

Introduction to PSC:

The “Top Ten” Issues for Patients and Families

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#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 autoimmune disease (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 can I interpret the reports of
Blood Tests,
ERCP,
PTC, and
Liver Biopsy?**

“Knowledge is power.”

Francis Bacon

Evaluation

Liver injury

	<u>Patient</u>	<u>Normal Range</u>
Enzymes:	AST 47	< 40
	ALT 78	< 40
	Alk Phos 423	< 112
Bx:	Grade 2	0
	Stage 2	0

Liver Function

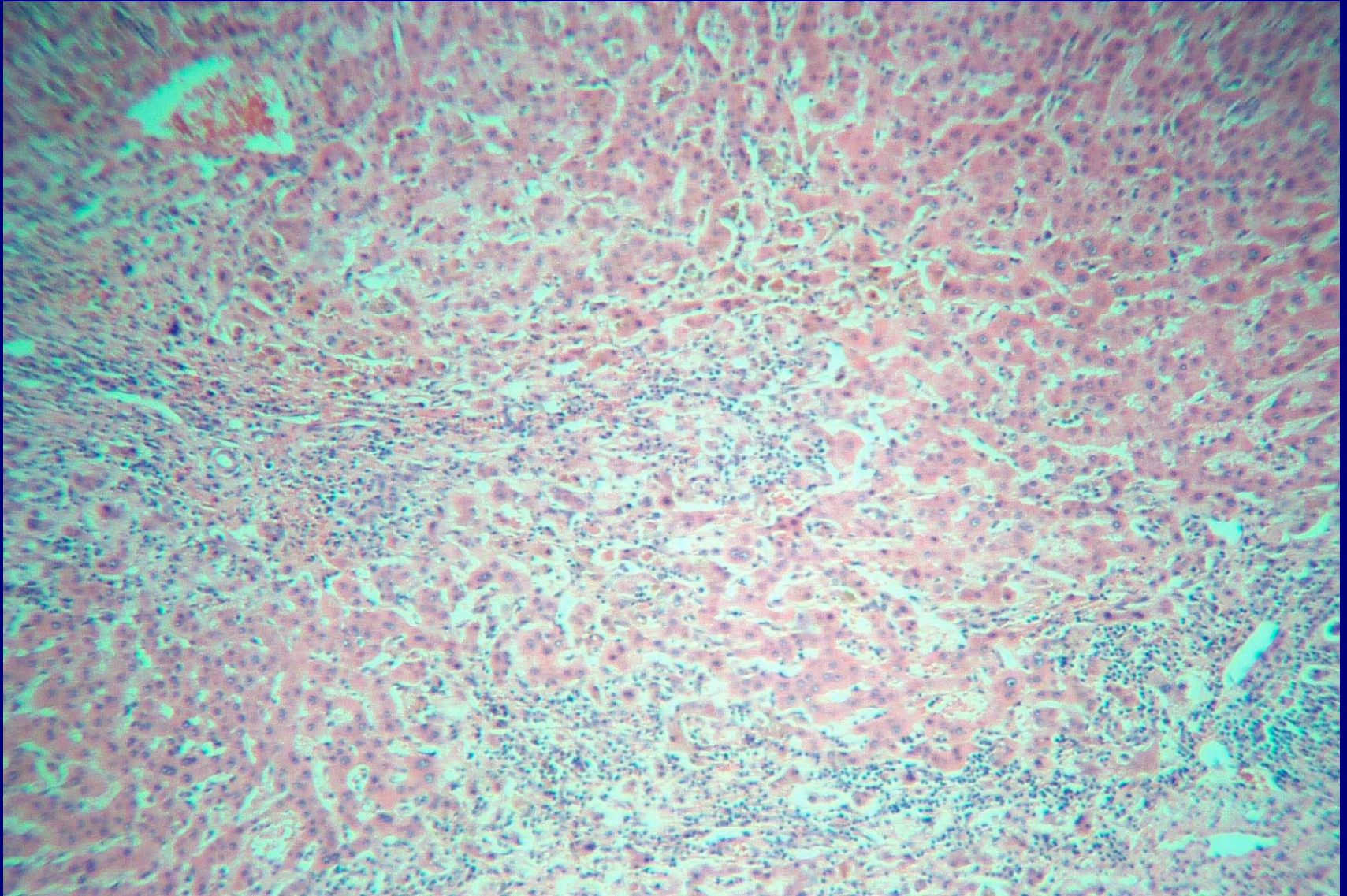
Synthesis:	INR 1.3	1
	Albumin 3.2	> 3.5
Excretion:	Bilirubin 2.1	< 1.2

Metabolism, Blood Flow, Hepatocyte Mass

Ductal Injury

ERCP/PTC	Strictures	None
	Intrahepatic	
	Extrahepatic	
	Stones	None
	CCA	None

Liver Biopsy in PSC



#3

**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

#4

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

Xanthelasma



Xanthomata



#5

What are guidelines for Healthy Nutrition?

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

Turkish proverb

Avoid Alcohol



GUINNESS

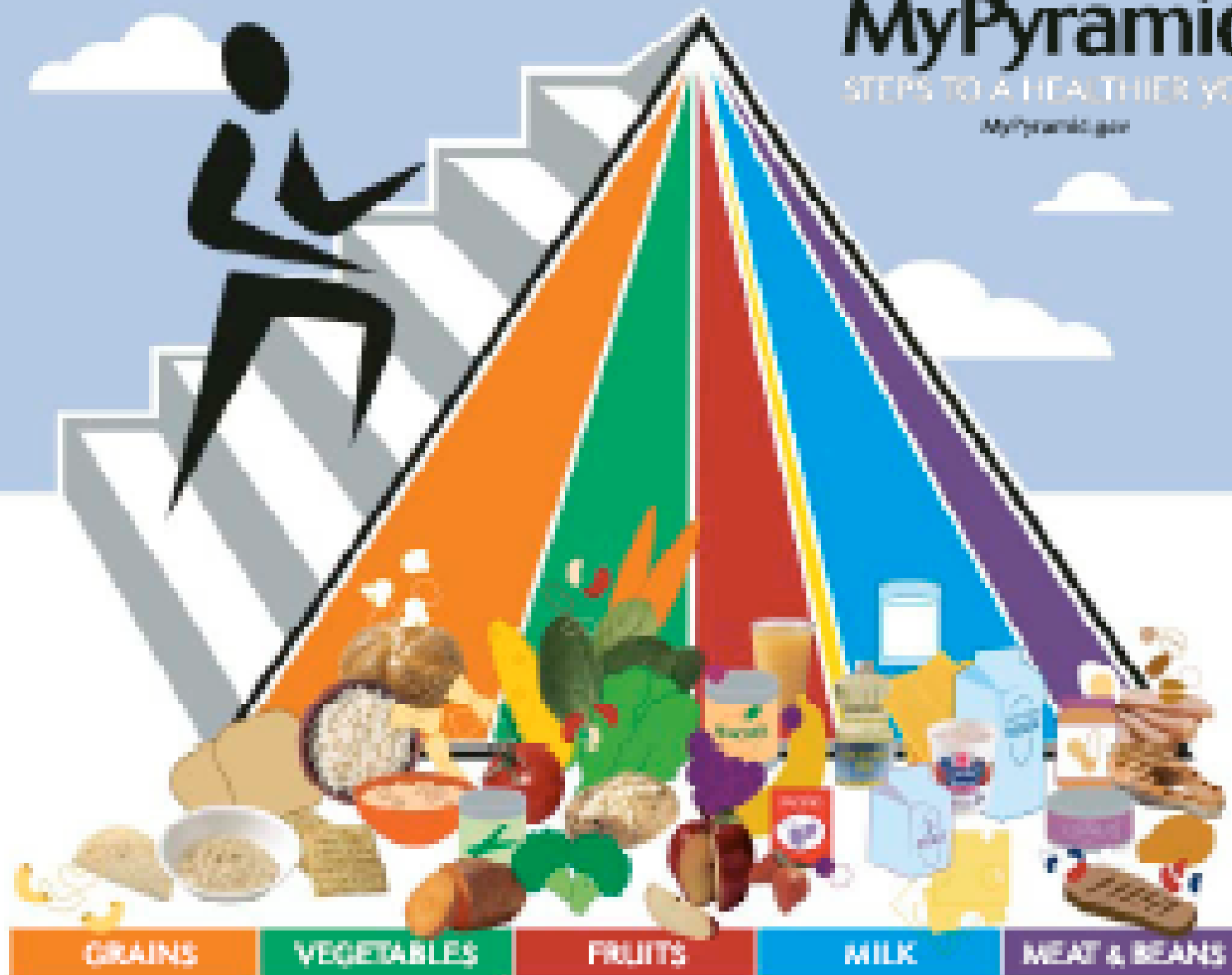
FOR STRENGTH



MyPyramid

STEPS TO A HEALTHIER YOU

MyPyramid.gov



#6

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

#7

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
4. Anti-pruritic agents

#8

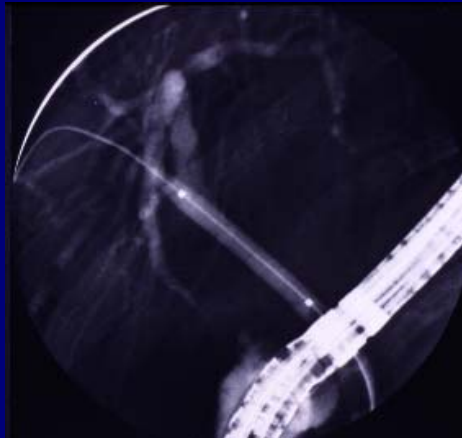
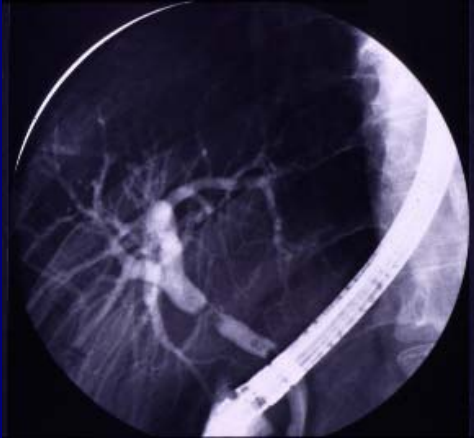
**What do you mean I “need”
ERCP or PTC?**

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

Lao-tzu

Stricture Management

Endoscopic Approaches



Radiologic Approaches



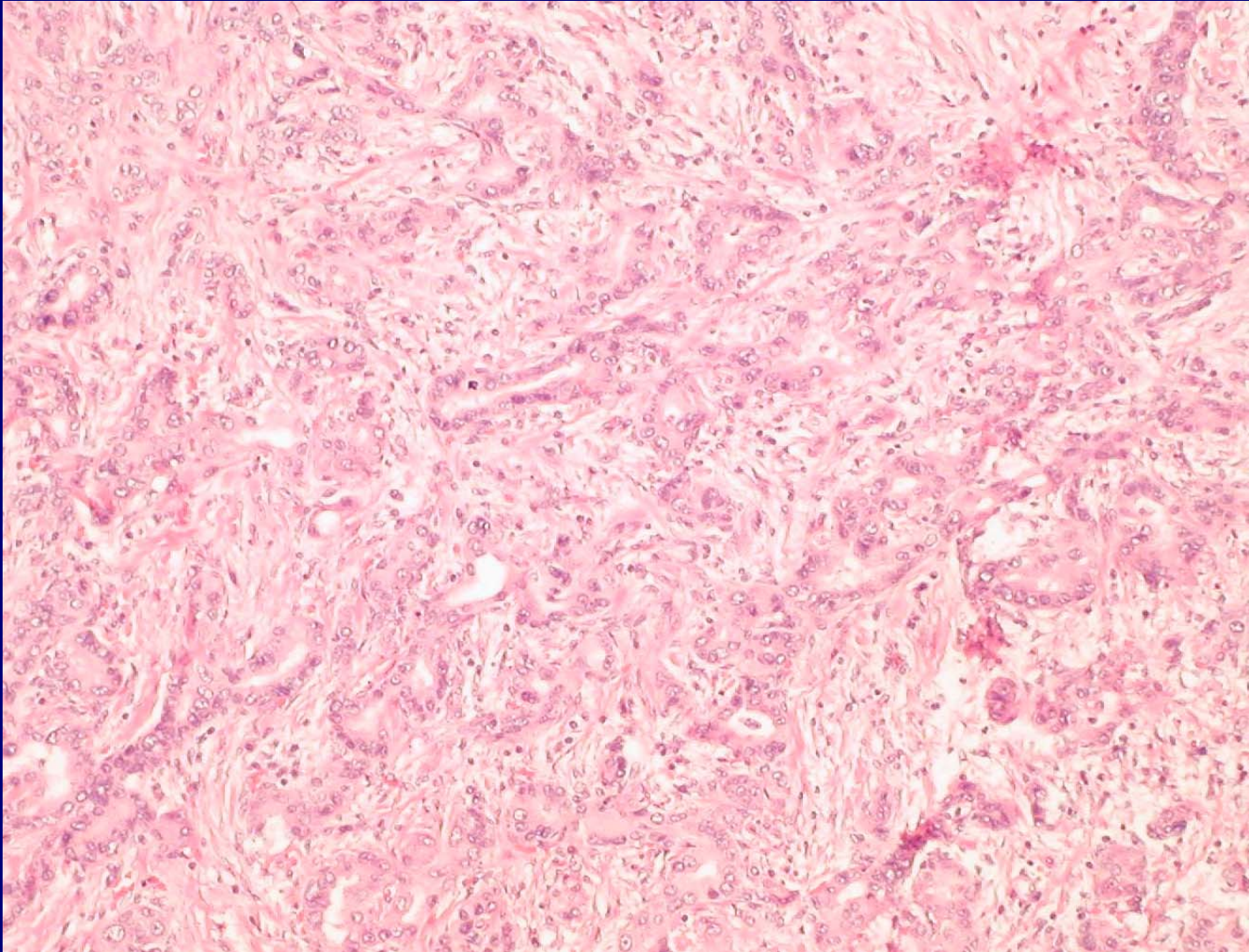
#9

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

“The first wealth is health.”

Emerson

Cholangiocarcinoma complicating PSC



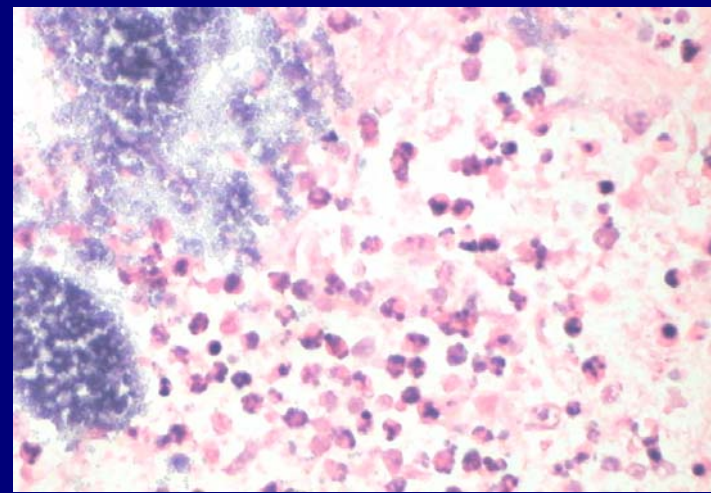
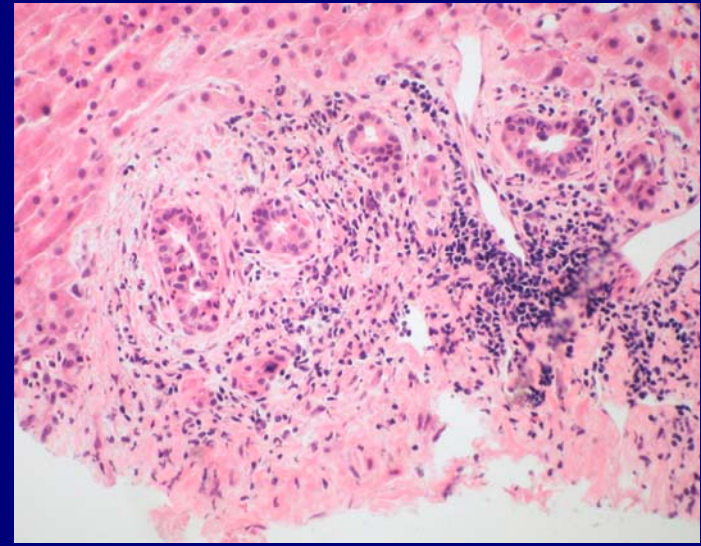
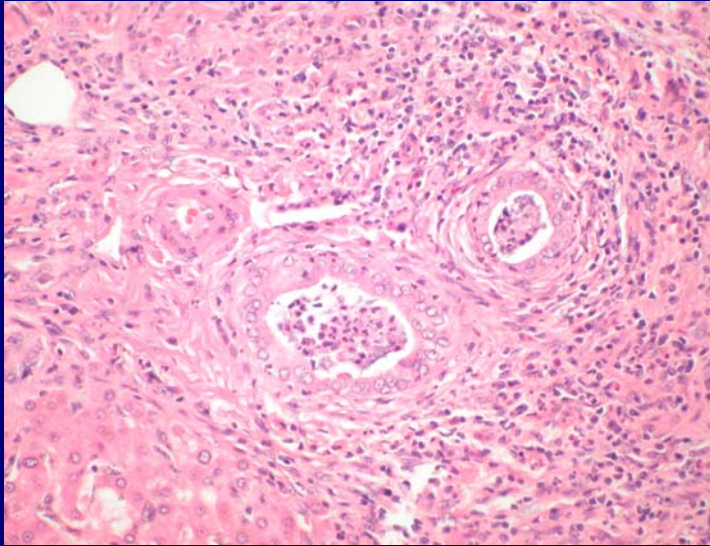
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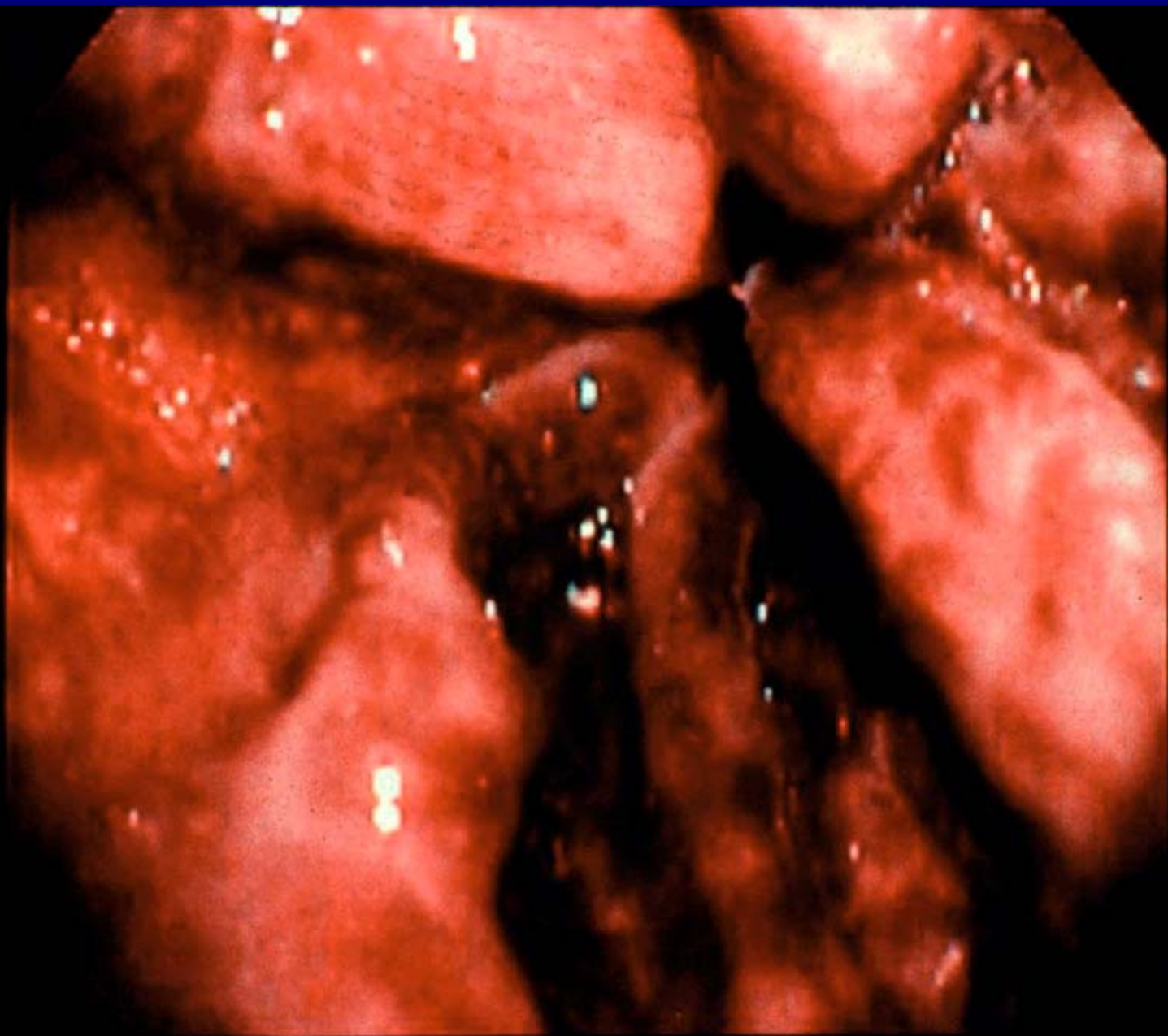
When is it time to consider Liver Transplantation?

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

Louis Pasteur

Features of Bacterial Cholangitis





Prognostic Model for PSC

- *Useful in PSC only and not generalizable to other liver diseases and only predictive once lab tests become abnormal*
- *Model:*
 - $R = .535\ln(\text{bili}) + .486(\text{Stage on Bx}) + .041(\text{age}) + .705(\text{spleen})$
 - *Survival is calculated from R*
- *Both internally (Mayo) and externally (other centers) validated to predict survival in patients with PSC*