irritating not to find ‘thalassemia’ or ‘hemoglobinopathy’, though ‘globinopathies’ appears. The defects are relatively minor and the book as a whole succeeds admirably in its object. There can be few geneticists who would not find it of the utmost value in understanding the very stuff of their science. The book is produced to the very high standard one has come to expect of this publisher.

L. I. WOOLF


Pseudocholinesterase is an extraordinary enzyme which has diverse functions. The activity in the serum is measured to determine liver function and is also investigated to assess the degree of poisoning by anticholinesterases. It is of importance for the anaesthesit and the surgeon because it attacks succinylcholine. It is for this reason that this muscle relaxant is short acting and has become most useful in anaesthetics. In psychiatry, suxamethonium is used to prevent the side-effects of electro-convulsion therapy. The physiological function of pseudocholinesterase is still under discussion. It is of particular interest in this connexion that pseudocholinesterase in the brain, in the intestines, and in the nerve endplates of muscle is always found side by side with true cholinesterase which is the truly physiological enzyme hydrolysing acetylcholine. High substrate concentrations of acetylcholine inhibit the true cholinesterase, whereas pseudocholinesterase can act even on unphysiologically high concentrations of acetylcholine. Possibly the function of pseudocholinesterase is the protection of the vital cholinesterase against accumulation of acetylcholine at high concentration. Pseudocholinesterase was the first serum protein for which a genetic control was reported. It exists in several allelic forms, and one of them, the so-called ‘silent’ gene variant, is of interest because its gene expresses itself by producing no enzyme activity at all in vivo. However, the authors of this book suggest that there is in fact some small residual activity present.

The volume deals with these many aspects of pseudocholinesterase in detail. In its introduction, it discusses the biochemical interpretation of pharmacogenetic phenomena as a whole, and then describes the biochemistry of pseudocholinesterase by defining that of the enzyme itself and differentiating it from the biochemistry of alyleresterases, carboxylesterases, lipases, atropinestersases, and other cholinesterases. It then discusses the usual common variant of the enzyme, its inhibition, reactivation, and different techniques of measuring activity. Other chapters deal with the dibucaine resistant variant, the fluoride resistant variant, and the silent gene, giving a number of hypotheses for their inheritance. In addition to the usual pseudocholinesterase and its variants, there is found associated with them a so-called C₃ component which is related to pseudocholinesterase rather than haemoglobin A₂ is related to normal haemoglobin A. Enzyme kinetics are discussed in detail, as is the connexion between the structure of substrates and enzyme action. One chapter is devoted to the use of pseudocholinesterase estimation as a liver function test and another is given over to the role of pseudocholinesterase in anaesthetics, the management of apnoeas, and the occurrence of apnoeas in different context. There is a useful section on techniques at the end. This book is to be highly recommended.

H. LEHMANN


This book is a continuation of McKusick’s catalogue of X-linked traits, patented in 1962. A similar catalogue on autosomal recessives was begun in 1962, and a year later computer methods were initiated, making it possible to begin a catalogue of dominant phenotypes in 1964. This resultant volume deals with 837 dominants, 531 recessive, and 119 X-linked traits. Each entry carries a brief description of the phenotype, a summary of genetic information available, and key references. A scholarly introduction indicates the value of this formidable undertaking as a contribution to genetics and to the clarification of the specific significance of hereditary affections. An authors’ index of some 6000 entries and a surgical index with about 3000 entries add to the value of this unique encyclopedia. Because of its comprehensiveness and accuracy, this book is assured of a place in every medical genetics department.

ARNOLD SORSBY


This beautifully organized volume breaks radically with the purely descriptive accounts of mental disorders. This is seen at its best in the 4th and 5th of the eight sections into which this book is divided, and these deal with the hereditary mental defects induced by metabolic disorders and those associated with chromosomal anomalies. Both these sections, the one contributed by H. Bickel and H. Cleve and the other by W. Lenz, have the further merit of covering critically a scattered literature not readily available in summary form. Over 40 metabolic disorders and almost as many chromosomal defects are discussed. Mental deficiency is dealt with on a more clinical basis in two further sections, idiopathic disorders being considered by E. Zerbin-Rüdin, while
variety syndromes in which mental deficiency figures is considered by G. Koch. Koch also contributes a full section on the epilepsies, and this covers the genetic findings in electroencephalography, and there are also clinical and pathological studies on laboratory animals. A shorter section on neuroses and psychopathies contributed by E. Strömgren is sufficiently up to date to include the reports on the XXY anomaly in criminal offenders. E. Zerbin-Rüdin, who contributed the section on idiopathic mental deficiency, is also responsible for the sections on atrophies of the brain and on endogenous psychoses: the genetic aspects in these difficult spheres are discussed critically.

As in the earlier volumes of this Handbook, the production is worthy of the contributors' and the editor's outstanding efforts.

Arnold Sorsby


Dr. Pratt's book fills a gap in English medical literature. Over the past century, clinicians have provided a rich but chaotic account of heredodegenerative and familial disorders affecting the nervous system, and a treatise was badly needed that would provide both a systematic account and a critical discussion of relevant world literature. Here we have both.

This monograph is essentially a catalogue of most of the known genetically determined disorders as they affect the central nervous system, but inevitably a measure of selection has been applied—thus a discussion of muscular dystrophies is included because it comes within the purview of the neurologist, but inborn metabolic errors, such as Hartnup's disease, are dealt with very briefly because of their absorbing interest to workers in other disciplines and because they are dealt with adequately elsewhere. And yet perhaps these metabolic disorders are a paradigm for our understanding of most of the conditions described here, since they are probably also biochemically determined. Dr. Pratt deals with his subject from the standpoint of the physician and not from that of the biochemist, but in doing so is somewhat unreserved in his acceptance of the clinical assertions of others. Of course, this is inevitable in reviewing ancient descriptions of obscure disorders (and in contemporary medicine, 'ancient' means anything more than 15 years old, the subject matter of at least one-third of the contents of the bibliography).

Most of the clinical descriptions are deliberately brief, yet for an intelligent understanding of what is written here, a clinical appraisal is essential, and actual first-hand experience very desirable. The reader is not spared the need to read original articles but this is facilitated by a comprehensive bibliography of over 2800 references. This synoptic approach is wholly justifiable otherwise the treatise would have been very lengthy and perhaps turgid.

I have no major disagreement to cite. In the discussion of epilepsy, it is a pity that the more recent studies of Ounsted and his colleagues (Clinics in Developmental Medicine, No. 22, 1966) relating temporal lobe epilepsy in childhood to febrile convulsions are not mentioned, because they may prove to be important, not only from the therapeutic standpoint, but also in genetic counselling in this disease which is still bemused by old wives' tales. It is a pity, too, that, particularly in the so-called abiotrophic disorders, there is no discussion of the age-specific data available for estimating the risk to siblings, but it must be recognized that often there is an abyssmal lack of reliable information for such computations, anyway.

Inevitably there are difficulties over classification and nomenclature, arising out of the differing interpretation of others' findings. It is becoming abundantly clear that the traditional clinical and even neuropathological features are not sufficiently discriminatory to provide any fundamental differentiation. Yet these parameters have provided the material for wearingly uncritical discussion for over half a century, and of course Dr. Pratt has been obliged (for the time being) to work within this very limited framework. One hopes that biochemical advances in the next two or three decades will be such that, in future classifications of hereditary neurological disorders, we will have the advantage of a more basic understanding of the various disease processes, and that this monograph will thus be eclipsed (but not redundant). The duration of its pre-eminence will be a useful measure of the rate of progress in neurology. In the meantime, diseases that are genetically determined, directly or indirectly, form such a large and often unsuspected part of neurological practice, that all neurologists should possess this book—I assume that clinical geneticists will have it already.

John Wilson


McKusick's little volume is an excellent introduction to human genetics in general. Two introductory chapters deal with the basic aspects of chromosomes and genes in inheritance. Of the remaining chapters two deal with the gene in individuals—bringing in oral matters as protein structure and enzyme deficiencies—and genes in differentiation and development. Two further chapters deal with the gene in families and in populations; the necessary elementary mathematics being most adequately covered. The three concluding chapters deal with genetics in relation to evolution, medicine, and society. Both the arrangement of the text and the presentation of the matter are imaginative and attractive.