precursors. Although erythrocyte protoporphyrin is also increased in iron deficiency anemia and lead intoxication, the protoporphyrin is more avidly bound to hemoglobin in these two conditions and does not diffuse into plasma and tissues, which may be why photosensitivity does not occur.

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In contrast to the hepatic porphyrias, protoporphyria usually occurs in children younger than four years; males are predominantly affected. Symptoms are acutely precipitated by sunlight and may be limited to burning and itching, often accompanied by edema, erythema, or urticaria. Less frequently, blisters and ulcers occur. Excoriations secondary to scratching may be present. Recurrence of these lesions as a result of chronic sun exposure creates scarring, altered pigmentation, lichenification, and premature aging of the skin.

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Increased amounts of protoporphyrin are also deposited in the liver. Although mild liver function abnormalities are the rule, cirrhosis, hepatosplenomegaly, and death from liver failure have also been reported. Gallstones containing protoporphyrin are seen in a minority of patients, and a mild hypochromic, microcytic anemia is found in almost half. Some hemolysis may also be present. A more severe form of protoporphyria occurs if both parents are heterozygous and the offspring inherit two defective genes .

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Diagnosis is suggested by the combination of history and familial occurrence and is confirmed by increased levels of fecal and erythrocyte protoporphyrin. Erythrocytes or normoblasts fluoresce on exposure to the Soret band.

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Avoidance of sunlight to prevent irreversible scarring is desirable but difficult. Ingestion of á-carotene, with the aim of reaching blood levels of 500 æg/dl, diminishes photosensitivity. The use of pyridoxine in dosages of up to 1 g/day has also been advocated, but this drug does not alter porphyrin metabolism. (ref 31) The effects of treatment may not be evident for months. Because hepatic protoporphyrin synthesis may also contribute to the clinical picture, a high-carbohydrate diet may be of value, even though the acute neuropathic attacks characteristic of AIP do not occur in this disorder.

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In one patient, oral iron therapy reduced protoporphyrin levels in stool and erythrocytes. (ref 32) Resolution of anemia and improved results on liver function tests occurred within two to four months. It was postulated that this approach increased the intracellular level of iron and that heme was produced when the iron combined with protoporphyrin either nonenzymatically or by the catalytic action of the residual ferrochelatase. Heme formation in turn could have