

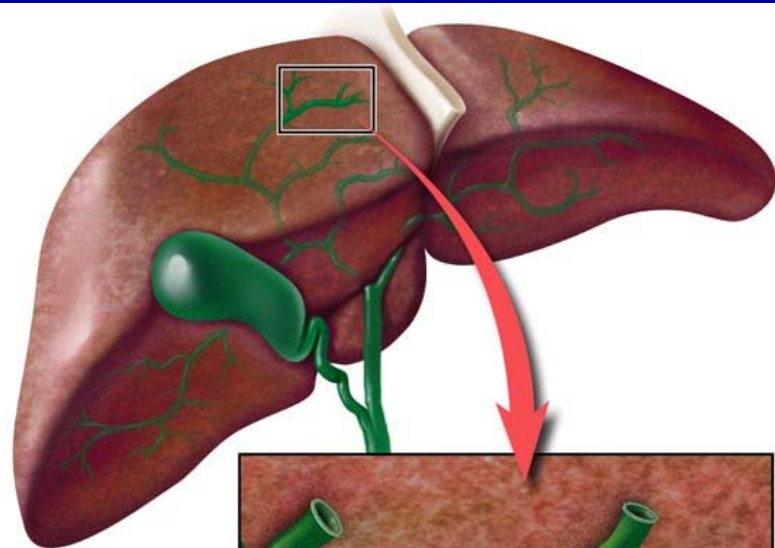
# An Introduction to PSC: Primary Sclerosing Cholangitis

Denise M. Harnois  
Department of GI and Hepatology  
Department of Transplant  
Mayo Clinic  
Jacksonville, FL

# What is PSC?

**“The beginning of wisdom is to call things by their right names.”**

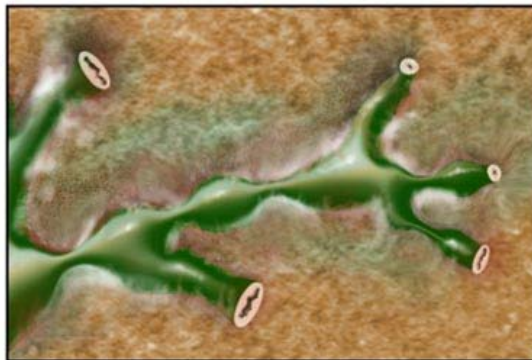
**Chinese proverb**



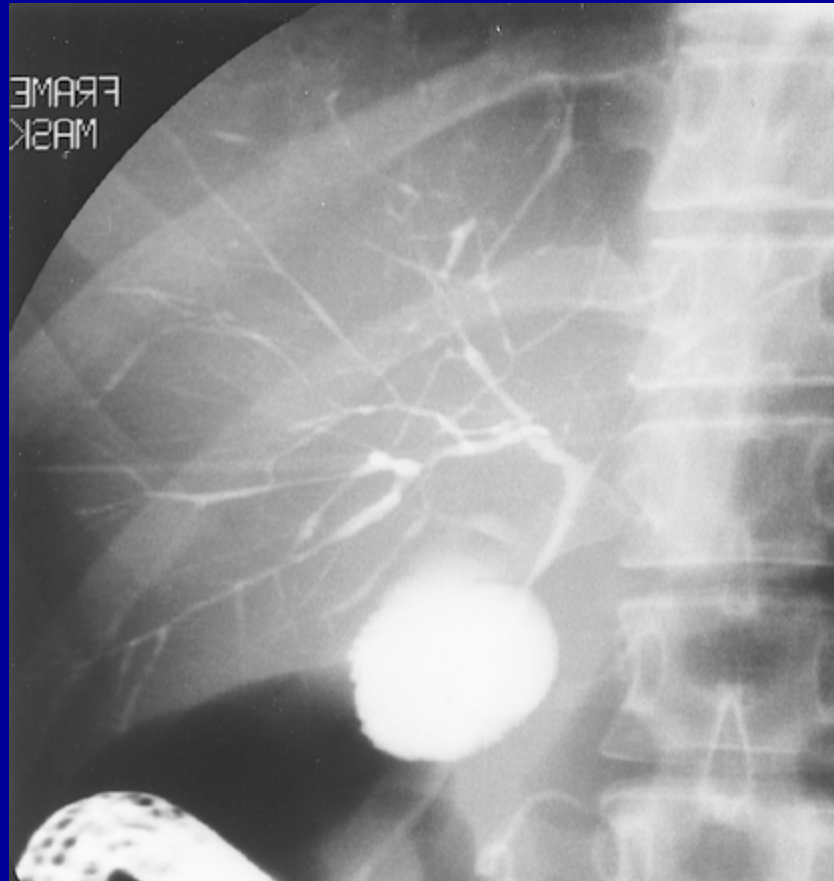
**Normal bile ducts**



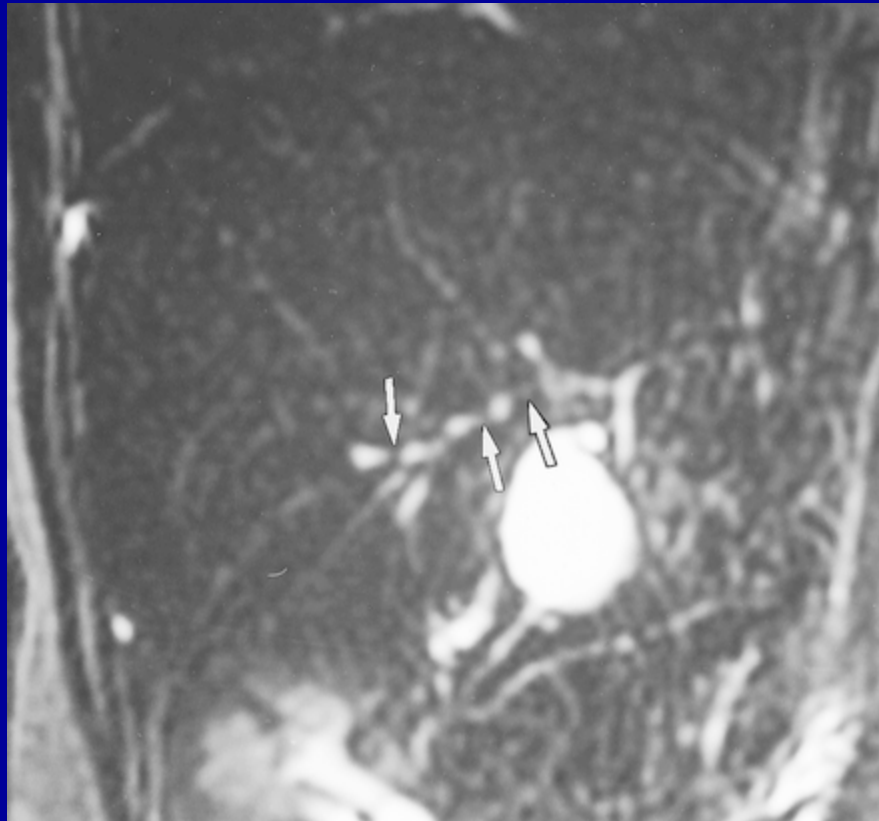
**Inflammation and scar tissue destroy ducts**



# PSC: images on ERCP



# PSC: images on MRI/MRC



# How did I or my family member get PSC?

“We learn geology the morning after the earthquake.”

Emerson

# The causes of PSC are unknown

- Genetic predisposition
- Immunologic abnormalities
- Retention of toxic bile acids may play a role in development of disease

# How is PSC diagnosed?

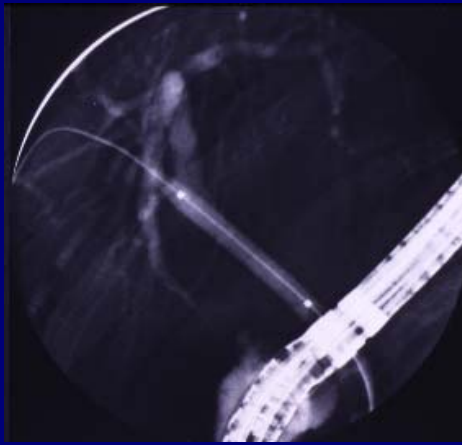
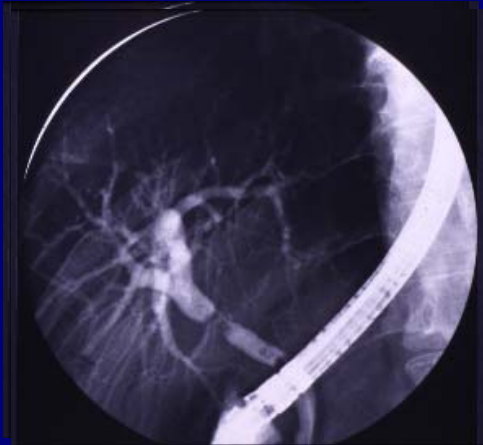
**“A journey of a thousand miles must begin with a single step.”**

**Lao-tzu**



# Stricture Diagnosis and Treatment

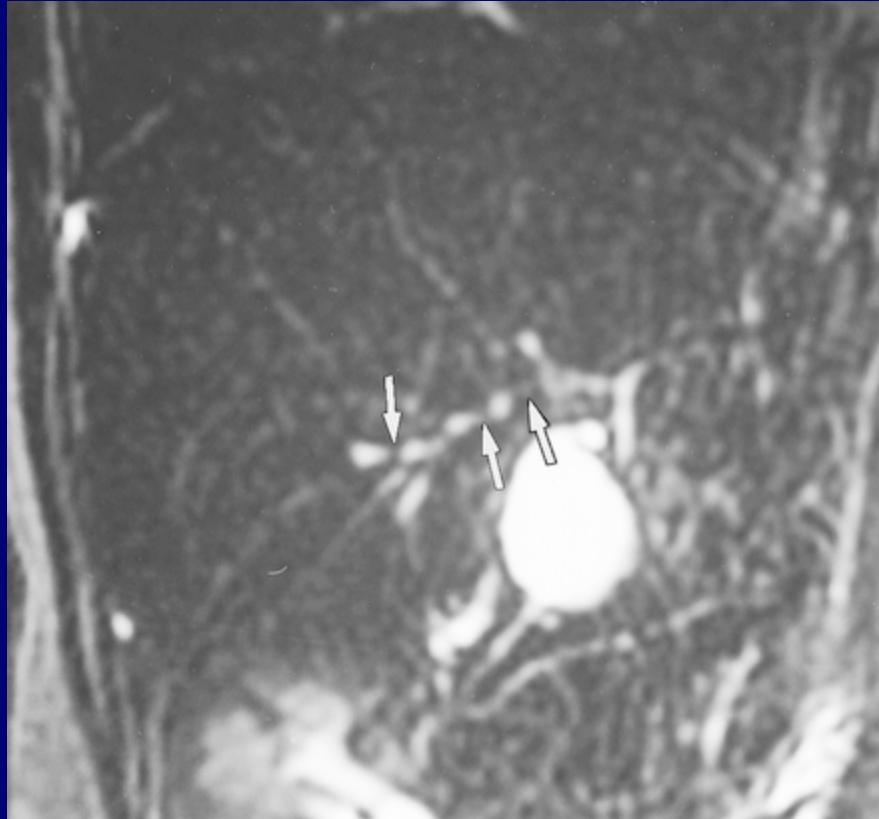
## *Endoscopic Approaches*



## *Radiologic Approaches*



# MRI/MRC: the new kid on the block



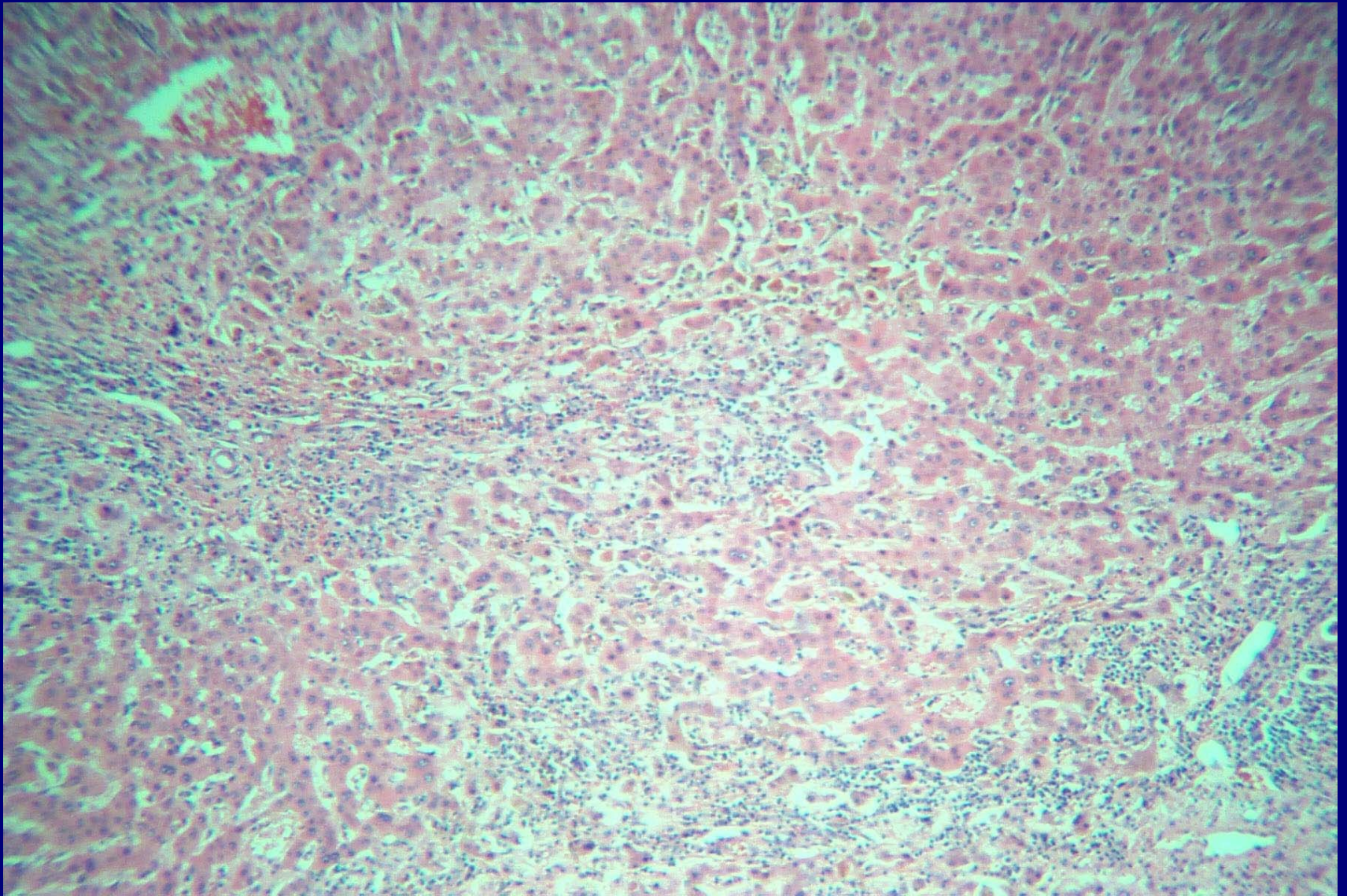
# Is a liver biopsy necessary?

“Knowledge is power.”

Francis Bacon

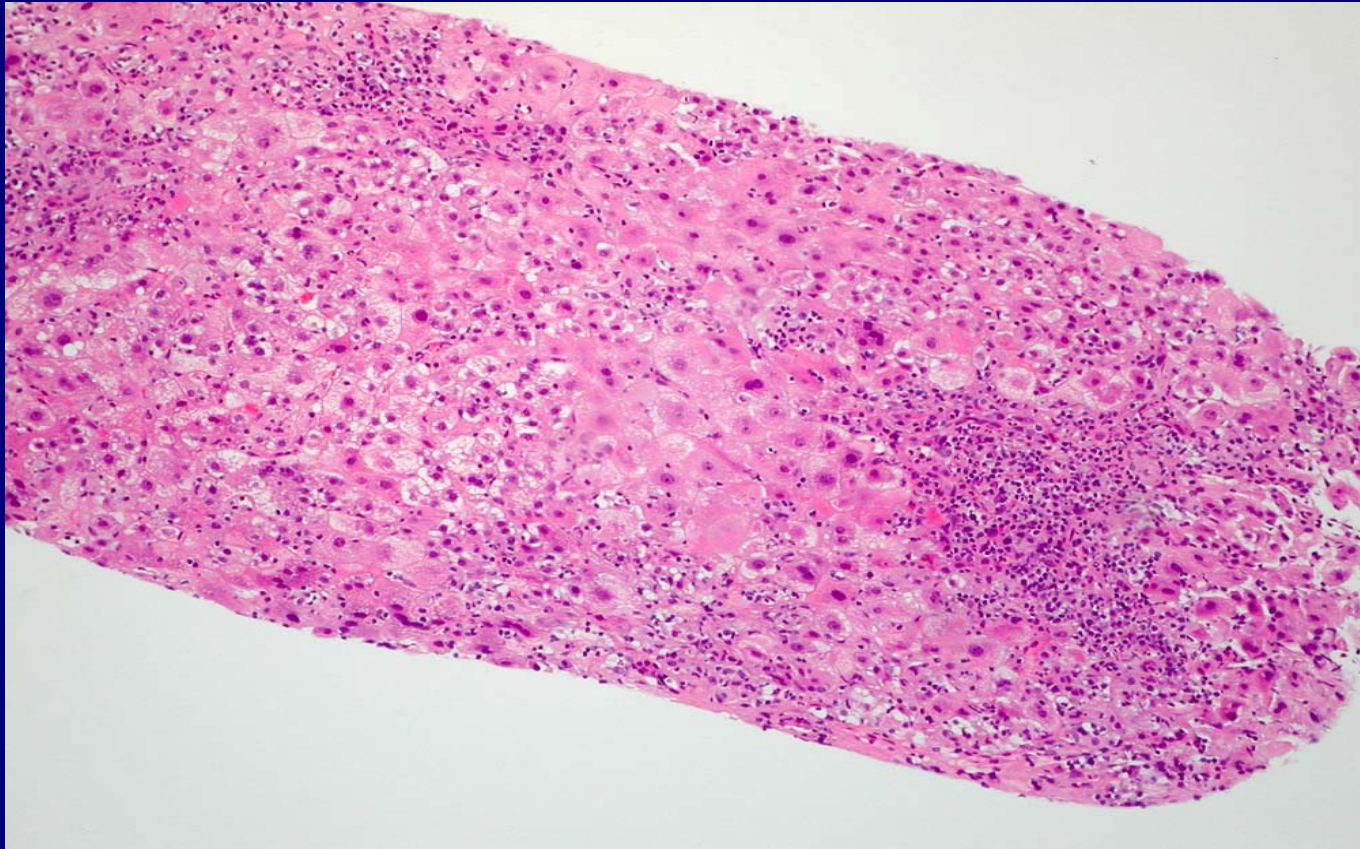


# Liver Biopsy in PSC: Staging





# Liver biopsy to exclude overlap with Autoimmune Hepatitis



# **What is the function of the Liver?**

**“The beginning of health is to know the disease.”**

**Miguel de Cervantes**

# Functions of the Liver

1. **Regulates metabolism**
  - a. **Fat**
  - b. **Carbohydrate (sugar, starch)**
  - c. **Protein**
  - d. **Cholesterol**
  - e. **Vitamins**
2. **Maintains coagulation**
3. **Clears toxins**
4. **Produces enzymes and proteins**
5. **Excretes bile, pigments, and chemicals**
6. **Metabolizes drugs and chemicals**

# Measurements of function

- Laboratory testing:
  - INR: marker of coagulation
  - Albumin: marker of metabolism of liver
  - Total bilirubin
- Clinical symptoms
  - Jaundice and cholangitis (infection)
  - Pruritus (itching) and fatigue
  - Encephalopathy (confusion)
  - Ascites (abdominal fluid accumulation)



# How is PSC treated?

**“No physician, insofar as he is a physician, considers his own good in what he prescribes, but the good of his patient.”**

**Plato**

There is much we still do not  
understand or know.

?

# Treatments

## focus on complications:

Antibiotics for infections

ERCP/PTC

- a. Stones
- b. Strictures

Anti-pruritic agents

URSO therapy (placebo controlled trial)

# **When is it time to consider Liver Transplantation?**

**“..... chance favors only the prepared mind.”**

**Louis Pasteur**

# Models: PSC model vs MELD

- **PSC Model**: Useful in PSC only and not generalizable to other liver diseases:
  - Age
  - Bilirubin (jaundice) on blood testing
  - AST and albumin
  - Presence of varices
  - Estimates Long-term Survival (1 - 4 yr)
- **MELD score**: Generalizable to all liver disease – current method for allocation of livers for transplantation:
  - Bilirubin (jaundice) on blood testing
  - INR (coagulation system) on blood testing
  - Creatinine (kidney function) on blood testing
  - Survival (90 days) is calculated from R
- **MELD SCORE is current standard for allocation of livers to patients with PSC – same as for other diagnosis**

