

12

INCIDENCE AND DISTRIBUTION OF ACQUIRED PORPHYRIA IN AFRICA

By JAMES MARSHALL, M.D.

*Head of the Department of Dermatology, University of Stellenbosch,
South Africa*



Reprinted from *The Practitioner*
August 1963, Vol. **191**, pages 198-200

INCIDENCE AND DISTRIBUTION OF ACQUIRED PORPHYRIA IN AFRICA

By JAMES MARSHALL, M.D.

Head of the Department of Dermatology, University of Stellenbosch, South Africa

SOUTH Africa is noted for a high incidence of hepatic porphyria in its white, negro (Bantu) and coloured (mixed) populations (Dean and Barnes, 1955; Eales, 1960). The porphyria of the Whites is a familial disease, inherited in dominance, whilst that of the Negroes is an acquired disease. Coloured people may show either variety.

Cutaneous signs (porphyria cutanea tarda) are found in both groups. In the White cutaneous lesions may or may not be associated with episodes of acute porphyria, there is a variable degree of uroporphyrinuria, and the faecal porphyrin concentration is markedly raised. In the Negro the disease is purely cutaneous; uroporphyrinuria is gross and the faecal porphyrin concentration is normal or only slightly increased. Many of the cases of acquired porphyria reported in the Bantu have abnormal 'liver function tests' and high serum globulin concentrations, and 30 out of 100 patients studied by Lamont and his colleagues (1961) showed fibrotic lesions of the liver (28 biopsies, 2 necropsies). Acquired porphyria of the type seen in the Bantu occurs sporadically in all races throughout the world. It is commonly associated with liver damage and is often ascribed thereto. This ascription may be valid, but factors other than 'liver damage' alone must operate; otherwise porphyria would be even commoner than it is in the South African Bantu who, without developing this disease, often have clinical and/or biochemical signs of liver damage such as are found in porphyrics.

GEOGRAPHICAL DISTRIBUTION

A clue to the factors involved may come from studies of the geographical distribution of cases. It appears from the literature and from replies to a questionnaire on dermatoses in Africa which I have circulated that acquired porphyria is common in the Bantu of South Africa and Southern Rhodesia. Elsewhere in Africa south of the Sahara (Negro Africa) it appears to be rare or non-existent.

The porphyria belt seems to stop fairly sharply at the borders of Southern Rhodesia. A correspondent, Dr. J. E. P. Thomas from Kitwe, Northern Rhodesia, reports that 'porphyria is extremely common in Southern Rhodesia and most cases have liver disease. In Northern Rhodesia, in spite of a higher incidence of liver disease, I have found only one case in two years'. Dr. R. Ashworth from Lilongwe, Nyasaland, writes: 'I have been on the lookout for porphyria but have not seen a case in Nyasaland although I saw the condition in South Africa and Southern Rhodesia.'

North of the Sahara occasional cases have been reported from Morocco and along the Mediterranean coast down into Ethiopia; these, of course, concern people of Hamitic or Semitic stock.

Within South Africa porphyria appears to be commoner among the urban Bantu than in country people. These general findings are confirmed

by Professor L. Eales of Cape Town who has independently been inquiring into the distribution of the disease.

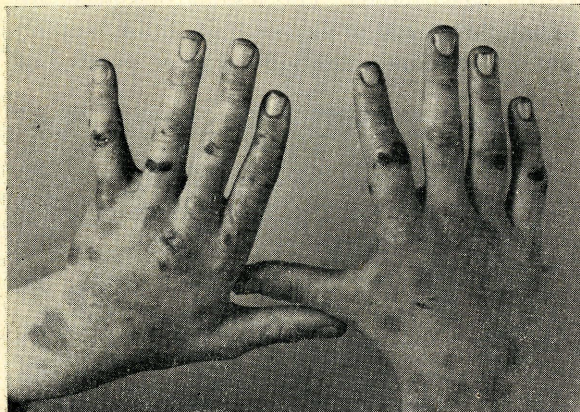


FIG. 1.—Cutaneous porphyria. Hæmorrhagic bullæ, erosions, scars and hyperpigmented remains.



FIG. 2.—Cutaneous porphyria. Dried bullæ, erosions and depigmented remains.

in Southern Africa and elsewhere, so that an inquiry into drinking habits is indicated. What little information I have suggests that the urban Negro uses hard liquor more than country dwellers who are far from teetotal but take milder brews.

Disturbance of iron metabolism may be involved, and Lamont and his colleagues (1961) report that most of the livers from the cases they examined showed the presence of iron, but its extent and distribution differed from

ETIOLOGY

So far in only one instance has a specific cause for acquired porphyria been suspected. In the recent epidemic of porphyria in Turkey described by Çam (1960), it appears that chronic poisoning with a fungicide containing hexachlorobenzene (used on seed wheat) may have been the cause. Schmid (1962) reports that prolonged ingestion of hexachlorobenzene by rats results in a profound disturbance of pyrrole and porphyrin metabolism in the liver.

There is a fairly consistent history of malnutrition and excessive use of spirits in patients with acquired porphyria,

that usually found in South African Negroes. Hæmoglobin levels were normal or elevated, and plasma iron was considerably increased in the majority of patients.

CARDINAL SIGNS

The cardinal signs are demonstrated in figures 1 to 3. The exposed skin is fragile and easily damaged by trivial trauma. The lesions produced are superficial erosions or bullæ containing clear or hæmorrhagic fluid. Healing may be complete or leave scars (sometimes showing milia) or patches of hyper- or de-pigmentation. Sites of election, in rough order of frequency of involvement, are the back of the fingers and hands, the face, the feet, arms, legs, scalp, neck, finger tips, palms, and under the nails. Increased pigmentation of the exposed skin is common in all races and may be the sign which causes the patient to seek attention.

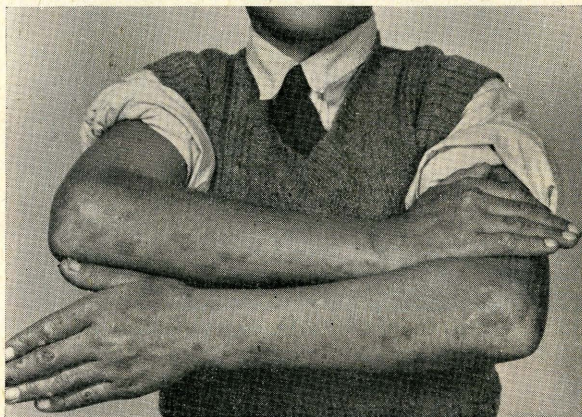


FIG. 3.—Cutaneous porphyria. Erosions and hyperpigmented remains.

Hypertrichosis is common and easiest to see in women. The temporal area is most commonly involved but the forehead and cheeks may also be covered with downy hair.

Patients usually pass red, pink or very dark urine periodically, but may not volunteer this information.

AN APPEAL

To help solve the problem of the causes of acquired porphyria more accurate information is needed about its incidence in Africa and elsewhere. I should therefore be grateful if anyone recognizing cutaneous porphyria could please communicate with me at Simon's View, Sorrento Road, St. James, C.P., South Africa.

This request extends not only to those practising in parts of Africa where the disease seems to be uncommon but also to those in country districts of Southern Africa.

References

- Çam, C. (1960): *Bull. Soc. Méd. Paris.*, **76**, 1305.
 Dean, G., and Barnes, H. D. (1955): *Brit. med. J.*, **ii**, 89.
 Eales, L. (1960): *S. Afr. J. Lab. clin. Med.*, **6**, 63.
 Lamont, N. M., et al. (1961): *Quart. J. Med. N.S.*, **30**, 373.
 Schmid, R. (1962): *Excerpta med. (Amst.)*, International Congress Series, **52**, 53.