minor points. Dr Davison's section is concluded by three appendices, containing invaluable records of all case summaries and histories in the first two, with some of the pedigrees in the third.

The second part (21 pages) of the monograph is an aetiological investigation by Drs Swift, Benson, and Studdy of those patients hospitalized in Leybourne Grange Hospital who have an IQ of 50 or less, and at least one sib with an IQ of 60 or less. They found 66 such patients from 30 families and divided them into diagnostic groups of known aetiology, those with similar physical, but no biochemical, abnormalities among the affected sibs, those with no physical or biochemical abnormalities and a group of four families, in each of which a new biochemical error had been found (both the affected sibs in these four families were excreting excessive amounts of tyrosin, beta-alanin, L-methylhistidin, and hydroxylysin, respectively). Column chromatography was employed in the biochemical investigations and the advantage of this technique is discussed. Case histories, listing physical and other abnormalities, are given, thus providing a valuable source of comparison for other workers in the field. The four new amino-acidurias provide scope for the study of treatment and prevention.

This small monograph should be carefully studied by all workers in the field of genetics and mental subnormality and an example taken from the methods and techniques used.

RENATA LAX


Immunogenetics is a fundamentally important subject for at least two reasons. Firstly, immunology although in a state of near transcendental euphoria, is undoubtedly concerned with some of the most important processes in biology. In addition to resisting the incursions of pathogenic micro-organisms, important roles are claimed in differentiation, in fetal development and in oncology. Secondly, the combination of immunochemistry and formal genetics has already yielded an astonishing variety of information about eukaryotic gene action. Autosomal allelic exclusion and the demonstration that the 'one gene, one polypeptide' rule did not apply in immunoglobulin synthesis are good examples.

The term immunogenetics has been used to include the study of any genetic polymorphism providing immunological methods were used. However, one can distinguish within this definition a much more homogeneous area involving the genetics of immunity, that is the genetics of the immune response and of the structure of immunoglobulins irrespective of whether immunological techniques are used in their elucidation. Professor Fudenberg and his colleagues have produced a remarkable review of a field which incidentally has been greatly enhanced by their own activities. They have properly concentrated on the genetics of immunity. The book begins with a gentle introduction in the form of a wide ranging essay. Chapter 2 deals with immunoglobulin structure and evolution and provides an adequate background for a remarkably comprehensible discussion of the genetics of immunoglobulin molecules covered in Chapter 3 and of antibody variability covered in Chapter 4. In Chapter 5 the genetics of histocompatibility, a brief review of T and B cells and their interaction, antigen receptors on lymphoid cells, immune deficiency states in man and animals, autoimmunity and lymphoid neoplasias, and finally the genetic control of specific immune response are reviewed in only 27 pages. In Chapter 6 the authors consider human blood groups and to some extent depart from the narrower definition of immunogenetics by including systems which, although they may have been detected by immunological (usually serological) techniques, presumably have, with one or two exceptions, little to do in vivo with the immune response. Finally, in three appendices, blood group terminology, standard abbreviations for immunological terms (immunology must be 'Top of the Pops' so far as jargon goes) and a list of methods for detecting antibody (with an indication of sensitivity) are given.

This book can be highly recommended not only 'for the graduate and medical student with a fair knowledge of biochemistry and genetics' as the authors modestly put it, but also for more advanced workers, including physicians, who want an up-to-date, brief, and understandable overview of this rapidly advancing and still slightly anarchic field.

R. HARRIS


The value of this series of publications in enhancing medical communication in the birth defects field has been proven many times over. The present volume should be stimulating for all concerned with the diagnosis, treatment, and counselling of congenital heart disorders.

Several articles are of outstanding interest. Dr van Praagh's segmental approach to the diagnosis of congenital heart disease does indeed simplify the seemingly unlimited types of cardiac malformation. Dr Mitchell writes interestingly of her large prospective study of over 50,000 pregnant women with regard to risk factors in the mother and cardiovascular malformations in the infant. Mothers with congenital heart disease themselves have the greatest risk of producing a baby with congenital heart disease, mothers with diabetes have the second highest risk and their babies are apt to have defects of the great vessels, while mothers producing babies with transposition of the great vessels often have an unusual history of oestrogen deficiency. Increased maternal age also
increases the risk of congenital heart disease in the offspring. Dr Reynolds found that in a group of children with congenital heart disease, aortic stenosis, usually subvalvular, was about 2\(\frac{1}{2}\) times more common in those with intrauterine retardation than in those without it, suggesting that intrauterine growth retardation of children with cardiac defects is due to intrauterine organ hypoplasia. Idiopathic endocardial fibroelastosis is the topic of three authors (Hutchins, Rosenquist and colleagues, and Rose) who, respectively, feel that it is a disease secondary to interstitial myocarditis in many cases, that it is probably not an immunologic disease, and that after the birth of the first affected child, the sib recurrence risk is 17.7%. Fascinating discussions of hereditary cardiovascular malformations in dogs by Drs Patterson and Mulvihill stress the value of research using these available animal models.

Several new syndromes with cardiovascular components are described, and many known syndromes are further elucidated, including the controversial Noonan syndrome, with pedigrees suggesting a dominant inheritance discussed by Nora and colleagues. The Holt-Oram syndrome, the surdo-cardiac Jervell and Lange-Nielsen syndromes and the genetic mucopolysaccharidoses are reviewed in detail. Genetic counselling, presentation, and cardiac manifestations of the familial form of hyperlipoproteinaemia are lucidly presented. A most useful contribution is Nora's and Spangler's excellent summary of known recurrence risks for cardiovascular anomalies.

We are reminded by Dr German that chromosomal analysis has not revealed aetiological changes in isolated congenital heart disease; however application of techniques with greater resolution will test for possible minute imbalances. Dominant forms of hereditary familial cardiomyopathy and hereditary ventricular hypertrophy are presented, representative of the few congenital heart disorders inherited in Mendelian fashion.

The many case presentations are most instructive and the appendix, compiled by Dr Char, of heritable congenital heart disease types with a recent reference for each, is a most valuable feature for all readers.

In short, this useful volume recommends itself as a reference text for the personal or unit libraries of all clinical geneticists and cardiologists.

Anne H. Child


Miss Ruth Wynne-Davies writes in her preface regarding the orthopaedic and genetic aspects of this book: ‘Working in the no-man's land between the two disciplines, I am no expert at either.' This is modest but untrue; she is, if I may say so without frivolity or offence, that very rare bird, an expert in both. Indeed, there is no one in this country who has contributed more to the knowledge of orthopaedic genetics by personal and widely organized field studies, and to produce valid results the worker must have a first-class background in clinical orthopaedics as well as in genetics, lest he or she be deceived in the diagnostic field.

This is an excellent little book. It starts with a primer of genetics and practical advice on planning a survey and genetic counselling, all directed primarily at the orthopaedic reader. Miss Wynne-Davies knows the limits of her clinical colleagues and does not try to dazzle them with the (to us) complex mathematics, a knowledge of which is essential to the academic geneticist. She makes it all sound easy, but I trust that the orthopaedic surgeons nevertheless will be chary of trying to give genetic counselling except in the most clear-cut circumstances. She is brief and not unduly didactic; only in some of the clinical descriptions does she sometimes get a little too carried away. The text is leavened with outline drawings, some of which are brilliant, most of them good, and only a few of which miss the mark.

But for a book which is necessarily something of a synopsis she has managed, while writing with authority and clarity, to produce a most readable text. It is not an academic book for geneticists; the few well-chosen references are almost all clinical, and the every-day working orthopaedic surgeons will be most grateful to Miss Wynne-Davies for giving us just what we have been waiting for.

T. J. Fairbank