the skin lesions are identical to those seen in hereditary coproporphyria and porphyria cutanea tarda (see below), VP must be differentiated from these two conditions. The presence of neuropathic lesions without cutaneous manifestations necessitates differentiation from AIP, the symptoms of which may be identical to those of VP. When both cutaneous and neuropathic lesions occur in an individual or a kindred, hereditary coproporphyria or one of the dual porphyrias (see below) should be considered in the differential diagnosis.

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Biochemical differentiation rests on the fact that VP is caused by an enzyme block distal to hepatic protoporphyrinogen synthesis. Therefore, excess protoporphyrin (levels greater than those of coproporphyrin) is present in the stool. In contrast, AIP and hereditary coproporphyria are caused by diminished enzymatic function proximal to protoporphyrinogen and are therefore associated with normal or near-normal levels of stool protoporphyrin. Because stool porphyrins that result in fluorescence can arise from heme-derived protoporphyrin, patients should avoid eating red meat before and during fecal collections; in addition, the guaiac (Hemoccult) test should be performed to prevent analyzing samples that contain occult blood.

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The precipitating factors, principles of prophylaxis, and treatment of VP are the same as those for AIP. The absence of a readily accessible assay for the defective enzyme in VP, protoporphyrinogen oxidase, makes it necessary to obtain urine and stool samples from family members and to test these samples for porphyrins to determine the diagnosis and permit genetic counseling. Negative tests should not be considered definitive unless the family member has reached puberty. The treatment of skin lesions is discussed elsewhere .

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Hereditary Coproporphyria

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Even less common among the hepatic porphyrias is HCP. It is inherited as an autosomal dominant trait and can be traced to insufficient activity of coproporphyrinogen oxidase, leading to overproduction of coproporphyrin and its precursors. Like AIP and VP, HCP is exacerbated both chemically and clinically by induction of ALA synthetase.

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When symptoms occur, increased levels of ALA and PBG are present in the urine, producing a positive Watson-Schwartz test. Levels of urinary porphyrin are elevated, and coproporphyrin levels in both the urine and stool are elevated. There also may be small increases in stool protoporphyrin levels (although coproporphyrin levels are higher), but these levels are considerably lower than those