

442e Electrodiagnostic Studies of Nervous System Disorders: EEG, Evoked Potentials, and EMG

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ELECTROENCEPHALOGRAPHY

The electrical activity of the brain (the electroencephalogram [EEG]) is easily recorded from electrodes placed on the scalp. The potential difference between pairs of electrodes on the scalp (bipolar derivation) or between individual scalp electrodes and a relatively inactive common reference point (referential derivation) is amplified and displayed on a computer monitor, oscilloscope, or paper. Digital systems allow the EEG to be reconstructed and displayed with any desired format and to be manipulated for more detailed analysis and also permit computerized techniques to be used to detect certain abnormalities. The characteristics of the normal EEG depend on the patient's age and level of arousal. The rhythmic activity normally recorded represents the postsynaptic potentials of vertically oriented pyramidal cells of the cerebral cortex and is characterized by its frequency. In normal awake adults lying quietly with the eyes closed, an 8- to 13-Hz alpha rhythm is seen posteriorly in the EEG, intermixed with a variable amount of generalized faster (beta) activity (>13 Hz); the alpha rhythm is attenuated when the eyes are opened (Fig. 442e-1). During drowsiness, the alpha rhythm is also attenuated; with light sleep, slower activity in the theta (4–7 Hz) and delta (<4 Hz) ranges becomes more conspicuous.

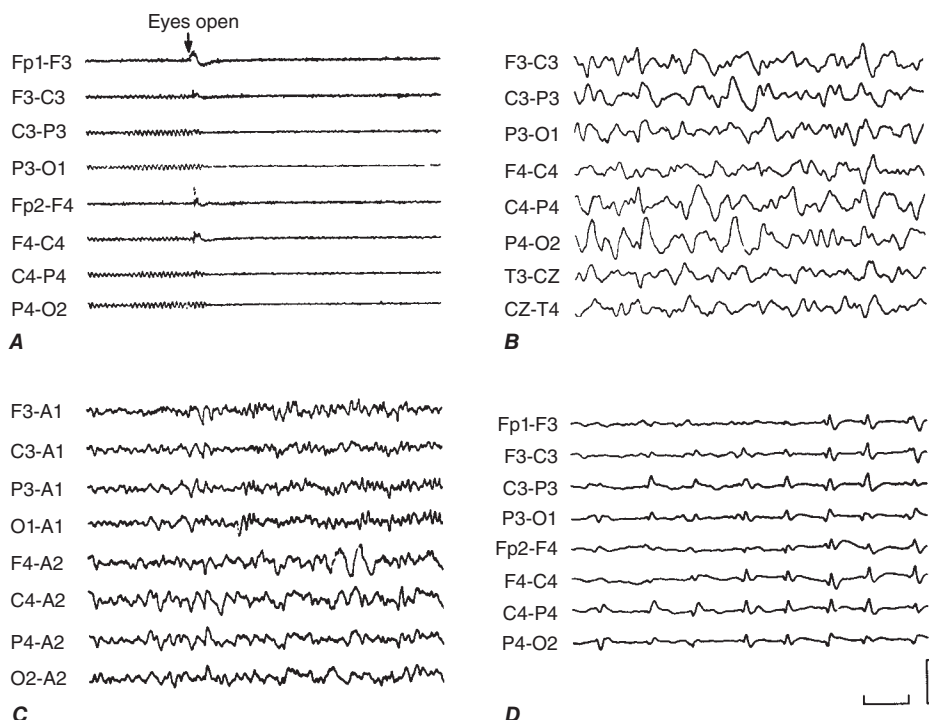


FIGURE 442e-1 **A.** Normal electroencephalogram (EEG) showing a posteriorly situated 9-Hz alpha rhythm that attenuates with eye opening. **B.** Abnormal EEG showing irregular diffuse slow activity in an obtunded patient with encephalitis. **C.** Irregular slow activity in the right central region, on a diffusely slowed background, in a patient with a right parietal glioma. **D.** Periodic complexes occurring once every second in a patient with Creutzfeldt-Jakob disease. Horizontal calibration: 1 s; vertical calibration: 200 μ V in A, 300 μ V in other panels. In this and the following figure, electrode placements are indicated at the left of each panel and accord with the international 10:20 system. A, earlobe; C, central; F, frontal; Fp, frontal polar; P, parietal; T, temporal; O, occipital. Right-sided placements are indicated by even numbers, left-sided placements by odd numbers, and midline placements by Z. (From MJ Aminoff [ed]: *Aminoff's Electrodiagnosis in Clinical Neurology*, 6th ed. Oxford, Elsevier Saunders, 2012.)

Activating procedures are generally undertaken while the EEG is recorded in an attempt to provoke abnormalities. Such procedures commonly include hyperventilation (for 3 or 4 min), photic stimulation, sleep, and sleep deprivation on the night prior to the recording.

Electroencephalography is relatively inexpensive and may aid clinical management in several different contexts.

THE EEG AND EPILEPSY

The EEG is most useful in evaluating patients with suspected epilepsy. The presence of electrographic seizure activity—i.e., of abnormal, repetitive, rhythmic activity having an abrupt onset and termination and a characteristic evolution—clearly establishes the diagnosis. The absence of such electrocerebral accompaniment to an episodic behavioral disturbance does not exclude a seizure disorder, however, because there may be no changes in the scalp-recorded EEG during certain focal seizures. With generalized tonic-clonic seizures, the EEG is always abnormal during the episode. It is often not possible to obtain an EEG during clinical events that may represent seizures, especially when such events occur unpredictably or infrequently. Continuous monitoring for prolonged periods in video-EEG telemetry units has made it easier to capture the electrocerebral accompaniments of such clinical episodes. Monitoring by these means is sometimes helpful in confirming that seizures are occurring, characterizing the nature of clinically equivocal episodes, and determining the frequency of epileptic events.

The EEG findings in the interictal period may show certain abnormalities that are strongly supportive of a diagnosis of epilepsy. Such *epileptiform activity* consists of bursts of abnormal discharges containing spikes or sharp waves. The presence of epileptiform activity is not specific for epilepsy, but it has a much greater prevalence in epileptic patients than in normal individuals. However, even in individuals with epilepsy, the initial routine interictal EEG may be normal up to 60% of the time. Thus, the EEG cannot establish the diagnosis of epilepsy in many cases.

The EEG findings have been used in classifying seizure disorders and selecting appropriate anticonvulsant medication for individual patients (Fig. 442e-2). The episodic generalized spike-wave activity that occurs during and between seizures in patients with typical absence epilepsy contrasts with focal interictal epileptiform discharges or ictal patterns found in patients with focal seizures. These latter seizures may have no correlates in the scalp-recorded EEG or may be associated with abnormal rhythmic activity of variable frequency, a localized or generalized distribution, and a stereotyped pattern that varies with the patient. Focal or lateralized epileptogenic lesions are important to recognize, especially if surgical treatment is contemplated. Intensive long-term monitoring of clinical behavior and the EEG is required for operative candidates, however, and this generally also involves recording from intracranial (subdural, extradural, or intracerebral) electrodes.

The EEG findings may indicate the prognosis of seizure disorders: In general, a normal EEG implies a better prognosis than otherwise, whereas an abnormal background or profuse epileptiform activity suggests a poor outlook. The EEG findings are not helpful in determining which patients with head injuries, stroke, or brain tumors will go on to develop seizures, because in such circumstances epileptiform activity is

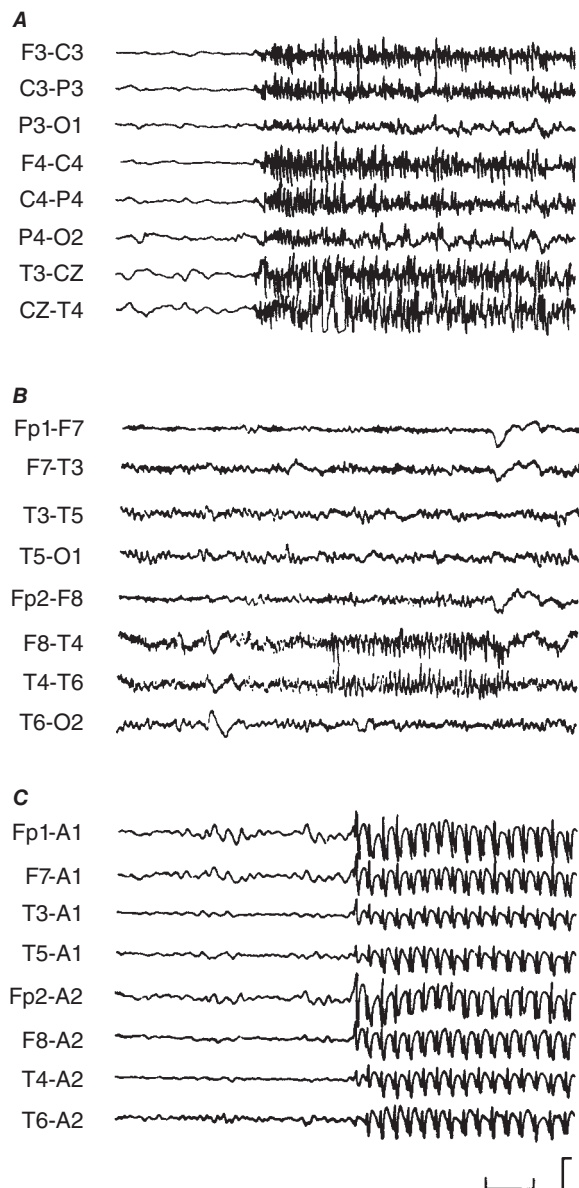


FIGURE 442e-2 Electrographic seizures. **A.** Onset of a tonic seizure showing generalized repetitive sharp activity with synchronous onset over both hemispheres. **B.** Burst of repetitive spikes occurring with sudden onset in the right temporal region during a clinical spell characterized by transient impairment of external awareness. **C.** Generalized 3-Hz spike-wave activity occurring synchronously over both hemispheres during an absence (petit mal) attack. Horizontal calibration: 1 s; vertical calibration: 400 μ V in A, 200 μ V in B, and 750 μ V in C. (From MJ Aminoff [ed]: *Aminoff's Electrodiagnosis in Clinical Neurology*, 6th ed. Oxford, Elsevier Saunders, 2012.)

commonly encountered regardless of whether seizures occur. The EEG findings are of limited utility in determining whether anticonvulsant medication can be discontinued after several seizure-free years. Further seizures may occur after withdrawal of anticonvulsant medication despite a normal EEG or, conversely, may not occur despite a continuing EEG abnormality. The decision to discontinue anticonvulsant medication is made on clinical grounds, and the EEG is helpful only for providing guidance when there is clinical ambiguity or the patient requires reassurance about a particular course of action.

The EEG has no role in the management of tonic-clonic status epilepticus except when there is clinical uncertainty about whether seizures are continuing in a comatose patient. In patients treated by drug-induced coma for refractory status epilepticus, the EEG findings indicate the level of anesthesia and whether seizures are occurring.

During status epilepticus, the EEG shows repeated electrographic seizures or continuous spike-wave discharges. In nonconvulsive status epilepticus, a disorder that may not be recognized unless an EEG is performed, the EEG may also show continuous spike-wave activity ("spike-wave stupor") or, less commonly, repetitive electrographic seizures (focal status epilepticus).

THE EEG AND COMA

The EEG tends to become slower as consciousness is depressed, regardless of the underlying cause (Fig. 442e-1). Other findings may suggest diagnostic possibilities, as when electrographic seizures are found or a focal abnormality indicates a structural lesion. The EEG generally slows in metabolic encephalopathies, and triphasic waves may be present. The findings do not permit differentiation of the underlying metabolic disturbance but help to exclude other encephalopathic processes by indicating the diffuse extent of cerebral dysfunction. An EEG responsive to external stimulation is helpful prognostically because electrocerebral responsiveness implies a lighter level of coma than a nonreactive EEG, and thus a better prognosis. Serial records provide a better guide to prognosis than a single record and supplement the clinical examination in following the course of events. As the depth of coma increases, the EEG becomes nonreactive and may show a burst-suppression pattern, with bursts of mixed-frequency activity separated by intervals of relative cerebral inactivity. In other instances there is a reduction in amplitude of the EEG until eventually activity cannot be detected. Such electrocerebral silence does not necessarily reflect irreversible brain damage, because it may occur reversibly in hypothermic patients or with drug overdose. The prognosis of electrocerebral silence, when recorded using an adequate technique, therefore depends on the clinical context in which it is found. In patients with severe cerebral anoxia, for example, electrocerebral silence in a technically satisfactory record implies that useful cognitive recovery will not occur.

In patients with clinically suspected brain death, an EEG recorded using appropriate technical standards may be confirmatory by showing electrocerebral silence, but disorders that may produce a similar but reversible EEG appearance must be excluded. The presence of residual EEG activity in suspected brain death fails to confirm the diagnosis but does not exclude it. The EEG is usually normal in patients with locked-in syndrome (Chap. 446), and helps in distinguishing this disorder from the comatose state with which it is sometimes confused clinically.

THE EEG IN OTHER NEUROLOGIC DISORDERS

In developed countries, computed tomography (CT) scanning and magnetic resonance imaging (MRI) are used as a noninvasive means of screening for focal structural abnormalities of the brain, such as tumors, infarcts, or hematomas (Fig. 442e-1). The EEG is still used for this purpose in many parts of the world, however, although infratentorial or slowly expanding lesions may not be recognized. Focal slow-wave disturbances, a localized loss of electrocerebral activity, or more generalized electrocerebral disturbances are common findings but do not indicate the nature of the underlying pathology.

In patients with an acute encephalopathy, focal or lateralized periodic slow-wave complexes, sometimes with a sharpened outline, suggest a diagnosis of herpes simplex encephalitis, and periodic lateralizing epileptiform discharges (PLEDs) are commonly found with acute hemispheric pathology such as a hematoma, abscess, or rapidly expanding tumor. The EEG findings in dementia are usually nonspecific and do not distinguish reliably between different underlying causes except in rare instances when the presence of complexes occurring with a regular repetition rate ("periodic complexes") supports a diagnosis of Creutzfeldt-Jakob disease (Fig. 442e-1) or subacute sclerosing panencephalitis. In most patients with dementia, the EEG is normal or diffusely slowed, and the findings alone cannot indicate whether a patient is demented or distinguish between dementia and pseudodementia.

CONTINUOUS EEG MONITORING

The brief EEG obtained routinely in the laboratory often fails to reveal abnormalities that are transient and infrequent. Continuous monitoring over 12 or 24 hours or longer may detect abnormalities or capture

clinical events that otherwise would be missed. The EEG is often recorded continuously in critically ill patients to detect early changes in neurologic status. Continuous EEG recording in this context has been used to detect acute events such as from nonconvulsive seizures or developing cerebral ischemia, to monitor cerebral function in patients with metabolic disorders such as liver failure, and to manage the level of anesthesia in pharmacologically induced coma.

MAGNETOENCEPHALOGRAPHY AND MAGNETIC SOURCE IMAGING

Recording the magnetic field of the electrical activity of the brain (magnetoencephalography [MEG]) provides a means of examining cerebral activity that is less subject to distortion by other biologic tissues than the EEG. MEG is used in only a few specialized centers because of the complexity and expense of the necessary equipment. It permits the source of activity to be localized and coregistered with the MRI in a technique that is known as *magnetic source imaging*. In patients with focal epilepsy, MEG is useful in localizing epileptogenic foci for surgery and for guiding the placement of intracranial electrodes for electrophysiologic monitoring. MEG has also been used for mapping brain tumors, identifying the central fissure preoperatively, and localizing functionally eloquent cortical areas such as those concerned with language.

EVOKED POTENTIALS

SENSORY EVOKED POTENTIALS

The noninvasive recording of spinal or cerebral potentials elicited by stimulation of specific afferent pathways allows the functional integrity of these pathways to be monitored but does not indicate the pathologic basis of lesions involving them. Such evoked potentials (EPs) are small compared to the background EEG activity, and the responses to a number of stimuli are therefore recorded and averaged with a computer to permit their recognition and definition. The background EEG activity, which has no fixed temporal relationship to the stimulus, is averaged out by this procedure.

Visual evoked potentials (VEPs) are elicited by monocular stimulation with a reversing checkerboard pattern and are recorded from the occipital region in the midline and on either side of the scalp. The component of major clinical importance is the so-called P100 response, a positive peak having a latency of approximately 100 ms. Its presence, latency, and symmetry over the two sides of the scalp are noted. Amplitude changes are less helpful for the recognition of pathology. VEPs are most useful in detecting dysfunction of the visual pathways anterior to the optic chiasm. In acute severe optic neuritis, the P100 is frequently lost or grossly attenuated; as clinical recovery occurs, it is restored but with an increased latency that generally remains abnormal indefinitely. The VEP findings are therefore helpful in indicating previous or subclinical optic neuritis. They may also be abnormal with ocular abnormalities and with other causes of optic nerve disease, such as ischemia or compression by a tumor. Flash-elicited VEPs may be normal in patients with cortical blindness.

Routine VEPs record a mass response over a relatively large cortical area and thus may be insensitive to localized waveform abnormalities. A newer technique, *multifocal VEP*, measures responses from 120 individual sectors within each affected eye and is more sensitive than routine VEP.

Brainstem auditory evoked potentials (BAEPs) are elicited by monaural stimulation with repetitive clicks and are recorded between the vertex of the scalp and the mastoid process or earlobe. A series of potentials, designated by roman numerals, occurs in the first 10 ms after the stimulus and represents in part the sequential activation of different structures in the pathway between the auditory nerve (wave I) and the inferior colliculus (wave V) in the midbrain. The presence, latency, and interpeak latency of the first five positive potentials recorded at the vertex are evaluated. The findings are helpful in screening for acoustic neuromas, detecting brainstem pathology, and evaluating comatose patients. The BAEPs are often normal in coma due to metabolic/toxic disorders or bihemispheric disease but are typically abnormal in the presence of brainstem pathology.

Somatosensory evoked potentials (SSEPs) are recorded over the scalp and spine in response to electrical stimulation of a peripheral (mixed or cutaneous) nerve. The configuration, polarity, and latency of the responses depend on the nerve that is stimulated and on the recording arrangements. SSEPs are used to evaluate proximal (otherwise inaccessible) portions of the peripheral nervous system and the integrity of the central somatosensory pathways, especially in patients who are comatose or suspected to be brain dead.

Clinical Utility of EPs EP studies may detect and localize lesions in afferent pathways in the central nervous system (CNS). They have been used particularly to investigate patients with suspected multiple sclerosis (MS), the diagnosis of which requires the recognition of multifocal white-matter lesions. In patients with clinical evidence of a single lesion, the electrophysiologic recognition of abnormalities in other sites helps to support the diagnosis but does not establish it unequivocally. Multimodality EP abnormalities are not specific for MS; they may occur in AIDS, Lyme disease, systemic lupus erythematosus, neurosyphilis, spinocerebellar degenerations, familial spastic paraplegia, and deficiency of vitamin E or B₁₂, among other disorders. The diagnostic utility of the EP findings therefore depends on the circumstances in which they are found. Abnormalities may aid in the localization of lesions to broad areas of the CNS, but attempts at precise localization may be misleading because the generators of many components are unknown.

The EP findings are sometimes of prognostic relevance. Bilateral loss of cortically generated SSEP components implies that cognition may not be regained in posttraumatic or postanoxic coma, and EP studies may also be useful in evaluating patients with suspected brain death. In patients who are comatose for uncertain reasons, preserved BAEPs suggest either a metabolic-toxic etiology or bihemispheric disease. In patients with spinal cord injuries, SSEPs have been used to indicate the completeness of the lesion. The presence or early return of a cortically generated response to stimulation of a nerve below the injured segment of the cord indicates an incomplete lesion and thus a better prognosis for functional recovery than otherwise. In surgery, intraoperative EP monitoring of neural structures placed at risk by the procedure may permit the early recognition of dysfunction and thereby permit a neurologic complication to be averted or minimized.

Visual and auditory acuity may be determined using EP techniques in patients whose age or mental state precludes traditional ophthalmologic or audiologic examinations.

COGNITIVE EVOKED POTENTIALS

Certain EP components depend on the mental attention of the subject and the setting in which the stimulus occurs, rather than simply on the physical characteristics of the stimulus. Such “event-related” potentials (ERPs) or “endogenous” potentials are related in some manner to the cognitive aspects of distinguishing an infrequently occurring target stimulus from other stimuli occurring more frequently. For clinical purposes, attention has been directed particularly at the so-called P3 component of the ERP, which is also designated the P300 component because of its positive polarity and latency of approximately 300–400 ms after onset of an auditory target stimulus. The P3 component is prolonged in latency in many patients with dementia, whereas it is generally normal in patients with depression or other disorders simulating dementia. ERPs are, therefore, sometimes helpful in making this distinction when there is clinical uncertainty, although a response of normal latency does not exclude dementia.

MOTOR EVOKED POTENTIALS

The electrical potentials recorded from muscle or the spinal cord following stimulation of the motor cortex or central motor pathways are referred to as *motor evoked potentials*. For clinical purposes, such responses are recorded most often as the compound muscle action potentials elicited by transcutaneous magnetic stimulation of the motor cortex. A strong but brief magnetic field is produced by passing a current through a coil, and this induces stimulating currents in the subjacent neural tissue. The procedure is painless and apparently safe.

Abnormalities have been described in several neurologic disorders with clinical or subclinical involvement of central motor pathways, including MS and motor neuron disease. In addition to a possible role in diagnosis or evaluating the extent of pathologic involvement, the technique provides information of prognostic relevance (e.g., in suggesting the likelihood of recovery of motor function after stroke) and provides a means of monitoring intraoperatively the functional integrity of central motor tracts. Nevertheless, it is not used widely for clinical purposes.

ELECTROPHYSIOLOGIC STUDIES OF MUSCLE AND NERVE

The motor unit is the basic element subserving motor function. It is defined as an anterior horn cell, its axon and neuromuscular junctions, and all the muscle fibers innervated by the axon. The number of motor units in a muscle ranges from approximately 10 in the extraocular muscles to several thousand in the large muscles of the legs. There is considerable variation in the average number of muscle fibers within the motor units of an individual muscle, i.e., in the innervation ratio of different muscles. Thus the innervation ratio is <25 in the human external rectus or platysma muscle and between 1600 and 1700 in the medial head of the gastrocnemius muscle. The muscle fibers of individual motor units are divided into two general types by distinctive contractile properties, histochemical stains, and characteristic responses to fatigue. Within each motor unit, all of the muscle fibers are of the same type.

ELECTROMYOGRAPHY

The pattern of electrical activity in muscle, i.e., the electromyogram (EMG), may be recorded both at rest and during activity from a needle electrode inserted into the muscle. The nature and pattern of abnormalities relate to disorders at different levels of the motor unit.

Relaxed muscle normally is electrically silent except in the end-plate region, but abnormal spontaneous activity (Fig. 442e-3) occurs in various neuromuscular disorders, especially those associated with denervation or inflammatory changes in affected muscle. *Fibrillation potentials* and *positive sharp waves* (which reflect muscle fiber irritability) and complex repetitive discharges are most often—but not always—found in denervated muscle and may also occur after muscle injury and in certain myopathic disorders, especially inflammatory disorders such as polymyositis. After an acute neuropathic lesion, they occur earlier in proximal than distal muscles and sometimes do not develop distally in the extremities for 4–6 weeks; once present, they may persist indefinitely unless reinnervation occurs or the muscle degenerates so completely that no viable tissue remains. *Fasciculation potentials*

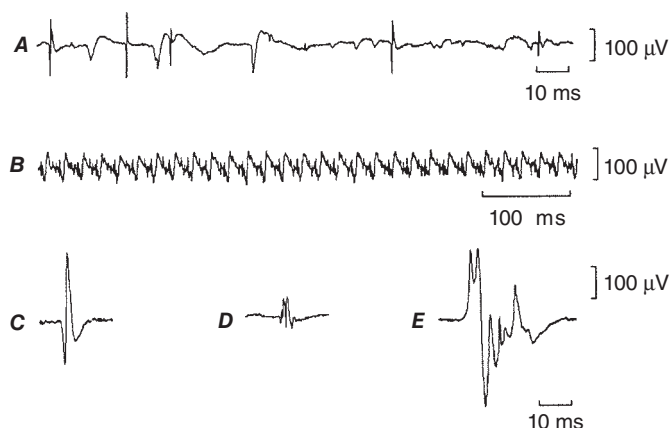


FIGURE 442e-3 Activity recorded during electromyography (EMG). **A.** Spontaneous fibrillation potentials and positive sharp waves. **B.** Complex repetitive discharges recorded in partially denervated muscle at rest. **C.** Normal triphasic motor unit action potential. **D.** Small, short-duration, polyphasic motor unit action potential such as is commonly encountered in myopathic disorders. **E.** Long-duration polyphasic motor unit action potential such as may be seen in chronic neuropathic disorders.

(which reflect the spontaneous activity of individual motor units) are characteristic of slowly progressive neuropathic disorders, especially those with degeneration of anterior horn cells (such as amyotrophic lateral sclerosis). *Myotonic discharges*—high-frequency discharges of potentials derived from single muscle fibers that wax and wane in amplitude and frequency—are the signature of myotonic disorders such as myotonic dystrophy or myotonia congenita but occur occasionally in polymyositis or other, rarer, disorders.

Slight voluntary contraction of a muscle leads to activation of a small number of motor units. The potentials generated by muscle fibers of these units that are within the pickup range of the needle electrode will be recorded (Fig. 442e-3). The parameters of normal motor unit action potentials depend on the muscle under study and age of the patient, but their duration is normally between 5 and 15 ms, amplitude is between 200 μ V and 2 mV, and most are bi- or triphasic. The number of units activated depends on the degree of voluntary activity. An increase in muscle contraction is associated with an increase in the number of motor units that are activated (recruited) and in the frequency of discharge. With a full contraction, so many motor units are normally activated that individual motor unit action potentials can no longer be distinguished, and a complete interference pattern is said to have been produced.

The incidence of small, short-duration, polyphasic motor unit action potentials (i.e., having more than four phases) is usually increased in myopathic muscle, and an excessive number of units is activated for a specified degree of voluntary activity. By contrast, the loss of motor units that occurs in neuropathic disorders leads to a reduction in number of units activated during a maximal contraction and an increase in their firing rate, i.e., there is an incomplete or reduced interference pattern. The configuration and dimensions of the potentials may also be abnormal, depending on the duration of the neuropathic process. The surviving motor units are initially normal in configuration but, as reinnervation occurs, they increase in amplitude and duration and become polyphasic (Fig. 442e-3).

Action potentials from the same motor unit sometimes fire with a consistent temporal relationship to each other, so that double, triple, or multiple discharges are recorded, especially in tetany, hemifacial spasm, or myokymia.

Electrical silence characterizes the involuntary, sustained muscle contraction that occurs in phosphorylase deficiency, which is designated a contracture.

EMG enables disorders of the motor units to be detected and characterized as either neurogenic or myopathic. In neurogenic disorders, the pattern of affected muscles may localize the lesion to the anterior horn cells or to a specific site as the axons traverse a nerve root, limb plexus, and peripheral nerve to their terminal arborizations. The findings do not enable a specific etiologic diagnosis to be made, however, except in conjunction with the clinical findings and results of other laboratory studies.

The findings may provide a guide to the severity of an acute disorder of a peripheral or cranial nerve (by indicating whether denervation has occurred and the completeness of the lesion) and whether the pathologic process is active or progressive in chronic or degenerative disorders such as amyotrophic lateral sclerosis. Such information is important for prognostic purposes.

Various quantitative EMG approaches have been developed. The most common is to determine the mean duration and amplitude of 20 motor unit action potentials using a standardized technique. The technique of macro-EMG provides information about the number and size of muscle fibers in a larger volume of the motor unit territory and has also been used to estimate the number of motor units in a muscle. Scanning EMG is a computer-based technique that has been used to study the topography of motor unit action potentials and, in particular, the spatial and temporal distribution of activity in individual units. The technique of single-fiber EMG is discussed separately below.

NERVE CONDUCTION STUDIES

Recording of the electrical response of a muscle to stimulation of its motor nerve at two or more points along its course (Fig. 442e-4)

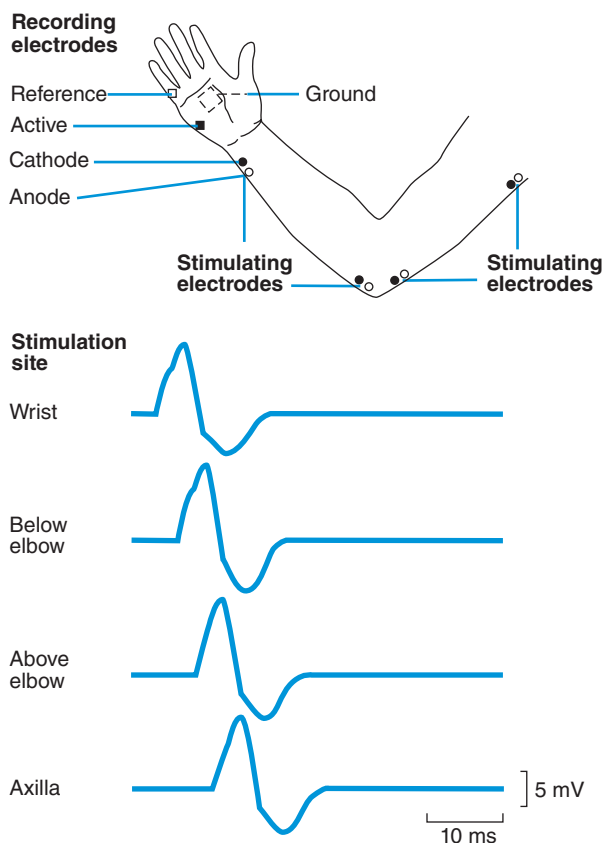


FIGURE 442e-4 Arrangement for motor conduction studies of the ulnar nerve. Responses are recorded with a surface electrode from the abductor digiti minimi muscle to supramaximal stimulation of the nerve at different sites, and are shown in the lower panel. (From MJ Aminoff: *Electromyography in Clinical Practice: Electrodiagnostic Aspects of Neuromuscular Disease*, 3rd ed. New York, Churchill Livingstone, 1998.)

permits conduction velocity to be determined in the fastest conducting motor fibers between the points of stimulation. The latency and amplitude of the electrical response of muscle (i.e., of the compound muscle action potential) to stimulation of its motor nerve at a distal site are also compared with values defined in normal subjects. Sensory nerve conduction studies are performed by determining the conduction velocity and amplitude of action potentials in sensory fibers when these fibers are stimulated at one point and the responses are recorded at another point along the course of the nerve. In adults, conduction velocity in the arms is normally between 50 and 70 m/s, and in the legs is between 40 and 60 m/s.

Nerve conduction studies complement the EMG examination, enabling the presence and extent of peripheral nerve pathology to be determined. They are particularly helpful in determining whether sensory symptoms are arising from pathology proximal or distal to the dorsal root ganglia (in the former instance, peripheral sensory conduction studies is normal) and whether neuromuscular dysfunction relates to peripheral nerve disease. In patients with a mononeuropathy, they are invaluable as a means of localizing a focal nerve lesion, determining the extent and severity of the underlying pathology, providing a guide to prognosis, and detecting subclinical involvement of other nerves. They enable a polyneuropathy to be distinguished from a mononeuropathy multiplex, which has important etiologic implications. Nerve conduction studies provide a means of following the progression and therapeutic response of peripheral nerve disorders and are used widely for this purpose in clinical trials. They may suggest the underlying pathologic basis in individual cases. Conduction velocity is often markedly slowed, terminal motor latencies are prolonged, and compound motor and sensory nerve action potentials may be dispersed in the demyelinating neuropathies (such as in Guillain-Barré syndrome, chronic inflammatory polyneuropathy, metachromatic leukodystrophy, or certain hereditary

neuropathies); conduction block is frequent in acquired varieties of these neuropathies. By contrast, conduction velocity is normal or slowed only mildly, sensory nerve action potentials are small or absent, and there is EMG evidence of denervation in axonal neuropathies such as occur in association with metabolic or toxic disorders.

The utility and complementary role of EMG and nerve conduction studies are best illustrated by reference to a common clinical problem. Numbness and paresthesias of the little finger and associated wasting of the intrinsic muscles of the hand may result from a spinal cord lesion, C8/T1 radiculopathy, brachial plexopathy (lower trunk or medial cord), or a lesion of the ulnar nerve. If sensory nerve action potentials can be recorded normally at the wrist following stimulation of the digital fibers in the affected finger, the pathology is probably proximal to the dorsal root ganglia (i.e., there is a radiculopathy or more central lesion); absence of the sensory potentials, by contrast, suggests distal pathology. EMG examination will indicate whether the pattern of affected muscles conforms to radicular or ulnar nerve territory or is more extensive (thereby favoring a plexopathy). Ulnar motor conduction studies will generally also distinguish between a radiculopathy (normal findings) and ulnar neuropathy (abnormal findings) and will often identify the site of an ulnar nerve lesion. The nerve is stimulated at several points along its course to determine whether the compound action potential recorded from a distal muscle that it supplies shows a marked alteration in size or area or a disproportionate change in latency, with stimulation at a particular site. The electrophysiologic findings thus permit a definitive diagnosis to be made and specific treatment instituted in circumstances where there is clinical ambiguity.

F-WAVE STUDIES

Stimulation of a motor nerve causes impulses to travel antidromically (i.e., toward the spinal cord) as well as orthodromically (to the nerve terminals). Such antidromic impulses cause a few of the anterior horn cells to discharge, producing a small motor response that occurs considerably later than the direct response elicited by nerve stimulation. The F wave so elicited is sometimes abnormal (absent or delayed) with proximal pathology of the peripheral nervous system, such as a radiculopathy, and may therefore be helpful in detecting abnormalities when conventional nerve conduction studies are normal. In general, however, the clinical utility of F-wave studies has been disappointing, except perhaps in Guillain-Barré syndrome, where they are often absent or delayed.

H-REFLEX STUDIES

The H reflex is easily recorded only from the soleus muscle (S1) in normal adults. It is elicited by low-intensity stimulation of the tibial nerve and represents a monosynaptic reflex in which spindle (Ia) afferent fibers constitute the afferent arc and alpha motor axons the efferent pathway. The H reflexes are often absent bilaterally in elderly patients or with polyneuropathies and may be lost unilaterally in S1 radiculopathies.

MUSCLE RESPONSE TO REPETITIVE NERVE STIMULATION

The size of the electrical response of a muscle to supramaximal electrical stimulation of its motor nerve relates to the number of muscle fibers that are activated. Neuromuscular transmission can be tested by several different protocols, but the most helpful is to record with surface electrodes the electrical response of a muscle to supramaximal stimulation of its motor nerve by repetitive (2–3 Hz) shocks delivered before and at selected intervals after a maximal voluntary contraction.

There is normally little or no change in size of the muscle response to repetitive stimulation of a motor nerve at 2–3 Hz with stimuli delivered at intervals after voluntary contraction of the muscle for about 20–30 s, even though preceding activity in the junctional region influences the release of acetylcholine and thus the size of the end-plate potentials elicited by a test stimulus. This is because more acetylcholine is normally released than is required to bring the motor end-plate potentials to the threshold for generating muscle fiber action potentials. In disorders of neuromuscular transmission this safety

factor is reduced. Thus in myasthenia gravis, repetitive stimulation, particularly at a rate of between 2 and 5 Hz, may lead to a depression of neuromuscular transmission, with a decrement in size of the response recorded from affected muscles. Similarly, immediately after a period of maximal voluntary activity, single or repetitive stimuli of the motor nerve may elicit larger muscle responses than before, indicating that more muscle fibers are responding. This postactivation facilitation of neuromuscular transmission is followed by a longer-lasting period of depression, maximal between 2 and 4 min after the conditioning period and lasting for as long as 10 min or so, during which responses are reduced in size.

Decrementing responses to repetitive stimulation at 2–5 Hz are common in myasthenia gravis but may also occur in the congenital myasthenic syndromes (Chap. 461). In Lambert-Eaton myasthenic syndrome, in which there is defective release of acetylcholine at the neuromuscular junction, the compound muscle action potential elicited by a single stimulus is generally very small. With repetitive stimulation at rates of up to 10 Hz, the first few responses may decline in size, but subsequent responses increase. If faster rates of stimulation are used (20–50 Hz), the increment may be dramatic so that the amplitude of compound muscle action potentials eventually reaches a size that is several times larger than the initial response. In patients with botulism, the response to repetitive stimulation is similar to that in Lambert-Eaton myasthenic syndrome, although the findings are somewhat more variable and not all muscles are affected.

SINGLE-FIBER ELECTROMYOGRAPHY

This technique is particularly helpful in detecting disorders of neuromuscular transmission. A special needle electrode is placed within

a muscle and positioned to record action potentials from two muscle fibers belonging to the same motor unit. The time interval between the two potentials will vary in consecutive discharges; this is called the *neuromuscular jitter*. The jitter can be quantified as the mean difference between consecutive interpotential intervals and is normally between 10 and 50 μ s. This value is increased when neuromuscular transmission is disturbed for any reason, and in some instances impulses in individual muscle fibers may fail to occur because of impulse blocking at the neuromuscular junction. Single-fiber EMG is more sensitive than repetitive nerve stimulation or determination of acetylcholine receptor antibody levels in diagnosing myasthenia gravis.

Single-fiber EMG can also be used to determine the mean fiber density of motor units (i.e., mean number of muscle fibers per motor unit within the recording area) and to estimate the number of motor units in a muscle, but this is of less immediate clinical relevance.

BLINK REFLEXES

Electrical or mechanical stimulation of the supraorbital nerve on one side leads to two separate reflex responses of the orbicularis oculi—an ipsilateral R1 response having a latency of approximately 10 ms and a bilateral R2 response with a latency in the order of 30 ms. The trigeminal and facial nerves constitute the afferent and efferent arcs of the reflex, respectively. Abnormalities of either nerve or intrinsic lesions of the medulla or pons may lead to uni- or bilateral loss of the response, and the findings may therefore be helpful in identifying or localizing such pathology.