LETTER TO THE EDITOR

Palmoplantar hyperkeratosis and deafness

I found the paper written by Sharland *et al*¹ most interesting. However, in addition to the reference to Bititci² they mentioned, the combination of palmoplantar keratoderma and deafness has been described before, without widespread ectodermal dysplasia, in two atopic brothers,³ in two brothers in a later paper by Hatamochi *et al*,⁴ and in three female members of a family in two of whom there was evidence of minor atopy.⁵

The extent of the keratoderma may vary and there does seem to be a wide range of onset of keratoderma from infancy to late childhood. This compares with the usual early infancy onset of dominant palmoplantar keratoderma when it occurs as an isolated abnormality.

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- 1 Sharland M, Bleach NR, Goberdhan PD, Patton MA. Autosomal dominant palmoplantar hyperkeratosis and sensorineural deafness in three generations. *J Med Genet* 1992;29:50-2.
- 2 Bititci OO. Familial hereditary, progressive sensori-neural hearing loss with keratosis palmaris and plantaris. *J Laryngol Otol* 1975;89:1143-6.
- 3 Frentz G, Everberg G, Wulf HC. Congenital perceptive hearing loss and atopic dermatitis. *Acta Otolaryngol* 1976;82:242-4.
- 4 Hatamochi A, Nakagawa S, Ueki H, Miyoshi K, Iuchi I. Diffuse palmoplantar keratoderma with deafness. *Arch Dermatol* 1982;118:605-7.
- 5 Verbov J. Palmoplantar keratoderma, deafness and atopy. *Br J Dermatol* 1987;116:881-2.

BOOK REVIEWS

All titles reviewed here are available from the BMJ Bookshop, PO Box 295, London WC1H 9TE. Prices include postage in the UK and for members of the British Forces Overseas, but overseas customers should add 15% to the value of the order for postage and packing. Payment can be made by cheque in sterling drawn on a UK bank, or by credit card (Mastercard, Visa, or American Express) stating card number, expiry date, and full name.

Eugenics, Human Genetics and Human Failings: The Eugenics Society, its Sources and its Critics in Britain. Pauline M H Mazumdar. (£40.00.) London: Routledge. 1991.

Historians have in recent years uncovered a rich seam in the history of human genetics and, in particular, eugenics. This book analyses the history of the Eugenics Society in Britain from the time of its inception in 1907 to its recent demise (at least in its original form). It is based mainly on unpublished papers held by the Society itself, which often give a more vivid reflection of the attitudes of

those involved than do their publications. The author hopes that her account will not upset those in the Society who gave her free access to its records, but I expect that it will, at least those for whom the concept of eugenics still holds an attraction.

The Eugenics movement in Britain, led by the Eugenics Society, was in many respects a mirror for the views and prejudices of society, or more accurately the narrow segment of society which was predominant up to the end of the Second World War. The opening section shows how the Society was preoccupied by class, by the dangers that poverty and social injustice posed in threatening to overthrow the established order, and how its proponents attempted to provide a valid biological basis to underpin their strategies. 'Pauperism', 'the residuum', 'the dangerous class' are terms that recur in the agenda of problems to be combatted.

The author traces the closely intertwined development of eugenics and human genetics; the use of pedigrees, especially in relation to mental illness and mental handicap, was often more polemic than scientific, though in contrast to America (and later Nazi Germany), the medical profession and politicians were never to accept the view proposed, that society as a whole would degenerate unless legal measures for isolation and sterilisation of affected people were adopted. Especially fascinating to geneticists is how the extraordinary talent in human genetics between the two world wars, including Haldane, Hogben, Fisher, and Penrose, was arrayed on both sides of the eugenics debate. Even the most convinced anti-eugenicists, such as Hogben and Penrose, nevertheless developed their mathematical approaches and main research areas around the problems dictated by the eugenics movement. The insights into the personalities of these remarkable scientists are also valuable in understanding how their attitudes were formed: Hogben and Penrose as conscientious objectors in the First World War, with Hogben imprisoned during the later part of it; Haldane embracing Marxism just as Hogben was becoming disillusioned with it; Fisher and his 17 year old bride farming a smallholding before his move to the plant breeding station at Rothamstead; what an amazing group of people these were!

In Britain the eugenicists, and the Eugenics Society, largely lost their battle. The irony of Penrose, holder of the Galton Chair, stating that during his tenure "nobody taught eugenics and the Galton Professor of Eugenics was not a eugenicist", and his changing the titles of his department and its journal from *Eugenics* to *Human Genetics*, can be seen as the end of eugenics as a coherent philosophy and policy in Britain, with the change of the Society's name to 'Galton Institute' in 1989 as the epilogue to a long drama. It is now for the members of the new society to ensure that its policy is as new as its constitution.

In the closing pages of this valuable book, the author points out the dangers posed by the human genome project in providing the possibility of a 'new eugenics'. These dangers are real, but, in Britain at least, I wonder whether we will be more likely to see eugenic policies again proposed as the answer to the deep seated social problems of deprivation, inequality, homelessness, and their attendant medical and educational aspects. A walk through the London of today can give pictures not so far removed from those of the Victorian England that gave rise

to the deeply flawed science and damaging social philosophy of eugenics.

PETER S HARPER

(1) **Facts about Cornelia de Lange syndrome.** (2) **Facing the Challenges. A Parents' Guide to Cornelia de Lange Syndrome.** (3) **Cornelia de Lange Syndrome Foundation Album 1989-90.** USA: Cornelia de Lange Syndrome Foundation. 1991.

These three publications are produced and distributed by the de Lange Syndrome Foundation in the USA. Cornelia de Lange syndrome is a rare developmental malformation syndrome and most clinical geneticists will only come into contact with a few cases in their working life. Publications like these are therefore of great value for providing information for families to take home and read at their leisure.

The fact sheet gives a comprehensive summary of the syndrome and would be a useful guide for relatives, teachers, speech therapists, and anyone else who comes into contact with an affected child. A large part of the sheet gives information on the American Foundation and a request for donations which is not appropriate to the UK which has its own well organised parent support group.

The Parents' Guide is a beautifully written booklet, and is by far the most useful publication of the three. The introduction states that it is designed to give both factual information and emotional support to parents facing the challenge of raising a child with the syndrome. Inserted throughout the text are quotes of parents' own experiences of how they felt or coped with different challenges and problems. Sections on getting the diagnosis, medical problems commonly associated with the syndrome, language development, life expectancy, what to say to sibs, and how to cope with the reactions of grandparents and other adults are all included. This booklet contains the information that every new parent wants to know but will never find on the library shelves. The feelings and emotions expressed by the parents in this booklet make essential reading for anyone involved in counselling or caring for families with a child with any form of handicap.

The Foundation Album is a picture book of de Lange children whose families are members of the foundation. Under a photograph of each child is the family's address and telephone number and a short summary of the child written by the parents. It is useful for families who wish to contact others in the same state or country, but most of the children included are from the USA. It would, however, be a useful book to lend to parents of a newly diagnosed case.

M IRELAND

NOTICE

Dr R H Lindenbaum, Consultant Clinical Geneticist in Oxford, died on 10 April 1992. An obituary notice will appear in a future issue of the Journal.