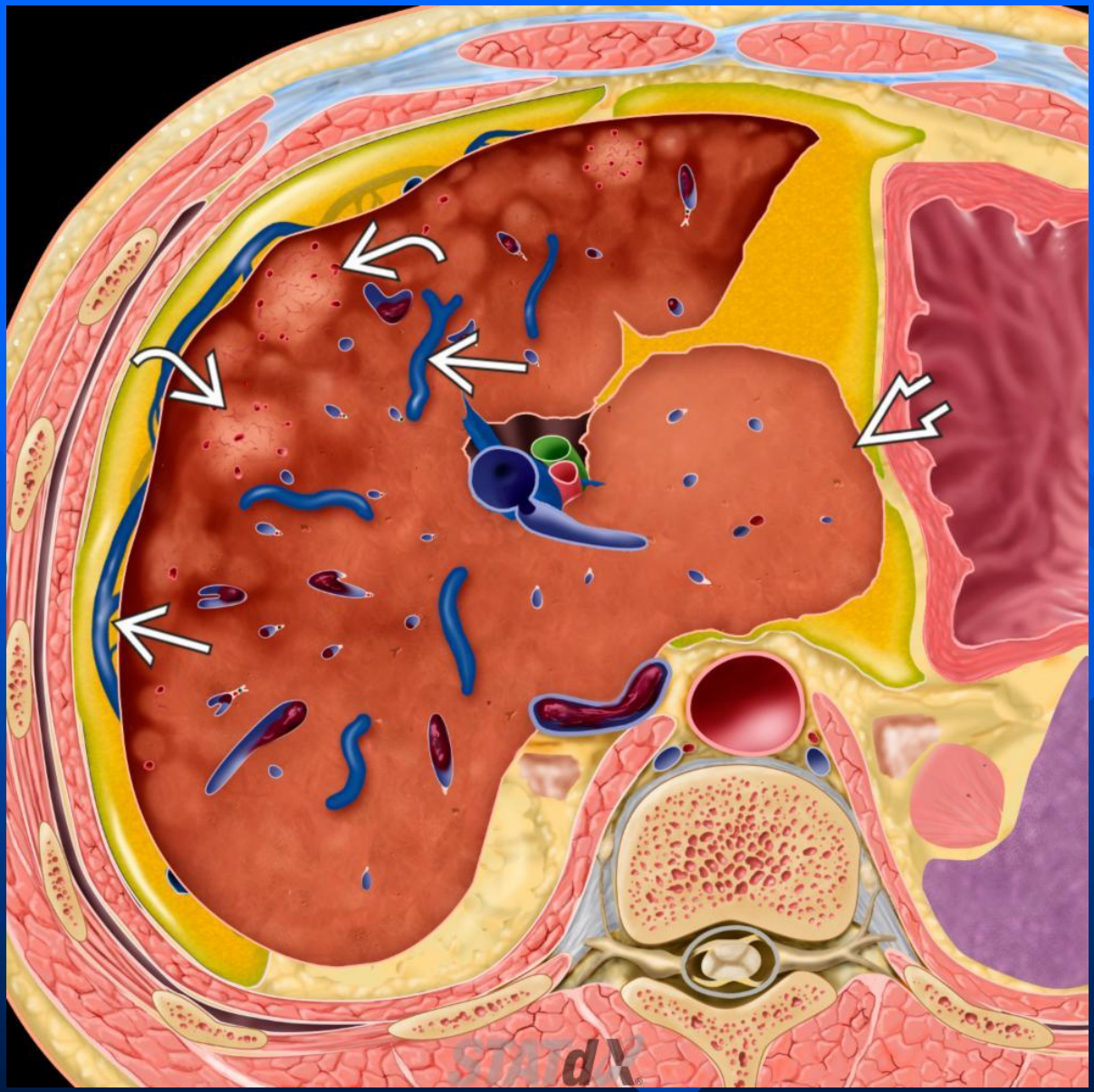
Budd-Chiari syndrome

- Global or segmental hepatic venous outflow or IVC obstruction.
- Etiology in western populations is usually a hypercoagulable condition.
- **Age**
  - Onset typically occurs in young adulthood
- **Gender**
  - M < F
- **Epidemiology**
  - Primary (congenital-membranous): Common in Japan, India, Israel, South Africa
  - Secondary (thrombotic): Most common in western countries, usually due to hypercoagulable state
  - Secondary (nonthrombotic): 2nd most common in western countries.
- **Consider**
  - Distinguish BCS from cirrhosis; do not mistake focal large regenerative nodules of BCS for HCC
- **Image Interpretation Pearls**
  - Recognize large benign regenerative nodules and caudate pseudotumor

Imaging

- Characteristic findings: Nodular regenerative hyperplasia in a dysmorphic liver with venous collateral and ascites
- CECT and MR show hypertrophied caudate lobe with atrophy and necrosis of peripheral liver
  - May simulate a large neoplasm within caudate lobe
- Intrahepatic and systemic venous collaterals bypass obstructed hepatic veins and IVC
  - Spider web pattern of hepatic venous collaterals on CT, MR, angiography
- Large regenerative nodules (form of nodular regenerative hyperplasia) are characteristic of chronic BCS
  - Imaging and histology similar to FNH
  - May have peripheral halo and central scar
  - Hypervascularity persists into venous phase without washout
  - Uniform or peripheral delayed retention (bright) on gadoxetate-enhanced MR
- Absent, reversed, or flat flow in hepatic veins; reversed flow in IVC on color Doppler US

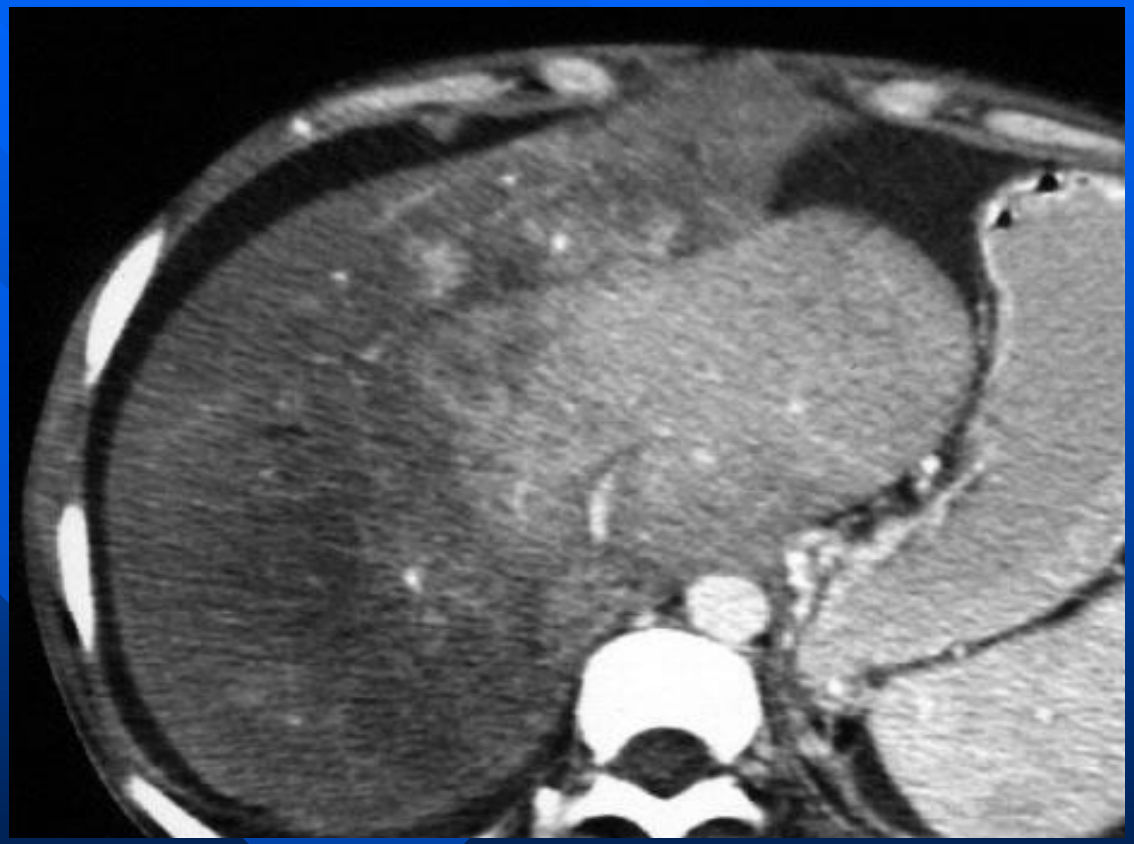


Axial anatomic illustration of Budd-Chiari syndrome demonstrates ascites, venous collaterals (white solid arrow), heterogeneous hepatic parenchyma due to centrilobular necrosis, and hypervascular regenerative nodules (white curved arrow). Note the sparing of the caudate lobe with hypertrophy (white open arrow), as well as the thrombosed IVC.

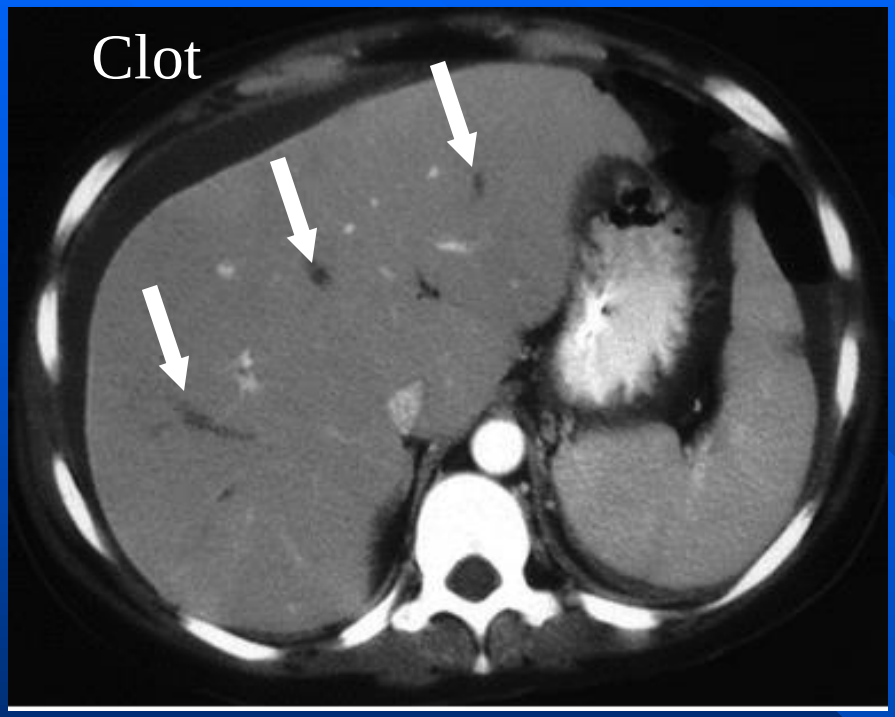
Axial CECT shows caudate hypertrophy, a large caudate collateral vein (white solid arrow), and peripheral atrophy and heterogeneity. The hepatic veins were occluded.

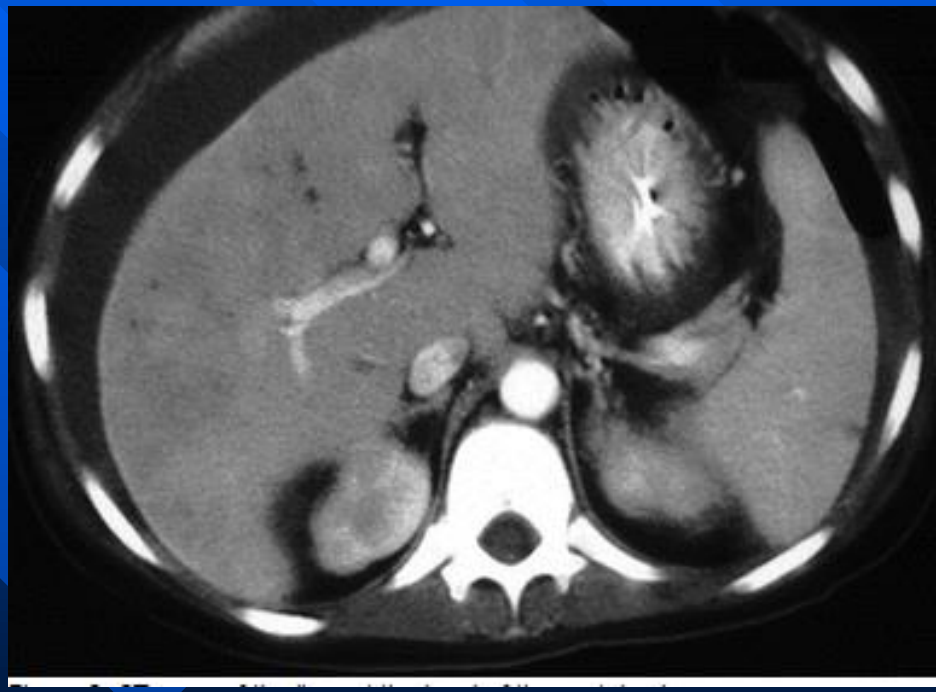
Budd-Chiari syndrome

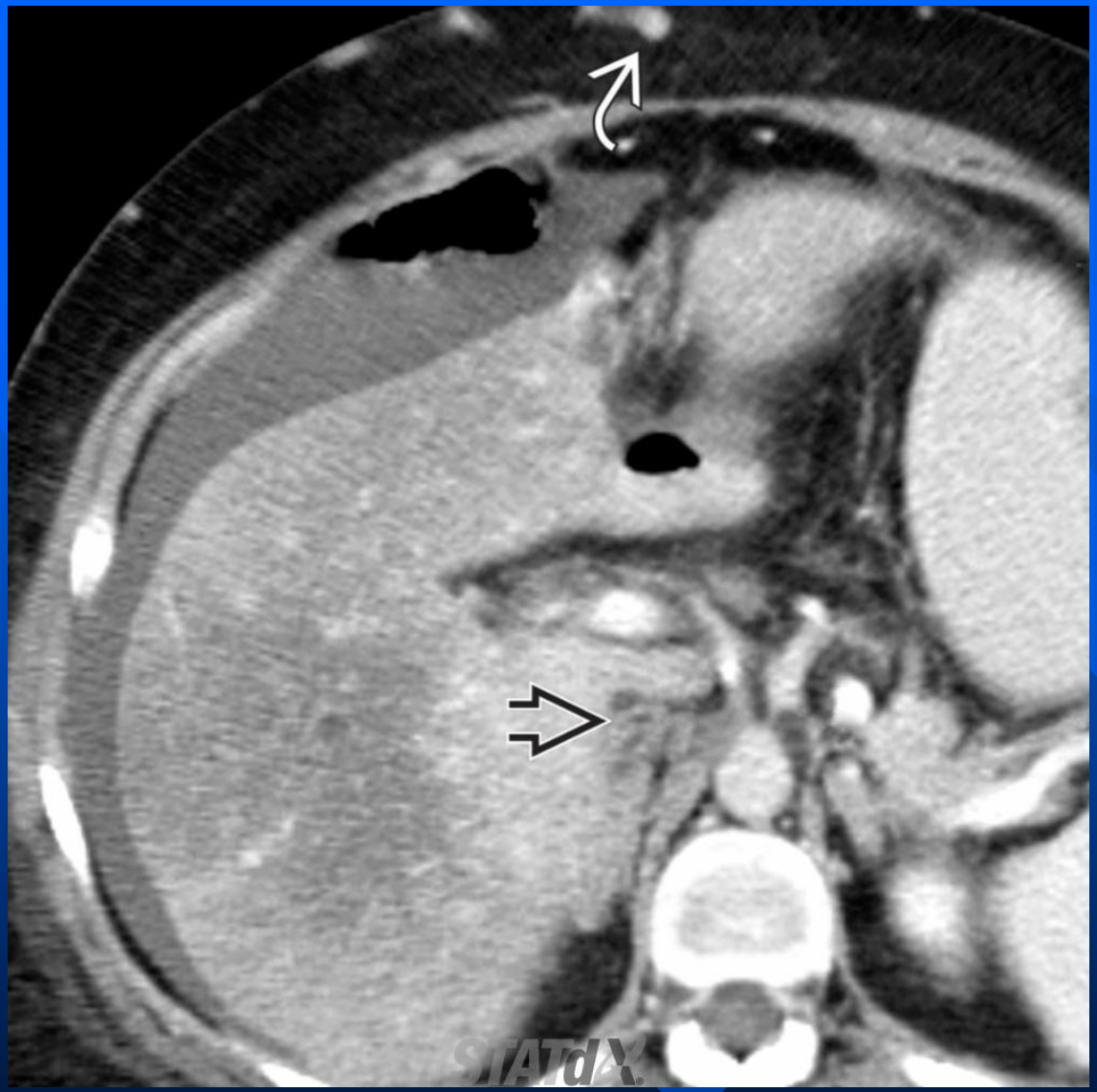
- Characterized on imaging
  - Ascites
  - Caudate hypertrophy
  - Peripheral atrophy
  - Prominent collateral veins.



Budd-Chiari syndrome







Axial CECT shows numerous collateral veins (white curved arrow) bypassing the obstructed IVC (black open arrow). Ascites is present, and the liver enhances heterogeneously. The central right and caudate lobe enhance normally, and the peripheral right and left lobes enhance poorly. These are typical findings of subacute BCS.