Electromyography in Clinical Practice

Electrodiagnostic Aspects of Neuromuscular Disease

Second Edition

Michael J. Aminoff

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Neuromuscular Disease

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Preface to Second Edition

Some 10 years have passed since I completed the manuscript of the first edition of this book, and these years have seen remarkable advances in the electrodiagnostic approaches used to study disorders of muscle and the peripheral nervous system. First, a more critical approach to the interpretation of the results of conventional studies has led clinicians to place greater reliance on electrophysiologic findings. Second, the development of new electrophysiologic techniques and the greater application of existing techniques to the study of neuromuscular disorders has broadened the scope of this form of investigation. In particular, the use of blink reflexes, H-reflexes, F-responses, and somatosensory evoked potentials has been helpful both for diagnostic and prognostic purposes and for providing new insights into the pathophysiologic basis of various disorders. Third, the increasing use of computers has made possible the use in clinical practice of techniques that were once of purely academic interest, and the development of techniques for studying conduction in different subpopulations of nerve fibers in individual nerve trunks now promises to become of major clinical significance. Fourth, the application of both new and more conventional techniques to the evaluation of patients with disorders of the central nervous system—an approach sometimes referred to as central EMG—has provided new means of analyzing these disorders.

These developments have made necessary this second edition, in which I have made numerous additions to the text and completely rewritten some sections. Inevitably, the book has become larger. However, I have kept very much in mind the primary purpose of the book as outlined in the preface to the first edition, namely, to provide a concise and practical account of the subject rather than a comprehensive account of all its aspects, which is now anyway available from larger, more encyclopedic texts. Techniques used solely for research have deliberately not been considered.

I am grateful to Dr. R.K. Olney for reading a number of the chapters and offering helpful suggestions and comments on the text, and to Ms. Susan Shaddick for her good-humored secretarial assistance. Furthermore, the encouragement and understanding of my wife, Jan, and our three children made it possible for me to find the time to bring this book to its conclusion. Finally, I am grateful to Churchill Livingstone for undertaking the publication of this second edition, and in particular to Mr. Robert Hurley and his staff for their help and advice during its production.

Preface to First Edition

Although the term electromyography refers strictly to methods used to record the electrical activity of muscle, it has come to have a wider meaning which encompasses also the electrodiagnostic techniques used to study the functional integrity of peripheral nerves and the neuromuscular junction. In recent years, the application of refined neurophysiologic techniques to the study of disorders of muscle and the peripheral nervous system has proved to be of considerable diagnostic value to the physician; this, in turn, has stimulated the further develour and expansion of this branch of clinical neurophysiology into a separate specialty of its own. Perhaps because of its rapid advance, however, many clinicians are not fully aware of the scope and limitations of the investigative procedures in current use, and are uncertain about the interpretation and emphasis to be placed on the information obtained from them in relation to individual clinical problems. At the same time, an increasing number of those responsible for undertaking these procedures, or training in this field, have felt the need for a convenient summary of the physiologic basis of the specialty and the accumulated clinical experience of recent years, and for a resumé of those techniques which are most useful in practice.

Accordingly, the purpose of this book is to review the manner in which electromyography may be of value in the investigation of patients with neuromuscular disorders, and to provide a concise, practical guide to those procedures in current clinical use. The book contains an account of the fundamental physiologic principles on which this form of investigation depends, and describes the equipment and methods used in its practice. It also discusses the significance of observations made at electromyography and reviews the findings in various clinical disorders. It is not intended, however, to provide a comprehensive account of all aspects of electromyography, and those techniques developed and used solely for purposes of research are not considered.

The book is aimed at medical practioners concerned with the management of patients with neuromuscular disorders, and at clinical neurophysiologists. Sufficient clinical details are provided to enable those readers without formal neurologic training to grasp the nature of the diagnostic problems confronting the clinician, and the manner in which electromyography can contribute to their solution. Pertinent anatomic details are included in the text, but this book is written primarily for those who already have some knowledge of human anatomy.

The book was written partly in California, and partly in England while I was senior registrar at The National Hospitals for Nervous Diseases. I am grateful to a number of friends and colleagues in London and San Francisco who helped in its preparation, and particularly to Dr. P.C. Gautier-Smith, Dr. M.J.G. Harrison,

and Dr. R.A. Jaffe for their advice, to Dr. J.B. Pickett for providing some of the material used for illustrations, and to Miss Beryl Laatz for her invaluable secretarial assistance. Finally, I wish to thank my wife, Jan, for her encouragement and help, and the staff of the Addison-Wesley Publishing Company for their patient cooperation and advice during the production of this book.

Michael J. Aminoff, M.D.

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Electrodiagnostic Methods for the Study of Nerve and Muscle

Electrophysiologic methods for studying the function of nerve and muscle were first introduced in the nineteenth century, but only within the last 30 to 40 years have they come to be used widely in a clinical context. Their widespread clinical application came about because of improvements in apparatus design that reduced the practical difficulties of using electrophysiologic methods and permitted the introduction of more refined techniques, thereby expanding considerably the scope of this form of investigation.

The accumulated experience of recent years has shown that electrophysiologic methods have a definite place in the investigation and diagnosis of certain categories of clinical disorder, as is discussed in general terms in the latter part of this chapter. In addition, these methods have enabled the physiologic characteristics of muscle and the peripheral nervous system to be studied in health and disease, and have provided insight into the pathophysiologic basis of many neurologic symptoms. Such studies have also led to both a re-examination of traditional concepts which, until recently, were accepted without question and, in

some instances, to the more precise definition of neuromuscular disorders.

The application of electrophysiologic techniques to the study of neuromuscular disorders has also contributed to a general resurgence of interest in these conditions. In recent years, considerable new information has been accumulated regarding the ultrastructural, histochemical, biochemical, pharmacologic, and physiologic aspects of these disorders, and attempts have been made to integrate these data with observations made by clinical neurophysiologists. A multidisciplinary approach of this sort offers the greatest hope for ultimately finding a solution to fundamental problems concerning the development, prevention, and treatment of neuromuscular disorders.

CLINICAL APPLICATIONS

The electrophysiologic examination of patients with suspected neuromuscular disorders is no more than an extension of the clinical examination. As such, it should be performed by a physician who is experi-

enced in the clinical evaluation of neuromuscular disorders, familiar with the anatomic intricacies of the peripheral nervous system, and appreciative of the scope and limitations of the methods in use. Because the procedure is more than a laboratory test, it does not provide a simple, unqualified answer to the clinical problem that has prompted referral, but instead yields information that must then be integrated with that already available.

Patients are referred to the laboratory for electrophysiologic evaluation for a variety of reasons. Some patients who are referred have clinical problems or questions that cannot fully be assessed or answered by the electrophysiologic techniques in current use. In many instances, referral is merely a means of obtaining a second clinical opinion when there is uncertainty about the diagnosis or management of a disorder. In other cases, a referral is made to obtain information that may help to establish the diagnosis or provide a guide to prognosis. to serve as a baseline or a means of comparison in following the course of a disorder, or to provide supportive evidence in a medicolegal context. It is important for the clinical neurophysiologist to determine the reason for each patient's referral if a satisfactory service is to be provided. An adequate electrophysiologic examination cannot even be planned unless the clinical history and physical findings are known. Unlike a simple laboratory test, the procedure for electrophysiologic examination does not follow a rigid, predetermined protocol, but is modified according to the clinical context in which it is requested, as well as by the initial findings that are obtained.

Electromyography

Electromyography, in the strict sense of the word, refers to methods of studying the electrical activity of muscle. It had little direct relevance to clinical neurology until the introduction of the concentric needle electrode in 1929.¹ By inserting this electrode directly into a muscle, electrical activity within circumscribed regions of that muscle and, in particular, the electrical events accompanying the activation of muscle fibers and single motor units, could be recorded. The pattern of electrical activity in normal muscle thus came to be characterized, and this permitted departures from normal activity to be recognized and correlated with the presence of disorders of the motor unit. Electromyography is discussed in detail in later chapters, but it is appropriate to consider here its clinical usefulness.

The term motor unit, first used in 1925 by Liddell and Sherrington,2 describes a single lower motor neuron and the muscle fibers that it innervates. The term therefore encompasses the cell body of the lower motor neuron in the spinal cord; its axon, which emerges from the cord to become a constituent fiber of a nerve root and, ultimately, of a peripheral nerve; its terminal arborizations and the neuromuscular junetions with which they are associated, and all of the muscle fibers that are innervated by it. The concept of the motor unit is a physiologic one because the activity of a muscle cell cannot properly be considered without reference to the nerve cell governing it.

The function of the normal motor unit can be disturbed at several sites. A lesion affecting either the cell body of a lower motor neuron or its axon may prevent the activation of the muscle fibers of that unit. Alternatively, the function of a motor unit may be disturbed because of some disorder affecting its constituent muscle fibers. Each of these conditions may give rise to characteristic changes in the electrical activity recorded from the affected muscle. Electromvography is often helpful, therefore, in detecting disorders of motor units and in determining the site of the underlying lesion. Furthermore, if electrical evidence of a neurogenic lesion is present, the pattern of affected muscles and other electrophys-

iologic findings may indicate whether it is at the level of the root, plexus, or peripheral nerve.

As emphasized by Lambert,3 such information may sometimes be obtained by electromyography when it cannot be obtained by clinical examination, either because the disease is only mildly advanced, because the presence of emotional overlay or of other symptoms and signs (such as pain or deformity) makes clinical evaluation difficult, or because clinical signs, such as weakness in individual muscles, are obscured by the compensatory action of unaffected, synergistic muscles. It must again be stressed, however, that electromyography is not a substitute for detailed clinical examination, but is complementary to it. Indeed, an adequate electromyographic study cannot be performed unless the clinical background of the patient and the diagnostic problems facing the clinician are known. The patient's history and physical signs govern which muscles are examined first, whereas the related diagnostic problem determines which other muscles are subsequently examined.

In some patients with weakness that is predominantly upper motor neuron in type but of uncertain etiology, the presence of a coexisting lower motor neuron deficit affects the diagnostic possibilities. Similarly, in patients with clinical evidence of a focal lower motor neuron deficit, the presence of more widespread denervation (even though subclinical) may profoundly influence the diagnostic considerations. In such circumstances, then, the findings obtained by the needle examination may be of special significance.

Electromyographic findings do not, in themselves, provide a clinical diagnosis, for there are no potentials that are pathognomonic of specific diseases.3 The pattern of activity detected by electromyography does, however, provide a guide to the site and, in some instances, to the nature of the lesion. In arriving at a final diagnosis, the information so obtained should be considered in relation to the other available clinical and laboratory data. Conclusions based solely upon electromyographic findings must never be relied upon to the extent that clinical judgment is neglected.

Information derived from electromyography may also provide a guide to prognosis. For example, in patients with Bell's palsy, electromyographic evidence of denervation of the facial muscles indicates a less favorable prognosis than when denervation has not occurred. Similarly, when used to determine whether denervation is partial or complete, electromyographic sampling may reveal surviving motor units in a muscle that appears clinically to be totally inactive, 4 which would be of prognostic significance. Indeed, the findings and results of nerve stimulation may govern whether a peripheral nerve is explored after injury.

Electrical Stimulation of Peripheral Nerve

THE MUSCLE RESPONSE EVOKED BY NERVE STIMULATION

In addition to recording the electrical activity of muscle at rest or under voluntary control, the response of a muscle to supramaximal electrical stimulation of its motor nerve can be studied with surface electrodes. The size of the electrical response evoked in this way normally varies widely in different individuals and in different muscles, and depends on many factors, including the precise recording arrangements. Within these limitations, however, size of the electrical response correlates with the number of muscle fibers that can be activated by excitation of the nerve. Thus, an abnormally small response indicates loss or disease of motor units, although a response of amplitude within the normal range does not exclude such pathology. Accordingly,

this procedure may provide evidence of motor unit pathology—an evoked response that is abnormally small—in patients with symptoms that might otherwise be attributed to nonorganic causes or to disease of upper motor neurons.⁵ In such circumstances, needle electromyography is necessary to determine whether the underlying pathology is in the muscle itself or in its nerve supply.

Examination of the electrical response of a muscle to supramaximal stimulation of its motor nerve is also useful in defining the site of a focal lesion of the nerve, in determining whether axonal degeneration has occurred, in studying the completeness of the lesion after a peripheral nerve injury, and in detecting anomalous patterns of muscle innervation in the extremities.3 Furthermore, when repetitive stimuli are used, changes in the size of the evoked response may enable disorders of transmission at the neuromuscular junction to be detected and classified. Such disorders include myasthenia gravis, Lambert-Eaton syndrome, and botulism.

NERVE CONDUCTION STUDIES

By recording the electrical response of a muscle to stimulation of its motor nerve at two points along its course, conduction velocity in the fastest conducting motor fibers along the intervening segment of the nerve can be calculated. This is of considerable value in distinguishing between disorders of the peripheral nerves and those affecting primarily either muscle or the cell bodies of the lower motor neurons. Similarly, the conduction velocity and amplitude of the action potential in sensory nerves can be measured by stimulating sensory fibers at one point and recording from them at another. Such a procedure is helpful in determining whether sensory symptoms are attributable to a disturbance of peripheral

nerve function or to a lesion proximal to the dorsal root ganglia.

Nerve conduction studies thus provide a means of demonstrating the presence and extent of a peripheral neuropathy, and are particularly helpful when clinical examination is difficult, such as when evaluating young children. These studies are useful in a number of different contexts. First, they may be used to determine whether patients with a mononeuropathy have an underlying, subclinical polyneuropathy. Second, in patients who do have a mononeuropathy, the site of the focal lesion may be identified. Third, nerve conduction studies, in combination with needle electromyography, are an important means of determining the severity of a focal nerve lesion, and thus may guide management. Fourth, they provide a means of distinguishing between a polyneuropathy and mononeuritis multiplex when the two are clinically indistinguishable. Fifth, in patients with an established polyneuropathy, they may be used to determine the extent to which the disability relates to the superadded compressive focal neuropathies that are likely to occur in this context and that are potentially reversible. Sixth, nerve conduction studies provide a means of following the course of peripheral nerve disorders and of determining a patient's prognosis. The findings, especially when considered in conjunction with the results of needle examination, may indicate the extent of axonal loss and the presence or absence of reinnervation. Finally, nerve conduction studies may provide a clue to the underlying pathology. In recent years, studies on human neuropathies and on experimental neuropathies in animals have revealed that the predominant pathologic change may be either axonal degeneration or segmental demyelination, depending on the etiology of the disorder. A distinction between these two pathologic processes may not be possible on clinical grounds. However, conduction velocity is usually markedly reduced in primary demyelinative

neuropathies, whereas it is either unaffected or only mildly reduced in axonal neuropathies. Other electrophysiologic distinguishing features include the occurrence of dispersed muscle and nerve action potentials, areas of focal conduction block, and markedly prolonged terminal latencies in demyelinative neuropathies. By contrast, in axonal disorders, the muscle response to stimulation of motor nerves and the action potentials recorded from sensory fibers may be unrecordable or markedly reduced in amplitude, and needle examination of affected muscles reveals evidence of denervation. On the basis of the electrophysiologic changes, therefore, it is frequently possible to predict the underlying pathology, thereby reducing the number of possible etiologies to be considered, without resorting to biopsy.

The measurement of conduction velocity has been used in genetic and epidemiologic studies as a means of detecting hereditary disorders of the peripheral nerves prior to the development of clinical signs, and as a means of detecting heterogeneity in clinically similar cases. For example, it has been shown that patients with peroneal muscular atrophy may have either a markedly reduced or a relatively normal conduction velocity in motor nerves. This distinction is probably genetically determined since values for conduction velocity are consistent within families.7-9

The development of techniques for evaluating nerve conduction has thus added a new dimension to the study of disorders of the peripheral nervous system. Practical details of the techniques concerned, as well as their application to specific clinical problems, are discussed in detail in later chap-

H REFLEX

The H reflex is a monosynaptic reflex that is easily recorded from the soleus muscle in adults in response to low-intensity stim-

ulation of the tibial nerve. As discussed in later chapters, the response may be delayed or lost in a number of peripheral neuropathies, even though the results of conventional motor and sensory conduction studies are normal. 10 The response may also be abnormal in patients with S1 radiculopathies. Although H reflexes have been used for some years to study motor neuron excitability in different contexts, this use has no immediate clinical application and will be considered only briefly.

F RESPONSE

The F response may be useful in the evaluation of patients with peripheral neuropathies, radiculopathies, or plexopathies. It is sometimes abnormal even when conventional nerve conduction studies are normal, and it is especially useful in assessing the function of those proximal portions of the peripheral nervous system, such as the roots or plexuses, that are not easily evaluated by conventional techniques because of their inaccessibility. Unlike the H reflex. F responses can be recorded without difficulty from most skeletal muscles by stimulation of their motor nerves. These responses occur as a result of the discharge of a few anterior horn cells by antidromic impulses reaching the cell bodies along their axons from the site of nerve stimulation.

BLINK REFLEX

The blink reflex, which is well known to clinical neurologists, can also be elicited by an electrical stimulus to a branch of the trigeminal nerve, and the response of the orbicularis oculi muscles on each side may be recorded on an oscilloscope. This technique has been used, especially by Kimura, 11 to monitor function of the trigeminal and facial nerves when an isolated lesion of these nerves is suspected, or in patients with polyneuropathies. It has also been used to provide evidence of pontine dysfunction when this is not clinically evident, as in patients with suspected multiple sclerosis. The blink reflex has been studied in a variety of other contexts, but the findings have then been notable more for their academic interest than for any immediate clinical relevance.

SOMATOSENSORY EVOKED POTENTIALS

Electrical stimulation of an accessible sensory or mixed peripheral nerve elicits potentials that can be recorded from selected locations over the scalp and spine, as well as peripherally. These somatosensory evoked potentials have been used to identify and localize lesions involving the somatosensory pathways, especially those segments that traverse the central nervous system. Their major application has been in the detection of subclinical lesions in patients with suspected multiple sclerosis. Their utility in the evaluation of spinal injuries, in predicting the outcome of posttraumatic coma, and in monitoring patients undergoing spinal surgery is less clear. 12 These potentials have also been used to evaluate lesions of the peripheral nervous system, especially when inaccessible proximal portions of peripheral nerves or a limb plexus are involved.

Electrical Stimulation of Muscle

There is a difference in the excitability response of innervated and denervated muscle to electrical stimulation. This difference forms the basis of a simple test for determining whether or not a particular muscle is denervated and, if the test is repeated at intervals, whether the extent of denervation is increasing or reinnervation is occurring. The most satisfactory form of

this test involves determining the relationship between the intensity and the duration of current necessary to produce a minimal contraction of the muscle, with the duration being shortened incrementally. Strengthduration curves were first applied in a clinical context by Adrian. 13 Their use has declined with the development of needle electromyography and methods for measuring nerve conduction velocity, and in many laboratories they are no longer undertaken. Although they provide no clue to the site of the underlying lesion, they can nevertheless be used to follow the course of a disorder and to determine whether or not it is progressing.

OTHER APPLICATIONS

The preceding discussion emphasizes the clinical value of electrophysiologic methods for studying the neuromuscular system. These methods have also been used in a wider context to examine the activity of individual muscles in the maintenance of posture and during normal movement. Such studies have provided information of considerable academic interest and have advanced knowledge of normal physiologic mechanisms. They are not, however, of immediate clinical relevance, and so are bevond the scope of this book. The application of similar methods to the analysis of various movement disorders and their underlying pathophysiology is of unquestionable clinical utility, and electrophysiologic techniques are also being widely used to study the functional integrity of the autonomic nervous system. These applications are, therefore, reviewed briefly in Chapter 15.

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Aspects of Nerve and Muscle Physiology

Modern electrodiagnostic techniques involve recording the electrical activity of striated muscle and peripheral nerves. It is of some importance, therefore, to appreciate the physiologic basis of such activity, which is discussed in this chapter. The physiology of muscle and the peripheral nervous system is also considered in relation to certain other factors that have direct relevance for those undertaking electrodiagnostic procedures in clinical practice. However, in view of the particular importance of the functional organization of the motor unit to the interpretation of the electromyogram, this is discussed separately in the next chapter. Throughout this chapter and the next, deliberate emphasis is placed not only on current physiologic concepts, but also on the experimental observations that have led to their development.

THE ELECTRICAL ACTIVITY OF NERVE AND MUSCLE

The nature and ionic basis of the electrical activity of striated muscle fibers are qualitatively similar to that of nerve cells. The following account, therefore, refers to both types of cell.

The Resting Membrane Potential

relevant to any discussion of membrane no-

The inside of a muscle or nerve cell is electrically negative with respect to its exterior. This potential difference across the cell membrane is called the resting membrane potential, and is on the order of -90 mV for striated muscle cells and -70 mV for lower motor neurons, the sign indicating the polarity of the inside of the cell relative to its exterior. The resting membrane potential primarily reflects differences in ionic concentration that exist across the cell membrane and the selective permeability of the cell membrane.

The cell membrane separates two aqueous solutions—intracellular and interstitial fluids—each of which contains ions, particles that carry an electrical charge. The intracellular and interstitial fluids are in osmotic equilibrium with each other, and in each the total number of anions and cations is equal, so that both are electrically neutral. However, the distribution of ions between these two solutions is unequal. The intracellular concentration of potassium is relatively high, and the concentration of sodium and chloride is relatively low compared with the concentration of these ions

outside the cell. These inequalities in ionic concentration are created and maintained by a transport mechanism requiring the expenditure of energy by the cell, and are related also to the varying degree of permeability that the cell membrane has for different ionic species.

In discussing the manner in which these differences in ionic concentration lead to a potential difference across the cell membrane, the effect of the active transport mechanism will initially be neglected for the sake of simplicity. There are, however, two important passive forces-forces not requiring the expenditure of energy—that affect the movement of ions, and these are relevant to any discussion of membrane potentials. First, particles unequally distributed in a solution will tend to move by diffusion from areas of high concentration to areas of low concentration, moving down their concentration gradients until these are abolished. Second, the movement of ions will be influenced by voltage gradients, with positive ions being attracted down the electrical gradient toward the cathode, and negative ions moving similarly toward the anode.

It was originally proposed by Bernstein that only one species of ion—potassium could diffuse across the resting cell membrane. If this were the case, potassium ions would tend to move out of the cell and down their concentration gradient, thereby producing an excess of anions within the cell and of cations outside of it. These excess ions of opposite polarity would attract each other across the cell membrane, collecting in the vicinity of the membrane. This would then set up an electrical gradient across the membrane, opposing further outward movement of potassium ions. Eventually, this gradient would become large enough to cause an influx of potassium ions in a number just sufficient to counteract their efflux down the concentration gradient. The electrical potential difference needed to counteract completely the tendency of potassium ions to diffuse out of the cell along their concentration gradient—that is, to maintain in a steady state the unequal distribution of potassium ions on either side of the membrane—is called the equilibrium potential for potassium. Its magnitude at 37°C can be calculated from the Nernst equation, a convenient form of which is

$$E_k = 61 \log \frac{[K]_0}{[K]_1}$$

where E_k is the equilibrium potential for potassium, expressed in millivolts, and $[K]_O$ and $[K]_I$ are the potassium concentrations outside and inside the cell, respectively. It should be noted that the Nernst equation can actually be applied to any permeant ionic species unequally distributed across the cell membrane.

Bernstein's theory provided an explanation for the existence and polarity of the resting membrane potential, and for the changes in potential that appeared to occur when a nerve or muscle cell is excited. However, experimental studies subsequently provided data that could not easily be reconciled with his theory. The magnitude of the resting membrane potential was found to differ from that predicted by the Nernst equation for the unequal potassium distribution. Furthermore, it was discovered that the cell membrane is permeable to other ions, including sodium and chloride, and that excitation of the cell led not to abolition of the resting membrane potential, as was originally supposed, but to its reversal.

The demonstration, with radioisotopes, that the cell membrane is permeable to sodium, and that this ion is in a continuous state of flux across it, was of particular significance in the development of present concepts concerning the origin of the resting membrane potential. Passive electrochemical factors alone could not account for the efflux of sodium ions from the cell because this efflux must occur against both the concentration and the electrical gra-