The hepatic porphyrias: experience with 105 cases Las porfirias hepáticas: experiencia con 105 casos.

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Hepatic porphyria is a rare metabolic syndrome caused by abnormal enzyme activity in heme biosynthesis. Between 1974 and 1991; 105 patients have met criteria for diagnosis of hepatic porphyria based on typical clinical findings and/or laboratory abnormalities. According to type, 42% had porphyria cutanea tarda, 21% porphyria variegate, 15% protoporphyria, 6.7% acute intermittent porphyria, 6.7% coproporphyria and 1.9% porphyria due to porphobilinogen deficit. A proper classification was not established in 6.7% of patients. Porphyria cutanea tarda was more common in males (70%) and porphyria variegata, in females (90%). A family history of the disease was present in 33% of patients; 20% of patients were of European descent and 4% of Mapuche descent. Diagnosis was usually established in the third decade, somewhat later in porphyria cutanea tarda (45 years of age) and very early in protoporphyria. 10% of patients were asymptomatic and 29 patients developed at least one porphyric crisis. T