

	15 - 44%
Symptomatic	
Fatigue	75
Pruritus	70
Jaundice	30-69
Hepatomegaly	34-62
Abdominal pain	16-37
Weight loss	10-34
Splenomegaly	30
Multiple Ascending cholangitis	5-28
Hyperpigmentation	25
Variceal bleeding	2-14
Ascites	2-10

* IBD 60-80% of PSC (2:1) UC > Crohn

* 4-5% of UC have PSC.



- Diagnostic criteria \gg History of IBD + Cholestatic symptoms + $\uparrow ALP$ (2-3 folds) > 6 months

(mostly used)
Non-invasive
- **MRCP** \leftarrow radiation cost-effective

\leftarrow ERCP (invasive, risk of pancreatitis) \leftarrow stent balloon biopsy

- Liver tests: Always elevated: ALP / AST & ALT \uparrow / Bilirubin, albumin, PTT (normal)

\downarrow
when liver cirrhosis \Rightarrow impairment

- AutoAb: \uparrow p.AUCA (80%) note [UC = ANCA +ve, Crohn's = ASKA +ve]

\uparrow ANA (not specific)

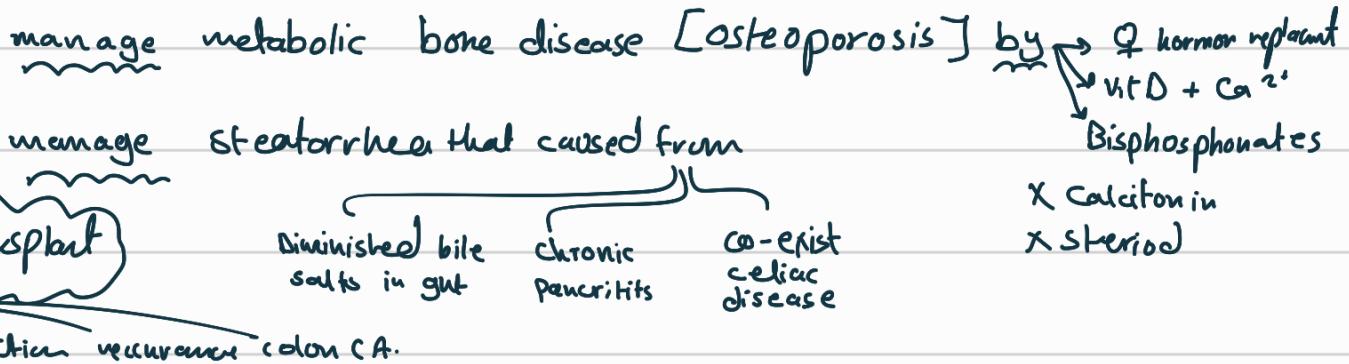
- Histology: onion skin like appearance [periductal fibrosis]

5% of PSC, can progress to PSC

∴ Small duct PSC $\left\{ \begin{array}{l} \text{Normal cholangiogram, Biopsy +ve} \\ \text{many or not with UC} \end{array} \right.$

• Therapy: Dilation of stricture, IV antibiotics, manage vit. Def. (AKED)

imp.
↓



• Cancer risk: CholangioCA / Hepatocellular CA / Colorectal CA / Pancreatic CA.

Must be screened yearly to rule out colon CA.

► Primary Biliary Cirrhosis [AMA↑, IgM↑, Middle age ♀]

- Cholestatic liver disease confirm diagnosis → Biopsy by ERCP → **Florid duct sign**.

Biochemical: ↑ ALP 3x4 folds / ↑ AST, ALT / Bilirubin rise late / ↑ cholesterol / ↑ IgM

- Associated with autoimmune thyroid disease / Sicca syndrome / smoking

• Presentation:

use Cholestyramine to treat

	Asymptomatic	~50%
Symptomatic		
Fatigue (most common)	+++	
Pruritus	++	
Sicca symptoms	+++ Dry mouth, Dry eyes mucus membranes dryness including vagina	
Hepatomegaly	+	
Splenomegaly	+	
Jaundice	uncommon	
Xanthelasma	uncommon	

Extrahepatic Autoimmune Diseases

	(%)
① Sicca syndrome	70 ✓
② Thyroid disease	40 ✓
③ Arthritis	20
④ Scleroderma	15
⑤ Raynaud's phenomenon	10
⑥ CREST syndrome	5

Sicca syndrome [Keratoconjunctivitis + xerostomia] needs ↑ fluid intake, oral sialogogues, artificial tears, vaginal lubricants

Metabolic bone disease [osteopenia, osteoporosis - osteomalacia] → treat same as PSC

Portal HTN, most common in cirrhotic.

⇒ UDCA: decrease liver damage (the only useful drug)

► Autoimmune hepatitis

- Progressive inflammatory disease, ↑ IgG ↑ ALT-AST, +ve ANA-SMA

- peri-portal hepatitis ✓ Respond to corticosteroids

- Middle age, ♀, non drinker, no viral hepatitis

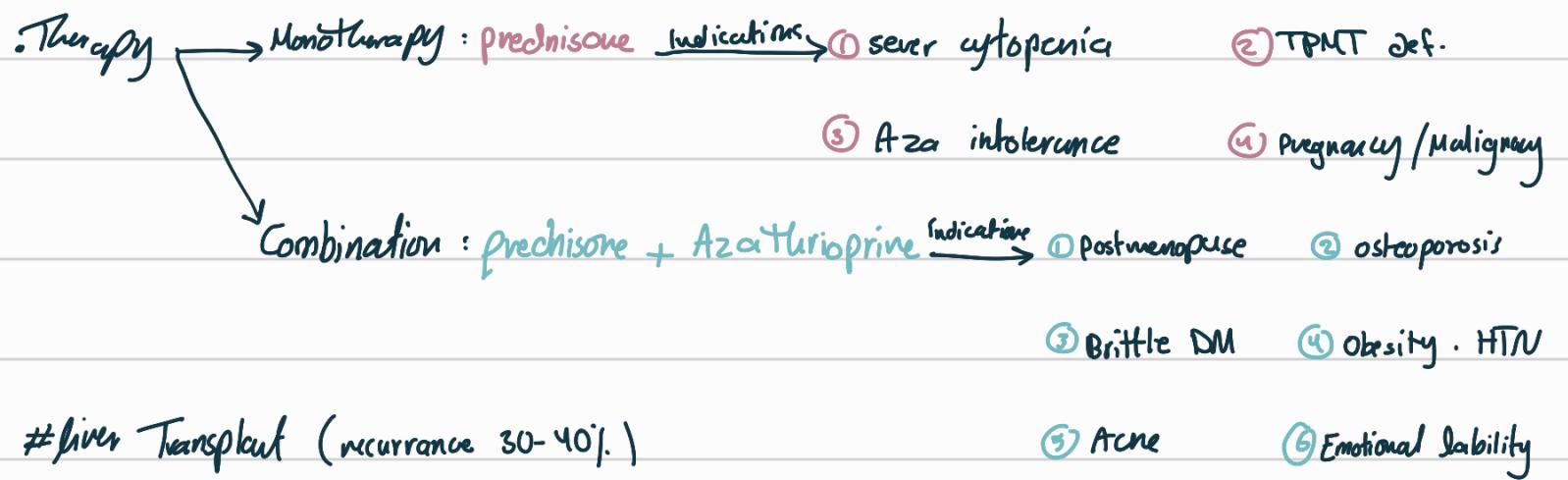
• Clinical feature → fatigue / antinflammatories / oligomenorrhea / jaundice

• Histo → Interface hepatitis with lymphoplasmacytic infiltrate

Type 1	Type 2
Age at Presentation More common (80%)	Any age more with adults
Female:Male both are female-predominant	4:1
Ig G Levels	Elevated IgG
Ig A Levels	Normal
Auto-antibodies	ANA, SMA
Cirrhosis at 3 yrs	~ 40%
	~ 80% worse prognosis

Autoimmune hepatitis Type 1 → +ve ANA, ASMA
Type 2 → +ve LKM-1, SLA/LP

Indications for treatment AST ≥ 10x normal
AST ≥ 5x & γ globulin ≥ 2x
Bridging necrosis



Liver Transplant (recurrence 30-40%)

⑤ Acne ⑩ Emotional lability

Sunny :-

	Primary sclerosing cholangitis	Primary biliary cholangitis
Epidemiology	<ul style="list-style-type: none"> More common among middle-aged men 	<ul style="list-style-type: none"> More common among middle-aged women
Pathophysiology	<ul style="list-style-type: none"> Progressive chronic inflammation of both intrahepatic and extrahepatic bile ducts 	<ul style="list-style-type: none"> Progressive destruction of only intrahepatic small and medium-sized bile ducts
Clinical presentation	<ul style="list-style-type: none"> Pruritus Fatigue Jaundice Hepatomegaly 	<ul style="list-style-type: none"> Similar to PSC Potentially xanthomas and xanthelasma
Laboratory tests	<ul style="list-style-type: none"> pANCA ↑ ALP, GGT, and conjugated bilirubin 	<ul style="list-style-type: none"> Anti-mitochondrial antibodies (AMA) ↑ ALP, GGT, and conjugated bilirubin
Associated conditions	<ul style="list-style-type: none"> Ulcerative colitis and cholangiocarcinoma 	<ul style="list-style-type: none"> Autoimmune conditions

Differentiating PSC from PBC

	PSC	PBC
Cholestasis	+	+
History of colitis	+	-
AMA	-	+
Liver biopsy	onion skin fibrosis	florid duct lesion
Cholangiogram	abnormal	normal

↓

Ig M

Autoimmune hepatitis → IgG

, Respond to corticosteroids.