Portal Hypertension in Congenital Hepatic Fibrosis:

Diagnosis and Treatment

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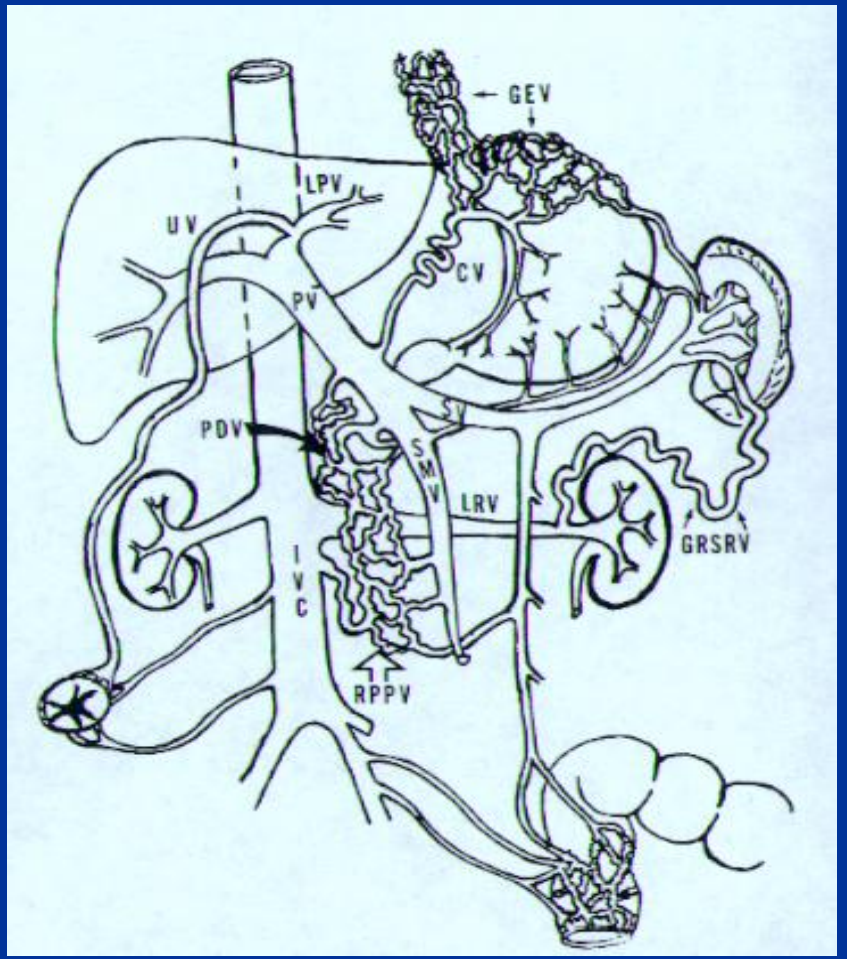
(with many thanks to Dr. Piccoli for sharing slides!)

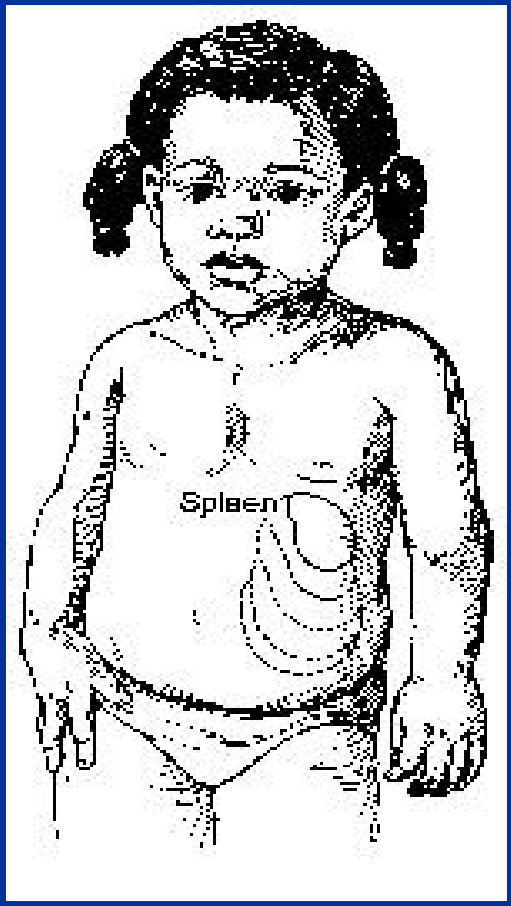
What is portal hypertension?

- Increased pressure in portal venous system causes backup of blood into spleen and other vessels
  - Enlarged spleen
  - Low white blood cell count
  - Low platelet count

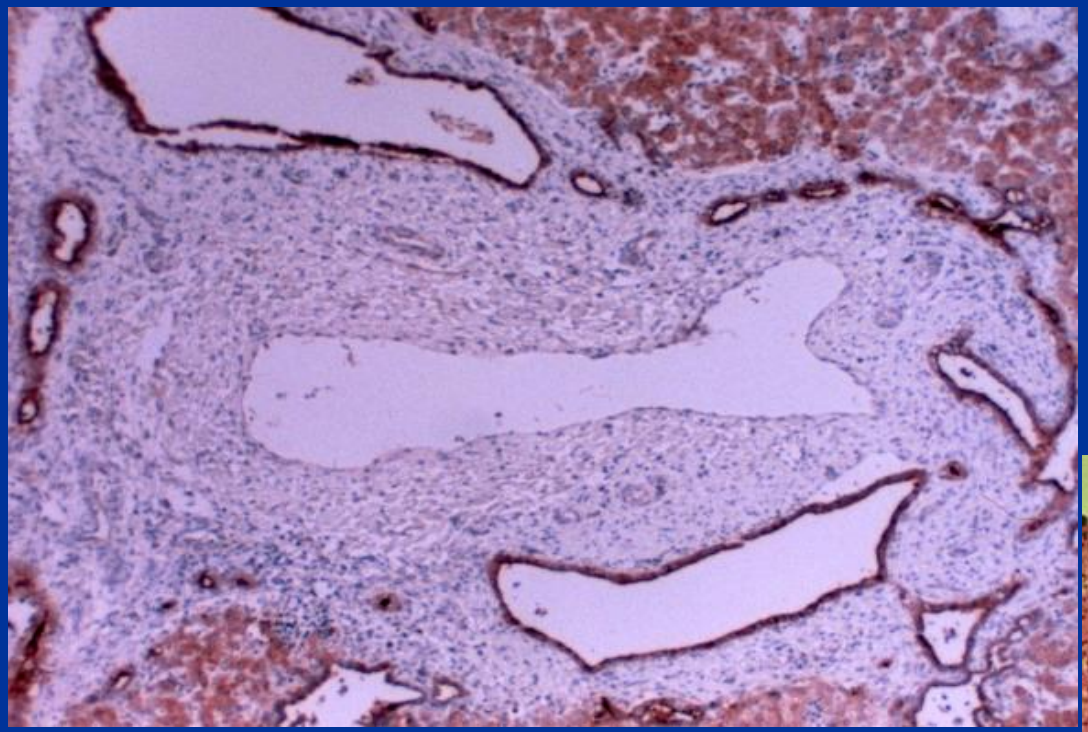


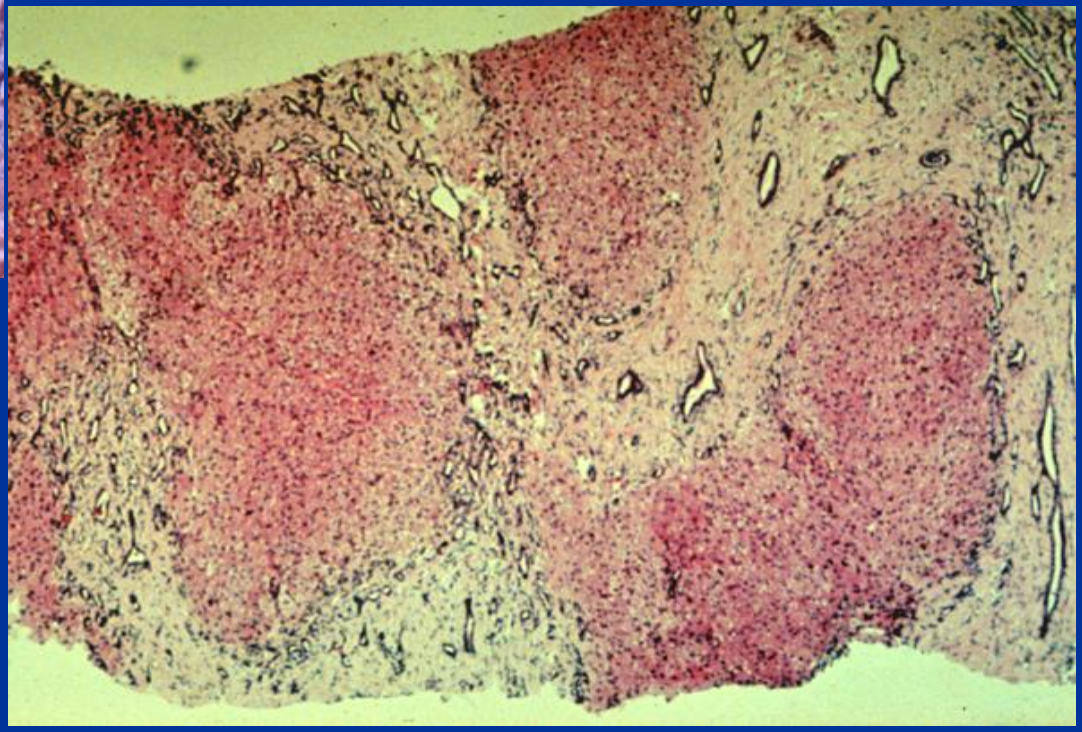
How do we diagnose portal hypertension?

- Not usually possible to measure portal pressures directly
- Define portal hypertension as:
  - Enlarged spleen (2 cm below left rib margin)
  - Low platelet count (< 150,000)



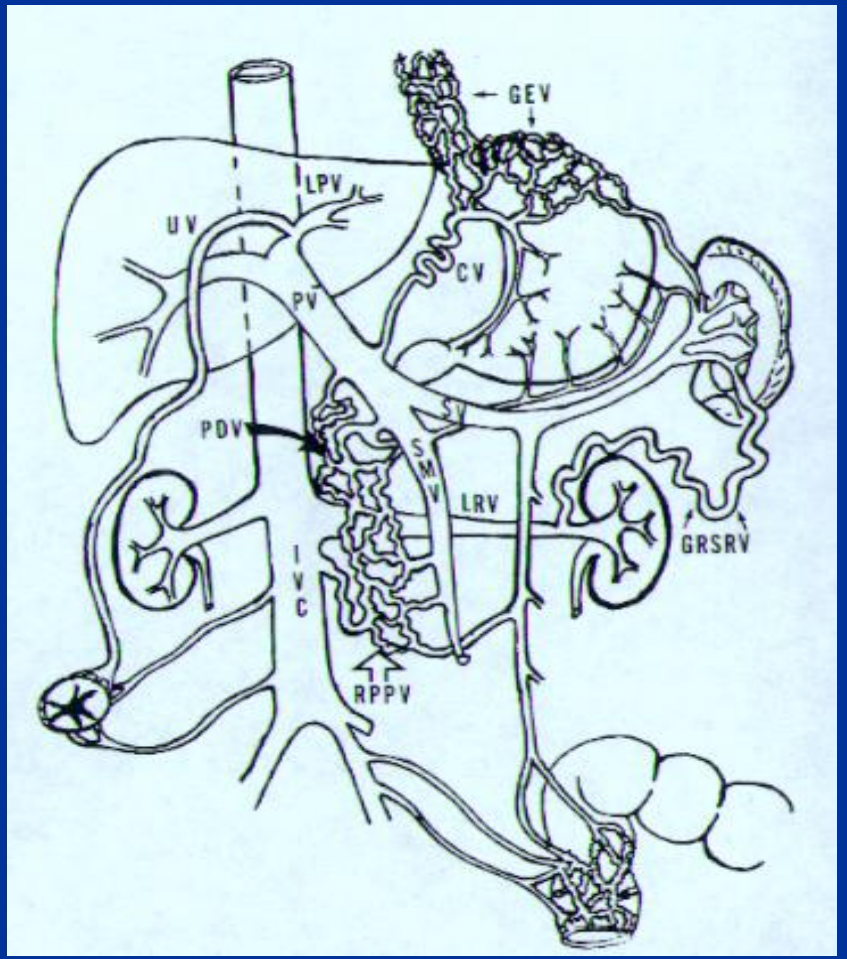
Why do CHF patients get portal hypertension?





Complications of Portal Hypertension in CHF

- Splenomegaly
- Ascites/peritonitis
- Varices
- Portal hypertensive gastropathy
- Hepato-pulmonary syndrome
- Caput medusa
- Hemorrhoids



Spleen guard

Therapy For Portal Hypertension with GI Bleeding

- Medical therapy - designed to decrease the pressure and volume in the portal system
- Endoscopic therapy – injection or banding of the esophageal varices to clot them or thicken the wall - so that flow will go in a different direction
- Surgical shunts- taking the high pressure portal or splenic blood flow and plugging it into low pressure vessels
- Radiologic shunts - TIPS - making a connection between the high pressure portal vein and the low pressure hepatic vein
- Liver transplantation

Initial Stabilization – Local Emergency Room!!

- ABC's (airway, breathing, circulation)
- Adequate intravenous access
- Fluid resuscitation
  - Initially with normal saline
  - Packed red blood cell transfusion
  - Clotting factors and platelets as needed
- Vitamin K IV
- Acid blockade IV
  - Ranitidine, pantoprazole, lansoprazole

- Nasogastric (NG) tube is essential

NG Tube Placement And Lavage

- Identify location of bleeding
  - Verify bleeding with Gastroccult
- Monitor amount of ongoing losses
- Does not by itself stop bleeding
- No role for iced-saline lavage
- Balance the gastric infusion/suction (I/O) to avoid fluid overload

Medical Management of Variceal Upper GI Bleed

- GI / PICU consults
- Acid blockade with IV H2 blockers, PPIs
- Octreotide (1 mcg/kg bolus, then 1 mcg/kg/hour)
  - Follow blood sugars
- Vasopressin less commonly used
  - Side effects include SIADH, seizures, peripheral ischemia
- Packed red blood cell, plasma and platelet transfusions
  - May need large amounts
- Avoid sodium overload

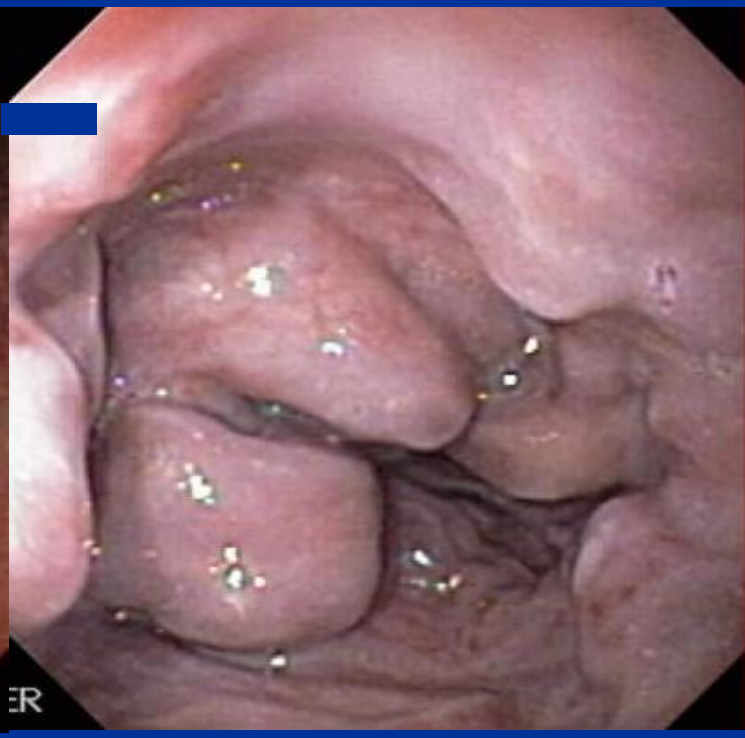
Role of Upper Endoscopy in UGI Bleeding

- Diagnostic
  - Identifies the location and type of bleeding in up to 90%
- Therapeutic
  - Sclerotherapy
  - Variceal band ligation
  - Bipolar coagulation for ulcers
  - Other techniques

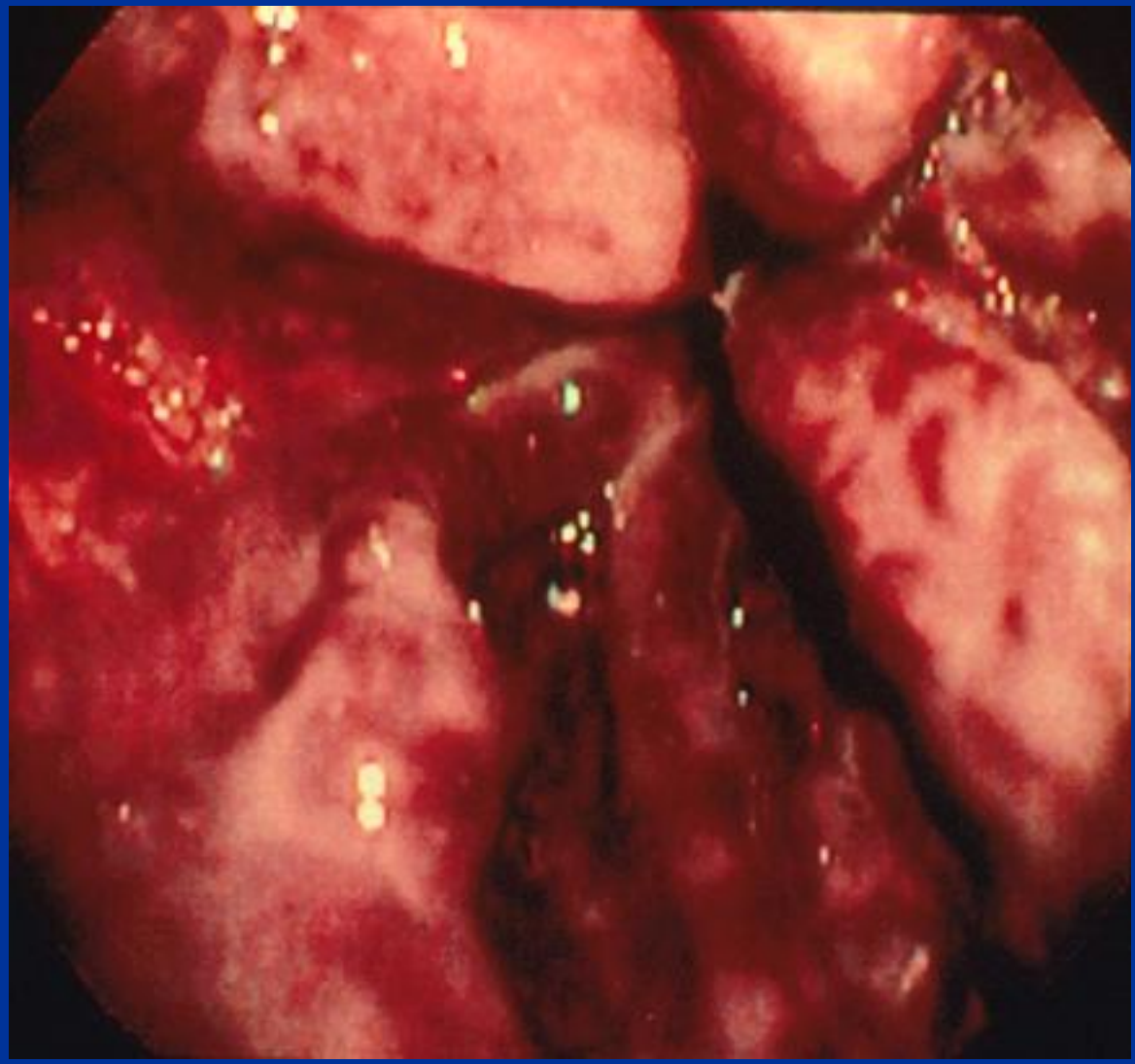
Esophageal Varices

Normal

Varices



Bleeding Esophageal Varices

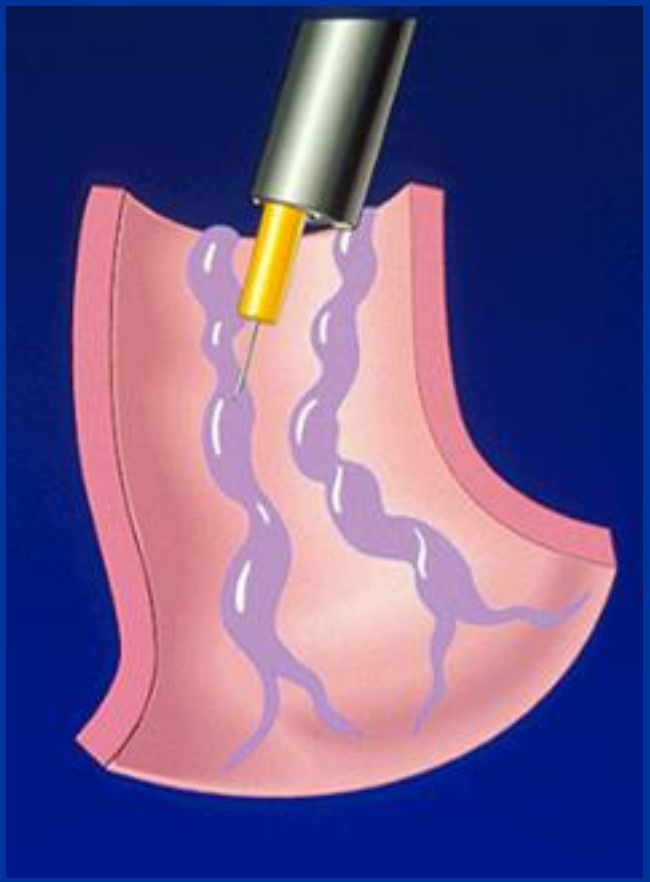


Sclerotherapy

- Injection of varices with a sclerosing agent
- Indications and uses
  - Active variceal bleeding
  - Therapy following a bleed
  - Prophylactic therapy
- Role in pretransplant therapy

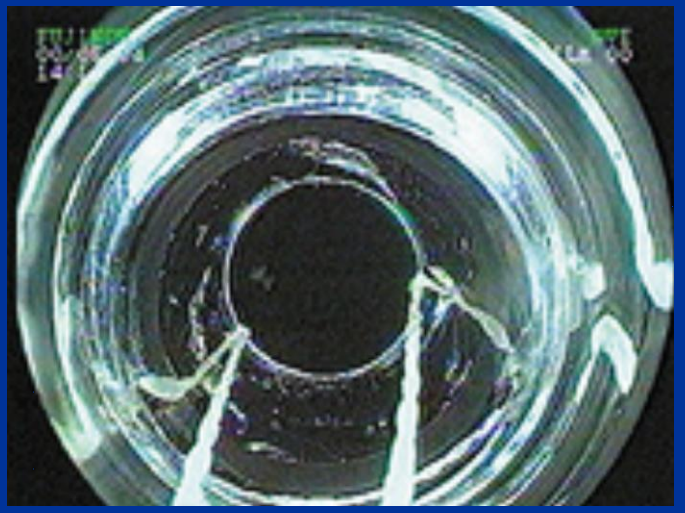
Sclerotherapy

- Bleeding controlled in 80-95%
- Effect on rebleeding rate uncertain
- No effect on survival in adults
- Major complications in 10-20%
- No large scale studies in children



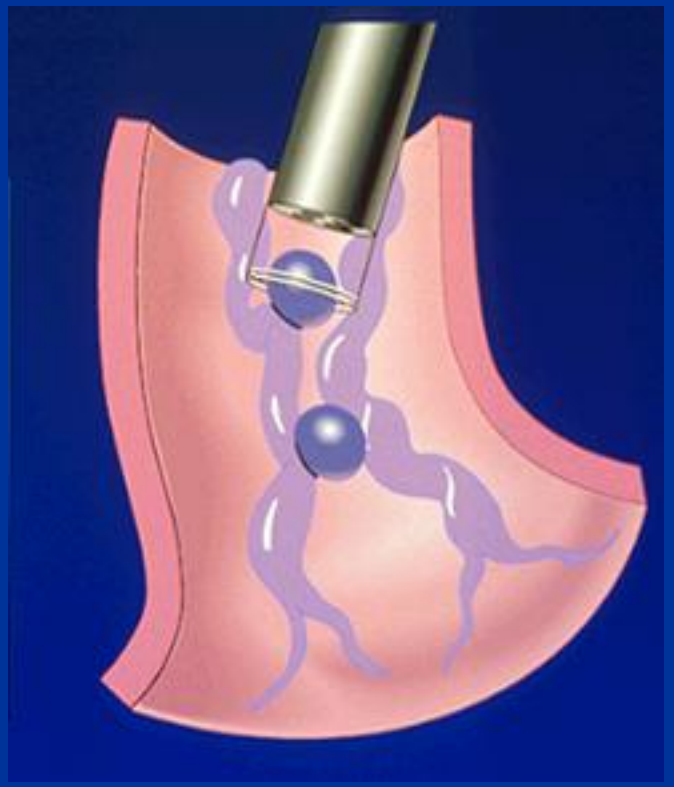
Variceal Band Ligation

- Acute or chronic care of esophageal varices
- Spring loaded multiple rubber bands
- Technique
  - Upper endoscopy
  - Suction to pull varix into chamber
  - Firing a band over the neck of the varix
- Band occludes varix
- Necroses (dies) and falls off after several days
- Minimizes many of the risks of sclerotherapy
- No large scale studies in children
- Chamber cannot fit into the esophagus of infants and young children

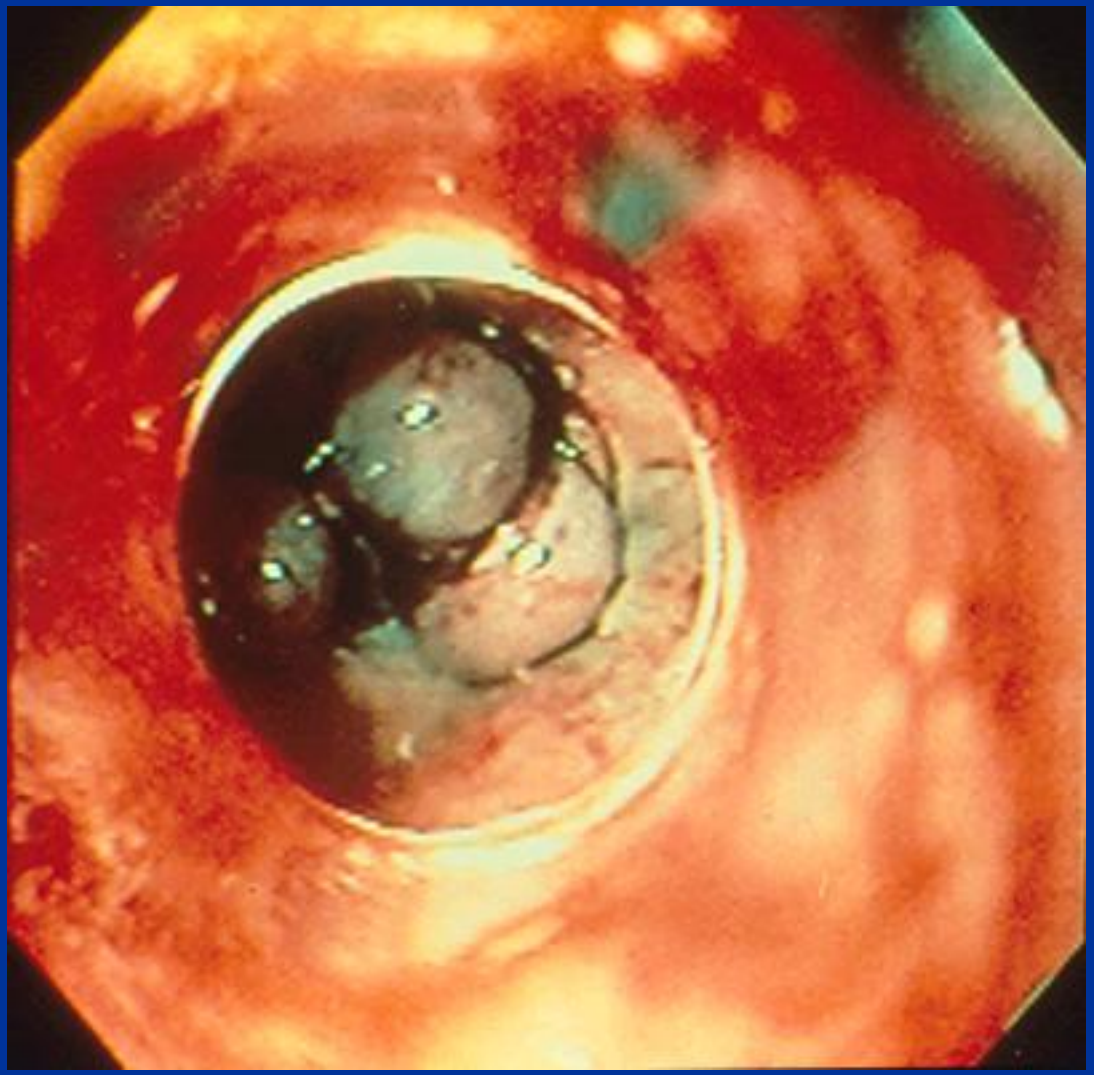


Variceal Ligation - Banding

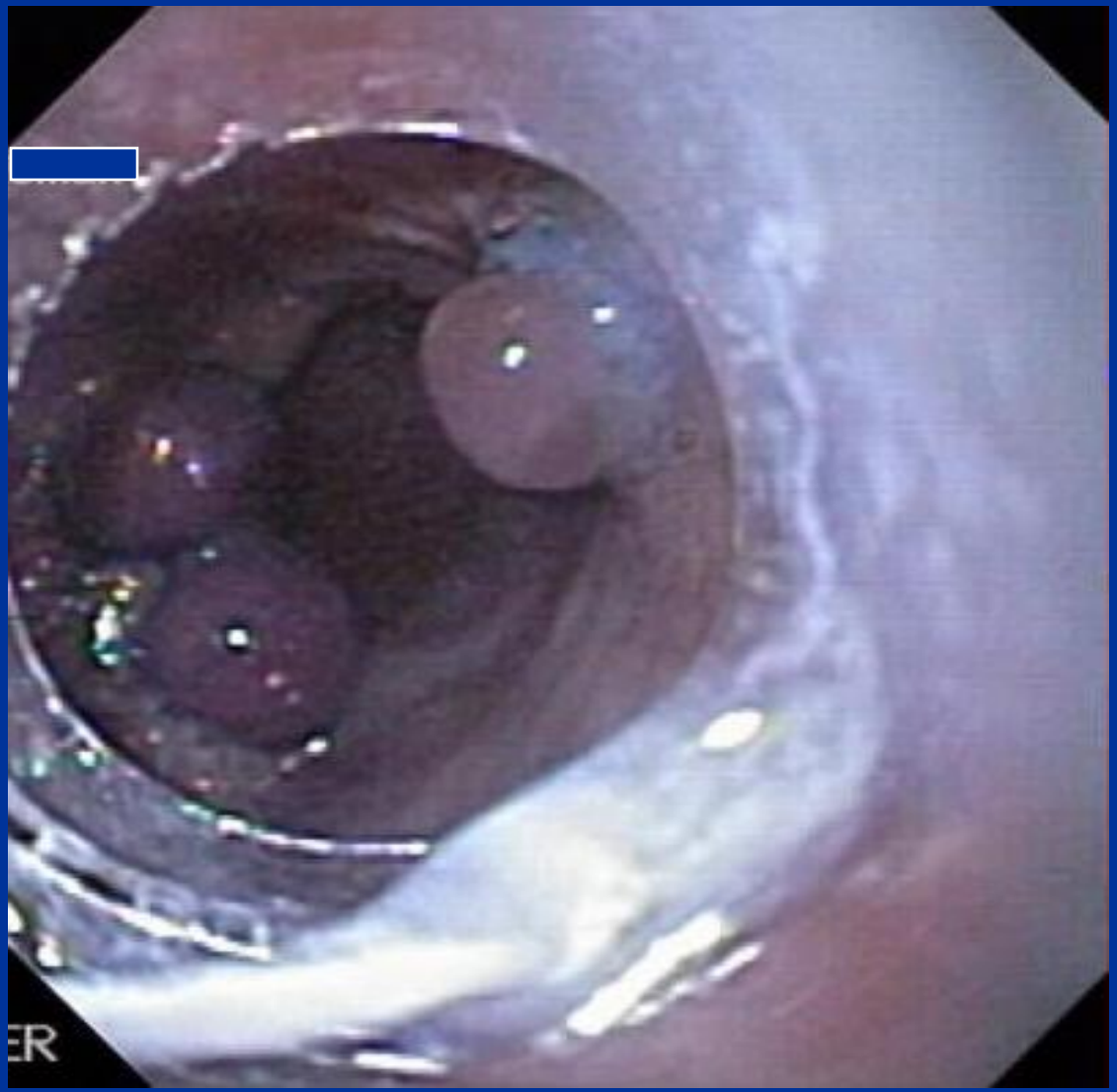
- Bleeding controlled in 90%
- Rebleeding rate reduced to 30%
- Compared with sclerotherapy
  - Less rebleeding
  - Lower mortality
  - Less complications
  - Fewer treatment sessions



Variceal Bleeding with Band Ligation



Post Band Ligation



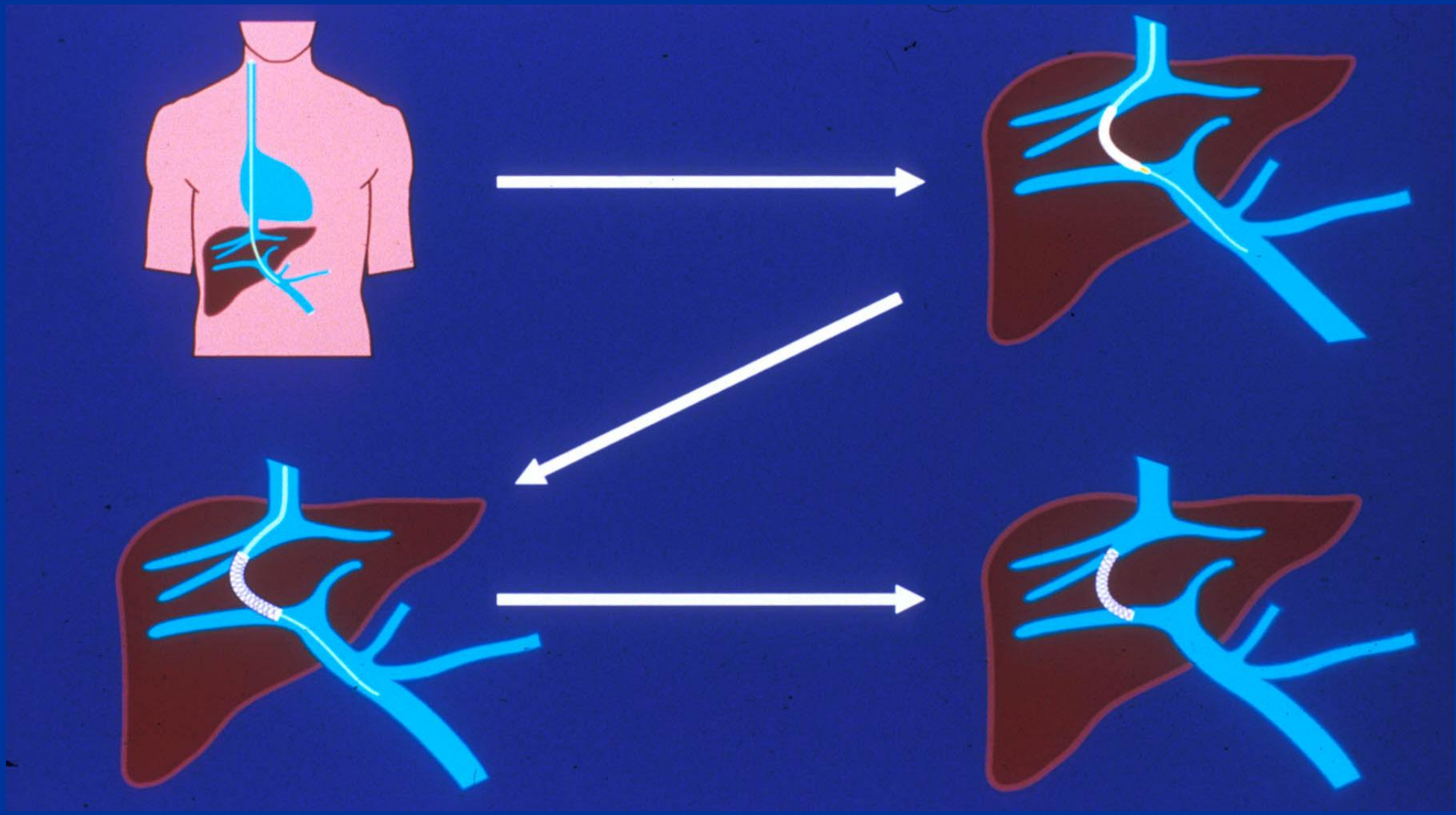
Sengstaken-Blakemore Tube

- Purpose - to occlude venous flow
- 3 and 4 lumen pediatric tubes
  - gastric port
  - gastric balloon
  - esophageal balloon
  - (esophageal port)
- 3 lumen tubes require esophageal drainage with a second tube
- Endotracheal intubation
- Requires fixation with weights or a football helmet

Emergency or Elective TIPSS

- Transvenous (jugular) intrahepatic portosystemic shunt performed by interventional radiologists
- Technique
  - Access the hepatic veins
  - Perforate through to the portal veins
  - Dilate the channel
  - Place a stent and then dilate the stent
- Advantages
  - Can be performed emergently without surgery
  - Does not require surgery of prehepatic vessels
- Disadvantage - may occlude

Transjugular Intrahepatic Portosystemic Shunt

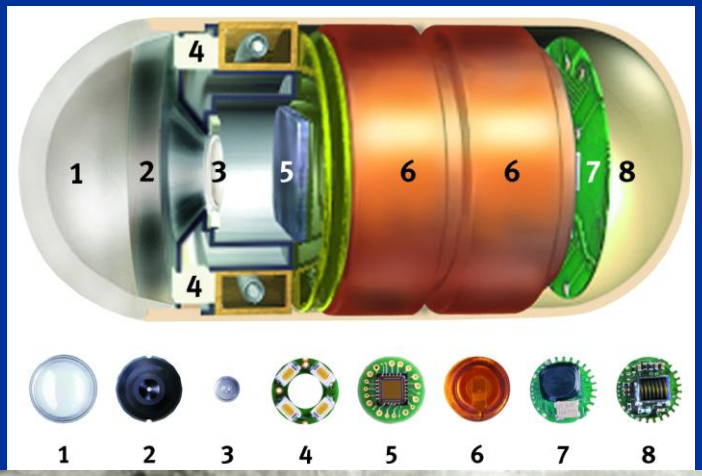


Portosystemic Surgical Shunts

- Techniques
  - Portocaval shunt
  - H-type mesocaval shunt
  - Distal or proximal splenorenal shunt
- Disadvantages
  - Major surgery
  - Rearranges prehepatic vessels pre-transplant
  - May clot with time
  - Difficult or impossible in small infants
  - Post-shunt encephalopathy may occur

Endoscopic Therapies for Other Sources of GI Bleeding

- Injection
- Coagulate
  - Heater probe
  - Bipolar probe
  - Laser coagulator
- Mechanical
  - Hemoclips
  - Banding



Long term Management of Esophageal Varices

Endoscopic Therapy

- Sclerotherapy or banding
- More frequently at first to eradicate varices
- Repeat scope at intervals to prevent bleeding

Medical management

- β-blockers can decrease portal pressure
- Not used in infants and young children

Other management

- IR or surgical portosystemic shunts
- Transplantation

More unanswered questions!

- Why does fibrosis happen in CHF?
  - Different than other liver diseases
- When will varices bleed?
  - Not all varices will bleed
  - When should we intervene?
- How can we treat and reverse fibrosis?
  - Many drugs under study in adults
- How can we measure portal hypertension?

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Subharmonic Aided Pressure Estimation (SHAPE)

- New study starting here at CHOP in 2019
- Using special ultrasound technique with IV contrast to measure portal pressures
  - Will study patients with and without portal hypertension
  - Will study whether the SHAPE technique can determine portal pressures associated with risk of bleeding or other complications
- Stay tuned!

Summary

- Patients with CHF often develop portal hypertension
- Some patients will develop complications, most commonly GI bleeding.
- We have effective therapies for acute treatment and long term management
- If these therapies are ineffective, surgical shunts or liver transplantation may be required.
- Ongoing research will lead to novel therapies for fibrosis and portal hypertension