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Myotonic Dystrophy: Majors Problems in Neurology.
By Peter S. Harper. Philadelphia, PA: W.B. Saunders; 2001. 436 pages.

Considerable advances have taken place in the understanding of myotonic dystrophy since the second edition of this book was published in 1989. There is now a much clearer understanding of the varied clinical features of this disorder. Remarkable progress has been made in understanding the molecular genetics of myotonic dystrophy during the last decade.

The book consists of 14 chapters. The first chapter introduces the reader to the earliest descriptions of the disorder and reviews the significant historical contributions to the understanding of myotonic dystrophy. It also summarizes the advances that have taken place in recent years. Chapters 2 through 9 describe the neuromuscular manifestations in considerable detail, including the differential diagnoses and other disorders associated with myotonia. Myotonic dystrophy involves many organ systems, in addition to neuromuscular involvement. Cardiovascular, respiratory, endocrine, and systemic abnormalities, eye findings, and changes in the brain are all clearly described in detail. Chapter 9 deals with myotonic dystrophy in infancy and childhood. The next chapter deals with diagnostic tests, including electrophysiological studies and muscle pathologic examinations, and also describes experimental myotonia.

When the last edition was published in 1989, the concept of unstable DNA and trinucleotide repeats was unknown. Chapters 11 and 12 deal with the genetic basis and molecular and cell biology of myotonic dystrophy. The chapters are written in such a way that even readers who are not well versed in molecular genetics will find it easy to understand. The next chapter presents genetic counseling by using applied molecular genetics. The final chapter deals with management and therapy of myotonic dystrophy.

Myotonic dystrophy has several systemic manifestations in addition to the neuromuscular involvement and is therefore of interest not only to the neurologist but the cardiologist, pulmonologist, pediatrician, and ophthalmologist. A chapter is devoted to nervous system involvement with images of the brain. Although many specialists may find this book to be invaluable, it may be of only limited relevance to the neuroradiologist.

The book is well organized, the style of writing is very pleasing, and all aspects of myotonic dystrophy are covered in considerable detail. The book is easy to read, and even patients and their family members will find it to be of value. The references are up-to-date and exhaustive.

Clinicians and researchers who deal with myotonic dystrophy should keep this volume in their personal library. It is an excellent source of information on all aspects of myotonic dystrophy.