Vascular diseases

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- No financial disclosures

Topics

1) Pre-sinusoidal disorders (portal vein thrombosis, congenital intrahepatic portosystemic shunts, hereditary hemorrhagic telangiectasia, etc.)

2) Post-sinusoidal disorders (Budd-Chiari syndrome, sinusoidal obstruction syndrome, effect of congestive heart failure on the liver, etc.)

3) Vascular lesions seen in the context of vascular diseases of the liver, including large regenerative nodules

Liver vascular diseases

- Vascular diseases are common
- From asymptomatic to life-threatening
- Classification:
  - Diseases of outflow / inflow / sinusoids

Hepatic Blood Supply

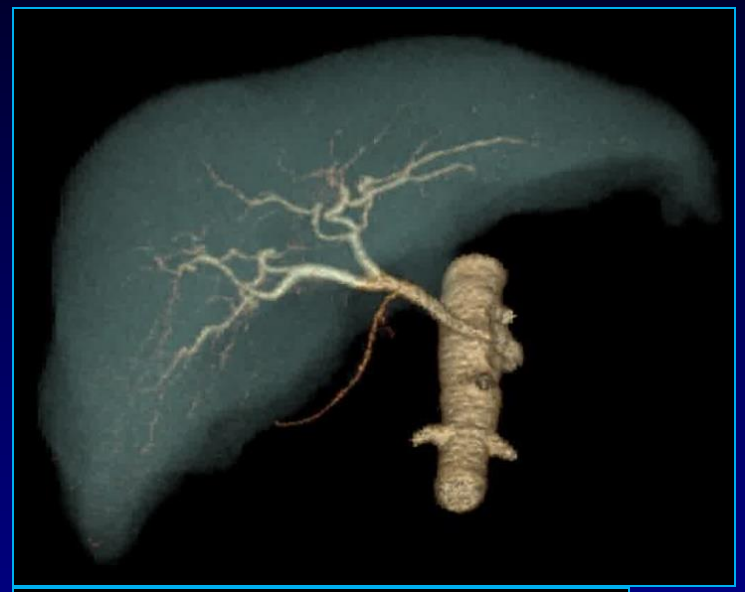
The liver has a dual blood supply

Hepatic artery:

- 25% of hepatic inflow
- ±10cc/100g/min
- 120/70 mm Hg
- 55% Type I anatomy

Portal vein:

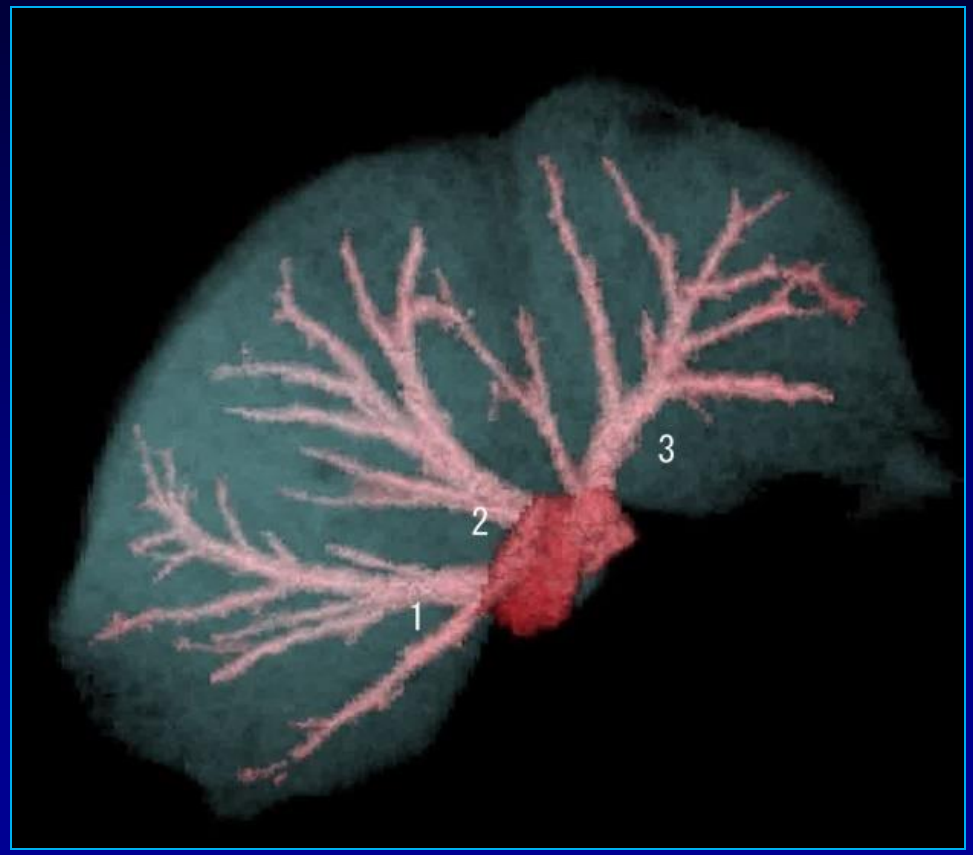
- 75% of hepatic inflow
- ±60cc/100g/min
- 10 mm Hg
- 80-94% Type I anatomy



Hepatic venous drainage

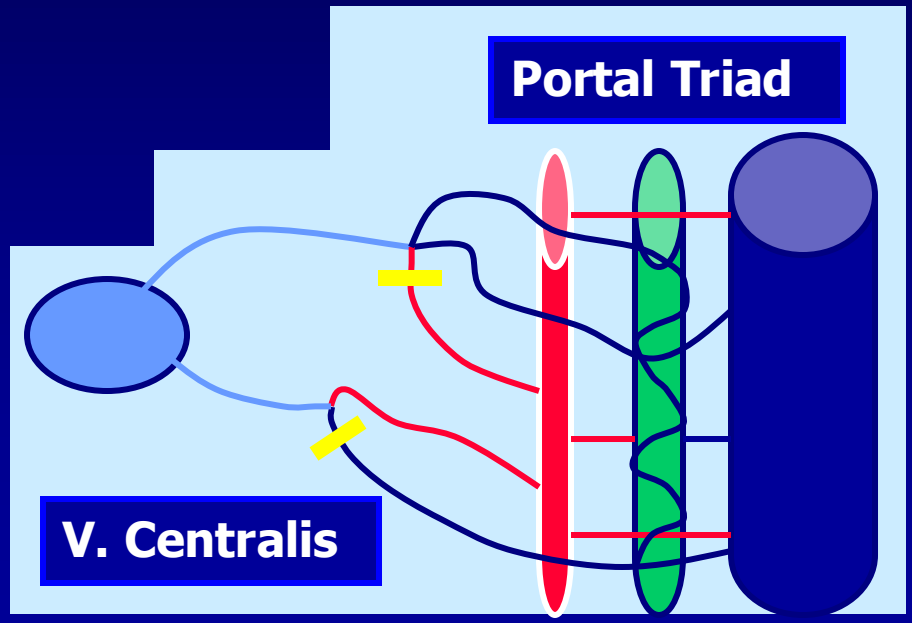
Single venous drainage:

- Hepatic veins
  - Right– middle - left
  - 5-20 dorsal hepatic veins
    - Drain paramedian tissue
  - 85%: common trunk



Hepatic sinusoids

- Hepatic lobules
  - polygonal units (1,000,000)
  - contain 1 central vein
  - supplied by Glisson's triad
  - flow from periphery to center
- Sinusoids
  - arterioportal communications
  - dynamic microcirculation
  - arteriolar inlet sphincter
  - venular sphincter
  - angiogenic modulators

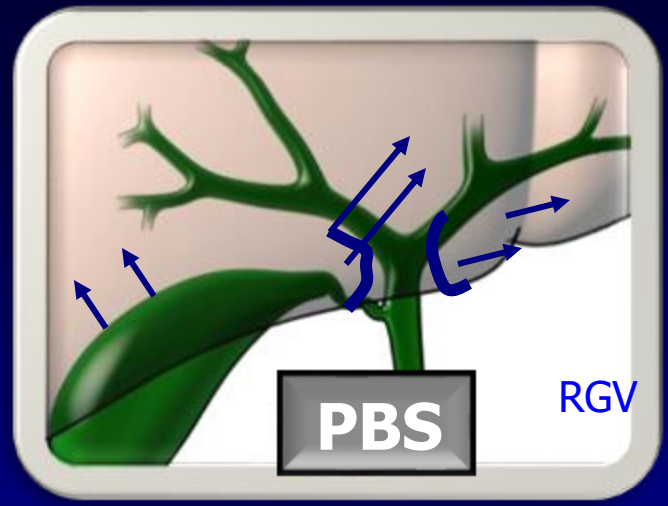


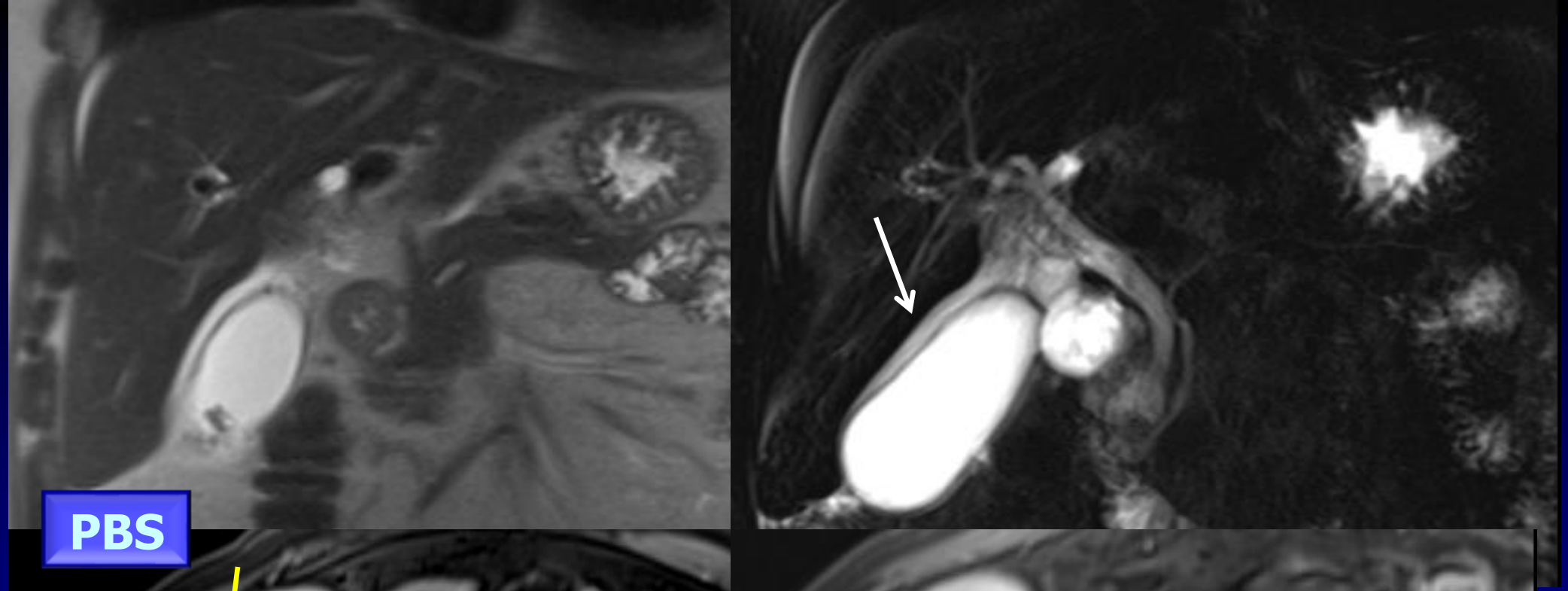
Third inflow

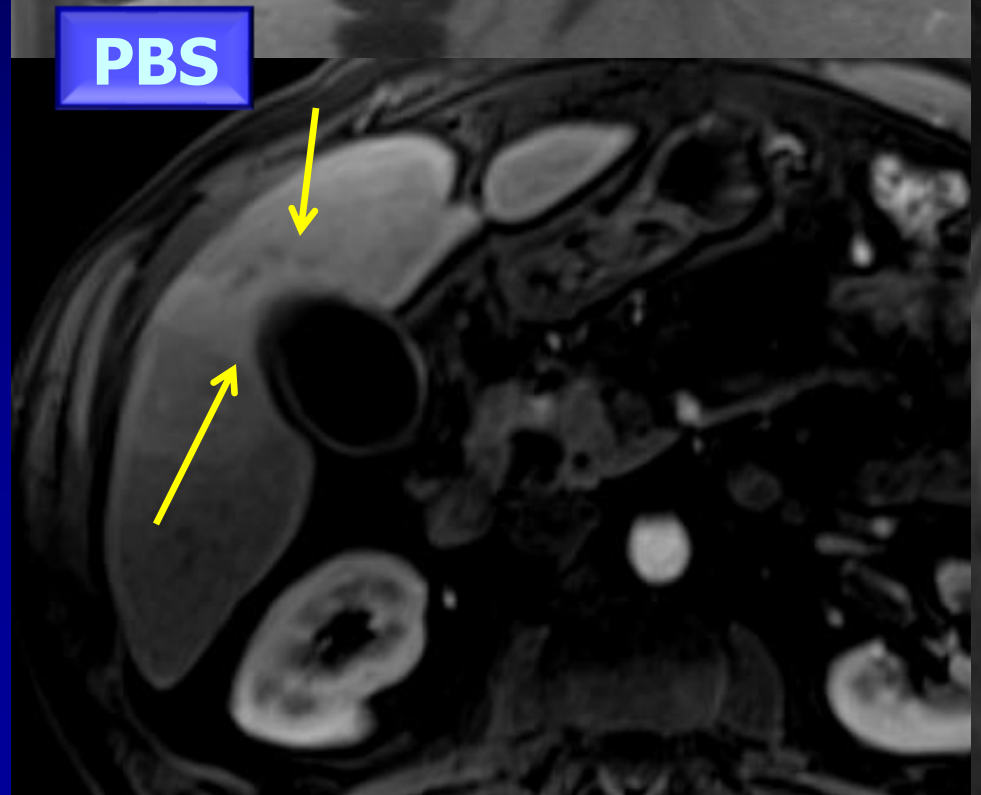
Anomalous Venous Inflow

Parabiliary Venous System

- R gastric vein (S 4)
- Pancreatico-duodenal veins (S 4)
- Cholecystic veins (S 5)



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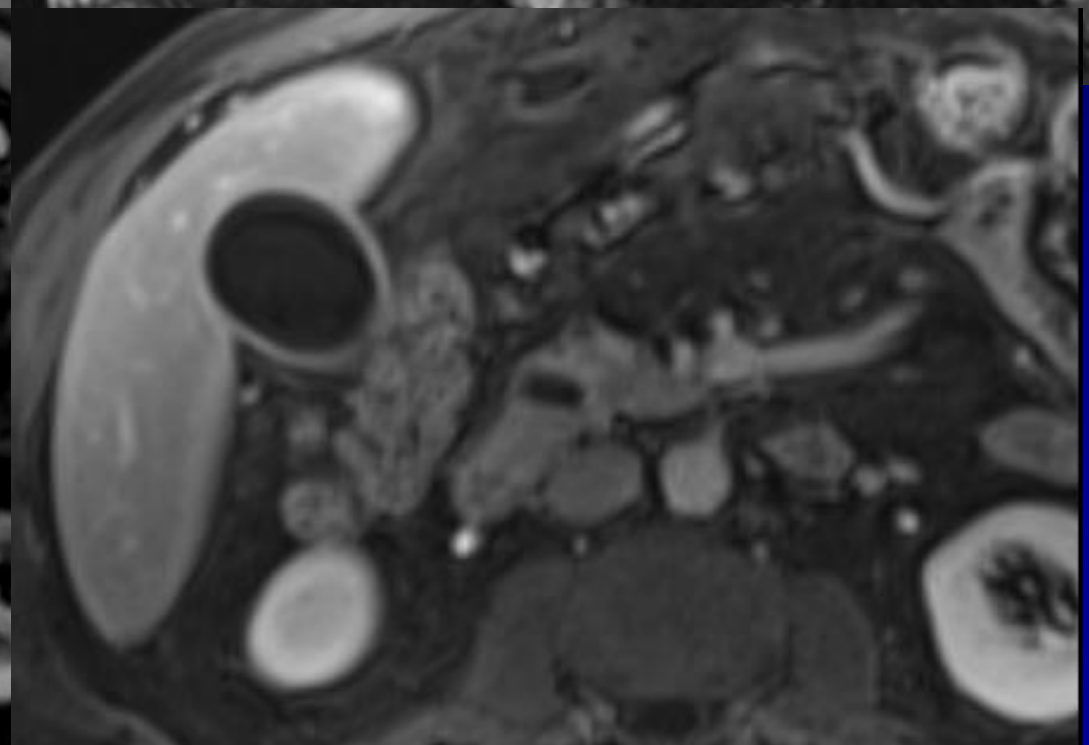

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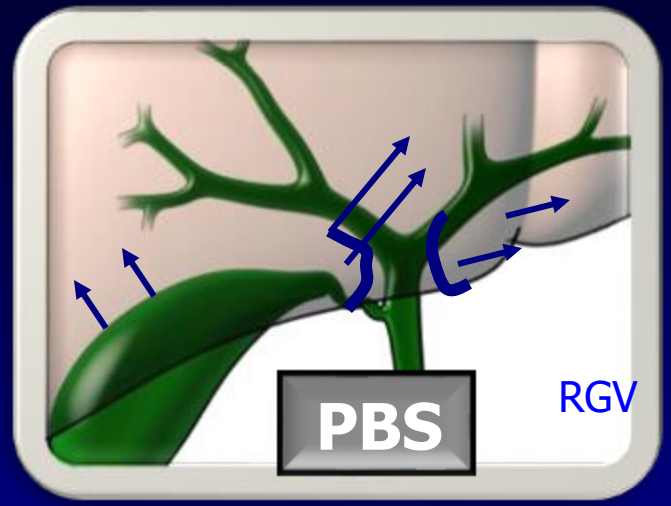
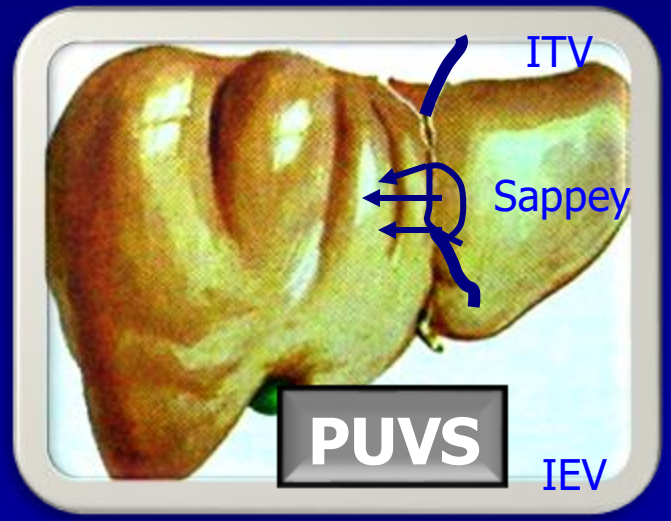


Paraumbilical Venous System

- Internal thoracic vein
- Inferior epigastric vein
- Veins of Sappey
- S 4b (pons hepatis)



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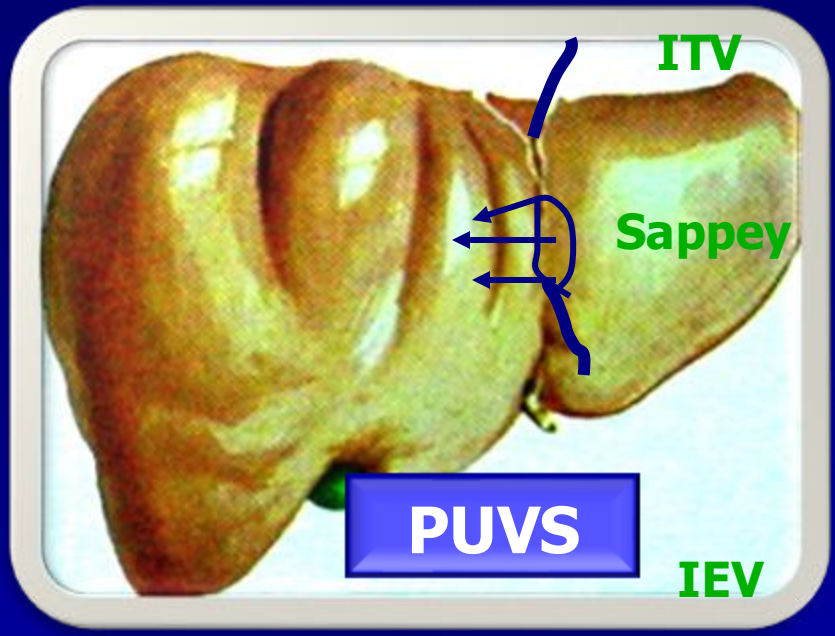

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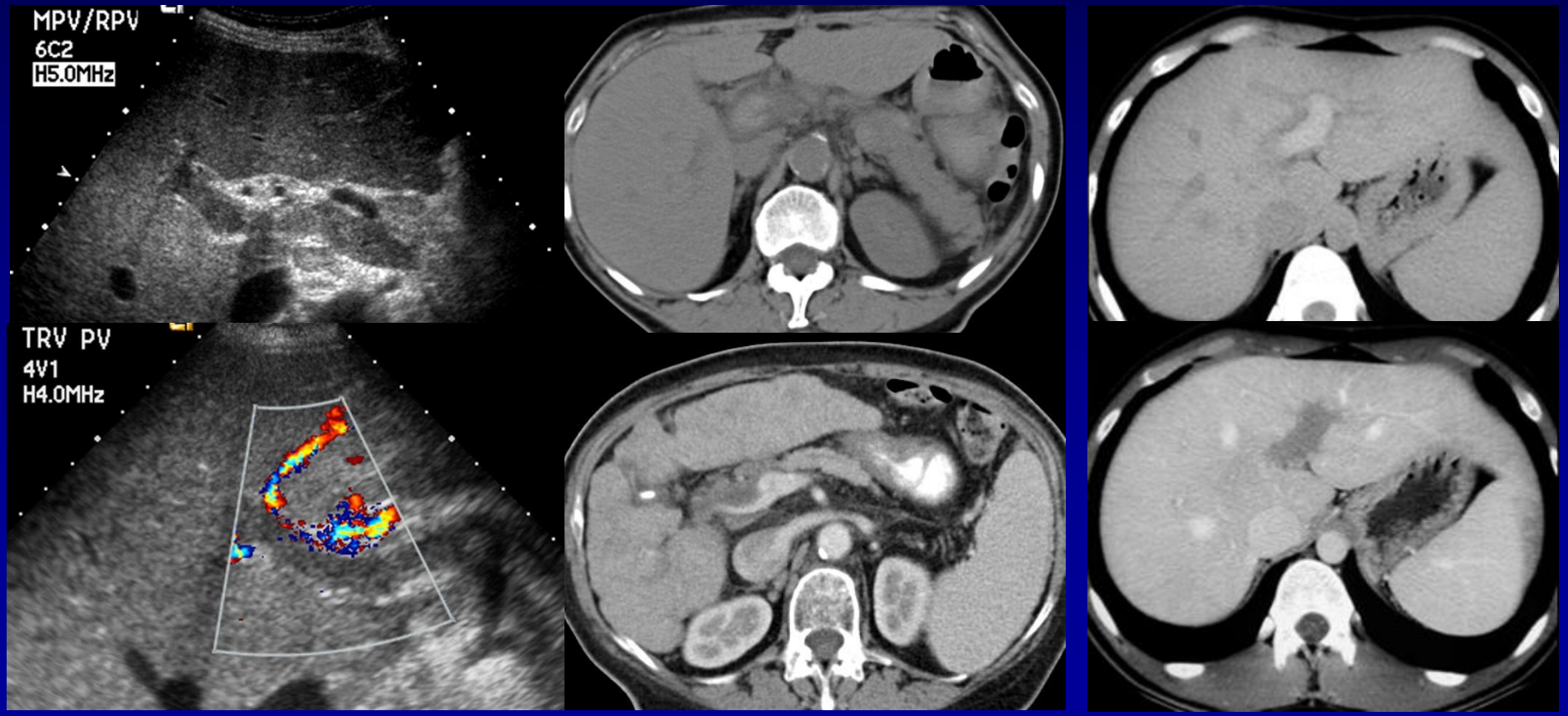
Portal vein thrombosis

- **Etiology**
  - Systemic or local causes
    - Hereditary thrombogenic factors (protein S deficiency)
    - Acquired thrombogenic factors (myeloproliferative Dx)
    - Portal vein injury
    - Portal venous stasis (cirrhosis, venous outflow)
    - Inflammatory conditions (appendicitis, diverticulitis, pancreatitis)
    - Neoplastic conditions (HCC, mets, pancreas ca)
- **Impairment**
  - Upstream: mesenteric venous arches prevent gut ischemia
  - Downstream: protection due to « buffer response »
    « cavernoma formation »

Acute Portal Vein Thrombosis

- Echogenic or high attenuation material in PV
- PV dilated
- Bland thrombus* or tumor thrombus

  * no enhancement versus arterial waveform

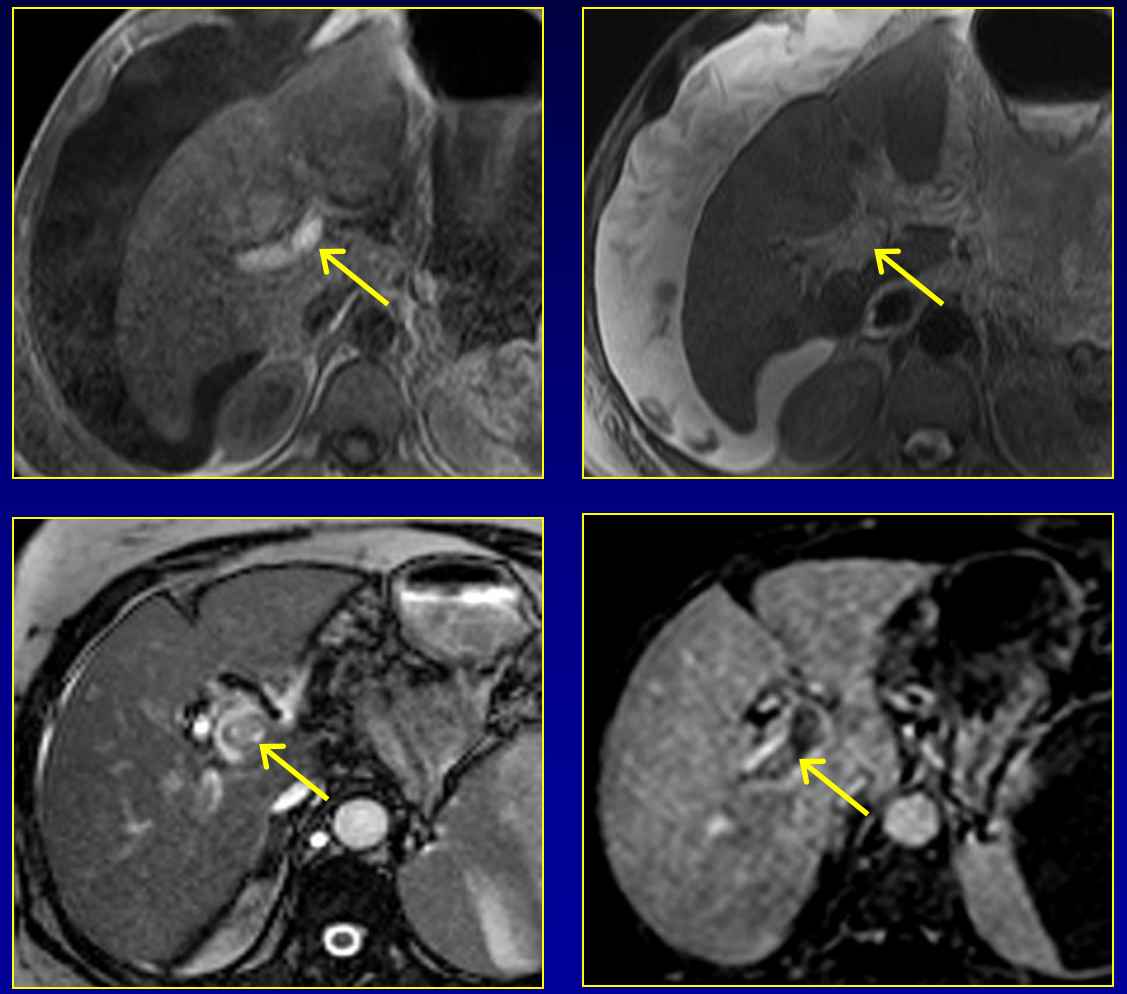


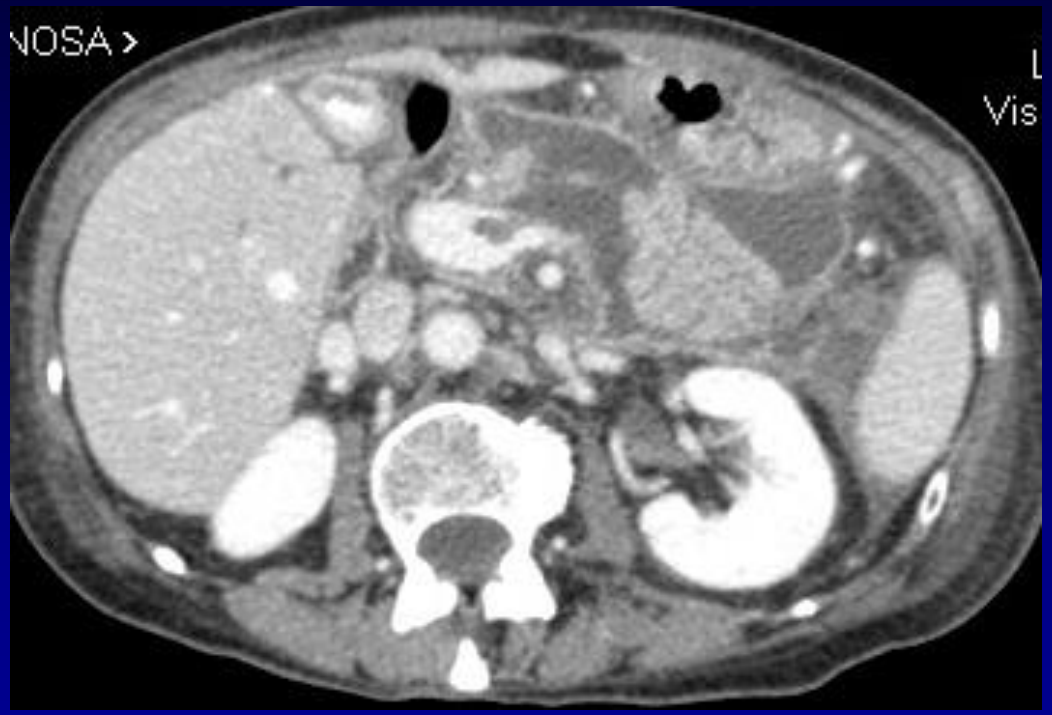
Portal vein thrombosis: direct signs

Isointense/hyperintense in “dark blood” sequences

Hypointense in “brightblood”

No enhancement

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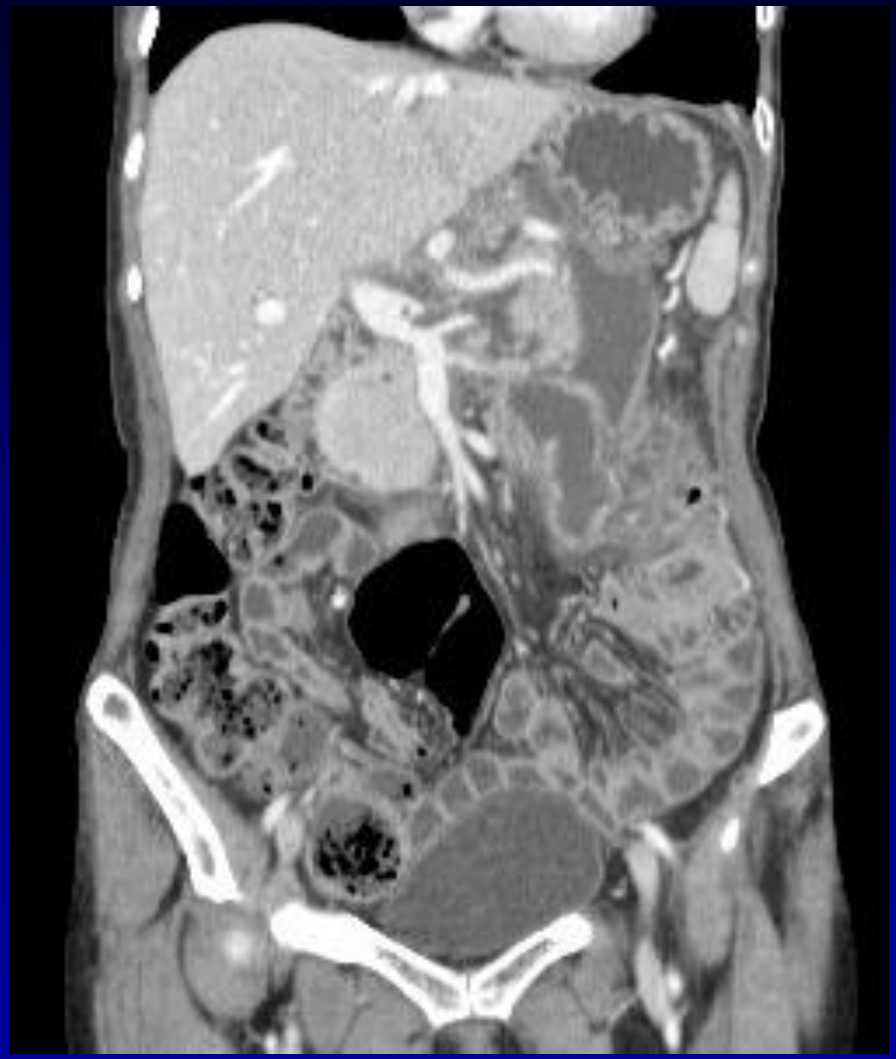
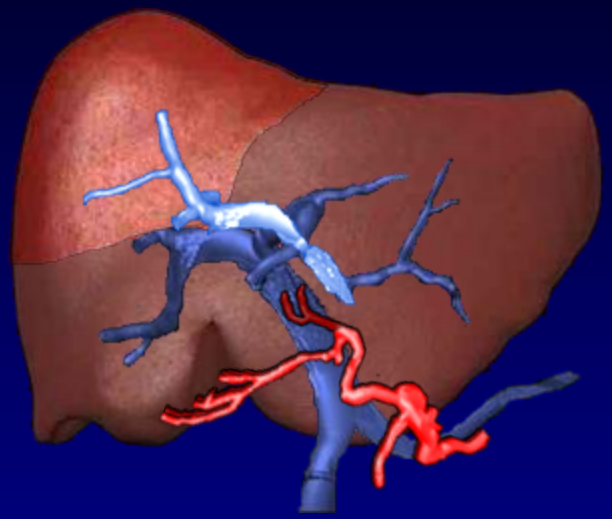
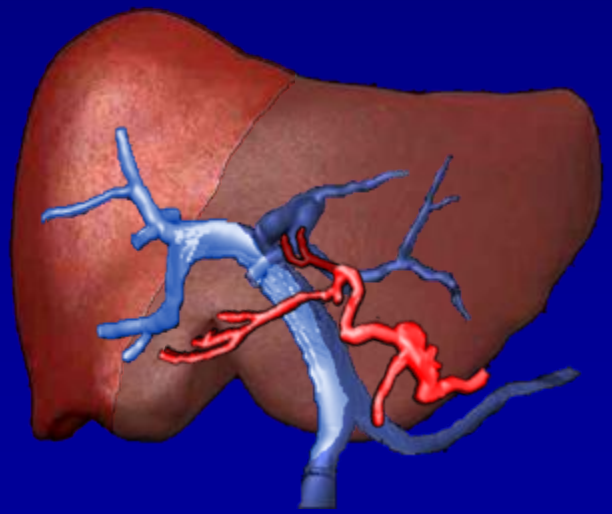
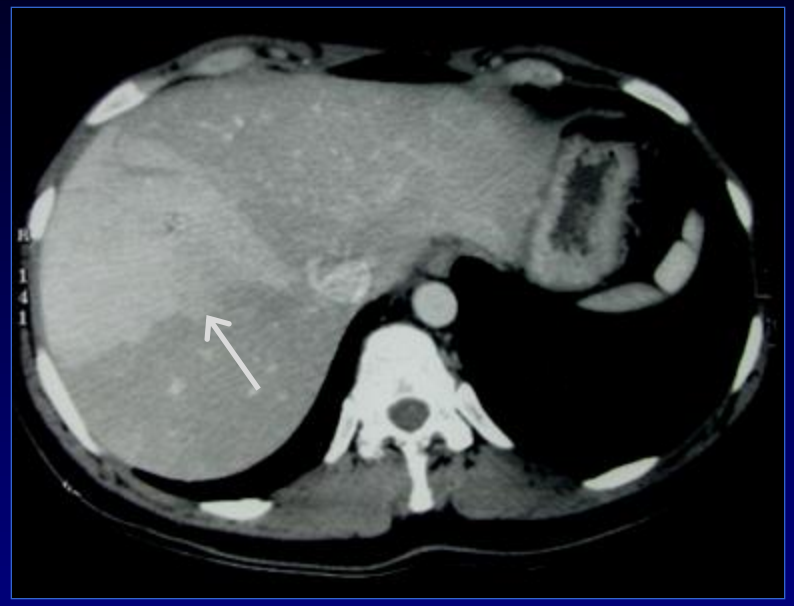
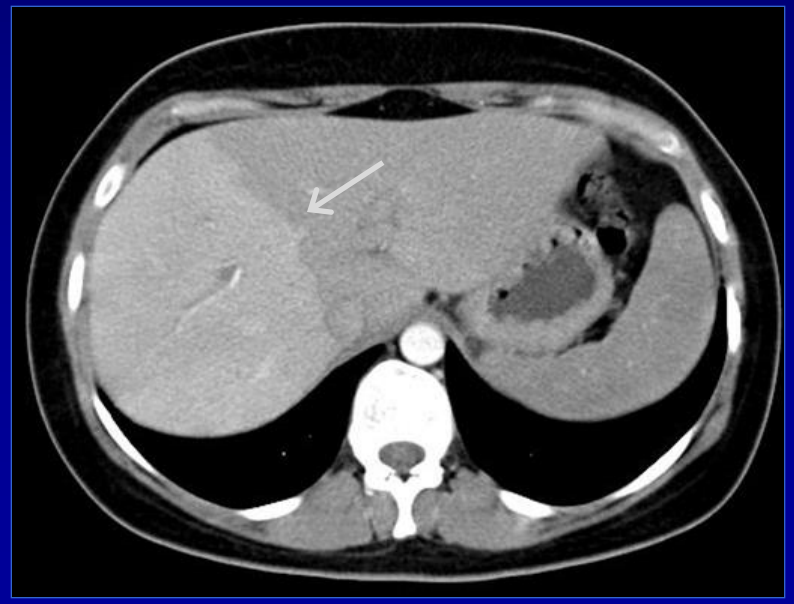


Acute PV thrombosis: perfusion abnormalities



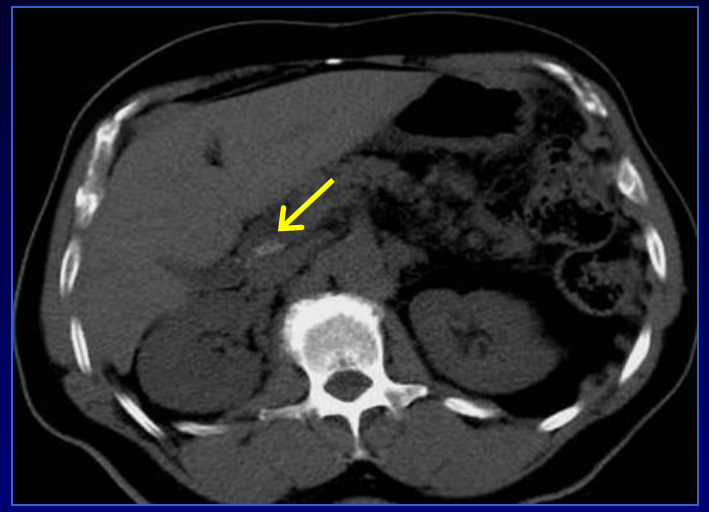


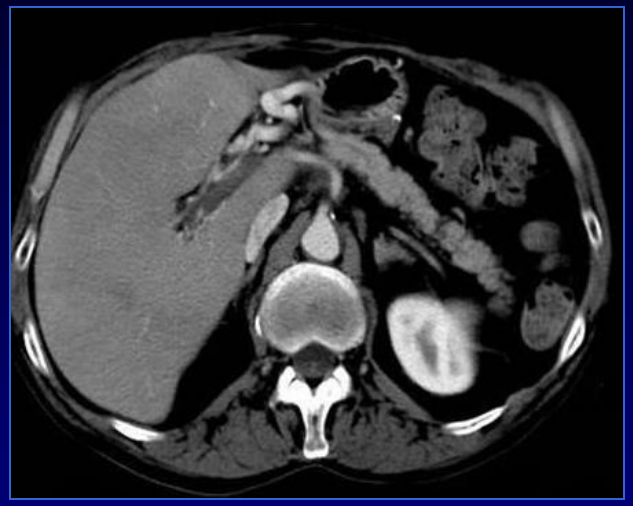




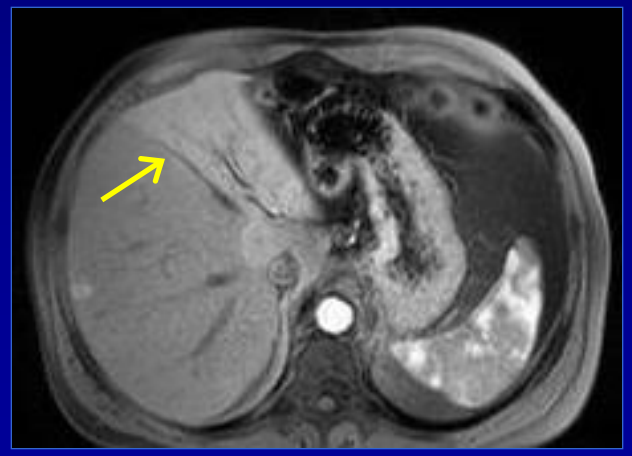
Chronic PV thrombosis

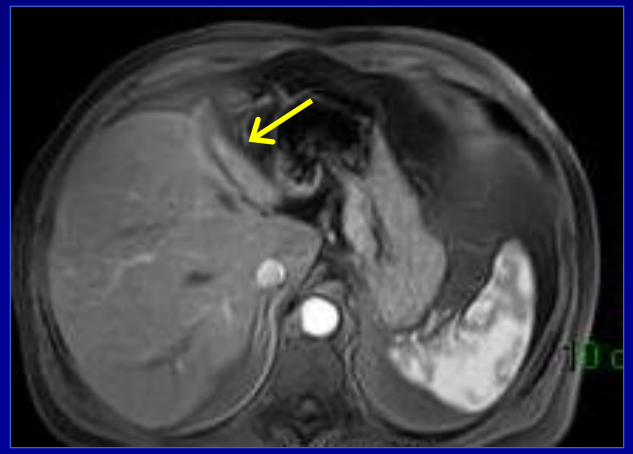
Fibrosis and calcifications of the obstructed branch



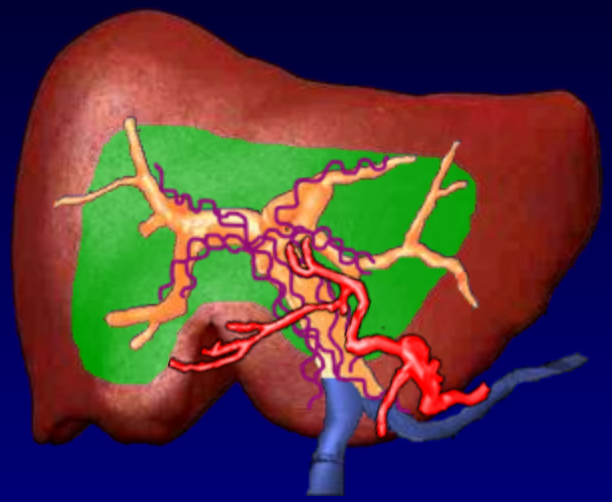


Parenchymal atrophy



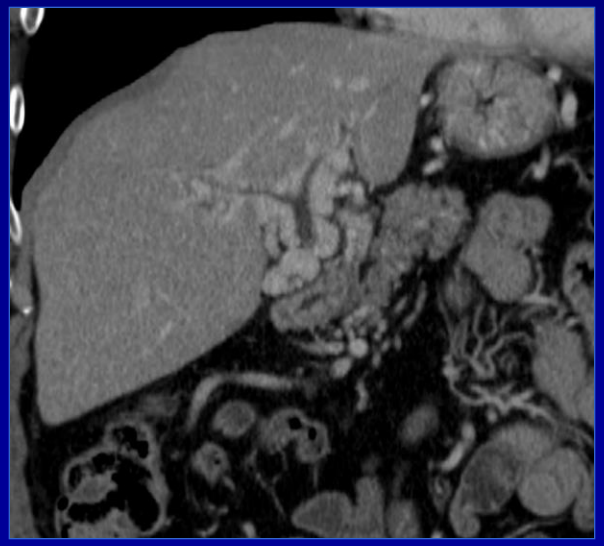


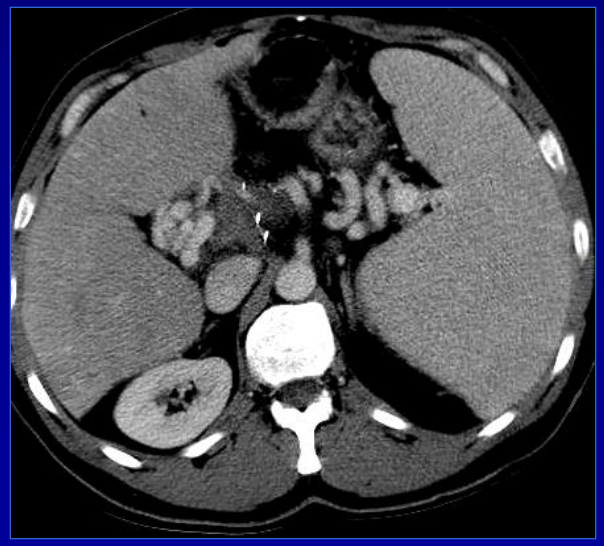
Chronic PV thrombosis: portal cavernoma

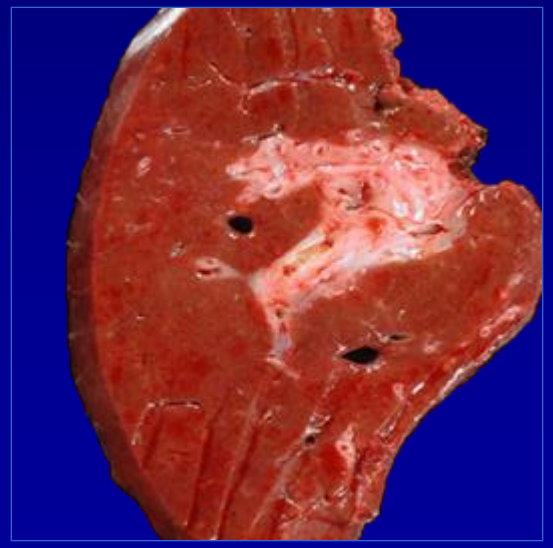


Cavernoma: network of capillaries around the hepatic pedicle

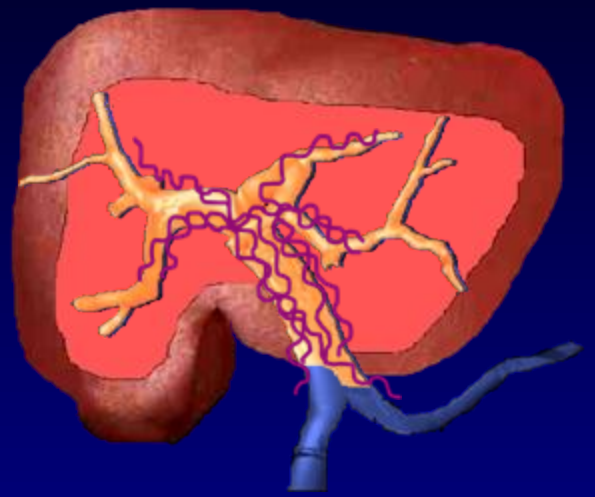
pericoledocic
pericolecystic
peripancreatic







Chronic PV thrombosis: changes in morpholgy



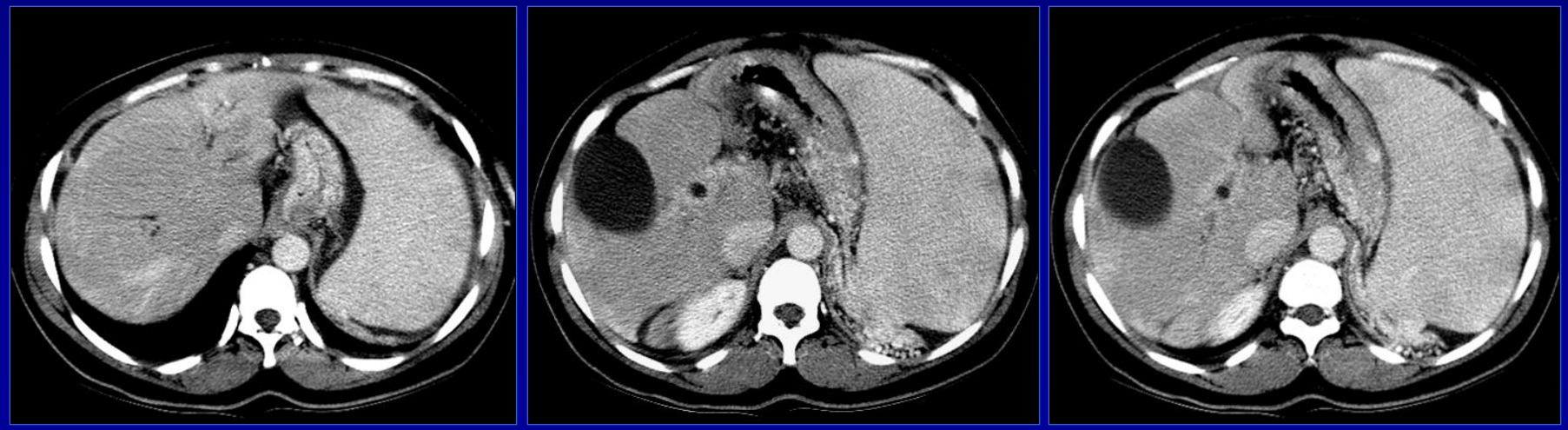
No fibrosis at biopsy!

May mimic cirrhosis because

- Caudate lobe hypertrophy
- Right lobe atrophy

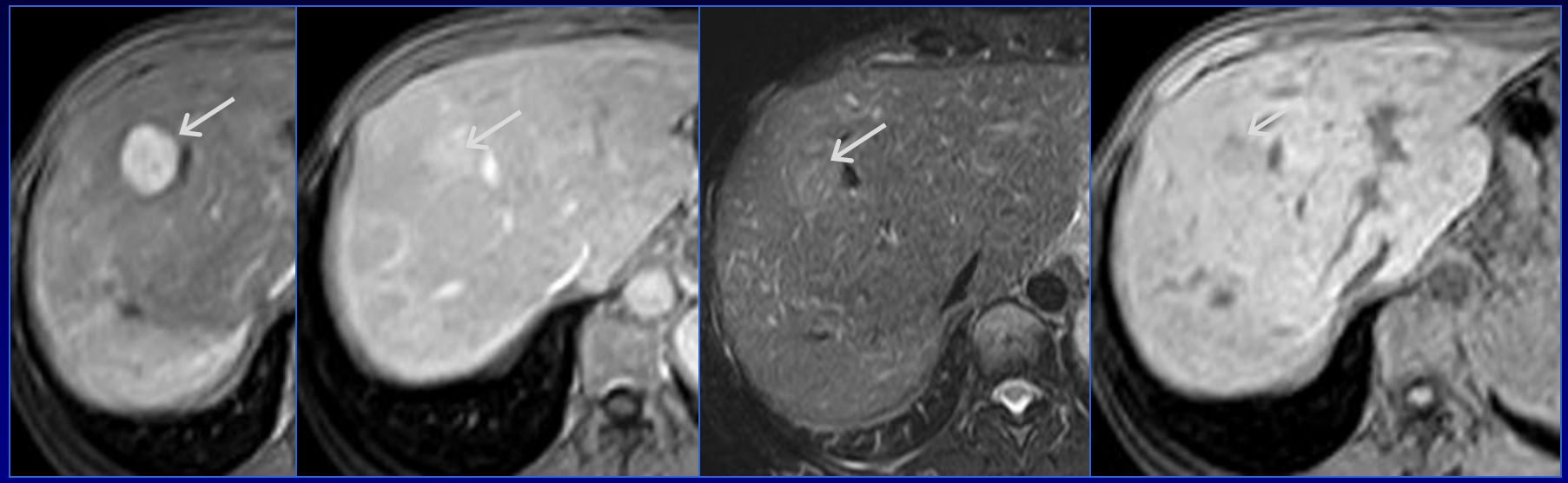
but

- Sharp margins
- Left lobe atrophy
- S4 normal or hypertrophic



Chronic PV thrombosis

Regenerative nodules



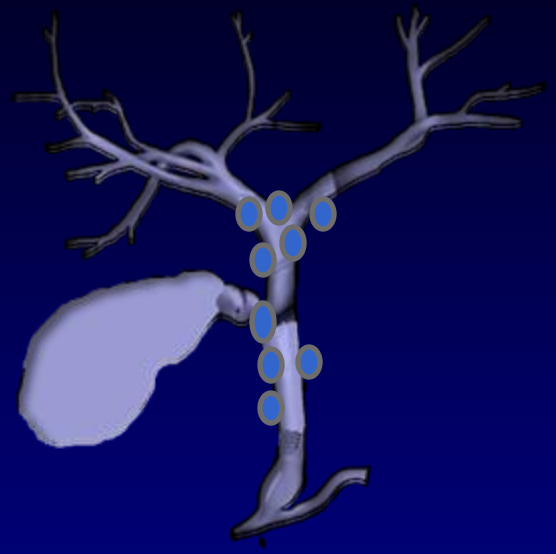
single/multiple nodules

hypervascular

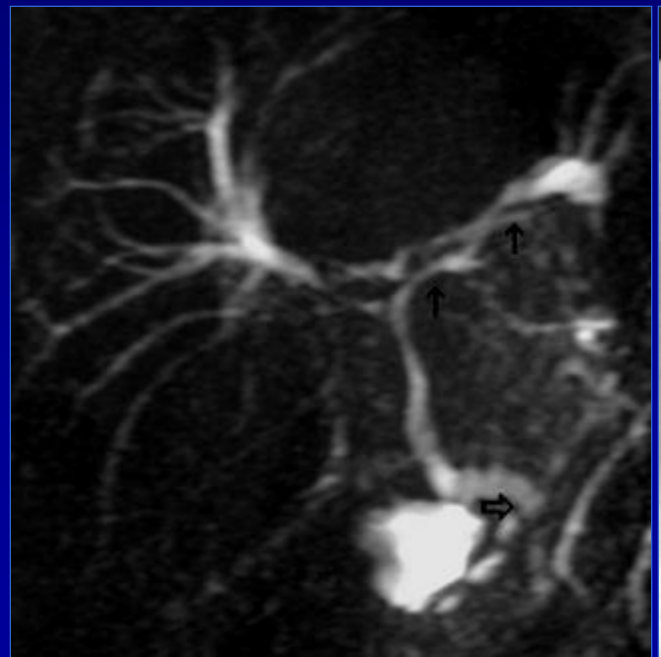
mean diameter ~ 16 mm (5-40 mm)

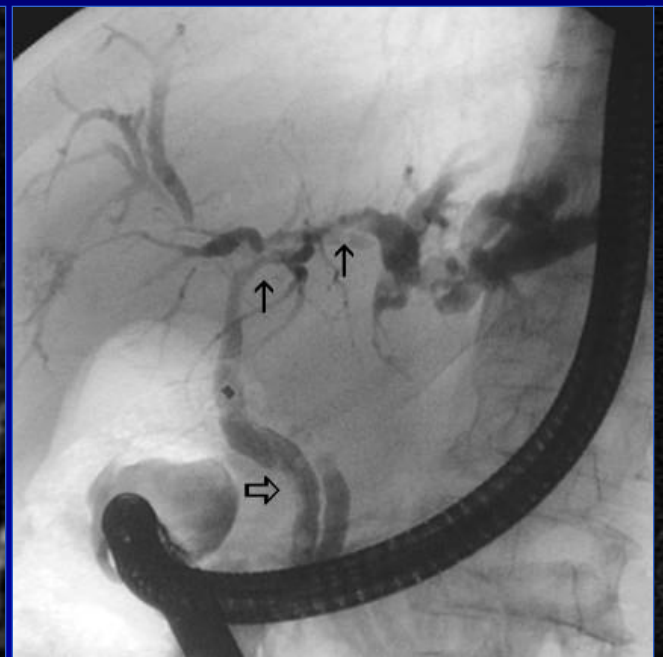
benign, FNH-like, confirmed at biopsy

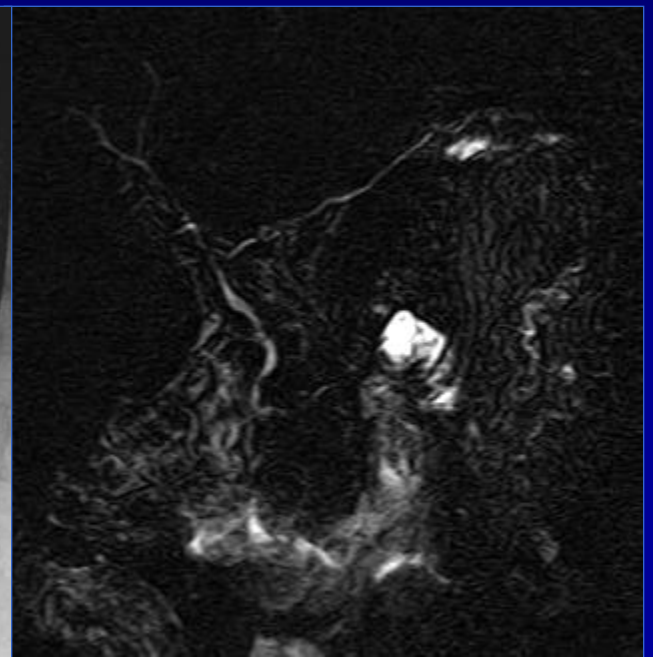
Chronic PV thrombosis: portal cholangiopathy



Stenotic, irregular, angulated bile ducts, compressed by cavernoma







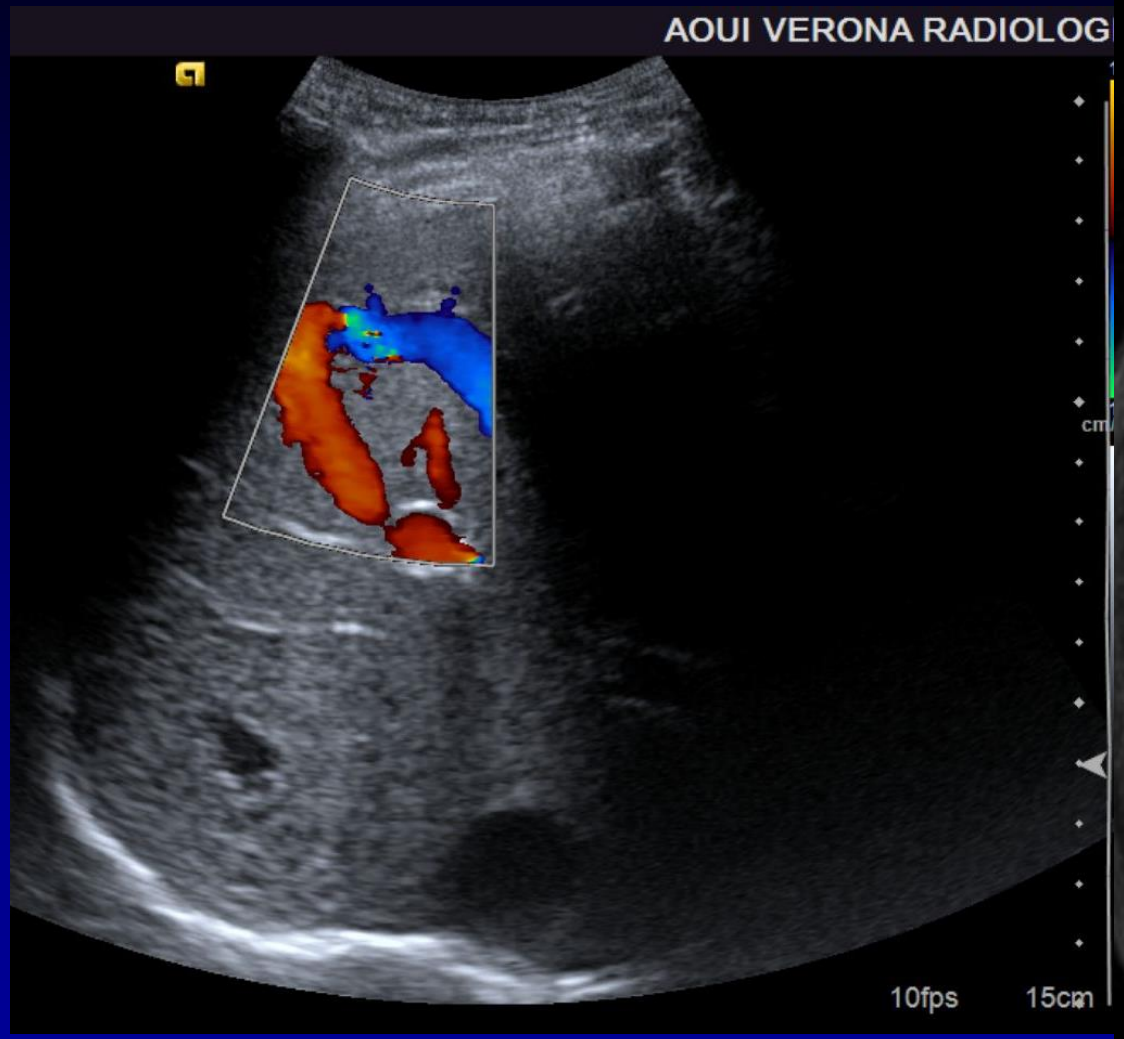
Intrahepatic portosystemic shunts

Prevalence up to 7%

4 types:

1. Single vessel connecting right portal vein and IVC
2. Segmental shunt, with one or more connections between peripheral portal branch and peripheral venous branch
3. Like 2, but with focal varix
4. Multiple connections between peripheral portal and venous branches in both lobes

Park et al AJR September 1990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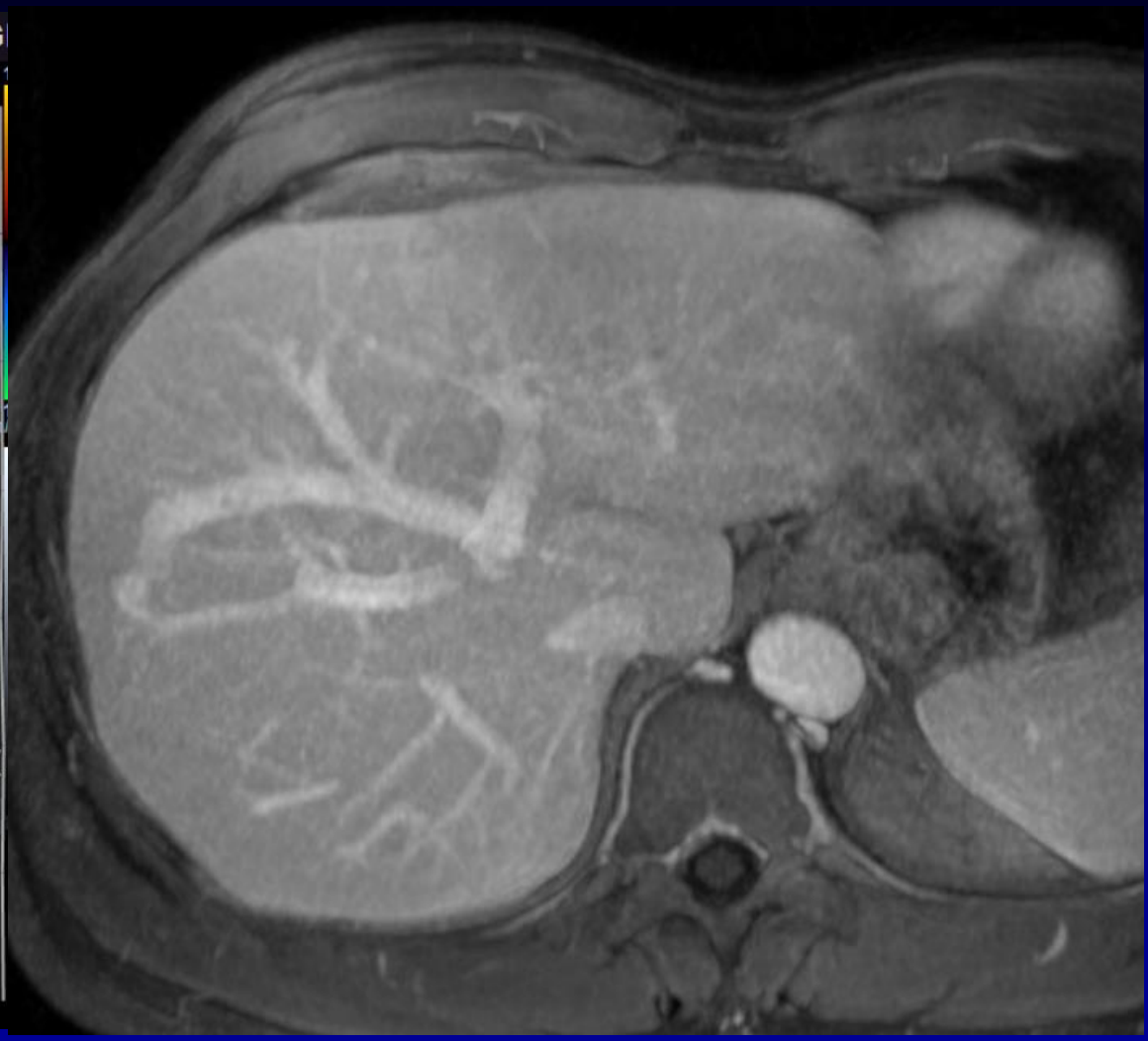
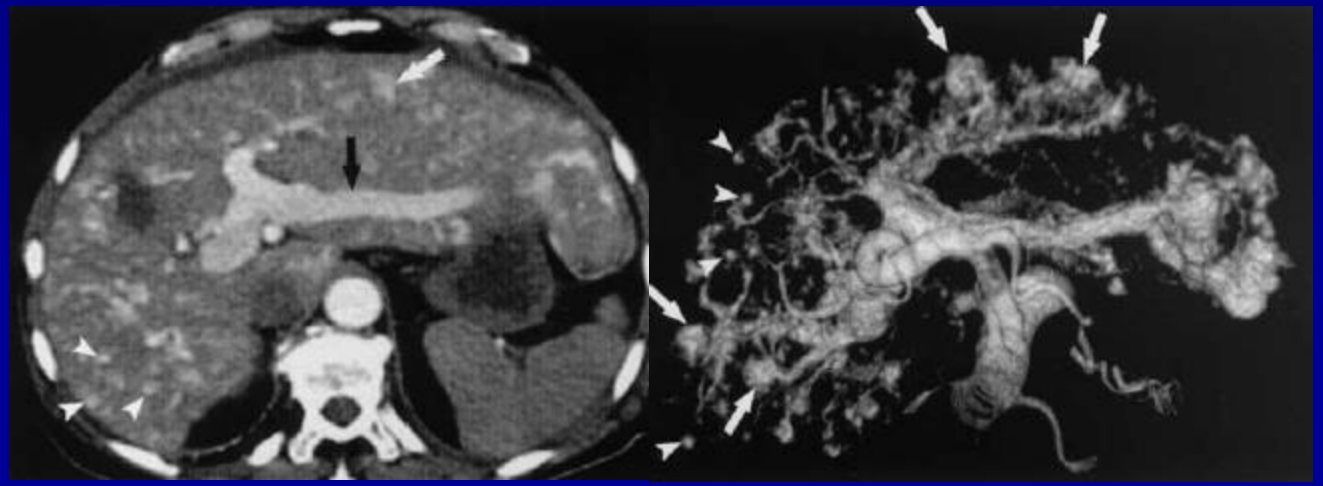


Hemorrhagic telangiectasia

Rendu-Osler-Weber disease

- autosomal dominant (1-2/10,000)
- mucocutaneous and visceral angiodysplastic lesions
- hepatic involvement = 8-31%
- types of lesions
    * intrahepatic shunts (A-P, A-S, P-S)
    * telangiectases, NRH, portal hypertension



Memeo M, et al. Hepatic involvement in hereditary hemorrhagic telangiectasia: CT findings. Abdom Imaging 2004;24:211-220.

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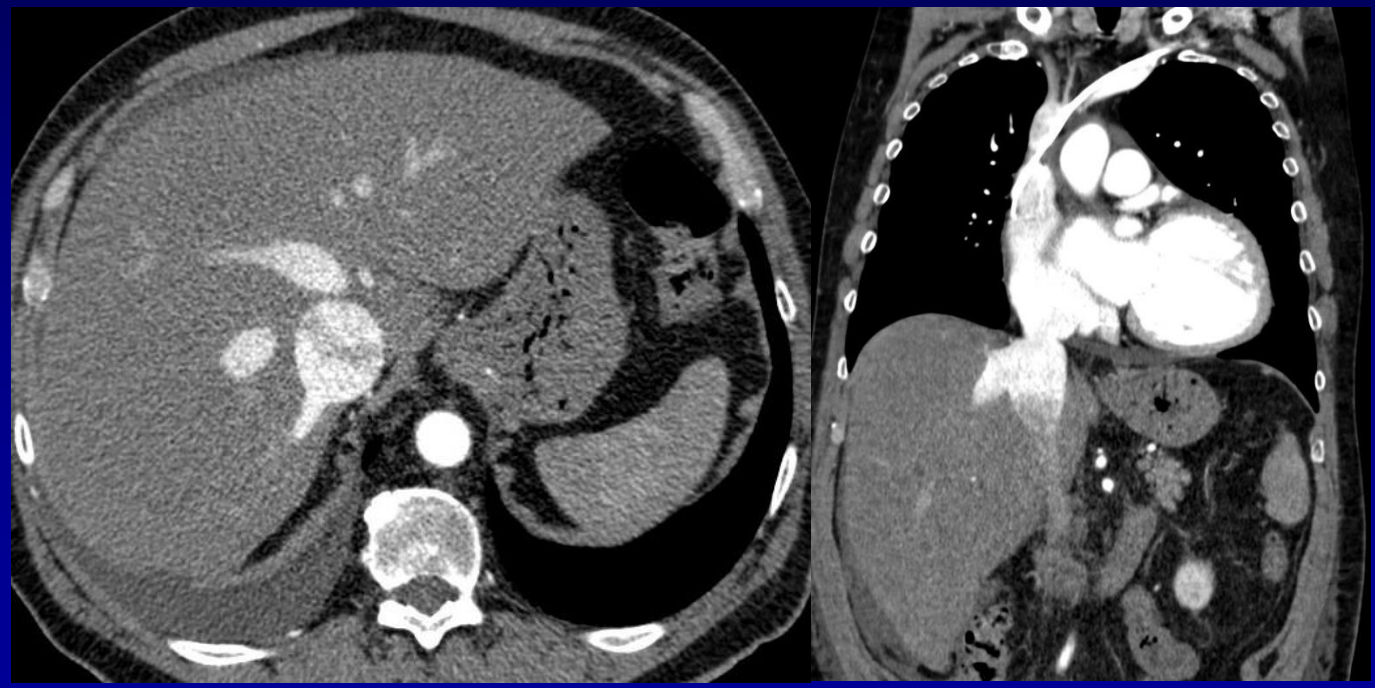
Hepatic vein disorders
Passive hepatic congestion

- **Pathophysiology**
  - Due to increased CVP
  - Causes centrilobular congestion (jaundice) and hepatic cell death due to hypoxia associated with stasis
  - Among most common diffuse liver diseases
- **Etiology**
  - Congestive heart failure
  - Constrictive pericarditis
  - Tricuspid insufficiency
  - Cardiomyopathies
  - Mitral and aortic valve disease

Giallourakis CC, et al. The liver in heart failure. Clin Liv Dis 2002;6:947-967

Hepatic vein disorders
Passive hepatic congestion

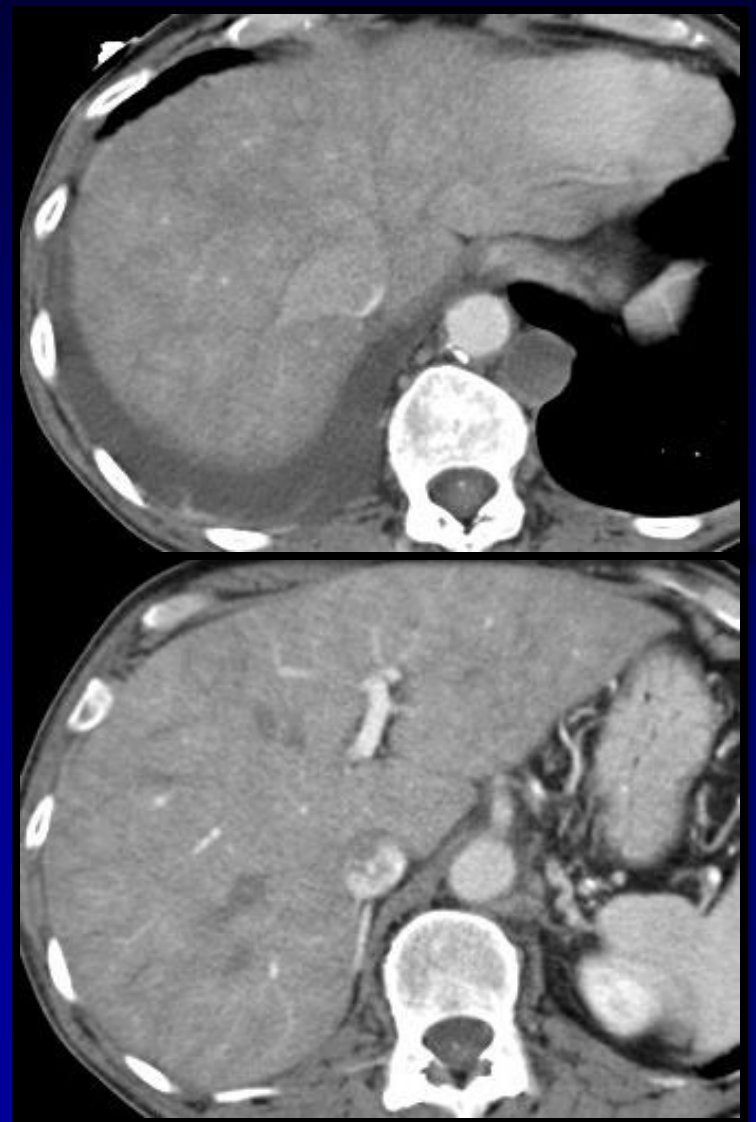
- Dilated IVC and hepatic veins
  - RHV=9-13 mm (normal=5-6 mm)
- Early reflux of contrast into IVC



Gore RM, et al. Passive hepatic congestion: cross-sectional imaging features. AJR 1994;162:71-75

Hepatic vein disorders
Passive hepatic congestion

- Dilated IVC and hepatic veins
  - RHV=9-13 mm (normal=5-6 mm)
- Early reflux of contrast into IVC
- 'Mosaic' enhancement
- Hepatomegaly
- Periportal & GB edema
- Ascites and effusions
- Increased pulsatility of PV
- Unidirectional, low-velocity, flow in IVC and hepatic veins



Gore RM, et al. Passive hepatic congestion: cross-sectional imaging features. AJR 1994;162:71-75

Hepatic vein disorders
Budd-Chiari syndrome

- **Pathophysiology**
  - Obstruction of the hepatic venous outflow tract
  - Small or large hepatic veins or IVC (suprahepatic)
  - Causes centrilobular congestion and ischemic necrosis
- **Primary**
  - Coagulation disorders
  - Myeloproliferative disorders
  - Membranonus obstruction (Asia)
- **Secondary**
  - Tumoral invasion
  - Vascular compression by infectious process (hydatid, amoeba)

Van Wettere et al, Abdom Radiol 2017

Hepatic vein disorders
Budd-Chiari syndrome

Clinical manifestations

- Acute forms (7%): abdominal pain, hepatomegaly, ascites, LFT
- Chronic (45%) or subacute forms (48%): impaired liver function
- Asymptomatic forms: discovered fortuitously

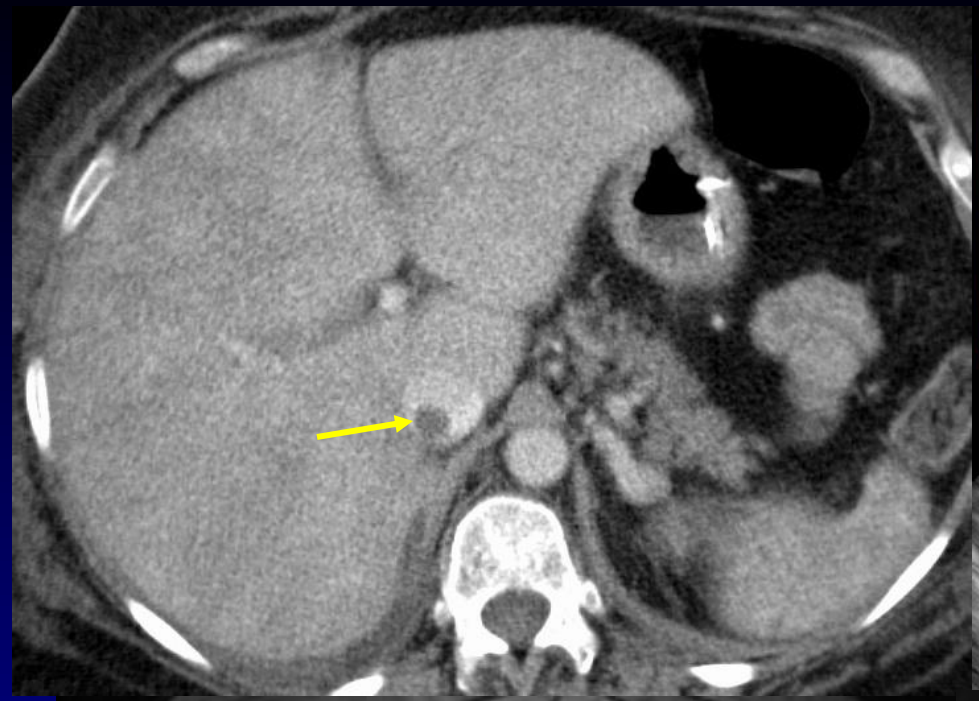
Van Wettere et al, Diagnosis of Budd-Chiari Syndrome; Abdom Radiol 2017

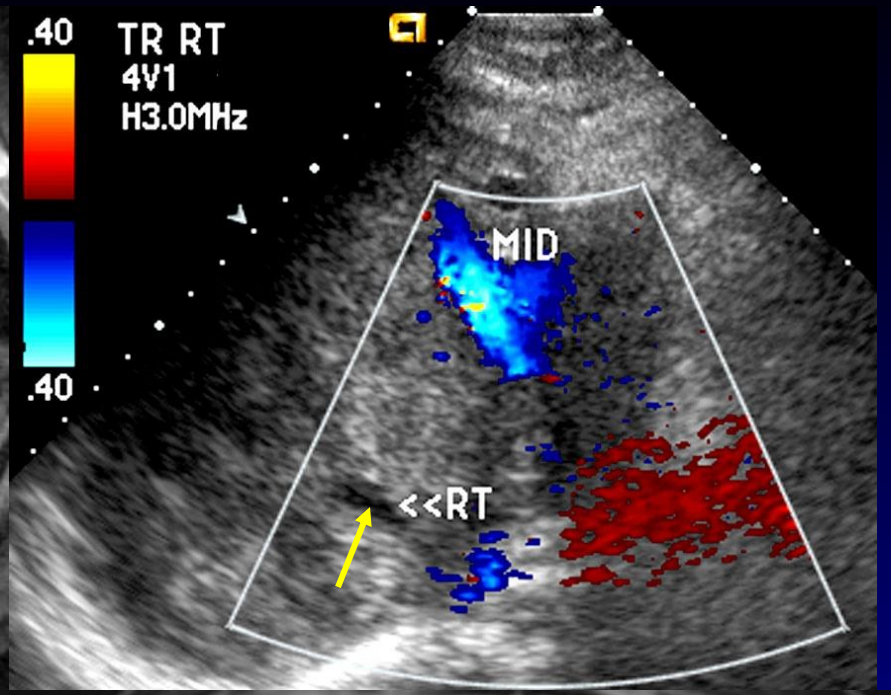
Hepatic vein disorders
Budd-Chiari syndrome

Imaging Features

- direct diagnostic findings
  - intraluminal material in hepatic veins or IVC (web, thrombus, tumor)
  - abscence of hepatic vein flow or localized flow disturbances (Doppler-US, MRA)
  - presence of intraparenchymal collaterals (‘comma sign’)
  - narrowing or nonvisualization of the H veins or IVC
- indirect diagnostic findings > morphologic changes

Camera L, et al. Triphasic helical CT in Budd-Chiari syndrome: patterns of enhancement in acute, subacute, and chronic disease. Clin Radiol 2006; Van Wettere et al, Diagnosis of Budd-Chiari Syndrome; Abdom Radiol 2017

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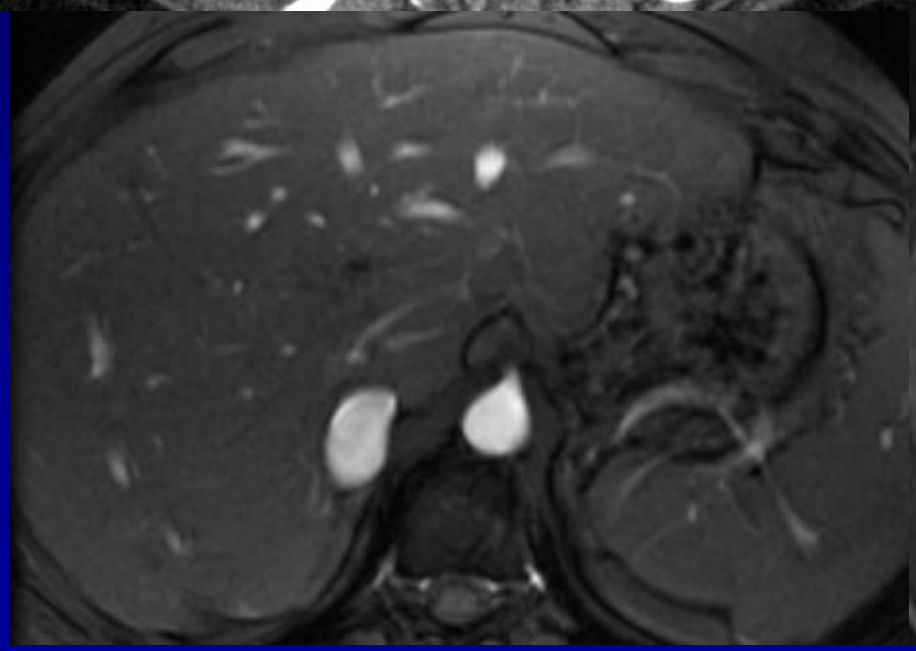
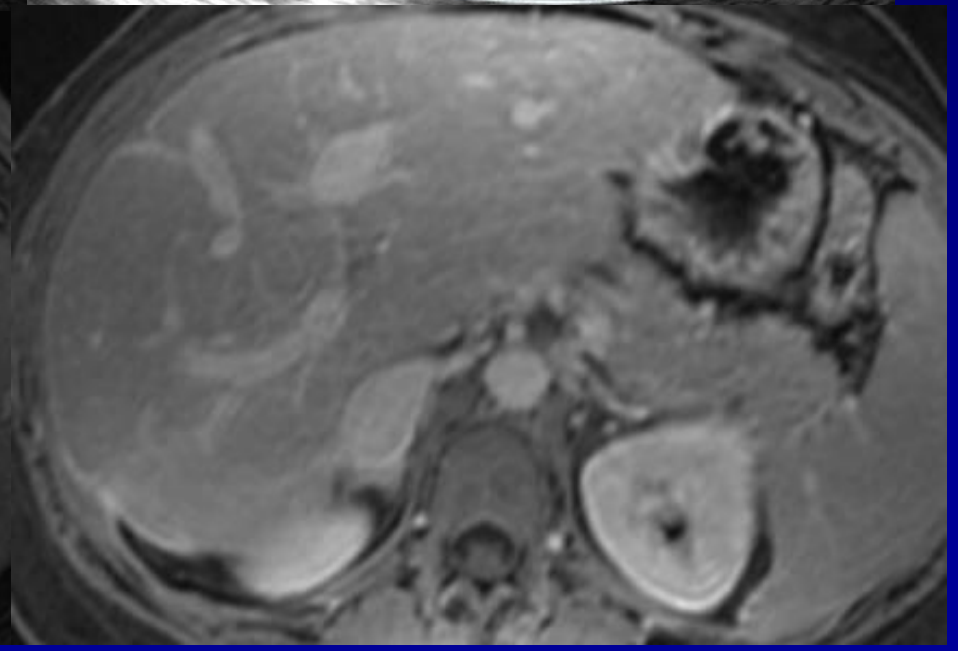
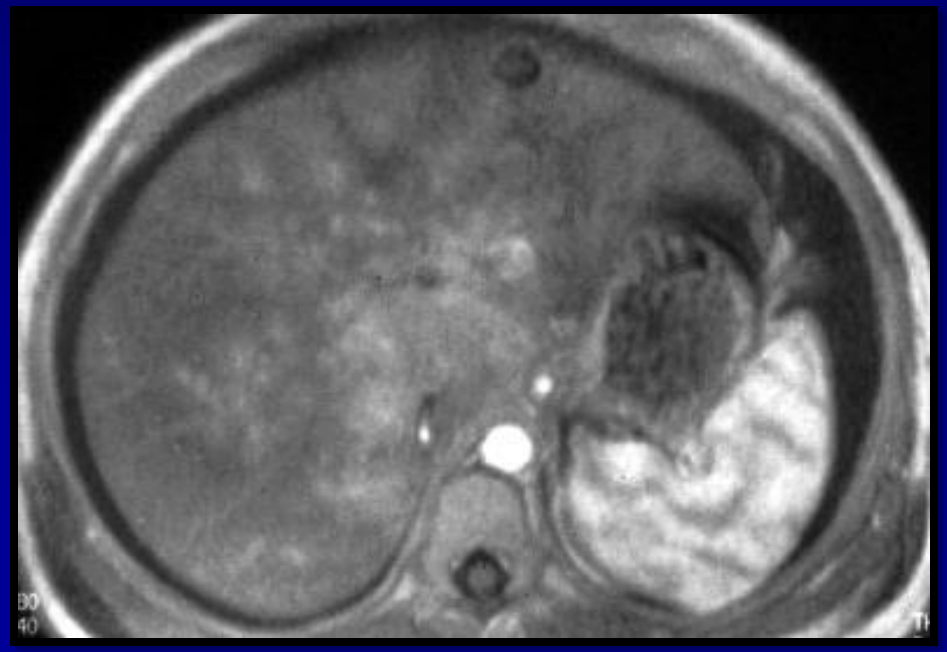


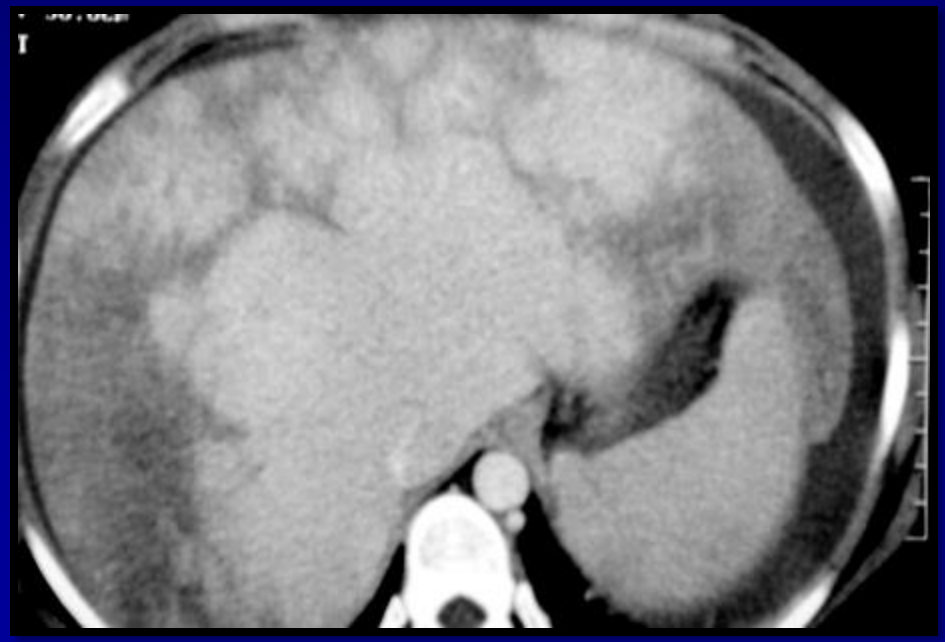


Budd-Chiari syndrome

morphologic changes: ACUTE

- delayed enhancement pattern of areas drained by the obstructed veins
- hypodense CT & edematous high SI on T2WI
- « fan-shaped » pattern of enhancement CL
- hepatomegaly without CL hypertrophy

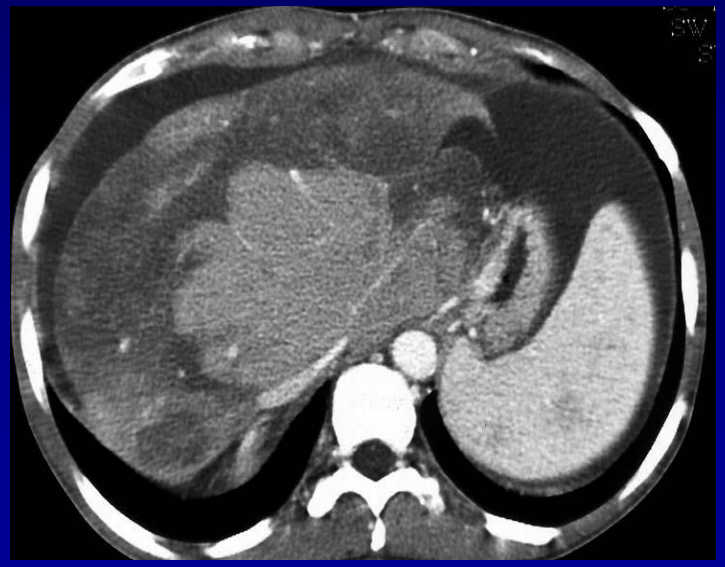


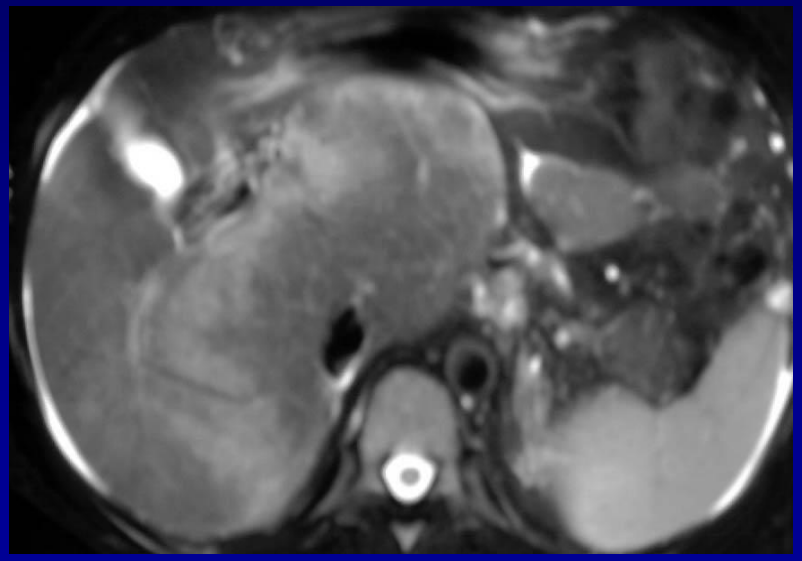


Budd-Chiari syndrome

morphologic changes: CHRONIC

- affected peripheral areas become fibrosed & atrophic
- caudate lobe hypertrophy
- HCC (6%) & benign regenerative nodules (25%)



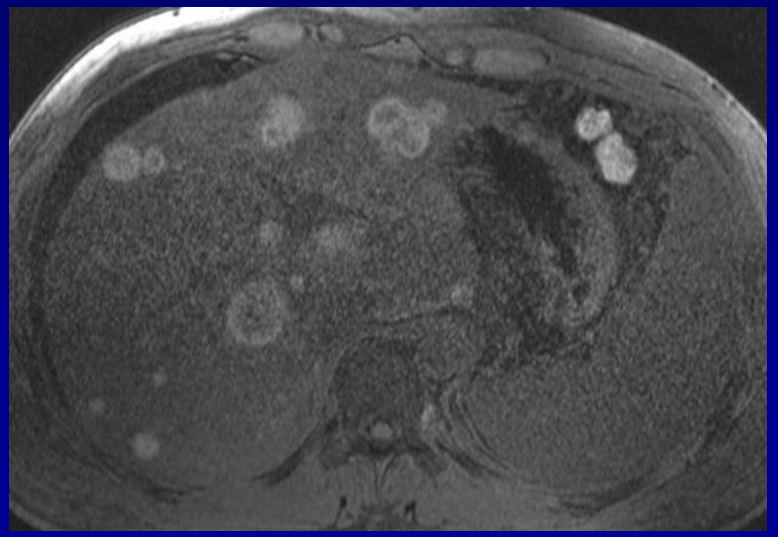


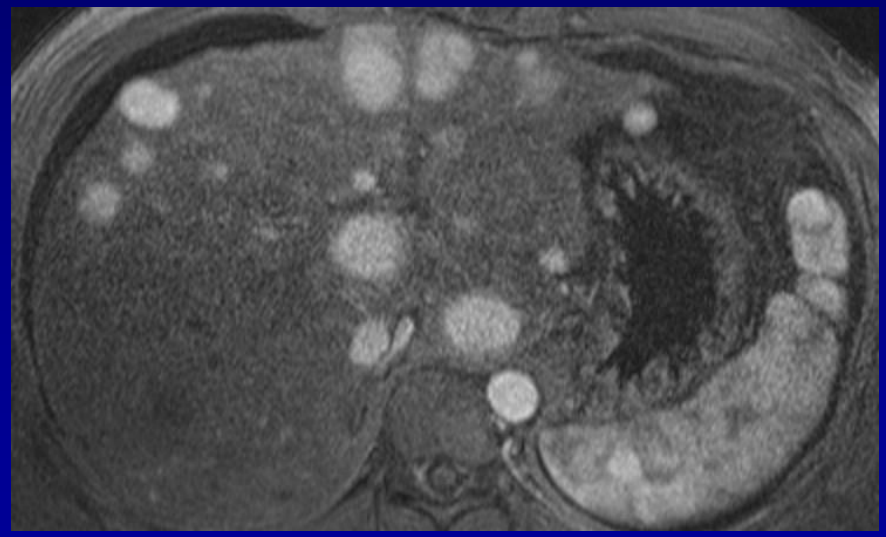
Courtesy of K Mortele

Budd-Chiari syndrome

benign regenerative nodules (FNH-like):

- homogeneous
- hypervascular
- hyperintense T1WI
- variable on T2WI
- Usually >10 lesions and <3 cm
- DDx: HCC, HCA





Vilgrain et al, Benign and malignant hepatocellular lesions in patients with vascular liver diseases; Abdom Radiol 2017

Sinusoidal obstruction syndrome

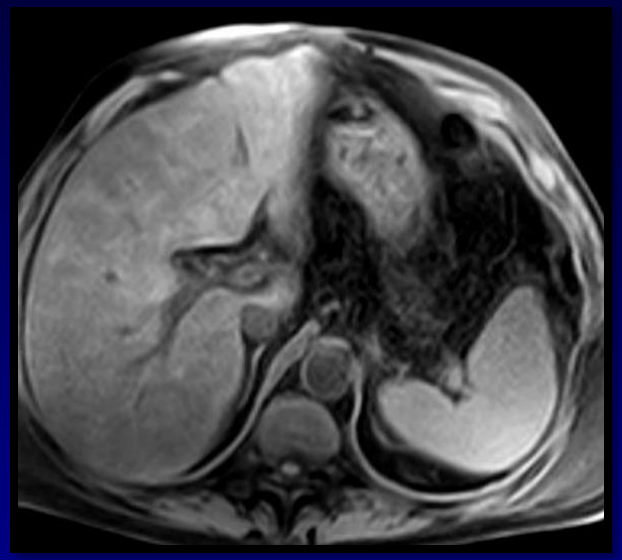
- Non-thrombotic obstruction of sinusoids w/wo centrilobular vein involvement
- Acute toxic damage to sinusoid endothelial cells
- Causes stop of the post-sinusoidal efflux (VOD)
- causes
  * After myeloablative therapy (10-30 days)
  * Post-radiation
  * Toxic substances (alkaloids, azatioprine, platinum cht)
  * herbal supplements (“bush tea”)
- symptoms: like Budd-Chiari
- DD post-BMT: GVHD, sepsis

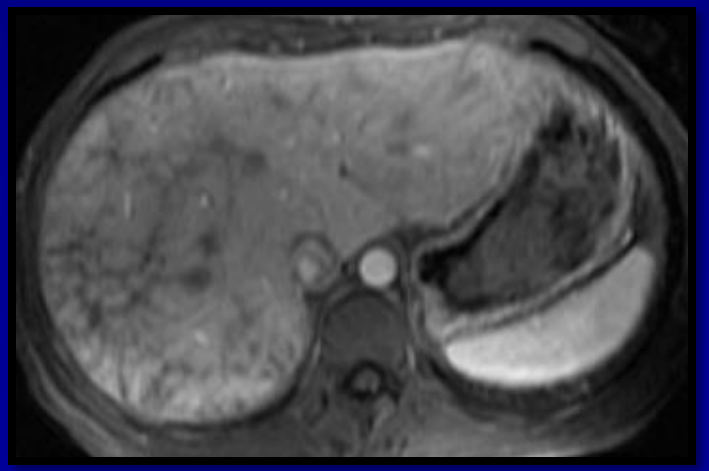
Mortele KJ, Abdom Imaging 2002
Rubbia-Brandt, Ann Oncol 2004
Lassau, Radiology 2000
Lassau, Trasnplantation 2002
Erturk SM, AJR 2006

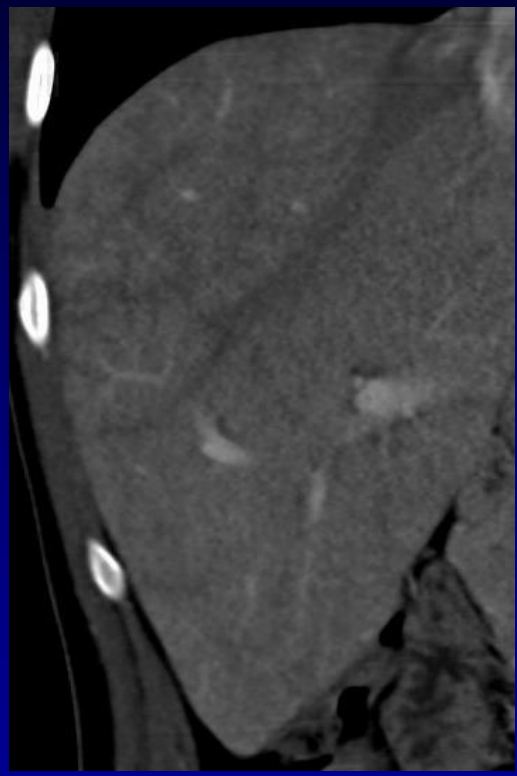
Sinusoidal obstruction syndrome

Imaging Features

- Narrowed IVC & hepatic veins
  - RHV=2-3 mm
- delayed enhancement HV & IVC
- ‘Mosaic’ enhancement
- Ascites and effusions
- Hepatomegaly
- Periportal & GB edema
- no preferential enhancement or hypertrophic changes caudate lobe







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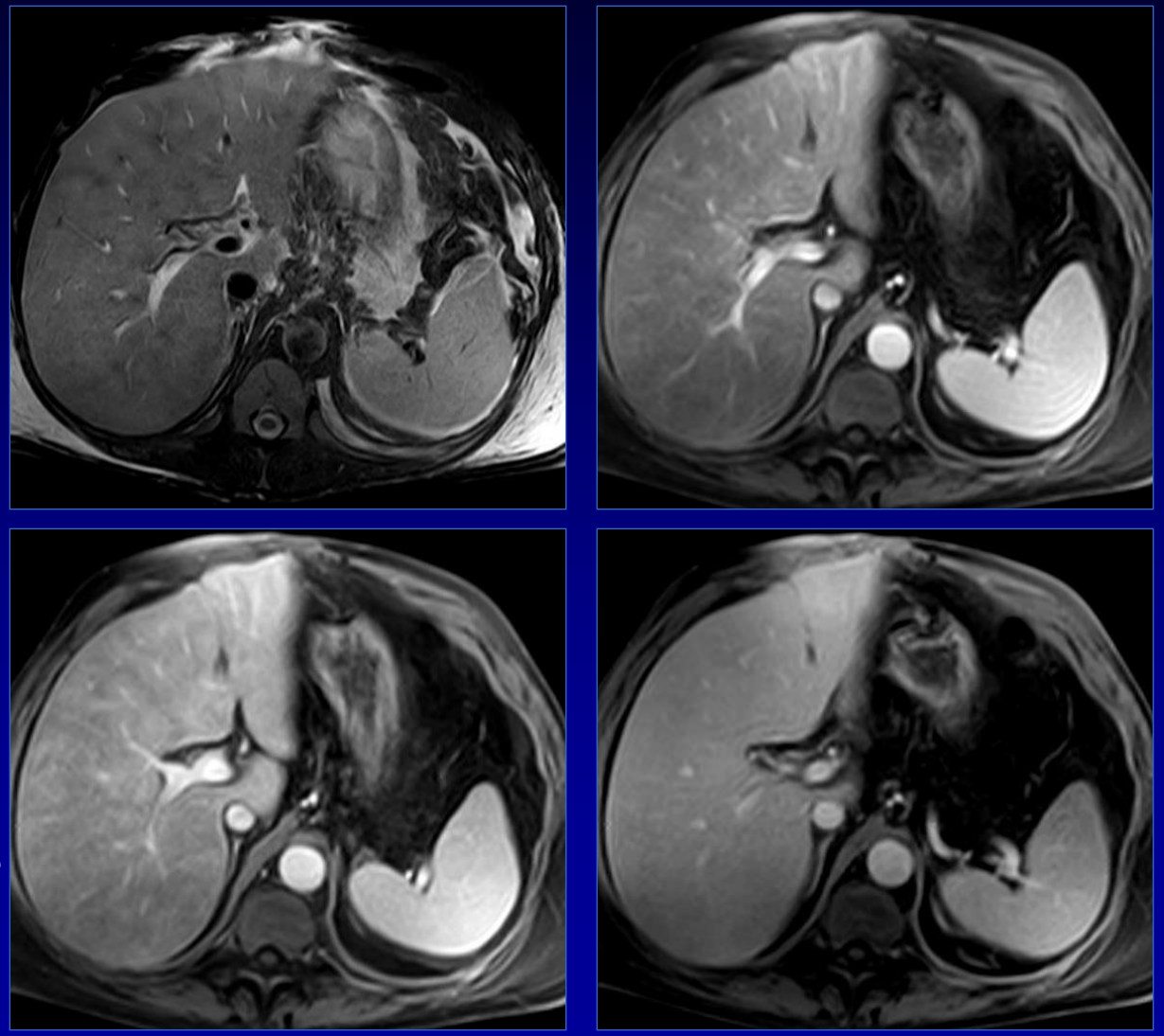


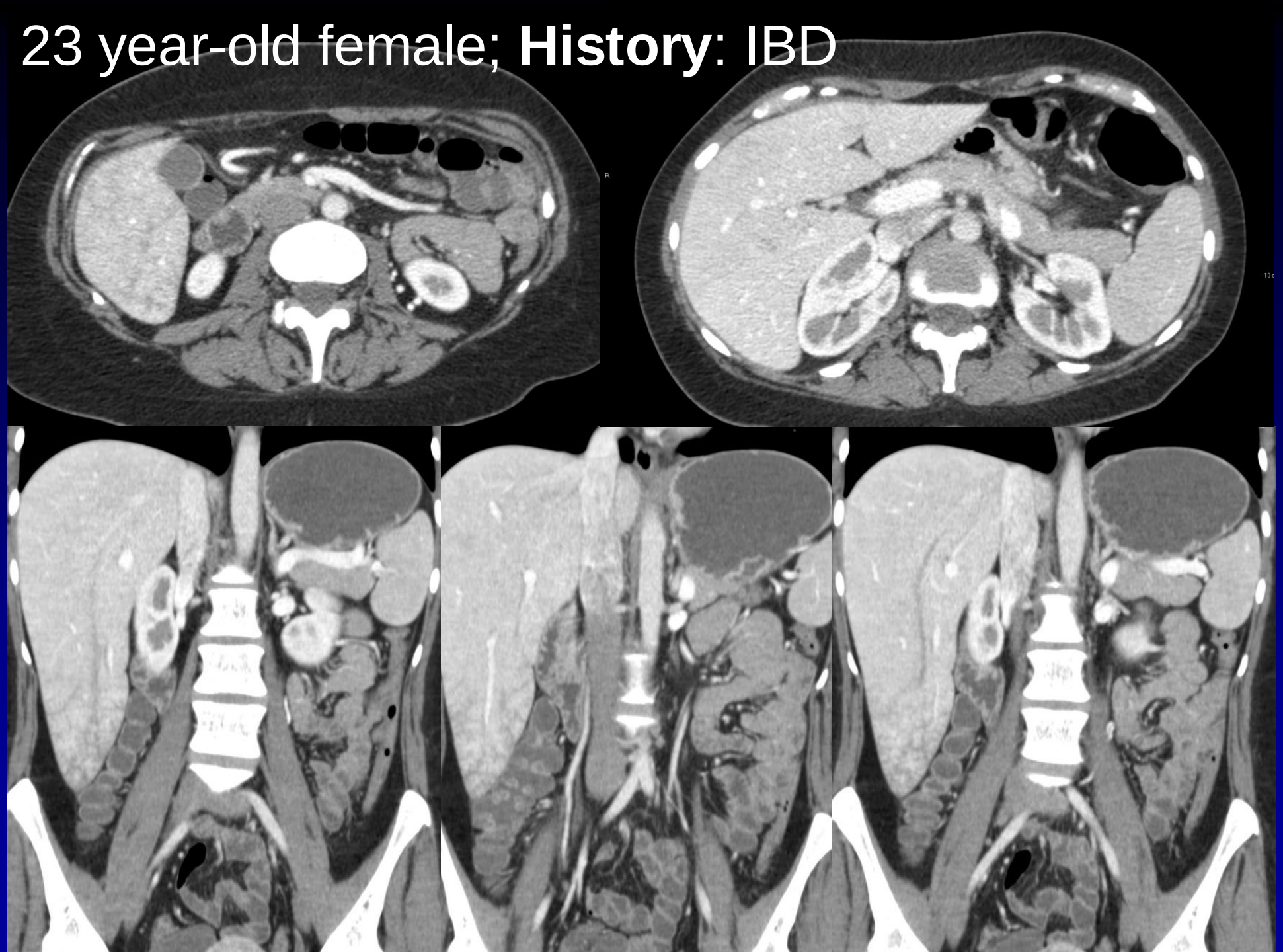
Sinusoidal dilatation

Causes

1) Venous outflow obstruction (heart failure, pericardial dx, BCS, SOS)
2) Non-obstructive (acute inflammatory/infectious conditions, OCP, chronic conditions)

Brancatelli et al, Hepatic sinusoidal dilatation; Abdom Radiol 2017

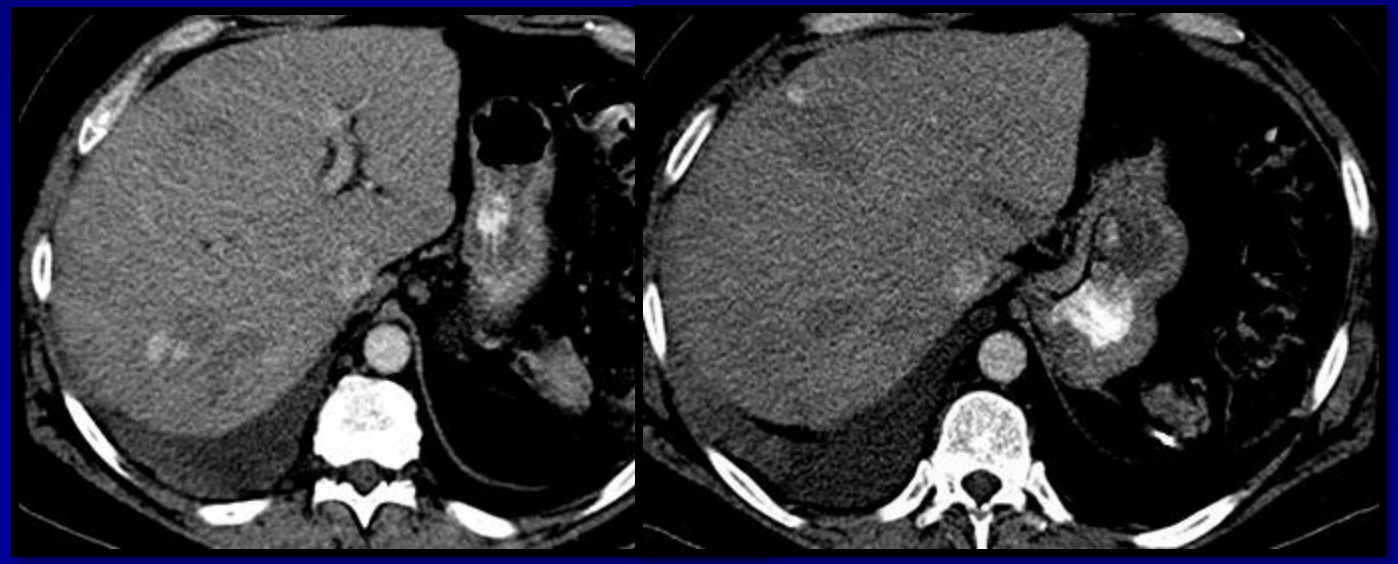
23 year-old female; **History**: IBD



Peliosis hepatis

Blood lakes with lack of endothelium

- Predisposing factors:
  - chronic debilitating conditions (anabolic steroids)
  - *Bartonella henselae* infection (immunodeficiency)
  - SOS
  - Hematologic disorders



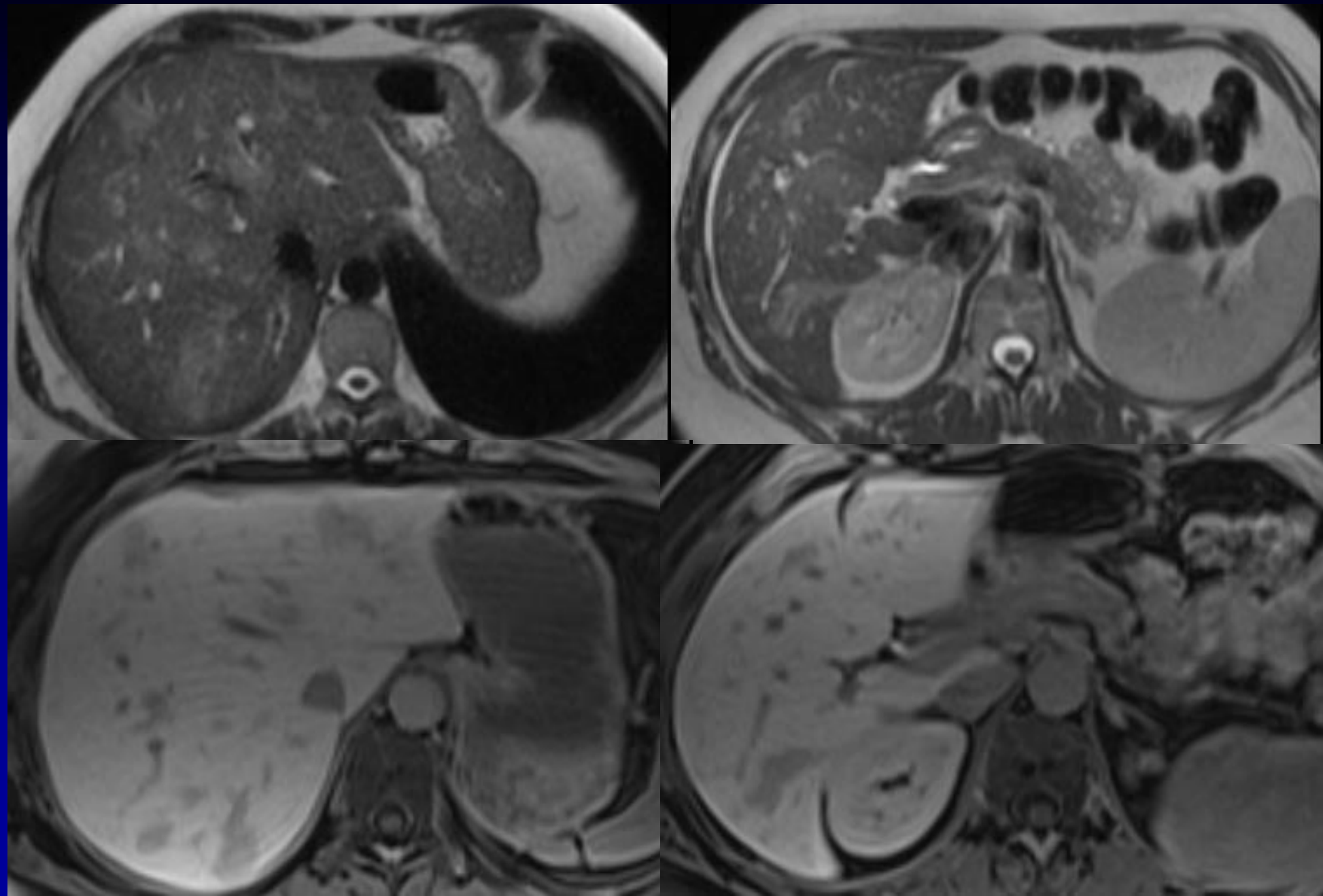
< 3 cm

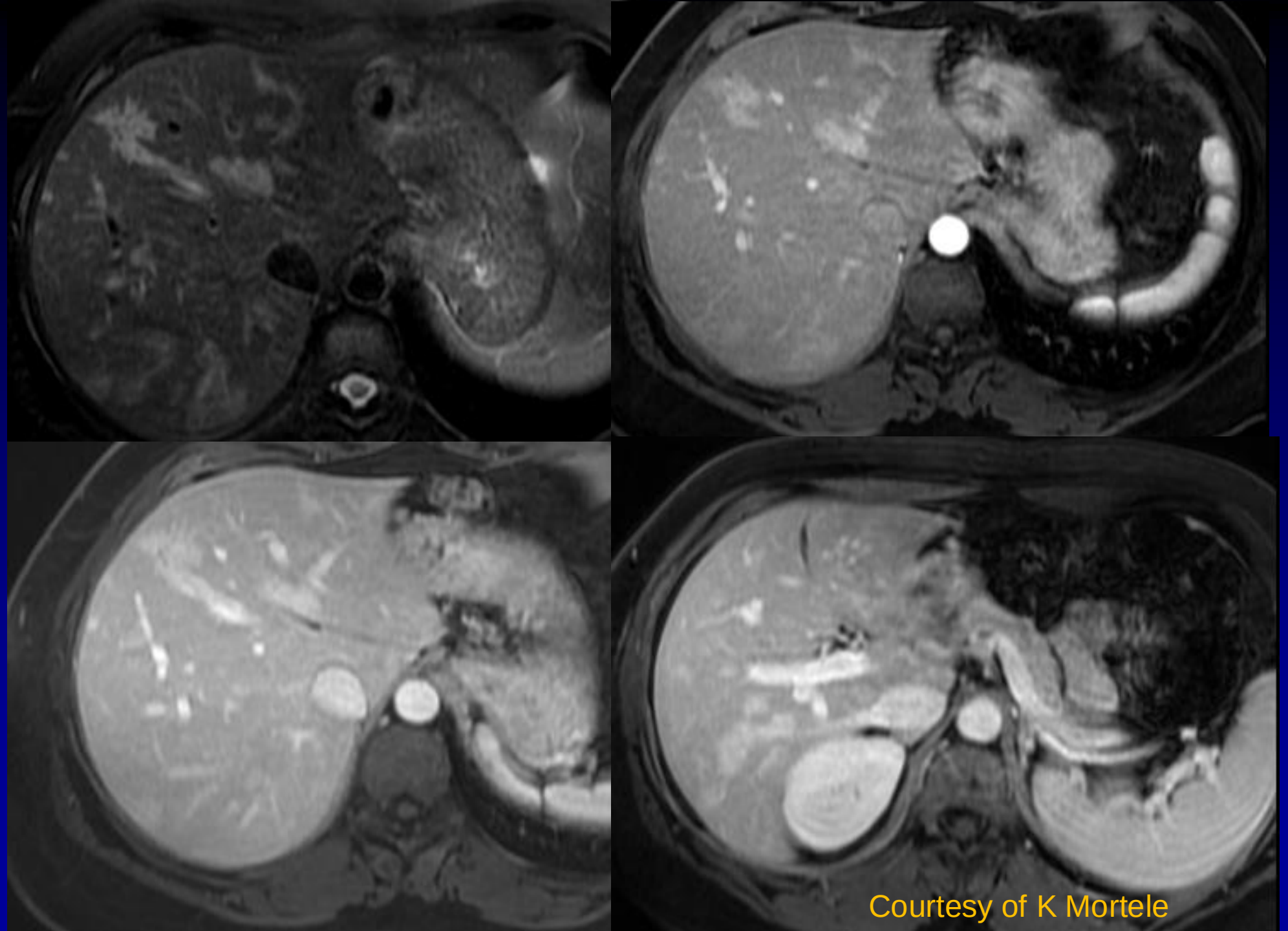
hypodense

hypervascular foci

no mass effect

47 year old male with lung carcinoid

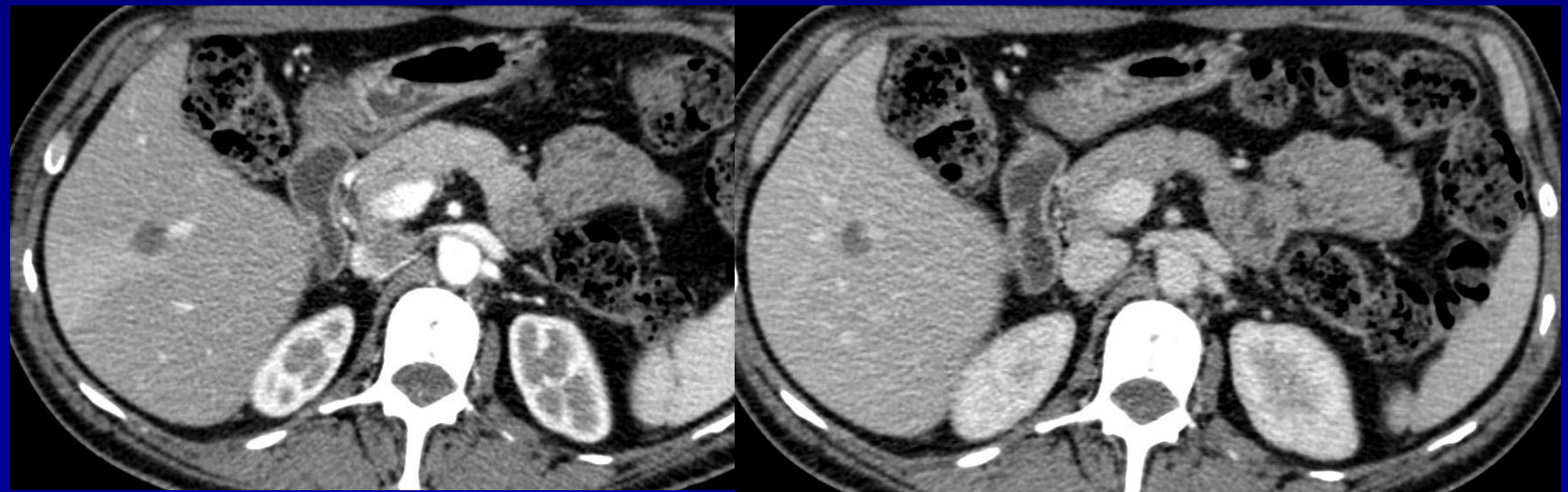


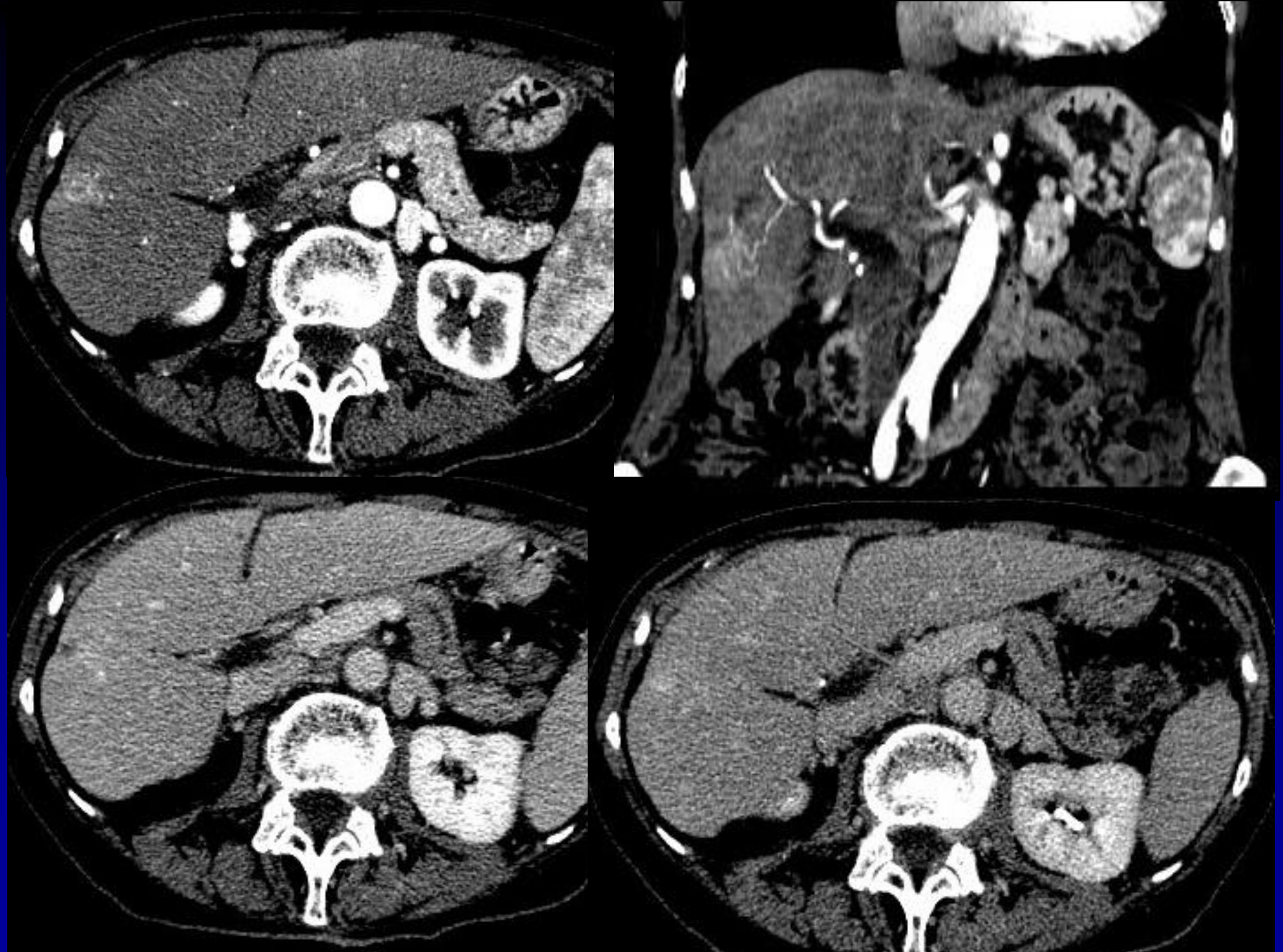

Courtesy of K Mortele

THAD & THID

- **THAD**: transient hepatic attenuation difference
- **THID**: transient hepatic intensity difference

- **Imaging Features**
  - wedge-shaped (90%), portal venous territory
  - late arterial phase: hyperenhancing
  - portal venous phase: isodense or isointense





- Vascular diseases are common
- From asymptomatic to life-threatening
- Classification:
  - Diseases of outflow/inflow/sinusoids
- Ddx with hypervascular tumors
- Imaging useful for more accurate diagnosis >> less need for further imaging

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