Book reviews


During the middle decades of the 19th century the familial form of progressive adult chorea associated with dementia was recognized but, for reasons that are not altogether apparent, it was George Huntington's comparatively late account of the disorder which was first to be widely publicized. To celebrate the centenary of this description, a symposium was held during March 1972, near where Huntington used to live, in Columbus, Ohio and the various reviews and papers read at this meeting make up the present volume. As it took a long time for the disease to be widely recognized, so knowledge concerning the condition has accumulated gradually, and indeed if the symposium had been held 15 to 20 years ago, there would have been little to report that would have surprised even Huntington himself. The considerable amount of work done in recent years, and the rather more modest advances in knowledge, justify the publication of a large work on the disease, although it may be somewhat unexpected to those not directly concerned with the condition, that it has been possible to produce 826 pages on this comparatively obscure disorder.

In this book most aspects of the disease are covered in detail, few are treated in an incomplete or unbalanced manner, and virtually everything that is worth saying about the condition is said. As there are many authors there is inevitably a lot of repetition which, whilst it is sometimes annoying, is not really the bad thing that it is sometimes said to be, for occasionally a better perspective can be obtained by reading several accounts of the same subject. The book is divided into a number of sections, each containing one or more review articles as well as several smaller papers. The subjects dealt with include 'History', 'Clinical variants and differential diagnosis', 'Genetics and epidemiology', 'Early detection', 'Pathology', 'Biochemistry', 'Experimental models', 'Behaviour and social aspects', and 'Management'.

The section on history contains some of the other early descriptions, as well as that by Huntington and it also contains several detailed references to one aspect of the history of the disease which is now in serious doubt, without there being any acknowledgment of the existence of these doubts. This concerns the story of the emigration from Essex in 1630 of some of the alleged ancestors of a number of American choreics, a story that is based on genealogical work which has recently been called into question. The subject of variants is dealt with in the appropriate section and is also touched upon in the section on pathology. The clinical and pathologically features of these variants are outlined several times and their genetic characteristics are described. The question as to whether all of these variants are really distinctive is also raised. The section on preclinical diagnosis is headed by a thoughtful and provocative review and amongst the papers that follow are two on the laevodopa load test, this being a test in which subjects at risk are given oral laevodopa to establish whether or not involuntary movements can be provoked, the assumption being that those with the abnormal gene will develop such movements whereas those without will not. Contained in the lead review is a plea for answers to the many difficult questions posed by the possible preclinical diagnosis of such a grave disease, but this aspect is either ignored or at best only mentioned in passing in subsequent papers. The section on pathology contains several papers on the variety of changes that can occur in the classical form of the disease as well as in some of the variants. The value of cerebral biopsies is also discussed. The lead review on biochemical by Barbeau is immensely detailed and contains 268 references, most of which are to studies that have yielded negative or equivocal results. Included also is the paper by Perry and colleagues in which they first report their finding of low levels of gamma aminobutyric acid in the brains of choreics, an observation that has since been followed up by the finding of changes in the amounts of glutamic acid decarboxylase activity in similar brains. This is one of the very few occasions when the book can be seen to be already out of date. The section on experimental models contains information on a number of drug induced and naturally occurring animal disorders; amongst the latter being the endearing but, in this context, probably irrelevant acrobatic rabbits and the quaint hyperkinetic episodes of Scottie dogs. The section on management touches on many aspects including the management of the family as well as the care of the choreic individual, but the parts dealing with the drug treatment of chorea are a little eccentric to British eyes, for the two drugs that are used widely in this country—tetrabenazine and thiopropazate—are either mentioned very briefly or, in the case of the latter drug, not at all.

Notwithstanding the criticisms, this book is the best and most detailed account of this disorder that is currently available and it should be on the shelves of every major library used by neurologists, geneticists and psychiatrists. The problem is that it is enormous, and as it is hardly practical to read it from cover to cover, a newcomer to the disease would find it difficult to obtain an overall view of the disease without a lot of effort. It is also very expensive, the United Kingdom price being well over £20, which means that it will be caught by comparatively few private individuals and, one suspects, few small medical libraries either. Thus, the two major contemporary sources of detailed information on the condition—this book and the even more expensive sixth volume 'Handbook of clinical neurology'—will not be available to most of those who might wish to consult them. Something smaller and cheaper is needed.

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DAVID L. STEVENS
Huntington's Chorea, 1872-1972

David L. Stevens

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Updated information and services can be found at:
http://jmg.bmj.com/content/12/1/117.citation

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