placed in refrigerated opaque containers, and delivered to a qualified laboratory for quantitative analysis of ALA, PBG, and porphyrins.

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Because the other inducible hepatic porphyrias (ALA dehydratase deficiency, HCP, and VP) may produce identical neuropathic syndromes without photosensitive lesions and may be marked by excess excretion of PBG, ALA, or both, definitive diagnosis should be sought by stool examination and the erythrocyte PBG deaminase assay. The PBG deaminase assay is 95 percent sensitive when combined with an assay that shows increased activity of leukocyte ALA synthetase (ref 14) and is accurate during an acute attack as well as during the latent period, when urinary biochemical abnormalities may be absent. It is also useful for screening asymptomatic family members, who may have the latent condition, but it may be falsely elevated (i.e., normal) if the r

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eticulocyte count is high. (ref 3) Early identification of the etiology of porphyric symptoms will prevent unnecessary tests and permit earlier treatment. Screening of family members should be recommended even though screening may cause anxiety and difficulty in obtaining health insurance or employment. It is most important to avoid drugs known to precipitate attacks [see Tables 2a and 2b]. A high-carbohydrate diet, with reduction of fat intake to maintain isocaloric intake, is also desirable, as is genetic counseling.

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Therapy consists of the use of medications to treat the symptoms of the disease, such as meperidine and phenothiazines for pain relief. Intravenous infusion of glucose at a rate approaching 20 g/hr (400 to 500 g/day) is recommended. If excessive hyperglycemia or glucosuria is present, small doses of insulin may be administered. Beta blockers have been advocated for the treatment of marked tachycardia and hypertension.

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If rapid symptomatic improvement does not occur within 24 hours or if motor neuropathy is present, heme therapy should be begun and the patient's vital capacity should be measured at frequent intervals. Because there is a great risk of death caused by rapidly progressing ventilatory insufficiency, skilled facilities for tracheal intubation and respiratory assistance should be immediately available. Proper management of fluid and electrolyte infusions may also be necessary to treat hyponatremia. Nasogastric intubation or parenteral nutrition may be required if bulbar symptoms prevent swallowing.

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The use of hematin, the ferric hydroxylated form of heme,