Surveys, Symposia, and Transactions


The aims of the Muscular Dystrophy Group of Great Britain are first to form a friendly link between sufferers and their families, and second to make available funds for medical research in muscular dystrophy and other muscle diseases. Recently the Research Committee decided to support all disorders affecting muscles, whether of the muscle cell itself or of its innervation, and such conditions as myasthenia gravis and spinal muscular atrophy are now also its concern.

This volume contains the 33 papers (collected into 5 chapters) presented at the Fourth Symposium held in January 1968, and also an edited version of the discussion which followed the individual papers. Chapter I consists of the Joan Vincent Memorial Lecture, delivered in memory of the first secretary of the Group and given by F. J. Nattrass, who classifies the 'pure' muscular dystrophies, reviews modern developments, and gives some fascinating biographical details about Duchenne—a strange, sauntering mariner-like figure—from Boulogne.

In Chapter II (clinical and genetic studies) there are four papers of particular interest to the reviewer. (1) The correlations between myasthenia gravis and disorders of the thyroid gland (J. A. Simpson)—and these are by no means confined to thyrotoxicosis; (2) second thoughts on classification of the muscular dystrophies (J. N. Walton and D. Gardner-Medwin) in which the non-progressive and ill-understood congenital myopathies (such as central core disease and nemaline myopathy with its affinities with Marfan's syndrome) are discussed in some detail and compared with the better known progressive types; (3) a description of the important benign X-linked muscular dystrophy (A. E. H. Emery); and (4) the exciting discovery that the cross-innervation of the two fibre types (fast and slow), the histochemical pattern of human skeletal muscle can be changed, suggesting that the dystrophies may prove eventually to be an aberration of neural influences rather than primary muscle disorders (Dubowitz).

In Chapter III (anatomical and pathological studies), two papers are selected for comment. The first questions whether the two major types of nerve lesion, Wallerian degeneration (as exemplified by isoniazid neuropathy) and segmental demyelination without axon damage (e.g. alloxan diabetic neuropathy), are in fact distinct entities, and the authors (Hildebrand, Joffroy, and Coers) suggest that the two forms may merely be the result of differences in the severity of the disease process. The second deals with the structure and fine structure of the intramuscular nerve endings in dystrophia myotonica, and the authors (Allen, Johnson, and Woolf) show that though the architecture of the motor end plates is largely normal, yet there is a tendency for them to be unusually elaborate, suggesting that the number of available binding sites for acetylcholine and the amount of acetylcholinesterase present would be increased. It is of great interest that somewhat similar findings are found in two quite different conditions, namely amyotrophic lateral sclerosis and myasthenia gravis, the latter in some ways being the reciprocal of dystrophia myotonica.

In Chapter IV (biochemical studies) there is a paper (W. H. S. Thomson) critically assessing the reliability of the biochemical identification of the carrier-state in X-linked muscular dystrophy, and there is emphasis on the importance of bearing in mind the diurnal variation of the enzyme creatine phosphokinase. When this is done the tests are satisfactory, for of 9 undoubted heterozygotes only 1 gave normal serum enzyme values (despite a suggestive EMG) in every circumstance. One of the heterozygotes investigated was found to have microsymptoms shown by incapacity to sustain rope skipping as a schoolgirl.

Finally in Chapter V (neuropathological studies) there are two papers (D. Gardner-Medwin and R. G. Willison) on the problems encountered when using electromyography to try and detect carriers of the Duchenne-type of dystrophy. These pitfalls should be widely known by registrars and others who ask for 'EMG studies, please' and feel disappointed when the reports are equivocal.

The book is in the usual good value Pitman style and is strongly recommended, being of use both to general physicians and also to neurologists, biochemists, and neuropathologists—and the discussions following the papers round off the topics particularly well.

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Since the war ended both the benefits and the hazards of medical treatment have been extended, under the National Health Service, to all persons with symptoms without reference to the cost to the individual. As some diseases are not responsive to therapy by the time they are manifest through symptoms (it is convenient to follow the classical usage of symptoms, by which a disease is manifest to a patient, and signs, by which it is manifest to others), various attempts have been made to increase the scope of treatment by inverting the normal order of discovery and seeking signs rather than symptoms, in the hope that earlier treatment would make life both longer and more pleasant.

In some organs, such as the lung, which can be displayed by x-ray to reveal early stages of tuberculosis