BUDD CHIARI SYNDROME

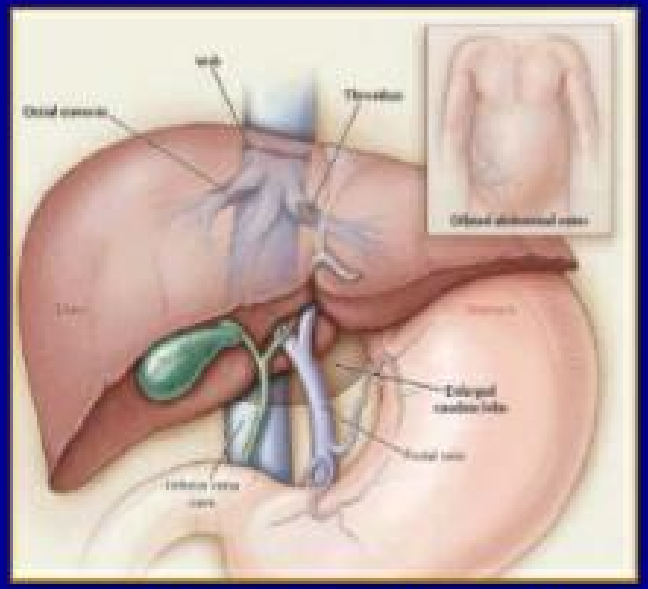
Definition

- The budd- chiari syndrome is a heterogenous group of disorders characterized by hepatic venous outflow obstruction at the level of the hepatic venules, the large hepatic veins, the inferior venacava or the right atrium.
- Where as Hepatic veno-occlusive disease refers to obstruction of hepatic venous outflow at the level of the central or sublobular hepatic veins, or both

Table 1. Causes of the Budd–Chiari Syndrome.

- **Common causes**
  - Hypercoagulable states
    - Inherited
      - Antithrombin III deficiency
      - Protein C deficiency
      - Protein S deficiency
      - Factor V Leiden mutation
      - Prothrombin mutation
    - Acquired
      - Myeloproliferative disorders
      - Paroxysmal nocturnal hemoglobinuria
      - Antiphospholipid syndrome
      - Cancer
      - Pregnancy
      - Use of oral contraceptives
- **Uncommon causes**
  - Tumoral invasion
    - Hepatocellular carcinoma
    - Renal-cell carcinoma
    - Adrenal carcinoma
  - Miscellaneous
    - Aspergillosis
    - Behçet's syndrome
    - Inferior vena caval webs
    - Trauma
    - Inflammatory bowel disease
    - Dacarbazine therapy
  - Idiopathic

Budd-Chiari syndrome



Occlusion of a single hepatic vein usually clinically silent

Two or three hepatic veins can be occluded without clear symptoms

Clinical features

- The clinical presentation of the Budd–Chiari syndrome depends on the extent and rapidity of hepatic-vein occlusion and on whether a venous collateral circulation has developed to decompress the liver sinusoids.
- Abdominal pain, hepatomegaly, and ascites is the clinical triad and present in almost all patients with the Budd–Chiari syndrome.
- It can be classified as- fulminant

  acute

  subacute

  chronic

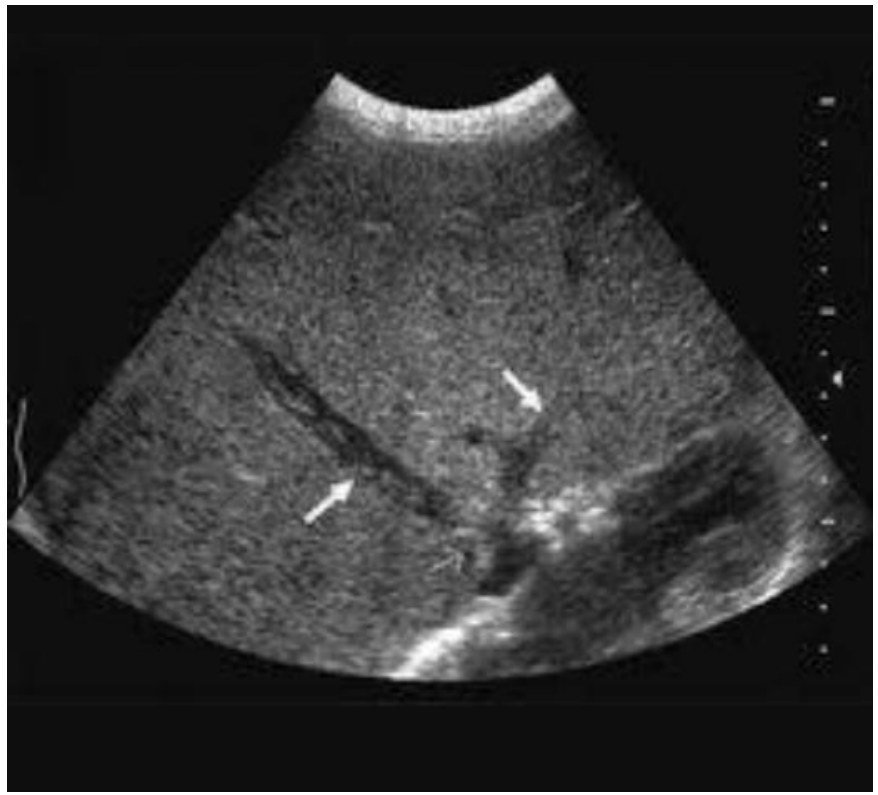
- Fulminant form : hepatic encephalopathy
- Acute form : short duration, intractable ascites, no venous collaterals.
- Subacute form: most common type,
  more insidious onset;
  portal hypertension and ascites
  hepatic necrosis may be minimal, sinusoids decompressed by a portal and hepatic venous collateral circulation.
- chronic form: It is manifested as cirrhosis.

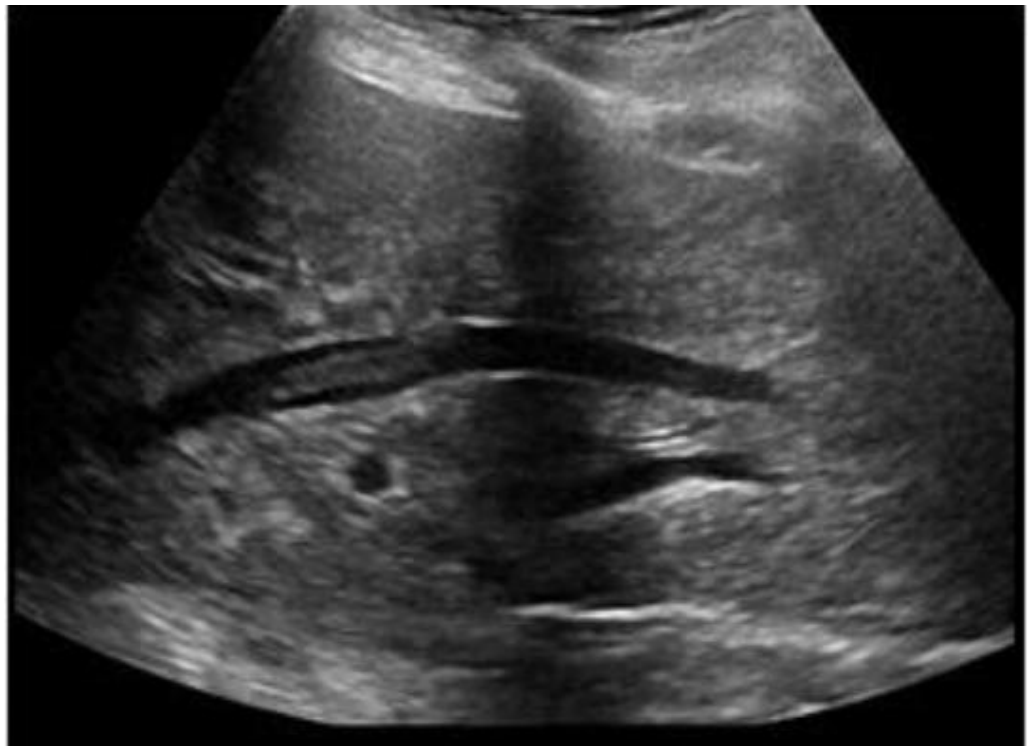
Imaging

- Doppler ultrasonography of the liver is the initial investigation of choice
- CT : Better delineation of the venous anatomy and the configuration of the liver when a transjugular intrahepatic portosystemic shunt is being considered.
- MR: when CT is contraindiacted
- ECHO : To r/o constrictive pericarditis , TR, Rt atrial myxoma
- Hepatic venography and Cavography

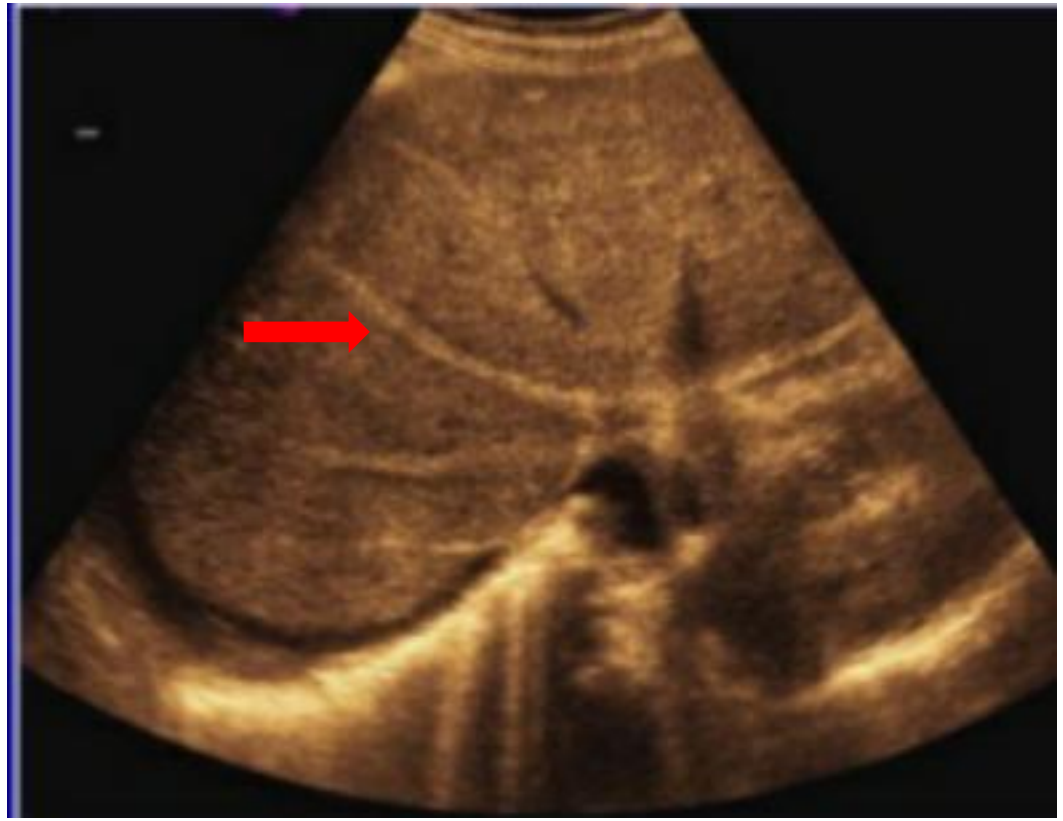
Doppler ultrasound

Obstruction &/or collaterals of HV or IVC*	
Obstructed HV	Presence of solid endoluminal material Hyperechoic cord replacing normal vein Reversed flow in large hepatic vein Dilatation of vein upstream to obstacle
HV collaterals	**Sipder web** in vicinity of HV ostia Subcapsular or HV to intercostal or HV veins Caudate lobe hypertrophy with dilated veins
IVC	Web – Thrombosis – Inversion of flow





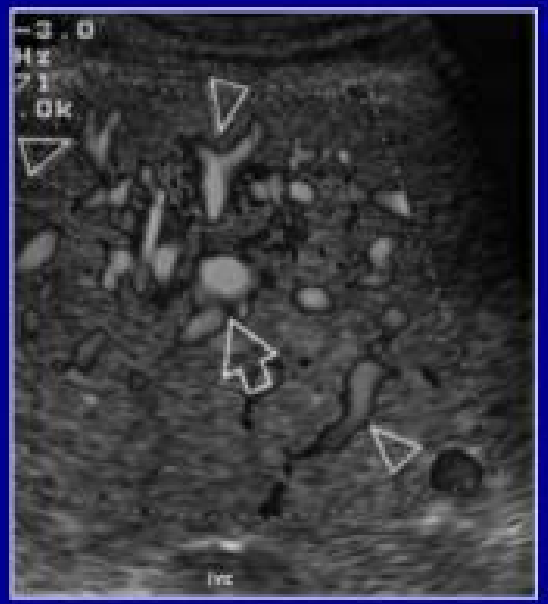
hyperechoic cords



Hepatic veins transformed to fibrous cords

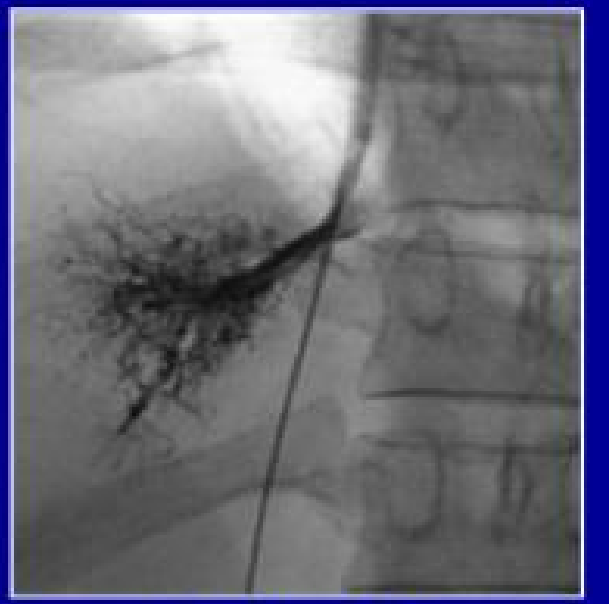
BCS / Sipder web in vicinity of HV ostia

Gray-scale US

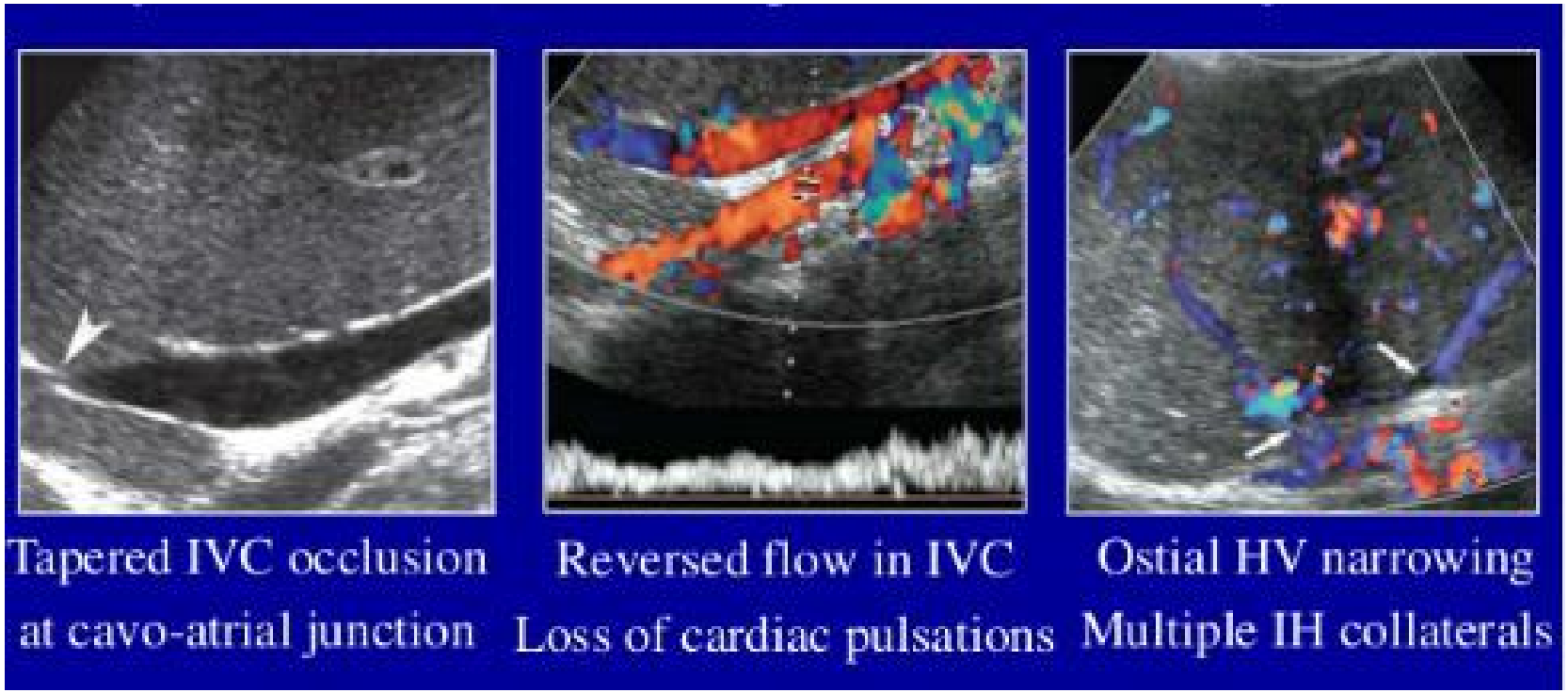


Small interwoven veins near IVC

Hepatic venogram

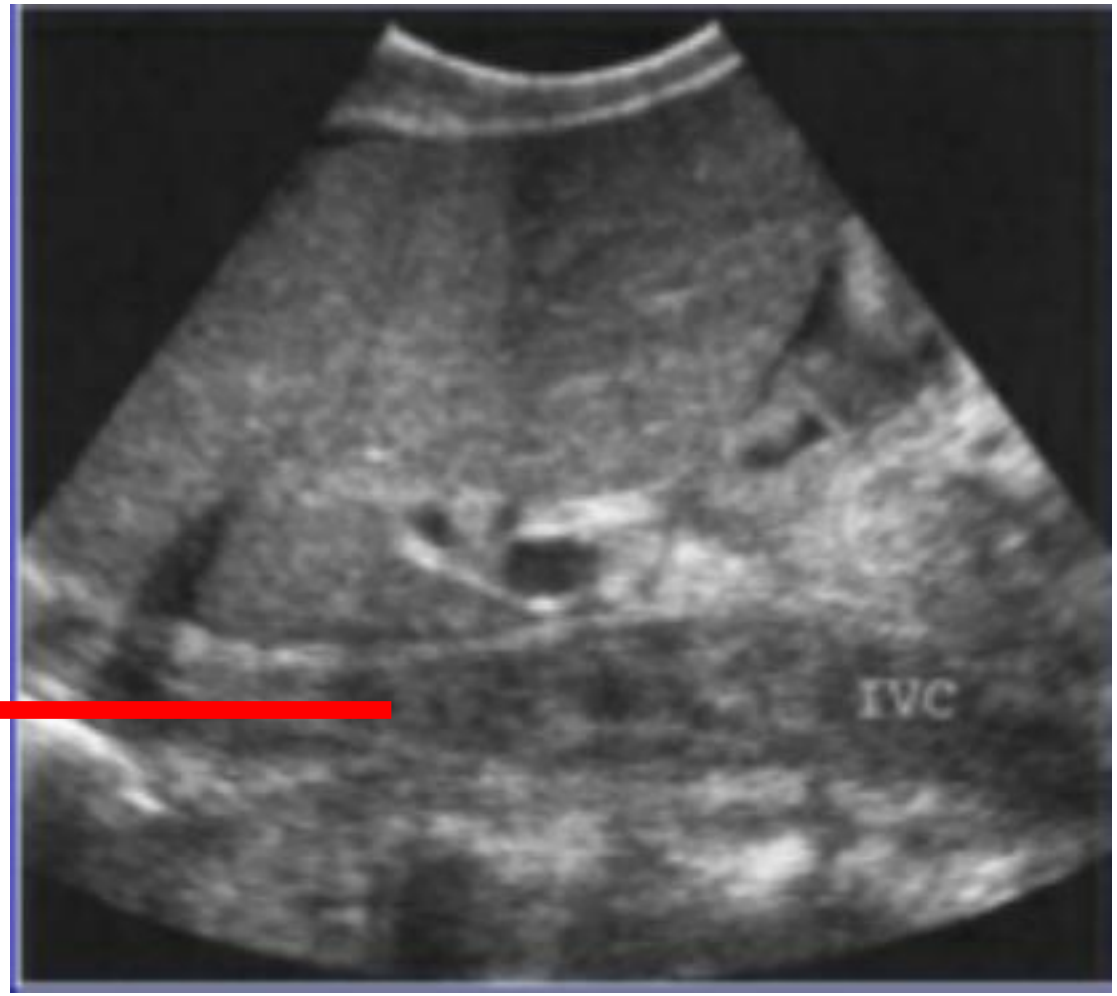


Typical **"spider web"** pattern

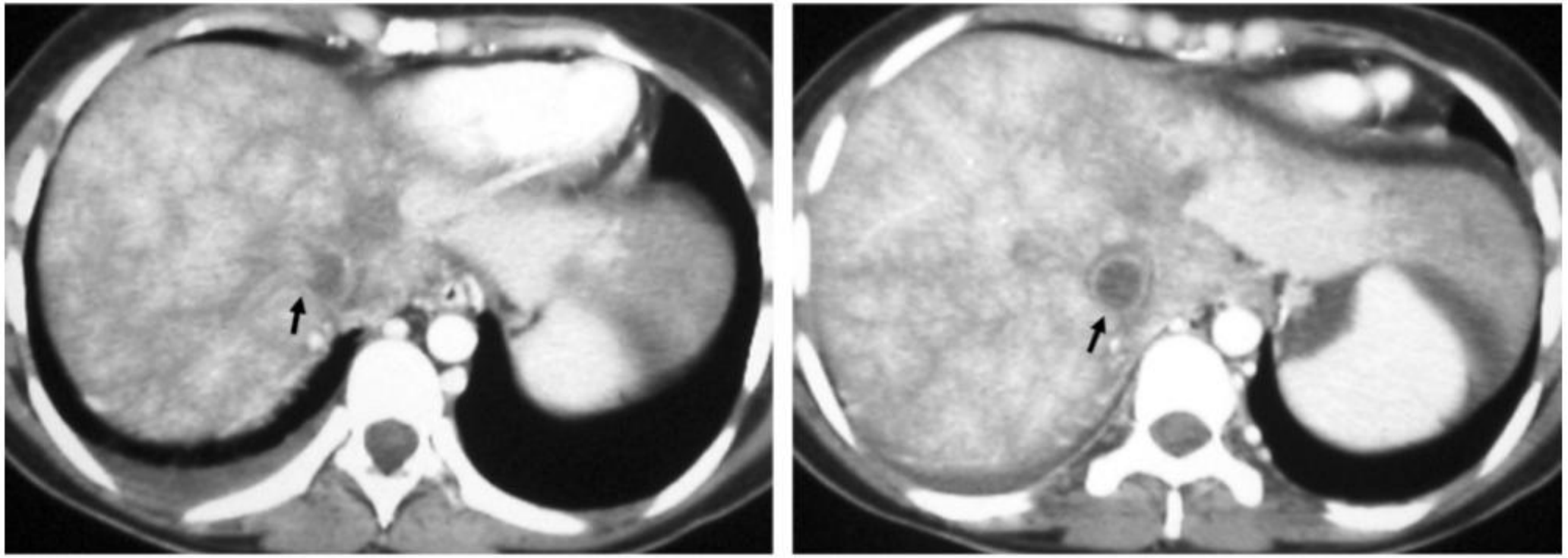

Tapered IVC occlusion
at cavo-atrial junction
Reversed flow in IVC
Loss of cardiac pulsations
Ostial HV narrowing
Multiple IH collaterals

Secondary BCS due to renal cell carcinoma

echogenic thrombus
In IVC



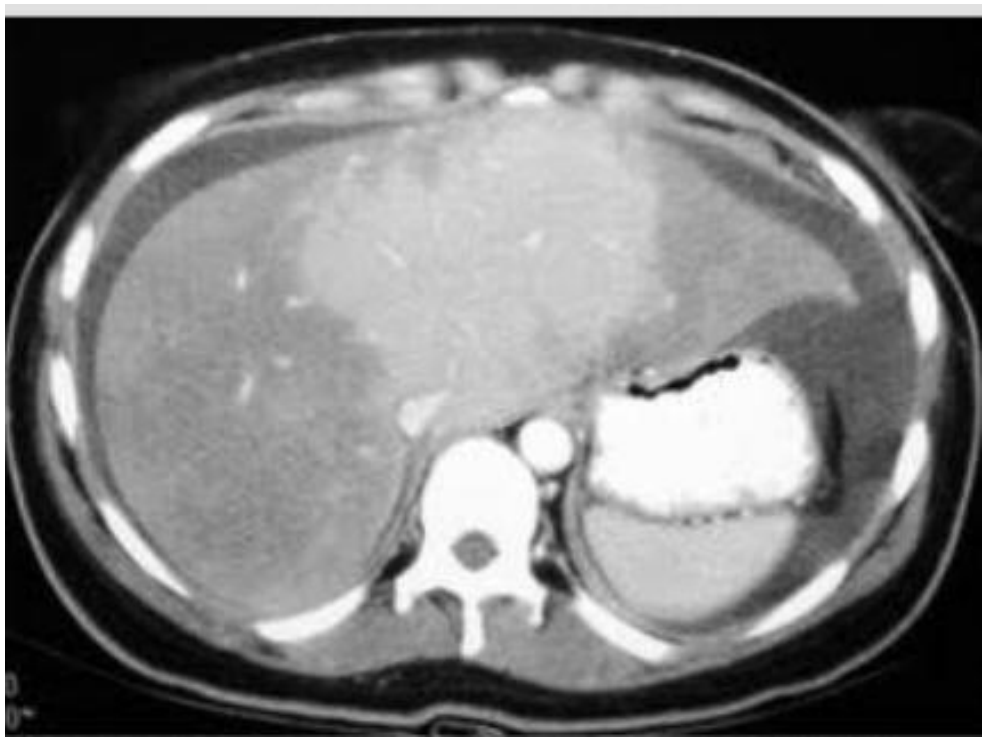
CECT

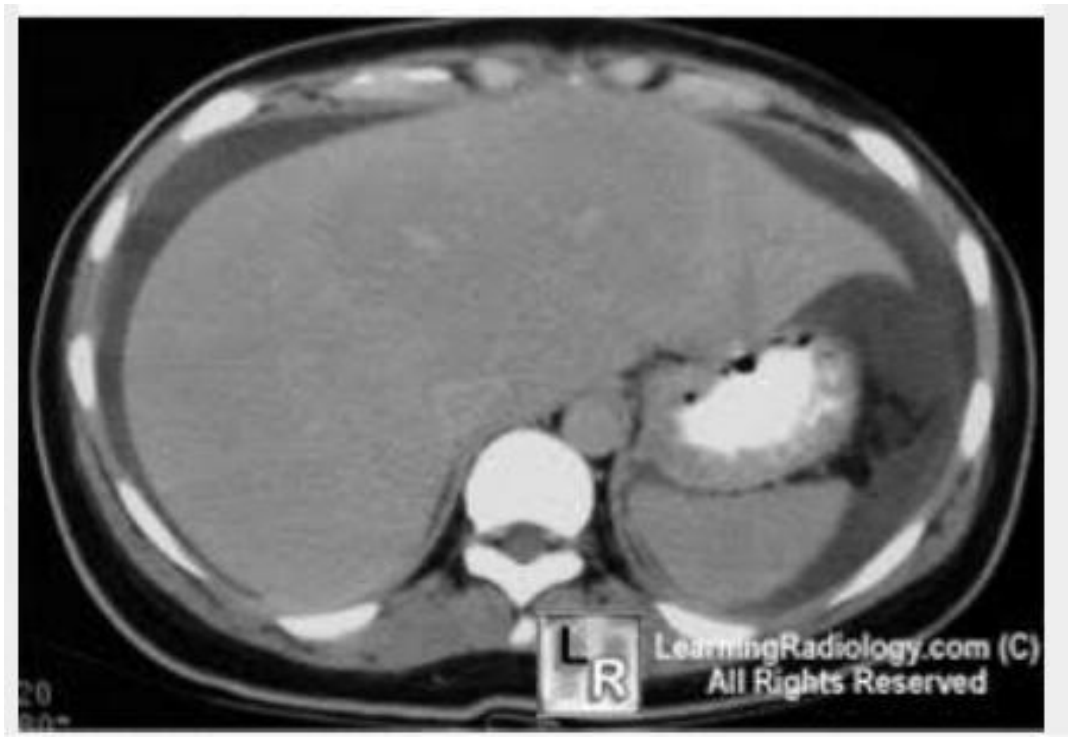


CECT shows patchy enhancement of the liver parenchyma, hypertrophy of the left hepatic lobe, and thrombosis of the hepatic veins and IVC (arrow).

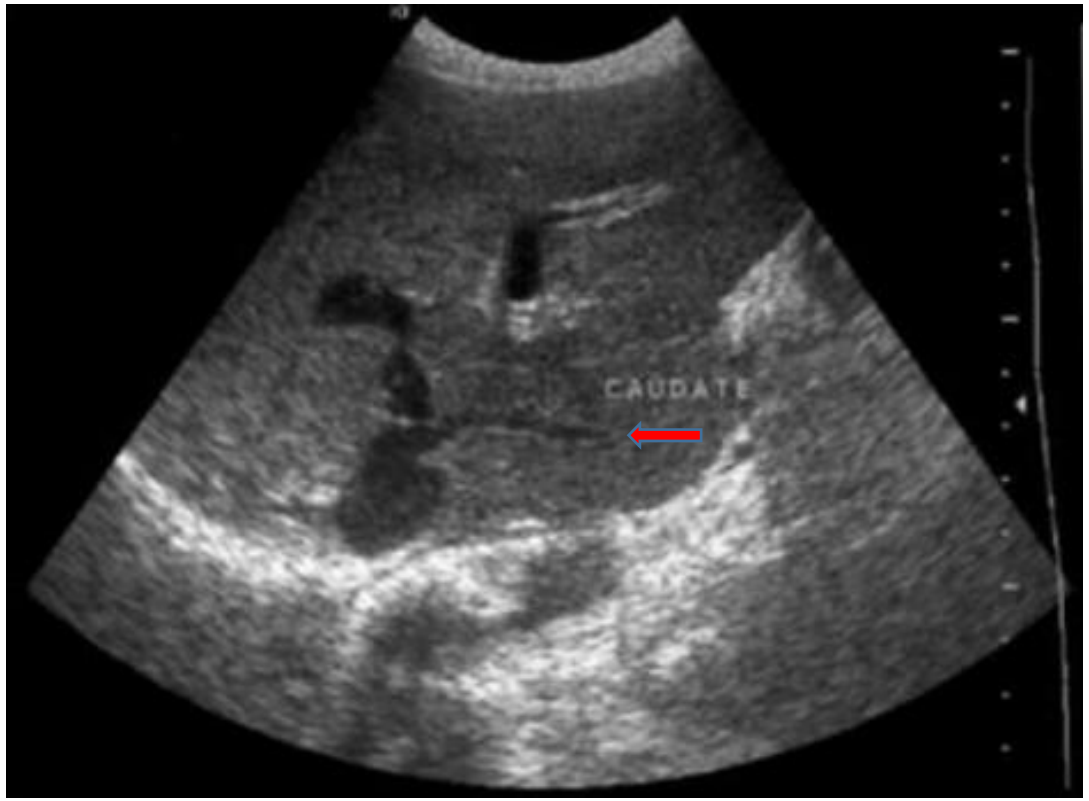
flip flop phenomenon

Early enhancement of the caudate lobe & central liver around IVC with delayed enhancement of the peripheral Liver with accompanying central low density

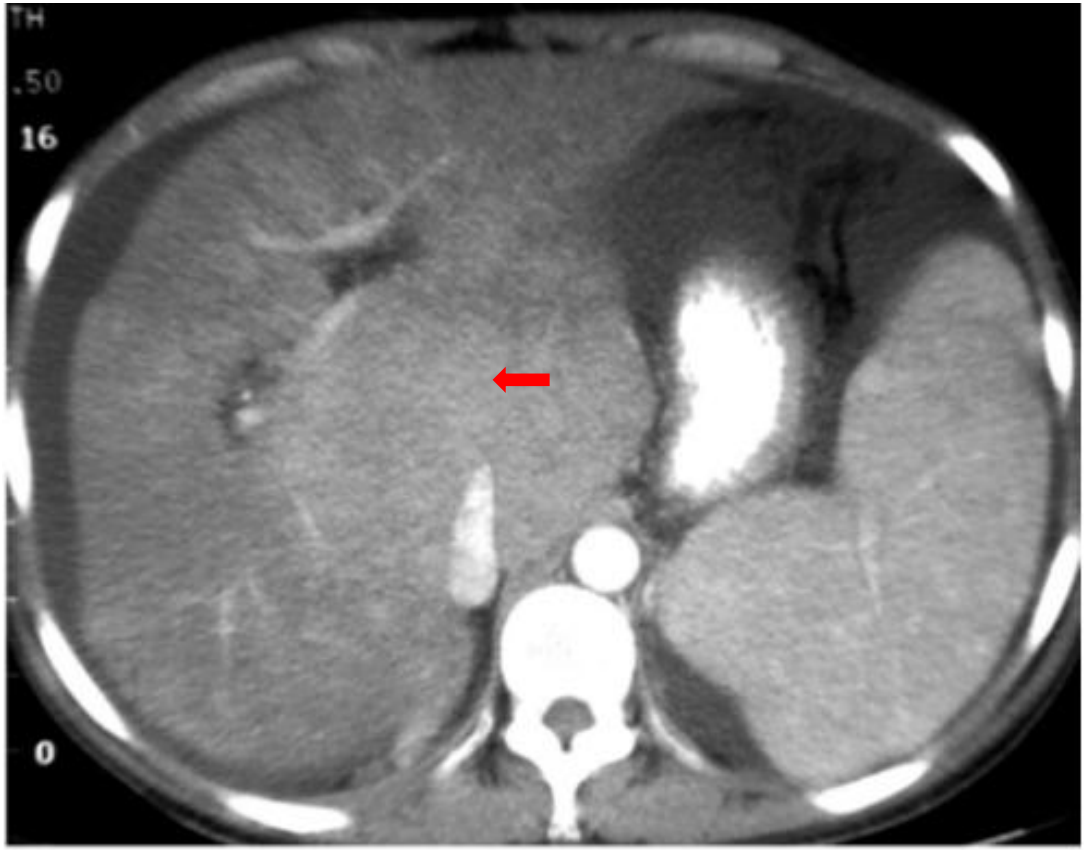




Acute form



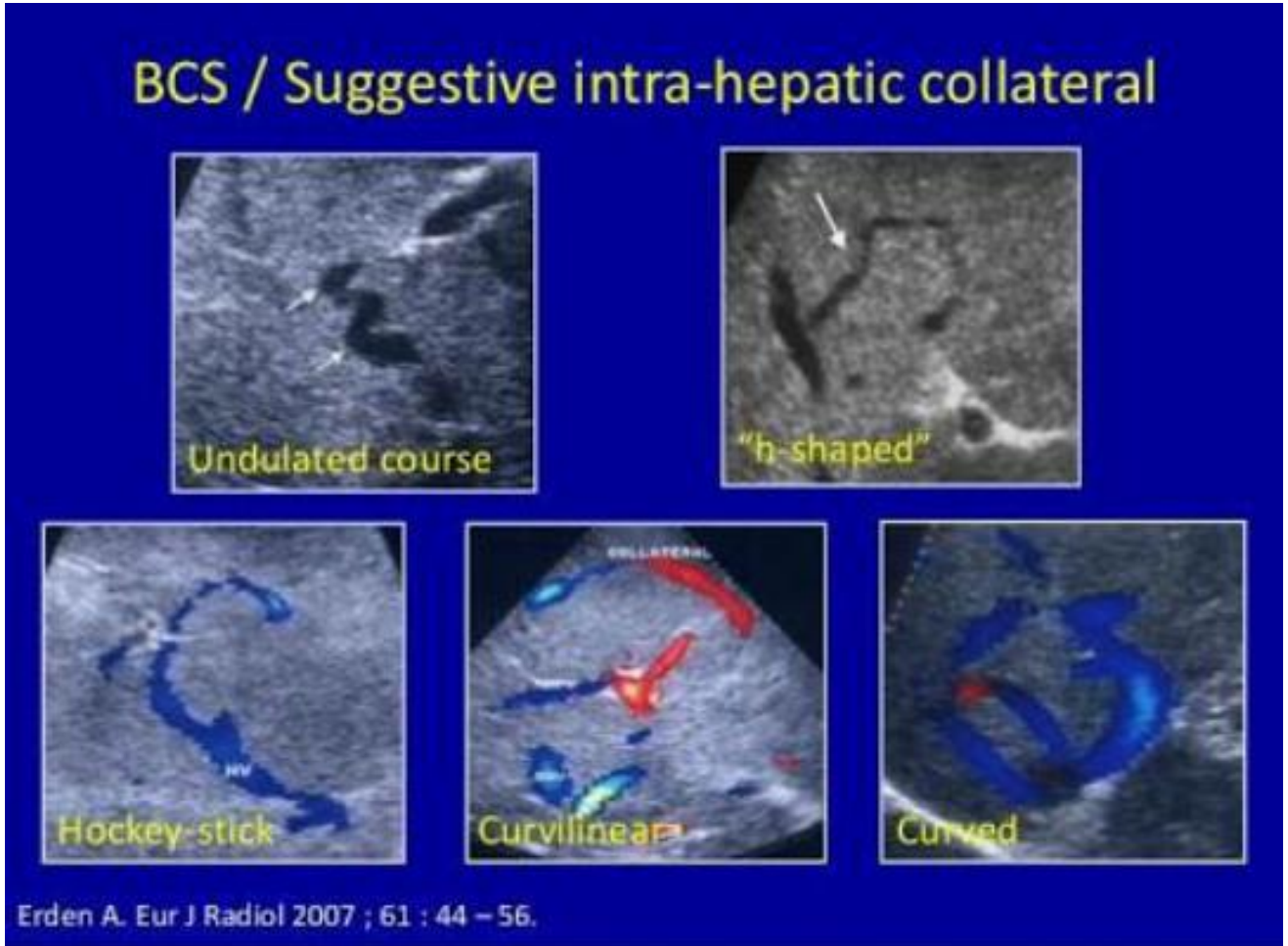
USG shows caudate lobe hypertrophy

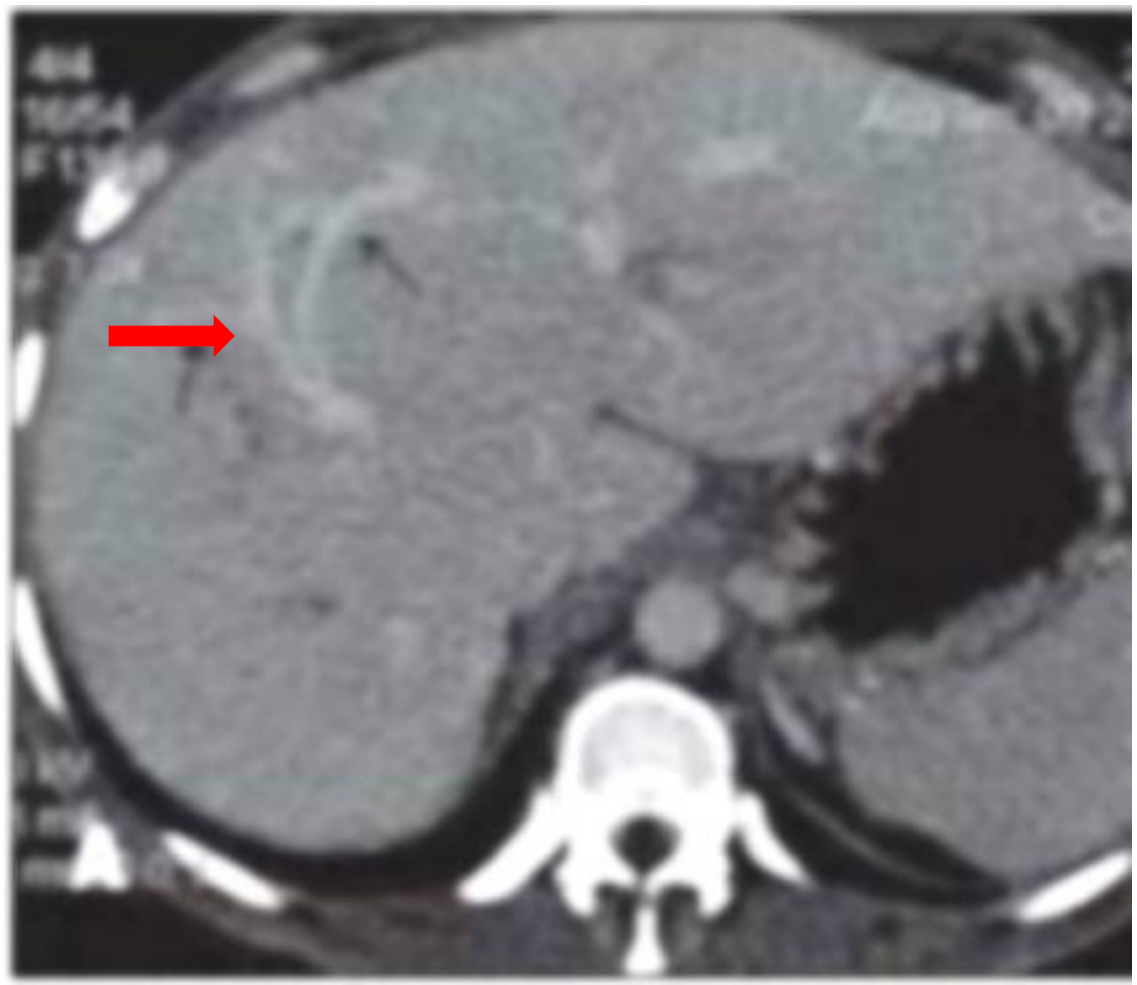


CECT shows ascites and stronger enhancement in the caudate lobe and central portion of the liver parenchyma than in the periphery.

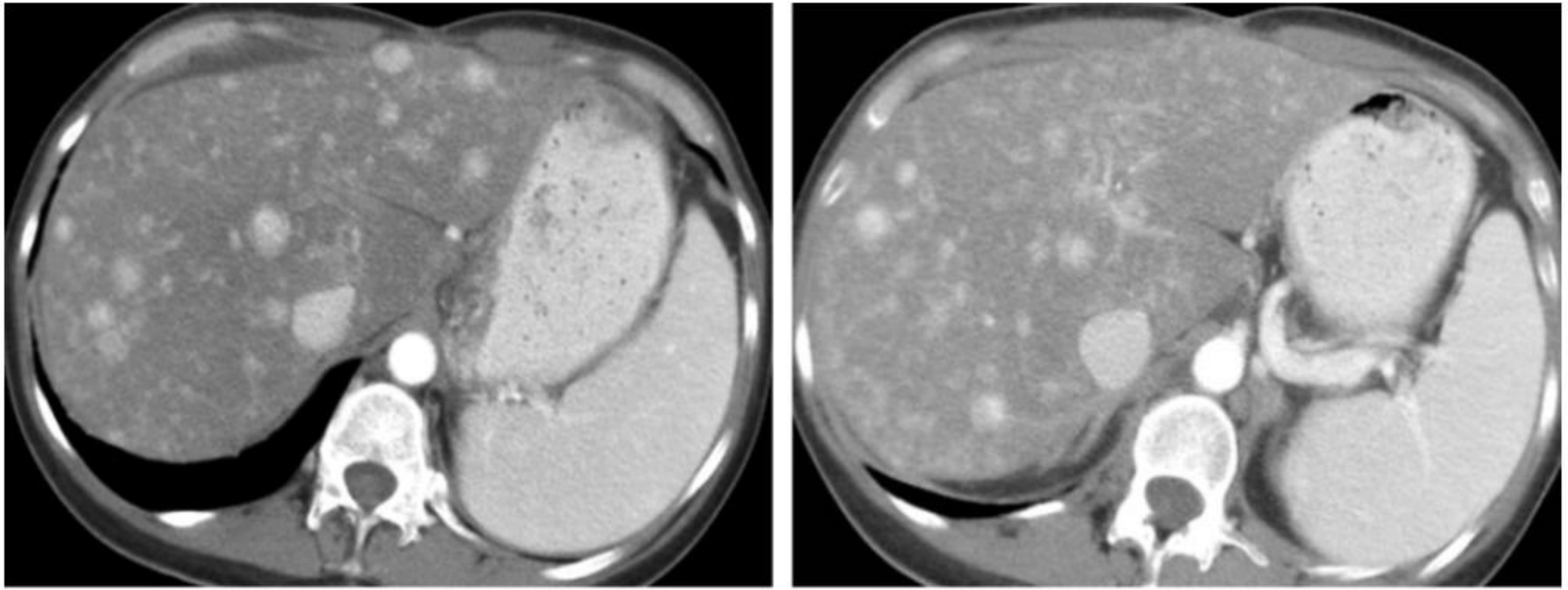
Sub-acute form

Characterized by development of multiple porto-systemic collaterals



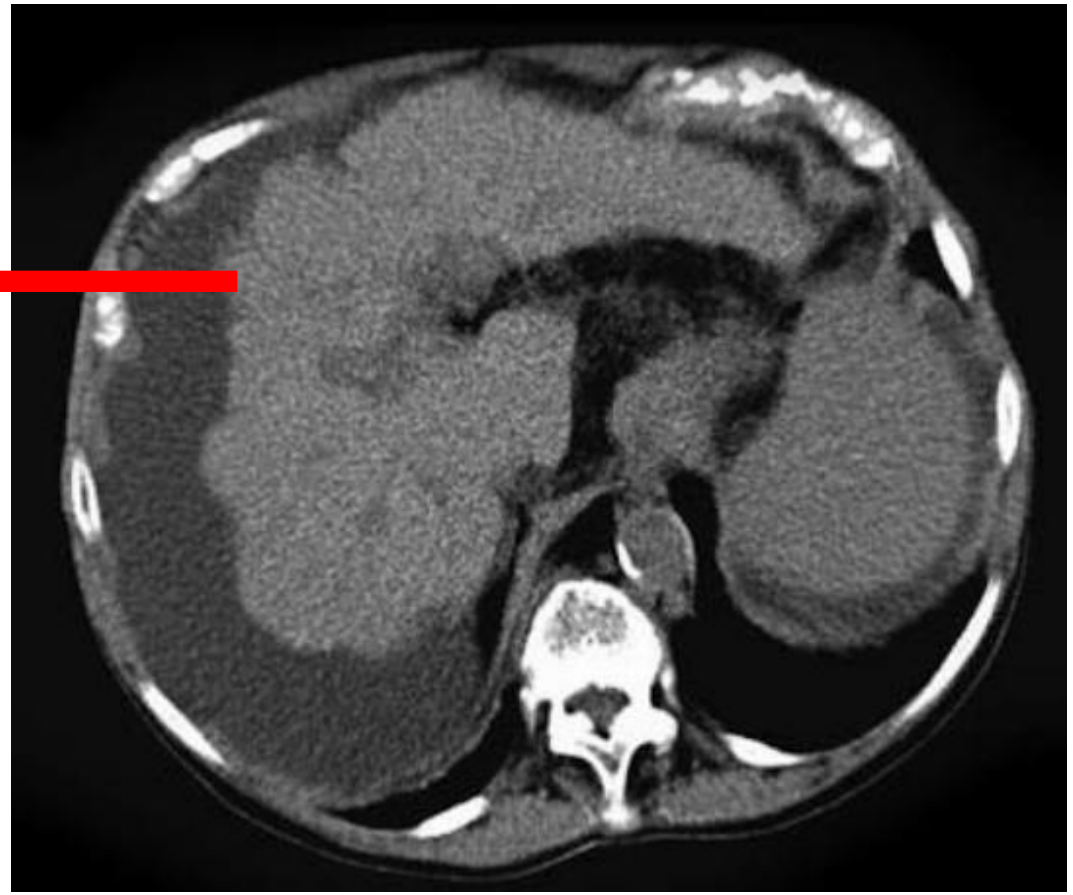


chronic form



CECT showing multiple regenerative nodules with marked homogenous enhancement

Chronic form developing into cirrhosis



Management

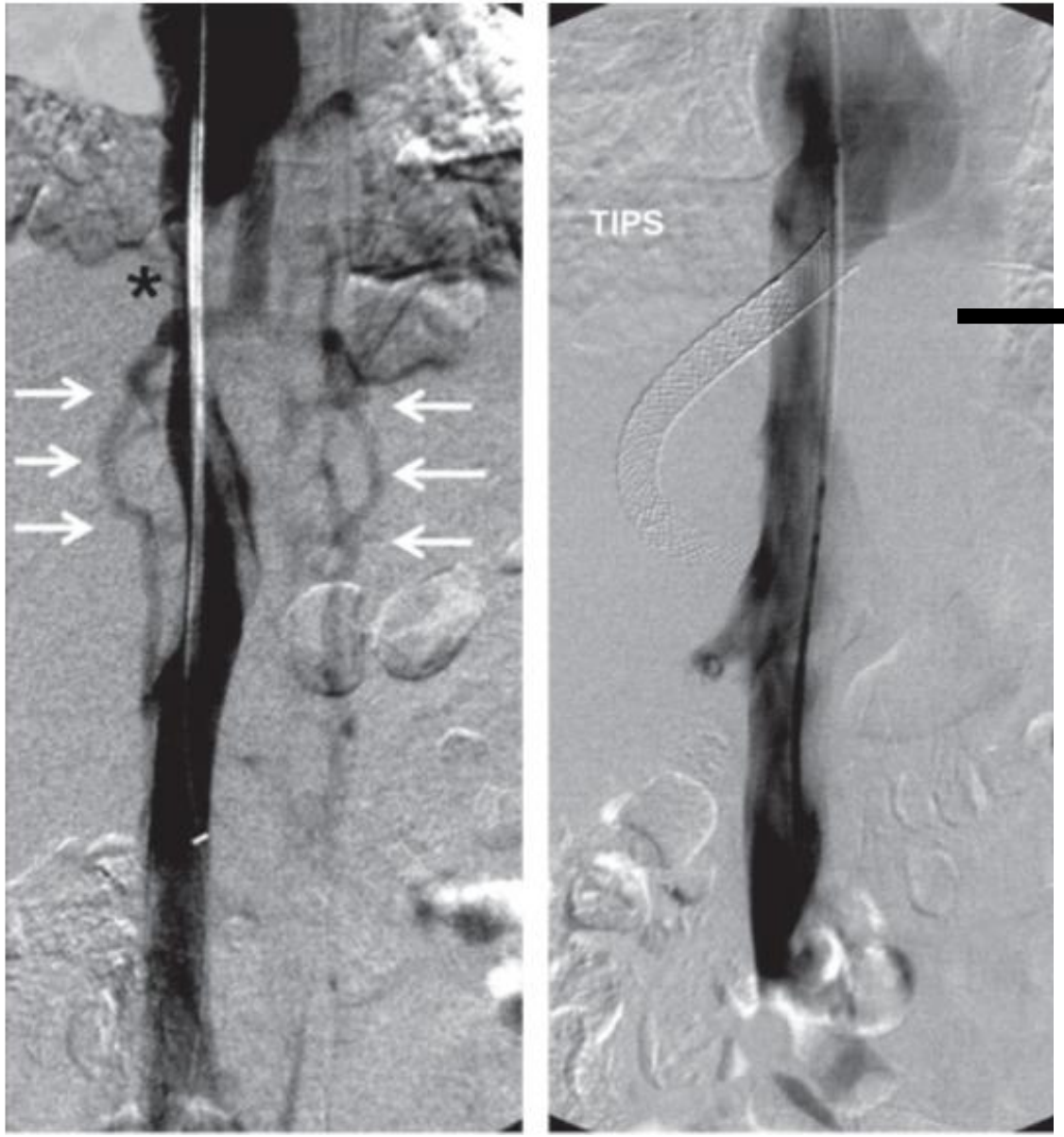
Table 2. Management of the Budd–Chiari Syndrome (BCS).*

Treatment	Indication	Advantages	Disadvantages
Thrombolytic therapy	Acute thrombosis	Reverses hepatic necrosis No long-term sequelae	Risk of bleeding Limited success
Angioplasty with and without stenting	IVC webs IVC stenosis Focal hepatic-vein stenosis	Averts need for surgery	High rate of restenosis or shunt occlusion
TIPS	Possible bridge to transplantation in fulminant BCS Acute BCS Subacute BCS if portacaval pressure gradient <10 mm Hg or occluded IVC	Low mortality Useful even with compression of IVC by caudate lobe	High rate of shunt stenosis Extended stents may interfere with liver transplantation
Surgical shunt	Subacute BCS Portacaval pressure gradient >10 mm Hg	Definitive procedure for many patients Low rate of shunt dysfunction with portacaval shunt	Risk of procedure-related death Limited applicability
Liver transplantation	Fulminant BCS Presence of cirrhosis in BCS Failure of portosystemic shunt	Reverses liver disease May reverse underlying thrombophilia	Risk of procedure-related death Need for long-term immunosuppression

* Information is from Ganguli et al.[26] IVC denotes inferior vena cava, and TIPS transjugular intrahepatic portosystemic shunt.

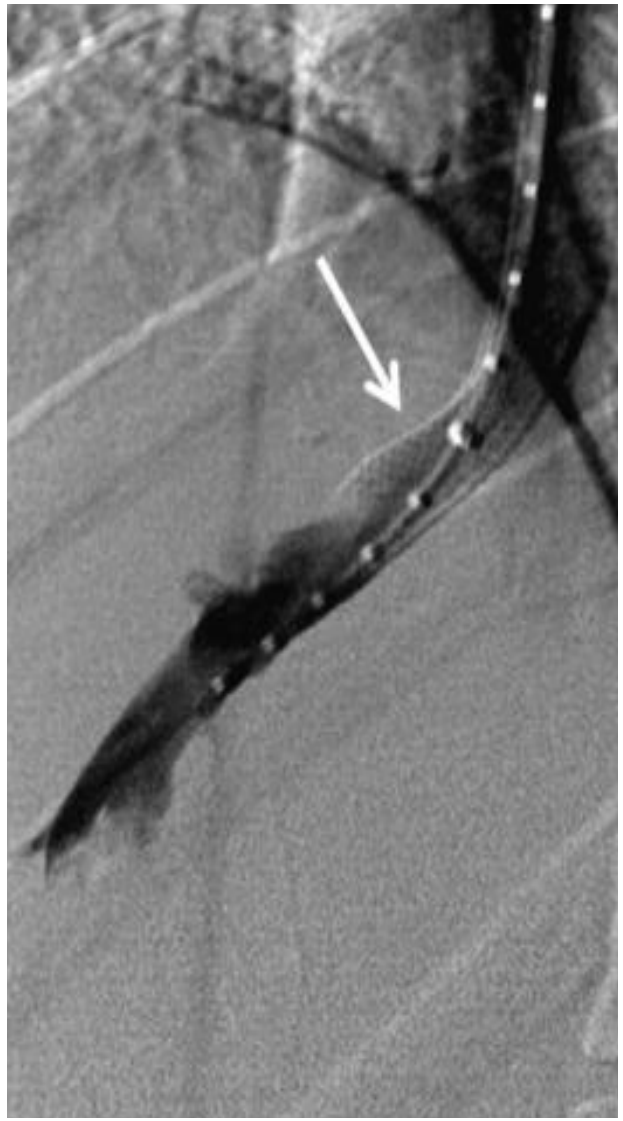
Cavogram before & after TIPS

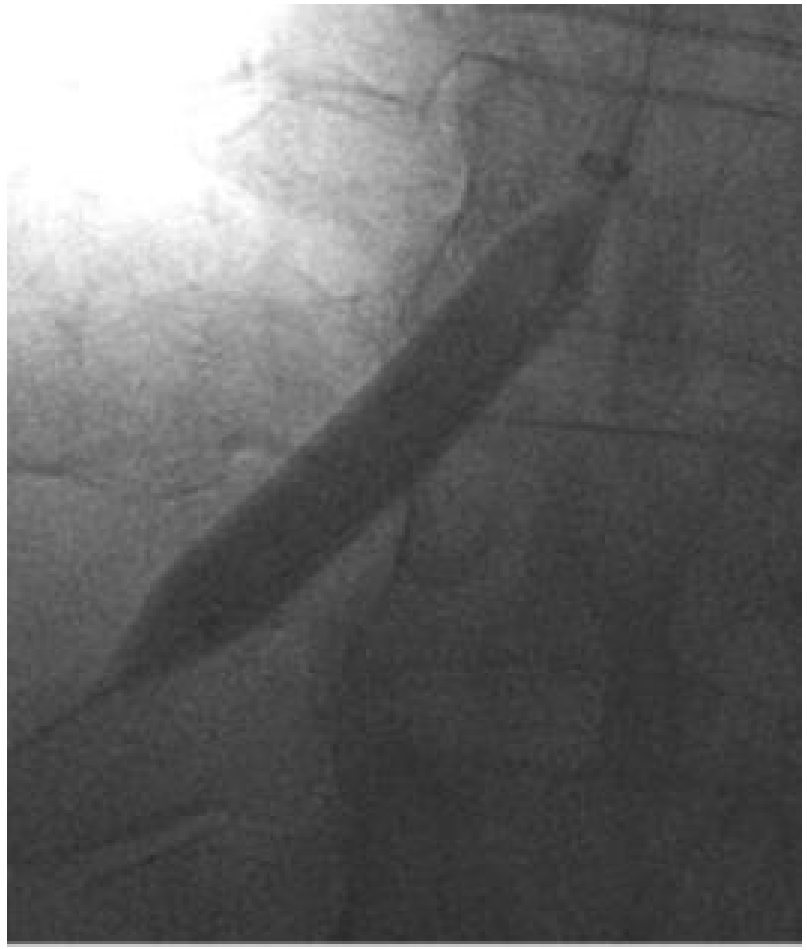
- intrahepatic IVC stenosis (*).
- perilumbar and azygos collateral veins (arrows)

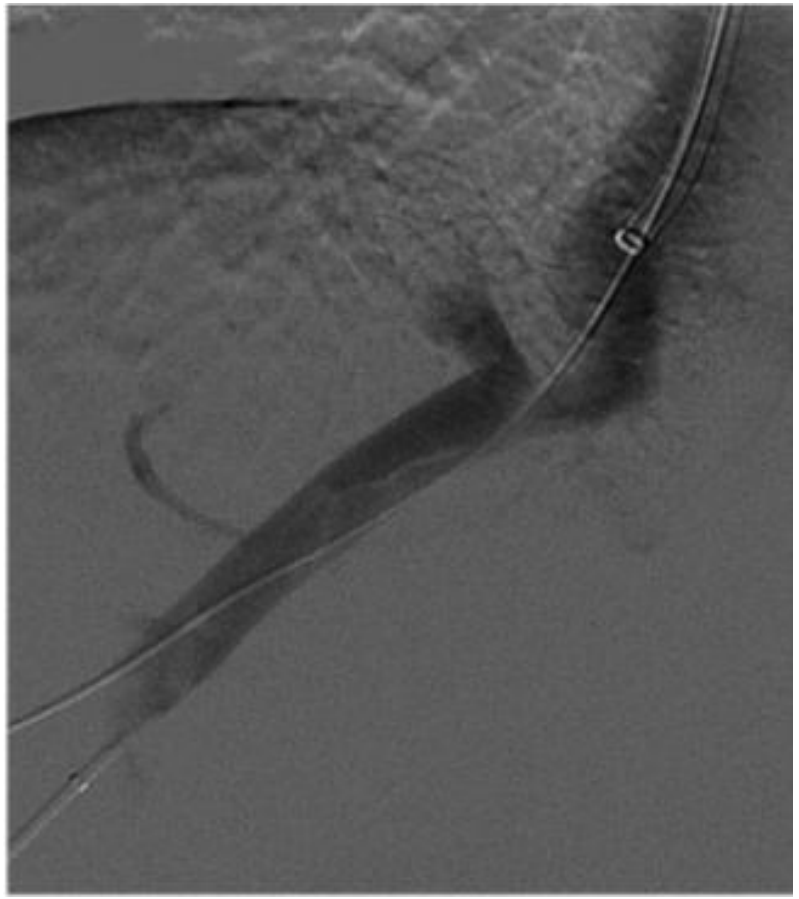


Cavogram obtained 2 months after TIPS creation shows resolution of the intrahepatic IVC stenosis and decompression of venous outflow

Hepatic venogram
Showing angioplasty
stent in situ







Comparison of hepatic venogram obtained before and after angioplasty shows a posttreatment increase in the luminal diameter of the vein

Complications

- Shock
- Portal hypertension
- Cirrhosis
- Hepatic failure
- Encephalopathy
- Hepatopulmonary syndrome

THANK YOU