

Introduction to PSC:

The “Top Ten” Issues for Patients and Families

**Gregory T. Everson, M.D.
Professor of Medicine
Director of Hepatology
UCHSC
Denver, CO**

#1

What is PSC?

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

Chinese proverb

PSC is:

Inflammation and destruction of intrahepatic and extrahepatic bile ducts (cholangitis). This leads to segmental scarring and strictures (sclerosing).

There is no known cause; it is an immune-mediated inflammatory disease focussed on bile ducts (primary).

PSC: Typical Presentations

<u>Gender</u>	<u>Age</u>	<u>Presenting with:</u>	<u>Referral for w/u for:</u>
Female	45	Asymptomatic	Abnl LFTs
Male	32	Jaundice	Biliary Obstruction
Male	55	Hepatic mass	Bile Duct Cancer
Female	48	GI Bleed	Evaluate for Varices and Cirrhosis
Male	49	Itching	Bile duct diseases
Female	59	Diarrhea, abnl LFTs	IBD, IBD-associated Liver Diseases
Male	72	Jaundiced, CUC	PSC, r/o Bile Duct cancer

#2

**How did I or my family
member get PSC?**

“We learn geology the morning after the earthquake.”

Emerson

Cause and Progression of PSC

Genetic Predisposition

Inciting agent

Inflammation and injury

Quiescence

Long-term Followup

#3

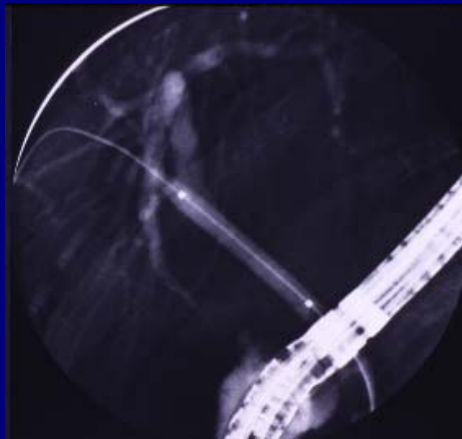
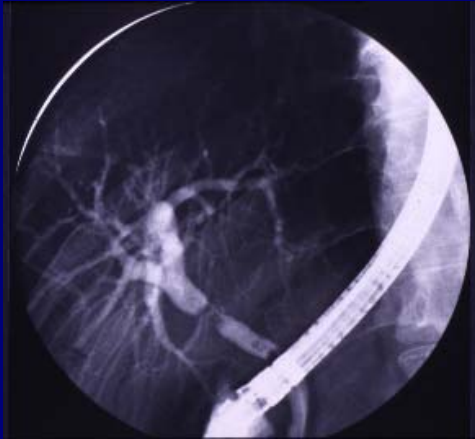
Why do I need an ERCP?

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

Lao-tzu

Stricture Diagnosis and Treatment

Endoscopic Approaches



Radiologic Approaches



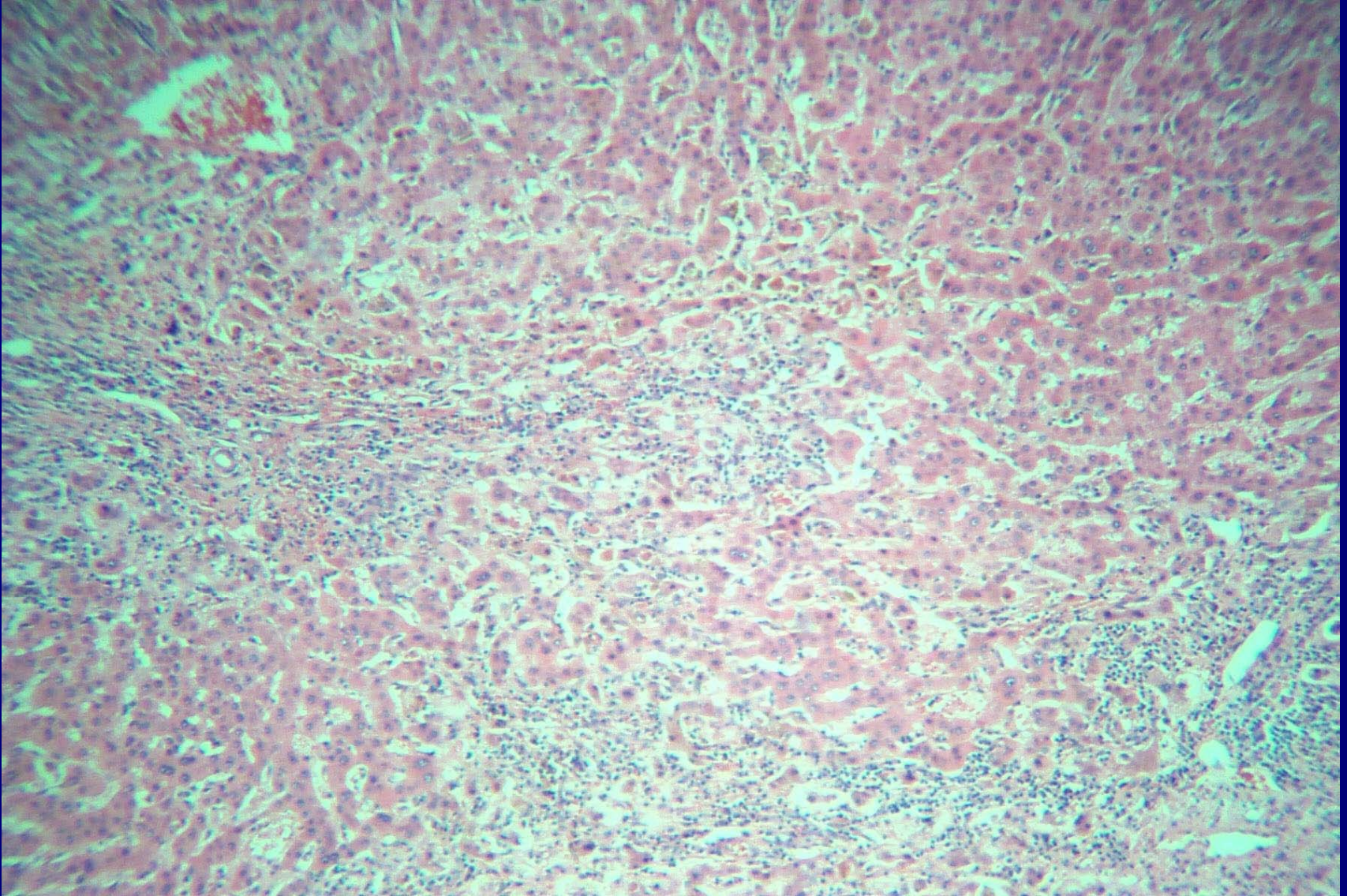
#4

Why do I need a Liver Biopsy?

“Knowledge is power.”

Francis Bacon

Liver Biopsy in PSC: Staging



#5

What are the functions 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 normal clotting
3. Clears toxins
4. Produces enzymes and proteins
5. Excretes bile, pigments, and chemicals
6. Metabolizes drugs and chemicals

#6

What are guidelines for Healthy Nutrition?

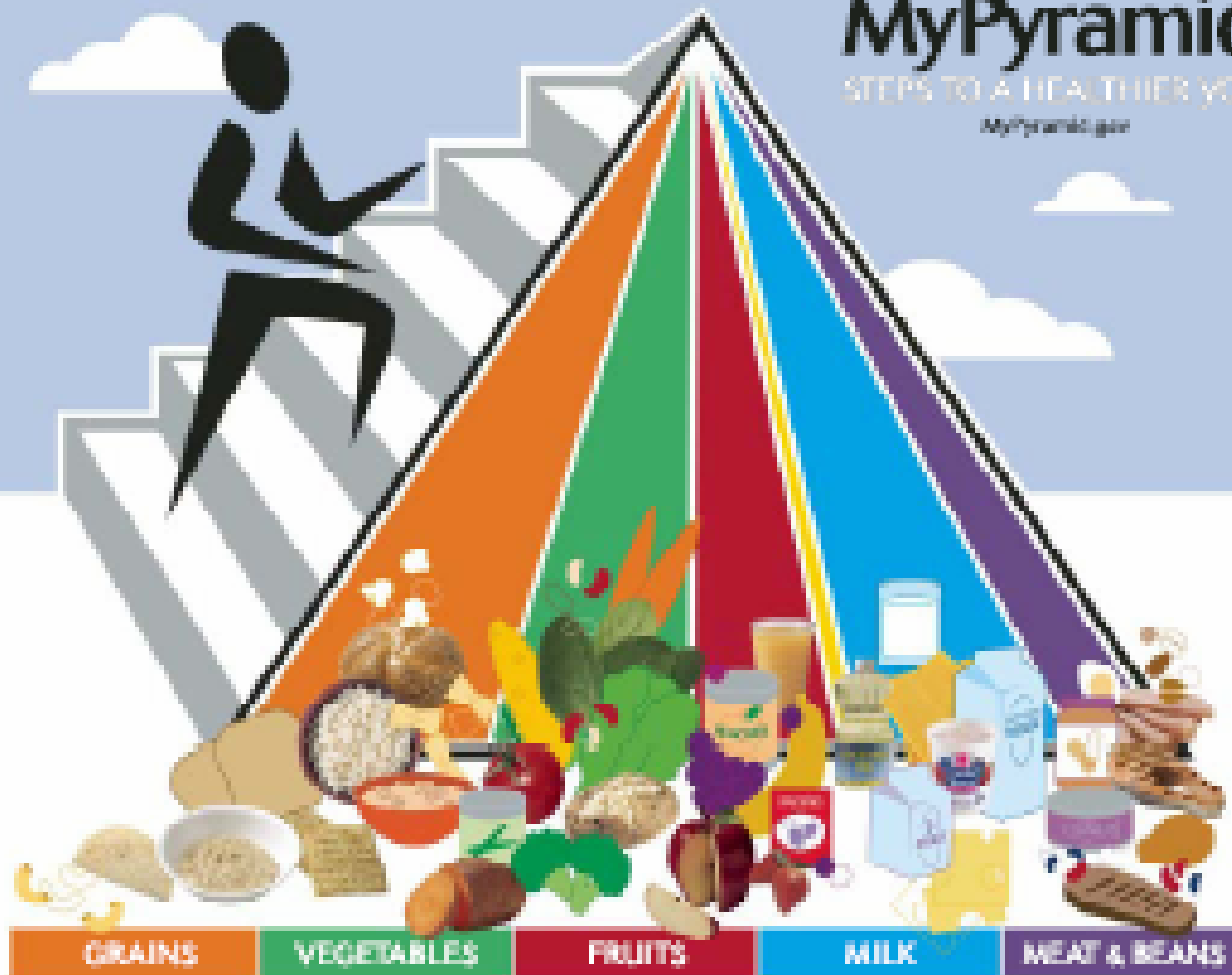
**“He who keeps on eating after his stomach is full
digs his grave with his teeth.”**

Turkish proverb

MyPyramid

STEPS TO A HEALTHIER YOU

MyPyramid.gov



#7

How can I best deal with the Emotional Roller-Coaster?

“There is no grief which time does not lessen and soften.”

Cicero

Support Systems

1. One's self
 - a. Education and knowledge
 - b. Know what to expect
 - c. Gain control
 - d. Developing coping skills
 - e. Maintain general health
2. Spouse or significant other
3. Family
4. Friends
5. Professionals
6. Medications

#8

What are the best Medical Treatments?

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

Plato

Medical Treatments

1. Antibiotics for infections
2. ERCP/PTC
 - a. Stones
 - b. Strictures
 - c. Bile Duct Cancers
3. UDCA – Pros and Cons
4. Anti-pruritic agents

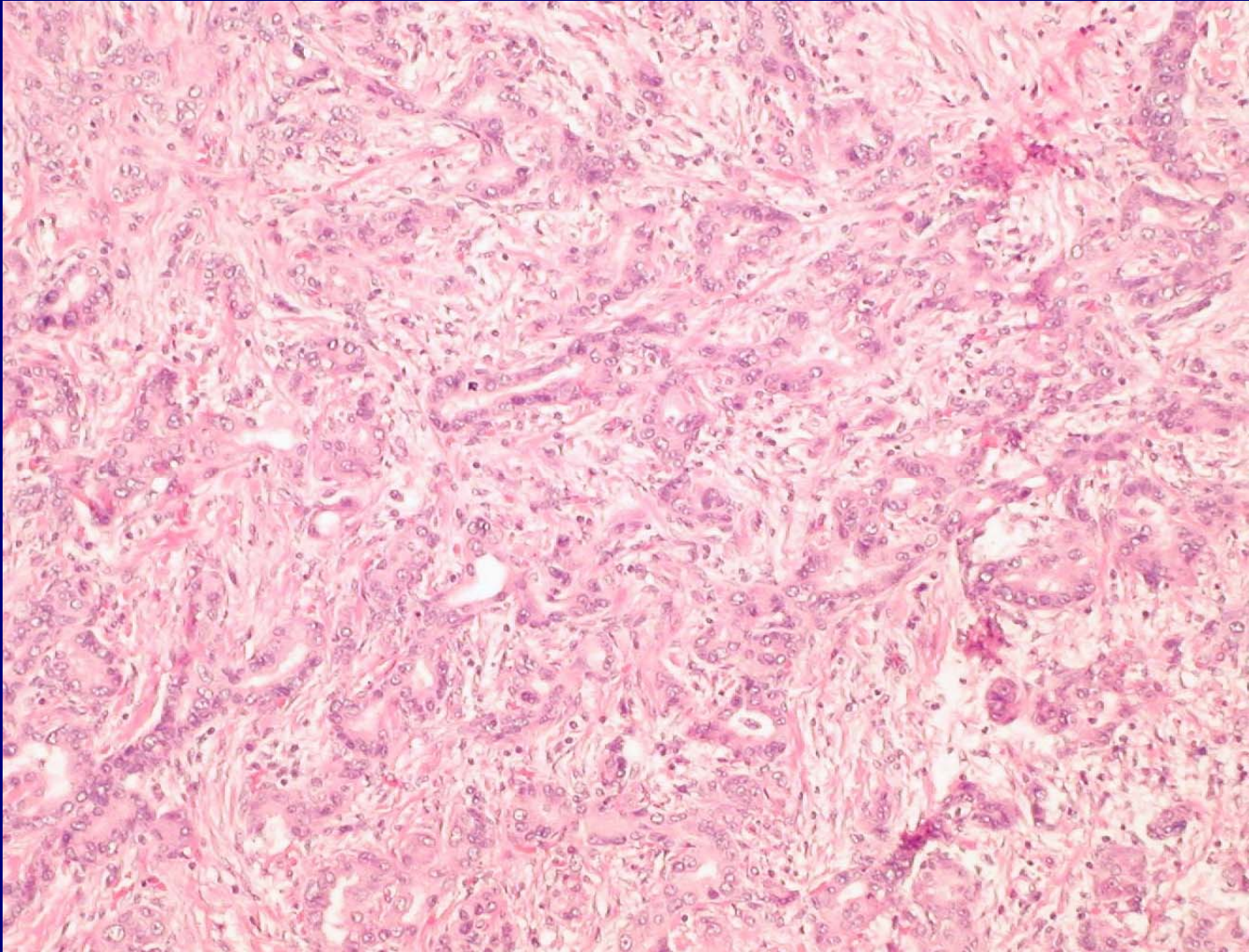
#9

**What is my risk of cancer?
Of Bile Duct?
Of Colon?**

“The first wealth is health.”

Emerson

Cholangiocarcinoma complicating PSC



#10

When is it time to consider Liver Transplantation?

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

Louis Pasteur

Models: PSC model vs MELD

- **PSC Model (Estimates 1 – 5 yr Survival)**: Useful in PSC only and not generalizable to other liver diseases:
 - Age
 - Bilirubin (jaundice)
 - Fibrosis Stage on Liver Biopsy
 - Spleen size on ultrasonography
- **MELD score (Estimates 90 day Survival)**: Generalizable to all liver disease:
 - Bilirubin (jaundice)
 - INR (coagulation system)
 - Creatinine (kidney function)
- **MELD SCORE is current standard for allocation of livers to patients with PSC – same as for other diagnosis**