Module 2

Neurology and Instrumentation

Motor paralysis

The term *paralysis* is derived from the Greek words *para*, "beside,off, amiss," and *lysis*, a "loosening" or "breaking up." In medicine it has come to refer to an abolition of function, either sensory or motor. When applied to motor function, *paralysis* means loss of voluntary movement due to interruption of one of the motor pathways at any point from the cerebrum to the muscle fiber.

Patterns of paralysis and their diagnosis:

The following subdivision, based on the location and distribution of the muscle weakness:

1. *Monoplegia :* refers to weakness or paralysis of all the muscles of one leg or arm. This term should not be applied to paralysis of isolated muscles or groups of muscles supplied by a single nerve or motor root.
2. *Hemiplegia*: It is the commonest form of paralysis, involves the arm, the leg, and sometimes the face on one side of the body. With rare exceptions, mentioned further on, hemiplegia is attributable to a lesion of the corticospinal system on the side opposite to the paralysis.
3. *Paraplegia:* indicates weakness or paralysis of both legs. It is most often the result of diseases of the thoracic spinal cord, caudal equina, or peripheral nerves, and rarely, both medial frontal cortices.
4. *Quadriplegia (tetraplegia) :*denotes weakness or paralysis of all four extremities. It may result from disease of the peripheral nerves, muscles, or myoneural junctions; gray matter of the spinal cord; or the upper motor neurons bilaterally in the cervical cord, brainstem, or cerebrum. *Diplegia* is a special form of quadriplegia in which the legs are affected more than the arms. *Triplegia* occurs most often as a transitional condition in the development of or partial recovery from tetraplegia.
5. Isolated paralysis of one or more muscle groups.
6. Nonparalytic disorders of movement (apraxia, ataxia, etc.).
7. Muscular paralysis without visible changes in motor neurons, roots, or nerves.
8. Hysterical paralysis.

1. Monoplegia

The examination of patients who complain of weakness of one limb often discloses an asymptomatic weakness of another, and the condition is actually a

hemiparesis or paraparesis. Or, instead of weakness of all the muscles in a limb, only isolated groups are found to be affected. Ataxia, sensory disturbances, or reluctance to move the limb because of pain must not be misinterpreted as weakness. Parkinsonism may give rise to the same error, as can rigidity or bradykinesia of other causation or a mechanical limitation due to arthritis and bursitis. The presence or absence of atrophy of muscles in a monoplegic limb is of particular diagnostic help, as indicated below.

Monoplegia without Muscular Atrophy

This is most often due to a lesion of the cerebral cortex. Only infrequently does it result from a subcortical lesion that interrupts the motor pathways. A cerebral vascular lesion (thrombotic or embolic infarction) is the commonest cause; a circumscribed tumor or abscess may have the same effect. A small cortical lesion may paralyze half the hand or even just the thumb. Multiple sclerosis and spinal cord tumor, early in their course, may cause weakness of one limb, usually the leg. Monoplegia due to a lesion of the upper motor neuron is usually accompanied by spasticity, increased reflexes, and an extensor plantar reflex (Babinski sign); exceptionally, a small lesion of the motor cortex will not result in spasticity.

.

Monoplegia with Muscular Atrophy

This is more frequent than monoplegia without muscular atrophy. Long-continued disuse of one limb may lead to atrophy, but it is usually of lesser degree than atrophy due to lower motor neuron disease (denervation atrophy). In disuse atrophy, the tendon reflexes are retained and nerve conduction studies are normal. With denervation of muscles, there may be visible fasciculations and reduced or abolished tendon reflexes in addition to paralysis. If the limb is partially denervated, the electromyogram shows reduced numbers of motor unit potentials (often of large size) as well as fasciculations and fibrillations. The location of the lesion (in nerves, spinal roots, or spinal cord) can usually be determined by the pattern of weakness, by the associated neurologic symptoms and signs, and by special tests—magnetic resonance imaging (MRI) of the spine, examination of the cerebrospinal fluid (CSF), and electrical studies of nerve and muscle. When present in an infant, it should suggest brachial plexus trauma from birth; in a child, poliomyelitis or other viral infection of the spinal cord; and in an adult, poliomyelitis, syringomyelia, amyotrophic lateral sclerosis, or a brachial plexus lesion. Crural (leg) monoplegia is more frequent than brachial monoplegia and may be caused by any lesion of the thoracic or lumbar cord—i.e., trauma, tumor, myelitis, multiple sclerosis, progressive muscular atrophy, late radiation effect,

etc.

2.Hemiplegia

This is the most frequent form of paralysis. With rare exceptions (a few unusual cases of poliomyelitis or motor system disease), this pattern of paralysis is due to involvement of the corticospinal pathways. The site or level of the lesion i.e., cerebral cortex, coronaradiata, capsule, brainstem, or spinal cord can usually be deduced from the associated neurologic findings. Diseases localized to the cerebral cortex, cerebral white matter (corona radiata), and internal capsule usually manifest themselves by weakness or paralysis of the leg, arm, and lower face on the opposite side. The occurrence of seizures or the presence of a language disorder (aphasia), a loss of discriminative sensation (astereognosis, impairment of tactile localization, etc.), anosognosia, or a homonymous visual field defect suggests a contralateral cortical or subcortical location.Damage to the corticospinal and corticobulbar tracts in the upper portion of the brainstem also causes paralysis of the face, arm, and leg of the opposite side.

The lesion in such cases may in some patients be localized by the presence of a third nerve palsy (Weber syndrome) or other segmental abnormality on the same side as the lesion (opposite the hemiplegia). With low pontine lesions, an ipsilateral abducens or facial palsy is combinedwith a contralateral weakness or paralysis of the arm and leg (Millard-Gubler syndrome). Lesions in the medulla affect the tongue and sometimes the pharynx and larynx on one side and the arm and leg on the other. Even lower in the medulla, a unilateral infarct in the pyramid causes a flaccid paralysis followed by slight spasticity of the contralateral arm and leg, with sparing of the face and tongue. Some motor function may be retained, as in the case described by Ropper and colleagues; interestingly, in this case and in others previously reported, there was considerable recovery of voluntary power even though the pyramid was almost completely destroyed. Rarely, an ipsilateral hemiplegia may be caused by a lesion in the lateral column of the cervical spinal cord. In this location, however, the pathologic process more often induces bilateral signs, with resulting quadriparesis or quadriplegia. A homolateral paralysis that spares the face, if combined with a loss of vibratory and position sense on the same side and a contralateral loss of pain and temperature, signifies disease of one side of the spinal cord As indicated above, muscle atrophy that follows upper motor neuron lesions never reaches the proportions seen in diseases of the lower motor neuron. The atrophy in the former cases is due to disuse. When the motor cortex and adjacent parts of

the parietal lobe are damaged in infancy or childhood, normal development of the muscles as well as the skeletal system in the affected limbs is retarded. The limbs and even the trunk are smaller on one side than on the other. This does not happen if the paralysis occurs after puberty, by which time the greater part of skeletal growth has been attained. Trauma (brain contusion, epidural and subdural hemorrhage) ranks second. Other important causes, less acute in onset, are, in order of frequency, brain tumor, brain abscess, demyelinative diseases, and the vascular complications of meningitis and encephalitis. Alternating transitory hemiparesis may be due to a special type of migraine .From time to time, hysteria is found to be the cause of a hemiplegia

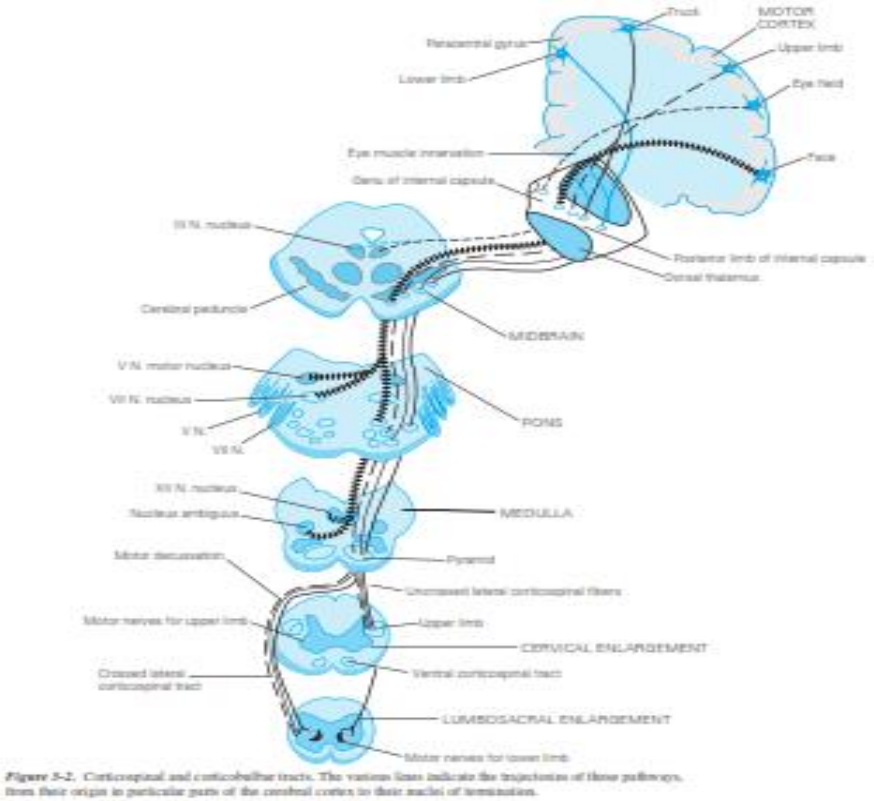


Figure 3–2. Corticospinal and corticobulbar tracts. The various lines indicate the trajectories of these pathways, from their origin in particular parts of the cerebral cortex to their nuclei of termination.

Paraplegia

Paralysis of both lower extremities may occur with diseases of the spinal cord, nerve roots, or, less often, the peripheral nerves. If the onset is acute, it may be difficult to distinguish spinal from neuropathic paralysis because of the element of spinal shock, which results in abolition of reflexes and flaccidity. In acute spinal cord diseases with involvement of corticospinal tracts, the paralysis or weakness affects all muscles below a given level; usually, if the white matter is extensively damaged, sensory loss below a particular level is conjoined (loss of pain and temperature sense due to spinothalamic tract

damage, and loss of vibratory and position sense due to posterior column involvement). Also, in bilateral disease of the spinal cord, the bladder and bowel and their sphincters are usually affected. These abnormalities may be due to an intrinsic lesion of the cord or to an extrinsic mass that narrows the spinal canal, both types of lesion being evident on MRI.

In peripheral nerve diseases, motor loss tends to involve the distal muscles of the legs more than the proximal ones (exceptions are certain varieties of the Guillain-Barre´ syndrome and certain types of diabetic neuropathy and porphyria); sphincteric function is usually spared or impaired only transiently. Sensory loss, if present, is also more prominent in the distal segments of the limbs, and the degree of loss is often more for one modality than another. For clinical purposes it is helpful to separate the acute paraplegias from the chronic ones and to divide the latter into two groups: those beginning in adult life and those occurring in infancy. The most common cause of acute paraplegia (or quadriplegia if the cervical cord is involved) is spinal cord trauma, usually associated with fracture-dislocation of the spine. Less common causes are hematomyelia due to a vascular malformation, an arteriovenous malformation of the cord that causes ischemia by an obscure mechanism, or infarction of the cord due to occlusion of the anterior spinal artery or, more often, to occlusion of segmental branches of the aorta (due to dissecting aneurysm or atheroma, vasculitis, and nucleus pulposus embolism).

In adult life, multiple sclerosis and tumor account for most cases of subacute and chronic spinal paraplegia, but a wide variety of extrinsic and intrinsic processes may produce the same effect: protruded cervical disc and cervical spondylosis (often with a congenitally narrow canal), epidural abscess and other infections (tuberculous, fungal, and other granulomatous diseases), syphilitic meningomyelitis, motor system disease, subacute combined degeneration (vitamin B12 deficiency), syringomyelia, and degenerative disease of the lateral and posterior columns of unknown cause. In pediatric practice, delay in starting to walk and difficulty in walking are common problems. These conditions may indicate a systemic disease (such as rickets), mental deficiency, or, more commonly, some muscular or neurologic process

Quadriplegia (Tetraplegia)

All that has been said about the spinal causes of paraplegia applies to quadriplegia, the lesion being in the cervical rather than the thoracic or lumbar segments of the spinal cord. If the lesion is situated in the low cervical segments and involves the anterior half of the spinal cord, as typified by the syndrome resulting from occlusion of the anterior spinal artery (but occurring also in some cases of myelitis and fracture-dislocations of the cervical spine). In all these processes, the paralysis of the arms may be flaccid and areflexic in type and that of the legs, spastic. There is usually pain in the neck and shoulders and numbness of the hands; elements of ataxia from posterior column lesions accompany the paraparesis. Compression of the C1 and C2 spinal cord segments is caused by dislocation of the odontoid process. Rheumatoid arthritis and Morquio disease are causes of special note; in the latter, there is pronounced dural thickening.

A progressive syndrome of monoparesis, biparesis, and then triparesis is caused by tumors and a variety of other compressive lesions in the region of the foramen magnum and high cervical cord. Bilateral infarction of the medullary pyramids from occlusion of the vertebral arteries or their anterior spinal branches is a very rare cause of quadriplegia. Repeated strokes affecting both hemispheres may lead to bilateral hemiplegia, usually accompanied by pseudobulbar palsy. In infants and young children, aside from developmental abnormalities and anoxia of birth, certain metabolic cerebral diseases (metachromatic and other forms of leukoencephalopathy, lipid storage disease) may be responsible for a quadriparesis or quadriplegia, but always with severe psychomotor retardation. Congenital forms of muscular dystrophy and muscular atrophy (Werdnig-Hoffmann disease) may be recognized soon after birth or later and may progress slowly.

Triplegia Paralysis that remains confined to three limbs is observed only rarely; more often the fourth limb is weak or hyper-reflexic, and the syndrome is really an incomplete tetraplegia. As indicated earlier, this pattern of involvement is important, because it may signify an evolving lesion of the upper cervical cord or cervicomedullary junction. A meningioma of the foramen magnum, for example, may begin with spastic weakness of one limb, followed by sequential involvement of the other limbs in an “around the clock”

pattern. There are usually bilateral Babinski signs early in the process, but there may be few sensory findings. Also this pattern is seen in patients with multiple sclerosis and other intrinsic inflammatory and neoplastic lesions. These same diseases may produce triplegia (or triparesis) by a combination of paraplegia from a thoracic spinal cord lesion and a separate unilateral lesion in the cervical cord or higher that results in a hemiparesis.

Paralysis of Isolated Muscle Groups

This condition usually indicates a lesion of one or more peripheral nerves or of several adjacent spinal roots. The diagnosis of an individual peripheral nerve lesion is made on the basis of weakness or paralysis of a particular muscle or group of muscles and impairment or loss of sensation in the distribution of the nerve. Complete or extensive interruption of a peripheral nerve is followed by atrophy of the muscles it innervates and by loss of tendon reflexes of the involved muscles; abnormalities of vasomotor and sudomotor functions and trophic changes in the skin, nails, and subcutaneous tissue may also occur. Knowledge of the motor and sensory innervation of the peripheral nerve in question is needed for a satisfactory diagnosis. It is not practical to memorize the precise sensorimotor distribution of each peripheral nerve, and special manuals, such as Aids to the Examination of the Peripheral Nervous System, should be consulted.

In addition, it is important to decide whether the lesion is a temporary one of electrical conduction alone or whether there has been a structural interruption of nerve fibers, requiring nerve regeneration or corrective surgery for recovery. Electromyography and nerve conduction studies are of great value here. If there is no evidence of upper or lower motor neuron disease but certain movements are nonetheless imperfectly performed, one should look for a disorder of position sense or cerebellar coordination or for rigidity with abnormalities of posture and movement due to disease of the basal ganglia .In the absence of these disorders, the possibility of an apraxic disorder should be investigated by the methods outlined earlier

Hysterical Paralysis

Hysterical paralysis may involve one arm or leg, both legs, and all of one side of the body. Tendon reflexes are retained and atrophy is lacking in hysterical paralysis, features that distinguish it from chronic lower motor neuron disease. Diagnostic difficulty arises only in certain acute cases of upper motor neuron disease that lack the usual changes in reflexes and muscle tone. The hysterical gait is often diagnostic .Sometimes there is loss of sensation in the paralyzed parts and loss of sight, hearing, and smell on the paralyzed side—a pattern of sensory changes that is never seen in organic disease of the nervous system. When the hysterical patient is asked to move the affected limbs, the movements tend to be slow, hesitant, and jerky, often with contraction of agonist and antagonist muscles simultaneously and intermittently ("give-way" weakness). Lack of effort is usually obvious, despite facial and other expressions to the contrary. Power of contraction improves with encouragement.

The weakness is inconsistent; some movements are performed tentatively and moments later another movement involving the same muscles is performed naturally. Hoover's sign and the trunk-thigh sign of Babinski are helpful in distinguishing hysterical from organic hemiplegia. To elicit Hoover's sign, the examiner places both hands under the heels of the recumbent patient, who is asked to press the heels down forcefully. With organic hemiplegia, downward pressure will be felt from the nonparalyzed leg. The examiner then removes his hand from under the nonparalyzed leg, places it on top of the nonparalyzed one, and asks the patient to raise that leg. In true hemiplegia, no added pressure will be felt by the hand that remained beneath the heel of the paralyzed leg. In hysteria, the heel of the supposedly paralyzed leg may press down on the examiner's hand. Or, more useful in our experience, the normal leg fails to demonstrate downward pressure when the hysteric is asked to elevate the supposedly paralyzed one, thereby indicating a lack of voluntary effort. To carry out Babinski's trunkthigh test, the examiner asks the recumbent patient to sit up whilekeeping his arms crossed in front of his chest. In the patient with organic hemiplegia, there is an involuntary flexion of the paretic lower limb; in paraplegia, both limbs are flexed as the trunk is

flexed; in hysterical hemiplegia, only the normal leg may be flexedand in hysterical paraplegia, neither leg is flexed.

Muscular Paralysis and Spasm Unattended by Visible Changes in Nerve or Muscle

A discussion of motor paralysis would not be complete without some reference to a group of diseases in which muscle weakness may be profound but there are no overt structural changes in motor nerve cells or nerve fibers. Almost any disease of the neuromuscular junction and many diseases of muscle may cause this combination. This group comprises myasthenia gravis; inflammatory sympathies, the muscular dystrophies, and myotonia congenita(Thomsen disease); familial periodic paralysis; disorders of potassium, sodium, calcium, and magnesium metabolism; tetany; tetanus; poisoning by Clostridium botulinum; black widow spider bite; and the thyroid and other endocrine myopathies. In these diseases, each with a fairly distinctive clinical picture, the abnormality is essentially biochemical; their investigation requires special biochemical and histochemical tests and electron microscopic study.

Electrodiagnosis of neuromuscular disease

It was long ago discovered that muscle would contract when a pulse of electric current was applied to the skin, near the point of entrance of the muscular nerve (motor point). The electrical pulse required is brief, less than a millisecond, and is most effectively induced by a rapidly alternating (faradic) current. After denervation, an electrical pulse of several milliseconds, induced by a constant electrical (galvanic) stimulus, is required to produce the same response. This change, in which the galvanic stimulus remains effective after the faradic one has failed, was the basis of Erb's reaction of degeneration, and varying degrees of this change were later plotted in the form of strength-duration curves. For decades, this was the standard electrical method for evaluating denervation of muscle. Though still valid, it was replaced long ago by nerve conduction studies and by the needle electrode examination. The latter test, based on the sherringtonian concept of the "motor unit" is accomplished by recording the firing characteristics of evoked motor unit potentials (CMAPs) and by the insertion into muscle of needle electrodes to measure spontaneous and voluntarily evoked muscle fiber activity. The terms

electromyography and electromyogram (EMG) were used originally to describe the needle electrode examination but are now a common shorthand designation for the entire electrodiagnostic evaluation, including the nerve conduction studies, described below.

Studies of Nerve Conduction

The main laboratory technique for the study of peripheral nerve function involves the transcutaneous stimulation of motor or sensory nerves and recording of the elicited action potentials in the muscle (CMAP) and the sensory nerve action potential (SNAP). The results of these motor and sensory nerve conduction studies, expressed as amplitudes, conduction velocities, and distal latencies, yield certain quantitative information and additional qualitative observations regarding the waveform and dispersion of electrical impulses. Hodes and coworkers in 1948 were the first to describe nerve conduction studies in patients and the techniques used currently are not much changed. An accessible nerve is stimulated through the skin by surface electrodes, using a stimulus that is large enough to recruit all the available nerve fibers.

The resulting action potential is recorded by electrodes on the skin over the muscle distally in the case of motor fibers stimulated in a mixed or motor nerve (CMAP), over the nerve proximally, using orthodromic techniques for sensory fibers stimulated in the digital nerves, over the nerve more distally, using antidromic techniques for sensory nerve conduction studies (this has technical advantages over orthodromic techniques), and over the nerve more proximally for mixed nerve conduction studies. These techniques are the ones used most often in clinical work. An alternative but much more demanding technique uses “near nerve” needle electrodes to record action potentials as they course through the nerve. The main characteristics of the conventional nerve conduction studies are described below

Distal (Terminal) Latencies, Conduction Times, and Conduction

Velocities The conduction times from the most distal stimulating electrode to the recording site, in milliseconds, as determined by the latency from the stimulus artifact to the onset and to the peak of the CMAP, are termed the distal (or terminal) and peak motor latencies, respectively (see Fig.). The

former is the one used more often as a reflection of conduction time in routine work. A stimulus may then be applied to the nerve at a second site more proximally (or if recording electrodes can be placed more proximally in the case of sensory fibers), and a conduction time can be measured over a longer segment of nerve. When the distance (in millimeters) between the two sites of stimulation is divided by the difference in conduction times (in milliseconds), one obtains a conduction velocity (in meters per second), which describes the maximal velocity of propagation of the action potentials in the largest diameter and fastest nerve fibers. These velocities in normal subjects vary from a minimum of 40 or 45 m/s to a maximum of 65 to 75 m/s, depending upon which nerve is studied (e.g., slower in the legs than in the arms; Table45-1). Values are lower in infants, reaching the adult range by the age of 2 to 4 years and decline again slightly with advancing age. They are routinely diminished also with exposure to cold—a potentially important artifact if these recordings are taken when the patient's skin is cool; measurement of skin temperature therefore is routinely made prior to performing the conduction tests.

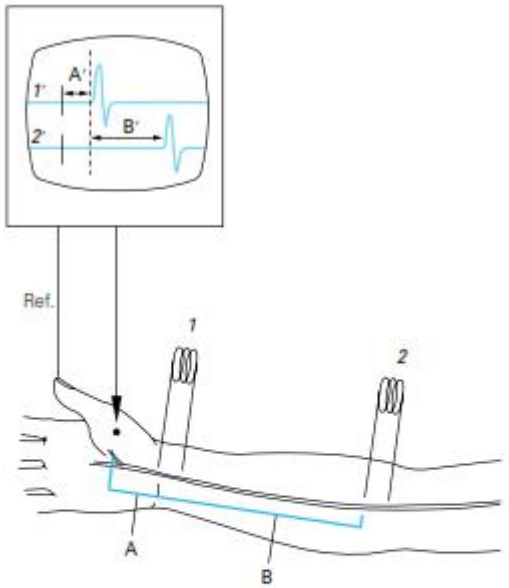


Figure 45-3. The median nerve is stimulated percutaneously (1) at the wrist and (2) in the antecubital fossa with the resultant compound muscle action potential recorded as the potential difference between a surface electrode over the thenar eminence (*arrow*) and a reference electrode (Ref.) more distally. Sweep 1′ on the display depicts the stimulus artifact followed by the compound muscle action potential. The distal latency, A′, is the time from the stimulus artifact to the take-off phase of the compound muscle action potential and corresponds to conduction over distance A. The same is true for sweep 2′, where stimulation is at 2 and the time from the artifact to the response is A′ + B′. The maximum motor conduction velocity over segment B is calculated by dividing distance B by the time B′.

Normal values have been established for distal latencies from the various sites of stimulation on various mixed nerves to the appropriate muscles. When one stimulates the median nerve at the wrist, for example (see electrode 1 and segment A in Fig. 45-3), the latency for motor conduction through the carpal tunnel to the median-innervated thenar muscles is always less than 4.5 ms in healthy adults.

Similar normal values have been compiled for orthodromic and antidromic sensory conduction velocities and distal latencies (see Table 45-1) in all the main peripheral nerves.

Normal Values for Representative Nerve Conduction Values at Various Sites of Stimulation (Mean Values +/− 2 SD for Adults 16 to 65 Years of Age)

MOTOR NERVE CONDUCTION STUDIES

NERVE	DISTAL STIMULATION SITE	OTHER STIMULATION SITES	RECORDING SITE	ONSET LATENCY (ms)	AMP (mV)	CV (m/s)	DISTANCE (cm)	F-WAVE LATENCY (ms)
Median	Wrist	Elbow	APB	<4.2	>4.4	>49	6–8	<31
Ulnar	Wrist	BG, AG	ADM	<3.4	>6.0	>49	5.5–7.5	<32
Radial	Forearm	Elbow, SG	EIP	<5.2	>4.0	>50	10	NA
Peroneal	Ankle	BFH, AFH	EDB	<5.8	>2.0	>42	6–11	<58
Peroneal	BFH	AFH	TA	<3.0	>5.0	>42	10	NA
Tibial	Ankle	PF	AH	<6.5	>3.0	>41	6–8	<59[a]

SENSORY NERVE CONDUCTION STUDIES[b]

NERVE	DISTANCE STIMULATION SITES	RECORDING SITE	ONSET LATENCY (ms)	PEAK LATENCY (ms)	AMP (μV)	CV (m/s)	DISTANCE (cm)
Median	Wrist	Dig2	<2.5	<3.5	>20	>52	13
Ulnar	Wrist	Dig5	<2.1	<3.0	>15	>52	11
Radial	Forearm	Wrist	<1.9	<2.8	>20	>48	10
Sural	Calf	Ankle	<3.2	<4.4	>6	>42	14

Key: AG, above ulnar groove; BG, below ulnar groove; AFH, above fibular head; BFH, below fibular head; SG, spiral groove; TA, anterior tibialis; EDB, extensor digitalis brevis; EIP, extensor indicis proprius; ADM, adductor digiti minimi; APB, abductor pollicis brevis; AH, abductor hallucis; PF, popliteal fossa.

[a] Tibial H reflexes: latency <35 ms; side-to-side difference <1.4 ms.

[b] Sensory studies are performed antidromically; amplitudes are measured from baseline to negative peak of nerve potential.

Table 45-1

Disease processes that preferentially injure the fastest-conducting, large-diameter fibers in peripheral nerves reduce the maximal conduction velocity because the remaining thinner fibers conduct more slowly. In most neuropathies, all of the axons are affected either by a fairly uniform “dying-back” phenomenon or by wallerian degeneration, and nerve conduction velocities are then less informative. This is true for typical alcoholic-nutritional, carcinomatous, uremic, diabetic, and other metabolic neuropathies, in which conduction velocities range from low normal to mildly slowed. In these so-called “axonal neuropathies,” the motor and sensory nerve amplitudes are diminished By contrast, demyelinating neuropathies of the acute (Guillain-Barre´, diphtheria) and chronic types such as chronic inflammatory, metachromatic leukodystrophy, Krabbe disease, and the common type of Charcot-Marie-Tooth disease show marked slowing of conduction and, in the case of the acquired demyelinating diseases, there is also dispersion of the action potential and conduction block

Amplitude of the Compound Muscle Action Potential In addition to the study of distal latency and conduction velocity, the amplitude of the evoked muscle action potential (CMAP) yields valuable information about peripheral nerve function. These amplitudes are a semiquantitative measure of the number of nerve fibers that respond to a maximal stimulus. Demyelinative lesions or axonal loss affecting the large, fast-conducting fibers may be detected by the finding of differential slowing among various calibre fibers that causes a dispersal of the CMAP response. Reduction in motor and sensory amplitudes is a more specific and sensitive indicator of axonal loss than is slowing of conduction velocity or prolongation of distal latencies. Conversely, prolonged distal latencies and slowed motor conduction velocities—as well as conduction blocks and dispersed responses—are the hallmarks of demyelinative lesions. The range of normal amplitudes for the CMAPs that are elicited by stimulation of the main motor nerves is shown in Table 45-1. It is usually possible to obtain a reliable motor conduction study as long as some functioning nerve fibers remain intact. The conduction velocities then reflect the status of the surviving axons, and the velocity may be normal despite widespread axonal degeneration. This is most apparent following incomplete transection of a nerve; the maximal motor conduction velocity may be normal in the few remaining fibers, although the muscle involved is almost paralyzed and the compound muscle potential recorded from it is very low.

Sensory Nerve Action Potentials When motor fibers in a mixed nerve are stimulated, an amplified CMAP of many hundreds of microvolts can easily be recorded from electrodes on the skin over the muscle. However, when one attempts to measure sensory potentials, where activity must be recorded from nerve fibers themselves, the “amplification” provided by many motor units is not available and electronic amplification is required. Sensory potentials are sometimes very small or absent even when powerful computer- averaging techniques are used, and sensory conduction measurements may then be difficult to determine. Table 45-1 gives the range of normal values for sensory nerve action potential amplitudes and velocities.

Conduction Block By stimulating a motor nerve at multiple sites along its course, it is possible to demonstrate segments in which conduction is partially “blocked” or is differentially slowed. From such data one infers the presence of

a multifocal demyelinative process in motor nerves. This contrasts with the findings in certain of the inherited demyelinating neuropathies, in which all parts of the nerve fiber are altered to more or less the same degree, i.e., there is uniform slowing of conduction and no conduction block. As a technical matter, conduction block is demonstrated by a reduction in the amplitude of the CMAP elicited from the proximal site along the motor nerve, compared to stimulation at a distal site. Generally, a 40 percent reduction in amplitude over a short distance of nerve, or 50 percent over a longer distance, qualifies as a block, one exception being along the tibial nerve, in which it is difficult to stimulate all the motor nerve fibers and in which some drop in amplitude is normally expected. It is also important to be sure that any reduction in amplitude along the course of the nerve is not due solely to dispersion of the waveform. One must also keep in mind that conduction block may be attributable simply to nerve compression at common sites (fibular head, across the elbow, flexor retinaculum at the wrist, etc.) rather than to an intrinsic disease of the peripheral nerves. The presence of a conduction block can also be inferred from the finding of poor recruitment of muscle action potentials in the absence of active denervation As mentioned, the finding of conduction block is a central feature of a number of acquired immune demyelinating neuropathies, including Guillain-Barre´ syndrome, chronic inflammatory demyelinating neuropathy, and multifocal conduction block associated with the GM1 antibody, all of which are discussed in

Focal compression of nerve, as occurs in the entrapment syndromes mentioned earlier, may produce localized slowing or blocks in conduction, perhaps because of segmental demyelination at the site of compression. The demonstration of such localized changes of conduction affords ready confirmation of nerve entrapment; for example, if the distal latency of the median nerve (see A, Fig. 45-3) exceeds 4.5 ms while that of the ulnar nerve remains normal, compression of the median nerve in the carpal tunnel is likely. Similar focal slowing or partial block of conduction may be recorded from the ulnar nerve at the elbow and from the peroneal nerve at the fibular head

Special Electrodiagnostic Studies of Nerve Roots and Spinal Segments (Late Responses, Blink Responses, Segmental Evoked Responses)

H Reflex Information about the conduction of impulses through the proximal segments of a nerve is provided by the study of the H reflex and the F wave. In 1918, Hoffmann, after whom the H reflex was named, showed that submaximal stimulation of mixed motor-sensory nerves, insufficient to produce a direct motor response, induces a muscle contraction (H wave) after a latency that is far longer than that of the direct motor response. This reflex is based on the activation of afferent fibers from muscle spindles (the same axons that conduct the afferent volley of the tendon reflex), and the long delay reflects the time required for the impulses to reach the spinal cord via the sensory fibers, synapse with anterior horn cells, and to be transmitted along motor fibers to the muscle (see Fig. 3-1). Thus the H reflex is therefore the electrical representation of the tendon reflex circuit and is especially useful because the impulse traverses both the posterior and anterior spinal roots. The H reflex is particularly helpful in the diagnosis of S1 radiculopathy and of other polyradiculopathies.

Its status generally parallels that of the Achilles reflex. However, it may be difficult to elicit an H reflex from nerves other than the tibial. Stimuli of increasing frequency but low intensity cause a progressive depression and finally obliteration of H waves. The latter phenomenon has been used to study spasticity, rigidity, and cerebellar ataxia, in which there are differences in the frequency-depression curves of H waves.

F Wave The F response, so named because it was initially elicited in the feet, was first described by Magladery and McDougal in 1950. It is evoked by a supramaximal stimulus of a mixed motorsensory nerve. After a latency longer than that for the direct motor response (latencies of 28 to 32 ms in arms, 40 to 50 ms in legs), a second small muscle action potential is recorded (F wave). The F wave is the result of the impulses that travel antidromically in motor fibers to the anterior horn cells, a small number of which are activated and produce an orthodromic response that is recorded in a distal muscle. The response is a more reliable test than the H wave of proximal nerve and root conduction in that the F wave traverses only the ventral root and can be elicited from any number of muscles. The combination of a normal F response and an absent H reflex is found in diseases of sensory nerves and roots. Both of these "late responses" find their main use as corroborative tests that must be interpreted in the context of the entire nerve conduction examination. The normal late response latencies are given in Table 45-1.

Blink Responses This special nerve conduction test is useful in the diagnosis of certain demyelinating neuropathies and in any process that affects the trigeminal or facial nerve. The supraorbital (or infraorbital) nerve is stimulated

transcutaneously and the reflex closure of both orbicularis oculi muscles is recorded with surface electrodes. Two CMAP bursts are observed: the first (R1) appears ipsilaterally 10 ms after the stimulus and the second (R2) ipsilaterally at 30 ms and contralaterally up to 5 ms later. The amplitudes of the responses vary considerably and are not in themselves clinically important. The first response is not visible as a muscular contraction but may serve some preparatory function by shortening the blink reflex delay. R1 is mediated by an oligosynaptic pontine circuit consisting of one to three neurons located in the vicinity of the main sensory nucleus; R2 utilizes a broader reflex pathway in the pons.

It has been established that R1 and R2 are generated by the same facial motor neurons. The elicitation of blink reflexes establishes the integrity of the afferent trigeminal nerve, the efferent facial nerve, and the interneurons in the pons (R1) and caudal medulla (related to the bilateral R2 response). The test may be also be helpful in identifying a demyelinating neuropathy when the facial and oropharyngeal muscles are affected and those of the limbs are relatively spared, leaving conventional nerve studies normal. In such cases, the blink responses are delayed ipsilaterally and contralaterally as a result of conduction block in the proximal facial nerve. Direct facial nerve stimulation often fails to demonstrate this block because only the distal segment of the nerve is amenable to study. Although this test is rarely necessary for diagnosis, most patients with hereditary neuropathy have blink response abnormalities. In Bell's palsy there is a delay or absence of R1 and R2 responses only on the affected side. Large acoustic neuromas may interfere with the afferent trigeminal portion of the pathway and give rise to abnormal responses on the affected side. Diseases of the brainstem have yielded inconsistent responses. It is noteworthy that the test is normal in patients with trigeminal neuralgia.

Segmental Motor, Cranial, and Somatosensory Evoked Potentials These techniques find use in diseases that affect spinal roots. By applying a magnetic stimulus, which induces an electrical impulse, or by a directly delivered electrical stimulus over the lower cervical or lumbar spine, it is possible to activate the motor (anterior) roots and to measure the time required to elicit a muscle contraction (Cros and Chiappa). These root stimulation tests can be quite uncomfortable for the patient because of the contraction of muscles

surrounding the stimulation site. Transcranial magnetic stimulation of the cerebral cortex permits measurement of the latency of muscle contraction after excitation of motor neurons in the cortex. Thus, the integrity of the entire corticospinal system, from the cortical motor neurons through spinal tracts, anterior horn cells, and the peripheral motor nerve can be determined. By combining this technique with the previously described root stimulation, it becomes possible to measure central and peripheral motor conduction times. These formsof testing have their main use in the study of amyotrophic lateral sclerosis (ALS) and related disorders. By applying repetitive electrical stimuli to a peripheral nerve, the sensory evoked responses can be recorded from sites along the nerve and plexus as well as in central pathways (the thalamus and somatosensory cortex). These evoked potential tests find their main use in the diagnosis of multiple sclerosis and in disorders of the sensory nerve roots . Other special nerve conduction techniques are available in many laboratories. For details of their performance and interpretation the reader is referred to specialized texts on the subject. Discussion of magnetic stimulation, collision techniques, quantitative EMG, etc., can be found in several monographs, such as the ones by Kimura, by Aminoff, and by Brown and Bolton

Repetitive Motor Nerve Stimulation (Jolly Test)

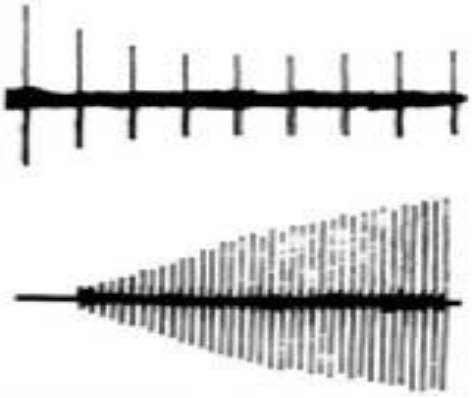


Figure 45-4. Compound action potentials evoked in hypothenar muscles by electrical stimulation of the ulnar nerve at the wrist. A. Patient with myasthenia gravis—typical pattern of decrement in first four responses followed by slight increment. At this rate of stimulation (4 per second), the decrement in response does not continue to zero. B. Patient with Lambert- Eaton syndrome and oat-cell carcinoma—typical marked increase toward normal amplitude with rapid repetitive stimulation (20 per second). Horizontal calibration: 250 ms.

This test of the function of the neuromuscular junction is based on Jolly's observation in 1895 that in myasthenia gravis the strength of muscular contractions progressively declines in response to a train of stimuli. By adjusting the amplitude of a stimulus over a nerve to supramaximal range, a maximal CMAP may be obtained for each stimulus. With repeated stimuli, each response will have the same waveform and amplitude until fatigue supervenes.

In a healthy individual, the response follows each stimulus even with rates of stimulation up to 25 per second for periods of 60 s or more before a decrement of the CMAP appears. In certain disorders, notably myasthenia gravis, a train of 4 to 10 stimuli at rates of 2 to 5 per second (optimally 2 to 3 per second), the amplitude of the motor potentials decreases and then, after four or five further stimuli, may increase slightly (Fig. 45-4A). A progressive reduction in amplitude is most likely to be found in proximal muscles, but theseare not easily stimulated for which reason the locations most commonly used for clinical testing are the accessory nerve in the posterior triangle of the neck (trapezius), the ulnar nerve (hypothenar muscle), the median nerve at the wrist (thenarmuscle), and the facial nerve and orbicularis oculi muscle.

A decrement of 10 percent or more denotes a failure of a proportion of the neuromuscular junctions that are being stimulated. The sensitivity of the procedure is improved by first exercising the tested muscle for 30 to 60 s. The induced failure of neuromuscular transmission in myasthenia is similar to the one produced by curare and other nondepolarizing neuromuscular blocking agents, and both cases can be partially corrected with anticholinesterase drugs such as neostigmine and edrophonium. Similar but lesser decremental responses may occur in poliomyelitis, ALS, and certain other diseases of the motor unit or motor nerve, particularly those resulting in the growth of reinnervating nerve twigs. The myasthenic syndrome of Lambert-Eaton, often associated with oat-cell carcinoma of the lung, is characterized by a presynaptic blockage of acetylcholine release and produces the opposite defect of neuromuscular transmission to the one recorded in myasthenia gravis.

During tetanic stimulation (20- to 50-per-second repetitive stimulation of nerve), the muscle action potentials, which are small or practically absent with the first stimulus, increase in voltage with each successive response until a more nearly normal amplitude is attained (see Fig. 45-4B). Exercising the muscle for10 s before stimulation will cause a similar posttetanic facilitation (200-fold increases are not uncommon). A less important decremental response to slow stimulation may occur, but it is difficult to discern because of the greatly diminished amplitude of the initial responses. Neostigmine has little effect on this phenomenon, but it may be reversed by guanidine and 3,4-

diaminopyridine, which stimulate the presynaptic release of ACh. The effects of botulinum toxin and of aminoglycoside antibiotics are similar, i.e., being active at the presynaptic membrane; they produce an incremental response at high rates of stimulation. The single-fiber EMG, discussed in a later section, is an even more sensitive method of detecting failure of the neuromuscularjunction.

Needle Examination of Muscle (Electromyography)

This technique requires the use of monopolar or concentric bipolar needle electrodes, which are inserted into the body of the muscle to record the electrical activity generated by contraction. With concentric needle electrodes, the tip of the wire that runs in the hollow of the needle is in proximity to many muscle fibers belonging to several different overlapping motor units; this is the active recording electrodeThe shaft of the needle, in contact over most of its length with intercellular fluid and many other muscle fibers, serves as the reference electrode. With monopolar electrodes, the uninsulated needle tip is the active electrode, while the reference electrode may be another monopolar needle electrode placed in subcutaneous tissue or a surface electrode on the skin overlying the muscle. Patients almost invariably find this portion of the test uncomfortable and should be prepared by a description of the procedure. Rapid and brief needle insertion by the skilled examiner makes the test more tolerable.

As the electrical impulse travels along the surface of the muscle toward the recording electrode, a positive potential is recorded on the oscilloscope, i.e., the recorded signal is deflected downward by convention (at A in Fig. 45-5). When the depolarized zone moves under the recording electrode, it becomes relatively negative and the beam is deflected upward (at B). As the depolarized zone continues to move along the sarcolemma, away from the recording electrode, the current begins to flow outward through the membrane toward the distant depolarized region, and the recording electrodebecomes relatively positive again (at C).

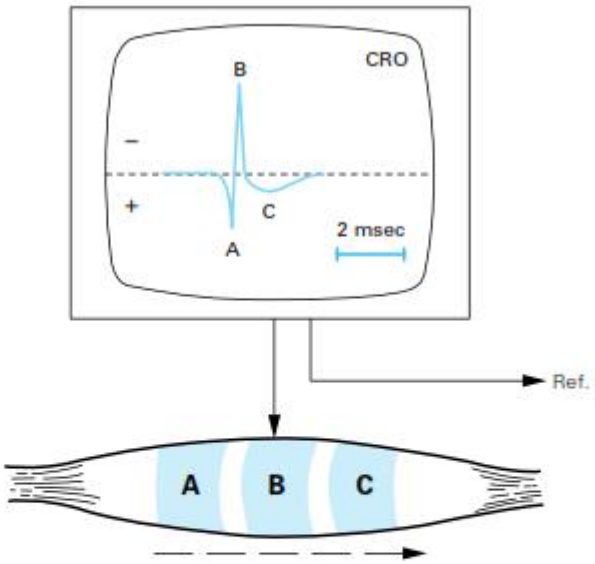


Figure 45-5. The shaded area represents the zone of the action potential, which is negative to all other points on the fiber surface. It is shown at three points in its course (*from left to right*) along the fiber. At each point, the correspondingly lettered portion of the triphasic muscle action potential displayed on the display screen reflects the potential difference between the active (*vertical arrow*) and reference (Ref.) electrodes. Polarity in this and subsequent figures is negative upward as depicted. The time calibration is on the screen.

It then returns to its resting isopotential position. The net result is a triphasic action potential, as in Fig. 45-5. This configuration is typical of the firing of a single fiber. The electrical activity of various muscles is recorded both at rest and during active contraction by the patient. As indicated earlier, muscle fibers do not normally discharge until activated together in motor unit activity. This involves the almost simultaneous contraction of all the muscle fibers innervated by a single anterior horn cell. Although the typical configuration of a motor unit potential (MUP) is triphasic, up to 10 percent of normal MUPs consist of four or more phases (polyphasic potentials); an excess of polyphasic potentials beyond this is pathologic. Normal muscle in the resting state should be electrically silent; the small tension spoken of as muscle tone has no EMG equivalent. There are, however, two closely related types of normal spontaneous activity and another that is induced by the insertion of the needle itself. One is a low-amplitude, 10- to 20-mV monophasic (negative) potential of very brief (0.5 to 1 ms) duration. These represent single or synchronized MEPPs due to the small number of ACh quanta that are being released all the time. They are normally sparse but are most evident when the recording needle electrode is placed near a motor end plate (“end-plate noise”).

Fortuitous placement of the needle electrode very close to or in contact with the end plate gives rise to a second type of normal spontaneous activity.

That is characterized by irregularly discharging high frequency (50- to 100-Hz) biphasic spike discharges, 100 to 300 mV in amplitude (i.e., large enough to cause an isolated muscle action potential). These potentials have been termed end-plate spikes and represent discharges of single muscle fibers excited by spontaneous activity in nerve terminals. They must be distinguished from fibrillation potentials finally; insertion of the needle electrode into the muscle injures and mechanically stimulates many fibers, causing a burst of potentials of short duration (300 ms). This is referred to as normal insertional activity. When muscle is voluntarily contracted, the action potentials of motor units begin to appear. One can observe a pattern of force build up by watching the progressive recruitment of MUPs; the initial ones, representing smaller motor units, firing at rates of 5 to 10 per second. With increased force of contraction, there is recruitment of larger, previously inactive motor units as well as an increased rate of firing (40 to 50 per second; Fig. 45-6A). Since individual MUPs can no longer be distinguished during maximal voluntary contraction, this activity is referred to as a complete interference pattern (Fig. 45-6A, right). This is seen not only as a summated signal pattern but is also heard as a mixed high frequency clicking when the electrical activity is made audible. As muscles relax, more and more units drop out. If a muscle is weakened by denervation or if electrical conduction is blocked, there will obviously be fewer MUPs, but the firing rate is still rapid (reduced recruitment, see Fig. 45- 6B). In contrast, with poor voluntary effort and with upper motor neuron lesions, the MUPs fire in decreased numbers, at slower rates, and often in an irregular pattern (termed poor activation). In the usual EMG examination, a plan for the study is made based on detailed knowledge of muscular innervation and focusing on the regions affected by weakness.

The Abnormal Electromyogram

Clinically important deviations from the normal EMG include

- increased or decreased activity upon insertion of the needle
- the occurrence of abnormal “spontaneous” activity during the relaxed state (fibrillation potentials, positive sharp waves, fasciculation potentials, cramp potentials, myotonic discharges, myokymic potentials)
- Abnormalities in the amplitude, duration, and shape of single MUPs

- a decrease in the number of MUPs and changes in their firing pattern
- variation in amplitude and number of phases of MUPs during voluntary contraction of muscle
- The demonstration of special phenomena, such as electrical silence during obvious shortening of the muscle (physiologic contracture).

Insertional Activity At the moment the needle is inserted into muscle, there is a brief burst of action potentials that ceases once the needle is stable, provided that it is not in a position to irritate a nerve terminal. Increased insertional activity is seen in most instances of denervation as well as in many forms of primary muscle disease and in disorders that dispose to muscle cramps.

Abnormal "Spontaneous" Activity With the muscle at rest, spontaneous activity of single muscle fibers and of motor units, known respectively as fibrillation potentials and fasciculation potentials, is abnormal. The two phenomena are often confused. Fibrillation is the spontaneous contraction of a single muscle fiber. It occurs when the muscle fiber has lost its nerve supply and is ordinarily not visible through the skin (but may be visible in the tongue). Fasciculation represents the spontaneous firing of an entire motor unit, causing contraction of a group of muscle fibers, and may be visible through the skin. The irregular firing of a number of motor units, seen as a rippling of the skin, is called myokymia

Fibrillation Potentials When a motor neuron is destroyed by disease or its axon is interrupted, the distal part of the axon degenerates, a process that takes several days or more. The muscle fibers formerly innervated by the branches of the dead axon—that is, the motor unit—are disconnected from the nervous system. For reasons that are still obscure, the chemosensitive region of the sarcolemma at the motor end plate "spreads" after denervation to involve the entire surface of the muscle fiber.

Figure 45-7. A. Fibrillations and positive sharp waves. This spontaneous activity was recorded from a totally denervated muscle—no motor unit potentials were produced by attempts at voluntary contraction. The fibrillations (above arrow) are 1 to 2 ms in duration, 100 to 300 mVin amplitude, and largely negative (upward) in polarity following an initial positive deflection. A typical positive sharp wave is seen above the star. B. Fasciculation. This spontaneous motor unit potential was recorded from a patient with amyotrophic lateral sclerosis. It has a serrated configuration and it fired once every second or two. Calibrations: 5 ms (horizontal) and 200 mV in A; 1 mV in B (vertical).

Fasciculation Potentials As stated earlier, a fasciculation is the spontaneous or involuntary contraction of a motor unit or part of a motor unit. Such contractions may cause a visible dimpling or twitching under the skin, though ordinarily they are of insufficient force to move a joint. Large distal fasciculations, however, can briefly displace a finger or toe; they occur irregularly and infrequently, and prolonged inspection of the skin overlying a muscle may be necessary to detect them

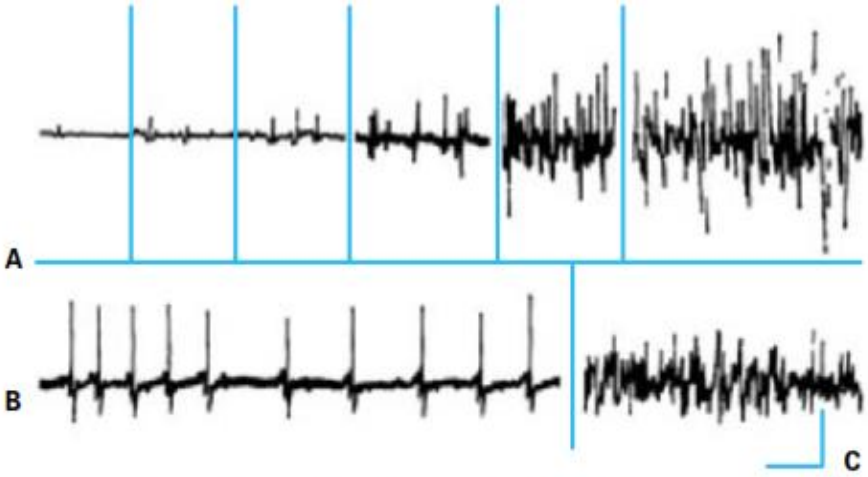


Figure 45-6. Patterns of motor unit recruitment. *A*. Normal. With each increment of voluntary effort, more and larger units are brought into play until, with full effort at the extreme right, a complete "interference pattern" is seen in which single units are no longer recognizable. *B*. After denervation, only a single motor unit is recorded despite maximal effort. It is seen to fire repetitively. *C*. With myopathic diseases, a normal number of units is recruited on minimal effort, though the amplitude of the pattern is reduced. Calibrations: 50 ms (horizontal) and 1 mV in *A* and *B*; 200 mV in *C* (vertical).

Less Common Types of Spontaneous Electrical Activity One of these is myokymia ,a persistent quivering and rippling of muscles at rest ("live flesh"). The EMG picture is distinctive. The spontaneously firing MUPs are called myokymic potentials or discharges and consist of groups of repetitive discharging units, each firing at its own rate, quasirhythmically, usually several

times per second, followed by an even briefer period of silence. The small motor unit discharges may occur singly or as doublets, triplets, or multiplets.

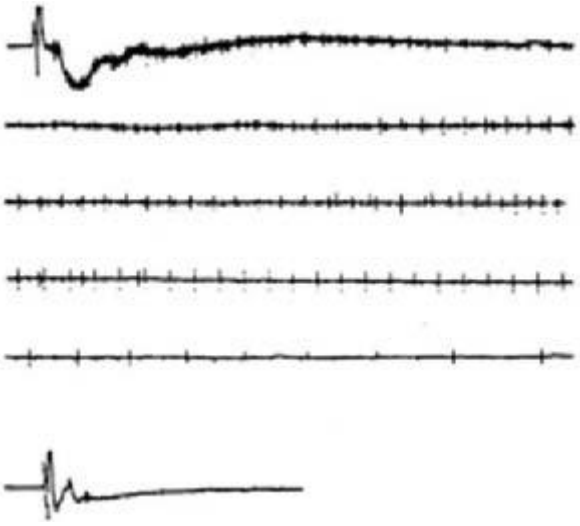


Figure 45-8. A. Myotonia congenita (Thomsen disease). The five lines are a continuous record of activity in the biceps brachii following a tap on the tendon. The initial response is within normal limits, but it is followed by a prolonged burst of rapid activity, gradually subsiding over a period of many seconds or minutes. B. Same electrode placement as in A. Response to the fifth of a series of tendon taps. "Warm-up" has occurred, and the characteristic prolonged myotonic activity is no longer evident.

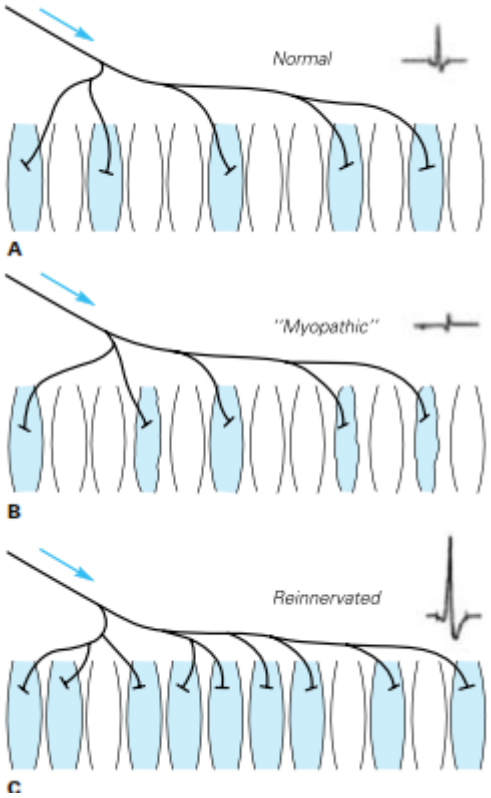


Figure 45-9. The shaded muscle fibers are functional members of one motor unit, whose axon enters from the upper left and branches terminally to innervate the appropriate muscle fibers. The action potential produced by each motor unit is seen in the upper right: its duration is measured between the two vertical lines. The normal-appearing but unshaded fibers belong to other motor units. *A*. Hypothetical situation, with five muscle fibers in the active unit. *B*. In this myopathic unit, only two fibers remain active; the other three (shrunken) have been affected by one of the primary muscle diseases. *C*. Four fibers which originally belonged to other motor units and had been denervated have now been reinnervated by terminal sprouting from an undamaged axon. Both the motor unit and its action potential are now larger than normal. Note that only under these abnormal circumstances do fibers in the same unit lie next to one another.

Abnormalities in Amplitude, Duration, and Shape of Motor Unit Potentials Figures 45-9 (schematically) and 45-10 depict the ways in which disease processes affect the motor unit and the appearance of the MUP in the EMG. **Motor Unit Potentials in Denervation** Early in the course of denervation, many motor units with functional connections to the spinal cord are unaffected, and though the number of MUPs appearing during contraction is reduced, the configurations of the remaining ones are quite normal

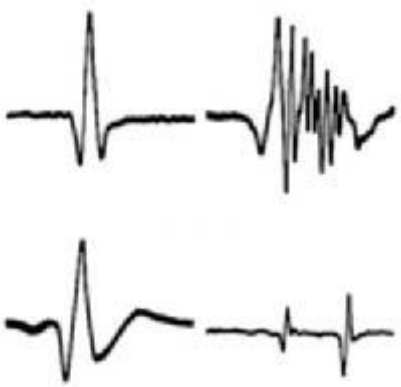


Figure 45-10. Single voluntary motor unit potentials. A. Normal. B. Prolonged polyphasic potential seen with reinnervation. C. "Giant unit normallynshaped but of much greater amplitude than normal. D. Brief, lowamplitude "myopathic" units. Calibrations: 5 ms (horizontal) and 1 mV in A and B; 5 mV in C; 100 mV in D (vertical).

Figure 45-10A,B,C,D

MUPs are to be differentiated from (1) polyphasic potentials of normal duration, which, as has been mentioned, make up as much as 10 percent of the total number of MUPs in normal muscle, and (2) polyphasic MUPs of short duration and low amplitude, which are characteristic of most myopathies and of myasthenia gravis and other disorders of neuromuscular transmission

The Motor Unit Potential in Myopathy Diseases such as polymyositis, the muscular dystrophies, and other myopathies that randomly destroy muscle fibers or render them nonfunctional obviously reduce the population of muscle fibers per motor unit, as shown in Fig. 45-9B.

Abnormalities of the Interference Pattern Diseases that reduce the population of functional motor neurons or axons within the peripheral nerve decrease the

number of motor units that can be recruited in the affected muscles. The decreased number of motor units available for activation then produces an incomplete interference pattern, which is manifest by a decreased number of units firing at a moderate to rapid rate

Motor Unit Counting This experimental technique, developed by McComas and colleagues, estimates the size of motor units and is thus exquisitely sensitive to changes of denervation and reinnervation. It is carried out by applying a weak stimulus to a motor nerve or motor point and increasing it gradually as the evoked muscle response is recorded

Single-Fiber Electromyography (SF-EMG) This is a special technique for the recording of single-muscle-fiber action potentials and is used to measure fiber density and so-called jitter. Fiber density is an index of the number and distribution of muscle fibers within a motor unit. Jitter is the variability of the interpotential interval of successive discharges of two single-muscle-fibers belonging to the same motor unit. This phenomenon is due largely to the very slight variability of delay at the branch points in the distal axon and by synaptic delay at the neuromuscular junction

Imaging of Muscle and Nerve

Imaging techniques—computed tomography (CT), magnetic resonance imaging (MRI), and ultrasound—enable one to measuremuscle volume and to recognize qualitative changes in muscle structure (see review of deVisser and Reimers). Such methods are finding increasing clinical and research use in the diagnosis of disorders of muscle and in gauging the effects of treatment. CT scans of dystrophic muscle show foci of decreased attenuation, representing masses of fat cells. The fatty masses spread gradually from multiple foci and eventually replace muscle fibers. The original shape of the muscle is retained; indeed, an enlarged weak muscle containing mostly fat confirms the clinical impression of pseudohypertrophy. In denervative atrophy, the muscles are obviously small and contain multiple punctate areas of decreased attenuation, which represent interstitial fat. Eventually, large portions of chronically denervated muscle may be replaced by fat. Blood, blood products, and calcium deposits are expressed by increased attenuation in CT. This may be helpful in

the diagnosis of muscle trauma, myositis ossificans, and polymyositis. In MR images, fat and bone marrow have a high signal intensity, while fascia, ligaments, and cortical bone lack signal intensity. In T1-weighted images, normal muscle has a low signal and dystrophic muscle, a slightly increased signal; in T2-weighted images, dystrophic muscle has a slightly enhanced signal. Given its sensitivity to these dystrophic changes in muscle, MRI is particularly effective in determining the distribution of muscle involvement in a dystrophy.

Transverse MR sections, for example, can help distinguish the topographic patterns of such disorders as the proximally predominant Becker dystrophy and the distally predominant Miyoshi myopathy or subtle subtypes of Emery-Dreifuss dystrophy (Mercuri). Spectroscopic MRI in metabolically determined myopathies has the capacity to quantitate levels of selected biochemical constituents of muscle, including intracellular pH and levels of metabolic intermediates such as phosphocreatine. This is particularly effective in demonstrating subnormal generation of intracellular acidosis after a limb is exercised in disorders of glycogenolysis and of glycolysis. Some individuals with mitochondrial disease of muscle will demonstrate rapid depletion of energy supplies and profound delays in recovery that can be quantified and used as an endpoint for treatments. New magnetic resonance techniques are also being developed that allow the imaging of nerves.

BIOPSIES OF MUSCLE AND NERVE

Muscle biopsy can be of great diagnostic value, but both surgical and microscopic techniques must be exacting. The muscle chosen for study should be accessible; there should be evidence that it has been affected but not totally destroyed by the disease in question; and it should not have been the site of a recent injection or EMG study, since the trauma of the needles produces focal necrotizing and inflammatory lesions. Muscle biopsy is helpful in distinguishing the following basic disorders in patients with neuromuscular disease.

Muscle Biopsy

- Denervation atrophy. Reduction in the size of muscle fibers with an accompanying enlargement of intact motor units (due to collateral reinnervation) and degenerative changes in some fibers are the main changes of denervation atrophy. Group atrophy denotes enlarged motor units where all the fibers in the group are reduced to the same size; this is typical of progressive denervation. Normally, the fibers of each motor unit are not clustered, so that when grouping occurs it means that some fibers of a denervated unit have been adopted by an adjacent intact motor unit. This change typifies axonal neuropathies and many spinal cord diseases that affect the anterior horn cell. A related change is particularly well shown in histochemical stains for ATPase, phosphorylase, and oxidases, where the normal mosaic pattern of fiber types is altered. The use of these stains reveals fiber type grouping, the most specific histologic evidence of denervation and reinnervation. Here, muscle fibers of similar histochemical type form groups of 15 or more fibers as a result of reinnervation by a single motor neuron. The diagnosis of denervation atrophy can usually be made from the clinical and EMG examinations; seldom is biopsy necessary for this purpose, but it is still utilized in cases of possible ALS, for example, where the diagnosis remains uncertain after other testing.
- Segmental necrosis of muscle fibers with myophagia and various manifestations of regeneration. These are the typical changes in idiopathic polymyositis (in combination with infiltrates of inflammatory cells), and infective polymyositis (in the presence of Trichinella, Toxoplasma). These changes may also be observed in more limited form in Duchenne and other rapidly progressive muscular dystrophies.
- Inflammation and vasculitis. Lymphocytic infiltration of the endomysium is most characteristic of polymyositis and in dermatomyositis it may be predominantly perimysial. The lymphocytic infiltrate is often florid in these two processes, whereas it tends to be less intense in inclusion body myositis. Lesser degrees of inflammation are common in the myopathies associated with Sjo¨gren syndrome, mixed connective tissue disease, and scleroderma. Numerous other processes—including the infections mentioned earlier and some dystrophies (especially the

fascioscapulohumeral type)—may be associated with an inflammatory reaction. There is usually acute myofibrillar destruction in regions of maximal lymphocytic infiltration.The muscle is a frequent site of inflammatory vascular destruction (vasculitis) in systemic diseases such as polyarteritis nodosa, and for this reason it is often useful to obtain a small sample of muscle adjacent to a nerve biopsy. The finding of a granulomatous myopathy may indicate the presence of systemic sarcoidosis.

- Alterations in the protein and histochemical composition of muscle fibers may be shown by special stains for enzymes, glycogen, and structural proteins that are implicated in disease. For example, it has become possible to detect the absence or deficiency of specific structural proteins of the muscle membrane that define each of the muscular dystrophies: dystrophin, sarcoglycan, laminin These tests require rapid freezing (in a cryostat) rather than formalin fixation. Also, a number of enzymatic deficiencies and intrafiber glycogen storage that lead to weakness and muscle fatigue may be detected by appropriate histochemical staining .
- Unusual changes of muscle fibers. Included here are sarcoplasmic masses and disorganized ring or serpentine collections of myofibrils and myofilaments (Ringbinden) in myotonic dystrophy; glycogen masses in glycogen storage diseases, rods (nemaline), central cores, aggregates of lipid bodies, and other cytoplasmic changes in certain congenital myopathies; and nuclear and cytoplasmic inclusions (and other changes) that characterize inclusion-body myositis. Application of histochemical methods and electron microscopy are the important techniques in the diagnosis of these disorders.
- Abnormalities of mitochondria. Several distinctive abnormalities are readily visualized in muscle biopsies and are virtually diagnostic of an entire class of mitochondrial diseases. Light microscopy of frozen sections of muscle stained with the Gomori trichrome stain show the main feature, so-called ragged red fibers, which are accumulations of subsarcolemmal mitochondria.
- Disorders of the neuromuscular junctions in which nerve fibers and muscle fibers appear to be intact. Here an abnormality may be revealed

by performing the demanding procedure of motor point biopsy (to include the motor end plate), and using electron microscopy and special staining techniques for nerve terminals, cholinesterase, and the outlining of acetylcholine receptors. Myasthenia gravis, botulism, Lambert-Eaton syndrome, and myasthenic syndrome with motor end-plate cholinesterase deficiency fall into this category.Biopsy is rarely necessary for the diagnosis of these disorders but it has added considerably to our understanding of them.

As a rule, the muscle biopsy procedure requires no more than a small cleanly excised block of muscle, 1.0 to 2.0 cm, which is prevented from contracting by a clamp or by tying at full length to a tongue depressor.

Nerve Biopsy Nerve biopsies are processed for study by conventional histologic methods and by thin-section phase and electron microscopy. These methods, sometimes supplemented by study of teased fiber preparation and immunologic studies, can provide valuable histopathologic data. With ordinary microscopy and staining, one may find evidence of focal inflammation, vasculitis, amyloidosis, leprosy, or sarcoid. These procedures are most valuable in the diagnosis of inflammatory, vasculitic, and amyloid neuropathies. In children, the nerve biopsy may reveal the histologic features of metachromatic or globoid leukodystrophy, giant axonopathy, or neuroaxonal dystrophy. However, nerve biopsy is relatively unhelpful in most other polyneuropathies and should be used prudently because the procedure is not without complications, such as the occasional occurrence of wound infections, painful stump neuromas, persistent dysesthesias of the lateral foot or heel, and thrombophlebitis. Indeed, in a prospective study of 50 consecutive cases with sural nerve biopsy reported by Gabriel and colleagues, nerve biopsy served only to confirm the clinical diagnosis in 70 percent of cases and altered the diagnosis in only 14 percent. Nevertheless, in some instances of idiopathic polyneuropathy that are not clarified by clinical and electrophysiologic testing, and particularly if a treatable chronic inflammatory polyneuropathy is suspected, many neuromuscular experts resort to sural nerve biopsy as a final diagnostic step. Although the sural nerve is typically chosen for biopsy, the superficial peroneal nerve and the adjacent peroneus brevis muscle gives a higher

yield in cases of vasculitis. In diseases that involve only the motor nerves, it is sometimes useful to sample a fascicle of the superficial radial nerve or the nerve to the extensor digitorum brevis, which may be taken with the muscle itself. Chronic inflammatory neuropathy and vasculitic neuropathy may be disclosed by this type of biopsy when the sural nerve is unaffected. Also, in selected circumstances we have undertaken biopsy of small radicles of upper lumbar roots (L1 or L2) to establish the diagnosis of an infiltrative lymphoma. Little deficit occurs from the removal of nerves from these alternative sites if the procedure is done by an experienced surgeon.

Other Laboratory Tests in the Study of Muscle and Nerve Disease None of the diagnostic procedures previously described may be taken as an infallible diagnostic index of a specific disease of muscle or nerve. Each procedure is subject to technical error and the findings to misinterpretation. A biopsy specimen may be taken from an unaffected muscle or portion of a muscle, and such a sampling error causes the sample to be normal in the face of obvious clinical evidence of disease. This is particularly true of the inflammatory myopathies that affect muscle in heterogeneous and spotty manner. Rough excision and improper fixation and staining may produce artifacts that may be misinterpreted as marks of disease when, in fact, the muscle (and nerve) is microscopically normal. Similarly, EMG study may fail to record fibrillations in obviously denervated muscle, particularly in slowly progressive disorders.

Also, in some of the muscles of the feet, fibrillations and fasciculations may be found in normal asymptomatic older individuals (Falck and Alaranta). As in the study of all disease, laboratory data have significance only if evaluated in the light of the clinical findings.

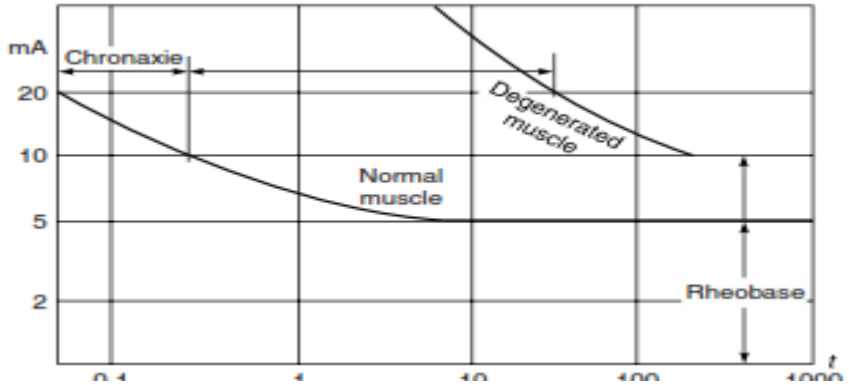
ELECTRODIAGNOSTIC /THERAUPETIC APPARATUS:

Electrodiagnosis:

If a normal muscle or motor nerve is stimulated with a current of adequate intensity, it results in its contraction. When there is disease or injury of a motor nerve or muscle, alterations are liable to occur in their response to electrical stimulation. The changed electrical response may be of considerable help in the diagnosis of certain diseases affecting them. Quantitatively, these changes

manifest themselves in that a higher or lower current intensity than normal is required to bring about a muscle contraction. It is, therefore, possible to determine the degeneration and regeneration processes in nerves and the muscle system by the use of the stimulation current technique.

Intensity-Time Curve (i-t Curves): In order to examine the conditions of excitability and to obtain a good picture of the degeneration and regeneration process of neuro-muscular units, modern stimulation current diagnosis plots the so called i-t curves based on the intensity of the stimulus and its duration. These curves are determined by means of rectangular and triangular pulses in such a manner that the threshold values are measured at progressively decreasing stimulation durations. The i-t curves have characteristic shapes and deviations from the standard form lead to an indication of the state of the tissues. In order to plot such curves, the tissue (muscle, nerve) to be examined is first stimulated
with long impulses (usually of 1 s pulse duration and then with shorter and shorter impulses, (down to say, 0.05 ms). For each impulse duration, the current intensity is adjusted until the stimulation threshold has been exceeded and the effect of the stimulation detected. Obviously, the current intensity has to be increased. The impulse duration is usually varied in stages such as 1000–300–l00–10–3–1–0.05 ms and the control of current is effected by a continuously adjustable resistance.



Typical intensity time curves of a normal muscle and degenerated muscle. The curve shows that decreasing excitability with progressive degeneration requires extended stimulation times and increased current strength for achieving successful stimulation

With degenerated muscle, the curve obtained is shifted to the right and upwards. The intermediate stages of degeneration and regeneration are characterized by curves lying in between these two limits.
The chronaxie and rheobase can be easily read from the i-t curves. The rheobase is the minimum intensity of current that will produce a response if

the stimulus is of infinite duration, in practice an impulse of 100 ms being adequate for estimating this. The chronaxie is the minimum duration of impulse that will produce a response with a current of double the rheobase. For example if the rheobase is 6 mA, the chronaxie is the duration of the shortest impulse that will produce a muscle contraction with a current of 12 mA. i-t curves with exponentially progressive current impulses can be drawn in the same way as rectangular impulses. The two curves differ considerably. A typical characteristic of these curves is that the stimulation threshold first decreases when the pulse duration is reduced and the rheobase is also missing. This is due to the phenomenon of the accommodability of the neuromuscular units.

Accommodation: Accommodation is the property of a neuro-muscular unit of being able to respond less strongly to a slowly increasing current impulse. In other words, the units exhibit a lower excitability and a higher stimulation threshold. The importance of accommodation from the diagnostic point of view lies in the effect that it gives an indication of the presence or alteration of a state of degeneration. In the representation of i-t curves, the determination of the accommodation consists of the comparison between the 100 ms points of the rectangular i-t curves and of the triangular i-t curves and is, in a way, analogous to the determination of the chronaxie, which is essentially a comparison of the two points of the rectangular i-t curves.

Electrotherapy: Electrotherapy, employing low-volt, low-frequency impulse currents, has become an accepted practice in the physiotherapy departments. The biological reactions produced by low-volt currents have resulted in the adoption of this therapy in the management of many diseases affecting muscles and nerves. The technique is used for the treatment of paralysis with totally or partially degenerated muscles, for the treatment of pain, muscular spasm and peripheral circulatory disturbances, and for several other applications.

Although some of the principles upon which low-volt therapy depends have been known since the end of the last century, it is only in recent years that it has started being widely used with the availability of safe and simplified apparatus required for the purpose.

Different types of waveforms are used for carrying out specific treatments. The most commonly used pulse waveforms are discussed below.

Galvanic Current: When a steady flow of direct current is passed through a tissue, its effect is primarily chemical. It causes the movement of ions and their collection at the skin areas lying immediately beneath the electrodes. The

effect is manifested most clearly in a bright red coloration which is an expression of hyperaemia (increased blood flow). Galvanic current is also called direct current.

Galvanic current may be used for the preliminary treatment of atonic paralysis and for the treatment of disturbance in the blood flow. It is also used for iontophoresis, which means the introduction of drugs into the body through the skin by electrolytic means. In general, the intensity of the current passed through any part of the body does not exceed 0.3 to 0.5 ma/sq cm of electrode surface. The duration of the treatment is generally 10–20 minutes.

Faradic Current: Faradic current is a sequence of pulses with a defined shape and current intensity. The pulse duration is about 1 minutes with a triangular waveform and an interval duration of about 20 minutes. Faradic current acts upon muscle tissue and upon the motor nerves to produce muscle contractions. There is no ion transfer and consequently, no chemical effect. This may be used for the treatment of muscle weakness after lengthy immobilization and of disuse atrophy.

Surging Current: If the peak current intensity applied to the patient increases and decreases rhythmically, and the rate of increase and decrease of the peak amplitude is slow, the resulting shape of the current waveform is called a surging current. The main field of application of the Faradic surge current is in the treatment of functional

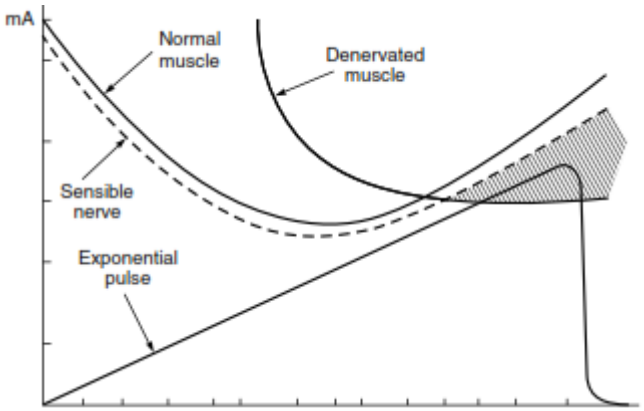
paralysis. The surge rate is usually from 6-60 surges per minute in most of the instruments. The ratio of interval to the duration of the surging is also adjustable so that graded exercise may be administered. This type of current is usually required for the treatment of spasm and pain.

Exponentially Progressive Current: This current is useful for the treatment of severe paralysis. The main advantage of this method lies in the possibility of providing selective stimulation for the treatment of the paralysed muscles. This means that the surrounding healthy tissues even in the immediate neighbourhood of the diseased muscles are not stimulated. The slope of the Exponential pulse is kept variable.

Biphasic Stimulation: The cell recovery from the effect of a stimulus current can be hastened by the passage of a lower intensity current of opposing polarity over a longer period so that the net quantity of electricity is zero. Such type of combination of positive and negative pulses is called biphasic stimulation. In a typical case, the stimulating pulse may be followed by a pulse of opposite polarity of one-tenth the amplitude and 10 times the width. Biphasic stimulation also helps to neutralize the polarization of the recording electrodes in case silver-silver chloride electrodes are not used. This means that there are no electrolytic effects, nor are any macroscopic changes

affecting either the skin or the electrodes observed. Also, there is reduced muscle fatigue, since each currentpulse is immediately followed by an opposite current phase of the same magnitude. The stimulation current intensity required during treatment is less as compared with monophasic currents.
Monophasic current forms, however, retain their importance in electro-diagnostic evaluation since the necessary pulse shapes are defined mono-phasically.

Principle of selective stimulation of the denervated musculature. Selective stimulation of the denervated muscle without irritation of the sensible receptors is possible in the shaded area of the graph.

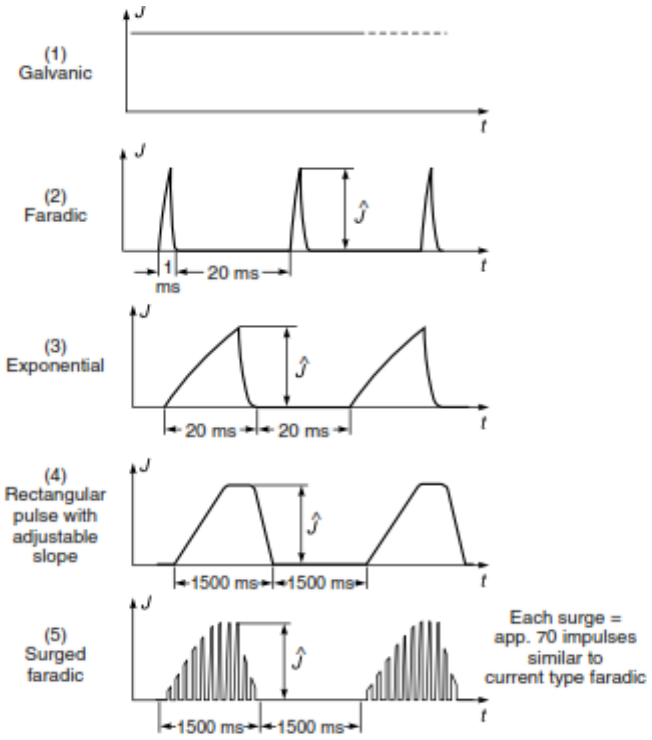


Types and functional block diagram:

Several types of commercial units are available which give specific output waveforms for specific applications. However, the trend is in favour of having a versatile apparatus which gives output current waveforms to cover the whole range of electro-diagnostic and therapeutic possibilities. The output waveforms generally required are shown in Fig Another important consideration is that the apparatus must be either of constant voltage or
constant current type. An instrument of indefinite intermediate principle can lead to unreliable results. Moreover, the apparatus must give reproducible and well-defined impulses that must correspond to the value set on the dials. For clinical practice, maximum tolerance permitted in the
pulse parameters is 15%. The instrument generally has a floating output and incorporates an isolation transformer in the output.
The typical specifications of an electro-diagnostic therapy unit are as follows:
• Galvanic current up to 80 mA, ripple less than 0.5% as constant current or surging current with adjustable surge frequency from 6 to 30 surges per minute
• Exponentially progressive current pulse sequences with continuously variable pulse duration from 0.01 to 1000 ms and independently adjustable interval

duration of 1 to 10,000 ms. The pulse form can be set continuously between triangular and rectangular forms.

• Faradic surging current with 25 surges per minute, upto 80 mA. Precision and constancy of the values set better than ±10%; peak current measurement facility. Constant current circuit, both poles earth-free.



➤ **Fig. 29.9** *Current waveforms normally employed in electrodiagnosis and electrotherapy*

Functional block diagram description:

The figure shows the block diagram of a versatile electro-diagnostic therapeutic stimulator. It makes use of a variable rate multi-vibrator (M1) to set the basic stimulus frequency. The output from the free running multi-vibrator triggers a monostable multi-vibrator (M2) circuit which sets

the pulse width. The output pulse from the monostable provides an interrupted galvanic output whose rate as well as duration can be independently controlled. Another astable multi-vibrator produces short duration pulses called faradic currents. Faradic currents are usually modulated at the frequency set by the multi-vibrator Ml, in a mixer circuit (M4). Since the modulation of Faradic pulses takes place with a slow rate of increase and decrease, the output of M4 is surged Faradic currents. By integrating the output of M2, the interrupted galvanic pulses can be modified to havean exponential rise and fall. The shape of these pulses is similar to a triangular waveform. Galvanic current is also made available by suitably tapping the DC supply. Finally, any one of the waveforms can be selected through a selector switch and fed either to an emitter-follower stage in order to provide a low

output impedance constant voltage output or to a high output impedance constant current stage. Usually the output impedance of a constant voltage stimulator is of the order of 100 W and that of a constant current type is greater than 100 kW. The output of a diagnostic/therapeutic stimulator is kept floating, i.e. it is isolated from earth. The usual method is to have an isolation transformer at the output of the stimulator. This transformer has floating terminals and is fitted with an electrostatic shield to reduce capacitive coupling with the earth. Another method of isolation of the output from earth is by the use of a radio frequency output stage.

The two methods have been widely used for providing isolation of the stimulator output, but they have some drawbacks. The simple transformer cannot transmit square waves without distorting the waveform and the method of radio-frequency is rather complex. Isolation can also be provided through the opto-isolation technique.

The question as to which type of stimulation impulses, whether constant voltage or constant current type, should be preferred in carrying out electro-diagnostic studies is still a matter of choice.However, most of the present-day instruments are of the constant current type because for a given electrode geometry, constant current stimulation will provide better reproducibility for a wide variation in preparation impedance.

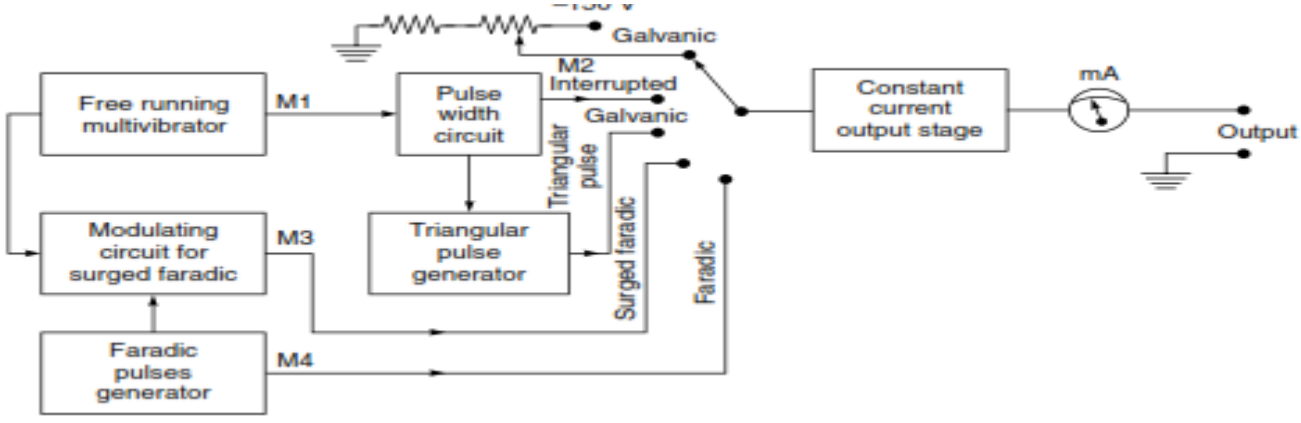
The advantages of constant current therapy are detailed below:

• The current flow is largely constant irrespective of the patient's resistance. The selected current intensity remains constant, even if the resistance in the tissue between the electrodes should vary, as a result of, say, changes in the blood circulation during treatment, or

After previous therapy.

• The current waveform is applied, and distortion-free, since micro-voltages between the electrode and the skin have no influence.

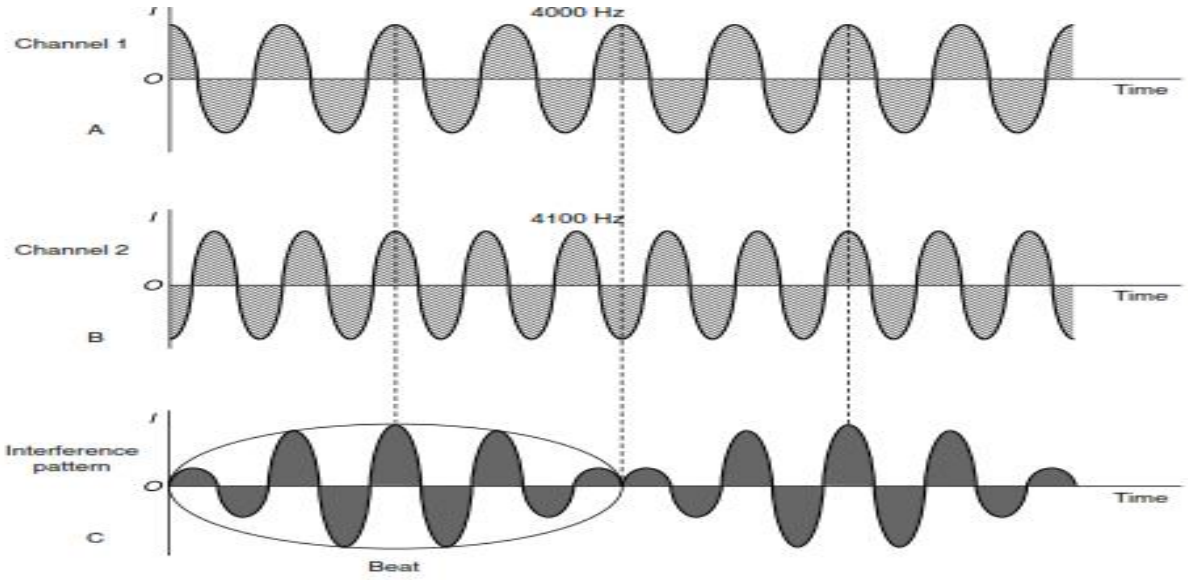
Schematic diagram of a diagnostic/therapeutic stimulating unit



• Current therapy avoids accompanying symptoms such as irritating stimulatory sensationsbetween electrodes by applying electrodes firmly to the skin and keeping them in one position.

Interferential current therapy:

Interferential electrical stimulation is a unique way of effectively delivering therapeutic currents to tissue. Conventional TENS (Transcutaneous Electrical Nerve Stimulator) and neuro-muscular stimulators use discrete electrical pulses delivered at low frequencies of 2–200 Hz. However, interferential stimulators use a fixed carrier frequency of 4,000 Hz and also a second adjustable frequency of 4,001–4,400 Hz. When the fixed and adjustable frequencies combine (heterodyne), they produce the desired beat frequency or interference frequency Interferential stimulation is concentrated at the point of intersection between the electrodes. This concentration occurs deep in the tissues as well as at the surface of the skin. Conventional TENS and neuromuscular stimulators deliver most of the stimulation directly under



electrodes. Thus, with interferential stimulators, the current perfuses to greater depths and over a larger volume of tissue than other forms of electrical therapy. When current is applied to the skin, the capacitive skin resistance decreases as pulse frequency increases. For example, at a frequency of 4,000 Hz (interferential range), the capacitive skin resistance is 80 times lower than with a frequency of 50 Hz (in the TENS range). Thus, interferential current crosses the skin with greater ease and with less stimulation of cutaneous nociceptors allowing greater patient comfort during electrical stimulation. In addition, because medium-frequency (Interferential) current is tolerated better by the skin, the dosage can be increased, thus improving the ability of the interferential current to permeate tissues and allowing easier access to

deep structures. This explains why interferential current may be most suitable for treating patients with deep pain. Interferential currents are produced by using two-channel stimulators and four electrodes. Each channel produces a sinusoidal, symmetrical alternating current at a high frequency (2000–5000 Hz). The electrodes are used in a quad-polar arrangement and the AC frequencies are set at slightly different frequencies but at similar amplitudes. The currents from the two waveforms interface with each other in the tissue, giving constructive (when the two waveforms add to each other) or destructive (when the circuits tend to cancel each other) interference.

Types of electrodes for theraupetic/electrodiagnostic applications:

Two methods of electrode systems are in common use. The mono-polar technique makes use of small active stimulation electrode. The indifferent or dispersive electrode is of larger area and is placed near to the active electrode. This technique is used for testing of the galvanic and Faradic excitability and for determining the chronaxie. For diagnostic purposes, a ball or plate electrode which is provided with a small thick muslin strip is mounted on a special handle. The handle carries a finger-tip switch to facilitate convenient control of output. Similarly, a small metal electrode can be secured on the motor point, particularly for therapeutic applications. For recording i-t curves, the bi-polar electrode technique is usually preferred. Both the electrodes are fixed to the body so that the hands of the operator are free to operate the apparatus. The active electrode in this method need not be a small as we deal with higher current intensitiesand small area electrode may cause unpleasant heat sensations. Suitably sized metal sheets are used as electrodes in this system. The electrodes are fastened to a moistened pad of about 1 cm thickness and 1 cm wider than the electrode sheet on all sides. The material used for pads is of good absorbancy and ordinary water can be used to moisten the electrodes. The electrodes are held in position by rubber straps. The stimulation current therapy can also be administered with suction electrodes. The current is conducted directly to the patient through the suction electrodes; the negative pressure necessary for adhesion is generated in the electrodes themselves by a compressed air flow. This method prevents liquids, dirt and bacteria from being drawn out from the electrodes into the pump/hose system of the unit. The adhesion strength can be adjusted to suit the application. Suction electrodes have been used by M/S Siemens in their therapy unit (model NEOSERV 824). Standard texts on muscle stimulation emphasize that successful muscle stimulation can only be achieved if the activating currents are properly applied. Physiotherapists are trained to

Principle of generation of interference currents

understand all about motor points and how to apply stimulation through these points.

Pain relief through electrical stimulation:

1.Transcutaneous electrical nerve stimulation:

Pain is man's oldest enemy and for centuries, medicine has searched for an innocuous, nondestructive, non-invasive, well-tolerated and effective way of relieving pain that is both efficient and practical. In the past few years, several workers have reported their success in using electrical Impulses to block the pathways of the transmission of pain. The impulses are produced in a battery powered pulse-generator to which a pair of electrode-tipped wires can be attached. Applied to the skin overlying any painful area of the body, these electrodes provide continuous, mild electrical stimulation. These signals seem to jam the pain signals travelling along the nerve pathways before they can reach the brain. The result is analgesia, often for hours after stimulation ends. The pain control is explained by:

• The Gate Control Theory which suggests that by electrically stimulating sensory nerve receptors, a gate mechanism is closed in a segment of the spinal cord, preventing paincarrying messages from reaching the brain and blocking the perception of pain; and

• The Endorsphin Release Theory which suggests that electrical impulses stimulate the production of endorphin and enkaphalins in the body. These natural, morphine-like substances block pain messages from reaching the brain, in a similar fashion to conventional drug therapy, but without the danger of dependence or other side-effects.

The electrical impulses required for electrotherapy to treat the pain are provided by an instrument called TENS (Transcutaneous Electrical Nerve Stimulator). Investigations on a great variety of electrical impulse parameters have indicated that two waveforms, the square wave and the spike wave are optimally and equally effective in relieving pain. Most stimulators feature adjustable settings to control the amplitude (intensity) of stimulation by controlling voltage, current and the width (duration) of each pulse. Electrodes are placed at specific sites on the body for treatment of pain. The current travels through the electrodes and into the skin stimulating specific nerve path- ways to produce a tingling or massaging sensation that reduces the perception of pain.

Typically, the stimulator is based around a 500 ms spike pulse, having adjustable amplitude of 0 to 75 mA and an adjustable frequency of 12 to 100 pulses per second. Instruments having similar specifications except that they produce square waveform, have a pulse frequency range of 20–200 Hz, pulse width from 0.1 to 1.0 ms and pulse amplitude of 0–120 V with maximum output current as 25 mA. The instrument powered by three standard flashlight batteries of 1.5 V each gives about 100 hours of continuous operation. Transcutaneous or skin surface application of electrical stimulus is accomplished by application of the conducting pads to various triggerzone areas, acupuncture sites or even peripheral nerves. Skin irritation at the site of electrode application is diminished by the use of carbonized rubber electrodes applied with a tincture of Benzoin interface.

The skin electrode system must be designed so as to minimize impedance variations with motion, to conform to the body surface to provide a uniform impedance across the surface of the electrode and to have an adequate surface area. The adequate surface area can be determined keeping in view the peak square-wave current at the threshold of thermal damage as a function of the electrode surface area. The thermal damage threshold varies widely with skin impedance, which is a function of skin preparation. Transcutaneous electrical nerve stimulation (TENS) electrodes are commonly moulded from an elastomer such as silicon rubber, loaded with carbon particles to provide conductance. Conformability is achieved by making the electrode thin.

2.Spinal chord stimulator:

Spinal cord stimulation is a term relating to the use of electrical stimulation of the human spinal cord for the relief of pain. This is accomplished through the surgical placement of electrodes close to the spinal cord, either with leads extending through the skin, or chronically, with the leads connected to an implanted source of electrical current. The applied electrical impulses develop an electrical field in and around the spinal cord, which then causes depolarization or activation of a portion of the neural system resulting in physiological changes. The stimulus source provides stimulation pulses at frequencies ranging from 10 to 1500 Hz, with pulse widths from 100 to 600 ms and controllable amplitude from 1 to 15 mA delivered into a load from 300 to 1500 W. These parameters can be controlled when one is using an implant that derives power and control through RF coupling from an externally power unit.

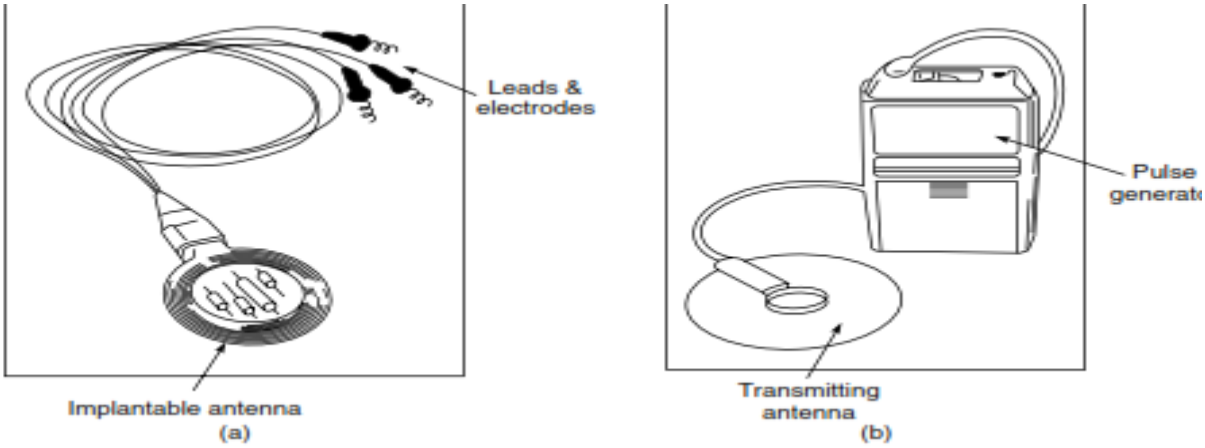
The figure shows the Medtronic Spinal Cord stimulation system, which has an implantable pulse generator and a hand-held programmer. Since a spinal cord stimulator is not a life-support system, there is no hazard associated with a

stimulator failing to provide an output. However, patients using ventricular inhibited or triggered pacemakers should not be exposed to nerve stimulation. Also, a patient entering a pulsed radio frequency field of the frequency to which the receiver is tuned would be in danger of having his stimulator activated by the field. Spinal cord stimulation has been shown to be of great benefit to some patients with multiple sclerosis and other neurological diseases; it is expected that the technique would be applied more and more in the near future.

Spinal cord stimulation system (Courtesy: M/s Medtronic, USA)
(A) ***Implantable pacemaker which includes electronic circuitry and power source***
(B) ***Magnet used by the patient to control the pulse generator's impulses within parameters set by the clinician***
(C) ***Hand held programmer which communicates with the pulse generator***
(D) ***Desk top programming console to programme the pulse generator and provides printouts of information from it***

The treatment of idiopathic scoliosis (lateral curvature of the spine) by electrical stimulation is described by Leonard (1980). The apparatus used for this purpose consists of an implanted radio receiver (Figa) and an external transmitter with an appropriate antenna (Fig. b). The transmitter is designed to generate pulses for muscle contraction lasting 1.6 seconds with a rest period of 9 seconds between contractions. The actual stimulation is not a single pulse but rather a burst of pulses consisting of individual pulses 220 ms wide, repeated 33 times every second. For transmission through the skin, the pulse bursts are modulated with a carrier frequency of 460 kHz. The receiver is a passive device designed to receive only signals from the transmitter. It de-modulates the signals and conducts them through the leads into the appropriate muscles to produce stimulation. The receiver circuit is embedded in an epoxy disc coated with silicon rubber for tissue compatibility. The receiver is attached to three leads of platinum-iridium wire terminating in platinum corkscrew electrodes. The electrodes are placed

over appropriate para-spinal muscles during surgery. The receiver is placed in a subcutaneous pocket on the convex side of the curve.The transmitting antenna is a flat disc which is taped on the skin over the subcutaneous receiver by disposable adhesive.



(a) ***Implanted radio receiver with leads***
(b) ***External transmitting unit with an antenna***

3.Magnetic Stimulation:

A problem with electric stimulation is that it is painful (Hallett and Leonardo, 1990). The pain is not very different from that induced by the stimulation of peripheral nerves, but it is sufficient to limit its clinical acceptability. It has been shown by Barker et al (1985) that it is possible to stimulate both the nerve and brain magnetically. A magnetic pulse is generated by passing a brief, high current pulse through a coil of wire. The technique has an advantage in that the stimulation is almost painless. Although a large number of studies have been carried out to study the effectiveness and safety of magnetic stimulation, the technique is still experimental and regulated in countries like the USA.