
Professor Wiedemann and his associates have contributed greatly to the delineation of syndromes over several decades, and this atlas reflects their experience and the value of careful documentation and follow up. The atlas is now in its second English edition and each entry follows a format with text on one page facing illustrations opposite. The text is telegraphic in style divided for each syndrome into section signs, diagnostic findings, aetiology, frequency, course, treatment, and references.

Many of the illustrations are old and some are taken against a dark background; consequently some appear indistinct and it is difficult to see abnormalities of skin texture or pigmentation. Some well delineated syndromes are not illustrated, such as VAR, TAR, MASA, SDR, E Pall-Scheinker, and CHARGE associations, though I did wonder whether the "unknown" syndrome on page 44 could be this latter disorder. The inclusion of the authors' "unknowns" spread throughout the atlas is a sympathetic and does not help the organisation of the book, which is not explicit anyway.

This atlas, although enlarged in this edition, has not been extensively rewritten and as a consequence recent knowledge about the genetic basis and mechanisms underlying some of the syndromes is not included. For example, osteogenesis imperfecta types IIA–C are referred to as usually recessive disorders whereas recurrences in some families have now been shown to be the result of parental germinal mosaicism.

Most dysmorphologists like to have all the available "syndrome books" to see different cases illustrated at varying ages. Although this atlas would not be my first choice, it is a good value, reasonably comprehensive, and constitutes a helpful contribution to the body of publications.

DIAN DONNAI


The recent advances in understanding some of the ataxias exemplify the different ways in which genetic and other studies can help to elucidate the pathogenesis. This interesting volume presents many new and exciting aspects of ataxic conditions, some of which are not yet fully explained. It is an enjoyable and stimulating book.

First there is a clear account of the classification of ataxic disorders, which emphasises the recognition of those few syndromes that are treatable. An improved delineation of cerebellar syndromes can be made with modern imaging, for MRI techniques show how the cervical cord is atrophied in patients with Friedreich's ataxia while the cerebellum appears normal, whereas in early onset spastic ataxia, the cervical cord is normal but there is atrophy of all parts of the cerebellum. In adult onset ataxia type I (ADCA type I) MRI scanning shows atrophy of the entire cerebellum together with atrophy of thepons, middle cerebellar peduncle, medulla, and upper cervical cord. However, MR-findings in pure cerebellar atrophy (ADCA type III) show atrophy confined to the cerebellum. Patients with the spastic ataxia associated with neuropathy (Charlevoix-Saguenay atrophy) of the superior cerebellar vermis and atrophy of the cervical and thoracic segments of the spinal cord.

Gene mapping has been successfully accomplished in Friedreich's ataxia, ataxia telangiectasia, and some families with ADCA type I. In Friedreich's ataxia there is substantial evidence that only one gene locus is involved; its localisation is a tribute to international cooperation and a lot of hard work and it was the third disease gene locus to be identified. However, it is proving difficult to pinpoint the gene more accurately, owing to its position near the centromere on chromosome 9 and the uninformative nature of markers used in the early studies. Recent identification of CpG doublets (which usually lie alongside coding sequences) and the use of yeast artificial chromosomes to clone segments of DNA from the region are valuable strategies. Moreover, Chamberlain et al are screening cDNA libraries obtained from fetal and adult brain and cerebellum. The prospects for finding and sequencing the gene look promising. Ataxia telangiectasia is an interesting condition in which the type of gene that could cause such varied effects on the immune system and on chromosome stability is unknown. The ataxia itself is a relatively minor feature of the disease and of the many other problems that can occur in patients with ataxia telangiectasia. In recent years recombination in the region of the ATM gene has been shown, and this has raised the possibility of a new approach to treatment. For example, a hard mutation is present in individuals with ataxia telangiectasia, and this is associated with the development of cancer. If we can find a way to introduce normal ATM into cancer cells, this could provide a new approach to treatment of these patients. The recent improvements in respiratory, feeding strategies, and dietary management will hopefully continue to improve the prospects for patients with this condition.

SARAH BUNDEY


A useful addition to this series, Genetic Engineering provides an excellent introduction to the technological wizardry behind the science of molecular genetics. Little previous knowledge is assumed and the text

In the preface to this book, David Brock states that its purpose is to inform clinicians about the impact of molecular genetics without either "blinding them with science in the first few pages or frustrating their intellects with immature sociology". He suggests that no book that successfully steers this path has previously been written, but that seems more than a little unfair on David Weatherall's The New Genetics and Clinical Practice (NGCP), published by Oxford University Press and now in its (1991) edition. In fact, the aims of the two books appear superficially fairly similar, making comparison inevitable. So, should one favour the Cambridge or the Oxford version? With the dubious record that I have attended both universities and still find the Boat Race a complete bore, I trust that I can be considered impartial. Professor Brock's is the shorter book by about 100 pages and it sticks closely to its stated brief of describing the application of molecular studies to genetic disease. The text is arranged in nine chapters, covering the basic genetics of disease, DNA technology, types of mutation, gene tracking, molecular cytogenetics, multifactorial disorders, and future developments. It is well illustrated with numerous clear line drawings and photographs, although lacking colour the overall impression is less attractive than NGCP. On strictly genetic issues it is very comprehensive, more so than NGCP, and this doubtless reflects both the author's own background and the impact of developments in the field. Thus the interested clinician wishing to know more about very contemporary topics such as imprinting/uniparental disomy, use of the polymerase chain reaction in DNA diagnostics, fluorescence in situ hybridisation (illustrated with the only colour photograph!), X inactivation, the molecular basis of the fragile X syndrome, and screening for cystic fibrosis would find a full and entertaining treatment in Brock's book, together with appropriate references for further reading.

On the other hand the chapters on mutation and multifactorial disorders stick closely to classification and methodological aspects, and NGCP is likely to have more information on the molecular pathology of a disease of specific interest to the clinician. Brock also says little about the broader impact of the recombinant DNA revolution in fields such as infectious disease, cell biology, and biotechnology, but clearly he does not consider these to be part of his brief. Both books give only passing mention to germline or somatic mosaicism, a pity since these encompass some interesting genetic phenomena.

In summary, Molecular Genetics for the Clinician is very clearly written, although lacking for gene tracking, the back-to-basics approach, and vying for the same audience, Brock's book is better written. The nine chapters are devoted to the following topics:

1. Introduction
2. DNA technology
3. Types of mutation
4. Gene tracking
5. Molecular cytogenetics
6. Multifactorial disorders
7. Future developments

One can never really appreciate a subject without knowing its history and this is nowhere more true than in human genetics. Dronamraju has been interested in the subject for many years and has written extensively on J B S Haldane. At the International Congress on Human Genetics in 1991, a satellite meeting, under the chairmanship of Dronamraju, concerned itself with the history of human genetics and the current volume represents some of the presented papers as well as others added later. The result is very variable.

In an opening chapter, James Neel lists nine reasons for the amazing transformation of the subject over the past 40 years. Most of these he attributes to various technical innovations. But I would add (having once been a plant geneticist) that there was in the early years a reluctance amongst plant and animal geneticists to study humans because the subject was viewed as insufficiently challenging and unlikely to prove rewarding!

Other contributors to the volume address specific issues: the development of blood group genetics (Bodmer) and linkage analysis (Morton). These authors clearly could have written more extensively and in more detail on their chosen subjects and perhaps should have been encouraged to do so. Two chapters are by non-geneticists: Burian, a philosopher, on the development of human cytogenetics in France, and Olby, an historian, on the theory and practice of diagnosis. In my opinion these are the most rewarding contributions. They are well written and thought provoking.


For the rusty 'expert' or inquiring novice the first chapter introduces the ground rules of gene organisation and expression in man and bacteria providing a sensible foundation on which to introduce an array of elegant techniques. Chapters 3 and 4 describe methods of genetic analysis from low resolution mapping to gene structure and expression studies including such innovations as automated DNA sequence analysis, computer gene search analysis, and, of course, the ubiquitous PCR. Chapters 5 and 6 focus on the recent recombinant part of the technology merely 'walking' and 'jumping' all the way to the construction of genomic and cDNA libraries. The cystic fibrosis story provides an excellent example of positional cloning and general strategies used for identifying genes. The final chapters encompass more recent developments of cloning in higher organisms from YAC vectors to the generation of transgenic mice and the application of classical reverse genetics using in vitro gene targeting approaches for genomic and developmental studies. This leads naturally to a potential for application in gene therapy and the study and cure of human disease.

This book is a handy pocket guide to genetic engineering, ideally suited for clinicians, students, and researchers new to the subject or simply as a quick refresher for the specialist.

JOHN HARVEY


This extremely well presented book aims to give information about techniques for prenatal diagnosis of fetal abnormalities within a framework of up to date genetics and counselling, recognising that for many physicians and co-workers their knowledge of genetics may be dated. This is a multiauthor book of 25 chapters well organised into five main sections: general principles, risks in reproduction, clinical and laboratory diagnostic techniques, and management of fetal anomalies. Most readers will use it as a reference source, dipping into chapters, but it would certainly be feasible for a postgraduate student to read the whole book and emerge at the end having an up to date and comprehensive overview of reproductive genetics, counselling, and prenatal diagnosis.

The standard of all of the chapters is high. I would compliment particularly the chapter on genetic counselling which is of general value although organisationally written from a North American perspective. The first trimester ultrasound illustrations are clear and well annotated and throughout the book the line diagrams, illustrations, and general layout are good.

I have few adverse comments; the state- of-the-art is high and the references (Eldridge analysis is not widely available) is a little out of date, potentially teratogenic drugs are discussed in three sequential chapters, and the impact of the situs ambiguous syndrome on the preimplantation prenatal diagnosis are not discussed at all.

I would certainly recommend the book for obstetricians and geneticists in training, for genetic counselors, and as a reference source for specialists in these two fields. As is the nature of genetics, this book will soon be out of date but the references quoted are up to 1990 and it is an excellent foundation for future editions.

DIAN DONNAI

The remainder of the book is a collection of chapters, each of which traces the develop-