The Child with Multiple Birth Defects

This book provides a structured framework within which to approach dysmorphology. It starts with chapters on malformation, deformation and disruption, syndrome prototypes, and delineation (that is, lumping v splitting). Cohen then goes on to draw this together in a rational approach to syndrome diagnosis. Further chapters then cover dysmorphic growth and development and neoplastic aspects of syndromology. The important topic of psychosocial aspects of syndromes is also given considerable, and sympathetic, attention.

Cohen’s book was first published in 1982 and the major concepts it deals with are familiar to anyone with an interest in dysmorphology. However, it should also appeal to anyone new to this field, students, and doctors in other specialities, and would be a useful addition to any hospital library. It provides a framework in which one can begin to fit together what can otherwise seem confusing and unrelated findings. It is thought provoking and gives considerable insight into dysmorphology.

This is a delightful book and on the whole the style is lucid and very readable, but some of the illustrations and flow charts are confusingly laid out and not adjacent to the related text.

C McKeown

Leucocyte Typing III: White Cell Differentiation Antigens

Leucocyte Typing III is the third in a series of books devoted to human leucocyte differentiation antigens. It presents the proceedings of the 3rd International Workshop on Leucocyte Differentiation Antigens held in Oxford in September 1986 under the chairmanship of Professor Andrew McMichael and colleagues, who also edit the book. These workshops have been organised as a result of rapid developments in monoclonal antibody technology. They provide the main forum for the screening of leucocyte monoclonal antibodies and the nomenclature of human leucocyte differentiation antigens. The organisation, proceedings, and presentation of the workshop results, admirably summarised in this book, will be familiar to those with a interest in the HLA Histocompatibility Workshops. The book presents the results of patterns of monoclonal antibody reactivity on panels of different leucocytes, recognising 'clusters of differentiation' (CD). The first workshop defined 15 CD antigens; Leucocyte Typing III takes this to 45, with several others still undefined.

The book is organised into sections dealing with the CD antigens of the main leucocyte populations (T and B lymphocytes, myeloid cells), platelets, and antigens which are non-lineage restricted. Each section presents the analysis of a workshop study and is followed by a series of original research papers relevant to that section, using reagents to defined CD antigens. These address questions of epitope diversity, cell and tissue distribution, and functional significance of various leucocyte molecules. Some also use techniques such as somatic cell hybrids, gene cloning, and transfection of CD genes.

This book is essentially directed at those producing or using monoclonal antibodies to human leucocyte antigens. Nonetheless, the definition of the 45 CD antigens, as well as the listing of more than 900 monoclonal antibodies and their originators, makes it a valuable reference work for anyone interested in using monoclonal antibodies to analyse constitutional and acquired changes in leucocyte subpopulations in relation to human disease. Since much of the information is completely new it would be hard to find better value for over 1000 pages. The multi-authorship has been handled by careful editing to ensure that the information content is high. All of this information could be confusing, if the editors and publisher had not taken care over the layout, with clear uncluttered tables and diagrams. To present the most complete picture of current knowledge relating to human leucocyte antigens in a field of rapid change is one achievement. To have accomplished such a publishing feat in a short space of time is another.

G M Taylor

Molecular Approaches to Human Polygenic Disease

Recombinant DNA technology over the last few
years has revealed much about the molecular pathology of single gene disorders, and has led to new and precise methods of carrier detection and prenatal diagnosis for such disorders. There is now increasing interest in applying this technology to the commoner multifactorial disorders, and this book addresses this problem.

There are 18 chapters which are largely concerned with atherosclerosis (genetic factors, LDL receptors, apo-A, -B, and -E polymorphisms, and Lp(a) variations, etc); hypertension (genetic control of erythrocyte Na-Li countertransport); diabetes mellitus (HLA and insulin hypervariable region markers); psychiatric disorders (schizophrenia, Tourette syndrome, and affective disorders); and the molecular genetics of HLA related disorders. Gene therapy is discussed, and finally there is a very thoughtful contribution by an epidemiologist. The discussions after each chapter are particularly interesting, and clearly interest is largely centred on attempts to identify important 'candidate genes' for these disorders.

It seems that for some time yet the new technology may play little part in prevention through prediction of high risk groups. What seems much more likely, and particularly valuable, is that they may provide new insights into pathogenesis. It will be particularly interesting to many medical geneticists to see what is revealed about common congenital malformations, a subject which has so far remained largely untouched by the new technology.

**ALAN E H EMERY**

**Sickle Cell Disease**


Dr Serjeant is Director of the Medical Research Council Laboratory in the University of the West Indies, Jamaica, where he has carried out a life time study of sickle cell disease, and is running a long term cohort study to define its natural history. His book is intended to help clinicians with the practical management of patients with the disease, and admirably achieves this aim by crystallising the author's unique experience into a sort of encyclopaedia that is an indispensable reference volume for anyone involved in the day to day management of patients with sickle cell disease.

This book is primarily clinical. In it Dr Serjeant amalgamates his experience with that in the world's publications, to provide a comprehensive clinical guide. The book is divided into 37 (mostly short) chapters. The first three deal with the genetics and fundamental science of sickling of the red cell, and the next three contain a simple and highly readable account of its pathophysiology. Chapters 7 to 22 cover the potential implications of homozygous sickle cell disease for different organ systems one at a time, an approach that is necessary because of the great clinical variability of the disorder. These chapters include some very helpful evaluation and discussion; for instance, the section on the prevention of infection by the use of prophylactic antibiotics is particularly interesting. Chapter 23 is a review of sickle cell disease in pregnancy, and the next three chapters discuss the way that the pathological picture is modified in Hb S/C disease, sickle cell β thalassaemia, and other rarer genetic combinations. There are very useful discussions of sickle cell trait, the changing pattern of the homozygous disease with age, causes of death, and approaches to management, including blood transfusion and surgery. The final chapters on epidemiology and on the sickle cell clinic are interesting, but too short.

The general tone is critical and conservative: when the evidence to support a relationship or the value of a therapeutic approach is inadequate, Dr Serjeant says so.

The book has some limitations. For instance, its title suggests comprehensive coverage of the subject, but there is no epidemiological description of the disease in the population observed, or a summary of the approach to it at the community level, or systematic information on the relative frequency of different complications in the population. Though the Jamaican cohort study is alluded to, there is no description of its history, philosophy, or methodology, nor is there any discussion of genetic counselling or the role of prenatal diagnosis.

For the reader, these would be some of the most interesting aspects of the work that Dr Serjeant is uniquely qualified to describe. Also, the exact placement of the numerous references, often in the middle of sentences, fragments the prose and makes for a difficult read: it would have been better to cluster them at the end of sentences or paragraphs.

In short, this is an indispensable reference book for the clinician, but in the future, to satisfy his audience, Dr Serjeant should follow up with an account of the history of his unit and the development of his approach to the community at risk for sickle cell disease.

B MODELL.