Correspondence

Journal of Medical Genetics, 1977, **14**, 234

Heterogeneity among ectodermal dysplasias

SIR,

Settineri *et al.* (1976) reported a large kindred with several patients showing an ectodermal dysplasia which they claim to be the classical X-linked hypohidrotic syndrome, 'with some unusual features' (*sic*). The number of 'unusual features' is, however, so large that I have a suspicion that the authors are not dealing with the classical and very severe form (also known as the Christ-Siemens-Touraine syndrome) but rather with another X-linked condition, probably that described by Lenz (1963) and also characterised by hypohidrosis (E. Passarge, 1973, personal communication). I have called this mild condition Lenz syndrome (Freire-Maia, 1971).

Contrary to the opinion of Settineri *et al.* (1976), Christ-Siemens-Touraine syndrome is a well delineated ectodermal dysplasia whose aetiology is firmly established as caused by an X-linked gene. The fact that, in some families, the 'same' entity does not follow the X-linked pattern of inheritance (a situation already mentioned by Weech (1929) led to the recognition of another syndrome clinically similar to Christ-Siemens-Touraine syndrome—the autosomal recessive hypohidrotic ectodermal dysplasia— by Passarge *et al.* (1966) and Gorlin *et al.* (1970).

NEWTON FREIRE-MAIA
Department of Genetics (UFPr),
P.O. Box AA, 80 000 Curitiba, Paraná,
Brazil.

References

Freire-Maia, N. (1971). Ectodermal dysplasias. *Human Heredity*, **21**, 309-312.

Gorlin, R. J., Old, T., and Anderson, V. E. (1970). Hypohidrotic ectodermal dysplasia in females. A critical analysis and argument for genetic heterogeneity. *Zeitschrift für Kinderheilkunde*, **108**, 1-11.

Lenz, W. (1963). *Medical Genetics*. The University of Chicago Press, Chicago.

Passarge, E., Nuzum, C. T., and Schubert, W. K. (1966). Anhidrotic ectodermal dysplasia as autosomal recessive trait in an inbred kindred. *Humangenetik*, **3**, 181-185.

Settineri, Walkiria M. F., Salzano, F. M., and de Melo e Freitas, Maria J. (1976). X-linked anhidrotic ectodermal dysplasia with some unusual features. *Journal of Medical Genetics*, **13**, 212-216.

Weech, A. A. (1929). Hereditary ectodermal dysplasia (congenital ectodermal defect). A report of two cases. *American Journal of Diseases of Children*, **37**, 766-790.