14 Andrews O M Wilkie


This book is subtitled “Genetic and teratologic epidemiological studies” and describes a systematic and exhaustive study of nearly 1000 cases with congenital limb deficiency born in Hungary between 1975 and 1984. Hungary maintains a national computerized record of infant morbidities, and consanguineous pregnancies and terminated, because of prenatally diagnosed developmental defects, during the second and third trimester. Through this database all cases with a limb reduction deformity were ascertained and the diagnosis confirmed by personal examination or evaluation of medical records. Hungary has a birth rate of approximately 150,000 per year. Notification to the Hungarian Congenital Abnormality Register (HCAR) is the task of obstetricians and paediatricians and because notification is required every time an affected child is admitted to a medical institution, ascertainment is nearly complete.

The first chapter is a fascinating and well illustrated account of the history of limb defects spanning two and a half thousand years, from the clay tablets of Nineveh to the present day. There follows a description of the Hungarian systems for the evaluation of congenital abnormalities. The design of this study of congenital limb deficiency and the subdivision into isolated cases and those with other congenital abnormalities is described.

The morphological classification which forms the basis for further evaluation is then introduced. Each of the six morphological groups, terminal transverse, amniogenic, radial and cubital, and foot defects, is dealt with in detail and presents the reader with a wealth of clinical and morphological material.

The study’s strengths are the enormous volume of data amassed in a population with nearly complete ascertainment, and the painstaking and detailed manner in which the data have been collected and evaluated. The figures for the prevalence of various categories of congenital limb deficiency are a major contribution to the field. Its main weakness lies in the reliance on a morphological classification as the starting point for epidemiological evaluation of aetiology. Study of the range of limb defects that can be observed in different limbs of a single child, and hence presumably due to aetiology, casts intuitive doubt on the value of this approach. The eventual aetiological classification of isolated CLD groups ends by citing vascular disruption as an important aetiological factor in four of the six categories, with no recognised factors for atypical split hand and foot and early amnionic rupture, arguably itself a result of vascular disruption, cited in the sixth (amniogenic) group.

The disappointing news that emerges from this book is that despite a huge and thorough research effort, only a few environmental factors such as smoking emerge as potential aetiological factors. Even then, the relative risks observed are small.

The book has accompanied me now on two holidays, which suggests that it is not a very easy read! It will not find a place on the bookshelf of most practising clinical geneticists, nor I expect in the departmental libraries. It should, however, be required reading for anyone considering setting up a Congenital Abnormality Register, and will be of interest to everyone engaged in research into congenital limb defects.

HELEN FIRTH

This book is prefaced with the admirable statement that dermatology needed the new genetics as it had fallen into a rut. Genetic developments have swept through dermatology and the aim of the authors is to "bring the new genetics within the grasp of busy dermatologists".

To this end it has nine chapters dealing with skin development and its disorders, the epidermis and its disorders, the dermis and its disorders, cancer and premature aging, genetically determined benign tumours, genetic inflammatory skin disorders, immunological disorders, and chromosome disorders, ending with a short note on prenatal diagnosis and gene therapy. The chapters are well written and individual disorders are referenced with the MIM numbers. The content is well chosen, referenced, and indexed. I particularly appreciated the section on mosaicism and Blaschko's lines. There are occasional inaccuracies which may well be corrected in a subsequent edition. Colour illustrations are numerous and of good quality, an excellent aid to geneticists using the book as an aid to diagnosis. There is a six page glossary of genetic terms for the benefit of the non-geneticists for whom the book is primarily aimed.

Overall I feel this book will be useful to both dermatologists wishing to update themselves on genetic developments within the field of dermatology and geneticists counselling patients with dermatological features.

S J DAVIES

NOTICES

Human Genome Meeting '96

The Human Genome Meeting '96 will be held on 22-24 March 1996 in Heidelberg, Germany. For further information contact HGM '96 Secretariat, HUGO Europe, One Park Square West, London NW1 4LJ, UK. Tel: (44) (171)935 8085. Fax: (44)(171)935 8341.

5th European Meeting on Psychological Aspects of Genetics

This meeting will be held on 26-28 September 1996 in Rome, Italy. For further information contact Dr G Jacopini, Istituto di Psicologia, CNR, Viale Marx 15, 00137 Rome, Italy. Tel: 0039 6 86090278. Fax: 0039 6 824737.

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The Secretary, Royal Medical Benevolent Fund, 24, King’s Road, Wimbledon, London SW19 8QN
Tel: (0181) 540 9194 Fax: (0181) 542 0494

10th World Congress of the International Association for the Scientific Study of Intellectual Disabilities (IASSID)

This Congress will be held on 8-13 July 1996 in Finlandia Hall, Helsinki, Finland. For further information contact the Congress Secretariat, Kansalaiskonsultit-Congress Services, PO Box 762, FIN-00101 Helsinki, Finland. Tel: +358-0-440 822. Fax: +358-0-492 810.

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