

This is a book which clinical geneticists and those interested in malformations will enjoy looking at and reading. It is probably best bought together with the companion volumes rather than in isolation. It supplements but does not replace other illustrated catalogues which have been recently published on the malformation syndromes.

J. INSLEY

**Alzheimer's Disease and Related Conditions.** A Ciba Foundation Symposium. Edited by G. E. W. Wolstenholme and Maeve O'Connor. (Pp. xi+316; figures+tables. £4.) London: J. and A. Churchill. 1970.

Alzheimer's disease is a moderately infrequent form of presenile dementia, with a well-defined course and clinical picture, and characteristic cerebral pathology. Genetically it is interesting because of the indications of genetic heterogeneity within this clinico-pathologically delimited syndrome. A number of families have been described, in his contribution to the symposium Pratt says 'more than a dozen', in which a completely typical form of autosomal dominant inheritance is shown, with equal sex representation, and nearly complete penetrance. In comprehensive series, however, selected on a clinico-pathological basis, these typical families do not appear; and what one sees instead is a moderately raised familial incidence and a female preponderance of about 3:1, ie, the indications of a polygenic genetical basis. The genetical evidence, therefore, is strongly in favour of regarding the syndrome as being a mixture of at least two different and independent conditions. This is a conclusion which, for the most part, both clinicians and pathologists are unwilling to accept, as is made plain in these papers.

The symposium reported here was predominantly a pathological one, with interesting and original pathological work. Participants came from Belgium, Germany, the Argentine, Sweden, Hungary, the USA (7) and Britain (10). Active work is going on in histology, histochemistry, and electron-microscopy. Most promising, perhaps, is the study of the ultramicroscopic structure, ie the abnormal intracellular microtubules, which differentiate more than one kind of lesion, and may lead to a successful breaking down of the clinico-pathological continuum extending into normal senescence, senile dementia, and cerebrovascular disease, by which most workers are at present both obsessed and bewildered.

The reviewer would suppose that this book is an important one for the neuropathologist. Its genetical interest is not great. Apart from Pratt's paper reviewing the present state of genetical understanding, the main interest lies in the communication by Jacob (Marburg, Germany) of a family with 6 females in 3 generations showing a form of Alzheimer's disease with, in every case, the unusual feature of myoclonic twitchings appearing early in the course. The literature is reviewed by Jacob.

The book is well produced, with an abundance of good pathological illustrations, good papers, lively discussions, but rather inconclusive results.

E. SLATER

**Serendipity in St. Helena. A Genetical and Medical Study of an Isolated Community.** By Ian Shine. (Pp. xv+187; illustrated+tables. £3.75.) Oxford: Pergamon. 1970.

St. Helena shares with its near neighbour Tristan da Cunha many of the aspects of an isolated community. It is however an older settlement and has population of some 5000 against the 350 or so of Tristan da Cunha. A genetical survey of this highly inbred population with a culture of its own and its particular racial amalgam is a worth-while task which Dr. Ian Shine carried out single-handed while acting as medical officer to the island during his national service in the Army. The present book, based on an examination of almost the whole of the population, records his findings.

One study was devoted to hallux valgus. On St. Helena as elsewhere it was common among those who wear shoes and uncommon among the barefooted. Regression analysis showed however that an additional factor, probably genetic, was also operative. Furthermore some families seemed particularly susceptible to the deforming effect of shoes, while others developed the deformity in spite of their barefootedness. An equally detailed study on the frequency of ischaemic heart disease revealed a substantial incidence in the 1500 subjects over the age of 30 examined. This was unexpected, but clear aetiological conclusions were not possible.

In the course of the overall survey a large number of genetic disorders were observed, including some that are distinctly uncommon, such as albinism, retinitis pigmentosa, lithopaedion, ichthyosiform erythrodermia and Christmas disease. Undescribed disorders such as gross unilateral genu valgum, 'familial St. Helenian fever', and dwarfs with pseudobrachydactyly constituted a group of considerable interest. A high incidence of pterygium presented a puzzle, for the commonly accepted exciting factors, such as dust, wind, and dry air, could not be invoked as an explanation.

The genetic problems disclosed in this investigation are discussed in a critical penultimate chapter on inbreeding. Like the rest of the text this is competent, but this useful volume is not well served by its rather flippant title and somewhat jaunty style.

ARNOLD SORSBY

**The De Lange Syndrome.** By J. M. Berg, B. D. McCreary, M. A. C. Ridler, and G. F. Smith. (Pp. vii+127; figures+tables. £3.15; \$8.40.) Oxford: Pergamon Press. 1970.

The de Lange syndrome was first described by a Dutch paediatrician from Amsterdam in 1933. Since that time a wealth of data has accumulated on the clinical