Acta Dermatovener (Stockholm) 54: 501–502, 1974

LETTER TO THE EDITOR

Etiology of Erythropoietic Protoporphyria, and Protection of Photosensitive Subjects

We note that Krook & Haeger-Aronsen (7) referred to erythropoietic protoporphyria (EPP) as erythrohepatic protoporphyria. Lest your readers assume that it is now appropriate to rename this disease, we refer them to our current Letter to the Editor (3) in which we discuss the nomenclature of EPP. After reviewing published reports of radiotracer investigations on the basis of presently available information, we concluded that no single study may be interpreted as verifying primary hepatic involvement. The only definitive report that we are aware of is the one by Schwartz et al. (9), who observed that the source of excess protoporphyrin in their patient was erythropoietic tissue. In our reply (4) to Scholnick's critique of our letter, we stressed that the use of protoporphyria to denote the disease is also premature, because the only subheading which can be employed with confidence is erythropoietic. Our position is that some EPP patients may have primary hepatic involvement; but until such evidence is presented, the present terminology of EPP should be retained. It should be noted that formation of gallstones or development of hepatic damage due to massive protoporphyrin deposits do not constitute evidence that the liver is the major source of excess prophyrin. As pointed out in our Letter (3), excess protoporphyrin synthesized in any tissue must be excreted via the hepatic system, where the porphyrin may accumulate and produce the secondary symptoms of gallstones and cirrhosis.

Although oral β-carotene appears to be an effective therapy for EPP, it is of limited or no value in improving sunlight tolerance in other types of photosensitivities. It is surprising that the therapy is not even helpful in porphyria cutanea tarda, a disease with a photoreactivity spectrum similar to that of EPP. Swanbeck & Wennersten (10) cited beneficial effects of β-carotene in patients with polymorphous light eruptions (PMLE) on the basis of clearing of skin lesions. However, their patients were instructed "not to expose themselves more than usual to the sun". Nordlund et al. (8) employed oral β-carotene and topical sulisobenzone to protect a PMLE patient, and reported that the subject developed erythema in spots where the topical sunscreen washed off after sweating. Kobza et al. (6) observed that the carotenoid did not provide protection to persons with actinic reticuloid or solar urticaria.

It may be of interest to your readers to know that, for over a decade, we have successfully treated all types of photosensitivity with topical dihydroxyacetone/lawsone (Duoshield, Rowell Laboratories, Inc., Baudette, Minnesota 56623, USA). Our cumulative clinical experience through 1971 was recently reported (2). With respect to EPP, the sunscreen afforded a median 12-fold increase in sunlight tolerance (1). In contrast to the ease with which other sunscreens wash off, the components of dihydroxyacetone/lawsone bind chemically to the stratum corneum, and therefore resist even soap and water washing (5).

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Received May 14, 1974

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