BOOK REVIEWS

The Genetics of Neurological Disorders. 2nd ed. Oxford Monographs on Medical Genetics 18. M Baraitser. (Pp 733; paperback £30, hardback £60.) Oxford: Oxford University Press. 1990.

There is no doubt that if a neurologist or geneticist comes across an unusual combination of signs in a patient then he or she will look it up in Dr Baraitser's book, or alternatively in the Neurogenetic Database that he has compiled. *The Genetics of Neurological Disorders* is a thoroughly comprehensive account of neurological syndromes and is an invaluable source of reference. Where else could one learn about epilepsy with yellow teeth, or epilepsy with infantile balding? The purpose of this second edition is "to update the clinical delineation of genetically determined neurological disorders" in order to help neurologists make an accurate clinical diagnosis before using DNA probes. A secondary aim is "to incorporate new information about DNA".

Not only is Dr Baraitser's survey of neurological publications extensive, it is also critical and accompanied by experienced advice on empirical recurrence risks, although the latter have not changed much between the two editions. Pedigrees and anecdotes are helpful, such as those given for spastic paraplegia on page 278. There are definitions of dentatorubropallidoluysian atrophy and of olivopontocerebellar atrophy on page 298 which are particularly useful to non-neurologists. The application of DNA findings to genetic counselling is well described, particularly in the section on predictive testing for Huntington's chorea, and in carrier assessment in Duchenne muscular dystrophy. However, I was sorry not to read of the contribution made by DNA studies concerning our understanding of the origins or mechanisms of mutations, and the demonstration of gonadal mosaicism.

Any book in such a rapidly expanding subject will contain omissions, but in this edition there are some incon-

sistencies. For example, there is no account of dystrophin and its use in clinical diagnosis, yet the prion protein gene is mentioned. I think that the problem lies in the fact that a book of this size (508 pages of text and 207 pages of references) takes so long to revise; therefore, I recommend that a third edition should be divided into two volumes. It is an improvement to find photographs included in the text, although more would be welcome, particularly of neuroradiology, DNA blotting, or muscle pathology. It is also a relief to find pedigrees the right way up and labelled correctly. This book has a justifiably renowned international reputation as a work of reference and I can wholeheartedly commend this second edition to postgraduates working in the fields of neurology or genetics.

SARAH BUNDEY

POSSUM (Pictures of Standard Syndromes and Undiagnosed Malformations). Melbourne: Murdoch Institute. 1990.

POSSUM is a computer program which uses a microcomputer and videodisc to aid the diagnosis of dysmorphic syndromes and is produced by the Murdoch Institute at the Royal Children's Hospital, Melbourne. The initial versions of POSSUM mainly consisted of the better known syndromes, but subsequent versions have sought to be comprehensive and the latest version (2.5), released in January 1990, contains 1698 syndromes (160 more than version 1.5) and 890 traits for describing syndromes (155 more terms than in version 2.0). In a cursory glance through the syndrome lists in the accompanying manual, I could find only a few mistakes (I did notice that both cystinosis and Flannery syndrome were included twice). The manual is in the same attractive, readable, and easily accessible format as before. One new feature in version 2.5 is the inclusion of a specific field on the screen under the syndrome commentary indicating the chromosomal location of the gene, where this is known. There has been no alteration to the videodisc and version 2.5 is therefore used with the disc that was supplied with version 2.0.

POSSUM can be used in the same way as the London Dysmorphology Database in syndrome diagnosis. One or more traits can be chosen from the accompanying lists and a search made to include some or all of these traits. There is a further refinement in that each choice is weighted as either m (major trait), o (ordinary), or s (specific) and some factors can be excluded (for example, chromosomal abnormality) using the mode x. The search is much faster than the current London program.

The synopses accompanying the syndromes are brief and less thorough than in the London database and only a few selected references are included, compared to the extensive referencing which makes the London system invaluable.

The POSSUM system includes chromosomal anomalies which may occasionally prove useful. I am aware of a recent case where the diagnoses suggested by POSSUM included a chromosome anomaly which was subsequently detected on reviewing the karyotype with that specific chromosomal region in mind.

POSSUM can be used without the videodisc system but this really negates its greatest asset, which is its teaching potential. The pictures produced are of variable quality and the value of some is severely impaired by having the eyes blacked out. It is to be hoped that in future it may be possible to obtain consent from more patients or their parents for full facial photographs to be shown. There are numerous slides available for all of the common syndromes, including displaying the appearance at different ages. For Rett's syndrome there is even a moving video of the hand wringing movements. There is also a visual demonstration of what is meant by many of the traits, useful for easy demonstration of midface hypoplasia, triangular facies, etc. One question mark relates to the accuracy of some of the diagnoses shown in the pictures and there are some cases where there would be room to quibble, but the Melbourne group are to be congratulated on having the courage to display their findings and diagnoses to public scrutiny in such a difficult and subjective field.

It is because of this teaching potential in particular that I believe that the POSSUM system does have a place in genetics departments. It is complementary to the London Dysmorphology