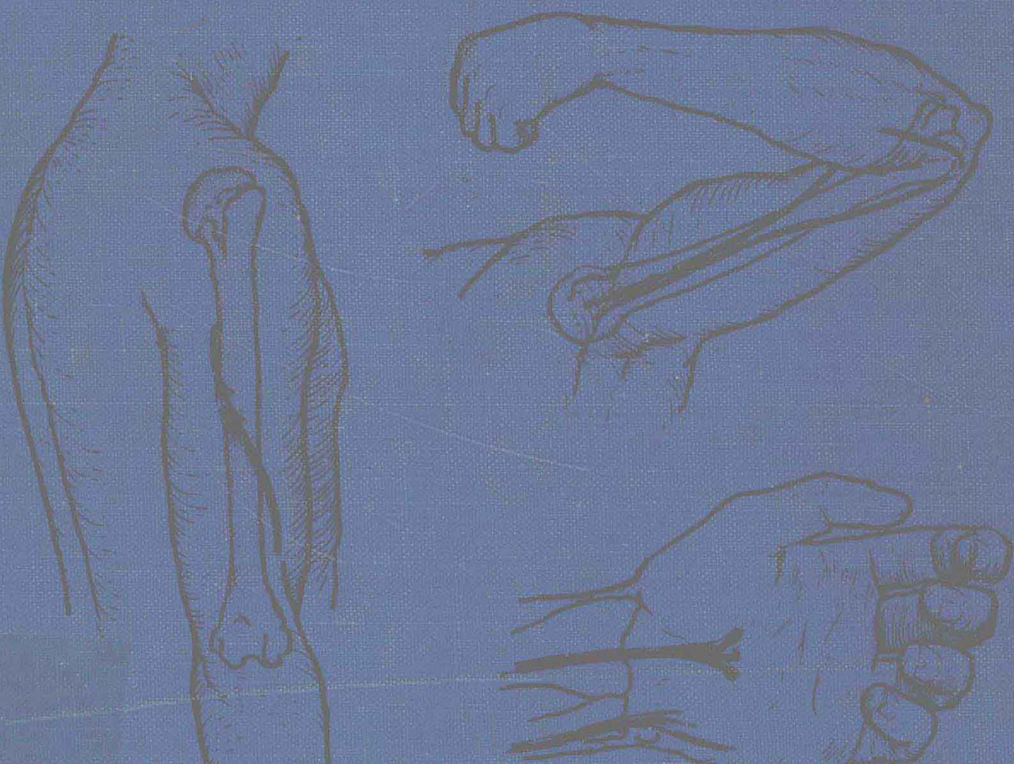
ELECTRODIAGNOSIS OF NEUROMUSCULAR DISEASES

2nd Edition

JOSEPH GOODGOLD, M.D.
ARTHUR EBERSTEIN, PH.D.



Electrodiagnosis of Neuromuscular Diseases, 2nd Edition

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Preface to the Second Edition

The primary objectives which guided the preparation of this second edition were, firstly, to improve the exposition of the material included in the first edition and, secondly, to incorporate new information and innovations relating to the practice of electrodiagnosis and neuromuscular physiology as well. Accordingly, revisions were made throughout the text. In particular, the chapter on motor and sensory nerve conduction measurements was substantially rewritten and illustrated with new drawings to provide a clear understanding of the various peripheral nerve lesions. In a similar manner, new material has been included in almost every chapter. Single fiber electromyography, the estimation of the number of motor units, computer analysis, and the latest explanation of muscle contraction are some of the topics particularly relevant to electromyography discussed in detail in this edition. A new chapter was also added, with extensive coverage of the blink reflex, the "F" loop, the H reflex, and the tendon vibration reflex.

The basic purpose of this book has not changed. This edition, as the first, presents a comprehensive and critical introduction to the practice of electromyography and nerve conduction studies at a level suitable for the serious student as well as the more advanced practitioner. Current knowledge regarding basic concepts and a rigorous review of modern techniques were intentionally integrated in one text. This approach enhances the acumen of the practitioner as well as demonstrates the logical reasoning and ultimate interpretation of the findings for the other interested physicians.

The expansion of this edition has made it necessary to add new contributors. We are deeply indebted to Bhagwan T. Shahani, M.D. and Robert R. Young, M.D. from the Department of Neurology, Harvard Medical School, Boston, Massachusetts, for the excellent discussion on the blink reflex. Again we would like to thank Goodwin M. Breinin, M.D., Chairman and Professor of the Department of Ophthalmology, for his contribution on Ocular Electromyography as well as Clark T. Randt, M.D., Professor and Chairman of the Department of Neurology, and Joseph Ransohoff, M.D., Professor and Chairman of the Department of Neurosurgery for their generous advice and assistance in the preparation of this text. We express our deep appreciation to Howard A. Rusk, M.D., Professor and Chairman of the Department of Rehabilitation Medicine, for his wise counsel and encouragement throughout the writing of both editions.

JG
AE
1977

Preface to the First Edition

Clinical neurophysiology is a relatively new area of specialization concerned with the diagnostic evaluation of neuromuscular diseases. The governing principles of the specialty are derived from applied electrophysiology, and the operational tools include sophisticated bioelectric, biomechanical, and computer instrument systems. If electroencephalographic laboratories are excluded (actually they are independent operations in most institutions), the number of full time departments of clinical neurophysiology in the United States or, for that matter, in the world, is still rather small.

When the specific laboratory tests which are carried out are considered, it is apparent that a certain degree of semantic clumsiness has involved their description. Why it has ever been implied that electromyography is nosologically distinct from electrodiagnosis and why electromyography and conduction studies should not be included under such a basic generic term as electrodiagnosis seems to be as illogical as the need for invention of new nomenclature, i.e., electroneuromyography. Basically we are dealing with the subject "Electrodiagnosis of Neuromuscular Diseases," which could be served with just a few such major subdivisions: (a) response of nerves and muscles to electrical stimulation, (b) recording of bioelectric potentials from nerves and muscles, and (c) miscellaneous studies, such as skin resistence measurements.

What is the definitive core of knowledge required of the specialist in clinical neurophysiology? It is obvious that the discipline is a hybrid with simultaneous overlap into many of the basic sciences and clinical subjects.

It seems equally obvious that medical training is an essential background if the implications of "clinical" are to be meaningful. Perfect comprehension and complete mastering of the technical aspects of the field may be attainable by exceptionally gifted individuals without such strong medical orientation, but there is an ultimate, albeit disguised, inadequacy because of the lack of background information required to view the patient and his illnesses in an integrated perspective. Just such a single simple bit of information such as that which relates carpal tunnel disease to hypothyroidism is intuitively considered by the well trained physician, whereas it may be, and usually is, beyond the informational resources of ancillary personnel. The exception is indeed an extremely rare, unusually motivated individualist.

The specific informational pillars which form the strong base upon which the specialty is built include at least: *Internal Medicine; Clinical Neurology;*

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Anatomy, Pathology, and Physiology of the Neuromuscular System; and *Electronics*.

These subjects do not uniquely fall within the absolute domain of any single medical specialty—neurology, neurosurgery, rehabilitation medicine, or orthopedics. Under normal circumstances, completion of formal training in any single one of these disciplines provides no more than one or two bits of the set of required knowledge of the clinical neurophysiologists. Successful completion of the set is keyed to a period of full time training in an established (and busy!) department which has been organized to accomplish this educational mission. With due regard to a candidate's background, at least 12 to 18 months of what really amounts to preceptorship seems to be minimal and essential. The American Association of Electromyography and Electrodiagnosis, in facing this issue, has recently organized an educational committee chaired by one of the authors (J. G.), and charged it with (a) formulation of the essentials of a training program for clinical neurophysiologists and (b) investigation of the feasibility of some process of certification as specialists. Whatever the outcome of the Association's study, it is apparent that although the specialty is young and changeable it is, however, already firmly established beyond the point when part time and piecemeal training can provide individuals who are capable of fulfilling the sensitive mission of consultation services to clinical and surgical colleagues.

The paucity of published texts in the field attest to the youthfulness of the specialty, whereas the radical changes and revisions of published opinions and "facts" throughout the years since the early 1940's attest to the vitality and progressive maturation of the field. This book has been conceived to meet the need for a comprehensive, critical, and modern introduction to basic concepts and to provide current information regarding neurophysiological evaluation of disorders of skeletal muscles and peripheral nerves. Discussion of some of the older methods of examination (chronaxie, etc.) have been intentionally minimized.

We have set about to fulfill these requirements by presenting the basic principles underlying electromyography and nerve stimulation studies in a form suitable for the physician planning to specialize as well as for the main group of physicians who rely on the results to augment clinical diagnosis—the neurosurgeon, neurologist, orthopedist, physiatrist and others. In this sense, the text furnishes a rigorous introduction which is neither elementary nor specialized. No prior training in electrodiagnostic technique is assumed. However, this is not a simple manual delineating methods of procedure. Instead, the subject is carefully developed from a fundamental level so that the reader may fully understand the logical reasoning and acumen behind the procedures carried out and arrive at an ultimate interpretation which has clinical significance.

A functional approach to the formulation of the concepts which are presented has been adopted throughout this book. The basis of the electrical activity recorded from muscles or nerves and a complete discussion of volume conduction are introduced in the first section, followed by a review of the instrumentation system necessary to perform the electrodiagnostic studies. This area is covered in a simple, descriptive manner and provides the reader with an understanding sufficient to select, utilize, and realize the limitations of the apparatus. After these instructional chapters, the technique and concepts of electromyographic examination and nerve conduction studies are developed. The order for presentations we have found best over the years of teaching these subjects proceeds from a discussion of normal to the findings in myopathy and neuropathy. Examples of cases which are unusually instructive have been included. Pitfalls and errors of procedure and interpretation have been presented and discussed for various abnormalities.

Important facts of neuroanatomy, pathology, neurology, or internal medicine are briefly reviewed whenever necessary to present a clear picture of the abnormal state and to define the purpose of the various testing procedures. The section on root compression lesions demonstrates how helpful this type of review may be to the clinician. If the electromyographer, for example, is not aware of the implications of lateral vs. medial herniation of an intervertebral disc, his value as an essential member of the diagnostic team is considerably weakened.

Perhaps it is regrettable that the Herculean task of an all-encompassing, exhaustive treatise has not been the limit set by the authors, but even if it were, it could not substitute for the prescribed educational process.

The authors express their deep appreciation for the wise counsel of numerous colleagues at the medical center, especially to Goodwin M. Breinin, M.D., Chairman and Professor of the Department of Ophthalmology for his invaluable contribution on Ocular Electromyography (Chapter 12) and to Clark T. Randt, M.D., Professor and Chairman of the Department of Neurology, and Joseph Ransohoff, M.D., Professor and Chairman of the Department of Neurosurgery, who have given generously of their time, experience, and wisdom, both philosophically and practically, in the preparation of this text.

Special thanks goes to Howard A. Rusk, M.D., Professor and Chairman of the Department of Rehabilitation Medicine whose foresight and encouragement fostered the organization of a full time Electrodiagnostic Department long before the highly specialized nature of this field was generally recognized as a needed and specific discipline.

It is our hope that our experiences detailed in this book will be helpful to others who are directing or organizing similar programs dedicated to clinical service, training and research.

The authors also acknowledge with gratitude the able and dedicated assistance of their scientific and secretarial colleagues.

It seems most befitting that a prologue such as this one conclude with a dedication of the book. This is especially true when it is to the two persons

Mildred Sylvia Goodgold

and

Marion Eberstein

whose understanding, devotion, and encouragement were ever present, from conception to completion of the manuscript.

JG
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1971

Contents

chapter 1

Anatomy of Nerve and Muscle— A Review

The anatomic system of primary interest in clinical electrodiagnosis consists essentially of the peripheral nerves, the myoneural junctions, and the skeletal muscles. During normal behavior, these three components interact with each other to bring about the contraction and relaxation of a muscle. Electromyography and nerve conduction measurements may be used to determine abnormalities occurring in the three subdivisions; however, the interpretation of the findings depends on a thorough understanding of basic neuromuscular anatomy and physiology.

THE FUNCTIONAL NERVOUS SYSTEM

The motor nerve fibers which innervate striated voluntary muscles except those in the head are axons of cells in the anterior gray matter of the spinal cord (Fig. 1.1). Those fibers which supply the head, such as the muscles of mastication, facial expression, and eye movement, emerge from the brain stem in close association with certain cranial nerves. In either case they are considered peripheral nerves because the peripheral nervous system is defined to include all of the nerves and associated ganglia.

Besides functioning as a receptor for nerve impulses, the muscles (as well as the tendons) contain sensory organs which serve as a *source* of nerve impulses. These proprioceptive receptors, spindles in the skeletal muscles and Golgi organs on the tendinous ends of the muscles, detect tension changes in the muscles or tendons and then, by way of the connecting sensory nerve fibers, send the information back to the central

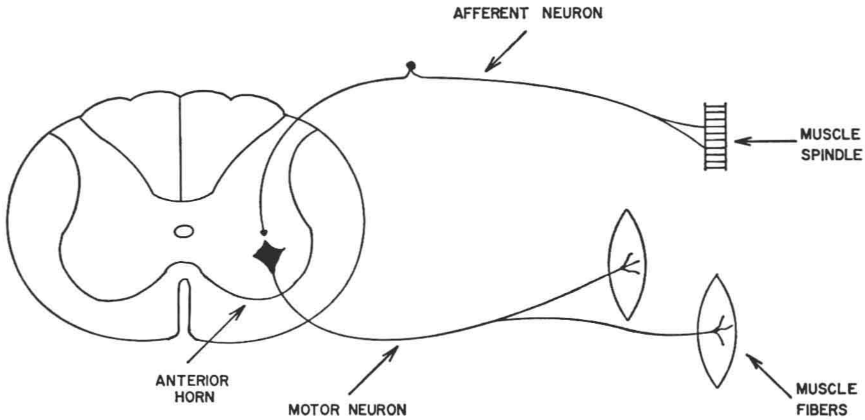

FIG. 1.1. Diagram showing innervation of skeletal muscle fibers by a motor neuron. Impulses are conducted away from the central nervous system in the motor neuron and toward the central nervous system in the afferent neuron.

nervous system. These sensory nerve fibers are, of course, also part of the peripheral nervous system.

The junction between the terminal branch of the nerve fiber and the muscle fiber is located at the midpoint of the muscle fiber and is called the motor end-plate (Fig. 1.2). Each terminal axon generally contributes to the formation of a single end-plate innervating one muscle fiber.

However, Coers (1, 2) showed 2.3% of the limb muscle fibers have double end-plates, and that these end-plates always come from the same nerve fiber. Coers and Woolf (5), in their extensive investigation of biopsy specimens from normal muscles, state that they never observed in human limb muscles the innervation of a single muscle fiber by two different axons. The only muscle fibers in man found to have multiple end-plates (i.e., more than two) are located in the extraocular muscles (3, 4).

Cholinesterase staining demonstrates the presence of two kinds of nerve endings: (a) large, heavily staining compact discs which are typical motor end-plates or *en plaque* endings and (b) smaller, lighter staining droplets arranged in clusters or chains along the single muscle fiber, which are classified as *en grappe* endings (4). It has not been established as yet whether the multiple junctions on one muscle fiber are derived from one neuron or from several neurons.

Myoneural junctions are not spread all over the muscle but are usually concentrated in confined zones. In the majority of muscles there is only one zone of innervation, the shape of which depends on the form and pattern of insertion of the muscle fibers on the tendon. For example, in muscles in which the fibers lie parallel to each other from one end to the other as in the soleus or peroneus brevis, the innervation zone runs in a line

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across the center and perpendicular to the muscle fibers (Fig. 1.3). In the pennate muscles, like the flexor carpi radialis or palmaris longus, the line of innervation is curved as it passes through the midportion of the muscle fibers (Fig. 1.3). In the sartorius and gracilis muscles, instead of one zone of innervation there appear to be numerous scattered bands (5). This does not necessarily indicate multiple innervation of individual fibers, because it has been shown that the fibers do not run the entire length of the muscle (6, 7). The scattered zones probably represent simple innervation of short fibers linked in series. Knowledge of the extent of the zone of innervation is important in the evaluation of certain normal spontaneous electrical activity.

The zone of innervation usually lies near the *motor point,* which is the point where the motor nerve enters the muscle. The motor point may be identified clinically as the site where a twitch may be evoked in response to minimal electrical stimulation. Localization of the motor point permits the innervation zone to be exposed and biopsied accurately in certain muscles

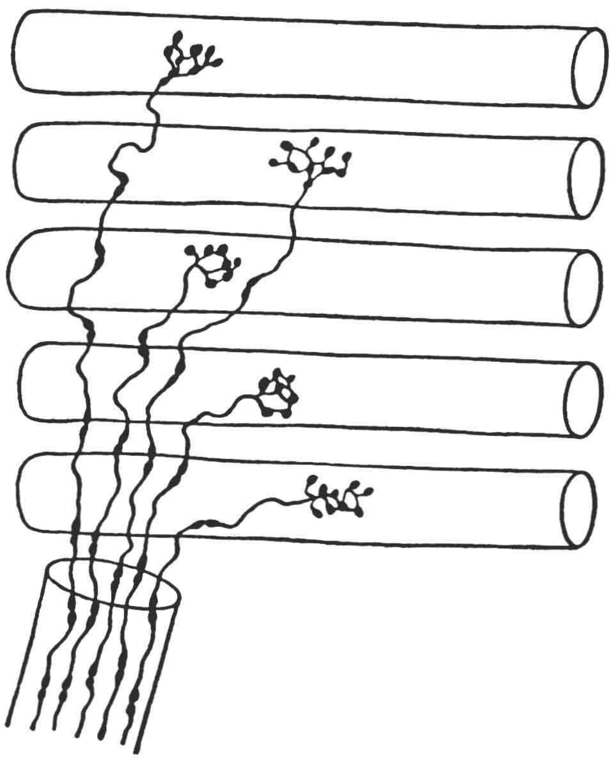

FIG.1.2. Diagram of normal terminal innervation pattern of skeletal muscle fibers. (From C. Cores and A. L. Woolf: In *The Innervation of Muscle,* Blackwell Scientific Publications, Ltd., Oxford, England, 1959.)

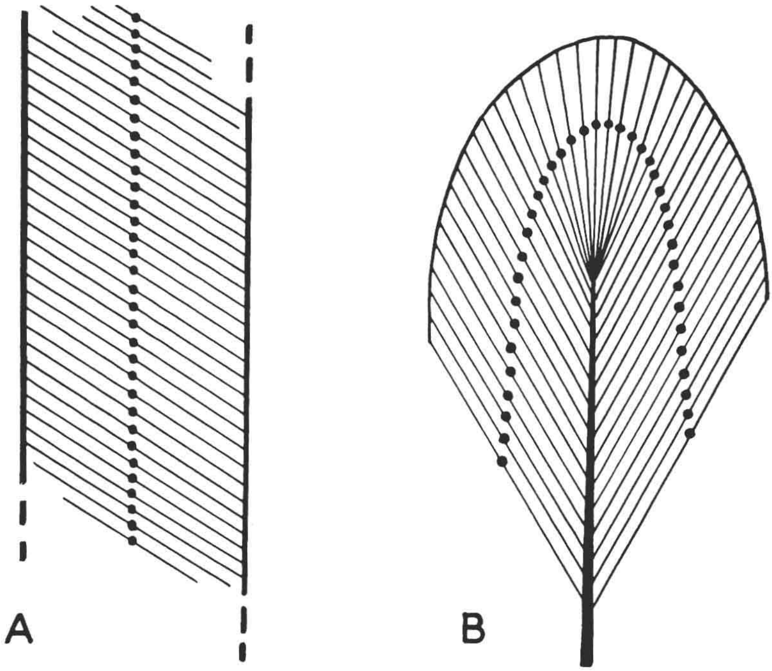

FIG. 1.3. Diagram of terminal innervation band distribution. *A*, muscle in which fibers run in a parallel manner; *B*, circumpennate muscle. (From Coers and A. L. Woolf: in *The Innervation of Muscle*, Blackwell Scientific Publications, Ltd., Oxford, England, 1959.)

with little difficulty. Coers (8), who developed a biopsy technique which depended on first finding the motor point by electrical stimulation, believes that in some muscles the motor point does not represent the entrance of the nerve into the muscle. It is the terminal branches of the nerve nearer the skin surface which are accessible for stimulation and correspond to the motor point. Whether the motor point represents the nerve entrance or the terminal branches, it is important to remember that the motor point is a fixed anatomic site.

SKELETAL MUSCLE

Each muscle is bound by a connective tissue sheath called the *epimysium* (Fig. 1.4). At various intervals the connective tissue passes from the surface into the muscle to form coarse sleeves, the *perimysium*. Smaller and smaller groups of muscle fibers are surrounded until ultimately the subdivisions of the perimysium result in the bundling together of about 12 or more muscle fibers into a discrete group, the muscle *fascicle*. The muscle fascicle is the smallest unit of the muscle that can be seen by the naked eye.

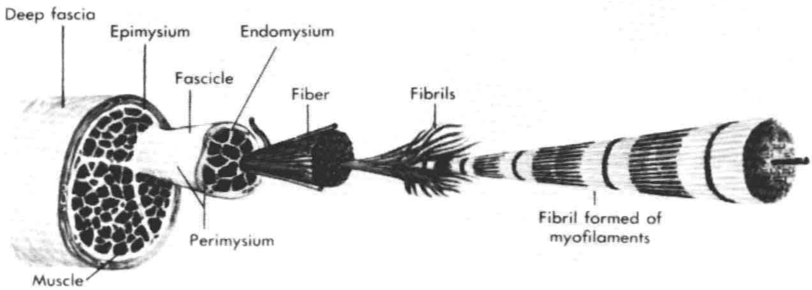

FIG. 1.4. Cross-section of skeletal muscle showing relationship of various anatomic structures. (From W. D. Gardner and W. A. Osburn: In *Structure of the Human Body*, W. B. Saunders Co., Philadelphia, 1967.)

The final distribution from the perimysium consists of a delicate network of fine connective tissue fibers which branches to surround each muscle fiber to form the *endomysium*, which serves to hold the capillaries and nerve fibers in place and secure the muscle fibers to each other.

Individual muscle fibers range from 0.01 to 0.1 mm in diameter and from 2 to 12 cm in length. The average fiber diameter increases from 0.01 mm in the newborn to about 0.05 mm in the adult (9). For short muscles, the muscle fibers extend the entire length of the muscle. For long muscles, however, a single fiber may extend only through a short distance of the total length. Within a fascicle, one end of the fibers terminates at a tendon and the other terminates in long tapering points which are overlapped by other muscle fibers and securely bound together by the reticular endomysium between them. Several muscle fibers may be attached end to end in this manner with the final fiber in the fascicle extending to the tendon at the other pole of the muscle. These fibers, tied together in series, act exactly as a single fiber of the same total length; it shortens by approximately one-half its length during contraction.

Histochemical studies have shown that human skeletal muscles do not consist of a grouping of homogeneous fibers but instead are composed of at least three types, each differing in enzymatic activity. Histochemically, the different fibers have been designated as Type I, Type II, and "intermediate."

Type I fibers are rich in mitochondrial oxidative enzymes, such as succinic dehydrogenase and cytochrome oxidase, but poor in phosphorylase, glycogen, and myofibrillar adenosine triphosphatase (ATPase). Type II fibers, conversely, are rich in phosphorylase, glycogen, and ATPase but poor in the oxidative enzymes (Table 1.1). Type II fibers also have a high content of mitochondrial α-glycerophosphate dehydrogenase (10, 11). Thus the two types of fibers contrast in energy metabolism; Type I fibers are concerned with aerobic metabolism, whereas Type II fibers are essen-

*Table 1.1. Relative Amount of Histochemical Staining within Human Muscle Fibers**

Reaction†	Muscle Fiber Reactivity	
	Type I	Type II
DPNH dehydrogenase	High	Low
TPNH dehydrogenase	High	Low
Succinate dehydrogenase	High	Low
Cytochrome oxidase	High	Low
Dihydroorotic acid dehydrogenase	High	Low
Benzidine peroxidase (probably myoglobin)	High	Low
Menadione-mediated α-glycerophosphate dehydrogenase	Low	High
DPN-linked lactate dehydrogenase (PMS, azide)‡	Low	High
DPN-linked α-glycerophosphate dehydrogenase, (PMS, azide)	Low	High
Phosphorylase	Low	High
Glycogen	Low	High
UDPG-glycogen transferase	High§	Low§
Argyrophil reaction	Medium	Medium
ATPase, myofibrillar	Low	High
ATPase, edetic acid low pH activated	High	Low
ATPase, "wet"	Medium	Medium
Antimyosin fluorescent antibody	Medium	Medium
Tyrosine	Medium	Medium
Esterase	High	Low
Osmium tetroxide	Medium	Medium
Oil red O	High	Low

* The relative amount of staining is consistent but does not necessarily represent the relative enzyme content of the two fiber types if technical factors exert a false localization influence.

† Abbreviations used are: DPNH, reduced diphosphopyridine nucleotide; TPNH, reduced triphosphopyridine nucleotide; DPN, diphosphopyridine nucleotide; PMS, phenazine methosulfate; UDPG, uridine diphosphate glucose; ATPase, adenosine triphosphatase.

‡ Reversed without PMS and azide.

§ Reversed in occasional specimens.

From W. K. Engel: Selective and nonselective susceptibility of muscle fiber types. Arch. Neurol. (Chicago), *22:* 98, 1970.

tially concerned with anaerobic metabolism. Fibers intermediate in enzyme activity between Types I and II have also been recognized.

In man, when the muscles are histochemically stained to exhibit these differential enzymatic characteristics, a cross-section presents a mosaic pattern of lightly and darkly stained fibers (Fig. 1.5); the different fiber types appear to be uniformly distributed through the muscle (12, 13). There does not appear to be any muscle composed entirely of one fiber type. In contradistinction, in animal muscles a particular histochemical fiber type may be concentrated in a single area of the whole muscle; for example, in the mouse, Type I and intermediate fibers are situated deeply, near to the bone in the normal triceps, tibialis anterior, and gastrocnemius, whereas Type II fibers are found in the more superficial part of the muscle.

In the soleus, the fibers are all of the Type I and intermediate classes (14).

In recent years, investigators (15, 16) have shown that the contractile characteristics of the various histochemical fiber types are also different. Isometric twitch measurements indicate that the contraction times (time from the start of the twitch to its peak tension) vary, with some fibers contracting much faster than others, so that a classification into slow twitch and fast twitch fibers is feasible. An example of twitch tensions recorded from a normal human rectus abdominis muscle biopsy showing both types of responses is given in Figure 1.6. There is also some evidence (15, 17) that the conduction velocities along the muscle fiber may be a function of fiber type.

All human skeletal muscle fibers are considered to be twitch fibers because they produce a mechanical twitch response for a single stimulus and generate a propagated action potential. This feature is quite distinct from the observations in frog muscle, where two major types of fibers are present, the fast or twitch fibers and the slow or tonic fibers. The extrinsic eye muscles represent a sole exception in humans, in that fibers with "tonic" characteristics may be present. Electron microscopic and cholines-

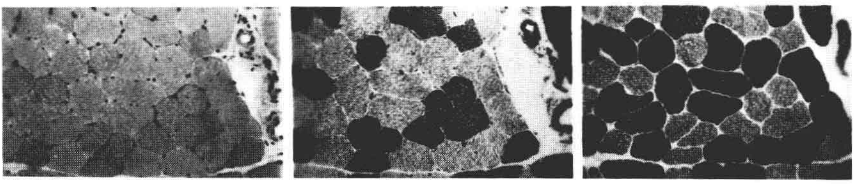

FIG.1.5. Serial sections of normal pectoralis major muscle. *Left,* hematoxylin and eosin stain; *center,* Type I fibers darker (DPN diaphorase); *right,* Type II fibers darker (adenosine triphosphatase). ×63. (Courtesy of John Pearson, M.D., Department of Pathology, New York University Medical Center, New York, New York.)

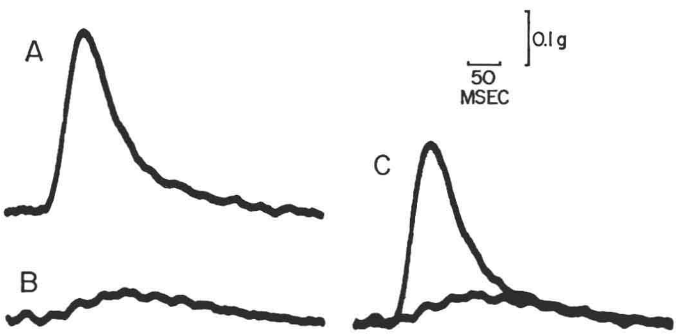

FIG.1.6. Twitch tensions of a rectus abdominis muscle biopsy showing fast twitch response (*A*) and slow twitch response (*B*). Response *A* and *B* are superimposed in *C*. (From A. Eberstein and J. Goodgold: Slow and fast twitch fibers in human skeletal muscle. Amer. J. Physiol., *215:* 539, 1968.)

terase staining studies show that some of these fibers have an afibrillar ultrastructure and multiple nerve endings.

THE MOTOR UNIT

Within a muscle, the axon from a single motor nerve cell arborizes into many terminal branches. Each branch is attached to an individual muscle fiber. The branching of the axon permits a single neuron to stimulate a group of muscle fibers. For example, an electrical impulse traveling along a single axon induces the contraction of approximately 2000 fibers in the gastrocnemius. The functional unit of the neuromuscular system thus differs from the structural units of the nerve and muscle systems which are, respectively, the neuron and the muscle fiber. The functional unit of the neuromuscular system is the *motor unit; it consists of the anterior horn cell, its axon, and all of the muscle fibers innervated by that axon* (Fig. 1.7). Modern study of the motor unit began when Liddell and Sherrington first used the term in 1925 (18). The concept was developed as a result of studies on the reflex activity of the spinal cord, the motor unit being considered the final common path of the nervous system.

The number of muscle fibers in a single motor unit varies widely for the different skeletal muscles (Table 1.2). A large muscle with many fibers which is involved in relatively gross movements may include hundreds of muscle fibers in a motor unit, whereas a muscle concerned with precise movements may have a small number of muscle fibers per motor unit. This is seen, for example, in the gastrocnemius and laryngeal muscles, which have 1934 and 2 to 3 muscle fibers per motor unit, respectively.

The number of muscle fibers per motor nerve fiber is expressed as the *innervation ratio*. This is usually computed by dividing the total number of muscle fibers in a muscle by the total number of motor nerve fibers. It is difficult to determine the exact number of motor nerve fibers in the nerve

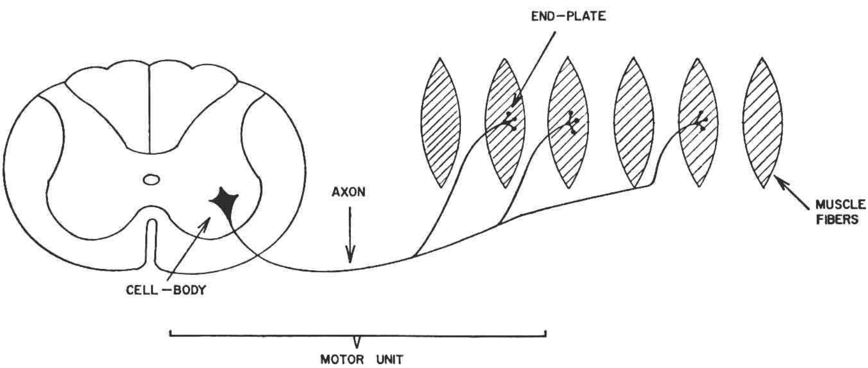

FIG. 1.7. Diagram indicating the single motor unit: the anterior horn cell, axon, and all of the muscle fibers innervated by the axon.

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