**Book reviews**

**Oncology and Immunology of Down Syndrome**


The National Down Syndrome Society of America has sponsored two previous conferences, one on the molecular structure of chromosome 21 and the other on the neurobiology of Down syndrome. This third conference, held in December 1986, was concerned with two very important aspects of the disorder, namely the increased risk of leukaemia, particularly in early childhood, and the increased susceptibility to infection which still remains the main cause of death. The 14 edited chapters of this book address these matters.

Persons with Down's syndrome (DS) are some 10 to 20 times more likely to develop acute leukaemia than others. Both DS and acute leukaemia, quite independently, increase in incidence with maternal age and with radiation exposure in utero. About 40% of the leukaemia in DS may possibly be of a particularly rare type referred to as acute megakaryoblastic leukaemia. However, which DNA sequences on chromosome 21 are involved in this process is still not clear. Furthermore, lymphocytes in DS are hypersensitive to the effects of X irradiation as well as DNA damaging agents, and it is suggested there may be a defect in DNA repair which might in some way be related to the increased frequency of leukaemia and perhaps even the neurodegeneration which occurs.

With regard to the immunology of DS, complement and B cell number and function usually appear normal. In contrast there are defects in the polymorphonuclear-phagocytic system and in T cell number and function. The specific T cell functional abnormalities are documented in detail by several contributors. Interestingly, there are similarities in the haematological and immunological abnormalities in mouse trisomy 16 and human trisomy 21, the former therefore providing a useful model for DS. Altogether this is an interesting volume which will be of value to anyone concerned with this common cytogenetic abnormality.

ALAN E H EMERY

**Genetic Risk, Risk Perception, and Decision Making**


This book contains the proceedings of a conference held in Belgium in 1986, which brought together genetic counsellors and social scientists to discuss genetic risk and decision making. As expected from a collection of papers by authors from different backgrounds, it is rather a mixed bag. There are three papers, presumably given for the benefit of non-geneticists, which give a broad overview of chromosomal and Mendelian disorders and genetic linkage. There are also several papers by social scientists reviewing decision making theory in some detail. It is no doubt a reflection of the paucity of collaboration between these disciplines that only one group had practical experience of applying analytical decision making techniques to genetic counselling (Drs Pauker and Pauker from Boston who have used a 'decision analytic model' when counselling couples about amniocentesis). The remaining three chapters discuss the results of studies of attitudes towards predictive testing in Huntington's chorea and prenatal diagnosis for cystic fibrosis, Down's syndrome, and neural tube defects.

The tenet of the symposium on which these proceedings are based was to provide a 'platform for geneticists, psychologists and social scientists, to explain their respective disciplines to each other'. However, the editors note that at the discussion period, at the end of the meeting, there was antagonism between those doing genetic counselling and those developing theoretical models. Unfortunately, in the main, the social scientists did not help their cause by presenting papers using unnecessary jargon and failing to show the relevance of analytical decision theory to families faced with choices involving genetic risk. At least one exception is a paper by Ch Vlcek, which gives a clear overview of risk analysis, perception, and decision making theory. The concept of a 'good' decision in a genetic context is thoughtfully developed, and the author points out that any use of formal decision analysis in genetic counselling must not be at the expense of more psychological approaches. While most people involved in genetic counselling would be familiar with the use of Bayesian theory in the derivation of genetic risks, they might be reluctant to accept that Bayes (in the guise of rational decision analysis) can provide 'right' answers about appropriate courses of action for families at genetic risk. However, we should be open to examining the ways in which we
help families make these decisions. This book will be of undoubted interest to people involved with research into genetic decision making, but I would be surprised if it succeeds in having much impact on the way in which genetic counselling is practised.

L Kerzin-Storrar

Cystic Fibrosis: The Facts

This attractively presented book, hardback, which has been prepared “for parents and all those who care for children with cystic fibrosis”, contains clear clinical descriptions of the disease, assisted by numerous charts and diagrams. There are illuminating passages written by parents and the CF children themselves, which give valuable insights on the problems of day to day living. It is a little surprising that a low fat diet is still advocated for many patients at the Manchester Clinic, but no-one would deny that cystic fibrosis is a disease with variable clinical expression and few proven, beneficial lines of treatment.

The section on genetics is concise and interesting, reflecting the major interests of the authors. The structure of DNA is clearly described. Information is given on the inheritance of the disease as it may affect the extended family, prenatal diagnosis including chorionic villus sampling and amniocentesis, and genetic counselling.

Research into the basic defect, tackled in the final major chapter, is of course a complex matter and much ground has been covered. One wonders if many parents with the exception of those biochemically and genetically trained, would understand it fully, and perhaps it should be made clear that the ‘CF protein’ described in this chapter is not the one coded for by the CF gene. However, this section does provide a comprehensive and encouraging look at the broad spectrum of cystic fibrosis research. There is an informative glossary and appendix.

I can recommend this book as a most useful and in many ways intriguing addition to the cystic fibrosis literature.

M Goodchild

Genetics and Malformations in Art

This book represents a valiant attempt on behalf of the editors to present an illustrated introduction to a fascinating aspect of art history along with a useful bibliography. Artists and craftsmen have often portrayed genetic disorders and malformations in their work, either because of their supposed mystical significance or merely for their intrinsic interest. However, the main problem is often distinguishing a presumed accurate reproduction of a defect by the artist from mere fantasy and artistic licence. Some of the contributors to this monograph do make this distinction and their comments are then that much more valuable, but unfortunately this is not always the case. Detailed knowledge and understanding of a work of art, its creator, and the times in which it was produced are important if more than fanciful guesswork is to be achieved. Nevertheless, many of the illustrations will no doubt be reproduced as interest catching introductions to lectures. The serious student, however, will have to search elsewhere for critical and more in-depth appreciation of the subject.

Alan E H Emery