Book Reviews

Books and Monographs

Blood Groups in Man. 5th ed. By R. R. Race and Ruth Sanger. (Pp. xxviii + 599; illustrated + tables. 110s.) Oxford: Blackwell Scientific Publications. 1968.

Race and Sanger's book on *Blood Groups in Man* was first published in 1950 and is now coming out in its fifth edition in English, besides other editions which appeared in Spanish and German. In 1950 Sir Ronald Fisher wrote in the foreword that he was glad that Race and Sanger had been induced to undertake the heavy labour of preparing a modern book on the blood groups giving 'special attention to inheritance'. The fact that it is now possible even to think of blood groups without relating them to genetics indicates the development of the past 18 years in which Race and Sanger's work has played such an important part. In the present edition they state that while in 1950 the title 'Blood Groups' seemed appropriate for a book dealing almost entirely with antigens of the red cells, they have felt for some years that they owe an apology to those other inherited factors found in blood, the various haemoglobins, serum groups, the enzyme variants, and to the so 'splendidly complicated' antigens of the white cells. The reader, however, will be grateful that they have kept to what one might call classical blood groups and not let their book overflow into an indeterminate compendium.

One of the recent exciting developments is the finding that one of the blood groups, Xg, is linked with the X chromosome, just as haemophilia and other X-linked characters. This makes it necessary for the authors to discuss chromosomes at some length, and on page 552 of this book there is a tentative map of the human X chromosome, probably of the short arm. On it are found the loci for the blood groups Xg, ichthyosis, ocular albinism, deuteran, glucose-6-phosphate-dehydrogenase, protan, and haemophilia. The distance between the various loci are given in centimorgans. There cannot be many books of this kind first published 18 years ago and still maintained by the same authors, which have so much of the original enthusiasm still in them. The authors and the publishers both have to be congratulated on the appearance of this book in its fifth edition.

H. LEHMANN

The Congenital Methemoglobinemias. Physiology and Pathophysiology of the Methemo-

globin Metabolism. Bibliotheca Haematologica No. 28. By Otmar Tönz. (Pp. v + 146; 26 figures + 6 tables. sFr./DM. 39. \$9.35. 78s.) Basel/New York: S. Karger; London: Academic Press. 1968.

This monograph deals with two groups of rare hereditary conditions which result in increased methaemoglobin levels and persistent cyanosis, namely Hb-M disease and enzymopenic methaemoglobinemia.

Hb-M disease was first described by Hörlein and Weber in 1947 and 1948 as a type of familial methaemoglobinemia characterized by dominant inheritance. Other families have subsequently been discovered in various parts of the world, and it is now known that at least five different molecular species of Hb-M exist, i.e. Hb-M Boston, Saskatoon, Iwate, Hyde Park, and Milwaukee: two of the haemoglobins have substitutions in the α chain and three in the β chain. Other possibly distinct types await exact analysis. It is suggested that the substitution of a larger amino acid for histidine or valine leads to a complex forming between trivalent haem iron and the reactive side chain of the amino acid, which constitutes an 'internal ligand'. This complexing of the trivalent iron probably explains its resistance to reduction by reducing agents. As in other haemoglobinopathies, the amount of abnormal component formed is smaller than the theoretically expected 50%: in Hb-M disease it has been reported to vary between 20–30%. Homozygosity has not been observed, and it is likely that the occurrence would be lethal, at least in the case of α chain substitutions. Dr. Tönz discusses the fact that in several cases Hb-M has been apparently absent in both parents of the propositus, and speculates on the possibility of a relatively high mutation rate. Clinically, affected patients have life-long cyanosis but without impairment of physical capacity or apparent fertility.

Congenital enzymopenic methaemoglobinemia was shown to be based on an enzyme defect for the first time by Q.H. Gibson in 1948—the enzyme concerned is NADH diaphorase. Most of the affected families have been of European ancestry, though 13 families have been observed in Eskimos and Alaska Indians. In contrast to the Hb-M diseases, the typical enzymopenic type is inherited as an autosomal recessive. (Rare dominant forms possibly exist.) Presumed heterozygotes have no symptoms and have normal methaemoglobin levels in their blood. However, measurement of the rate of methaemoglobin reduction and enzyme assays give values for heterozygotes intermediate between the normal and the very low values observed in