in patients with VP. Modestly increased urine coproporphyrin alone is a nonspecific finding and is seen in hepatic, hematologic, and toxic conditions.

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The symptomatic spectrum of HCP is indistinguishable from that of VP; it also overlaps with the pain and neuropathy seen in AIP and with the skin lesions seen in porphyria cutanea tarda. The decrease in coproporphyrinogen oxidase activity can be detected in lymphocytes by specialized laboratories. Values are usually 50 percent of normal in patients older than one year.

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Treatment and prophylaxis for HCP are the same as those for VP and AIP. The response to drugs is also the same [see Tables 2a and 2b].

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ALA Dehydratase Deficiency (Plumboporphyria)

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The least common form of hepatic porphyria is caused by a partial deficiency of hepatic ALA dehydratase. Because ALA dehydratase, which converts ALA to PBG, is also inhibited by lead (in Latin, plumbum), this disorder is also called plumboporphyria. Only a handful of cases have been reported. (ref 22, 23) It is inherited as an autosomal recessive trait and is marked by increased urinary excretion of ALA but not PBG or porphyrins. Therefore, despite typical neurovisceral symptoms (neurologic manifestations and abdominal pain), the usual screening tests (the Watson-Schwartz and Hoesch tests) are negative and a quantitative assay of 24-hour urinary levels of ALA is required. Lead poisoning should be excluded in the differential diagnosis. Treatment of ALA dehydratase deficiency is the same as that of AIP (see above).

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Dual Porphyrias

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When more than one enzyme in the biosynthetic sequence is affected within a single kindred, a person in the kindred may have a dual porphyria, which is characterized by a mixed profile of porphyrin excretion. In the case of so-called Chester porphyria, (ref 9, 10) members of one family in Chester, England, were shown to have defects both in PBG deaminase levels (mapped to chromosome 11) and in protoporphyrinogen oxidase levels (mapped to chromosome 14) that manifested as coincidental AIP and VP, respectively. Although no photosensitivity occurred, neurovisceral symptoms were present. The pattern of porphyrin excretion varied from individual to individual: some had a pattern that resembled AIP, whereas others had a pattern that resembled VP. This difference presumably reflected deficiencies in whichever enzyme was more rate-limiting.

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There have been reports of other families with dual