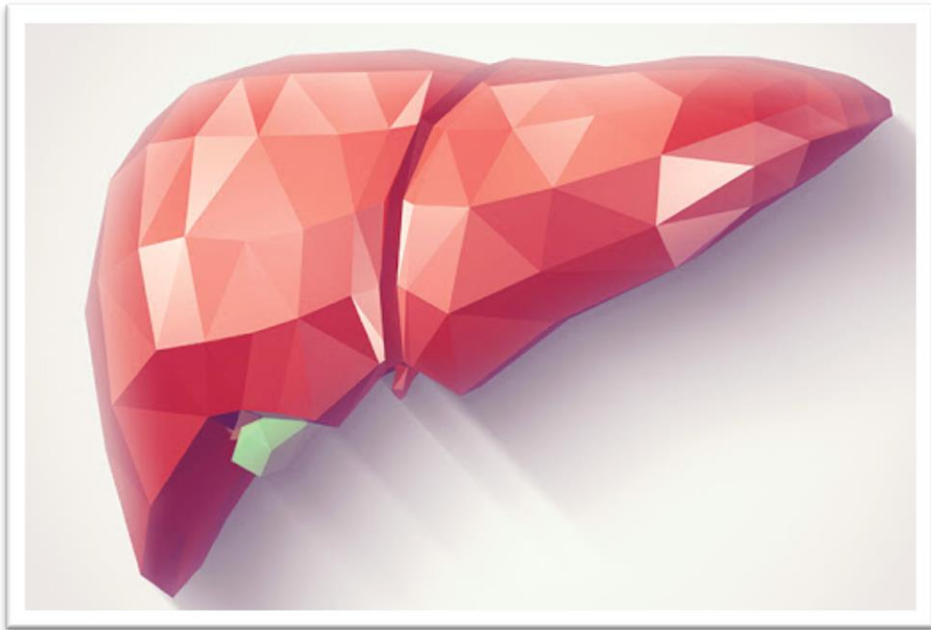


PORTAL HYPERTENSION



Department
of internal diseases

Used material: https://www.amboss.com/us/knowledge/Portal_hypertension and other.
Look at the information sources on the slides

Definition

- Portal hypertension is defined as a portal venous pressure of ≥ 6 mm Hg.
- Portal venous pressure > 10 mm Hg is clinically significant and > 12 mm Hg is associated with complications.

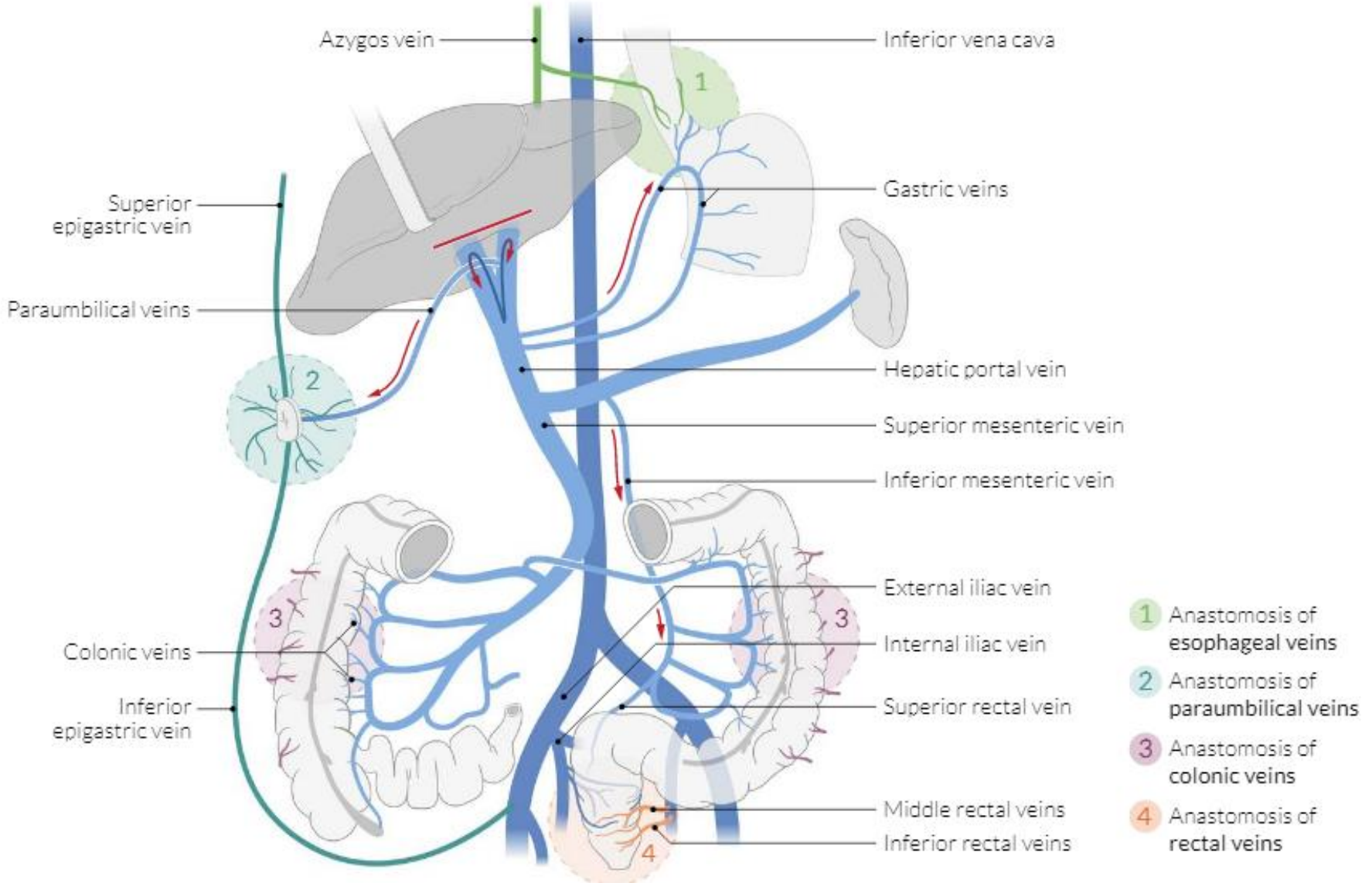
Etiology

- **Prehepatic**
 - Portal vein thrombosis
 - Splenic vein thrombosis
 - Hepatosplenic schistosomiasis
 - Atresia of portal vein
 - Phlebitis of portal vein
- **Intrahepatic**
 - Cirrhosis including fibrous proliferation (most common cause of portal hypertension in the US)
 - Massive hepatic metastases or tumors
- **Posthepatic**
 - Budd-Chiari syndrome
 - Right-sided heart failure
 - Constrictive pericarditis

Portocaval anastomoses

- Depending on the cause, portal [hypertension](#) may be either acute or chronic. Acute portal [hypertension](#) arises from acute [portal vein thrombosis](#), while chronic portal [hypertension](#) may be due to chronic [thrombosis](#), [cirrhosis](#), or [schistosomiasis](#).
- Signs and symptoms of the underlying disease (e.g., [cirrhosis](#), [right-sided heart failure](#))
- **↑ Blood flow via [portosystemic anastomoses](#)**
 - Via paraumbilical veins and epigastric [veins](#) → **caput medusae**
 - Via rectal [veins](#) → **hemorrhoidal or anorectal varices**
 - Via [veins](#) of the [gastric fundus](#) and [distal](#) 1/3 of the [esophagus](#), leading to:
 - **Esophageal varices**: risk of life-threatening [esophageal variceal bleeding](#) (hematemesis)
 - **Gastric varices**: [melena](#)
 - Consequences: impaired [liver](#) function (see [cirrhosis](#))
- **Congestive [splenomegaly](#)**, followed by signs of [hypersplenism](#) (e.g., [thrombocytopenia](#))
- [Upper gastrointestinal bleeding](#) from [portal hypertensive gastropathy](#), [gastrointestinal ulcers](#), or diffuse [lower gastrointestinal bleeding](#)
- **Transudative [ascites](#)**

Portocaval anastomoses



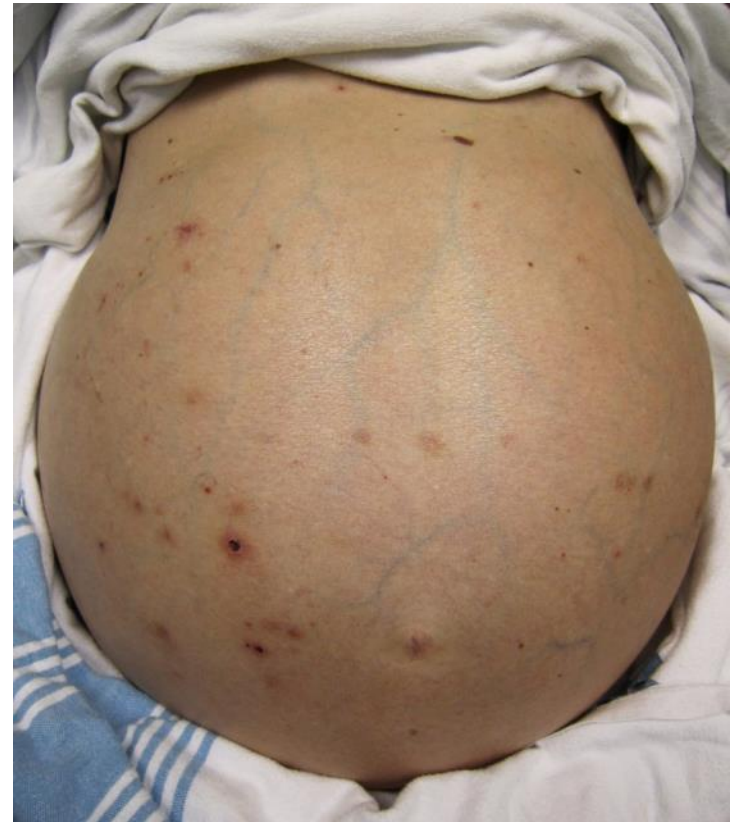
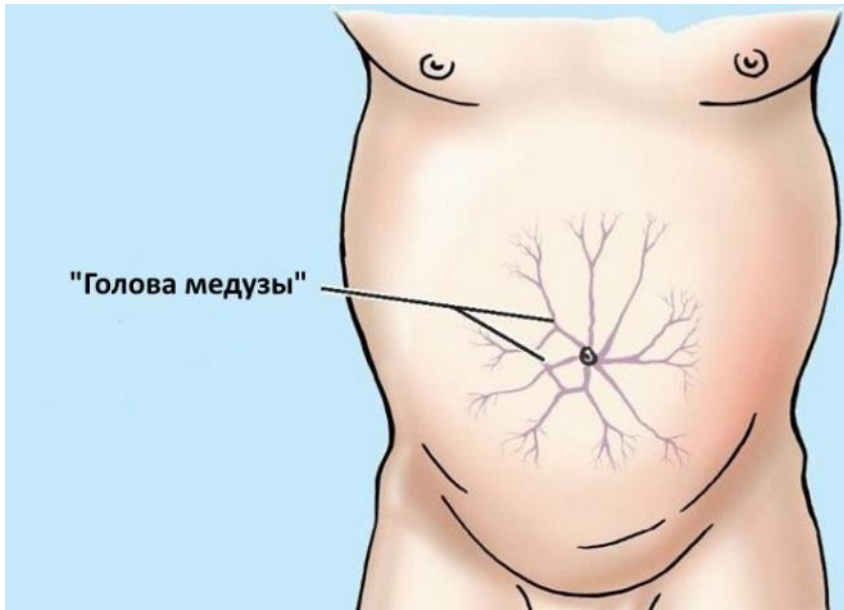
Portocaval anastomoses

- Portocaval anastomoses are anastomoses between veins of the portal venous system (veins that drain directly/indirectly into the portal vein) and veins of the caval venous system (veins that drain directly/indirectly into the superior or inferior vena cava).
- Physiologically, there is minimal/no blood flow through these anastomoses. In portal hypertension, increased portal venous pressure causes shunting of blood from the portal system into the caval system, which increases the pressure within the caval veins, causing variceal dilation of the caval veins at the sites of portocaval anastomoses.
- (1) Distal esophagus: anastomosis between the left gastric vein (portal system) and the azygous vein (caval system), which manifests as esophageal varices in portal hypertension

Portocaval anastomoses

- (2) Paraumbilical region: anastomosis between the paraumbilical veins (portal system) and the superior and inferior epigastric veins (caval system), which manifests as caput medusae in portal hypertension
- (3) Colon: anastomosis between the colonic veins (portal system) and the retroperitoneal veins (caval system), which manifests as colonic varices in portal hypertension
- (4) Rectum and anal canal: anastomosis between the superior rectal vein (portal system) and the middle and inferior rectal veins (caval system), which manifests as anorectal varices in portal hypertension

Portocaval anastomoses



There is distention of the abdomen (ascites) and periumbilical dilatation of subcutaneous veins (caput medusae).

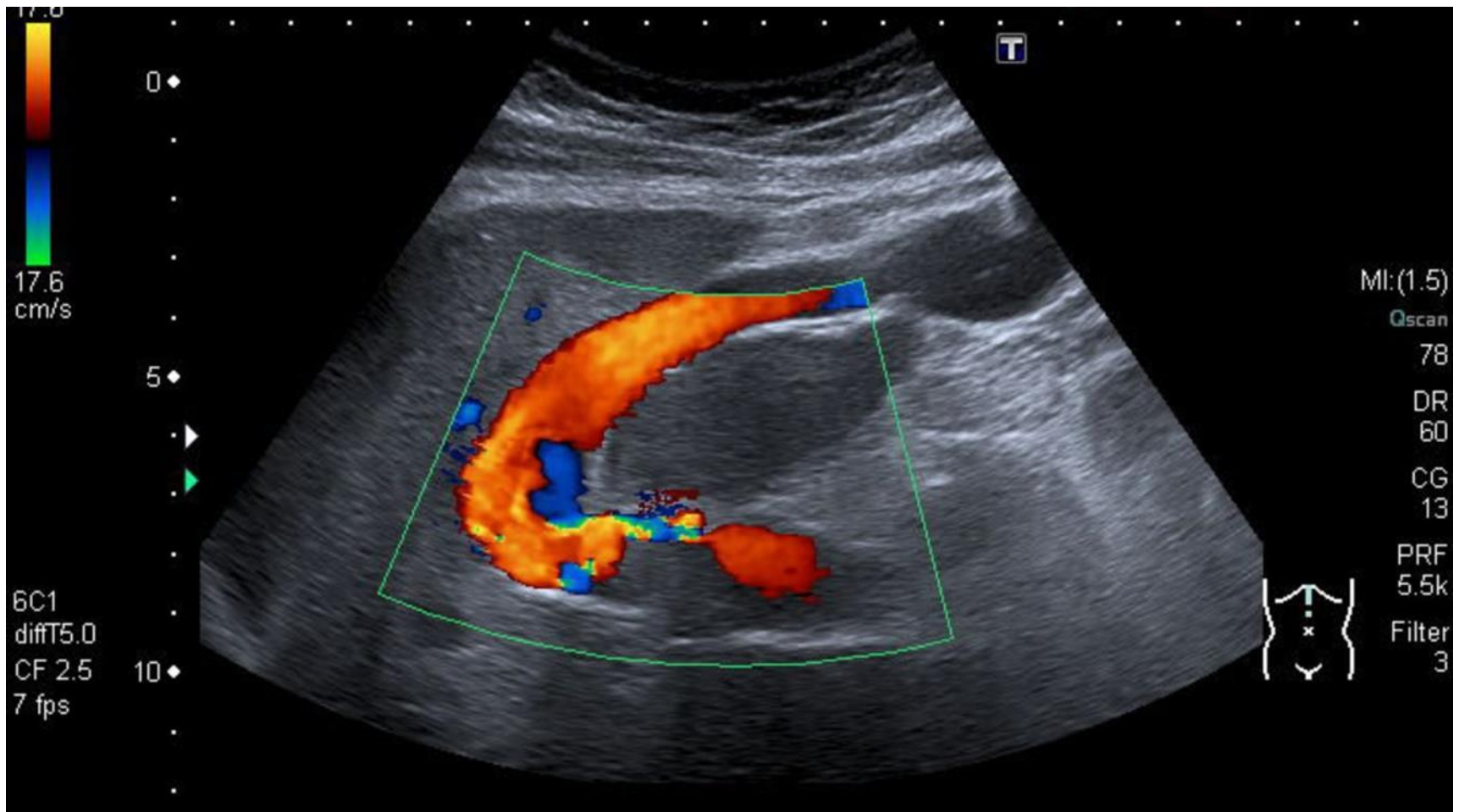
There are partially erosive, erythematous maculopapules (prurigo simplex).

Diagnosics

- Clinical manifestations of portal [hypertension](#) (e.g., [ascites](#)) in a patient with a known risk factor (e.g., [cirrhosis](#)) may already suffice for diagnosis. In addition to investigating underlying conditions, diagnostic steps may include:
- **Ultrasound**
 - **Specific findings:** on duplex ultrasonography, visualization of a **cavernous transformation** of the [portal vein](#) indicates (chronic) [portal vein thrombosis](#)
 - **Unspecific finding:** [portal vein](#) dilated to > **13 mm**
 - **Indirect indications**
 - Visualization of ↑ blood flow via [portosystemic anastomoses](#) using duplex ultrasonography
 - [Splenomegaly](#)
 - [Ascites](#)
- **Abdominal CT:** [portal vein thrombosis](#) is visible
- **Esophagogastroduodenoscopy (EGD):** assessment of and, if necessary, treatment of [esophageal varices](#)

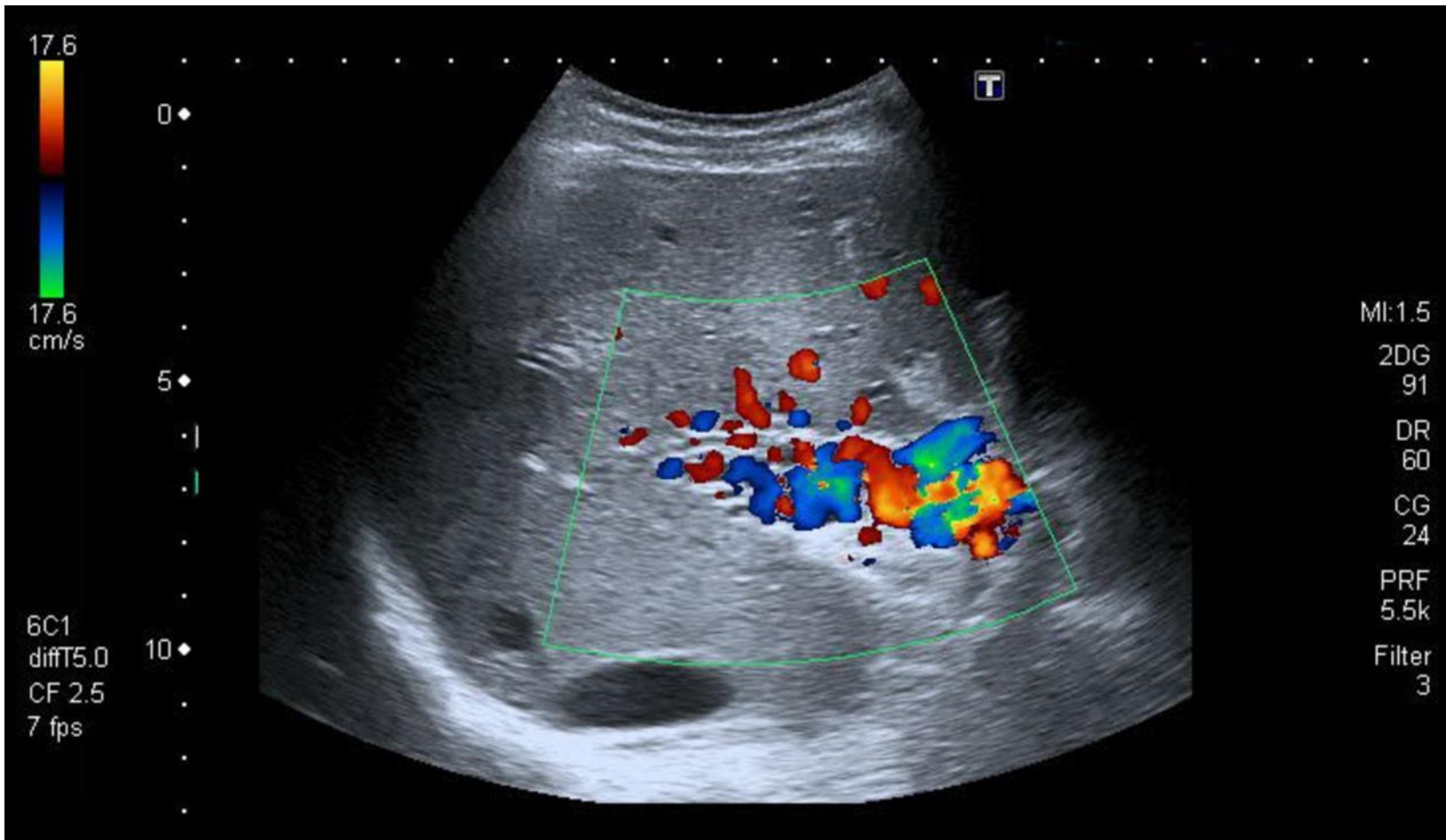
Carale J. Portal Hypertension. In: *Portal Hypertension*. New York, NY: WebMD. <http://emedicine.medscape.com/article/182098>. Updated November 20, 2016. Accessed March 29, 2017.

Recanalized umbilical vein in liver cirrhosis



The Doppler ultrasound depiction of the umbilical vein shows reopening of the vessel with pronounced blood flow from the liver towards the umbilicus as a sign of shunt formation in portal hypertension (Cruveilhier-Baumgarten syndrome).

Cavernous transformation of the portal vein

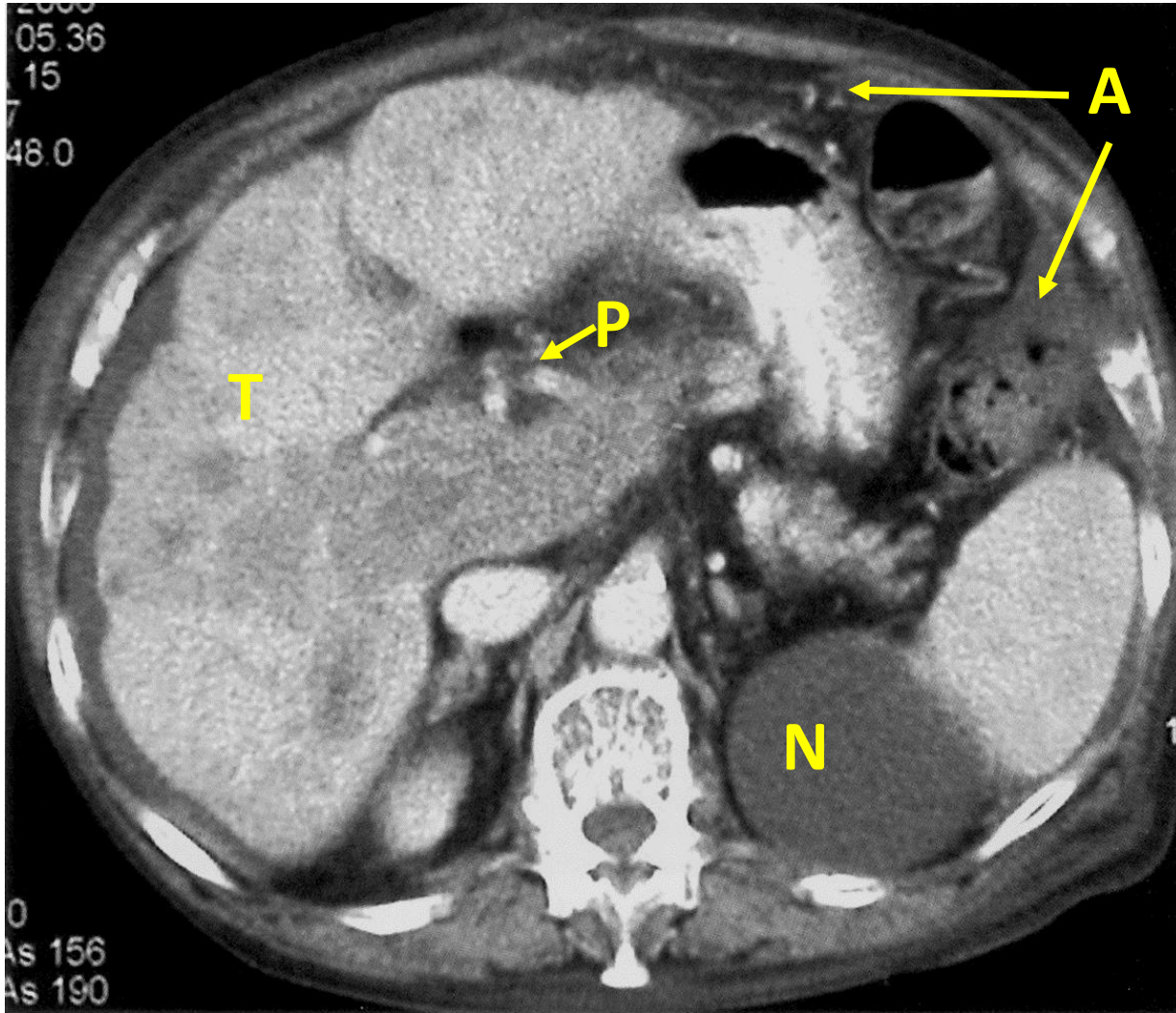


Color-coded duplex ultrasonography of the portal vein

There is a sponge-like network of collateral veins leading to (hepatopetal) and from (hepatofugal) the liver.

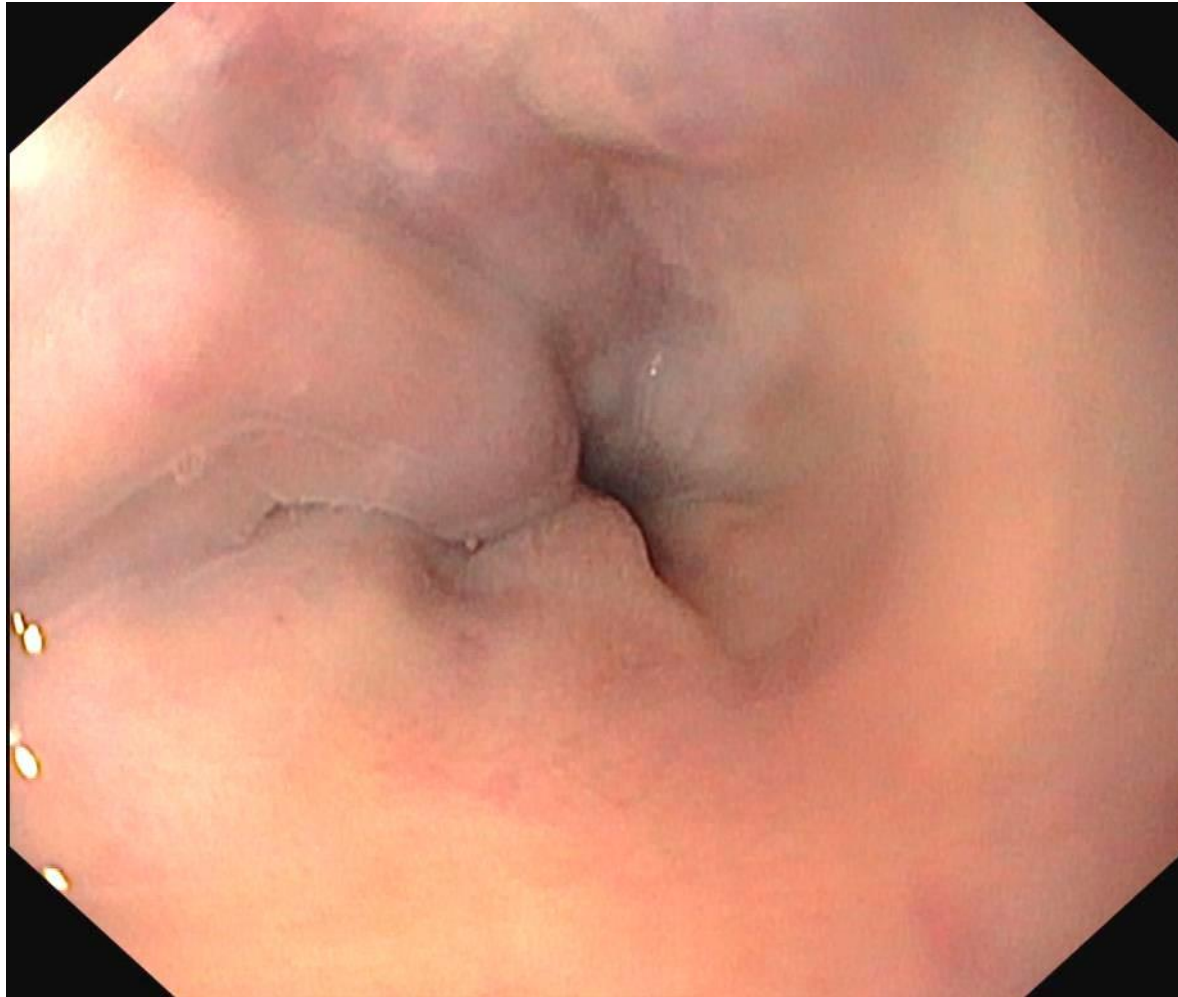
Cavernous transformation of the portal vein is a sequela of portal vein thrombosis. Following thrombosis, if the portal vein does not recanalize or does so only partially, collateral paraspportal veins may develop.

Abdominal CT scan of hepatocellular carcinoma in liver cirrhosis



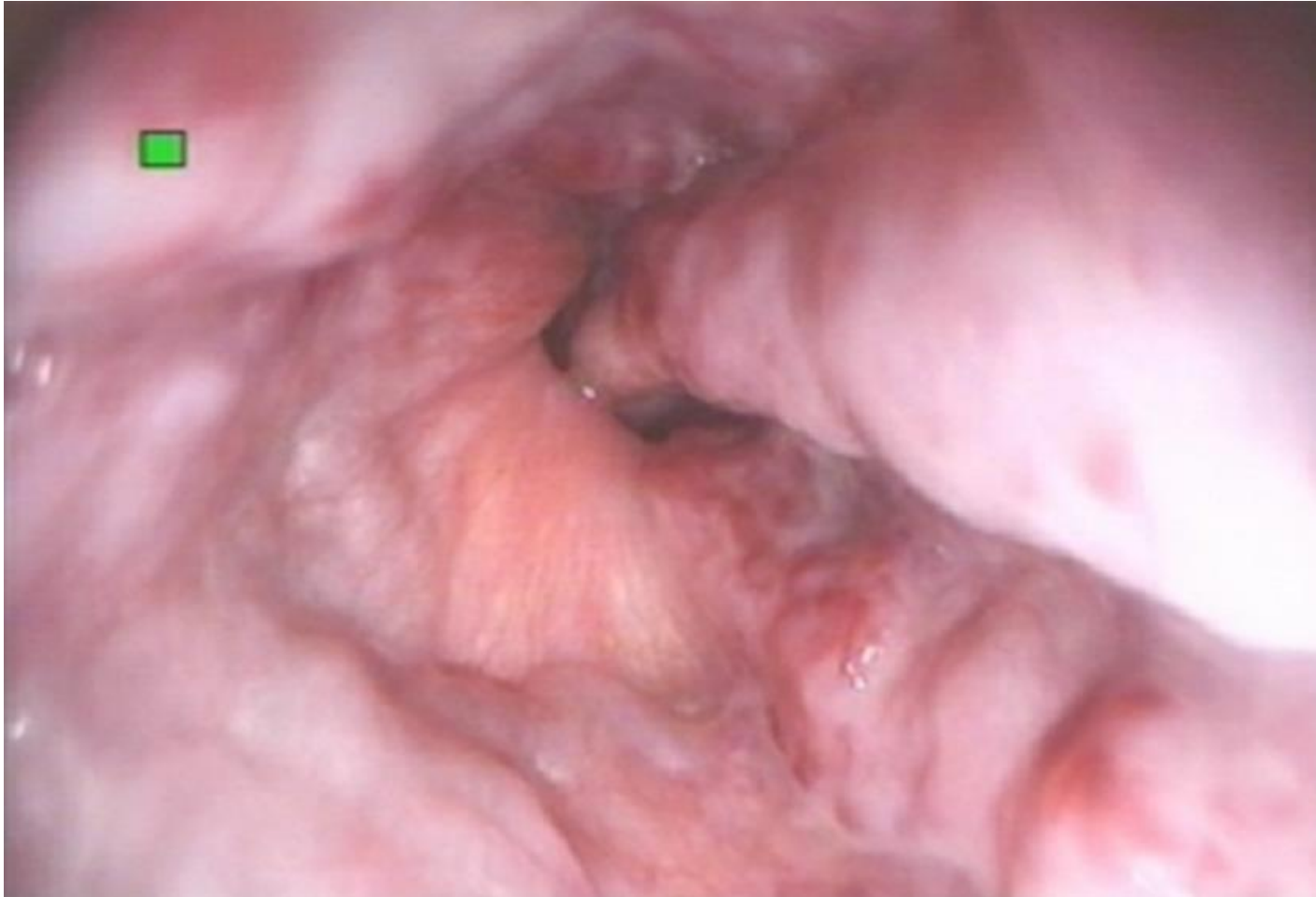
Axial abdominal CT scan with contrast medium: tumorous liver in hepatocellular carcinoma (T), liver cirrhosis, portal vein thrombosis (P), ascites (A), renal cyst (N).

Esophageal varices



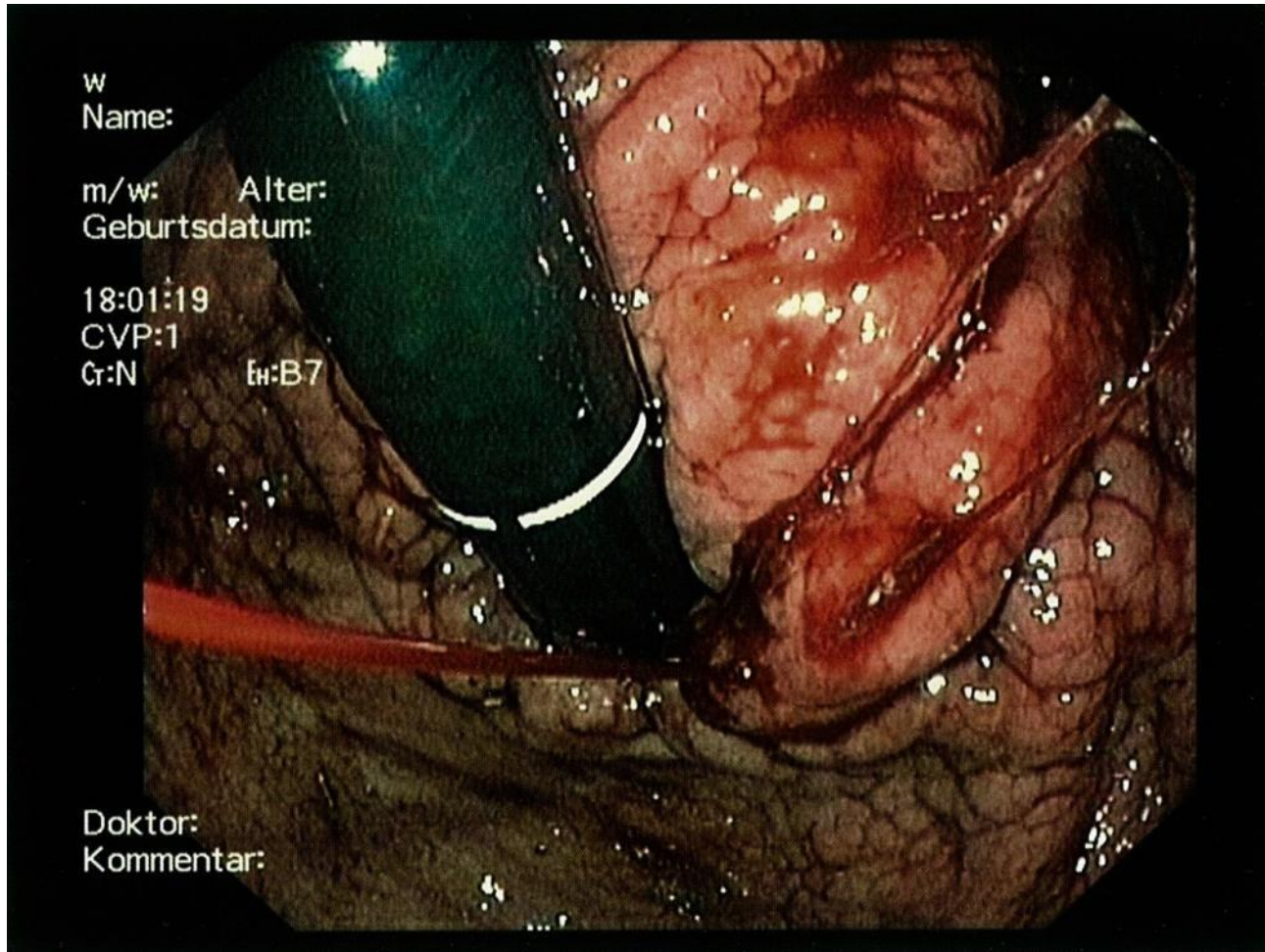
Dilated submucosal veins narrow the esophageal lumen (green overlay). The varices cannot be compressed with insufflation (Paquet grade II).

Esophageal varices



Dilated submucosal veins (varices) are ubiquitous, most prominently visible at the 1, 4, 7, and 11 o'clock positions of the esophageal lumen. Cherry-red spots, a marker of varices at risk of hemorrhage, can be seen on the variceal surface.

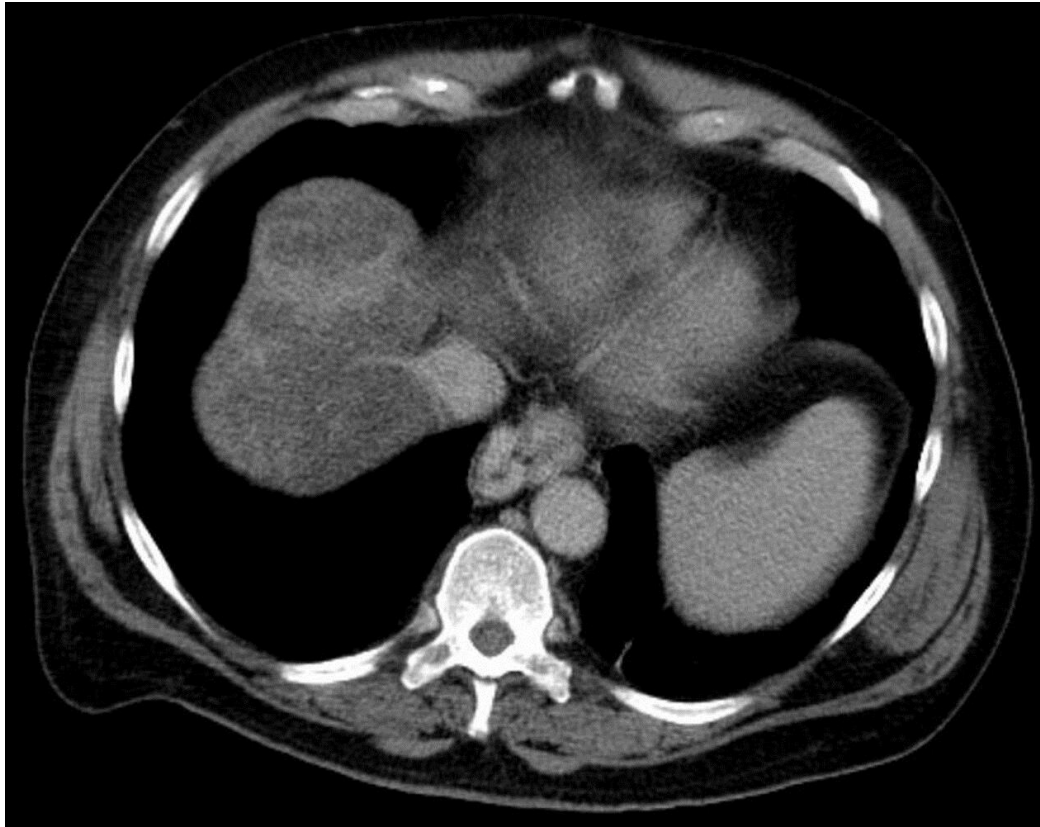
Fundal variceal hemorrhage in liver cirrhosis



Spurting hemorrhage (source at the center, jet running left) from a fundal varix

Esophageal varices

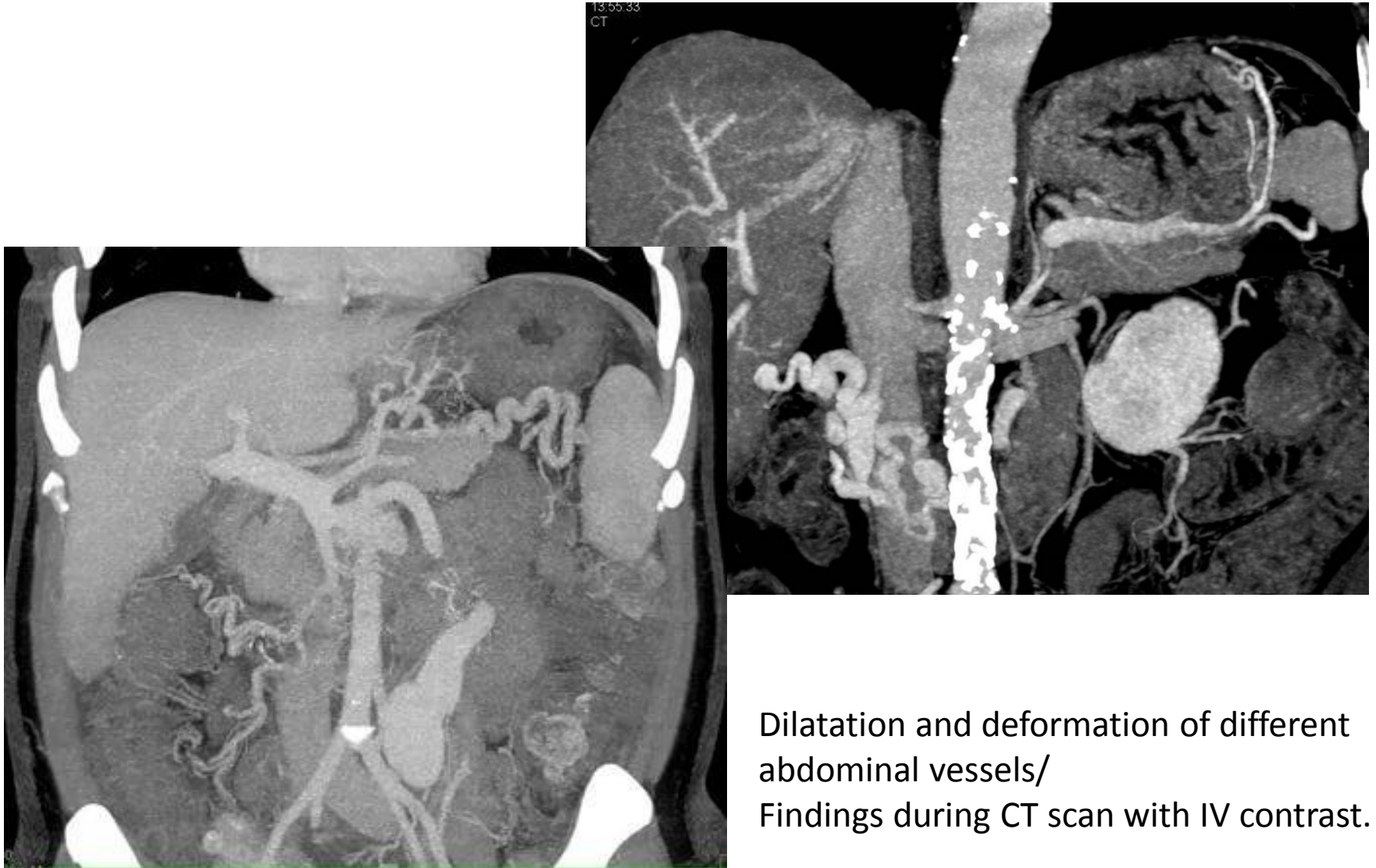
CT chest (oral and IV contrast-enhanced; axial section)



A bulbous, contrast-enhancing mass, not clearly separated from the esophageal wall, can be seen (green overlay). There is also a heterogeneous perfusion pattern (red overlay) seen in the liver, likely to be due to cirrhosis.

These findings are characteristic of portal hypertension with esophageal varices.

Portocaval anastomoses



Medical therapy

- **First-line medication:** nonselective [beta-blocker](#) (i.e., [propranolol](#) or [nadolol](#))
 - Effect: inhibits beta-2-adrenergic receptors in the [gastrointestinal tract](#) → splanchnic vasoconstriction → ↓ portal and collateral blood flow → ↓ portal [hypertension](#)
 - Also used to prevent variceal bleeding
 - At first line can be given beta-blockers with vasodilating effect
- [Beta-blocker](#) treatment in patients suffering from advanced [liver cirrhosis](#) ([Child class C](#)) may lead to circulatory dysregulation. If negative effects outweigh the benefits, [beta-blocker](#) treatment should be reconsidered!

Portosystemic shunts

- **Transjugular intrahepatic portosystemic shunt** ([TIPS](#) or [TIPSS](#))
 - **Indications**
 - Persistent, recurring, or treatment-resistant [upper gastrointestinal bleeding](#) resulting from portal [hypertension](#), e.g., from [esophageal varices](#)
 - Refractory [ascites](#)
 - Acute [thrombosis](#) of [portal vein](#)
 - Patients with [hepatorenal syndrome](#) who are not eligible for or are awaiting [liver transplantation](#) .
 - **Procedure**
 - A needle catheter inserted via the [internal jugular vein](#) → passed along to hepatic [vein](#) → pierced through [liver parenchyma](#) to intrahepatic branch of the [portal vein](#) → expandable metal stent is placed → side-to-side portocaval shunt
 - Assures blood drainage from the portal to the systemic system bypassing the [liver](#), thus lowering portal pressure

Sanyal AJ, Bajaj JS. Transjugular intrahepatic portosystemic shunts: Indications and contraindications. In: Post TW, ed. *UpToDate*. Waltham, MA: UpToDate. http://www.uptodate.com/contents/transjugular-intrahepatic-portosystemic-shunts-indications-and-contraindications?source=search_result&search=TIPS&selectedTitle=1~150. Last updated November 7, 2016. Accessed February 12, 2017.

Sanyal AJ. Primary and Pre-primary Prophylaxis Against Variceal Hemorrhage in Patients with Cirrhosis. In: Post TW, ed. *UpToDate*. Waltham, MA: UpToDate. <https://www.uptodate.com/contents/primary-and-pre-primary-prophylaxis-against-variceal-hemorrhage-in-patients-with-cirrhosis>. Last updated November 2, 2015. Accessed January 8, 2018.

Portosystemic shunts (part 2)

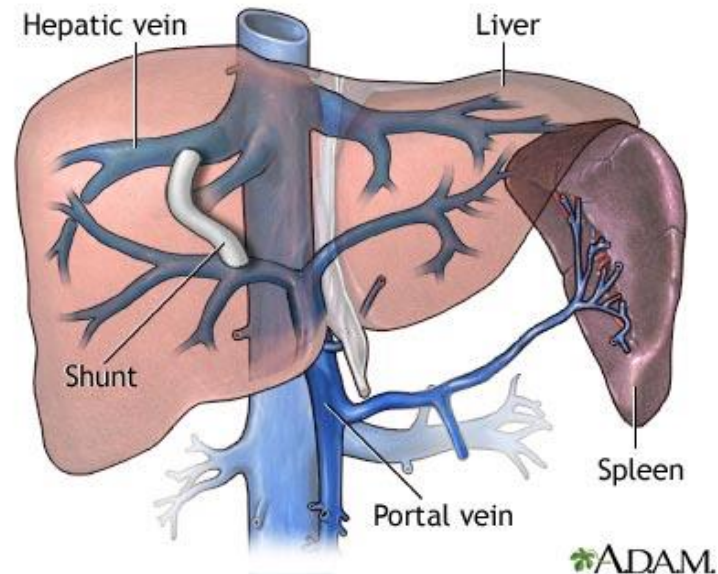
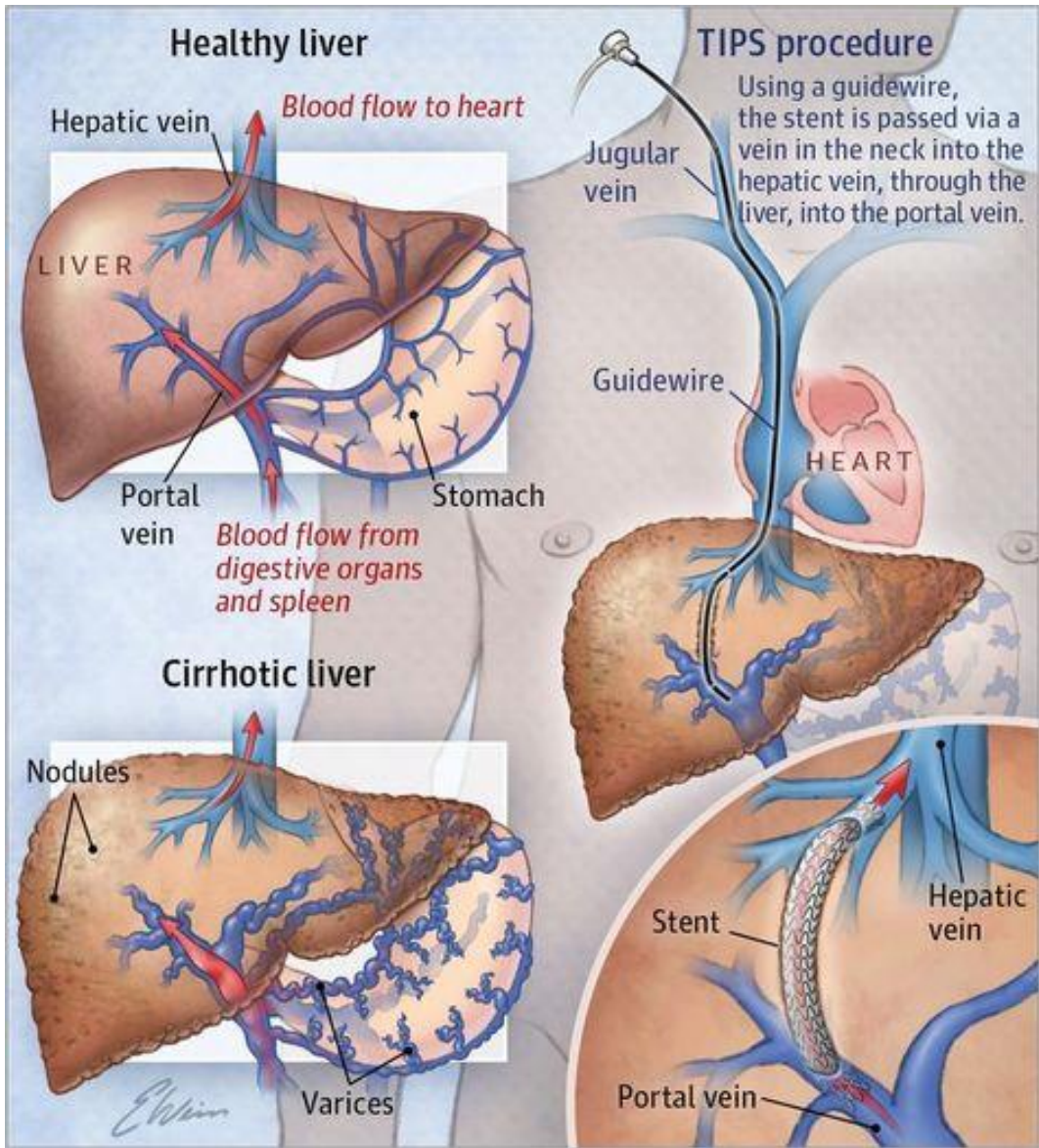
– Contraindications

- Pre-existing [hepatic encephalopathy](#) or [cirrhosis](#) (Child's class C): Shunt implementation results in reduced hepatic elimination of [ammonia](#) and worsening of encephalopathy.
- [Heart failure](#)
- Severe [pulmonary hypertension](#) > 45 mm Hg
- Uncontrolled systemic infection or [sepsis](#)
- [Hepatic cysts](#) or tumors
- Cavernous transformation of the [portal vein](#) following [thrombosis](#)
- **Total [portosystemic shunts](#)**: The [portal vein](#) is completely shunted to the vena cava, thereby reducing portal pressure.
- **Selective [portosystemic shunts](#)**: The [portal vein](#) is partially shunted to the vena cava, thereby reducing portal pressure. This partial shunt prevents varices while continuing to allow portal perfusion

Sanyal AJ, Bajaj JS. Transjugular intrahepatic portosystemic shunts: Indications and contraindications. In: Post TW, ed. *UpToDate*. Waltham, MA: UpToDate. http://www.uptodate.com/contents/transjugular-intrahepatic-portosystemic-shunts-indications-and-contraindications?source=search_result&search=TIPS&selectedTitle=1~150. Last updated November 7, 2016. Accessed February 12, 2017.

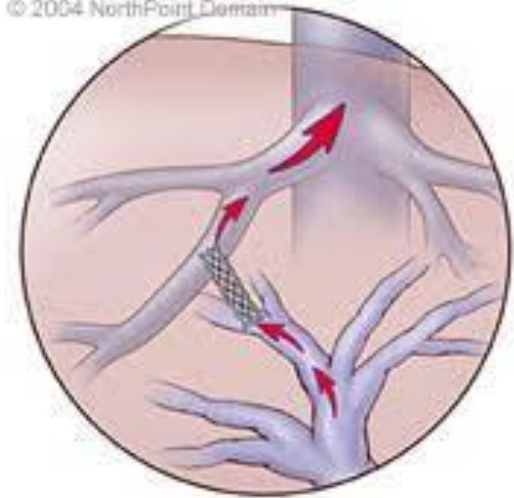
Sanyal AJ. Primary and Pre-primary Prophylaxis Against Variceal Hemorrhage in Patients with Cirrhosis. In: Post TW, ed. *UpToDate*. Waltham, MA: UpToDate. <https://www.uptodate.com/contents/primary-and-pre-primary-prophylaxis-against-variceal-hemorrhage-in-patients-with-cirrhosis>. Last updated November 2, 2015. Accessed January 8, 2018.

Transjugular intrahepatic portosystemic shunt



ADAM.

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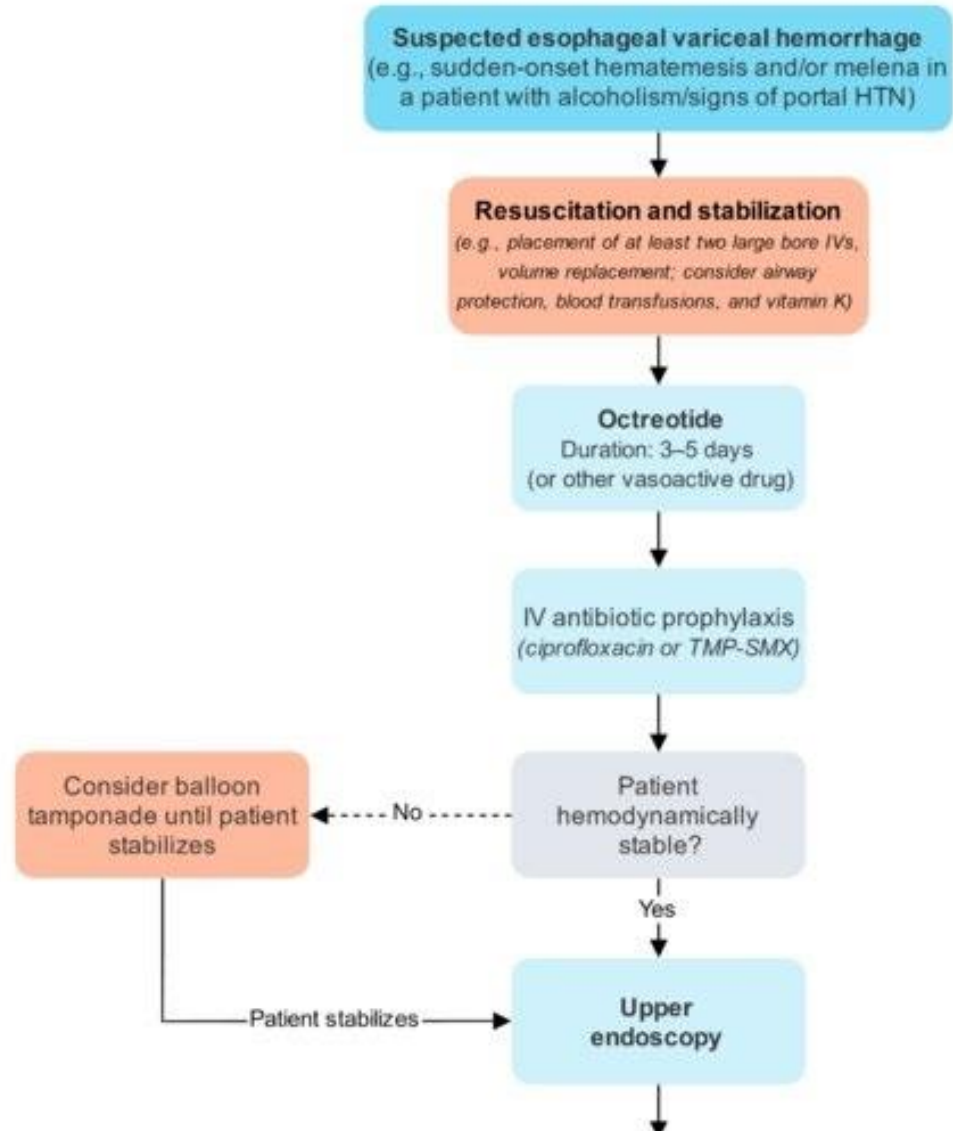


Metal stent in tunnel provides new blood flow and reduces pressure in varices

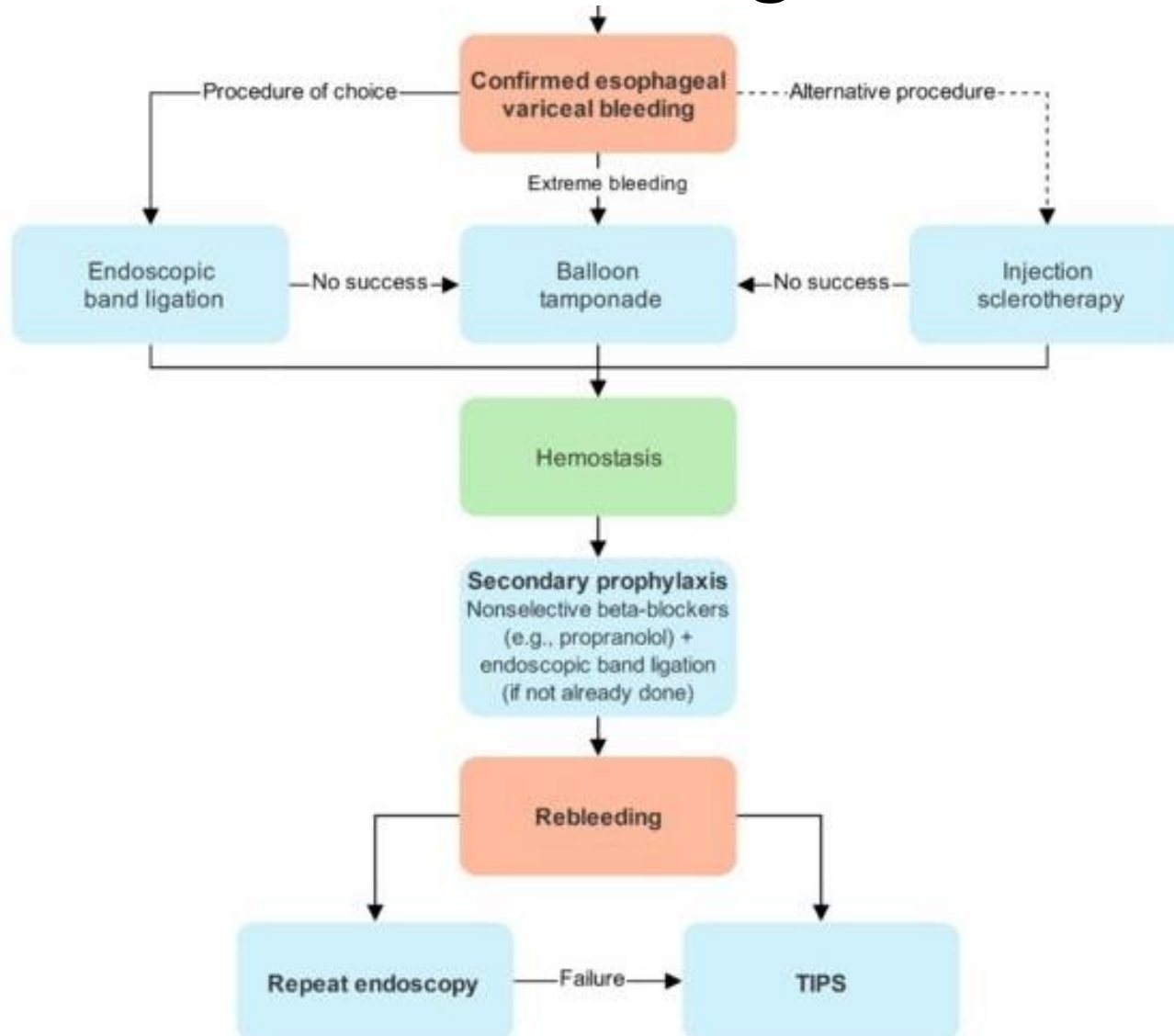
Complications

- **Esophageal variceal hemorrhage**
- **Definition**
- [Esophageal variceal hemorrhage](#) refers to the bleeding of dilated sub-mucosal [veins](#) (varices) of the [distal esophagus](#) and is a dangerous consequence of portal [hypertension](#). It is the most common form of upper gastrointestinal (GI) bleeding in patients presenting with [cirrhosis](#).
- **Clinical features**
- Symptoms of [upper GI bleeding](#) (see [clinical features of gastrointestinal bleeding](#)): signs of circulatory insufficiency, **hematemesis**, [melena](#), and/or **hematochezia**
- **Clinical diagnosis**
- Sudden onset of severe [upper GI bleeding](#) in a patient with signs of portal [hypertension](#), typically in combination with [liver](#) failure
- If bleeding occurs **following retching or vomiting**, consider a [Mallory-Weiss](#) tear as a differential diagnosis.

Acute management of variceal hemorrhage



Acute management of variceal hemorrhage



Acute management of variceal hemorrhage

- **Resuscitation and stabilization**

- Place (at least) two peripheral venous catheters
- Substitute [crystalloids](#) to maintain plasma volume
- Intubate patient (with decreased consciousness) to prevent the possibility of [aspiration](#) or [airway](#) obstruction
- Transfuse blood or blood products to stabilize coagulation, if indicated

- **Medical therapy**

- **Octreotide** for 3–5 days → inhibits secretion of vasodilative
- [hormones](#), e.g., glucagon → indirect splanchnic vasoconstriction → reduces splanchnic blood flow
- [Pituitrine](#) as vasopressor (as synthetic vasopressin leads to decreasing of pressure in esophageal veins and constriction of esophageal muscles)
- [Vitamin K](#) is indicated for patients with [coagulation disorders](#)
- Antibiotic prophylaxis for 7 days: prevention of infections (SBP, [urinary tract infection](#), [pneumonia](#)) and of [septic](#) complications due to [bacteremia](#), as well as [colon](#) decontamination to reduce [ammonia](#) production of gut flora
 - First-line: IV [ceftriaxone](#)
 - Alternative: oral [ciprofloxacin](#)

Acute management of variceal hemorrhage

- **Endoscopic management**
 - [Erythromycin](#) (a strong prokinetic agent) may be administered before gastroscopy.
 - **Procedures**
 - **Endoscopic band ligation** (procedure of choice)
 - [Hemostasis](#)
 - Used for primary prophylaxis and prevention of recurring hemorrhage
 - Alternative: **injection sclerotherapy**, absolute alcohol, and [fibrin](#) glue, as well as cyanoacrylate, to stop acute variceal bleeding
 - Balloon tamponade using a Sengstaken-Blakemore tube or Minnesota tube
 - Indication: alternative treatment in case of extreme hemorrhage, unsuccessful endoscopic treatment, or ineffective hemostatic medication; consider in hemodynamically unstable patients until they can be stabilized
 - Complication: risk of [decubitus gangrene](#) → prevention: deflation of balloon every 5 hours for 5 minutes
- **Interventional radiologic treatment:** See “[TIPS](#)” above.

Bleeding prevention

- **Primary prophylaxis**

- Medication to lower portal pressure, irrespective of variceal grading: nonselective [beta-blockers](#) (e.g., [propranolol](#), [nadolol](#))
- **Endoscopic esophageal [variceal ligation](#)**: only indicated for patients with a high risk of bleeding ([cirrhosis](#) Child B–C, [coagulopathy](#) or varices Paquet grade II)

- **Secondary prophylaxis**

- Combine [endoscopic variceal ligation](#) and medication (e.g., [propranolol](#)) to reduce portal pressure for residual varices
- If bleeding occurs despite secondary prophylaxis → placement of [TIPS](#)

Other complications

- Portal hypertensive gastropathy: which can cause [gastrointestinal ulcers](#) or diffuse [lower gastrointestinal bleeding](#).
- [Ascites](#)
- Spontaneous bacterial peritonitis
- [Hepatorenal syndrome](#)
- [Pulmonary complications of portal hypertension](#)
- [Cardiac cirrhosis](#)
- TIPS operation by itself will lead to hepatic encephalopathy (due to direct flowing of blood from intestine, that contain toxins including NO, into systemic blood flow)

Bajaj JS, Sanyal AJ. Methods to achieve hemostasis in patients with acute variceal hemorrhage. In: Post TW, ed. *UpToDate*. Waltham, MA: UpToDate. http://stage0www.uptodate.com/contents/methods-to-achieve-hemostasis-in-patients-with-acute-variceal-hemorrhage?source=search_result&search=octreotid&selectedTitle=11~150. Last updated November 12, 2015. Accessed February 12, 2017.

Reynaert H, Geerts A. Pharmacological rationale for the use of somatostatin and analogues in portal hypertension. *Aliment Pharmacol Ther*. 2003; 18(4): pp. 375–386. pmid: [12940922](#).

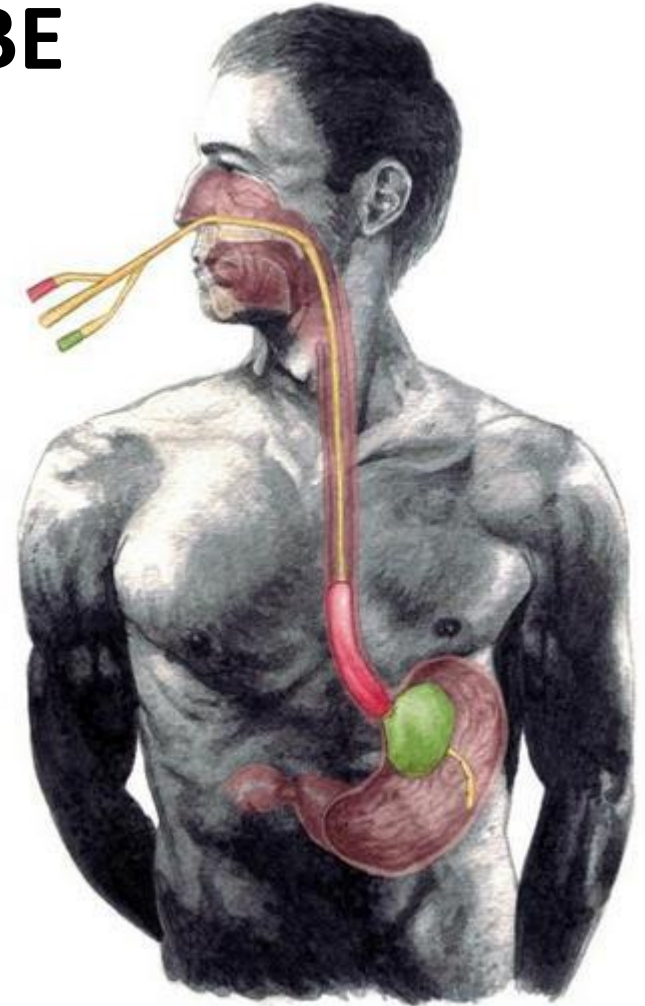
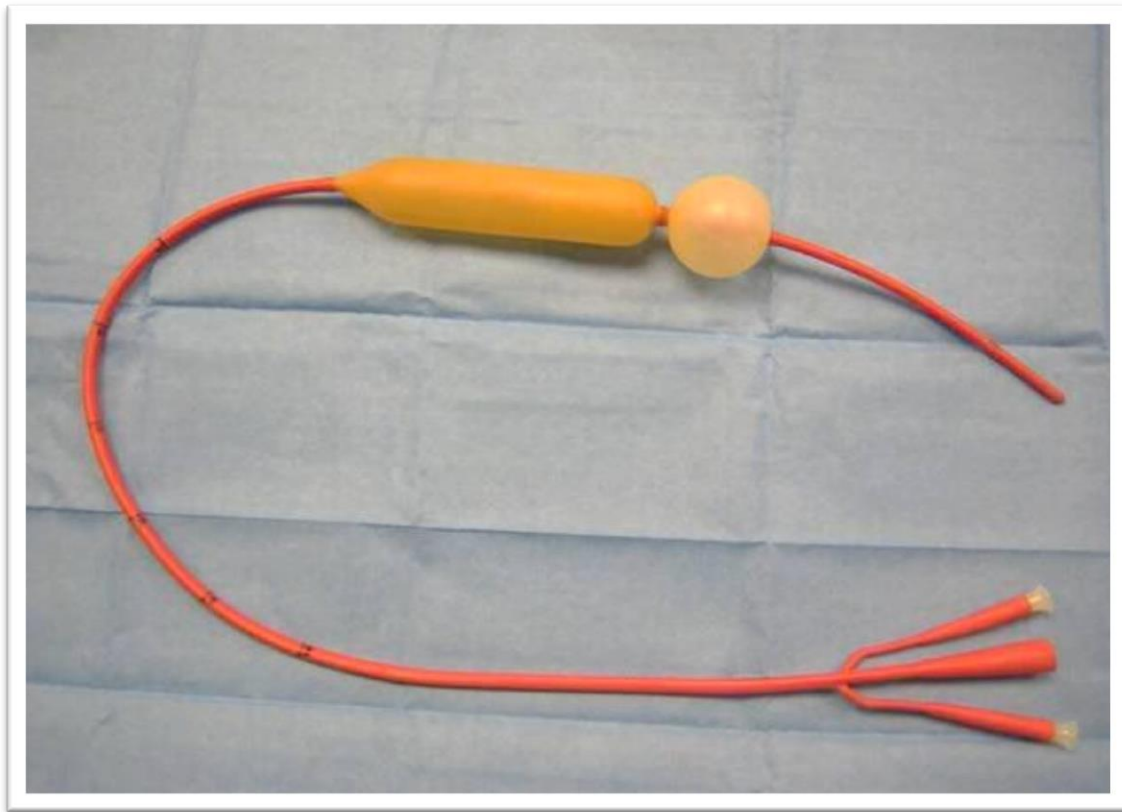
Sanyal AJ. General principles of the management of variceal hemorrhage. In: Post TW, ed. *UpToDate*. Waltham, MA: UpToDate. <http://www.uptodate.com/contents/general-principles-of-the-management-of-variceal-hemorrhage#H10>. Last updated December 22, 2016. Accessed February 12, 2017.

Questions for self preparation:

- 1. Syndrome of hypersplenism
- 2. Types of shunt operation in case of portal hypertension (selective, non-selective)
- 3. Tanners operation in case of esophageal varices
- 4. Grades of ascites
- 5. Blackmore probe

Extra

CORRECT STATEMENT OF THE BLACKMORE PROBE



Child–Pugh scoring system

	Points		
	1	2	3
Encephalopathy (grade)	None	1–2	3–4
Ascites	None	Slight	Moderate
Albumin (g/dL)	>3.5	2.8–3.5	<2.8
Prothrombin time prolonged (sec) or INR	<4 <1.7	4–6 1.7-2.3	>6 >2.3
Bilirubin (mg/dL)	<2	2–3	>3
for primary biliary cirrhosis	<4	4–10	>10

Class A:
5–6 points

Class B:
7–9 points

Class C:
10–15 points

SEGMENTS OF THE LIVER BY COUINAUD (1957)

Right lobe

Left lobe

Right (part of) liver

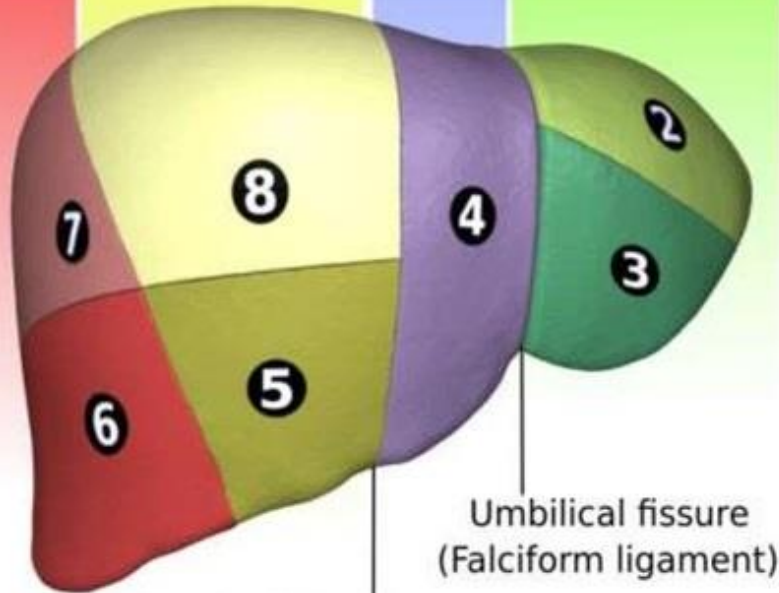
Left (part of) liver

Right posterior section

Right anterior section

Left medial section

Left lateral section



Cantlie's line

Anterior view

Left lobe

Right lobe

Left (part of) liver

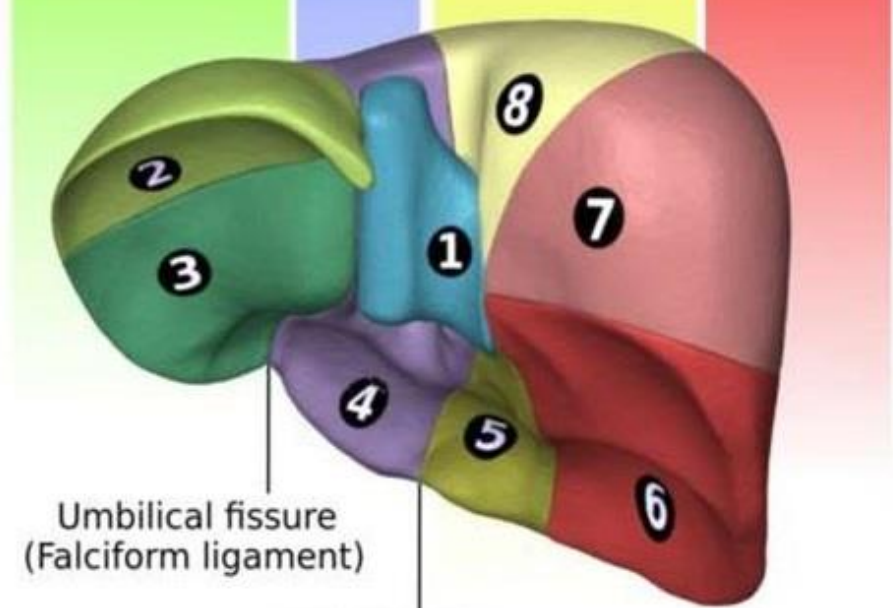
Right (part of) liver

Left lateral section

Left medial section

Right anterior section

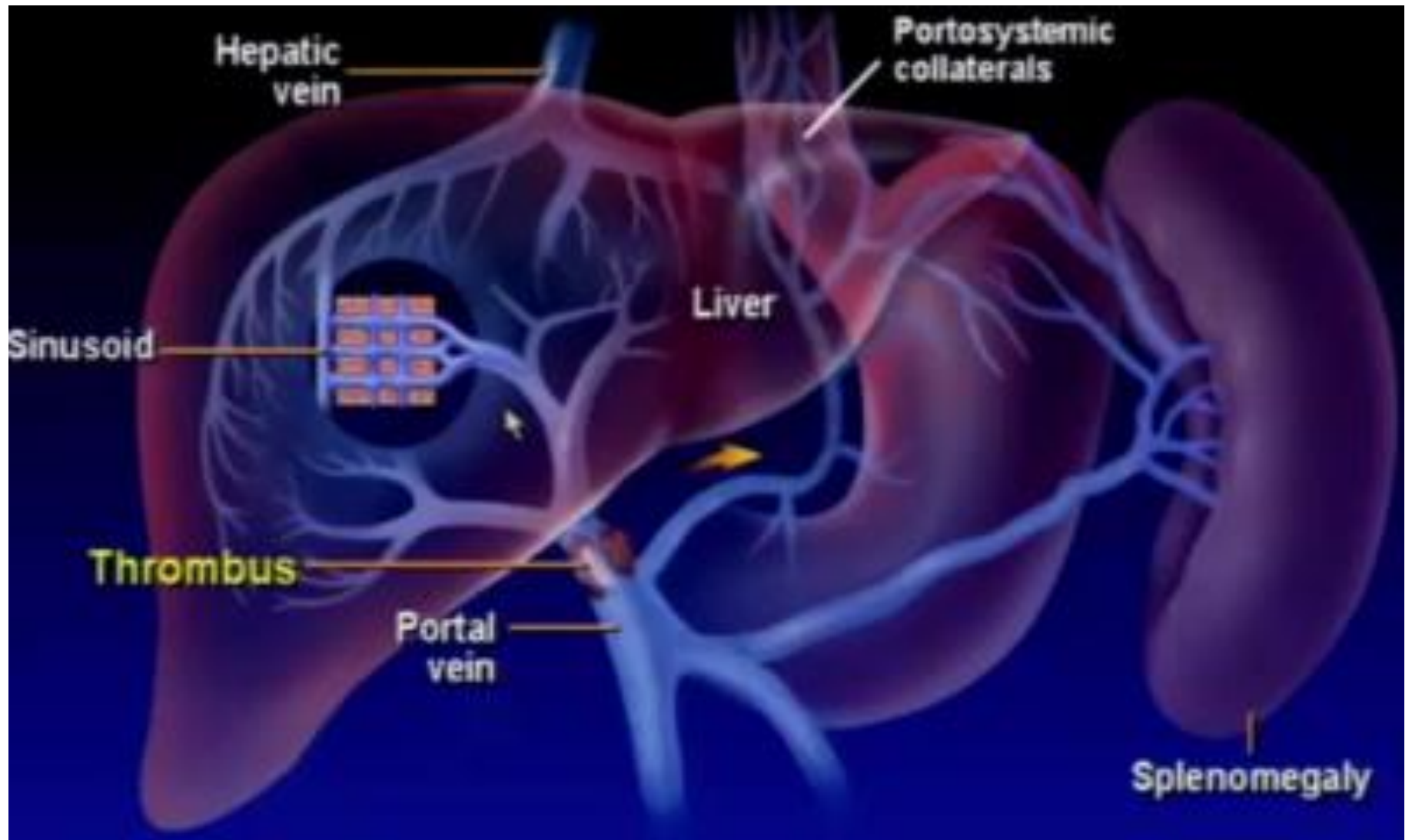
Right posterior section

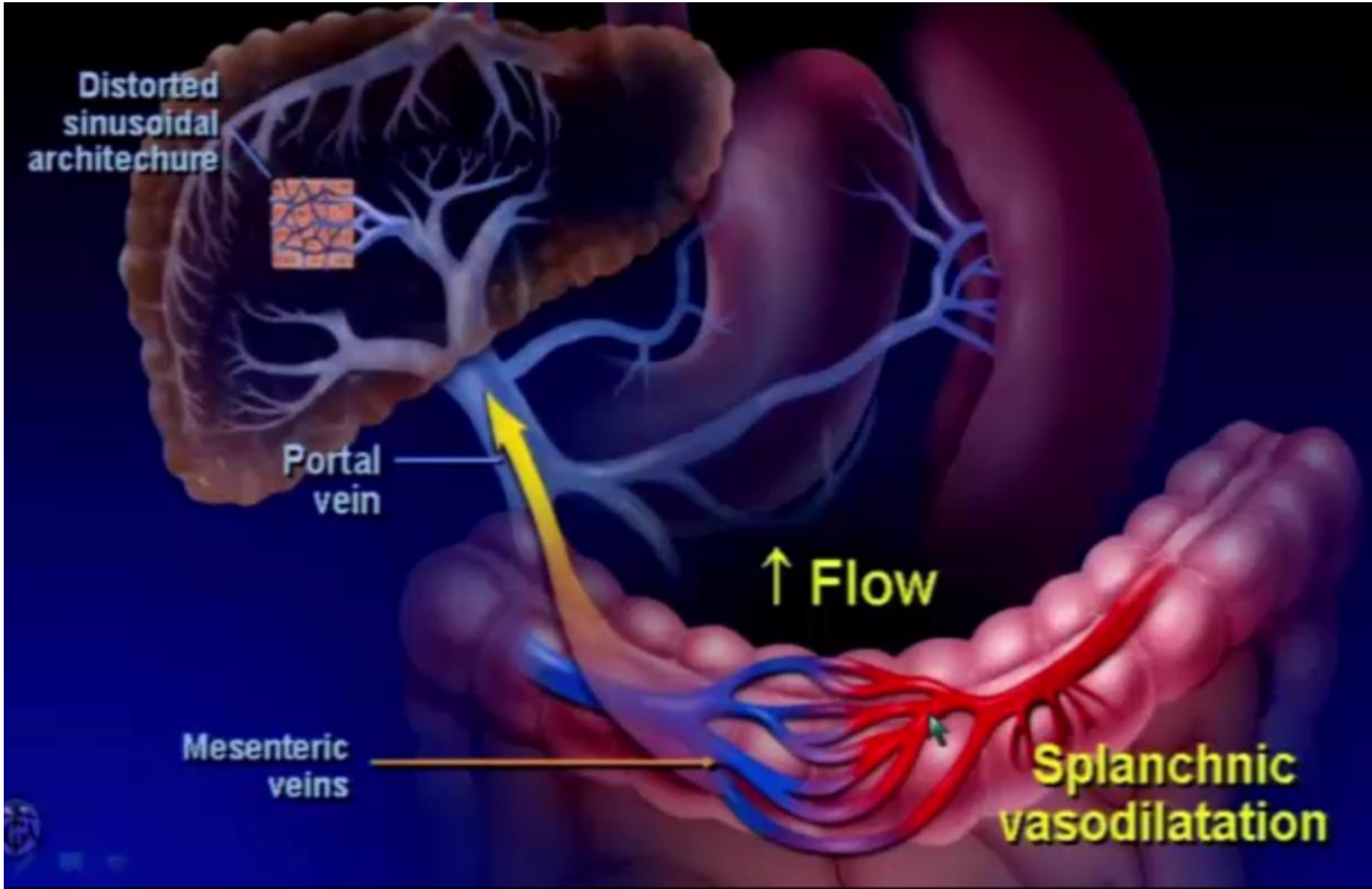


Umbilical fissure (Falciform ligament)

Cantlie's line

Posterior view





Distorted sinusoidal architecture

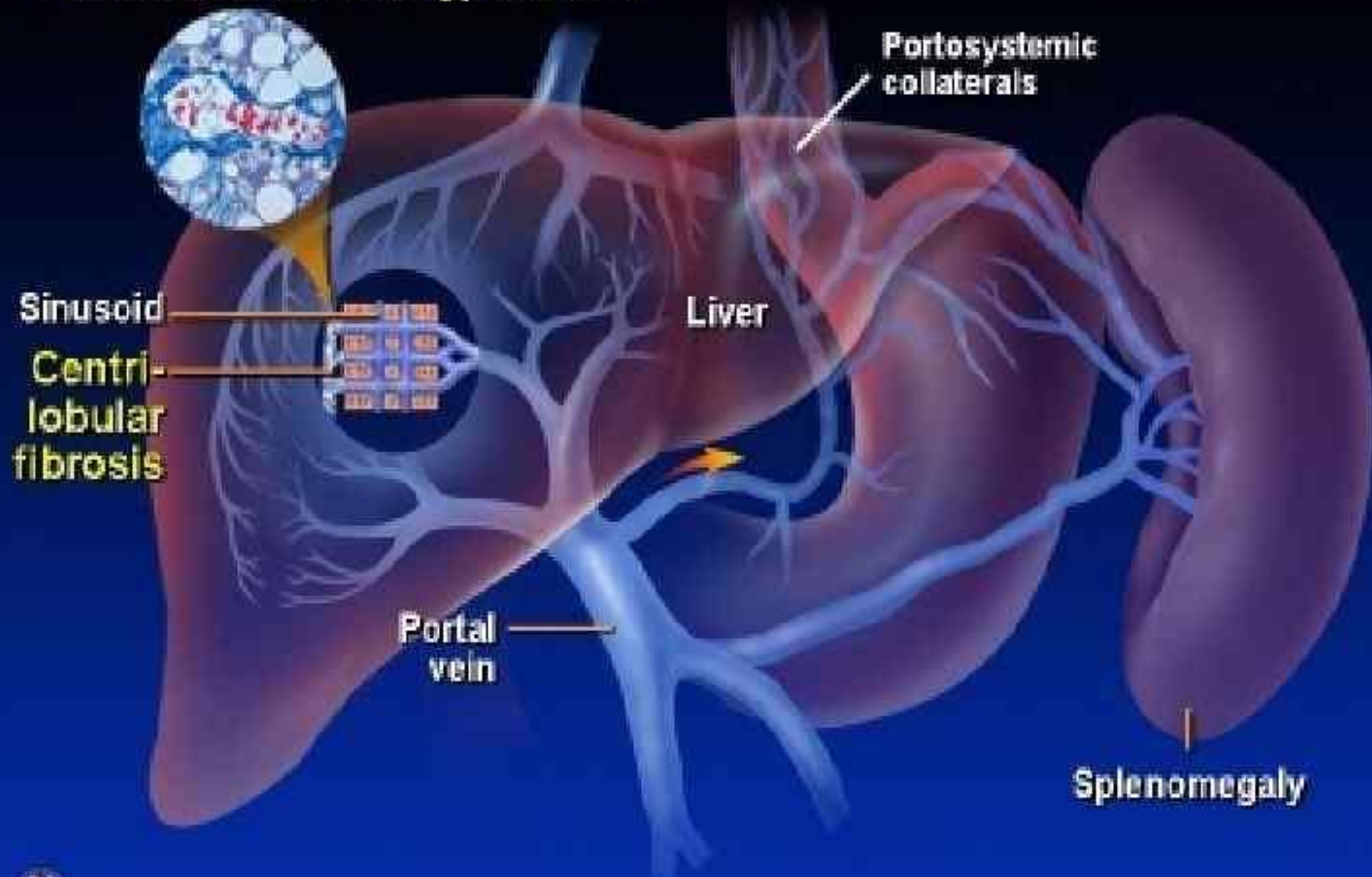
Portal vein

↑ Flow

Mesenteric veins

Splanchnic vasodilatation

Post-Sinusoidal Portal Hypertension



Transjugular Intrahepatic Portosystemic Shunt

Hepatic vein

TIPS

Portal vein

Splenic vein

Superior mesenteric vein

