ELECTRODIAGNOSIS OF NEUROMUSCULAR DISEASES THIRD EDITION

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

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JOSEPH GOODGOLD, M.D.
Director, Electrodiagnostic Service,
Institute of Rehabilitation Medicine and University Hospital,
New York University Medical Center

And the Howard A. Rusk Professor, and Chairman,
Department of Rehabilitation Medicine,
New York University School of Medicine

ARTHUR EBERSTEIN, Ph.D.
Professor, Department of Rehabilitation Medicine,
New York University School of Medicine

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This book is dedicated to

Mildred S. Goodgold
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for their continued understanding, devotion, and encouragement.

Preface to the Third Edition

The publication of the third edition of *Electrodiagnosis of Neuromuscular Diseases* has provided an opportunity to update the contents and to add new material that notably serves to reflect the dynamic nature of clinical electrodiagnosis.

The basic purpose of this book has not changed. This edition presents a comprehensive and critical introduction to the practice of electromyography, nerve conduction studies, reflexology, and evoked cerebral potentials 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 performance of these electrophysiological studies do not fall into the category of "laboratory" tests, carried out by technicians. Rather they are physician oriented and truly represent an extension of the history and physical examination used in clinical assessment of diseases of the neuromuscular system.

If meaningful interpretation of the electrodiagnostic observations is to prevail, a solid knowledge of medicine, especially in the neurological realm, must be engrafted on an equally concrete informational base of anatomy, pathology, physiology, and fundamental electronics. These subjects do not uniquely fall within the absolute domain of any single medical specialty, such as neurology, neurosurgery, rehabilitation medicine, or orthopaedics. Successful completion of training 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 seem to be a minimal, and essential, postresidency interval.

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 or omitted.

The principles underlying electromyography and nerve stimulation studies are presented 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 dignosis—the neurosurgeon, neurologist, orthopaedist, 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 fundamental 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.

In regard to content, 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 Reflexology; to Goodwin M. Breinin, M.D., Chairman and Professor of the Department of Ophthalmology, New York University Medical Center, for his contribution on Ocular Electromyography; and to Drs. Joan B. Cracco, Associate Professor, and Roger Q. Cracco, Professor and Chairman, Department of Neurology, State University of New York, Downstate Medical Center, Brooklyn, New York, for their comprehensive section on Somatosensory Evoked Potentials.

We are likewise indebted to the American Association of Electromyography and Electrodiagnosis for permission to reproduce as an appendix their publication "A Glossary of Terms Used in Clinical Electromyography."

Many sections of the previous editions have been altered and include updated comments on the resting potential, spontaneous electrical activity, motor unit recruitment, Guillain-Barré syndrome, H reflex, F wave, etc. There is a new chapter on Somatosensory Potentials, a new section on macro EMG, and an entirely rewritten chapter on Myasthenia Gravis.

Throughout the years many friends and colleagues have helped us by pointing out typographical, grammatical, photographic, and occasional substantive errors that have insidiously crept into the text. We are ever grateful to all, but particularly to David G. Simons, M.D., who meticulously reviewed and positively critiqued each and every chapter.

Joseph Goodgold, M.D.
Arthur Eberstein, Ph.D.

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. The Golgi tendon organs are highly specialized sensory receptors in series with the skeletal muscle fibers and detect tension applied to the tendon during muscle contraction or stretch; muscle spindles are sensory receptors which detect change in length of the muscle fibers and the rate of change in length. Signals from these receptors are sent back to the central

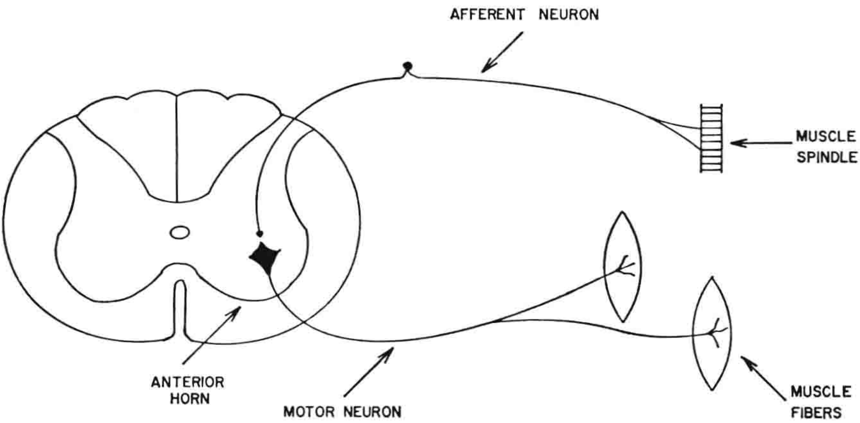

FIG. 1.1. Diagram showing innervation of skeletal muscle fibers by a motor neuron. Impulses are conducted away from the central nervous system in the motor neuron and toward the central nervous system in the afferent neuron.

nervous system via the sensory nerve fibers which are 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 (3), 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 (4, 5).

Cholinesterase staining demonstrates the presence of two kinds of nerve endings: (a) large, heavily staining compact discs which innervate the twitch fibers and are called *en plaque* endings; (b) smaller, lighter staining droplets arranged in clusters or chains along the single muscle fiber, which are classified as *en grappe* endings and innervate the tonic fibers (5). 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 across the

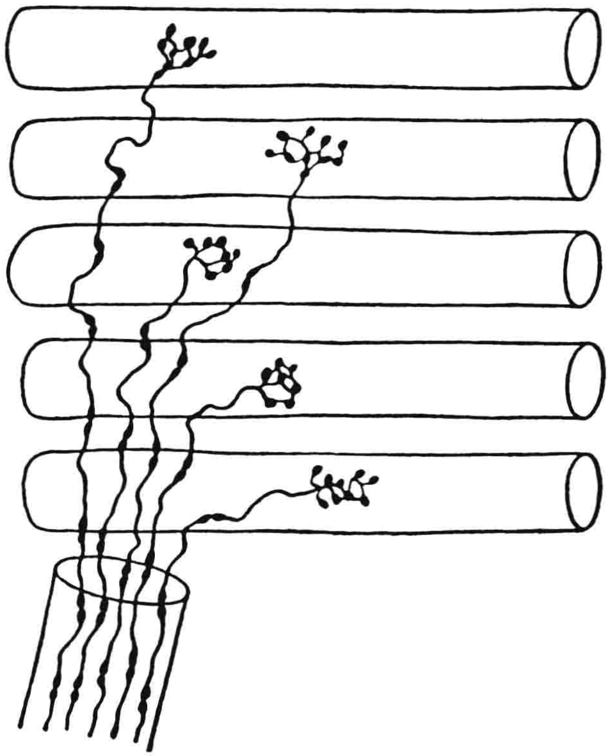

FIG. 1.2 Diagram of normal terminal innervation pattern of skeletal muscle fibers. (From C. Coers and A. L. Woolf: In *The Innervation of Muscle*. Blackwell Scientific Publications, Ltd., Oxford, England, 1959.)

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 (3). 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 biopsy specimens to be accurately

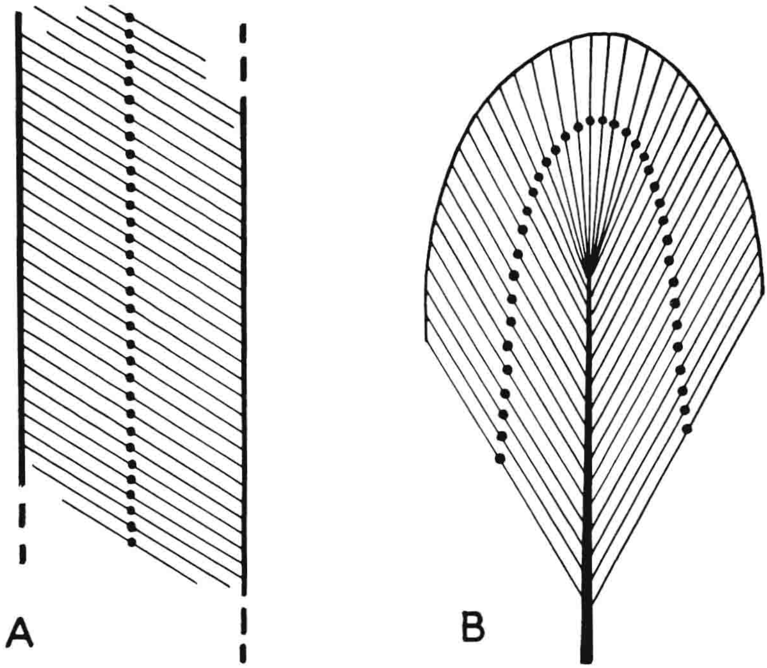

FIG. 1.3. Diagram of terminal innervation band distribution. *A*, muscle in which fibers run in a parallel manner; *B*, circumpennate muscle. (From Coers and A. L. Woolf: In *The Innervation of Muscle*. Blackwell Scientific Publications, Ltd., Oxford, England, 1959.)

obtained in certain muscles 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. The final

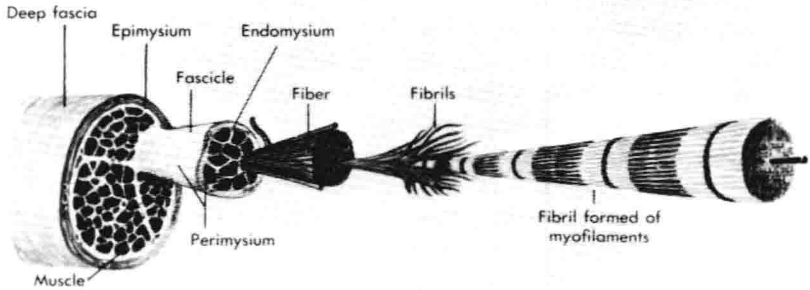

FIG. 1.4. Cross-section of skeletal muscle showing relationship of various anatomic structures. (From W. D. Gardner and W. A. Osburn: In *Structure of the Human Body*. W. B. Saunders Co., Philadelphia, 1967.)

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

Type I muscle fibers have long contraction times and are highly resistant to fatigue, whereas Type II fibers have short contraction times and fatigue rapidly. Type I fibers have a low threshold of activation and a firing rate of 8 to 10 per sec; Type II fibers have a higher threshold, are activated with a rapid vigorous contraction, and fire in short, irregular bursts at 16 to 50 per sec. A low level sustained contraction will stimulate the Type I fibers. The axons supplying the Type I motor units have smaller diameters and lower conduction velocities than those connected to Type II motor units.

In man, when the muscles are histochemically stained to exhibit these

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.

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). In any one region of a skeletal muscle, there may be an intermingling of approximately 10 different fiber types. 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 most 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. Tonic fibers do not respond to stimuli with twitches but with continuous, graded contractions. The extrinsic eye muscles represent a sole exception in humans, in that fibers with "tonic" characteristics may be present. Electron microscopic and cholinesterase staining studies show that some of these fibers have an afibrillar ultrastructure and multiple nerve endings.

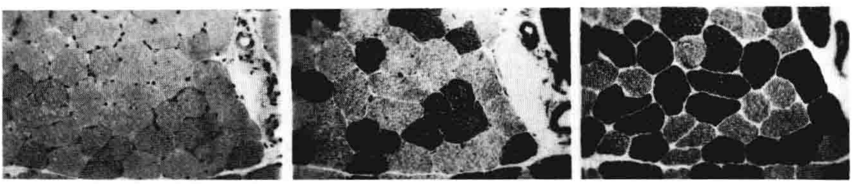

FIG. 1.5. Serial sections of normal pectoralis major muscle. *Left*, hematoxylin and eosin stain; *center*, Type I fibers darker (DPN diaphorase); *right*, Type II fibers darker (adenosine triphosphatase). ×63. (Courtesy of John Pearson, M.D., Department of Pathology, New York, University Medical Center,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

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

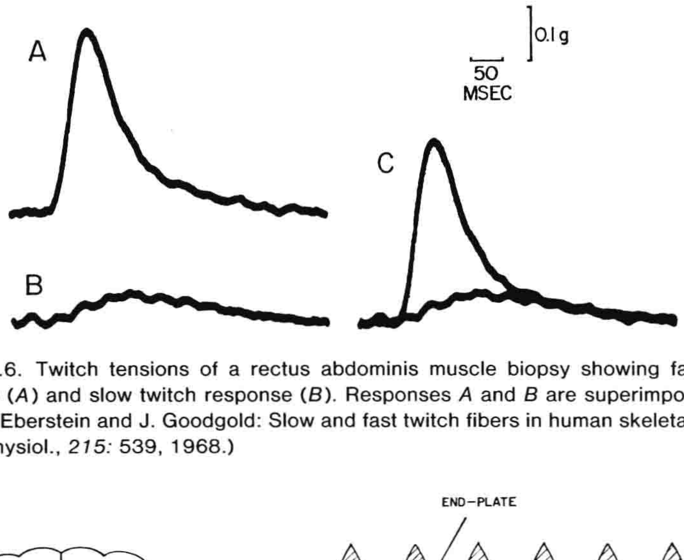

FIG. 1.6. Twitch tensions of a rectus abdominis muscle biopsy showing fast twitch response (*A*) and slow twitch response (*B*). Responses *A* and *B* are superimposed in *C*. (From A. Eberstein and J. Goodgold: Slow and fast twitch fibers in human skeletal muscle. Am. J. Physiol., *215:* 539, 1968.)

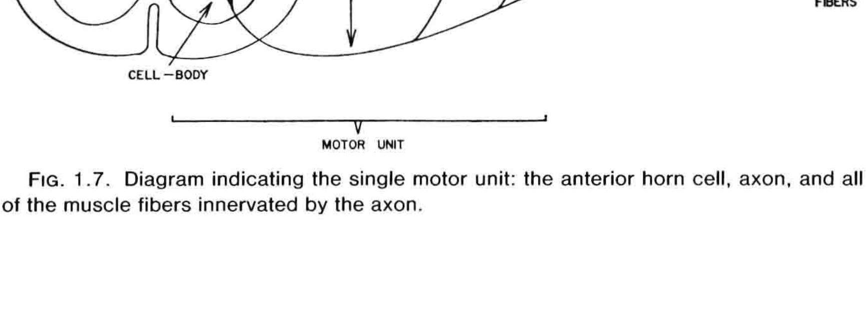

FIG. 1.7. Diagram indicating the single motor unit: the anterior horn cell, axon, and all of the muscle fibers innervated by the axon.

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 two to three 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 trunk because muscle afferent (sensory) and small motor nerve fibers supplying the intrafusal muscle fibers are also present. If only the large nerve fibers are considered (including both motor and sensory components), the calculated innervation ratio will be substantially lower than the correct value. Some investigators have attempted to increase the accuracy by assuming that 40% of the large-sized nerve fibers are afferent. One investigator (19) was able to determine the innervation ratio in the laryngeal muscles by actually observing the different twigs of the same branch of the nerve innervating the various muscle fibers.

It is generally agreed that the individual fibers of a motor unit are not all grouped together but rather involve more than single fasciculi. There is considerable intermingling of fibers derived from different motor neurons. Although direct evidence that this is true in human muscle is difficult to obtain and as yet not available, recent histochemical studies (22, 23) have clearly shown the diffuse anatomic distribution of the motor units in rat muscle. In these studies, single ventral root nerve fibers innervating the

TABLE 1.2. Number of Muscle Fibers per Motor Unit in Various Human Muscles

Muscle	No. of Large Nerve Fibers	No. of Muscle Fibers	Calculated No. of Motor Units	Mean No. of Fibers per Motor Unit	Reference
Platysma	1826	27,100	1096	25	20
First dorsal interosseous	199	40,500	119	340	20
Lumbricalis I	155	10,038	93	108	20
Anterior tibialis	742	250,200	445	562	20
Gastrocnemius, medial	965	1,120,000	579	1934	20
Laryngeal muscles				2–3	19
External rectus				9	20
Temporalis				936	21
Masseter				640	21