Book reviews

Oncology and Immunology of Down Syndrome
Progress in Clinical and Biological Research
Volume 246. Edited by E E McCoy and C J Epstein.
(Pp 242; £49.50.) New York: Alan R Liss. 1987.

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.

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Genetic Risk, Risk Perception, and Decision Making
Edited by G Evers-Kiebooms, J-J Cassiman, H Van den Berghe, and G d'Ydewalle. Birth Defects:

Journal of Medical Genetics 1988, **25**, 647-648

Original Article Series, Vol 23, No 2. (Pp 350; £42.00.) New York: Alan R Liss. 1987.

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 Paulker and Paulker 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