UNIPHARMA

Congenital extrahepatic porto-systemic shunt: a case report

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Unipharma Third Gastro Symposium

September 14 – 16, 2023 – Lattakia – Syrian Arab Republic

Case report

- 15-year-old female referred for abdominal pain of 5 month
- Medical history:
  Splenectomy for an unknown reason at 4 months of age
  Repair of atrial septal defect at 8 months of age
- No clinical evidence of jaundice or icterus
- Suspected mass in LUQ

Complete blood count

- WBC: 6 100/uL with normal differential
- RBC: 4.3 million/uL
- Hg: 11 g/dL
- Ht: 34%
- MCV: 79 fL
- Platelets: 247 000/mm^3
- Ferritin: 11 ng/mL

Liver function tests

- TB 0.58 mg/dL
- DB 0.2 mg/dL
- AST: 24 units/L
- ALT: 18 units/L
- ALP: 562 units/L (normal: < 279)
- Albumin: 4.33 g/dL
- PTT 41 sec
- INR 1.28
- Ammonia: 87 mcg/dL (normal: 19 – 82 for women)

UGI endoscopy

- **UGI endoscopy:**

  Absence of esophageal or fundic varices

  Severe antral congestion

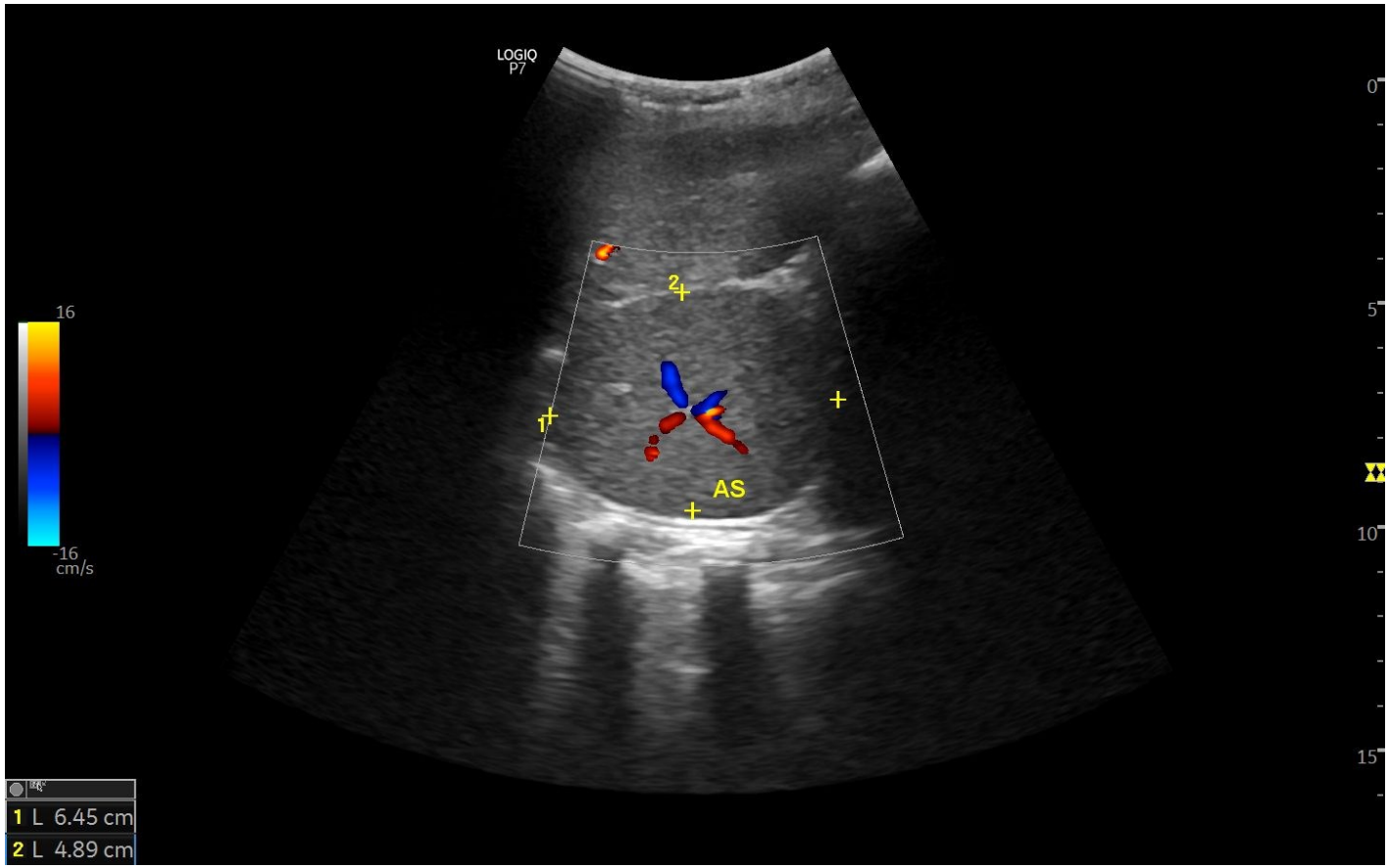
- **Biopsies:**

  Stomach: Chronic gastritis, Helicobacter pylori positive

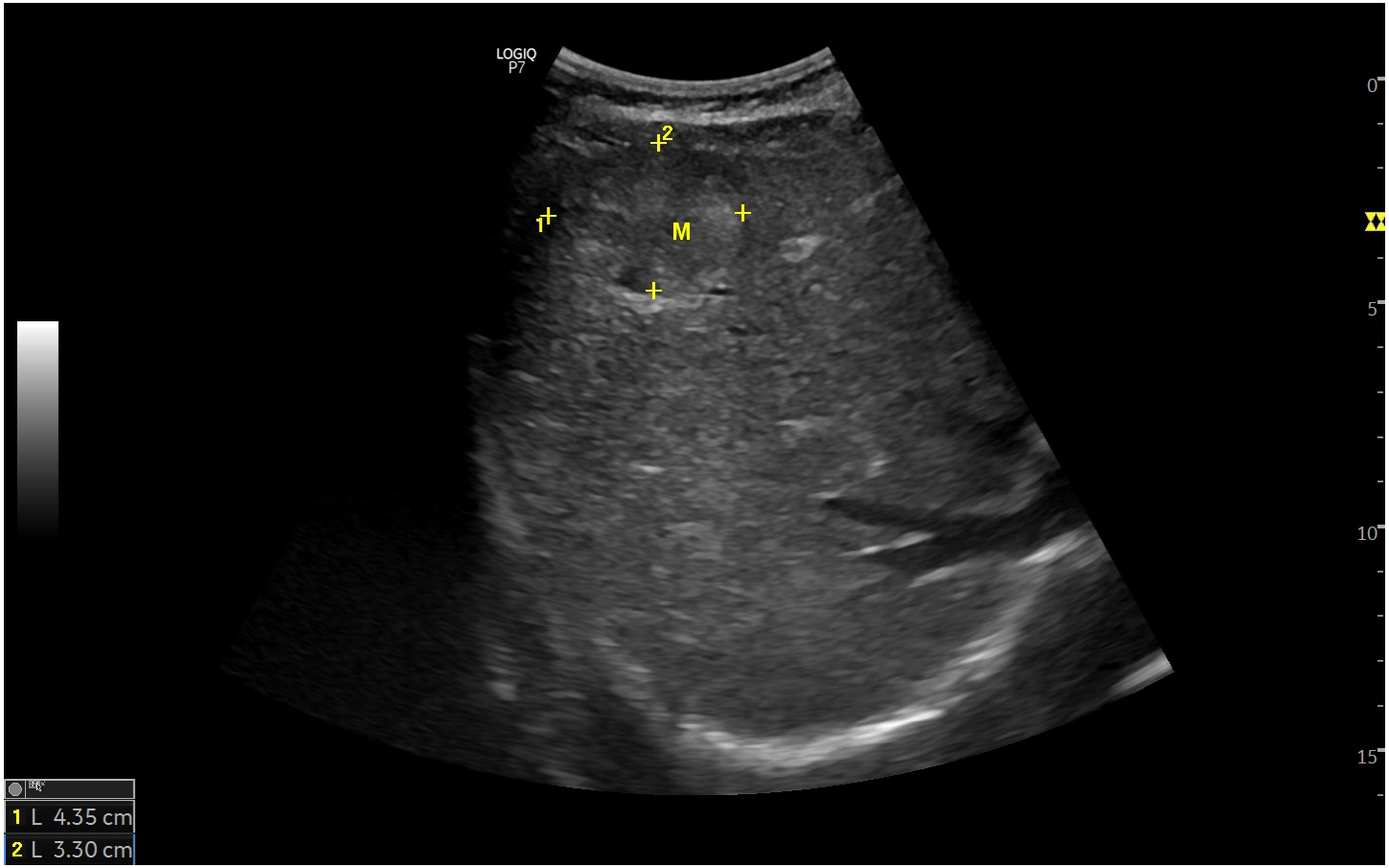
  Duodenum: Nonspecific inflammatory changes

  No signs of celiac disease

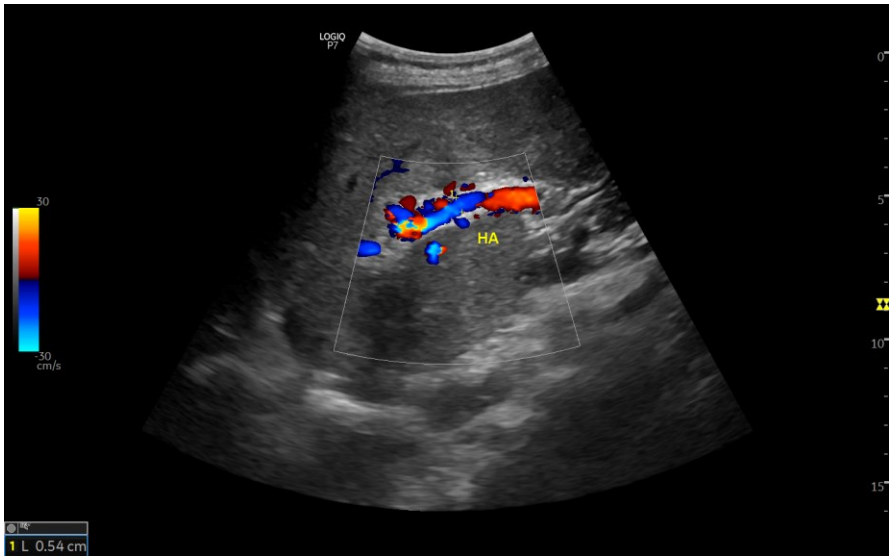
Accessory spleen in RUQ



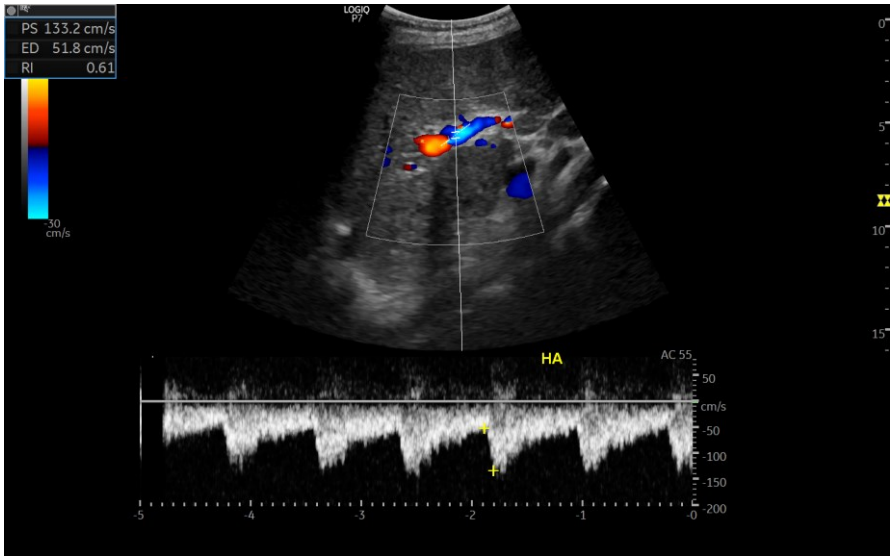
Nodular hepatic lesion on US



Doppler ultrasound of porta hepatis

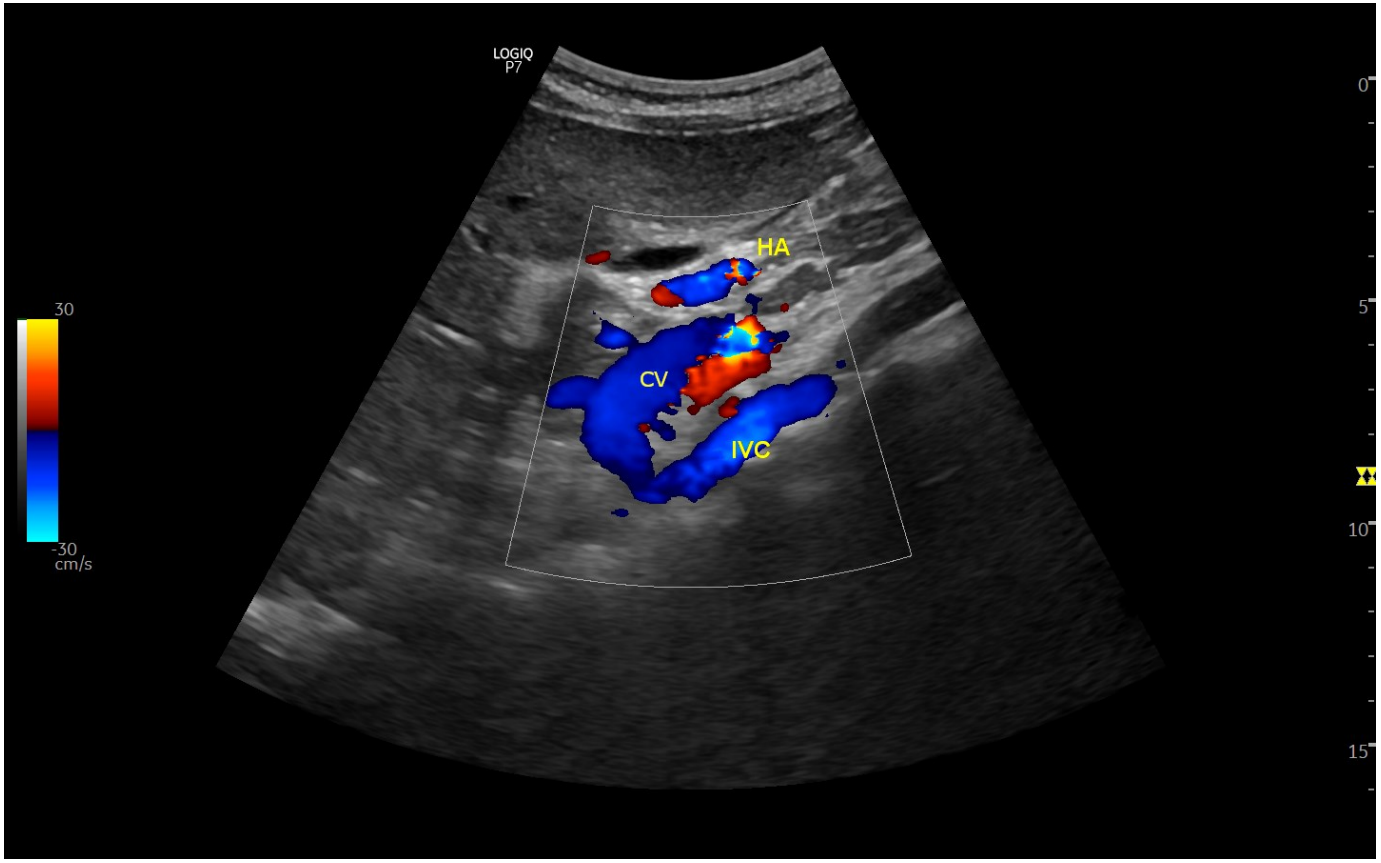


Absence of portal vein
Tiny venules in porta hepatis
Dilated hepatic artery (5.4 mm)



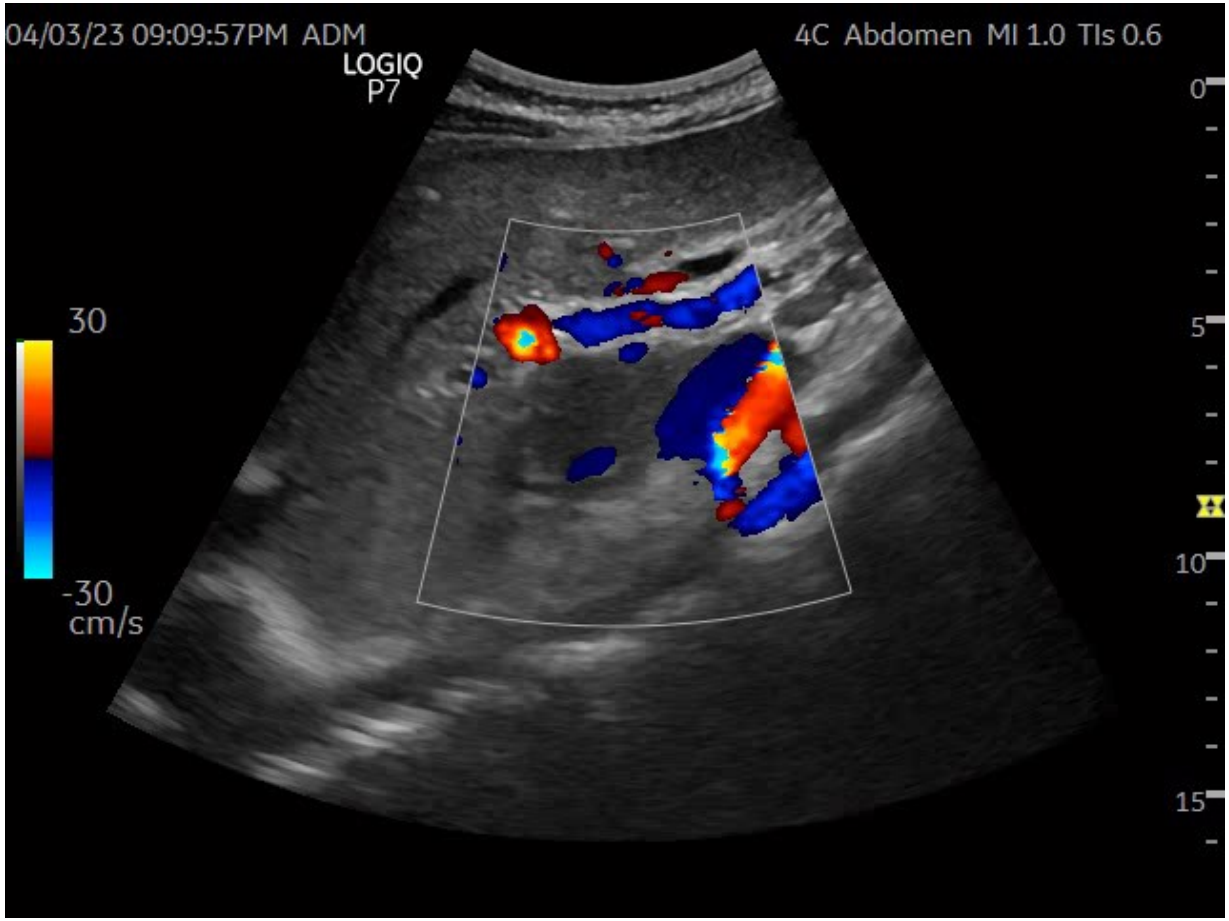
Pulsed Doppler of hepatic artery
Normal resistive index: 0.61

Abdominal color Doppler ultrasound



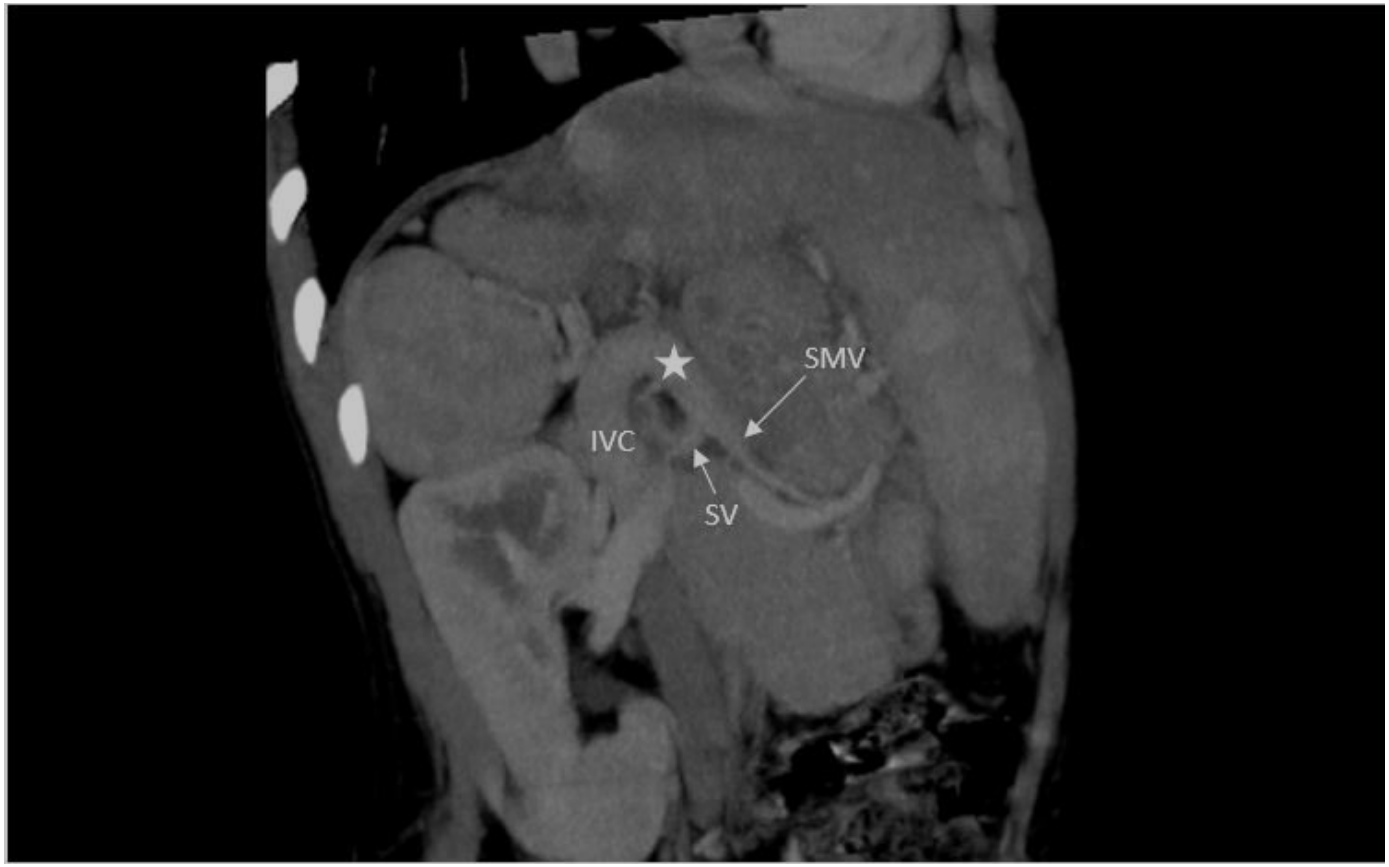
Venous trunk drained into IVC

Venous trunk drained into IVC



Contrast-Enhanced Computed Tomography

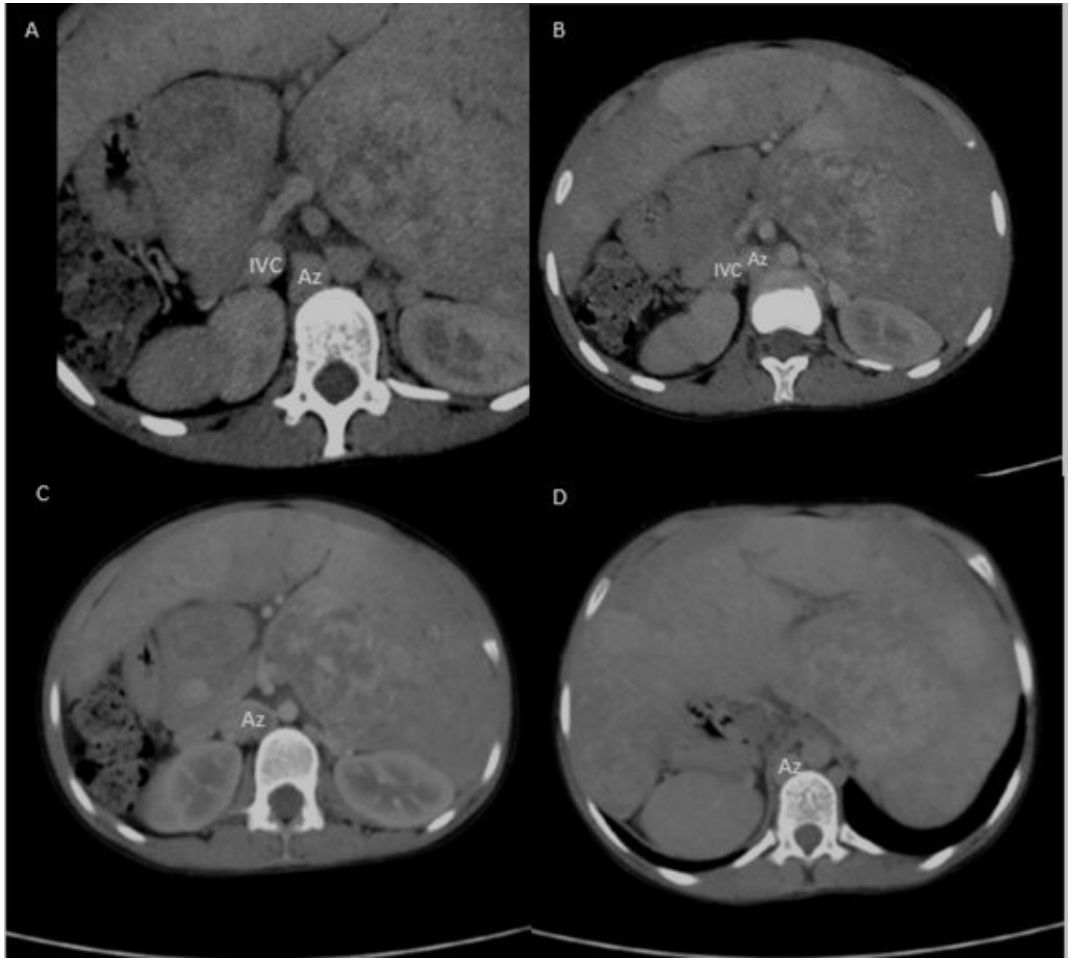
Oblique view



Splenic vein & superior mesenteric vein form common trunk (star) which drains into IVC

Contrast-Enhanced Computed Tomography

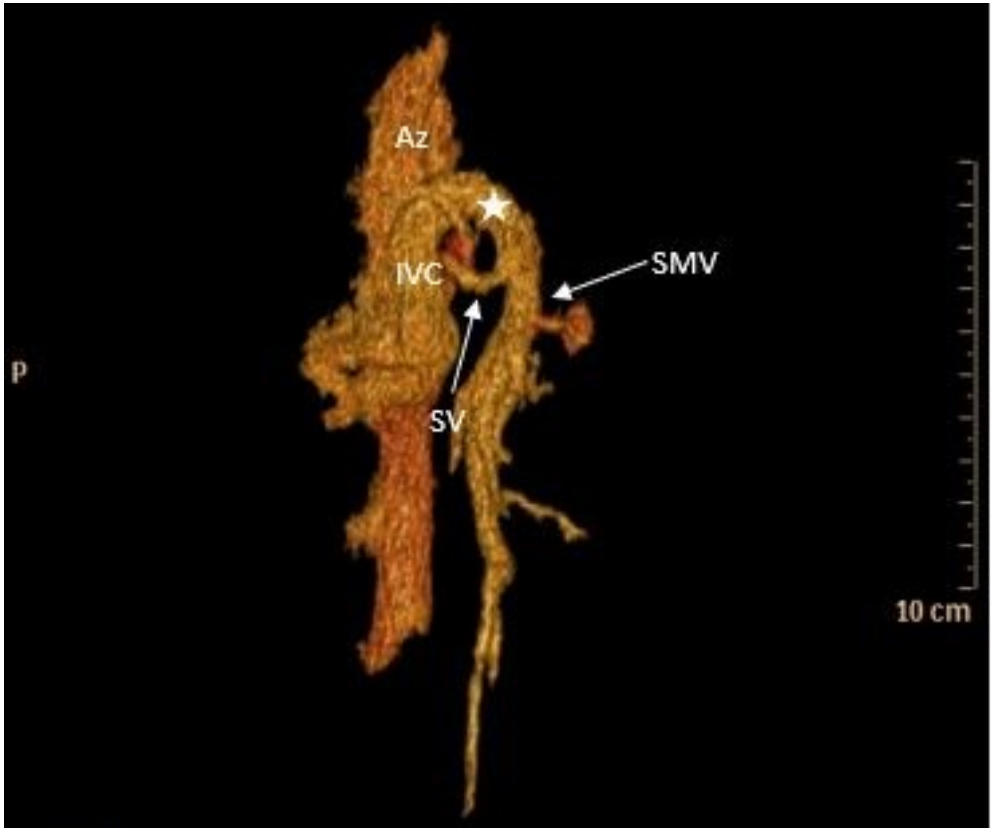
Axial view



Inferior vena cava draining into azygos vein which continues behind right crus of diaphragm

Contrast-Enhanced Computed Tomography

Reconstruction of abdominal venous system



Abernethy malformation type Ib

Drainage of IVC into azygos vein

Cardiac Ultrasound

- Normal mobility & volume of left ventricle
- Ejection fraction: 65%
- Moderate Tricuspid valve stenosis
  Pressure of 19 mm/Hg and median of 14 mm/Hg
- Closed atrial septal defect (ASD)
- Normal pulmonary valve
- Normal aortic arch

Abernethy malformation

Congenital extahepatic portosystemic shunt

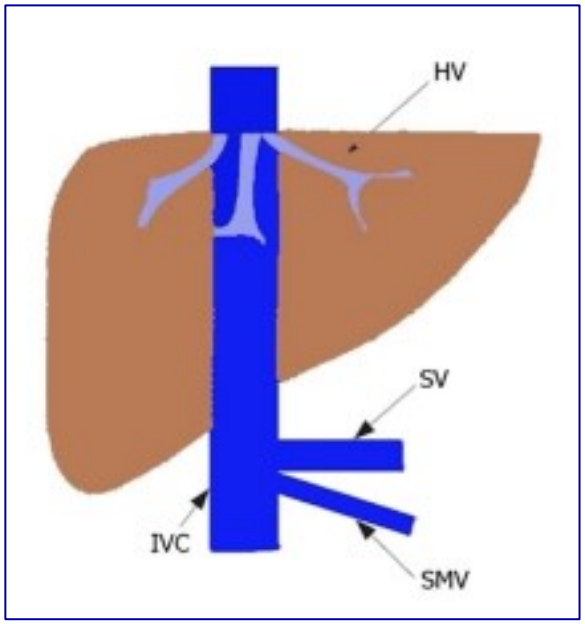
- First reported by John Abernethy in 1793
- Rare: less than 300 cases reported in literature
- Associated with several congenital anomalies
  Associated with multiple nodular liver lesions
- Spectrum of clinical variants: completely asymptomatic to severe forms of HE, hepatopulmonary syndrome & pulmonary arterial hypertension

Baiges A et al. Hepatology 2019;0:1–12 .

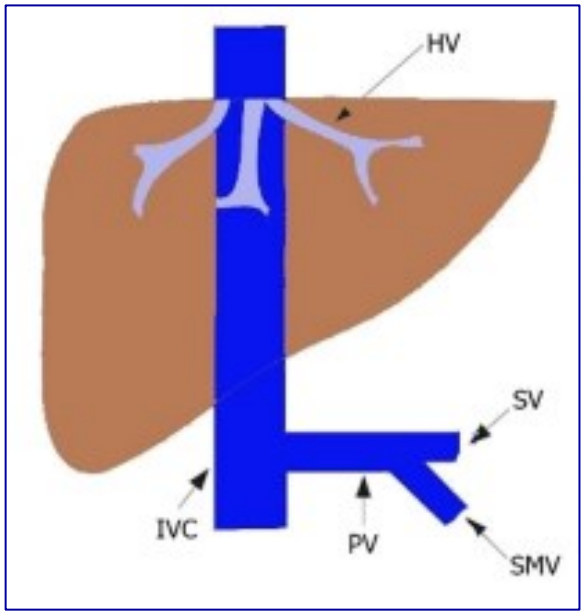
Classification of Abernethy malformation

Morgan & Superina classification

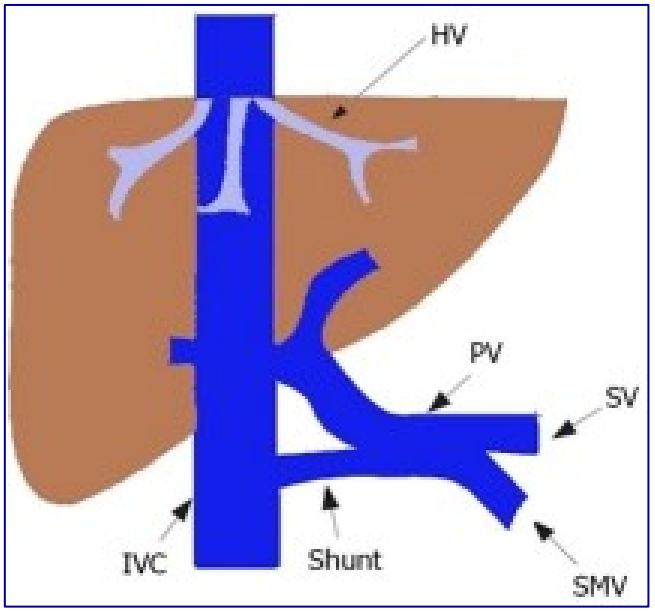
Type Ia



Type Ib



Type II



Morgan G. J Pediatr Surg 1994;29:1239 – 1241.

Concomitant cardiac abnormalities

Most common

- Atrial septal defect (ASD)
- Ventricular septal defect
- Patent foramen ovale
- Dextrocardia
- Patent ductus arteriosus
- Tetralogy of Fallot

Kumar P et al. Diagn Interv Radiol 2022; 28:21–2.

Concomitant hepatobiliary abnormalities

Hepatic abnormalities	Nodular liver lesions
• Biliary atresia • Choledochal cyst • Caroli's disease • Congenital hepatic fibrosis • Intrahepatic gallbladder	• Focal nodular hyperplasia • Hepatic adenoma • Nodular regenerative hyperplasia • Hemangioma • Hepatoblastoma • Hepatocellular carcinoma

Kumar P et al. Diagn Interv Radiol 2022; 28:21–2.

Concomitant gastrointestinal anomalies

- Esophageal atresia
- Tracheoesophageal fistula
- Intestinal malrotation
- Anal atresia

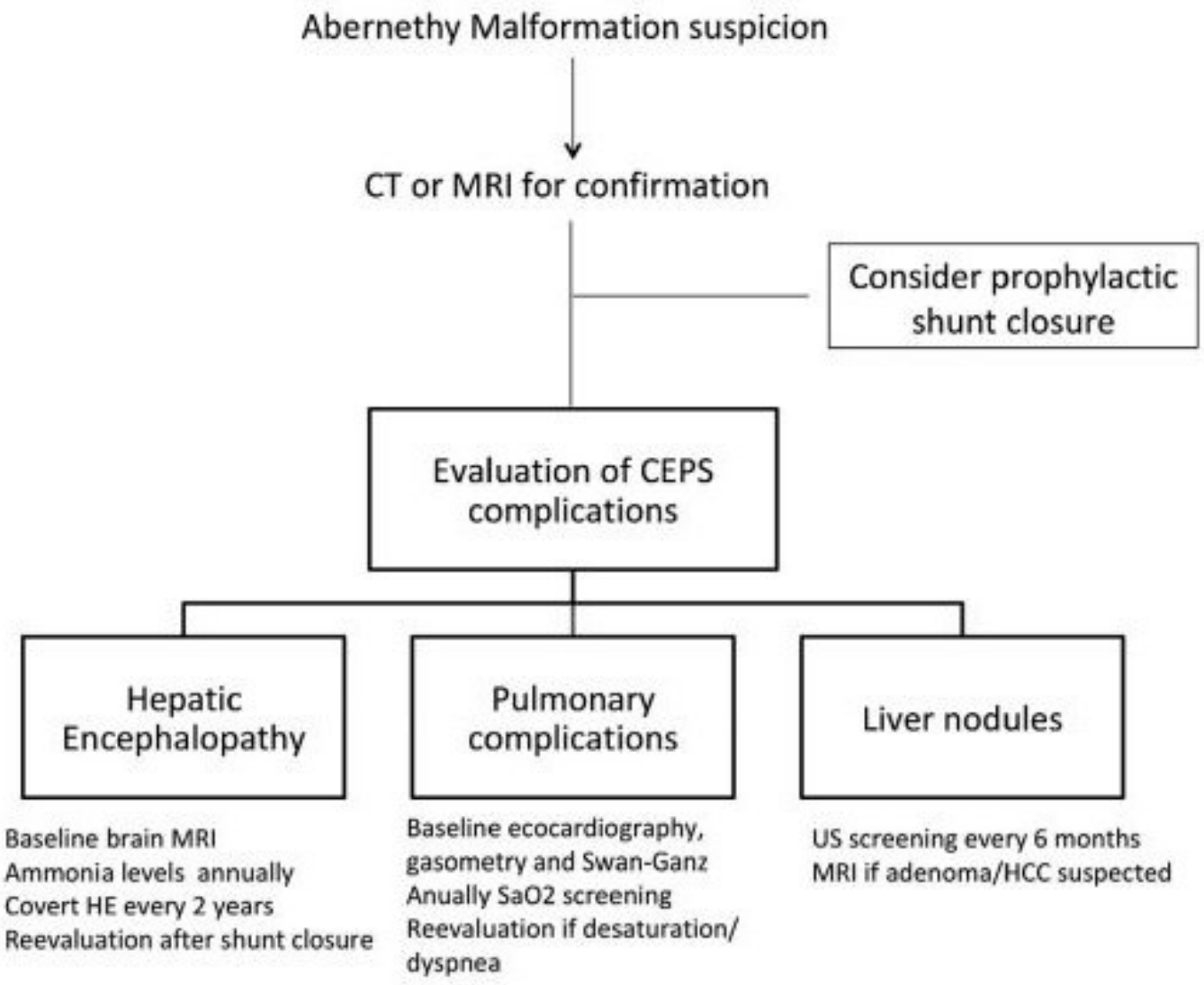
Kumar P et al. Diagn Interv Radiol 2022; 28:21–2.

Concomitant vascular abnormalities

- Agenesis or double IVC
- Azygos/hemiazygos continuation of inferior vena cava
- Double aortic arch
- Pulmonary artery stenosis

Kumar P et al. Diagn Interv Radiol 2022; 28:21–2.

Management algorithm



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Conclusion

- Abernathy malformations could be suspected by abdominal color Doppler ultrasound
- CEUS or MRI is recommended to confirm the diagnosis and to detect other congenital malformations

Thank You