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Summary

Itch in a hepatology outpatient clinic: A case-based approach

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A 26-year-old woman with PFIC type 1 previously controlled by biliary diversion developed severe cholestasis during pregnancy and achieved sustained biochemical and clinical remission with an IBAT inhibitor after delivery.

KEY POINTS

- The patient had surgical biliary diversion at age 4 for PFIC with complete response maintained into adulthood, and the diversion was left in place based on prognostic data linking serum bile acids under 102 micromole per liter with better native liver survival.
- During pregnancy she developed severe pruritus, serum bile acids rising to 195 micromole per liter, weight loss of 6 kg, and diarrhea up to 16 times daily; rifampicin was ineffective.
- Genetic testing revealed compound heterozygous mutations in the ABCB11 gene coding for the bile salt export pump, and the speaker changed the diagnosis to PFIC type 1.
- Treatment with an IBAT inhibitor during late pregnancy was followed by spontaneous delivery at 29 weeks with a healthy child.
- After stopping the IBAT inhibitor postpartum, cholestasis recurred with aminotransferases rising to 8 times the upper limit of normal and elastography worsening to 70.6 kilopascals, prompting reinitiation at 40 micrograms per kilogram body weight.
- Two months after restarting therapy the speaker reported resolution of pruritus, normalization of aminotransferases and elastography, and only slight elevation of serum bile acids, with the patient now considering a second pregnancy.

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