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Summary

ABERNETHY MALFORMATION – HOW TO TREAT THIS RARE TYPE OF PORTOSYSTEMIC SHUNT?

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A case report describes successful endovascular treatment of type 2 Abernethy malformation in a 58-year-old woman with recurrent hyperammonemic encephalopathy.

KEY POINTS

- Abernethy malformation is a rare congenital extrahepatic portosystemic shunt classified into type 1 (complete shunting, absent portal vein) and type 2 (partial shunting, hypoplastic portal vein).
- The patient presented with recurrent psychomotor slowing, disorientation, and elevated ammonia; CT scan revealed a type 2 shunt between left portal branch and inferior vena cava.
- After partial response to lactulose and rifaximin, endovascular single-step shunt occlusion was performed; hemodynamic studies showed hepatic venous pressure gradient of 14 mmHg, rising to 24 mmHg on balloon occlusion testing.
- At 1-year follow-up the patient remained asymptomatic with confirmed shunt closure, maintained inferior vena cava patency, portal reperfusion, and no endoscopic signs of portal hypertension.
- The authors note that no standardised management guidelines exist and position endovascular occlusion as a minimally invasive option for type 2 Abernethy malformation.

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