clinical description, followed by a critical analysis of the relevant genetic data and information concerning recurrence risks. The book lists all the key references for each group of disorders.

My only regret in the difference between this and the first edition is that the extra chapter on mitochondrial disease has meant that the appendices in the first edition have been omitted. Although the issues with regard to frequency of consanguineous matings and parents of patients with autosomal recessive disease (appendix 1, first edition) and Bayes' calculations for X linked disorders (appendix 2) are covered in other texts, I found the list of neurological disorders inherited in an X linked fashion a very useful reference source and I would hope that Dr Bundey might consider including it in any future editions.

Who will find this book useful? Within the field of clinical genetics, I think that it is a book that all trainees and consultant clinical geneticists should consider owning. In my own practice, I find that I use this book and Dr Baraitser's book, *The Genetics of Neurological Disorders*, to address different problems. For the commoner diseases, where I am unsure of the clinical features, I start with Dr Bundey's book and move on to Dr Baraitser's book if I want to cross check anything. For the rarer disorders and for an exhaustive review of published reports I use Dr Baraitser's book.

In addition to clinical geneticists, I think Dr Bundey's book will be of enormous use to consultants and trainees in paediatrics and neurology. The extensive re-revision of the text, particularly with regard to molecular genetic diagnosis of neurogenetic disorders, means that for all those who purchased a first edition, serious consideration should be given to buying the second edition.

SUSAN HUSON

An Atlas of Clinical Syndromes: A Visual Aid to Diagnosis. 2nd edition. H R Wiedemann, J Kunze, F R Grosse, H Dibbern. (Pp 564.) London: Wolfe Publishing Limited. 1992.

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 main signs, supplementary 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 VATER, HARD +/− E, Pena-Shokeir, and CHARGE association, 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 little idiosyncratic 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 good value, reasonably comprehensive, and constitutes a helpful contribution to the body of publications.

DIAN DONNAI

Inherited Ataxias. Advances in Neurology. Volume 61. Ed A E Harding, Thomas Deufel. (Pp 217; $119.00.) New York: Raven Press. 1993.

The recent advances in understanding some of the ataxias exemplify the different ways in which genetic and other studies can help to elucidate the causes of disease. 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 the pons, middle cerebellar peduncle, medulla, and upper cervical cord. However, MRI findings in pure cerebellar atrophy (ADCA type III) show atrophy confined to the cerebellum. Patients with the spastic ataxia associated with neuropathy (Charlevoix-Saguenay ataxia) have 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 uninformativeness 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 not known, but work with lymphoblastoid cell lines (Lindahl) suggests that a phosphodiesterase may be deficient.

Homologies with mouse diseases have been fruitful in a number of genetic disorders, and dominant ataxia is no exception. Desaiah *et al* have studied the phosphoinositide metabolism in the cerebellum of mice with the Lurcher mutation. Abnormalities were found in the cerebellums of Lurcher mice and of human patients with ADCA type I, and warrant further study. A long and detailed chapter on neurotransmitters in the cerebellum illustrates their complexity, but at present does not point the way towards effective treatment. Two chapters on regeneration of neurones in cell culture and on Purkinje cell transplants in mice indicate possible future therapeutic strategies.

It is useful to have in this book an up to date account of the fascinating group of neurodegenerative disorders known as prion diseases. The diseases are clinically variable, with ataxia and presenile dementia being the two commonest features. In humans the disease may be inherited as an autosomal dominant (five point mutations and two insertions in the prion protein gene have been described) or may be acquired (by the rare inoculation with an aberrant prion protein which then catalyses a post-transcriptional event so that further abnormal prion protein is derived from the host's genome) or, most commonly, by an unknown mechanism different from the first two. It is interesting that the host's genotype plays some part in the manifestation of disease. In inherited prion protein disease, homozygosity for a polymorphism of codon 129 leads to early onset of disease, while the few patients who have developed Creutzfeldt-Jacob disease after being given cadaveric derived growth hormone are predominantly homozygous for valine at this position. Patients with sporadic Creutzfeldt-Jacob disease are more often homozygous for either valine or methionine at codon 129 of the prion protein than would be expected from the distribution of these polymorphisms in the general population.

It used to be thought that the neurodegenerative disorders formed a depressing group of conditions because no prevention or treatment could be envisaged. However, this book illustrates that such a view is no longer true and that there are great possibilities for the future at least in regard to the ataxic conditions. This volume, therefore, in the series of Advances in Neurology, is of great interest to neurologists and to those geneticists interested in neurological diseases.

SARAH BUNDEY

Genetic Engineering. Medical Perspectives Series. J Williams, A Ceccarelli, N Spurr. (Pp 132; £14.00.) UK: Bios Scientific Publishers. 1993.

A useful addition to this instructive 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

is clear, well illustrated, and pleasantly 'readable'.

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 currently in use. Chapters 2 and 3 describe methods of gene 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 4 and 5 focus on the actual recombinant part of the technology merrily '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 ingenious gene targeting approaches for gene and developmental studies. This leads naturally to a potential for application in gene therapy and the study and cure of human diseases.

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

Molecular Genetics for the Clinician. D J H Brock. (Pp 289; £35.00.) Cambridge: Cambridge University Press. 1993.

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 3rd (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, cancer, 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 rapid 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 fuller exposition 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 germinal 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 the laid back style of NGCP. There are very few errors. For those wishing to acquaint themselves from scratch with the current state of human molecular genetics, it would certainly be an excellent buy; but for the broader canvas, NGCP would still get my vote.

ANDREW WILKIE

The History and Development of Human Genetics. Ed K R Dronamraju. (Pp 303; £56.00.) Singapore, London: World Scientific Publishing Co. 1992.

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 among most 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 of hereditary diathesis. In my opinion these are the most rewarding contributions. They are well written and thought provoking.

The remainder of the book is a collection of chapters, each of which traces the development of the subject in various countries: Canada, France, Italy, Hungary, Japan, Israel, Egypt, Chile, Brazil, and India. Here, dare I say, some degree of chauvinism is sometimes evident! And there are also some notable absences.

Though this can in no way be considered a comprehensive history of human genetics, searching does reveal some interesting facts. For example, the end of wartime censorship allowed Wald to publish his sequential analysis developed for quality control of bombs using a statistic subsequently referred to as lods. Also the discovery of trisomy 21 by Lejeune, Gautier, and Turpin was achieved under the most primitive working conditions in a small laboratory without either gas or water and unimproved since 1900! But finding such gems requires a thorough reading as there is no index. And in a subject such as a history, to publish it in the unattractive format of 'camera ready copy' would seem to be unnecessary.

ALAN EMERY

Reproductive Risks and Prenatal Diagnosis. Ed M I Evans. (Pp 369; £73.10.) Connecticut: Appleton and Lange. 1992.

This extremely well presented book aims to give information about techniques for prenatal diagnosis of fetal abnormality 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 statement that high resolution chromosome analysis is not widely available is a little out of date, potentially teratogenic drugs are discussed in three sequential chapters, and fluorescent in situ hybridisation and preimplantation prenatal diagnosis are not discussed at all.

I would certainly recommend the book for obstetricians and geneticists in training, for genetic associates, 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