

When should I refer a patient with PSVD to a center of expertise?

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UEG Postgraduate Teaching Programme Berlin 2025

Presentation

Hepatobiliary

October 5, 2025

Porto Sinusoidal Vascular Disorder (PSVD) is a rare vascular liver disease accounting for 5% of portal hypertension cases in Western countries that requires referral to expert centres for diagnosis and management due to its complexity and need for multidisciplinary evaluation.

KEY POINTS

- The speaker stated that PSVD diagnosis requires liver biopsy reviewed by an expert pathologist to exclude cirrhosis and identify characteristic histological signs, plus imaging interpretation by radiologists familiar with PSVD features such as low liver stiffness (typically around 10 kilopascals or less) despite portal hypertension and smooth liver surfaces.
- Up to 50% of PSVD patients have associated conditions including prothrombotic disorders, haematological diseases, toxin exposure, or genetic diseases that impact prognosis and require management by specialists in haematology, rheumatology, immunology, and other fields.
- The speaker reported that variceal bleeding is the first presentation in up to 40% of cases, one third develop ascites, and one third develop portal vein thrombosis within five years of diagnosis.
- Referral indications include difficult or uncertain diagnosis, need for specialized diagnostics, severe or refractory complications of portal hypertension requiring advanced interventions like TIPS, and consideration of liver transplantation.
- The speaker noted that outcomes after TIPS and liver transplantation are very good overall, but patients with uncontrolled associated conditions have worse long-term outcomes than those without comorbidities or with well-managed conditions.

- The speaker stated in discussion that there is currently no treatment for PSVD itself, only management of portal hypertension complications following standard guidelines and treatment of underlying associated conditions.

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