price is reasonable for such a monumental work, and it does define an enormous number of words in unexpected ways. I have no doubt that the majority of these terms are useful in their own disciplines.

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Myotonic Dystrophy. 2nd Edition. By PETER S. HARPER. Harcourt Brace Jovanovich Limited. 384 pages. Price £40.00. ISBN 0 7020 1364 1.

From many points of view myotonic dystrophy is a fascinating yet tragic condition. It is inherited as an autosomal dominant trait and extensive families with the condition are not uncommon. The manifestations are protean and include not only myotonia, which gives the condition its name and refers to a delayed relaxation after a muscle contracts, but also muscle weakness which affects mainly the facial as well as the more distal limb musculature, smooth and cardiac muscle involvement, some degree of mental deterioration, various endocrine abnormalities, cataracts, premature baldness and various skin and skeletal abnormalities. However, a gene carrier may sometimes demonstrate no more than very mild myotonia or symptomless cataracts and the demonstration of these defects in the past provided tests for presymptomatic diagnosis.

However the gene locus has been located on the long arm of chromosome 19 and several closely linked DNA markers (muscle creatine kinase MM, apolipoproteins C1 and C2) are now proving valuable for presymptomatic as well as prenatal diagnosis. But the gene itself has not yet been isolated, and the basic biochemical defect still remains unknown. Though drugs can be useful in controlling disabling myotonia there is as yet no effective treatment. Furthermore, several fascinating aspects still remain unexplained. These include the wide range in age at onset, the fact that some individuals may remain only slightly affected throughout their lives whereas others may be severely affected even from birth, particularly in the case of infants born to affected mothers. Finally, anticipation, which refers to progressively earlier onset and greater severity in successive generations, now seems likely to be a true phenomenon and not, as many once thought, merely a reflection of biases in sampling affected families.

All these aspects of the condition are reviewed in detail in this new edition, which is a well written, well referenced, up-to-date, and scholarly text, the author himself being a world authority on the disorder. It has no competitors and can be highly recommended to anyone interested in this fascinating condition.

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