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positive Watson-Schwartz tests. (ref 2) Avoidance of unnecessary surgery and noxious medications [see Tables 2a and 2b] diminishes the likelihood of an acute attack. If attacks do occur, sophisticated methods of intensive care for patients with respiratory failure and pneumonia increase the chances of survival. Finally, prompt use of hematin therapy may abort acute attacks before paralysis occurs and intubation becomes necessary. When full support and continuous respirator assistance are required, the prognosis for survival is probably less than 50 percent.

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Variegate Porphyria

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Variegate porphyria, also known as South African porphyria because it is relatively common in South African whites, is rarer than AIP in the United States. It is attributed to a diminution of protoporphyrinogen oxidase and therefore is associated with a persistent overproduction of protoporphyrinogen, coproporphyrinogen, and uroporphyrinogen (in descending quantitative order) and an increased excretion of their corresponding porphyrins. Protoporphyrin, which is the least hydrophilic, is excreted by the liver and must be sought in the feces. Coproporphyrin is found in both feces and urine. During acute attacks, ALA and PBG are excreted in abnormal quantities in the urine, and the Watson-Schwartz test is positive. During remissions or latent periods, ALA and PBG excretion may be normal. Increased excretion of these early intermediates reflects induction of ALA synthetase. Because the deficient enzyme, protoporphyrinogen oxidase, is situated in the mitochondria, mature red blood cells cannot be used for enzymatic assay. Normoblasts, leukocytes, and skeletal muscle and other tissues have been studied to define the biochemical abnormality.

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The presence of increased levels of circulating porphyrins predisposes patients to skin lesions. Typically, lesions appear in sun-exposed areas. Blisters as well as superficial ulcers in various stages of healing and scarring are present. Mechanical fragility is common, especially at sites of bony protuberances such as knuckles and ankles. Mild hypertrichosis of the face and temples is seen. Photosensitivity lesions can develop in patients with VP while they are indoors because the Soret band (with a wavelength of 400 to 410 nm) can penetrate ordinary window glass. In contrast, ordinary ultraviolet light (with a wavelength of about 300 nm) is blocked by this barrier. Transparent sunscreens with maximum protection against ordinary sunburn do not protect against photosensitivity lesions induced by the Soret band. Skin lesions and neuropathic lesions may occur separately or together. Members of the same kindred may have either or both. Because