Managing PSC symptoms

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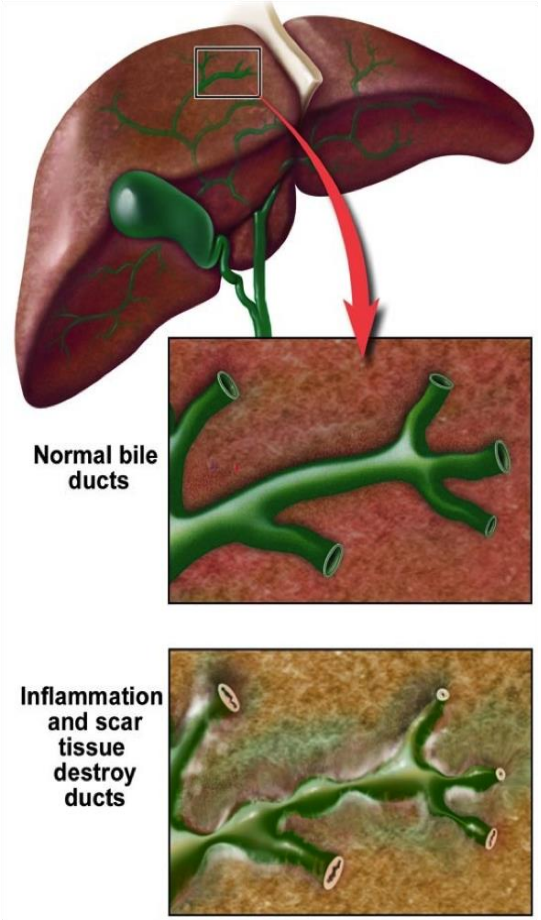
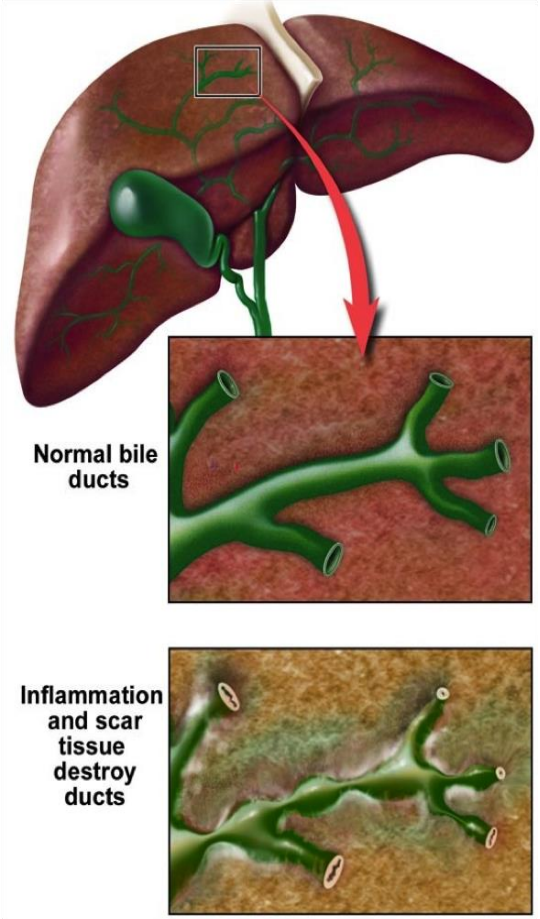
PSC Facts



- Chronic cholestatic liver disorder characterised by inflammation & fibrosis of intra/extra hepatic bile ducts
- Leads to progressive bile duct obliteration, liver fibrosis and biliary cirrhosis
- Complex disease of unknown etiology
- Incidence ≈ 0.9-1.3/100,000/yr;
  Prevalence ≈ 8.5-14.2/100,000
- Male : Female = 2 : 1
- > 90% of patients are non-smokers

PSC Facts



- 60-80% have co-existing IBD; ≈ 25% have auto-immune disease
- Diagnosed on the basis of cholangiography (ERCP/PTC/MRCP) and/or histology
- Variable/unpredictable disease course
- Main risk from progression to biliary cirrhosis
- Mortality due to liver failure and CCA
- 5th commonest indication for OLT

IBD in PSC – unique phenotype

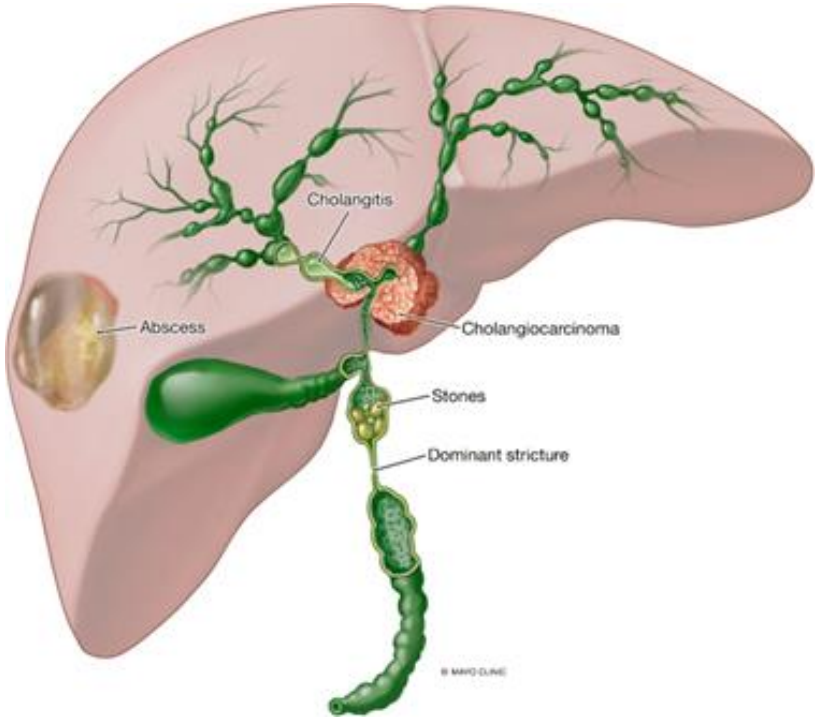
IBD phenotype in PSC
Pan-colitis with predominance of R sided activity
Rectal sparing; Backwash ileitis
Mild or Quiescent course
Increased risk of: - CRC ($2/3^{rd}$ R sided) - Increased risk of pouchitis in patients with IPAA - Increased risk of peri-stomal varices

Chapman et al, Hepatology 2010

Symptom burden in PSC

- Cholangitis
- Itch (pruritus)
- Jaundice
- Fatigue
- Fragility fractures
- Pain
- Impact on mental health

Cholangitis



- Defining cholangitis
- clinical suspicion
- blood test
- prompt antibiotics

- Imaging
- Role of biliary intervention (ERCP)

- recurrent cholangitis
- cyclical antibiotics

Fig: www.aasld.org/liver-fellow-network/core-series

Itch (Pruritus)

- Common but debilitating symptom
- Effect on QOL
- Common to all forms of bile duct disease (bile salt retention)

- General treatment:
  - emollient
  - cold shower
  - cooling menthol gels

Itch (Pruritus)

- **Medications:**
  - Colestyramine (bile salt sequestrant) – 4gm as needed
  - Rifampicin 150 – 300 mg daily
  - Naltrexone 12.5 – 50 mg daily
  - Bezafibrate 200mg twice daily
  - Sertraline 25 – 75 mg daily

Jaundice

- Obstructive jaundice – pale/clay stools; dark urine; eye discoloration
- Worsening itch
- Blood test and imaging
- Differentiate cholangitis vs tight bile duct narrowing vs bile duct stones

Fatigue

- Poorly understood mechanisms
- Treat contributing factors:
  - e.g. anaemia, thyroid, vitamin deficiencies
  - active colitis
- Central vs peripheral fatigue
- Graded exercise therapy

Reducing risk of fragility fractures

- Prevalence:
  - Osteoporosis ~ 11-15%
  - Osteopenia ~ 40%
- Risk factors: old age, low BMI, prolonged steroid use, presence of IBD
- Not dependent on presence of cirrhosis
- ↑ risk of fractures (especially post transplant)
- Improve awareness
- Vitamin D replacement if low +/- bone protection treatment if needed

Pain

- Abdominal pain:
  - IBD
  - cholangitis
  - biliary colic due to gall stones or bile duct stones
- Joint pains – IBD; autoimmune disease (e.g. rheumatoid arthritis, Sjogren's syndrome, psoriatic arthropathy)
- Generalised aches/pain –
- Bone pain

Pain

- Treatment tailored to underlying cause :
  - treat active inflammation (e.g. colitis/arthritis/cholangitis)
  - Buscopan
  - simple analgesia (paracetamol is safe; reduce dose if cirrhosis present)
  - Avoid NSAIDs (e.g. Ibuprofen, Diclofenac)
  - Avoid opioids (e.g. morphine, tramadol) if possible (especially in cirrhosis);
    - risk of itch, biliary dyskinesia and constipation; encephalopathy in those with cirrhosis

Impact on mental health

- Mental health impact: depression, anxiety, prognostic uncertainty, travel, insurance

  - support groups

- Anxiety/Uncertainty re: progression/prognosis

  – relationship with hospital team/GP and accessibility

Symptoms/complications related to cirrhosis

- Fluid retention – ascites, leg swelling
- Gastrointestinal bleeding
- Hepatic encephalopathy
- Jaundice

- Consideration for liver transplant

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