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 Bunday 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 Bunday'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 Bunday'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 Dibern. (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 edi-

tion, 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 uninformative 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. Desai *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