

Case-Based Diagnosis Training

Patient:

Gender: *Male*

Age: *2 years 6 months*

Submitted by:

A. Nair¹, V. Mouka¹, A. Giantsouli¹, V. Xydis¹, M. Rogalidou², E. Varveris², M.I. Argyropoulou¹

Departments of ¹Clinical Radiology; ²Pediatrics. Medical School, University of Ioannina, Greece.

Clinical history and working diagnosis on the referral:

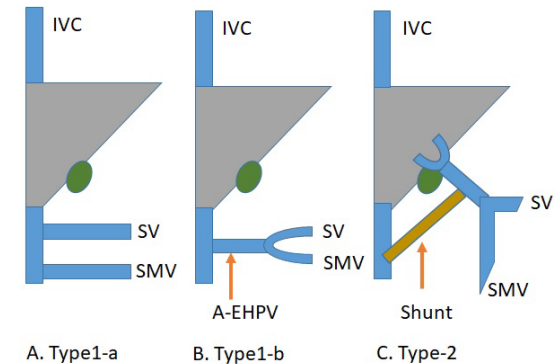
- The patient presented 10 months back with occasional nausea & vomiting. He was clinically unremarkable.
- Labs showed elevated liver enzymes (SGOT-991U/L; SGPT-651; ALP-225). **USG at this point was reported normal.**
- Repeat labs done 2 months later showed persistent elevated liver enzymes & elevated 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.



Picture 1: SV-Splenic vein; SMV- Superior mesenteric vein; A-EHPV- Anomalous extra-hepatic portal vein

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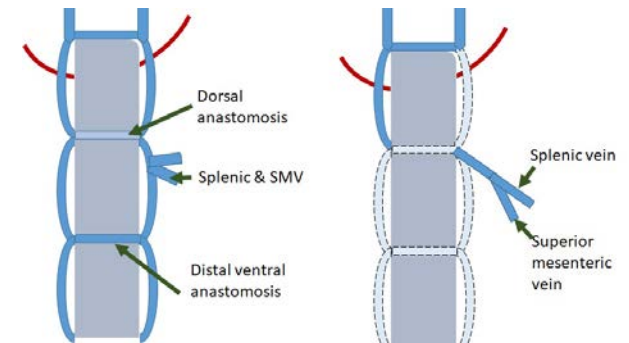
Additional information

Developmental abnormality of the portal vein, which carries an increased risk of hepatic encephalopathy, HCC, FNH & hepatoblastoma¹

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:

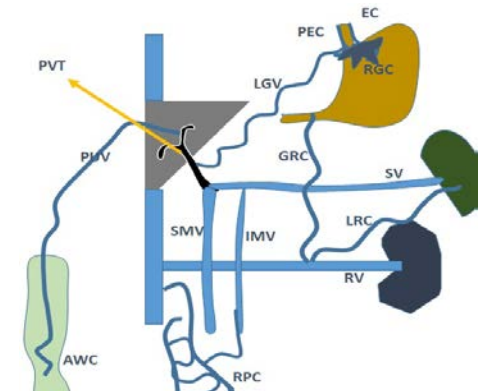
Underlying step in embryological development:

- 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



Embryological development of Portal vein

Picture 3:



Picture 4: PVT

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- Inferior mesenteric vein; EC- Esophageal collaterals; PEC- Paraesophageal collaterals; RGC- Retrogastric collaterals)

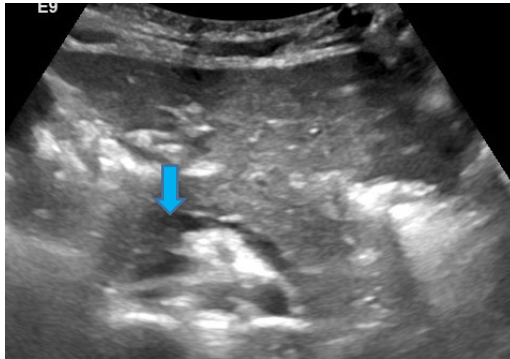
1. Tetsu Niwa, Noriko Aida, Katsuhiko Tachibana, et al. Congenital absence of the portal vein: clinical and radiologic findings. *Journal of computer assisted tomography*, 2002, 26.5: 681-686

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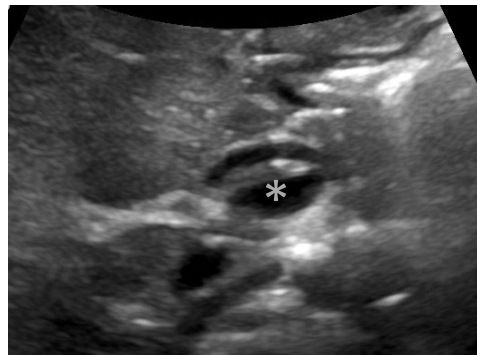
Learning point : Always check portal vein during routine pediatric USG. Abernathy malformation although rare is not uncommon, and can be misdiagnosed as normal!

Additional pictures

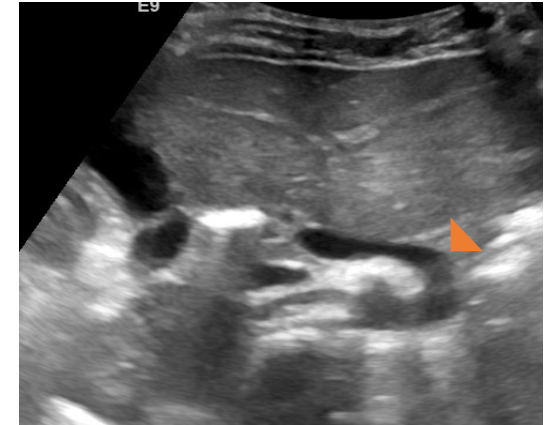
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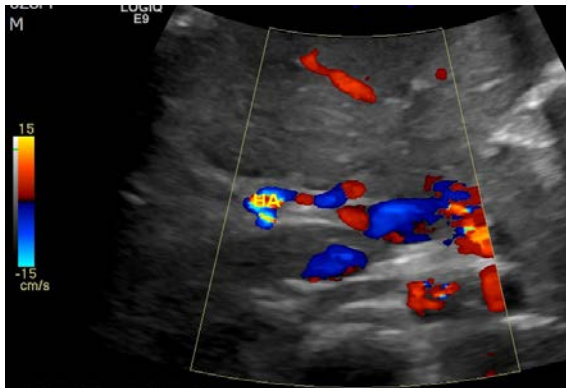
Picture 5: USG: SMV-Splenic vein confluence (arrow)



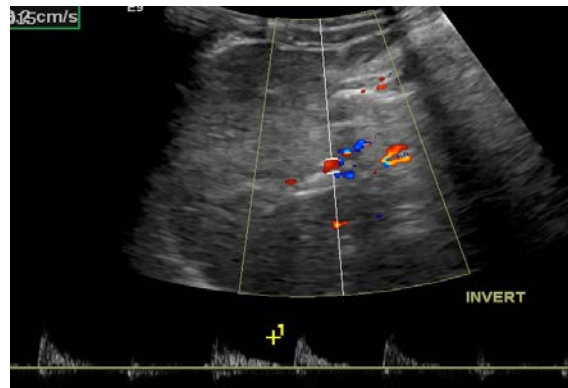
Picture 6: Confluence(asterisk), absent portal vein at porta hepatis



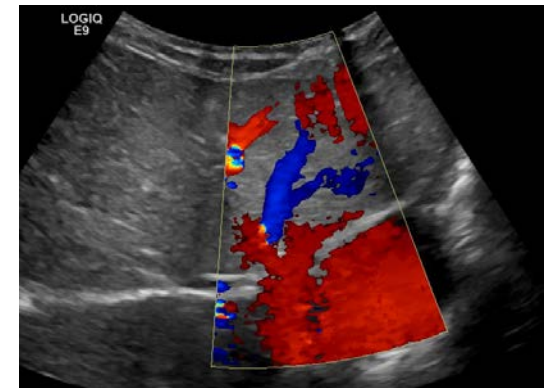
Picture 7: Anomalous vein draining the confluence (arrowhead)



Picture 8: CDU: HA flow at porta hepatis



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



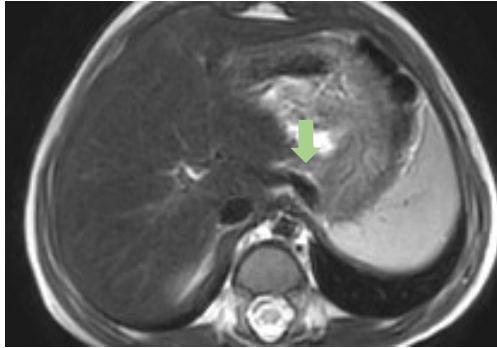
Picture 10: Patent hepatic veins (RHV, MHV, LHV) with normal spectra.

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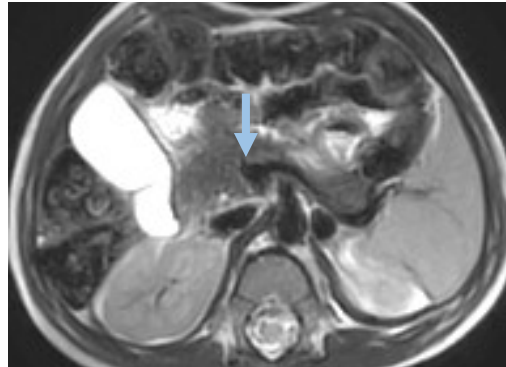
Learning point: Careful delineation of confluence, portal vein anatomy and CDU/Spectra is essential to suspect Abernathy malformation & differentiate it from PVT, Portal vein variant anatomy etc.

Additional pictures

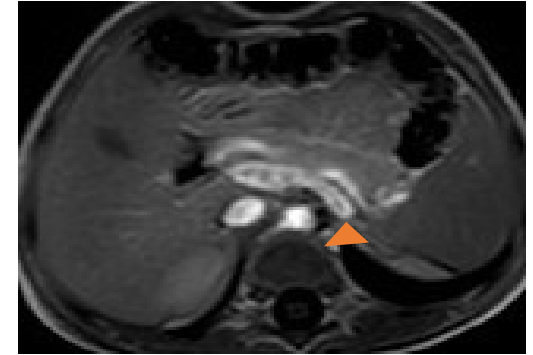
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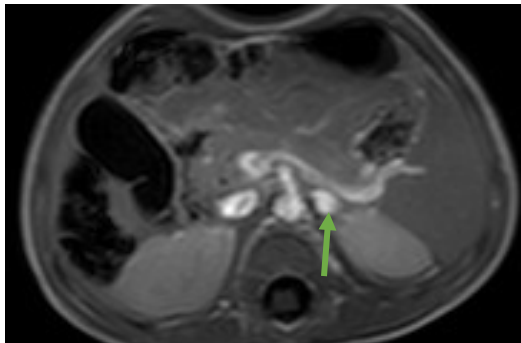
Picture 11: MR-T2W(Ax): A-EHPV draining confluence (arrow)



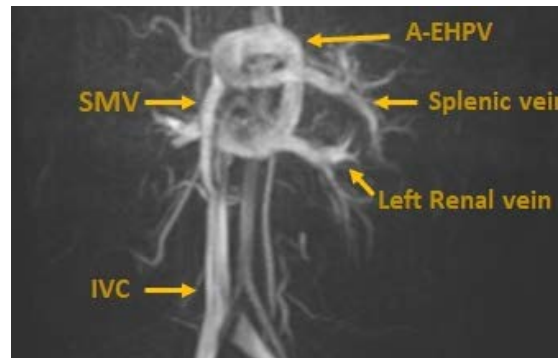
Picture 12: MR-T2W(Ax): SMV-SV confluence (arrow)



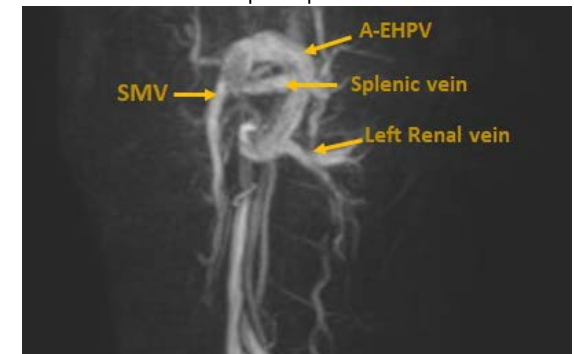
Picture 13: T1+C- Portal phase(Ax): A-EHPV (arrowhead) with absent intrahepatic portal vein



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 (ObI): A-EHPV with absent portal vein