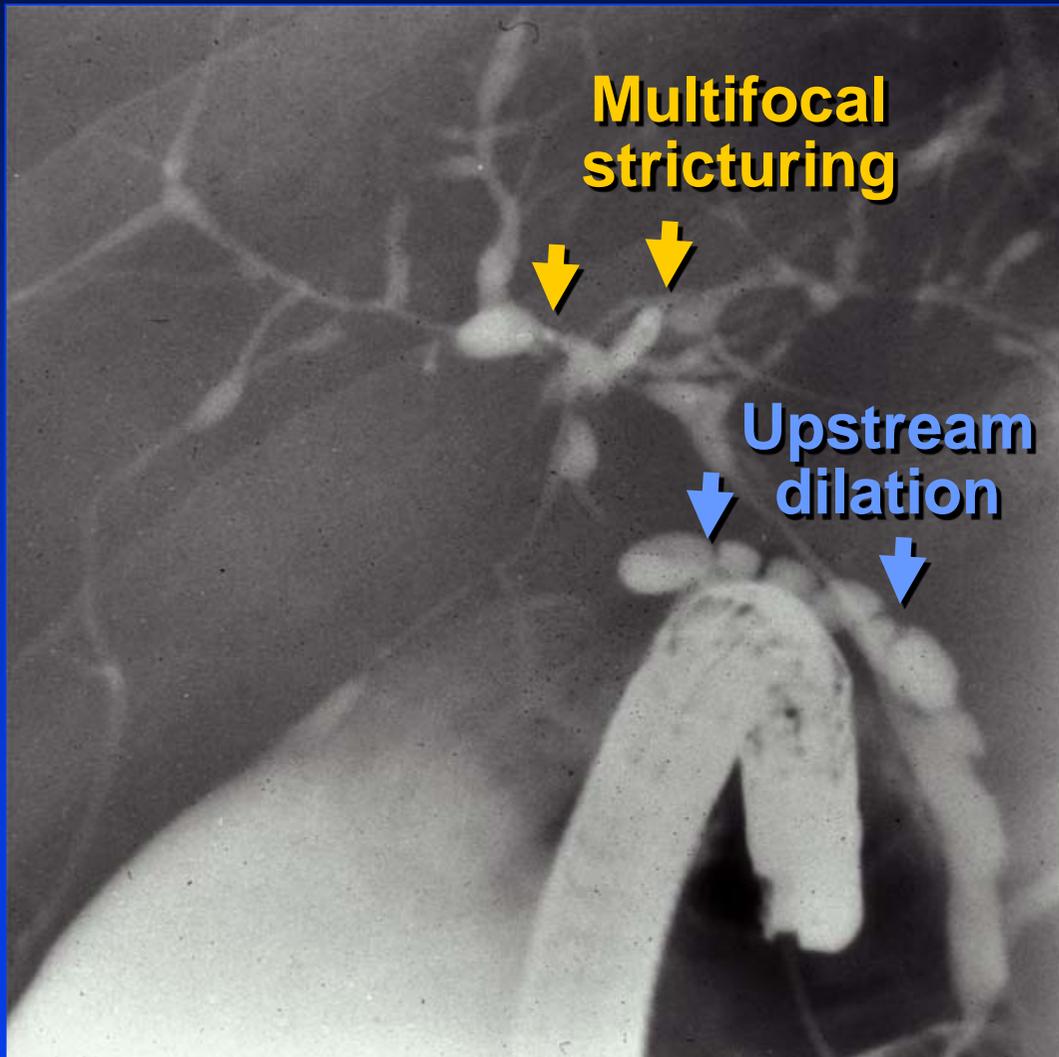


Overview of PSC

Jayant A. Talwalkar, MD, MPH
Associate Professor of Medicine
Mayo Clinic
Rochester, MN

2012 Annual Conference
PSC Partners Seeking a Cure
May 5, 2012

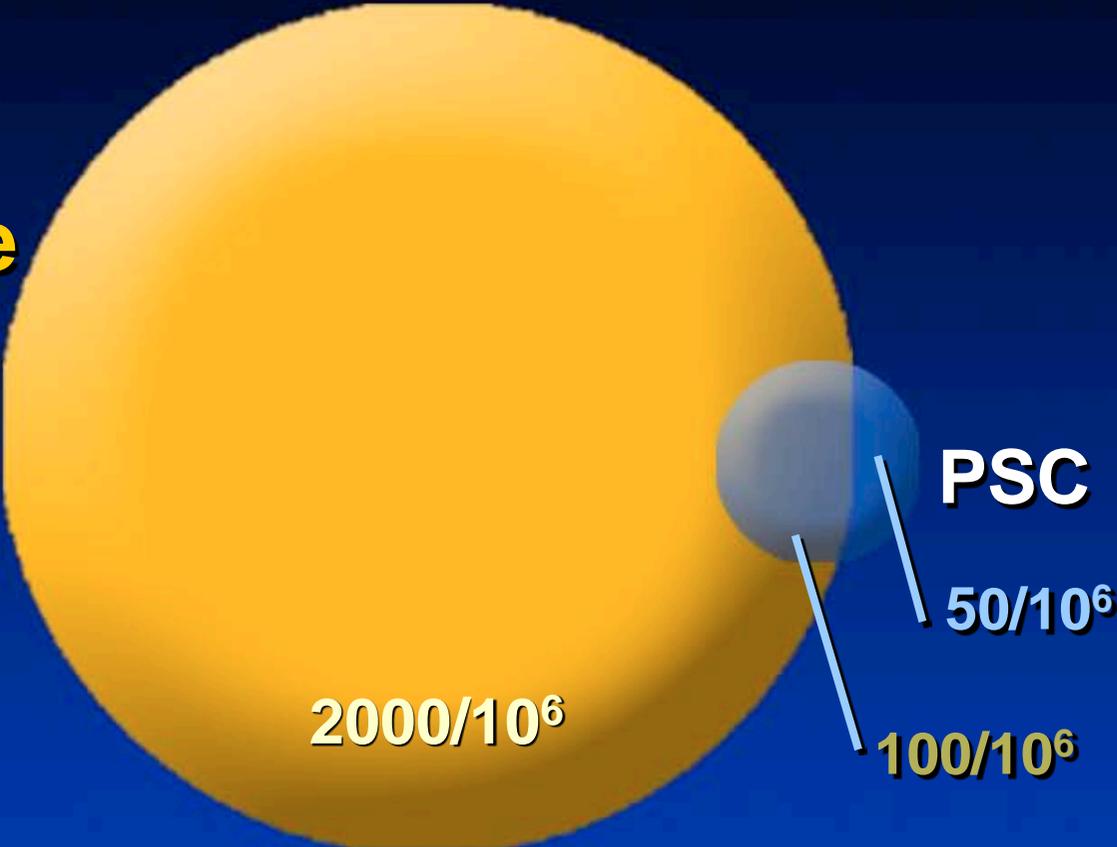
Primary Sclerosing Cholangitis



- Chronic cholestatic liver disease
- Unknown etiology, frequently associated with Inflammatory Bowel Disease
- Diffuse inflammation and fibrosis of the biliary tree
- Leads to biliary cirrhosis and portal hypertension

Primary Sclerosing Cholangitis in Colitis

**Chronic
ulcerative
colitis**



(Estimated prevalence)



Clinical Presentation

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

Serum Biochemical Tests

- Alkaline phosphatase nearly always elevated
- AST and ALT usually <5 times normal
- Bilirubin, albumin, prothrombin time usually normal at diagnosis



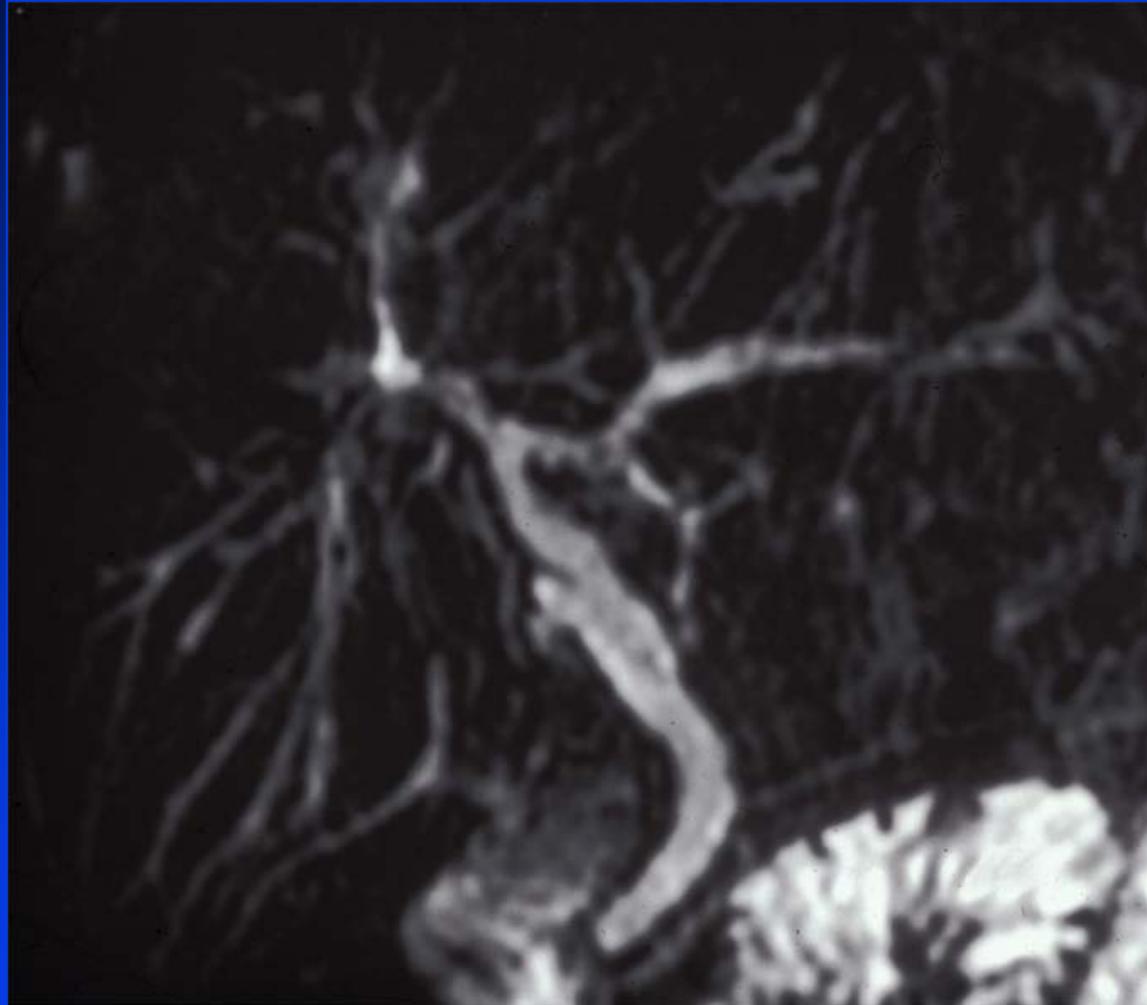
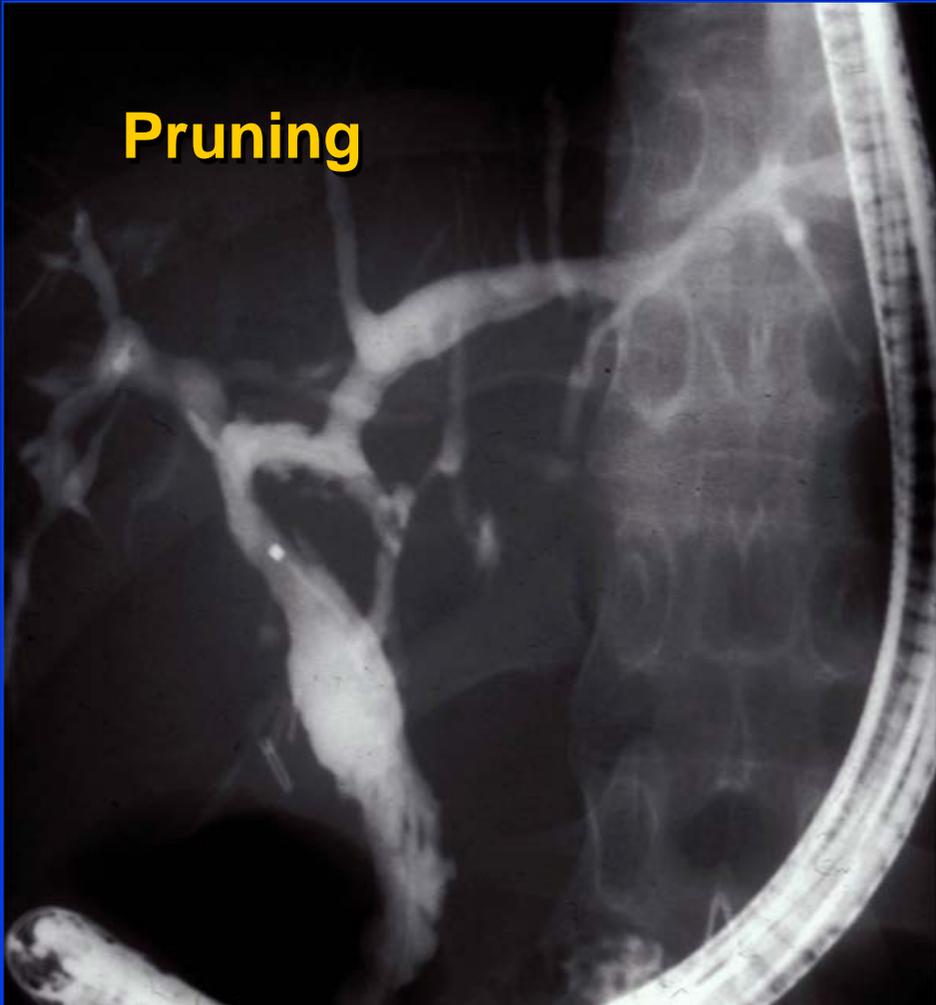
Diagnosis of PSC

Cholangiography

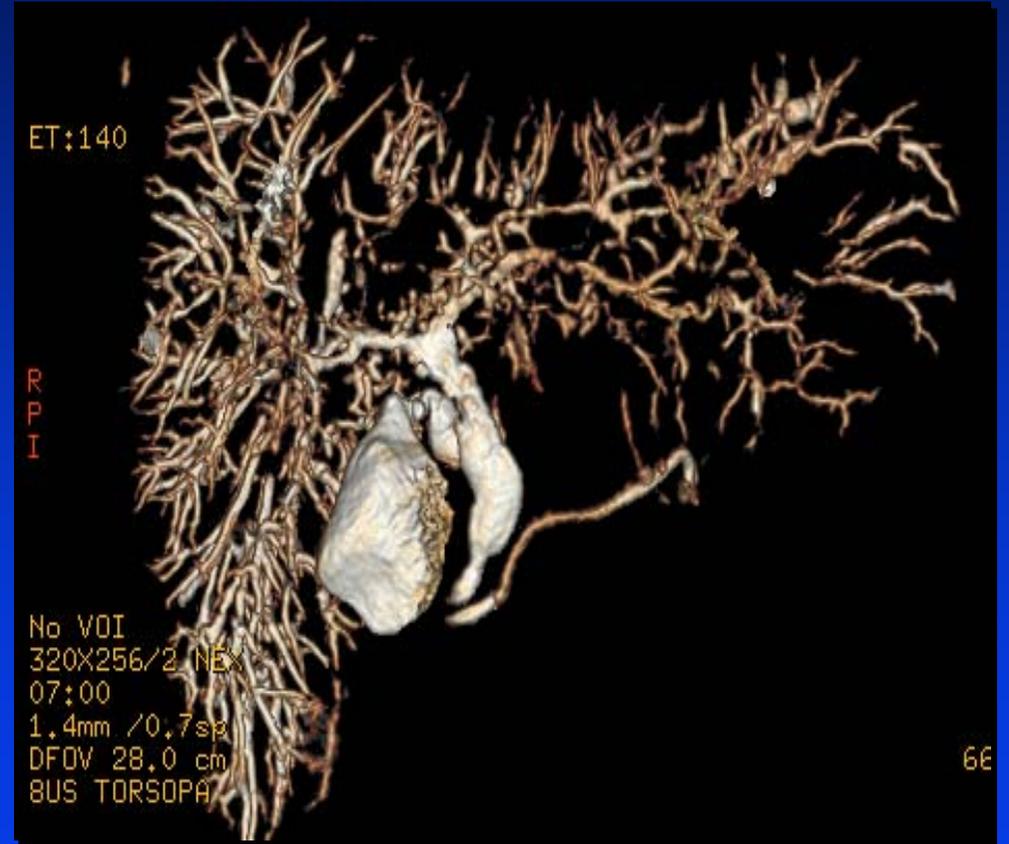
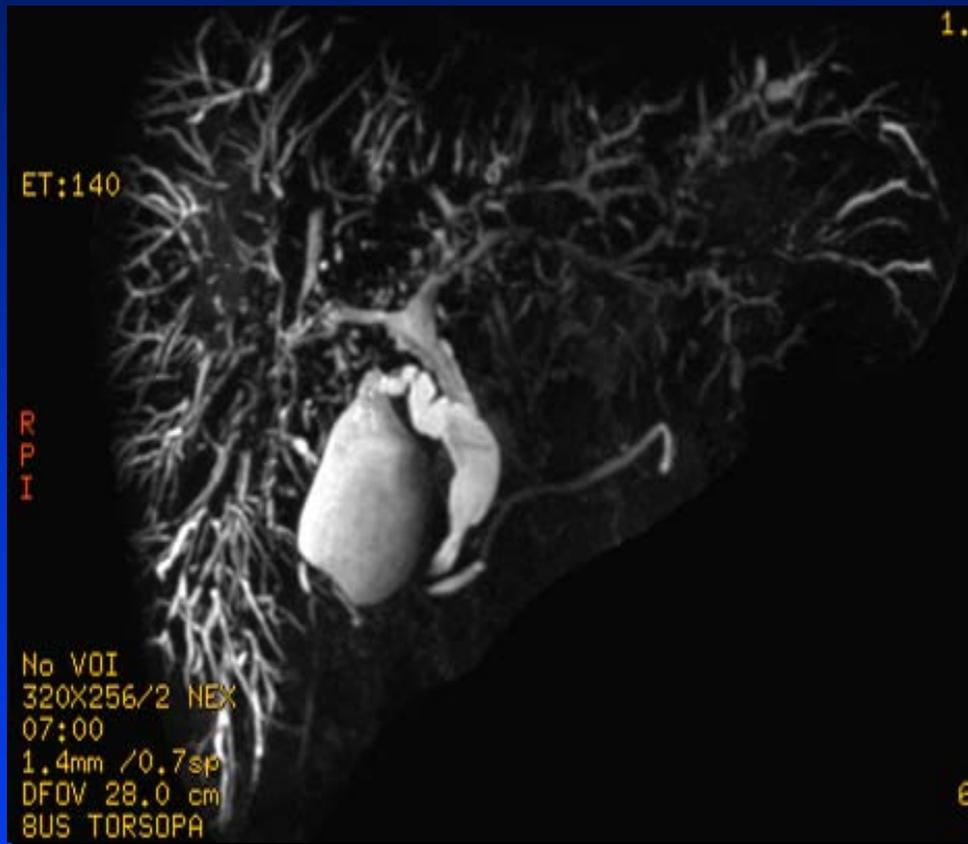
- ERCP commonly used in the past
- Percutaneous cholangiography infrequently used
- Magnetic resonance cholangiography
 - non-invasive
 - no radiation
 - cost-effective

Comparison of ERC and MRC in PSC

Pruning



Advances in MRCP and Image Acquisition for PSC



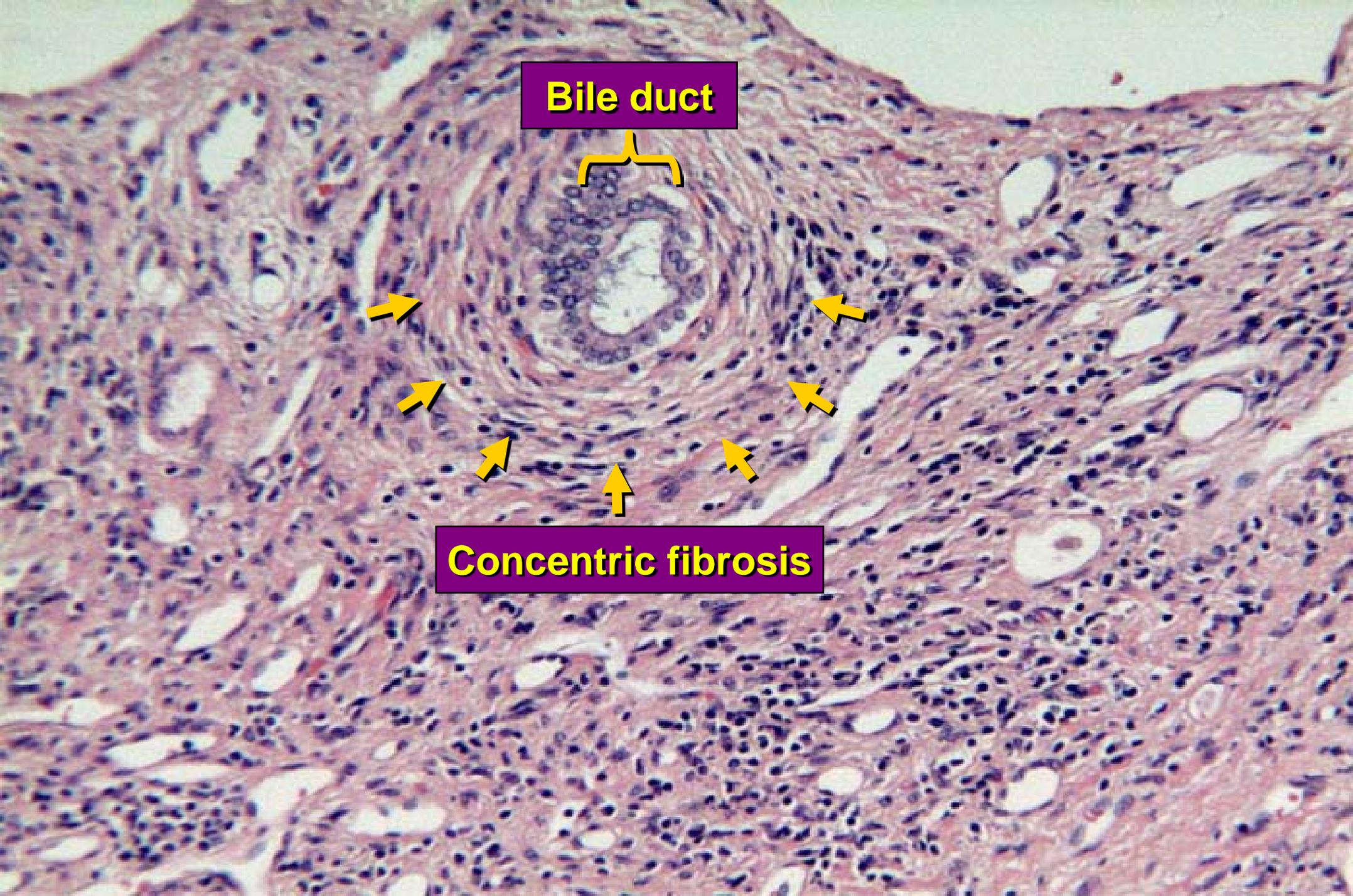
Role of Liver Biopsy in PSC

- **May help confirm diagnosis**
- **Can help exclude overlap with AIH**
- **Needed if cholangiogram normal – small duct PSC possible**
- **Can provide prognostic information**
- **May not be needed in all cases**

Bile duct



Concentric fibrosis



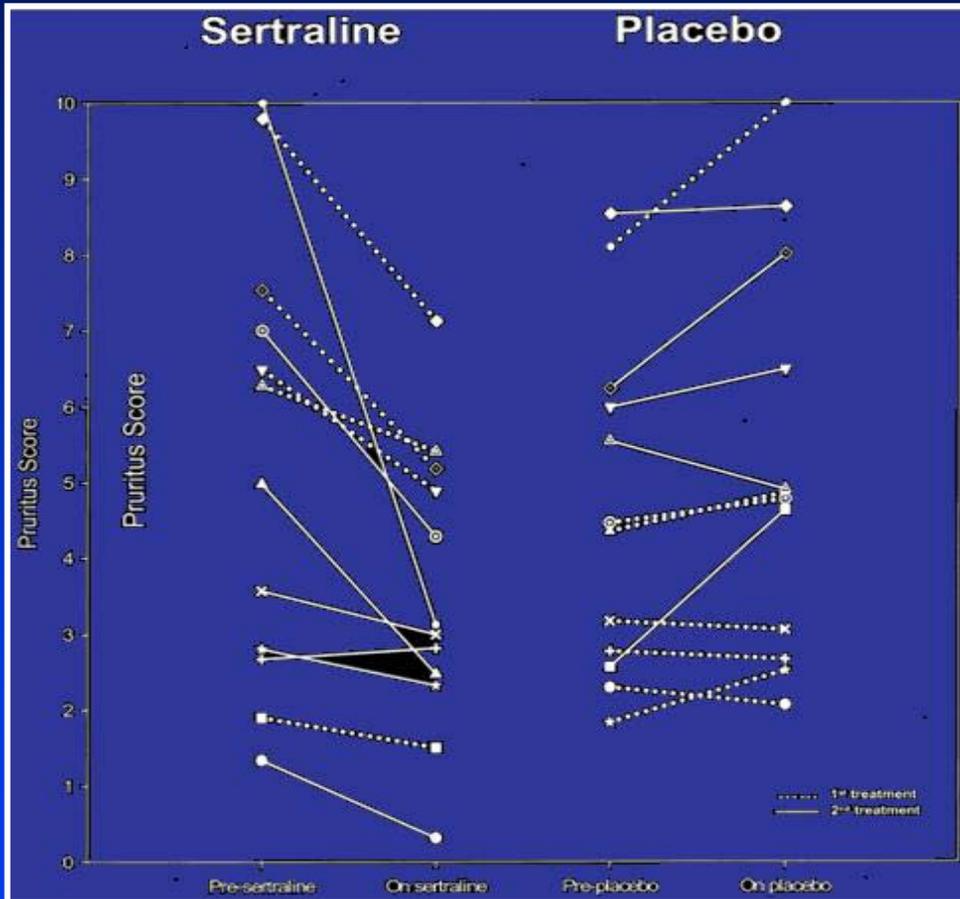
Fatigue in PSC

- **Common symptom**
- **Frequency 0 - 50%**
- **No association with age, sex, histological stage, bilirubin, and Mayo Risk score**
- **No identified effective therapy**
 - **SSRI, modafinil ?**

Pruritus in PSC

- **Frequency between 20-60% of cases**
- **May be intractable - ? dominant stricture**
- **No association with age, sex, histological stage, and Mayo Risk score**
- **Etiology unknown**
- **Treatment algorithm: cholestyramine, UDCA, sertraline, rifampin, naltrexone**

Sertraline for Pruritus in PSC



- Open-label dose escalation trial
- 12 patients in RCT after washout
- Itch scores better with active drug
- 75 to 100 mg well-tolerated

Management of Metabolic Bone Disease

Osteoporosis much more common than osteomalacia

- **Hormone replacement in women**
- **Calcium \pm vitamin D helpful**
- **Bisphosphonates may be helpful**
- **Steroid therapy may worsen bone disease**
- **Calcitonin not helpful**

Management of Biliary Stricture

- **Cholangitis, pruritus, pain**
- **Typically by ERCP vs. PTC**
- **Dilatation alone with response adequate for dominant strictures**
- **If stenting required, removal +/- replacement in 4-6 weeks preferred**
- **Post-procedures antibiotics**

Medical Therapy Tested to Date

Penicillamine

Colchicine

MMF

Cyclosporine

Methotrexate

Silymarin

Pentoxifylline

Budesonide

Tacrolimus

Nicotine

Pirfenidone

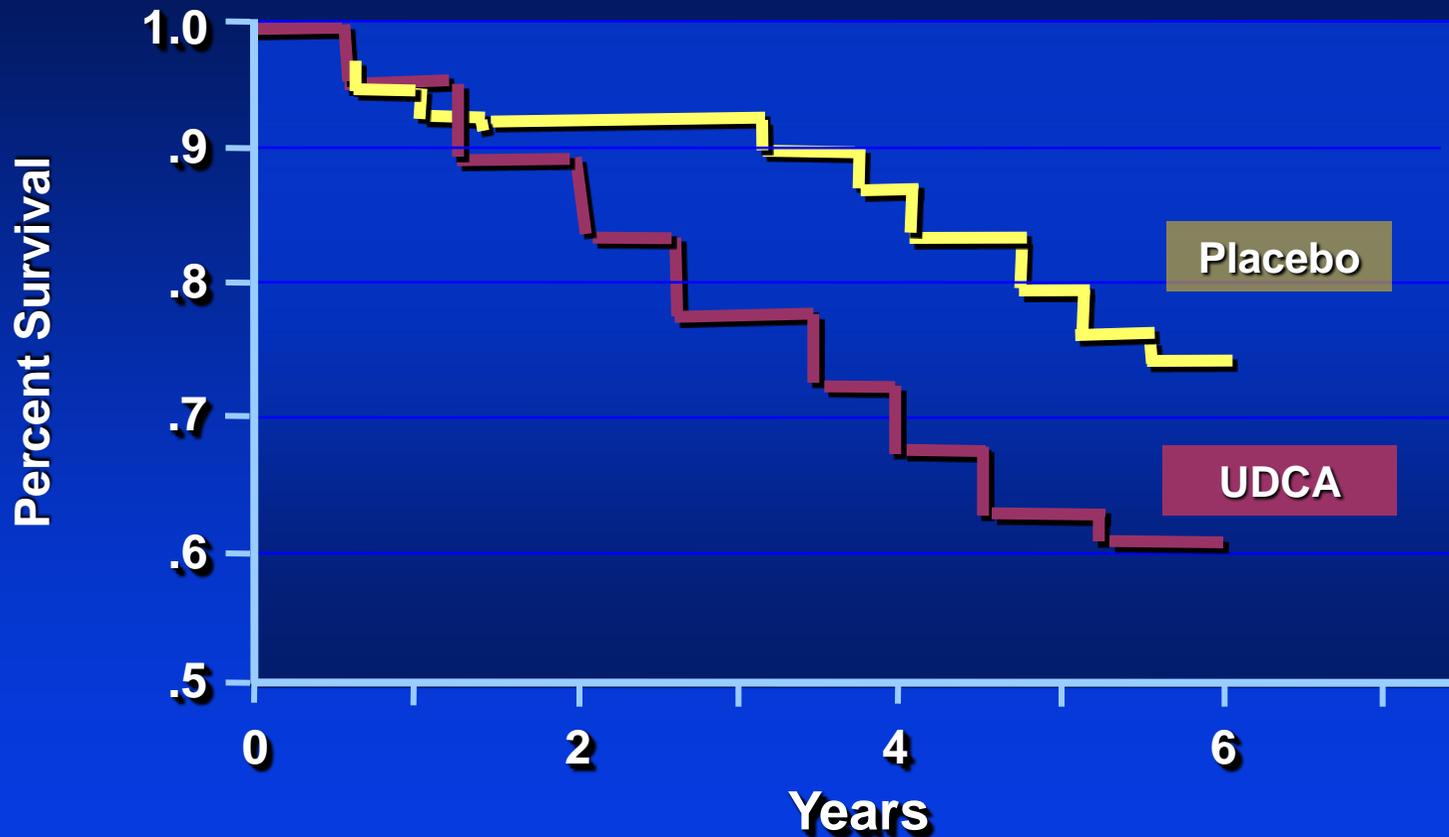
UDCA

Azathioprine

Etanercept

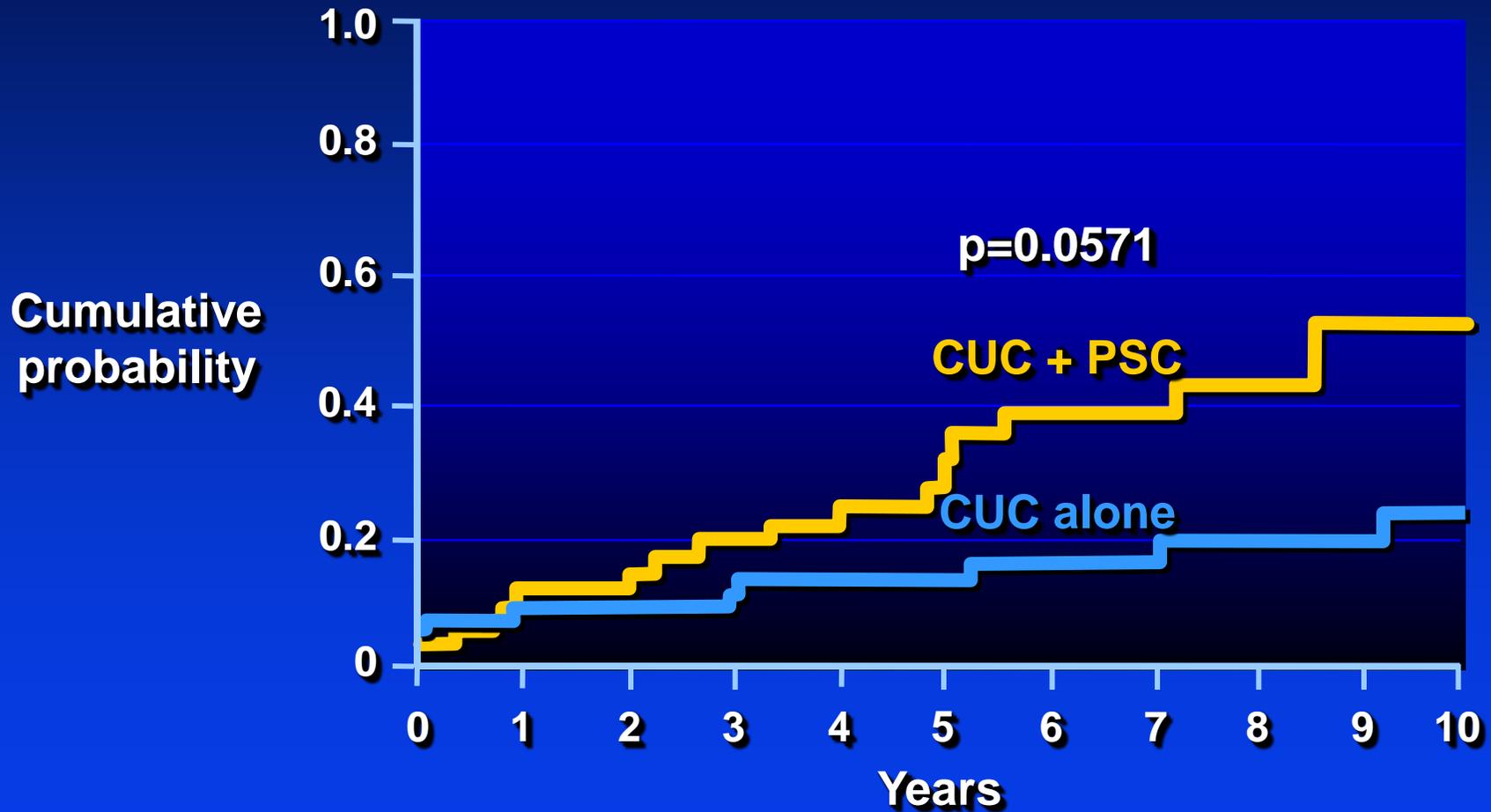
High-Dose UDCA for PSC

United States/NIH

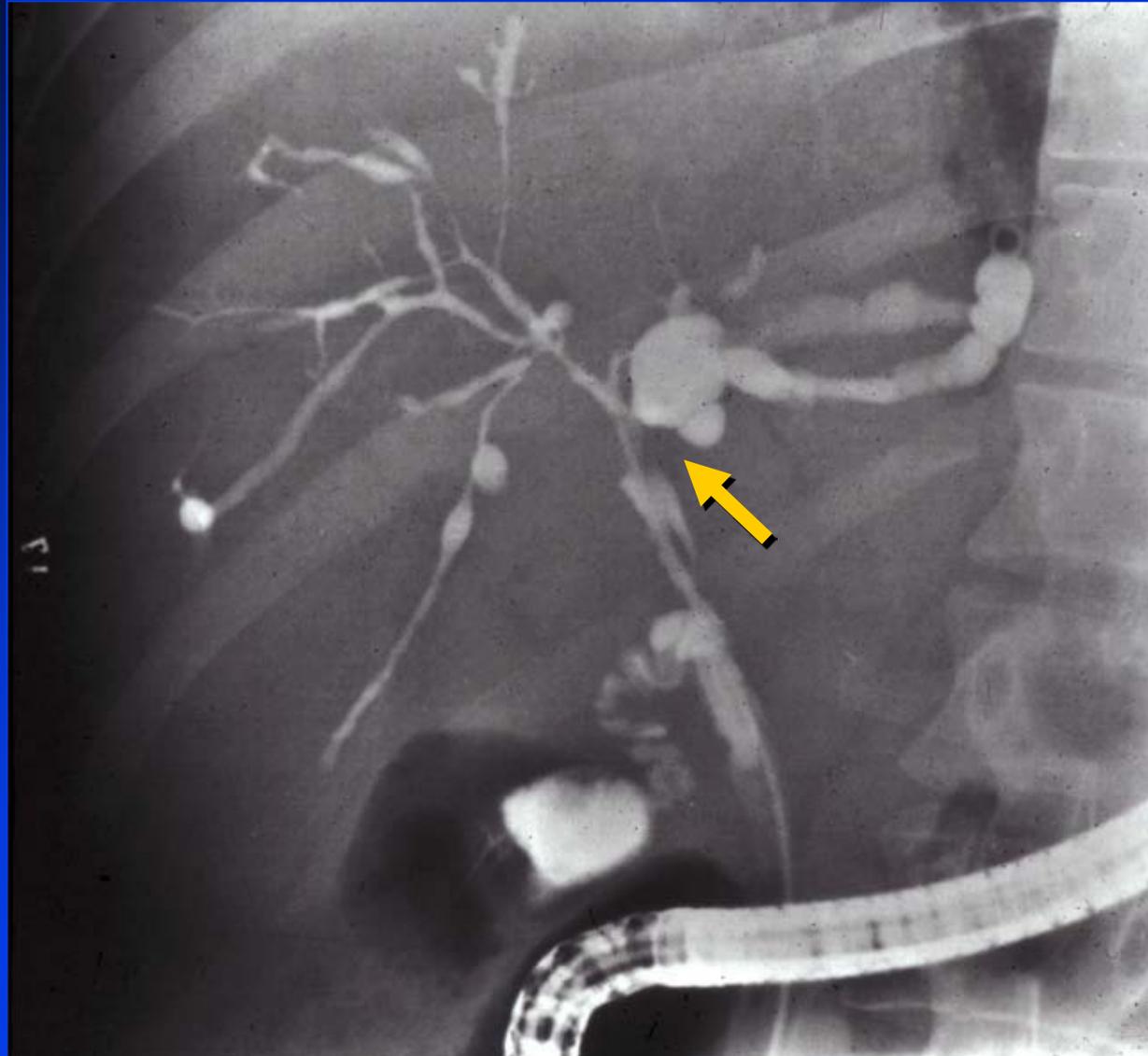


* UDCA (28-30 mg/kg/day)

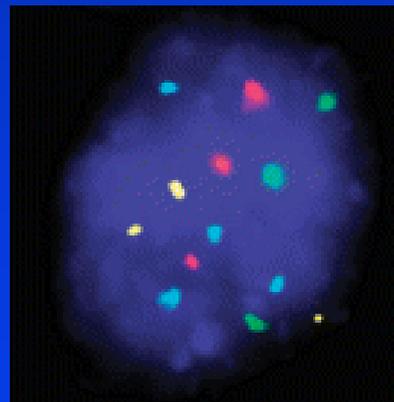
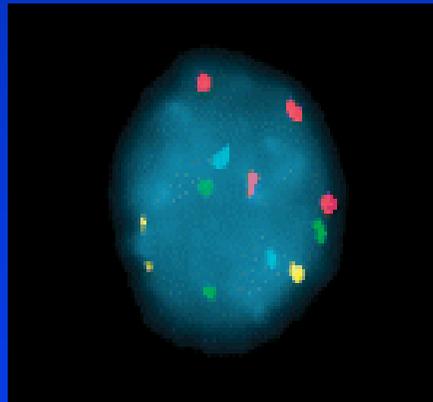
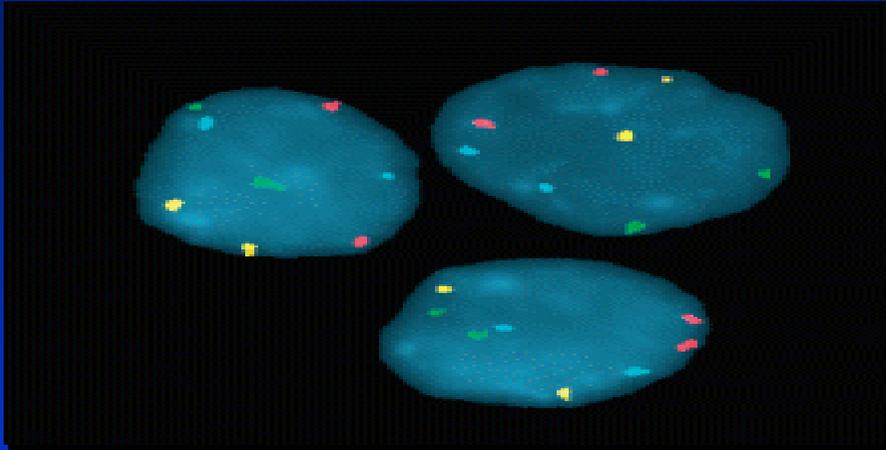
Risk of Colon Cancer in CUC and PSC Compared to CUC Alone



Cholangiocarcinoma



Diagnosis of Cholangiocarcinoma by FISH Technique



- Labeled DNA probes
- Polysomy = CA
- 233 patients (86 PSC)
- PSC and cytology
 - Sensitivity 4-20%
 - Specificity 100%
- PSC and FISH
 - Sensitivity 35-60%
 - Specificity 100%

Management of PSC

Evaluation	Interval
Clinical visit	6-12 months
Serum liver tests	3-6 months
US/MR + CA 19-9	Yearly
Colonoscopy (IBD)	Yearly
Bone density	Diagnosis, 2 years
Vitamin levels	If total bilirubin elevated

--- Thank you ! ---

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