

Guidelines for the management of acute porphyria: recommendations from the International Porphyria Network.

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Authors: Israeli Porphyria Center, Rabin Medical Hospital, Petach Tikva, Israel and Samson Assuta Ashdod Medical Center, Ashdod, Israel; Faculty of Health and Science, Ben-Gurion University in the Negev, Beer Sheva, Israel.

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Autosomal dominant acute porphyrias are rare inherited disorders of haem biosynthesis characterised by accumulation of potentially neurotoxic porphyrin precursors and attacks of severe abdominal pain with autonomic and neuropsychiatric features. Disease severity ranges from asymptomatic individuals to those with recurrent, life-threatening attacks. The International Porphyria Network invited 34 acute porphyria specialists from 17 countries to form an expert panel. The invited group included clinicians from diverse specialities (ie, internal medicine, haematology, endocrinology, gastroenterology, hepatology, neurology, and biochemistry), together with laboratory scientists and patient representatives. The panel met online (in 2023-25) to develop 15 evidence-based recommendations with the use of the Grading of Recommendations, Assessment, Development, and Evaluations framework addressing attack prevention, management of sporadic and recurrent attacks, long-term follow-up, surveillance for primary liver cancer, and family screening. The guidelines support safe, consistent clinical care and improved outcomes, recognising global variation in resources and access to high-cost drugs, and highlighting priorities for future research.