Case-Based Diagnosis Training

**Patient:**

**Gender:** Male

**Age:** 2 years 6 months

**Clinical history and working diagnosis on the referral:**

- The patient presented 10 months back with occasional nausea & vomiting. He was clinically unremarkable.
- Labs showed elevated liver enzymes (SGOT-991U/L; SGPT-651; ALP-225). **USG at this point was reported normal.**
- Repeat labs done 2 months later showed persistent elevated liver enzymes & elevated total bilirubin (7.22). Further evaluation was performed, revealing elevated tyrosine, methionine, alanine (blood & urine) & lactic acid possibly suggestive of hepatic dysfunction. There was no evidence of infectious viral markers/haemophilia/hemolysis/Pompe/Neimann Pick/Cystic fibrosis/Wilsons/Celiac disease.
- Repeat USG with CDU and CE-MRI were done revealing

**Normal variant:**

**Embryological malformation:** Abernethy Type 1- B ¹

1: End to end shunt (Congenital absence of the portal vein with complete diversion of portal blood into systemic veins)

B: The confluence of superior mesenteric vein (SMV) and splenic vein (SV) form a common trunk that drains into left renal vein.

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Additional information

Developmental abnormality of the portal vein, which carries an increased risk of hepatic encephalopathy, HCC, FNH & hepatoblastoma

Underlying step in embryological development:

- Part of right & left vitelline vein outside liver undergoes transformation (from 4th week)
- The portal vein is formed by:
  - Left vitelline vein between entry of SMV & SV
  - Dorsal anastomosis
  - Right vitelline vein between dorsal vein anastomosis & central vein anastomosis

Potential differential diagnostic entities:

- Portal vein thrombosis (Pic:4- PVT- Portal vein thrombosis; PUV- Paraumbilical vein; AWC- Abdominal wall collaterals; LGV- left gastric vein; GRC- Gastrorenal collaterals; SV- Splenic vein; RV- Renal vein; SMV- Superior mesenteric vein; IMV-Infierior mesenteric vein; EC-Esophageal collaterals; PEC-Paraesophageal collaterals; RGC- Retrogastric collaterals)


In case you want to submit further pictures, please add these (radiograph, ultrasound, CT or MR images) and schematic drawing of the developmental process if applicable by clicking on the symbols within the boxes below:
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Additional pictures

In case you want to submit further pictures, please add these (radiograph, ultrasound, CT or MR images) and schematic drawing of the developmental process if applicable by clicking on the symbols within the boxes below:

Learning point: Always check portal vein during routine pediatric USG. Abernathy malformation although rare is not uncommon, and can be misdiagnosed as normal!

**Picture 5:** USG: SMV-Splenic vein confluence (arrow)

**Picture 6:** Confluence (asterisk), absent portal vein at porta hepatis

**Picture 7:** Anomalous vein draining the confluence (arrowhead)

**Picture 8:** CDU: HA flow at porta hepatis

**Picture 9:** HA spectral pattern at porta hepatis; Absent portal vein

**Picture 10:** Patent hepatic veins (RHV, MHV, LHV) with normal spectra.
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Additional pictures

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**Learning point:** Careful delineation of confluence, portal vein anatomy and CDU/Spectra is essential to suspect Abernathy malformation & differentiate it from PVT, Portal vein variant anatomy etc.

**Picture 11:** MR-T2W(Ax): A-EHPV draining confluence (arrow)

**Picture 12:** MR-T2W(Ax): SMV-SV confluence (arrow)

**Picture 13:** T1+C- Portal phase(Ax): A-EHPV(arrowhead) with absent intrahepatic portal vein

**Picture 14:** T1+C- Portal phase-FS (Ax): SMV-SV confluence; A-EHPV seen left lateral to aorta (arrow)

**Picture 15:** MRV-TOF(Cor): showing anomalous-EHPV

**Picture 16:** MRV-TOF (Obl): A-EHPV with absent portal vein