preimplantation diagnosis and only a small number of babies have been born from such procedures. These brave pioneers must go through the early stages of a high failure rate and errors. It is important also to emphasise the pitfalls and dangers of the procedures. The main problems arise with the diagnostic tests, the problems of test contamination, extra sperm which may be present in the perivitelline space of the embryo, cellular mosaicism, and the presence of abnormal cells which may or may not be representative of the future fetus. Then, there are problems associated with the IVF procedure itself: low efficiency of pregnancy, multiple pregnancy, and the invasive treatments of the woman. Also, in all attempts at preimplantation diagnosis, a later prenatal diagnosis by chorionic villus sampling or amniocentesis is needed to check the accuracy of the earlier embryo diagnosis. Preimplantation diagnosis may be a safe and efficient approach to the prevention of genetic disease in the future but it has a long way to go. If you are interested to know how far along the road it has come since its recent beginnings just six years ago, this book is the best you will find to bring you up to early 1993.

MARILYN MONK


The editors describe the first edition of this book as being "at the cutting edge of the interface between clinical and basic neuroscience". This is reflected in their choices of contributors, all authorities in their fields and with high profile in the author lists in current neurology/genetics journals. The chapters are, in general, comprehensive, well written, and more than adequately referenced. The book is divided into sections, each containing between one and five chapters.

Part I, chapter 1, begins gently with descriptions of nucleic acids and DNA structure and translation, and progresses rapidly (in 17 pages) through DNA markers, restriction enzymes, and polymorphisms to linkage, cloning, and gene therapy. One small criticism is the use of the term "reverse genetics" where most would now use "positional cloning". In addition the section devoted to Lesch-Nyhan disease would seem superfluous in view of the full chapter coverage later.

Chapter 2 is a list of the chromosomal locations for genetic neurological diseases and genes of relevance to the nervous system. There are some minor omissions: tuberous sclerosis has entries at 9q34, 11q14, 12q22 (the last two are rather dubious localisations) but not chromosome 16 where there is a major locus. This is, however, mentioned in the chapter on TS. Similarly there are no listings for myotonia congenita and Becker's generalised myotonia which are associated with the skeletal muscle chloride channel at 7q32-qter. Some entries list the disorder alongside the gene product but this is not universal; dystrophin does not appear with Duchenne and Becker muscular dystrophies. Chapter 3 gives a small (three pages) taste of linkage analysis, computer programs for such, and the problem of genetic heterogeneity. It is perhaps a little too short for a reader with no previous knowledge of this field.

The disease specific chapters which follow contain many highlights and very few disappointments. The chapters on metabolic disorders admirably cover the clinical, biochemical, and molecular features of the appropriate diseases. In addition they offer therapeutic protocols where available or discuss potentials for therapy. Part XII contains a single chapter; a scholarly, multiauthour review of the prion diseases of humans and animals. Frusiner and colleagues also provide ample scope for further reading on this fascinating topic, with 311 references.

Part XIII contains reviews of the muscle dystrophies and, with the exception of the chapter on FSH/limb-girdle dystrophies which contains less than two pages of text, this section amply summarises the salient clinical and molecular features of these disorders. Roses (chapter 41) has been able to incorporate some of the rapidly appearing new data on the unstable trinucleotide repeat sequence associated with myotonic dystrophy and this chapter is well referenced up to 1992. At this time it was known that expansion of another trinucleotide repeat sequence, this time in the androgen receptor gene, was the causal mechanism of bulbospinal muscular atrophy (Kennedy's syndrome). There is no mention of this, either in this chapter or (that this reviewer could find) elsewhere in the book.

Part XIV contains, again, a single contribution. DiMauro's review of mitochondrial disorders is comprehensive, clearly written, well supplemented with explanatory diagrams, and excellently referenced. Part XV deals with "degenerative disorders" and begins with a chapter on heredity ataxias. Unfortunately this was a little disappointing as over half of this chapter reviews disorders given more comprehensive coverage elsewhere in the text. Part XIX (neuropathies and neuropathies) contains limited clinical information, the discussions focusing on gene mapping (for spinal muscular atrophy), mutations (transhydrogenin and amyloid neuropathies), and peripheral nerve physiology (axonal neuropathies). The review of the genetic epilepsies (part XX/chapter 63) is particularly thorough, including a description of the epileptic Mongolian gerbil! It also includes useful guidelines on risk estimates and prognosis for (in particular) relatives of persons with epilepsy. The final chapters (gene therapy and consequences of gene mapping) conclude this volume with general discussions of the future developments in the field of neurogenetic diseases, although specific examples are given for replacement therapy in animal models. The reviewer finds it surprising that there is no mention of the possible societal and political consequences of the Human Genome Project.

Is this the neurology equivalent of The Metabolic Basis of Inherited Disease? The answer must be "yes". The criticisms above are minor compared with the wealth of information provided overall and this volume will certainly fulfil its goal in being of use to clinicians caring for patients and for scientific investigators in this field. Every genetic/neurology unit should have access to this volume but most people will want their own copy and anyone with a colleague visiting North America would do well to persuade him/her to add the extra weight to their return luggage as the US price equates to £150.

JOHN MACMILLAN