IBD in PSC

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Objectives

- Understand current knowledge about the bowel disease associated with PSC
- Understand the relationship between IBD in PSC and colon cancer
- Understand current approaches to the medical and surgical treatment of IBD in PSC

IBD in PSC Fundamental Questions

 Is PSC an uncommon extra-intestinal manifestation of colitis (2-4%) ... or

 Is a colitis that is a lot like ulcerative colitis and a little like Crohn's colitis a very common extra-hepatobiliary manifestation of PSC (75%) ?

What Are the Features of the IBD Associated with PSC ?

- A lot like CUC
 - Confluent disease 'only' involving the colon
 No granulomas, + crypt abscesses
 - No fistulae or fissures
 - -+ pANCA in 80%; ASCA negative
- A little like Crohn's
 - Rectal sparing common (50% vs 5% in CUC)
 - Backwash Ileitis common (50% vs 5% in CUC)

What Are the Features of the IBD Associated with PSC?

- The bowel disease is:
 - Clinically mild
 - Responds to the same medicines as CUC (at least pre-liver transplant)
 - Steroids, colon specific 5-ASA compounds
 - Uncertain about immunomodulators
 - Is at greater risk (probably) of developing dysplasia and colon cancer

Dysplasia and Colon Cancer

- Most agree that the patient with PSC and IBD has an increased risk of developing colon cancer ... over and above the .5% per year in patients with CUC >8-10 years.
- In one study the risk was increased 4 fold.
- A possible explanation is the secondary bile acids at increased concentrations in the colon of the patient with PSC.

Dysplasia and Colon Cancer What to Do About It?

- Primary Prevention (All evidence is epidemiologic not prospective trials)
 - UDCA
 - 5-ASA compounds (Decreased inflammation?)
 - Folic acid (1 mg per day)
- Secondary Prevention
 - Surveillance colonoscopy (done properly) every year after having colitis > 8 years
 - Colectomy for documented dysplasia

Colectomy in the PSC Patient

- Total colectomy with ileostomy or ileoanal pull through with J pouch
 - Ileostomy Requires ostomy appliance, simple, lower risk. Risk of varices at the ostomy site in patients with cirrhosis.
 - Ileoanal pull through No ostomy or appliance. Good result is 5-6 continent BMs a day. However, inflammation of the pouch is increased in patients with PSC

Pouchitis in the Patient with PSC

- Pouchitis inflammation with ulcers of the J pouch that causes increased frequency of BMs, urgency, and incontinence.
- Cause unknown but likely a combination of the bacterial flora in the pouch and the immunologic set up of the patient.
- Over 75% of patients with PSC get pouchitis (as opposed to 30% CUC)

Pouchitis in the Patient with PSC

- Diagnosis requires endoscopic evaluation of the pouch with biopsies
- Treatment
 - Antibiotics metronidazole and ciprofloxacin are the mainstays but others are tried with varying success
 - Immunosuppressant Rx not helpful
- Prevention
 - Probiotics after surgery or after first episode
 - VSL #3 best data