Inherited Disorders of the Skeleton

There is a great need for monographs on the genetics of the various system specialities, so that both this book and the series of which it is the first example are to be welcomed. The book provides a thumbnail sketch of the clinical features and the genetics of over 120 different disorders affecting the skeleton. The 'inherited disorders' of the title is taken fairly literally, in that common but non-Mendelian conditions, such as congenital dislocation of the hip, talipes equinovarus, and idiopathic scoliosis are not discussed, though several rarer malformations and 'anomalads' with no known genetic basis are included. Roughly half of the conditions covered are illustrated by useful black and white photographs, which add greatly to the book's value. The vast majority of them appear to be of the author's own patients, attesting to his wide experience in this field.

The grouping of various types of disorder into chapters seems at times rather arbitrary. I suspect that this was done to make the chapters of manageable length, but it also causes difficulties in finding one's way around, especially in the first three chapters entitled 'Skeletal dysplasias without significant spinal involvement' (this, curiously, includes achondroplasia and chondrodysplasia punctata), 'Skeletal dysplasias with significant spinal involvement', and 'Miscellaneous skeletal dysplasias' (which includes several syndromes not usually thought of as skeletal dysplasias). Similarly, the three chapters entitled 'Osteoscleroses', 'Cranio-tubular dysplasias', and 'Cranio-tubular hyperostoses' separate some entities which, superficially, seem to have quite a lot in common.

Partly as a result of its organisation, the book is not as helpful as it might have been where the diagnosis is not known. This could have been remedied by including some flow diagrams for differential diagnosis in such clinical problems as short limbed dwarfism of pre- and postnatal onset, increased bone density, the child who appears to have a storage disorder, and so on, and by a more consistent attempt to lay down definitive criteria for each condition. Another useful innovation would be to show growth curves for some of the dwarfing conditions on a centile chart, indicating where possible the expected adult height.

Although the blurb on the jacket says that the book is 'based on the case histories of more than 1000 patients...examined by the author in genetic clinics in Southern Africa', and Professor Beighton's experience and authority are evident on every page, the book is in fact based, as it must be, at least equally on a review of the relevant literature. References up to 1977 are quoted and their content and bearing, particularly on the questions of genetics, expertly summarised. One occasionally wishes the author would allow his personal references and practices more play when discussing contentious or difficult issues. For example, how does he actually counsel the sporadic case of pseudoachondroplasia? How thoroughly would he want to examine the apparently normal parents of a child with Freeman-Sheldon or Larsen syndrome?

The references are unfortunately grouped under different topics at the end of each chapter. It would save a lot of page turning if the references for each topic were given immediately after the relevant section of text.

These are relatively minor quibbles compared with the positive value of the book. Professor Beighton has done clinical geneticists, paediatricians, and orthopaedic surgeons a service in presenting a great deal of information in reasonably short and readable form. It is hoped that he will keep the book up to date with frequent new editions. The publishers are to be congratulated on making the book portable without lowering of production.

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The Study of Man. An Introduction to Human Biology