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).

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