

LOW PHOSPHOLIPID-ASSOCIATED CHOLELITHIASIS SYNDROME: A NOT SO RARE CAUSE OF BILIARY DISEASE

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Poster

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LPAC syndrome is a genetic biliary disorder causing intrahepatic lithiasis that accounts for 1-5% of biliary pathology admissions and can be diagnosed by ultrasound findings and treated medically with ursodeoxycholic acid.

KEY POINTS

- LPAC syndrome results from ABCB4 gene mutation causing decreased biliary phospholipid secretion and MDR3 transporter dysfunction, leading to intrahepatic lithiasis
- Diagnosis requires two of three criteria: symptom onset before age 40, recurrence after cholecystectomy, or compatible ultrasound appearance showing intrahepatic hyperechogenic spots, comet tail images, or intrahepatic lithiasis with acoustic shadowing
- The ABCB4 gene mutation is not a diagnostic criterion as it appears in only 50% of patients
- A 23-year-old male with acute pancreatitis was diagnosed with LPAC based on ultrasound findings of multiple intrahepatic hyperechogenic spots and comet tail signs, treated with ursodeoxycholic acid, and remained symptom-free during follow-up
- Medical treatment with ursodeoxycholic acid is effective and cholecystectomy can be avoided

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