form, is discussed under the heading of primary diseases of cementum. These are, however, small criticisms of a generally excellent text which should be of considerable value to clinicians in many branches of medicine and dentistry.

G. B. WINTER

The Eye in Chromosome Duplications and Deficiencies

By Marcelle Jay. (Pp. xi + 249; Figures + Tables. SFr4.74.) Basel and New York: Marcel Dekker. 1977.

Ophthalmologists have long been interested in genetics. One of the first textbooks on genetics was written in 1934 by an ophthalmologist, Waardenburg, and the tradition has been carried on by Franceschetti and, in this country, by Sorsby. Some ophthalmic hospitals have a genetic clinic (the author helps her husband at the one at Moorfields, London), so that this book, though not written by a clinician, has its origins in the clinic.

Since the advent of banding techniques, many new chromosomal syndromes have been described. Clinicians are aware that eye anomalies appear in many different syndromes, so that the presence of hypertelorism, ptosis, strabismus, or epicanthus are not helpful diagnostic criteria. If less nonspecific features, such as optic atrophy or colobomata, were to be found in certain of the aneuploidies, these would be more useful clinical signs. The main purpose of this book is to describe and tabulate the clinical findings, with particular emphasis on the ocular anomalies in the partial deletions and trisomies. As the book has its origin in a thesis, the detail is excellent and there is a comprehensive coverage of references since the introduction of banding in 1972. In addition, there is a concise first chapter on the chromosome and a useful glossary of terms. The interesting association of retinoblastoma with partial deletion of chromosome 13 is also dealt with. Perhaps potential readers should be warned that duplications and deletions referred to in the title do not include whole chromosomes, so that trisomy 13, trisomy 18, and even Down's syndrome are not described. Surprisingly, an exception is made for Turner's syndrome. Nevertheless, this is a most useful reference work for those dealing with the 'funny looking kid' syndromes.

M. BARAITSER

Myotonia Congenita and Syndromes Associated with Myotonia. Clinical-Genetic Studies of the Nondystrophic Myotonias

By Peter Emil Becker, with contributions by Rainer Knusmann and Erick Kuhn. Translated by Mary F. Passarge. (Pp. x + 181; 146 Figures + 40 Tables. DM.78.) Stuttgart: Georg Thieme. 1977.

Myotonia refers to the phenomenon of delayed relaxation after either voluntary contraction or contraction induced by percussion. The first clear and thorough description of a dominantly inherited form of myotonia was by Dr Asmus Julius Thomsen 100 years ago, who observed the disease in himself as well as in other members of his family. Since then it has become increasingly clear that the inherited myotonic syndromes are a very heterogeneous group of disorders. In fact some 15 clinically distinct forms have now been described. Professor Becker has been particularly interested in these disorders, and this detailed monograph concerns his studies of 142 probands with established myotonia congenita and their relatives, including Thomsen's original family. His major contribution has been to show the clear distinction between dominant and recessive forms of myotonia congenita. The former tend to have an earlier onset, to be more variable, less severe, and less often associated with persistent weakness than the latter. There appear to be at least 3 clinically distinct forms of dominant myotonia congenita: Thomsen's disease itself, a form associated with myalgia, and another with marked cold dependence.

Becker observed the prevalence of dominant myotonia congenita in West Germany to be about 1 in 500 000, though this is clearly an underestimate for often symptoms are mild and ascertainment therefore not complete. In fact, he thinks the true prevalence may be as high as 1 in 23 000, about twice the prevalence of recessive myotonia congenita. Interestingly, malignant hyperthermia was not observed in any of the families he studied.

Besides presenting detailed clinical findings, the monograph concludes with 2 brief sections on biochemistry (by Kuhn) and multivariate analysis (by Knusmann). The results of the biochemical studies (serum and muscle enzymes, serum electrolytes, serum and muscle lipids) are unrevealing, apart from abnormalities in the lipid composition of muscle in dominant myotonia congenita, the relevance of which is not yet clear. With regard to the statistical analyses, an attempt was made to verify Becker's classification firstly by testing the significance between the myotonic types he described, and secondly by attempting to establish an independent classification from the original data. In general, both approaches tended to confirm Becker's intuitive classification.

Altogether this is a very valuable monograph and can be unreservedly recommended to anyone with an interest in this complex group of disorders. It also provides an excellent model for anyone embarking on a clinical-genetic study. It is gratifying to those with primarily clinical interests that this approach is still very worth while and can produce valuable results as well as being intellectually satisfying.

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