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PORPHYRIA TURCICA

(Cutaneous Porphyria Seen in Southeastern Turkey)

General Considerations

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The appearance of a new type of cutaneous porphyria in southeastern Turkey in, and since, the summer of 1955 has been of compelling interest to all students of metabolic disturbances and references to it have been made by several authors.¹⁻¹⁰

This syndrome is characterized by excretion of red urine; photosensitivity of the skin which results in severe blistering, scarring and mutilation of exposed parts of the body; hypertrichosis; hyperpigmentation; interphalangeal arthritis and enlargement of the liver with tenderness.¹⁰

The exact number of cases is not known but the most conservative estimates indicate that the numbers of those affected exceed 3,000. This large patient population in itself is outstanding. The fact that the overwhelming majority of the individuals afflicted

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consists of children makes this new disease type of particular interest to pediatricians, especially because, hitherto, except for rare cases of the congenital type, porphyria has generally been considered a disease of adult life.

In the beginning, authors describing this disease tried to fit it into the known forms of porphyria and some cases were published as «congenital porphyria,»¹ and others as «cutanea tarda type of porphyria.»² Soon it became apparent that a new form of porphyria was being dealt with. The fact that hexachlorobenzene intoxication plays a role in the porphyria seen in southeastern Turkey is now well established¹³ but recent study indicates that genetic predisposition is also a possibility and that hexachlorobenzene, probably, triggers the disease process in those who are predisposed or susceptible. This, if substantiated, may prove our cases to be analogous to Dean's cases^{11,12} in South Africa where a hereditary trait existed which passed benignly from generation to generation and became a problem only when the carriers of the trait were exposed to barbiturates.

In view of these factors it seems unwise to designate this new type of porphyria as a purely toxic one and «porphyria turcica» would appear to be a safer designation to use until such time as genetic and other possible agents of predisposition have been either ruled out or verified.

The author and his associates at the Hacettepe Children's Hospital have been greatly interested in the Turkish type of porphyria for the past five years. Nineteen cases have been studied as in-patients in Ankara for periods of one to two years and a porphyr laboratory, with animal experimentation, has been established at Hacettepe Medical Center of Ankara University. A two-part field survey was carried out, in 1959 and 1960, in southeastern Turkey in which 592 patients and over 3,000 normal controls in the area were examined and etiologic and epidemiologic studies were undertaken. And, finally, in order to be able to describe the clinical entity and the course of the disease in as complete a form as possible, 376 cases were clinically followed in the field for periods of from three to five years. For this purpose a mobile clinic was established in the town of Bismil, Diyarbakir Province, about 500 miles east of Ankara, where there has been a high incidence of porphyria. Reports of some of these studies appear in this issue of the *Turkish Journal of Pediatrics*.^{13,15}

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