

# Primary Sclerosing Cholangitis

- Immune-mediated disease causing progressive inflammation, fibrosis, and stricturing of intrahepatic and extrahepatic ducts.
- High association with other autoimmune disorders and inflammatory bowel disease (especially ulcerative colitis)
- Increased risk of **cholangiocarcinoma** and other malignancies (including gallbladder cancer)
- Most commonly seen in young (**30-40 years**) **males**, with ↑ incidence in Europe and North America
- Usually worsens progressively with little proven benefit to various medical, endoscopic, or surgical interventions
- Liver transplantation is curative, although primary sclerosing cholangitis (PSC) may recur in liver allograft

# General Features

## ■ Best diagnostic clue

- Multifocal biliary strictures, segmental ductal dilation, bile duct wall thickening, and irregular beading of intra- and extrahepatic bile ducts

## ■ Location

- Common bile duct (CBD) involved in > 90% of patients
- Involvement of both intra- and extrahepatic ducts in 87%
  - » Isolated involvement of intrahepatic (11%) or extrahepatic (2%) ducts is unusual
- Most severely affected segments of biliary tree are usually main right and left bile ducts
- Strictures can affect cystic duct and pancreatic duct

## ■ Morphology

- In patients with PSC-induced end-stage cirrhosis, liver is markedly deformed (to much greater extent than with other common causes of cirrhosis)
  - » Contour is grossly lobulated and rounded with peripheral atrophy and central hypertrophy
  - » Enlargement of central liver and caudate with peripheral atrophy described as "pseudotumoral" enlargement of caudate
  - » Atrophy/hypertrophy complex may even occur in absence of cirrhosis

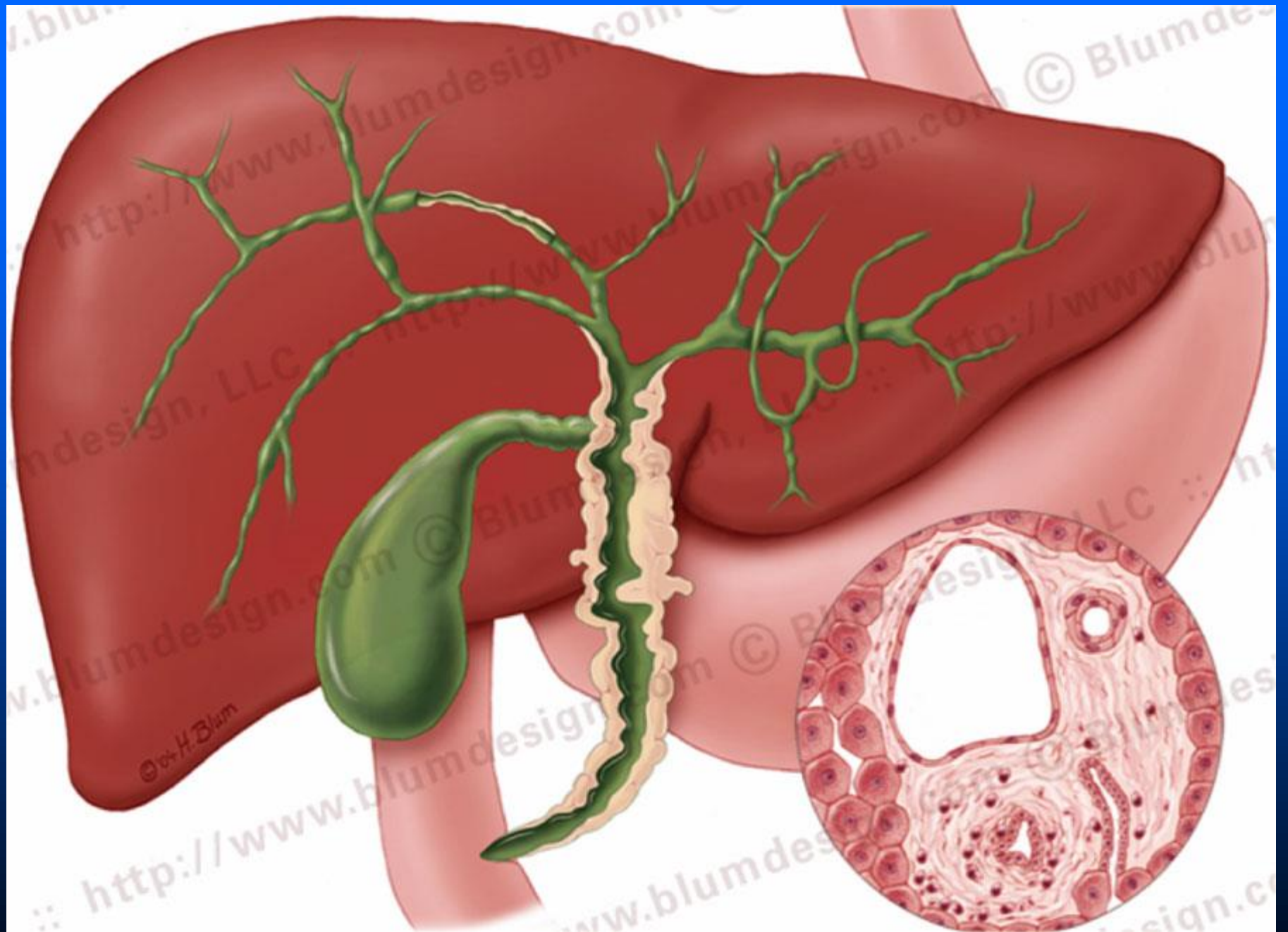
# MRCP

- ERCP remains the gold standard and also offers the ability to perform cholangioplasty if necessary.
- Characteristic of the biliary tree are
  - Multiple segmental strictures
    - » typically short segment
    - » intervening segments are of normal calibre or slightly dilated (beading)
  - Biliary dilatation: may be present in ~85% of cases<sup>9</sup>
    - » general: ~35%
    - » segmental: ~50%
  - Biliary diverticula
  - Mural irregularities
  - Distortion of the biliary tree due to associated cirrhosis

# Primary Sclerosing Cholangitis

- Secondary causes include
  - stone disease, recurrent infection and previous biliary surgery
- Differential diagnosis
  - Recurrent infectious cholangitis
  - Metastatic disease (especially breast cancer)
  - Diffuse sclerosing cholangiocarcinoma
  - Unusual biliary tumors (granular cell tumor)
  - Early Caroli disease
  - AIDS related cholangiopathy
  - Chemo
  - Iatrogenic bile duct strictures





# Primary Sclerosing Cholangitis



70% have underlying UC  
10-15% chance of  
cholangiocarcinoma

Chemo – “Flurourical” can  
occasionally mimic PSC

AIDS cholangitis can mimic  
However can get papillary  
stenosis with AIDS, rare with  
PSC

