subgroup of patients, the red blood cell assay yields false positive results. (ref 13)

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Although the defect in PBG deaminase activity in patients with AIP is unremarkable during normal circumstances, induction of ALA synthetase by one of the four "M's" leads to the accumulation of the precursors ALA and PBG, which are water soluble and are excreted in increased amounts in the urine. ALA is colorless, but PBG condenses into a brownish-red polymer (porphobilin) on standing and contributes to the characteristic dark-red color of the urine during acute attacks of the disease. Excess porphyrinogen production, and hence porphyrin production, is slight or absent. It is the excess levels of circulating porphyrins that are associated with cutaneous manifestations in some forms of porphyria. Because these excess levels are not found in AIP, photosensitivity does not occur.

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The prominent clinical and diagnostic features of AIP are varied [see Table 3]. Manifestations of neurotoxicity at the cortical level include mood changes, irritability, and frank psychosis. Hypothalamic lesions may cause the syndrome of inappropriate antidiuretic hormone secretion (SIADH) with hyponatremia, which in itself may cause seizures and psychiatric disturbances. Cranial nerve dysfunction may cause severe bulbar paresis with difficulty in swallowing and aspiration leading to pneumonia. Peripheral neuropathy causes pain and paresis. If intercostal and phrenic nerves are involved, ventilatory insufficiency may occur. Involvement of autonomic nerves may produce abdominal pain, ileus, tachycardia, hypertension, or, less commonly, diarrhea. Because signs and symptoms are of neuropathic origin, fever, leukocytosis, and other signs of inflammation are usually mild or absent.

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AIP should be included in the differential diagnosis of unexplained abdominal pain, acute psychiatric disturbances, and acute polyneuropathies. Of assistance in the diagnosis are a positive family history, the presence of pain in the back and extremities as well as in the abdomen, and the acute development of hypertension during the course of the attack. Suspicion should be greater if symptoms occur during menstruation or pregnancy, after exposure to certain drugs (including alcohol or anesthetics) [see Tables 2a and 2b], or during a weight reduction regimen.

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A urine sample can be rapidly screened for PBG by the Watson-Schwartz or the Hoesch test, both of which utilize Ehrlich's reagent as a chromogen. The Hoesch test may produce slightly more false positive results but is clinically as sensitive as the Watson-Schwartz test. In either case, 24-hour urine samples should be collected,