"Why is my skin so fragile?"

Catherine McCuaig, MD, FRCPC, DABD

A 50-year-old alcoholic presents with a one-year history of skin fragility of the hands (Figure 1), which is worse in the summer. He also presents with hypertrichosis of the forehead, as well as tiny blisters (Figure 2).

What does he have?

This patient has *porpyria cutanea tarda* (PCT).

What causes PCT?

PCT may be acquired or hereditary. The latter is due to an inherited deficiency of the enzyme uroporphyrinogen decarboxylase.

What are the clinical findings?

PCT blisters, crusts, milia, and scars cause hypertrichosis of sun-exposed skin. Hyperpigmentation, sclerodermoid changes, alopecia, dystrophic calcification, and photo-onycholysis may be present.

Precipitating factors include ethanol, estrogens, hydrocarbon hepatotoxins, increased iron stores, dialysis, hepatitis B or C, or human immunodeficiency virus (HIV).

Abnormal glucose tolerance, autoantibodies, and hepatocellular carcinoma may be found.

What are the features?

Histologic features of PCT include:

- Sub-epidermal blistering with minimal "cell-poor" dermal inflammatory infiltrate (H&E); and

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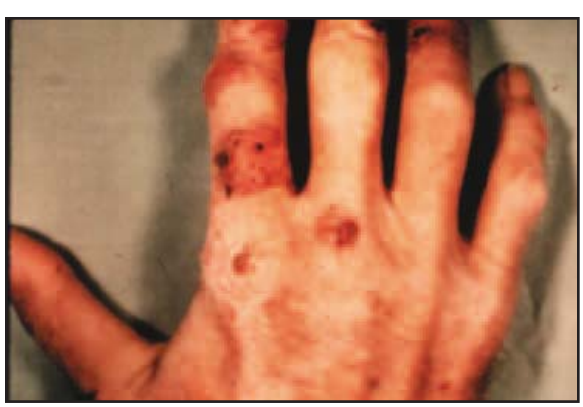

Figure 1. Skin fragility of the hands.

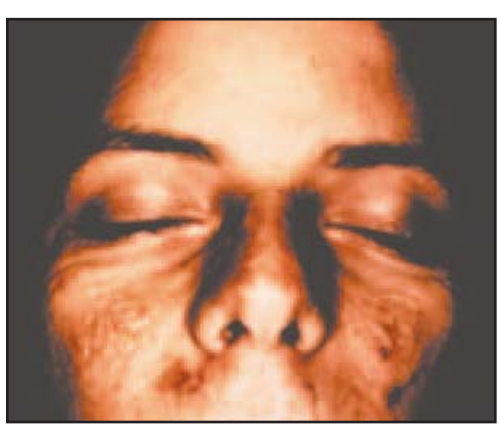

Figure 2. Hypertrichosis extending onto forehead and hyperpigmentation in sun-exposed areas.

- Festooning of the dermal papillae with periodic acid-Schiff (PAS) positive deposits within the walls of blood vessels (PAS stain).

What is the treatment?

Phlebotomies, which reduce excess hepatic iron stores; physical sunblocks; chloroquine; or hydroxychloroquine, 200 mg twice a week, and avoidance of the exogenous agents usually control the disease within six to 12 months. CME

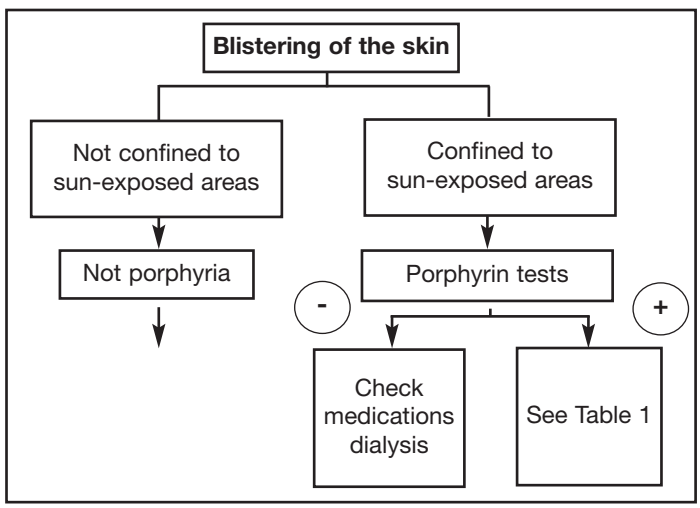

Figure 3. Approach to bullous porphyria.

Dr. McCuaig is an associate professor, Université de Montréal, Montreal, Quebec.

Table 1
Classification of the porphyrias

Acute porphyria	Non-acute porphyria
ALA-D defficient	PCT
Acute intermittent	Erythropoietic protorpophyria
Variegate	Congenital erythropoietic
Hereditary coproporphyria	

ALA-D: Aminolevulinic acid dehydrase
PCT: Porpyria cutanea tarda

Digital Camera Winner!!!

Dr. David Nelligan is a McGill graduate who has practised medicine on the West Island of Montreal since 1969. Until recently, Dr. Nellign had a private practice in Hudson, Quebec. Today, he continues to work at Stat-Care Medical Clinic in Pointe Claire, Quebec. On December 30, Dr. Nelligan became a proud grandfather, which will enable him to make good use of the digital camera he has won.

Dr. David Nelligan