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Summary

PRESENTING COMPLAINTS IN 250 PATIENTS WITH CONGENITAL PORTOSYSTEMIC SHUNTS

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Registry data from 256 patients with congenital portosystemic shunts show most are asymptomatic at diagnosis, with intrahepatic shunts diagnosed earlier and more likely to close spontaneously than extrahepatic shunts.

KEY POINTS

- Among 212 patients with known shunt type, 41.5% had intrahepatic shunts and 58.5% had extrahepatic shunts.
- Most patients were asymptomatic at diagnosis: 72% of those with intrahepatic shunts and 68% of those with extrahepatic shunts.
- Symptomatic intrahepatic shunt patients were diagnosed at a median age of 0.59 years, while symptomatic extrahepatic shunt patients were diagnosed later at a median age of 7.49 years.
- Spontaneous closure occurred in 37.5% of closed intrahepatic shunts (all before age 1), but in only 2 extrahepatic shunt patients.
- Closure methods varied by shunt type, with interventional radiology used in 52% of intrahepatic shunts and surgery in 54% of extrahepatic shunts, plus 7 liver transplants.
- This descriptive analysis from the ESPGHAN-sponsored International Registry of Congenital Portosystemic Shunts may help clinicians recognize this rare disorder.

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