35 Myopathy

In the evaluation of patients with suspected myopathy, molecular genetics has supplanted the need for electrodiagnostic (EDX) studies or muscle biopsy in many patients with inherited conditions. Moreover, in patients with suspected myopathy and no evidence of an inherited condition, a muscle biopsy ultimately will be required for definitive diagnosis, regardless of EDX studies. Despite these facts, EDX studies, especially the needle electromyography (EMG) examination, continue to play an important role in the evaluation of patients with suspected myopathy (Figure 35–1). EMG can often confirm the presence of a myopathy, as well as add diagnostic information if certain types of spontaneous activity are present. For example, fibrillation potentials and positive sharp waves in a myopathy suggest the possibility of inflammation or necrosis, whereas myotonic discharges suggest one of the myotonic muscle or periodic paralysis disorders (see Chapter 36), acid maltase deficiency, myotubular myopathy, or certain

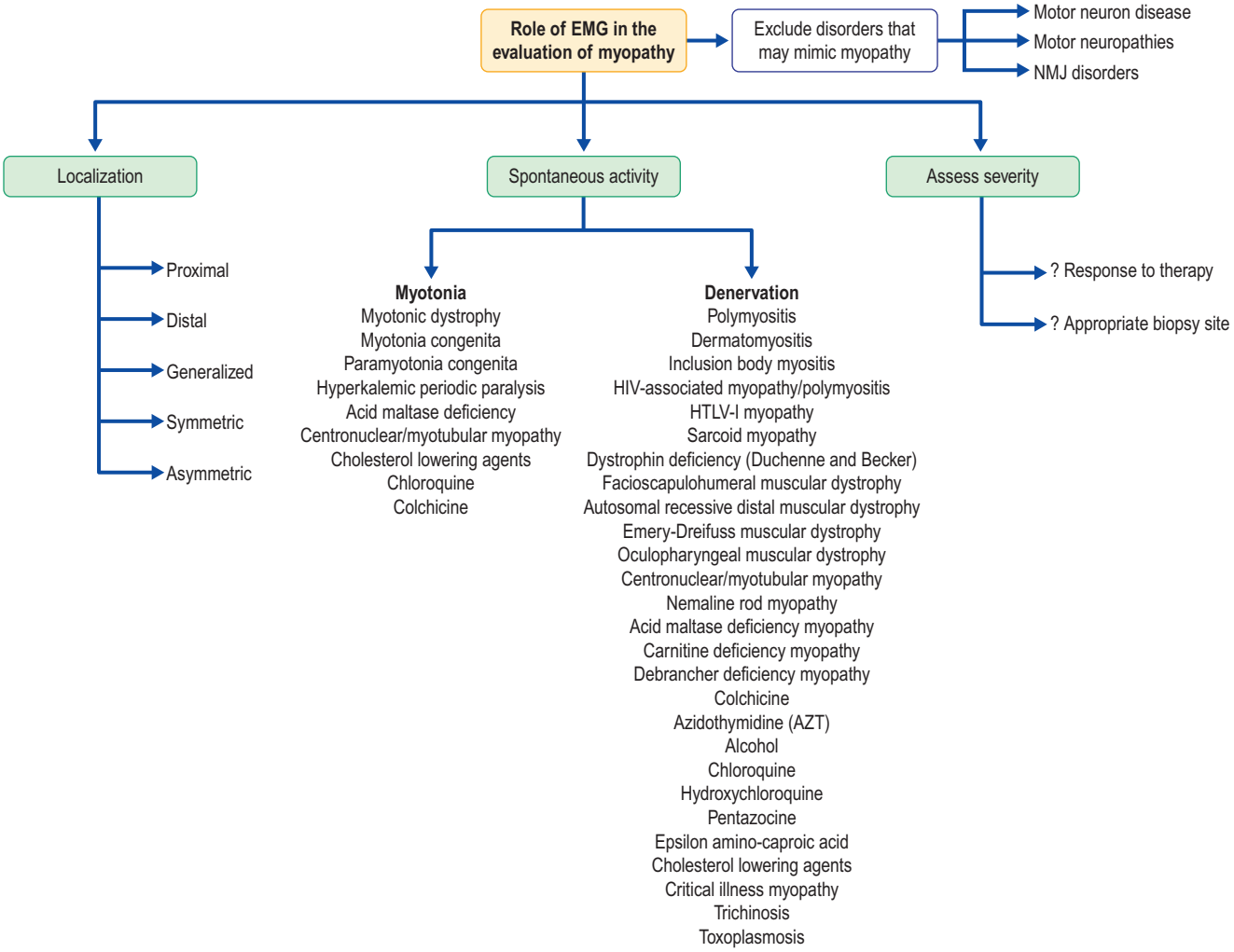

FIGURE 35–1 Role of electromyography in the evaluation of myopathy.

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toxic myopathies. Additionally, EMG may be helpful in suggesting alternate diagnoses that can mimic myopathy clinically.

EMG can also be useful in directing the site for a muscle biopsy in a patient with a myopathy. The EMG examination has the advantage that multiple muscles and sites can be sampled easily and often can suggest a suitable muscle to biopsy. It is always desirable to biopsy an unequivocally abnormal muscle yet one that is not end stage. However, biopsy is always recommended on the side contralateral to the EMG examination (see later).

Although the EMG examination may yield valuable information in the evaluation of suspected myopathy, mild cases may be especially difficult to interpret. Some myopathies, including steroid myopathy, may have minimal or no changes on EMG. In addition, some disorders of the neuromuscular junction (NMJ) may present with very similar clinical and EDX findings. Close attention to clinical detail, and often further EDX studies, including repetitive nerve stimulation and single-fiber EMG, may be required to differentiate between a myopathy and NMJ disorder.

CLINICAL

Myopathies present as pure motor syndromes without any disturbance of sensory or autonomic function. In most myopathies, symptoms tend to be bilateral and affect proximal muscles preferentially. Patients usually complain of difficulty rising from chairs, going up and down stairs, or reaching with their arms. Although most myopathies are symmetric and proximal, there are exceptions to both. For example, inclusion body myositis (IBM) and facioscapulohumeral muscular dystrophy may be very asymmetric. Myotonic dystrophy, distal hereditary myopathy, and IBM may preferentially affect distal more than proximal muscles. In some myopathies, ocular and bulbar muscles may be affected. Deep tendon reflexes are generally preserved or, if reduced, are in proportion to the degree of muscle wasting and weakness.

In evaluating a patient with suspected myopathy, it is important to determine whether symptoms are exercise induced. Such symptoms may manifest as fatigability, exercise-induced muscle cramps, or swelling. Patients who present with exercise-induced muscle cramps (see later) may develop frank weakness, swelling, and, if severe enough, myoglobulinuria. These latter symptoms suggest an inherited disorder of muscle energy metabolism. Note that although fatigability is certainly common in myopathies, frank muscle weakness that develops with exercise over a short period of time, if not accompanied by cramps, suggests a disorder of the NMJ rather than a myopathy. Additionally, patients with Lambert–Eaton myasthenic syndrome (LEMS) and some rare patients with myasthenia gravis (MG) present with isolated proximal muscle weakness mimicking a myopathy. In addition, adult onset spinal muscular atrophy, including X-linked bulbospinal muscular atrophy, usually presents with proximal muscle weakness and mimics the typical pattern of a myopathy.

Disorders of muscle can be simplified into the following categories: (1) muscular dystrophies, (2) inflammatory myopathies, (3) endocrine associated myopathies, (4) drug-induced and toxic myopathies, (5) metabolic myopathies, (6) congenital myopathies, and (7) myopathy associated with periodic paralysis.

Muscular dystrophies are inherited muscle disorders characterized by a progressive course and often an early onset, usually with a specific clinical and muscle biopsy pattern. In recent years, the chromosomal abnormality or specific gene product (e.g., dystrophin in Duchenne and Becker muscular dystrophy) has been discovered in several of these disorders. The more common muscular dystrophies include myotonic dystrophy, Duchenne muscular dystrophy, Becker muscular dystrophy, Emery–Dreifuss muscular dystrophy, facioscapulohumeral muscular dystrophy, oculopharyngeal muscular dystrophy, and limb girdle muscular dystrophies.

Inflammatory myopathies are associated most commonly with a presumed immunologic attack and include polymyositis (PM), dermatomyositis (DM), and IBM. Other types of inflammatory myopathy include those caused by muscle infection by parasites, viruses, or bacteria.

Endocrine myopathies are often seen in disorders of the thyroid and adrenal glands. In addition, myopathy can accompany some cases of acromegaly and parathyroid disease.

Drug-induced and toxic myopathies are becoming increasingly common. Examples of common drug-induced and toxic myopathies include those caused by steroids, alcohol, colchicine, azidothymidine (AZT), clofibrate, and many of the cholesterol-lowering agents.

Metabolic myopathies are disorders of muscle resulting from inherited enzyme deficiencies important in intracellular energy production. They may present in one of three ways: (1) as cramps and myoglobinuria, (2) as part of a more diffuse neurologic syndrome, often involving the central nervous system, or (3) as a typical clinical proximal myopathy. In patients with cramps and myoglobinuria, the genetic defect often is found either in the glycogen or lipid metabolism pathways. These patients may be completely normal at rest but become symptomatic during or after exercise. In patients with disorders along the lipid pathway, symptoms commonly occur after an episode of long or forced exercise (e.g., a long march or mountain climbing). In patients with disorders along the glycogen pathway, symptoms commonly occur after brief, intense isometric exercise. Muscle aches and fatigue may begin during the exercise, followed by frank myoglobinuria. Headache, nausea, and vomiting may occur. Muscles become painful and swollen. The creatine kinase (CK) level often is dramatically elevated into the thousands. The most common of these are caused by a deficiency of carnitine palmitoyltransferase (CPT) along the lipid pathway and myophosphorylase (McArdle's disease) along the glycogen pathway. Patients with defects in mitochondrial metabolism often present with a myopathy, as well as abnormalities involving other systems, including the central nervous system. Short

stature, hearing loss, seizures, cardiac abnormalities, learning disabilities, and stroke-like episodes are common. Lastly, some rare defects in metabolism (i.e., carnitine or acid maltase deficiency) may present as a typical clinical slowly progressive myopathy with proximal weakness.

Congenital myopathies are a group of myopathies in which each disorder has a fairly specific muscle biopsy finding on histochemical staining (e.g., nemaline rods, central cores, fiber type disproportion, myotubular myopathy). Typically, hematoxylin and eosin paraffin staining is normal or nonspecific. Although most patients present in the first few years of life, an occasional patient with a congenital myopathy presents in adulthood with one of these disorders. The clinical syndromes are nonspecific and tend to be slowly progressive or static. Muscle biopsy usually is needed for definitive diagnosis.

Myopathy associated with periodic paralysis occurs in the setting of hypokalemic and hyperkalemic periodic paralysis (see Chapter 36). Patients develop proximal weakness in the fifth or sixth decade. Even those patients with hypokalemic periodic paralysis who have never experienced episodic weakness, a common scenario in affected females, invariably will develop a proximal vacuolar myopathy in adulthood.

ELECTROPHYSIOLOGIC EVALUATION

Nerve Conduction Studies

Routine nerve conduction studies should always be done in patients with suspected myopathy (Box 35–1). Sensory nerve conduction studies are always normal, unless there is a coexistent neuropathy. Because most myopathies preferentially affect proximal muscles and routine motor nerve conduction studies record distal muscles, motor nerve conduction studies are also usually normal. If the myopathy is severe enough to affect distal and proximal muscles or is one of the rare myopathies that preferentially affects distal muscles, motor studies may show decreased compound muscle action potential (CMAP) amplitudes with normal latencies and conduction velocities.

The major reason nerve conduction studies must be performed is to exclude other motor disorders that may mimic myopathy (Box 35–2). Other than myopathy, pure motor disorders include motor neuron disease, rare cases of demyelinating polyneuropathy, and NMJ disorders. The nerve conduction studies in motor neuron disease and myopathies that affect distal muscles may be very similar. Differentiation is made based on the associated clinical features and needle EMG findings. Nerve conduction studies can easily differentiate demyelinating polyneuropathy from myopathy by the presence of conduction block or temporal dispersion, marked slowing of distal latencies and conduction velocity, or a combination of these findings.

Disorders of the NMJ present more of a challenge. NMJ disorders may present with proximal muscle weakness similar to myopathies. Postsynaptic disorders (e.g., MG) typically have normal CMAP amplitudes at rest. To demonstrate the NMJ abnormality, slow (3 Hz), repetitive nerve stimulation is required to demonstrate a decrement (see Chapter 34). Presynaptic disorders (e.g., LEMS) have a more characteristic nerve conduction pattern: CMAP amplitudes are low at rest with normal latencies and conduction velocities. Brief exercise (10 seconds) characteristically results in a marked increment of CMAP amplitude (typically >100% of baseline).

Box 35–1. Recommended Nerve Conduction Study Protocol for Myopathy

Routine studies:

1. At least one motor and one sensory conduction study and corresponding F responses from the upper extremity (e.g., median motor and sensory, median F responses)
2. At least one motor and one sensory conduction study and corresponding F responses from the lower extremity (e.g., tibial motor and sural sensory, tibial F responses)

Special considerations:

- If the compound muscle action potential (CMAP) amplitudes are decreased or borderline, exercise the muscle maximally for 10 seconds, then repeat a single supramaximal distal stimulation, looking for a significant CMAP increment (>100% of baseline), suggestive of the diagnosis of Lambert–Eaton myasthenic syndrome.
- If there is a clinical history of fatigability, repetitive nerve stimulation (3 Hz) of one distal muscle (e.g., ulnar nerve recording abductor digiti minimi) and one proximal muscle (e.g., the spinal accessory nerve recording the upper trapezius) should be performed. If a significant decrement is found with 3 Hz repetitive nerve stimulation of any muscle, then proceed with further testing, looking for a disorder of the neuromuscular junction (see Chapter 34, Box 34–2).

Electromyographic Approach

For the patient with suspected myopathy, the needle EMG examination must be individualized based on the distribution of the patient's symptoms (Box 35–3). Overall,

Box 35–2. Disorders that May Mimic Myopathy

- Motor neuron disease
  - Especially late-onset spinal muscular atrophy
  - X-linked bulbospinal muscular atrophy (Kennedy's disease)
  - Some cases of the progressive muscular atrophy variant of amyotrophic lateral sclerosis
- Neuromuscular junction disorders
  - Especially Lambert–Eaton myasthenic syndrome
  - Rare cases of restricted limb girdle myasthenia gravis
- Motor neuropathies
  - Usually demyelinating peripheral neuropathy (motor variants of chronic inflammatory demyelinating polyneuropathy; multifocal motor neuropathy with conduction block)
  - Rare cases of porphyric neuropathy preferentially affect proximal motor fibers
  - Diabetic amyotrophy (often affects proximal motor fibers, but usually with prominent pain)
- Central nervous system lesions
  - Bilateral middle cerebral artery–anterior cerebral artery watershed strokes

Box 35–3. Recommended Electromyographic Approach to Myopathy

Routine studies:

1. At least two distal and two proximal muscles in the lower extremity (e.g., tibialis anterior, gastrocnemius, vastus lateralis, iliacus)
2. At least two distal and two proximal muscles in the upper extremity (e.g., first dorsal interosseous, extensor indicis proprius, biceps brachii, medial deltoid)
3. At least one paraspinal muscle

Special considerations:

- Always try to study weak muscles. The number and distribution of muscles studied depend on the pattern of weakness.
- Try to study muscles that can easily be biopsied on the contralateral side (deltoid, biceps, vastus lateralis, gastrocnemius).
- If the motor unit action potential (MUAP) parameters are indeterminate, consider the following:
  Quantitative MUAP analysis: Accumulate 20 MUAPs from different locations within each muscle. Calculate the mean amplitude and duration and compare with age-matched controls for the muscle sampled.
  Single-fiber electromyography: If MUAP parameters, recruitment and activation pattern are normal when examined in weak muscles, then a neuromuscular junction disorder should be considered. Repetitive nerve stimulation should be performed first; if normal, single-fiber electromyography should be considered.

examining distal and proximal muscles in both the upper and lower extremities is indicated. Sampling the paraspinal muscles (the most proximal muscles) often is very useful. As most myopathies affect proximal muscles, the yield of finding abnormalities increases as progressively more proximal muscles are sampled. In adult-onset acid maltase deficiency myopathy, for instance, prominent changes may be seen only in the paraspinal muscles.

There are two other issues to keep in mind when performing EMG studies. First, measuring the serum CK immediately after the EMG examination probably is not wise. The CK level may rise slightly as a consequence of the EMG examination (typically 1.5× normal). The second issue is that of which muscle to biopsy, because patients with suspected myopathy often go on to muscle biopsy. The EMG can be very helpful in identifying an appropriate muscle to biopsy. One should biopsy a muscle that is abnormal but not at end stage. It usually is advisable to biopsy a muscle *contralateral* to the side sampled by the EMG needle. Because the EMG needle may induce transient inflammatory changes on the muscle biopsy, it is best not to biopsy muscles that have been sampled by the EMG needle. One would not like to diagnose an inflammatory myopathy and inappropriately place a patient on high-dose steroids based on spurious inflammation on a biopsy caused by the EMG needle.

Spontaneous Activity in Myopathies

Fibrillation potentials and positive sharp waves usually are associated with neuropathic disorders (i.e., neuropathy, radiculopathy, motor neuron disease). *However, denervating potentials occur frequently in many myopathic disorders*. They are thought to most likely occur as a consequence of segmental inflammation or necrosis of muscle fibers, separating a distal, healthy portion of the muscle fiber from the part attached to the endplate (Figure 35–2). Infarction of small intramuscular nerve twigs by surrounding interstitial inflammation also is speculated to be a possible cause of denervation in inflammatory myopathies. Although the presence of denervating potentials in a patient with myopathy often suggests the diagnosis of an inflammatory myopathy, denervating potentials can occur in a variety of myopathies (Box 35–4). In chronic myopathies, complex repetitive discharges may also be seen.

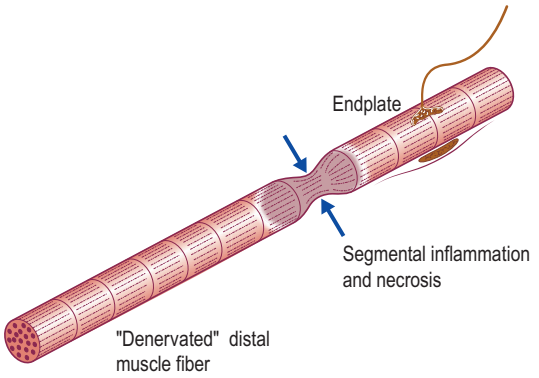

FIGURE 35–2 Generation of fibrillation potentials in inflammatory myopathies. Active denervation usually is associated with neuropathic disorders. However, active denervation also occurs frequently in many myopathies, especially those associated with inflammation or necrosis. Denervation is believed to occur as a consequence of segmental inflammation or necrosis of muscle fibers, separating a distal, healthy portion of the muscle fiber from the part attached to the endplate.

The presence of myotonic discharges yields additional information. A myotonic discharge is the spontaneous firing of a muscle fiber that waxes and wanes in both amplitude and frequency. The morphology of a myotonic discharge is either a positive wave or a brief spike potential. This morphology is the same as that of acute denervating potentials (i.e., fibrillation potentials and positive sharp waves). This should not be surprising because myotonic discharges are generated by muscle fibers as well. Myotonic discharges can be differentiated from fibrillation potentials and positive sharp waves by the waxing and waning of both firing frequency and amplitude. Remember that fibrillation potentials and positive sharp waves, in contrast, fire at a very regular rate. Myotonic discharges may be seen in myotonic dystrophy (types 1 and 2), myotonia congenita, paramyotonia congenita and hyperkalemic periodic paralysis. They can also be seen in other myopathies, including acid maltase deficiency (especially in the paraspinal muscles), myotubular (centronuclear) myopathy, some drug-induced myopathies (e.g., chloroquine, colchicine, cholesterol-lowering agents), and, occasionally, in PM.

Box 35–4. Myopathies with Denervating Features

- Inflammatory myopathies
  - Polymyositis
  - Dermatomyositis
  - Inclusion body myositis
  - Human immunodeficiency virus-associated myopathy/polymyositis
  - Human T-cell lymphotropic virus-1 myopathy
  - Sarcoid myopathy
- Dystrophies
  - Dystrophin deficiency (Duchenne and Becker)
  - Facioscapulohumeral muscular dystrophy
  - Autosomal recessive distal muscular dystrophy
  - Emery–Dreifuss muscular dystrophy
  - Oculopharyngeal muscular dystrophy
- Congenital myopathies
  - Centronuclear/myotubular myopathy
  - Nemaline rod myopathy
- Metabolic myopathies
  - Acid maltase deficiency myopathy
  - Carnitine deficiency myopathy
  - Debrancher deficiency myopathy
- Toxic myopathies
  - Colchicine, azidothymidine (AZT), alcohol, chloroquine, hydroxychloroquine, pentazocine, clofibrate, ε-aminocaproic acid, cholesterol-lowering agents, critical illness myopathy
- Necrotizing myopathy (non-inflammatory, immune-mediated)
- Amyloid myopathy
- Infectious myopathies
  - Trichinosis
  - Toxoplasmosis

The last type of "spontaneous activity" to recognize is a *contracture*, which is the complete absence of any EMG activity in a muscle while it is in the contracted state. Superficially, a muscle cramp and a contracture may appear similar clinically – the painful involuntary contraction of a muscle. However, during muscle cramps, which are neuropathic in origin, the EMG shows involuntary firing of motor unit action potentials (MUAPs) at a high frequency, whereas during a contracture, there is electrical silence. Contractures are seen only in rare metabolic myopathies (e.g., McArdle's disease, CPT deficiency) and occur as a result of insufficient energy available to break the actin–myosin bonds and return the muscle to a relaxed state. The "cramps" experienced by patients with metabolic myopathies such as McArdle's disease or CPT deficiency are, in fact, contractures.

Motor Unit Action Potential Analysis in Myopathies

Differentiating between myopathic and neuropathic disorders usually is primarily based on analysis of MUAP parameters (Figure 35–3). In most myopathies, there is dropout or dysfunction of individual muscle fibers that effectively decreases the size of the motor unit (Figure 35–4). In this situation, the actual number of motor units (i.e., anterior horn cells and axons) does not change. Only in the rare case of a very severe myopathy where every muscle fiber in a motor unit drops out does the effective number of motor units decrease.

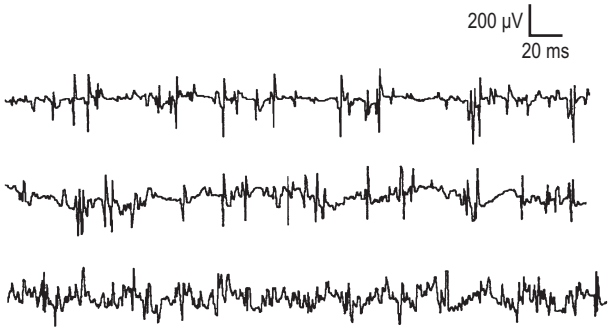

FIGURE 35–3 Myopathic motor unit action potentials. Short-duration, small-amplitude, polyphasic MUAPs with early recruitment are characteristic of myopathy. With little movement, many small, polyphasic MUAPs fill the screen and cannot be differentiated from each other **(lower trace).**

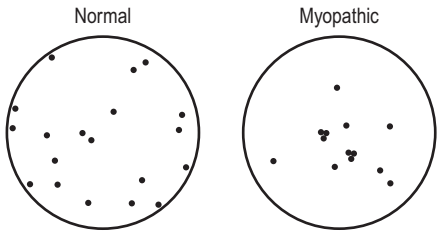

FIGURE 35–4 Motor unit territory in myopathy. In myopathy, motor unit territory typically decreases in size as individual muscle fibers drop out. Sometimes muscle fibers from the same motor unit are in close contact, either from muscle fiber splitting or after reinnervation in those myopathies associated with denervating features.
(Modified from Brown, W.F., 1984. The physiological and technical basis of electromyography. Butterworth, Boston, with permission).

Remember that there is a large normal variation in MUAP parameters. In borderline cases, it is advisable to measure at least 20 MUAPs and compare them with age-matched controls for the muscle sampled. Analysis of MUAPs, either subjectively or, more ideally, quantitatively, commonly allows a diagnosis of myopathy by noting specific changes in MUAP duration, amplitude, phases, and recruitment pattern.

MUAP duration is the most important parameter to measure in myopathy. Duration most closely reflects the total number of muscle fibers in a motor unit, including those muscle fibers at a distance from the recording electrode. The measurement of duration usually does not include linked potentials. In myopathy, duration characteristically decreases. The reduction in duration is best explained by the random dropout of muscle fibers (Figure 35–5). Of course, the finding of one brief MUAP does not make the electrodiagnosis of myopathy. Because there is a normal range of MUAP duration that varies depending on age and the muscle studied, one must sample several MUAPs to determine the *mean* duration. In myopathy, although the mean duration decreases, some of the MUAPs may be normal or possibly of long duration (Figure 35–6). In mild or equivocal cases, quantitative EMG of 10 to 20 MUAPs should be performed. In addition, it is important

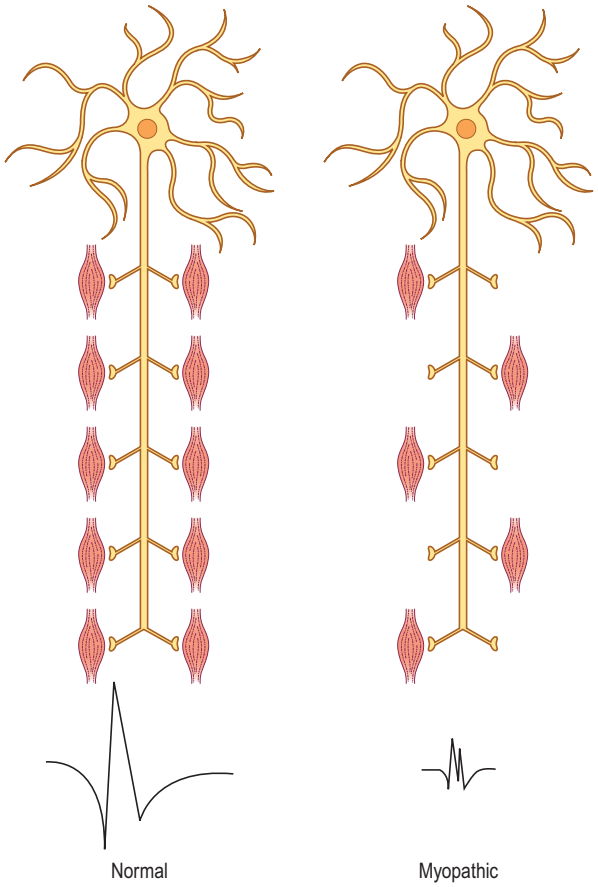

FIGURE 35–5 Model of the myopathic motor unit action potential. The MUAP in myopathy becomes short in duration, low in amplitude, and polyphasic from dropout and dysfunction of individual muscle fibers, whereas the motor neuron and its axon remain intact.

to remember that brief-duration MUAPs may be seen in conditions other than myopathy. Any disorder that effectively causes loss or dysfunction of individual muscle fibers (e.g., myopathy, NMJ disorders with block, disorders of the terminal axon) without affecting the motor neuron and its main axon can result in short-duration MUAPs (Box 35–5). A similar situation occurs in early reinnervation after severe denervation, when only a few fibers have successfully reinnervated, resulting in nascent (early reinnervated) motor unit potentials, which are also short and small. This point again emphasizes that the entire EMG examination must be taken as a whole and interpreted in light of the nerve conduction studies, as well as the history and examination, before a diagnosis is reached.

Somewhat surprisingly, chronic myopathies may have very long MUAP durations or frequently have linked or satellite potentials. These findings likely are secondary to fiber splitting or collateral sprouting from reinnervation in those myopathies associated with necrosis and subsequent denervation. In the chronic or late stage of a myopathy, it may be very difficult to distinguish a myopathic from a neuropathic disorder based on the duration of the MUAPs alone.

Box 35–5. Conditions Associated with Small, Short, Polyphasic Motor Unit Action Potentials

Myopathy
Neuromuscular junction disorders (myasthenia gravis, Lambert–Eaton myasthenic syndrome)
Early reinnervation after severe denervation (i.e., nascent motor unit potentials)
Periodic paralysis (during attack)
Disorders that selectively affect terminal axons (? paraneoplastic)

MUAP amplitude depends on just the few muscle fibers that are very close to the needle electrode. In myopathy, the amplitude commonly is decreased, but it can also be normal or increased if the needle electrode is placed near split or reinnervated fibers.

MUAP phases often are increased (>4 phases) in myopathy, but this is a nonspecific finding. The number of phases is primarily a measure of synchrony, and polyphasia may be seen in both myopathic and neuropathic disorders. Presumably, many of the remaining muscle fibers are dysfunctional and do not fire as synchronously as normal.

One of the most important findings in a myopathy is the presence of an early recruitment pattern. In myopathies in which there is dropout of individual muscle fibers from a motor unit, the motor unit becomes smaller and subsequently can generate less force. *Early recruitment refers to the inappropriate firing of many MUAPs to generate a small amount of force. In general, only the electromyographer performing the study can assess early recruitment.* Assessing early recruitment requires knowledge of how much force is being generated for the number of MUAPs that are firing. In myopathy, the number of MUAPs firing (recruitment) is appropriate for the firing frequency (activation); what is inappropriate is the number of MUAPs firing for the degree of force generated.

Only very rarely is the recruitment of MUAPs actually reduced in myopathy. This occurs only in the setting of end-stage muscle disease if all the muscle fibers of a single motor unit are lost, thereby causing an actual reduction in the number of motor units. This results in a reduced recruitment pattern on EMG. This situation is extremely uncommon but may arise in some very chronic myopathies that involve certain muscles severely, such as the quadriceps in IBM.

Single-fiber Electromyography in Myopathy

Single-fiber EMG (see Chapter 34) in patients with myopathy is commonly associated with increased jitter and blocking, especially in those myopathies associated with abnormal spontaneous activity. This finding emphasizes that increased jitter and blocking, although very sensitive to disorders of the NMJ, are not specific to these disorders. Any neuropathic or myopathic condition that involves any degree of denervation and reinnervation results in newly formed or dying NMJs, which in turn lead to abnormalities on single-fiber EMG. In differentiating a myopathy from a disorder

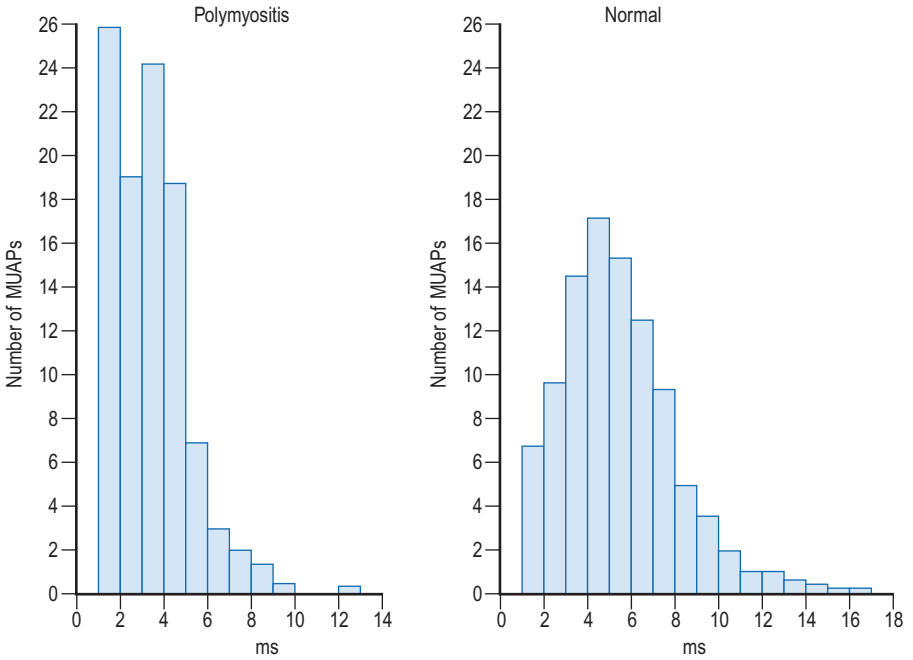

FIGURE 35–6 Motor unit action potential (MUAP) durations in myopathy. Patient with polymyositis **(left)** and a normal control **(right)** are shown. In the patient with polymyositis, there is a shift in mean duration to shorter MUAPs, although some medium and longer duration MUAPs are still present.
(Adapted from Buchthal, F., Pinelli, P., 1953. Muscle action potentials in polymyositis. Neurology 3, 424, with permission.)

of NMJ transmission, single-fiber EMG is most helpful in those cases where both nerve conduction and routine needle EMG findings are normal. In this setting, if the single-fiber EMG is abnormal, a disorder of NMJ transmission is more likely than a primary muscle disorder.

CLINICAL AND ELECTROPHYSIOLOGIC PATTERNS IN SELECTED MYOPATHIES

Polymyositis and Dermatomyositis

PM and DM are idiopathic inflammatory myopathies. Although they likely are autoimmune in nature, the disease mechanisms are not as clearly defined as in MG and LEMS, and at present the evidence is indirect. Muscle weakness may develop alone, which is the usual case. However, less commonly it may occur with a skin rash (DM), in association with one of a number of connective tissue diseases, or in the presence of a malignancy (usually occult). Approximately 20% of patients have an associated autoimmune or connective tissue disease (e.g., systemic lupus erythematosus, scleroderma, rheumatoid arthritis, Sjögren's disease, mixed connective tissue disease, polyarteritis nodosa).

The weakness in PM and DM usually develops subacutely but may be chronic and present over many months. Proximal muscles are predominantly affected in a symmetric fashion. The patient has difficulty getting out of chairs, the car, and bath; climbing stairs; and reaching above the head. In some cases, dysphagia occurs. Deep tendon reflexes are preserved unless the muscles are very weak, and atrophy is generally mild or does not occur. The neck muscles, especially the neck flexors, are commonly involved. However, the facial and extraocular muscles are generally spared, allowing fairly easy differentiation from MG. In a minority of patients, muscle swelling, tenderness, and myalgias occur. Also, arthralgias, joint stiffness, and Raynaud's phenomenon may occur, but true erosive arthritis is rare unless the patient has an associated connective tissue disorder. Cardiac involvement may occur in up to 40% of patients. This may range from minor electrocardiographic abnormalities to arrhythmias, pericarditis, and severe cardiomyopathy.

In DM, in addition to muscle weakness, the patient has a characteristic skin rash. This consists of a lilac-colored, reticulated, heliotrope-appearing rash on the upper eyelids. In addition, there is often erythema on the cheeks and over the shoulders and exposed upper chest. Erythematous, hyperkeratotic, scaly patches occur symmetrically on the extensor surfaces of the elbows, knuckles, and knees and over the medial malleoli. Periungual hyperemia and telangiectasias are common. In chronic DM, subcutaneous calcium deposits can occur and then rupture and drain.

Motor and sensory nerve conduction studies typically are normal. EMG typically shows prominent spontaneous activity (fibrillation potentials, positive sharp waves, and complex repetitive discharges). In acute and subacute cases, MUAPs are small, short, and polyphasic with early recruitment. In several studies of patients with biopsy-proven PM or DM, EMG abnormalities were seen in 89% of patients, with

fibrillation potentials present in 45 to 74% of patients. The diagnostic yield increased as more muscles were sampled. Of patients with fibrillation potentials, these were most commonly seen in the paraspinal muscles (94%), followed by the proximal shoulder and hip muscles (64–76%). With treatment, fibrillation potentials decrease or disappear. In chronic PM and DM (> one year), MUAPs become large and long in up to 50% of patients. They may occur in combination with small, short polyphasic MUAPs or rarely appear alone. The finding of large MUAPs, which most commonly are associated with neuropathic disorders, often creates confusion. In this case, assessment of the recruitment pattern is key. The finding of relatively preserved recruitment associated with large, long MUAPs must always suggest the possibility of a chronic myopathy.

Inclusion Body Myositis

IBM is an idiopathic inflammatory disorder of muscle that often is confused both clinically and on muscle biopsy with PM. IBM is now the most common inflammatory myopathy in individuals older than 50 years. Muscle biopsy shows inflammation and rimmed vacuoles, as well as intranuclear and cytoplasmic inclusions. Clinically, IBM presents as slowly progressive weakness. It is more common in men than in women (3 : 1). The age of presentation is commonly in the sixth decade. It is not unusual for patients to remain undiagnosed for many years after the onset of the first symptoms. Along with proximal muscle weakness, distal muscles are commonly involved. In some patients, the distal muscles actually are weaker than the proximal ones. Although the distribution of weakness is most commonly symmetric, asymmetric presentations also occur. *The disease has a predilection for certain muscles, especially the quadriceps and long finger flexors*. In addition, the iliopsoas, tibialis anterior, biceps, and triceps are commonly affected. Prominent muscle atrophy, especially of the quadriceps, is common. Facial and ocular weakness do not occur. The deep tendon reflexes tend to be depressed or absent early in the course, especially the quadriceps reflex. Fifteen percent of patients have other autoimmune diseases. A rare subset of patients has a familial form of IBM that spares the quadriceps muscles, and some forms are associated with Paget disease and frontotemporal dementia.

The initial diagnosis of IBM commonly is missed. It is not unusual for patients to be diagnosed initially with PM, which then fails to respond to immunosuppressive therapy. In addition, some patients with IBM and severe distal and proximal weakness and wasting, with depressed reflexes, initially are misdiagnosed with motor neuron disease. Rare patients with IBM present with dysphagia (in some patients, years before the onset of generalized weakness). Some patients present with dysphagia to solids and some also with dysphagia to liquids and nasal regurgitation. In these cases, the diagnosis of myopathy is rarely considered. The mechanism of the dysphagia in such patients has been shown to be paresis of the pharyngeal wall that precludes timely emptying of the pharynx before the upper esophageal sphincter closes, resulting in repetitive swallowing and choking.

Unfortunately, the electrophysiology often complicates the diagnosis of IBM. A distinct subset of patients demonstrates a mild sensory or sensorimotor polyneuropathy on nerve conduction studies (33–50% of patients). In addition, the needle EMG examination often is confusing. Prominent denervating potentials (fibrillation potentials and positive sharp waves) are common. The associated MUAP findings fall into one of three separate groups:

Group I	Small short MUAPs with polyphasia
Group II	Small short and large long MUAPs with polyphasia
Group III	Normal or large long MUAPs with polyphasia

As noted earlier, although large, long-duration MUAPs classically are associated with neuropathic disorders, they also are seen in myopathy, especially in chronic cases. In addition, patients with IBM can have an early, normal, or slightly reduced MUAP recruitment pattern, the latter finding usually associated with neuropathic processes. In group II, the small short and large long MUAPs may be found in the same muscle. The distribution of clinical weakness is most often proximal in group I and distal in groups II and III. Group III, although rare, is commonly mistaken for motor neuron disease (diffuse fibrillation potentials, large long-duration MUAPs with decreased recruitment). The heterogeneous profile of IBM makes the electrophysiologic diagnosis difficult. Many have commented that the combination of neuropathic and myopathic findings on EMG should suggest the diagnosis of IBM, although, in reality, this finding is simply consistent with a very chronic myopathy, as is usually the case in IBM.

Steroid Myopathy

Among drug-induced myopathies, steroids probably are the most common. The risk of steroid myopathy increases with the dose and duration of use. It typically is a proximal myopathy, preferentially affecting the hip girdle muscles. Motor and sensory nerve conduction studies are normal. The needle EMG typically is normal unless the myopathy is severe. In this situation, low-amplitude, short-duration MUAPs may be seen in the proximal muscles. *Of note, abnormal spontaneous activity is not seen*. This point is often very useful in differentiating PM from steroid myopathy. It is not uncommon for patients with PM to be treated with steroids, respond well initially, and then note a progression of weakness. In this case, it may be very difficult to differentiate recurrent or undertreated PM from steroid myopathy on clinical grounds. *The presence of abundant abnormal spontaneous activity strongly suggests PM, rather than steroid myopathy, as the cause of the weakness*.

Critical Illness Myopathy

Critical illness myopathy (CIM) is now a well-recognized and fairly common condition encountered in the intensive

care unit setting. It was first reported in patients who developed profound weakness after receiving intravenous steroids, usually high dose, after intubation. Nearly all of these patients were concurrently treated with NMJ-blocking agents. Patients with status asthmaticus have most often been reported. Typically, these are patients who are intubated and treated with several days of pharmacologic paralysis and high-dose intravenous steroids. After these measures are withdrawn, profound weakness is recognized or the patient fails to wean off the respirator. Often patients are flaccid and areflexic. Distal and proximal muscles are affected, and atrophy shortly ensues. Neck flexor weakness is common. Bifacial weakness can occur; however, extraocular weakness is unusual. Sensation usually is completely spared, an important finding that differentiates this condition from critical illness polyneuropathy and acute inflammatory demyelinating polyneuropathy. Laboratory testing frequently shows an elevated CK level, especially if the patient is tested early in the course of the weakness. Later CK levels may be slightly elevated or normal.

Nerve conduction studies demonstrate low CMAP amplitudes without any evidence of demyelination, *although in some patients the CMAPs are long in duration*. Conduction velocities and distal latencies are normal. Sensory studies are normal, unless there is a coexistent condition resulting in abnormal sensory potentials, such as critical illness polyneuropathy. Repetitive nerve stimulation studies are also normal. Needle EMG shows short-duration, low-amplitude MUAPs, often with denervation potentials, especially early in the course. Recruitment is normal or early, despite the profound weakness. On muscle biopsy, there is a characteristic loss of the thick (myosin) filaments on electron microscopy. In general, long-term prognosis is good, although most patients require not weeks, but usually several months, of rehabilitation to recover.

The pathophysiology of this disorder is not completely understood but most likely results primarily from toxicity of high-dose steroids in combination with NMJ blockers. In the majority of cases, patients have been exposed to both. Rarely, the myopathy develops with the use of steroids alone. The types of NMJ blockers vary, as do the type and dose of steroids. In general, patients treated with longer NMJ blockade (>24–48 hours) and longer- and higher-dose steroids appear to be more likely to develop this complication. The total dose of intravenous methylprednisolone usually is more than 1000 mg.

In recent years, there is growing evidence that CIM also occurs following the systemic inflammatory response syndrome (SIRS) that often accompanies sepsis, multi-organ failure, burns, trauma, and/or major procedures in the ICU. SIRS is felt to be present in the majority of patients hospitalized in the ICU for more than a week. In addition, many of the patients who develop CIM in the ICU also develop critical illness polyneuropathy as well, further complicating the clinical assessment, as well as the electrophysiologic evaluation (see Chapter 37).

EXAMPLE CASES

Case 35–1

History and Physical Examination

A 42-year-old woman was referred for progressive weakness of several months' duration. She had a long history of asthma treated with low-dose oral prednisone. Her initial symptoms were difficulty going up and down stairs and getting out of chairs. In addition, she developed mild difficulty with swallowing. The process was symmetric and progressive with little pain.

Neurologic examination showed mild proximal weakness in both upper and lower extremities. There was mild weakness of neck flexion with preserved neck extension. Muscle bulk and tone were normal. No facial or bulbar weakness was noted. Deep tendon reflexes and sensation were normal.

Summary

The history in this case suggests proximal muscle weakness. Difficulty going up and down stairs and difficulty getting out of low chairs are symptoms characteristic of proximal lower extremity weakness. On examination, proximal weakness in both upper and lower extremities, as well as mild weakness in the neck flexors, was found. Weakness of neck flexion is a key finding that indicates abnormalities above the cervical area. In some patients it may be difficult to differentiate whether upper extremity proximal weakness is due to myopathy or radiculopathy affecting the C5 and C6 roots. In such cases, examination of the neck flexors can be very helpful because they frequently are abnormal in myopathy.

The differential diagnosis of proximal weakness includes myopathy, polyradiculopathy, motor neuron disorders, NMJ transmission disorders, and unusual primarily motor demyelinating neuropathies. The absence of any sensory symptoms along with the intact reflexes argues against the possibility of a polyradiculopathy or demyelinating motor neuropathy. The absence of fatigability or weakness of the extraocular muscles makes the diagnosis of MG unlikely, although MG, along with LEMS, must still be considered. The history of long-term prednisone use may be important, because steroids are a common cause of myopathy.

Reviewing the nerve conduction findings, the right median, ulnar, tibial, and peroneal motor studies and F response studies are normal. All of the CMAP amplitudes, conduction velocities, and latencies are normal. Likewise, the median, ulnar, and sural sensory responses are intact. These normal motor, sensory, and F response studies effectively exclude a demyelinating polyneuropathy. In addition, the normal CMAP amplitudes at rest make the diagnosis of LEMS unlikely.

The EMG findings are very abnormal, with diffuse fibrillation potentials, especially in the proximal muscles. In addition, many of the MUAPs in the proximal muscles are of brief duration, low amplitude, and polyphasic with

CASE 35–1. Nerve Conduction Studies

Nerve Stimulated	Stimulation Site	Recording Site	Amplitude Motor = mV; Sensory = μV			Latency (ms)			Conduction Velocity (m/s)			F wave Latency (ms)		
			RT	LT	NL	RT	LT	NL	RT	LT	NL	RT	LT	NL
Median (m)	Wrist	APB	9.4		≥4	4.2		≤4.4				28		≤31
	Antecubital fossa	APB	8.9			8.5			64		≥49			
Ulnar (m)	Wrist	ADM	8.2		≥6	2.9		≤3.3				29		≤32
	Below elbow	ADM	8.2			6.5			60		≥49			
	Above elbow	ADM	8.2			8.2			60		≥49			
Median (s)	Wrist	Index finger	34		≥20	3.4		≤3.5	55		≥50			
Ulnar (s)	Wrist	Little finger	25		≥17	2.9		≤3.1	64		≥50			
Tibial (m)	Ankle	AHB	7.4		≥4	4.7		≤5.8				52		≤56
	Popliteal fossa	AHB	7.0			12.3			44		≥41			
Peroneal (m)	Ankle	EDB	4.2		≥2	4.8		≤6.5				51		≤56
	Below fibula	EDB	4.0			8.4			45		≥44			
	Lateral popliteal fossa	EDB	4.0			11.2			44		≥44			
Sural (s)	Calf	Posterior ankle	24		≥6	4.2		≤4.4	47		≥40			

m = motor study; s = sensory study; RT = right; LT = left; NL = normal; APB = abductor pollicis brevis; ADM = abductor digiti minimi; AHB = abductor hallucis brevis; EDB = extensor digitorum brevis.

Note: All sensory latencies are peak latencies. All sensory conduction velocities are calculated using onset latencies. The reported F wave latency represents the minimum F wave latency.

CASE 35–1. Electromyography

Muscle	Insertional Activity	Spontaneous Activity		Voluntary Motor unit Action Potentials				
		Fibrillation Potentials	Fasciculations	Activation	Recruitment	Configuration		
						Duration	Amplitude	Polyphasia
Right first dorsal interosseous	NL	0	0	NL	NL	NL	NL	NL
Right abductor pollicis brevis	NL	0	0	NL	NL	NL	NL	NL
Right extensor indicis proprius	NL	0	0	NL	NL	NL	NL	NL
Right biceps brachii	↑	+2	0	NL	Early	–2	–2	+2
Right pronator teres	↑	+1	0	NL	Early	–2	–2	+2
Right iliacus	↑	+1	0	NL	Early	–2	–2	+2
Right vastus lateralis	↑	+1	0	NL	Early	–1	–1	+1
Right tibialis anterior	↑	+1	0	NL	Early	–1	–1	+1
Right L5 paraspinal	↑	+2	0	NL	Early	–2	–2	+2
Right T6 paraspinal	↑	+2	0	NL	Early	–2	–2	+2

↑ = increased; NL = normal.

an early recruitment pattern. This MUAP profile of brief-duration, low-amplitude polyphasic MUAPs with early recruitment is characteristic of myopathic MUAPs. The prominent fibrillation potentials yield additional important diagnostic information, suggesting an inflammatory or necrotic muscle disease. Note that fibrillation potentials are not seen in steroid myopathy or in most cases of MG or LEMS.

When the nerve conduction studies and EMG examination are complete, the electrophysiologic impression can be formulated.

IMPRESSION: *There is electrophysiologic evidence consistent with a proximal myopathy with active denervating features.*

This case raises several important questions.

Does the Electromyography–Nerve Conduction–Clinical Correlation Make Sense?

There are several important correlations to note among the electrophysiologic study, the clinical history, and the neurologic examination. Looking first at the correlation between the motor nerve conduction studies and the needle EMG, the motor nerve conduction studies are quite normal, whereas the needle EMG findings are very abnormal. This paradox occurs because motor nerve conduction studies routinely record distal muscles, which are normal in most myopathies, whereas the needle EMG also can sample proximal muscles, which are abnormal in most myopathies. If CMAPs had been recorded from proximal muscles, where denervation is seen on the needle EMG, these would likely be abnormal with low amplitudes. It is not unusual for the routine motor conduction studies to be normal in typical proximal myopathies.

The next point to consider is the presence of the prominent fibrillation potentials, in light of the patient's history of steroid use. Although the patient takes steroids, her myopathy cannot be attributed to steroid use because of the active denervation. This EMG pattern is much more suggestive of an inflammatory myopathy such as PM.

The presence of brief-duration, low-amplitude, polyphasic MUAPs with early recruitment also eliminates the neuropathic disorders from the differential diagnosis, including amyotrophic lateral sclerosis, adult-onset spinal muscular atrophy, motor neuropathies, and polyradiculopathy. Early recruitment is characteristic of myopathic disorders. Because there is dropout or dysfunction of individual muscle fibers, each motor unit can generate less force. Therefore, more motor units than usual are needed to create a small amount of force. The EMG correlate of this underlying pathophysiology is the finding that the EMG screen fills very easily with many brief-duration, low-amplitude polyphasic MUAPs with a very small amount of force. To judge recruitment requires knowledge of how much force is being generated. Only the electromyographer can clearly assess early recruitment.

Which Muscle Should be Biopsied?

Muscle biopsy was performed on the contralateral vastus lateralis muscle. The contralateral side was chosen to avoid the possibility that minor inflammation caused by the EMG needle would be misinterpreted. Pathologic examination subsequently showed muscle fiber necrosis, with prominent mononuclear inflammatory infiltrates consistent with the diagnosis of PM. The patient was treated with high-dose prednisone and responded well.

Case 35–2

History and Physical Examination

A 75-year-old man developed progressive difficulty walking over a 2-year period. Initially, he noted that his gait was slightly unsteady. Later, he developed difficulty walking up stairs. His symptoms slowly worsened to the point that he would trip frequently when he walked quickly or walked on uneven ground. He noted no pain, numbness, or paresthesias in his legs and no bowel or bladder difficulties. He complained of no arm weakness, visual difficulties, or speech or swallowing problems.

Neurologic examination revealed normal cranial nerves and full strength in the neck flexors and extensors. He had 4/5 strength in the deltoids bilaterally and full strength in the biceps and triceps. More distally, the wrist extensors were 4/5, finger flexors were 3/5, and median and ulnar hand intrinsics were 4/5 bilaterally. Muscle bulk was near normal in the upper extremities, except for mild wasting of the proximal volar forearms. In his lower extremities, hip flexion was mildly weak bilaterally. There was prominent weakness of left knee extension (2/5) and bilateral foot dorsiflexion (3/5). Prominent wasting was noted in the left thigh and distally in both anterior calves. Deep tendon reflexes were 1+ in the upper extremities and at the right knee but were otherwise absent at the ankles and the left knee. Plantar responses were flexor bilaterally. Sensory examination, including vibration sense, light touch, position sense, and temperature sensation, was normal in the distal upper and lower extremities. Coordination was normal. He walked with a marked steppage gait.

Summary

The history and examination in this case suggest slowly progressive asymmetric weakness, predominantly affecting the lower extremities. There are no sensory complaints or sensory findings suggesting a radiculopathy or polyneuropathy. Examination shows asymmetric weakness and wasting, involving both proximal and distal muscles, but with a predilection for the left knee extensors and bilateral finger flexors. The diminished reflexes at the ankles and the left knee suggest a possible neuropathic process, although reduced reflexes may be seen with severe weakness from any cause.

CASE 35–2. Nerve Conduction Studies

Nerve Stimulated	Stimulation Site	Recording Site	Amplitude Motor = mV; Sensory = µV			Latency (ms)			Conduction Velocity (m/s)			F wave Latency (ms)		
			RT	LT	NL	RT	LT	NL	RT	LT	NL	RT	LT	NL
Median (m)	Wrist	APB		9.9	≥4		4.0	≤4.4					27	≤31
	Antecubital fossa	APB		9.8			7.9			51	≥49			
Ulnar (m)	Wrist	ADM		9.5	≥6		3.3	≤3.3					29	≤32
	Below elbow	ADM		8.9			6.9			55	≥49			
	Above elbow	ADM		8.8			8.9			50	≥49			
Median (s)	Wrist	Index finger		25	≥20		3.4	≤3.5		54	≥50			
Ulnar (s)	Wrist	Little finger		17	≥17		2.9	≤3.1		50	≥50			
Tibial (m)	Ankle	AHB		2.6	≥4		5.3	≤5.8					60	≤56
	Popliteal fossa	AHB		2.3			14.1			40	≥41			
Peroneal (m)	Ankle	EDB		1.1	≥2		5.1	≤6.5					56	≤56
	Below fibula	EDB		0.9			14.1			38	≥44			
	Lateral popliteal fossa	EDB		0.9			16.6			40	≥44			
Sural (s)	Calf	Posterior ankle		16	≥6		3.7	≤4.4		45	≥40			

m = motor study; s = sensory study; RT = right; LT = left; NL = normal; APB = abductor pollicis brevis; ADM = abductor digiti minimi; AHB = abductor hallucis brevis; EDB = extensor digitorum brevis.

Note: All sensory latencies are peak latencies. All sensory conduction velocities are calculated using onset latencies. The reported F wave latency represents the minimum F wave latency.

CASE 35–2. Electromyography

Muscle	Insertional Activity	Spontaneous Activity		Voluntary Motor Unit Action Potentials				
		Fibrillation Potentials	Fasciculations	Activation	Recruitment	Configuration		
						Duration	Amplitude	Polyphasia
Left tibialis anterior	CRD	+1	0	NL	Early	–1	NL	+2
Left medial gastrocnemius	CRD	+2	0	NL	Early	–1	NL	+1
Left vastus lateralis	CRD	+2	0	NL	↓	–1/+1	NL	+1
Left iliacus	CRD	+2	0	NL	Early	–1	NL	+1
Left L5 paraspinal	CRD	+1	0	NL	NL	–1	NL	+1
Left first dorsal interosseous	CRD	+2	0	NL	Early	–1	NL	+1
Left pronator teres	CRD	+1	0	NL	Early	–1	NL	NL
Left triceps	↑	0	0	NL	Early	–1	NL	+1
Left biceps brachii	↑	+1	0	NL	Early	–1	NL	+1
Left medial deltoid	CRD	+1	0	NL	Early	–1	NL	+1

CRD = complex repetitive discharge; ↑ = increased; ↓= slightly reduced; NL = normal.

Given the history and neurologic examination, the differential diagnosis includes motor neuron disease, a demyelinating motor neuropathy, or an unusual myopathy that is asymmetric and affects proximal and distal muscles. The prominent asymmetry and muscle wasting are not consistent with a disorder of the NMJ.

Moving on to the electrophysiology, the left median and ulnar motor and sensory nerve conduction studies and F responses are normal. In the left lower extremity, however, the peroneal and tibial CMAP amplitudes are decreased, with normal distal motor latencies and slightly slowed conduction velocities. The tibial F response latency is also slightly prolonged. The left sural sensory response is intact. The low motor responses with normal sensory responses in the lower extremity again suggest a predominant motor problem. The absence of markedly prolonged distal motor latencies or conduction velocity slowing, with no evidence of conduction block or temporal dispersion, effectively excludes a demyelinating motor polyneuropathy. After reviewing the nerve conduction studies, the possibility of motor neuron disease or an unusual disorder of muscle must still be considered.

The EMG shows prominent spontaneous activity, with frequent complex repetitive discharges and fibrillation potentials in most muscles tested. Most of the MUAPs, however, are short duration and polyphasic with early recruitment, consistent with a myopathy. The only exception is the left vastus lateralis, which has both long- and short-duration polyphasic MUAPs with slightly reduced recruitment.

After the nerve conduction and EMG studies, the electrophysiologic impression can be formulated.

IMPRESSION: *There is electrophysiologic evidence consistent with a chronic, asymmetric myopathy with denervating features.*

Several important questions can be addressed.

What is the Significance of the Complex Repetitive Discharges?

The presence of the complex repetitive discharges implies that the process is chronic. In addition, the finding of both long- and short-duration MUAPs in the vastus lateralis suggests a chronic process and, in the setting of small, short-duration MUAPs with early recruitment in other muscles, a chronic myopathy. Although myopathy is characteristically associated with small, short MUAPs, large, prolonged MUAPs also can be seen in chronic myopathies associated with denervating features (inflammatory and necrotic myopathies) in which reinnervation occurs as well.

What is the Most Likely Clinical Diagnosis?

This patient eventually had a biopsy of the right medial deltoid, a muscle that was clinically involved but had not been studied with the EMG needle. Pathologic examination showed marked variation in fiber size, marked mononuclear inflammatory infiltrates, numerous rimmed vacuoles, and intracytoplasmic inclusions. The pathologic diagnosis was IBM.

IBM usually presents in older men as a very slowly progressive muscle disorder often affecting both upper and lower extremity muscles. Many patients develop distal as well as proximal weakness. In some patients, weakness may be limited to the distal muscles. IBM often involves certain muscles preferentially, including the quadriceps, iliopsoas, tibialis anterior, biceps, triceps and the forearm, and long finger flexors. Focal atrophy of one of these muscles suggests the possibility of IBM. Occasional patients develop isolated dysphagia from IBM.

Electrophysiology often shows normal motor and sensory nerve conduction studies, although approximately one third of patients have mild slowing of motor and sensory conduction velocities. If distal muscles have been affected by the myopathy, the CMAP amplitudes also may be low. Fibrillation potentials are quite common, as are complex repetitive discharges, especially in long-standing cases. MUAPs may be brief or of long duration. A combination of large and small MUAPs may be present within the same muscle. In muscles that are severely affected by the myopathy, recruitment may actually be reduced. This occurs if every muscle fiber within a motor unit is lost, effectively leading to loss of the motor unit.

In end-stage muscle secondary to myopathy, it is not unusual to see fibrillation potentials, with large, prolonged MUAPs and a decreased recruitment pattern. These findings often incorrectly suggest a neuropathic illness, such as motor neuron disease. However, any EMG examination showing large, prolonged polyphasic MUAPs with a relatively normal or just slightly reduced recruitment pattern should suggest the possibility of a chronic myopathy. The only electromyographic clue that the disorder is myopathic in these cases is that the magnitude of the MUAP abnormalities appears too great for the mild degree of decreased recruitment. Indeed, there are occasional patients with chronic IBM in whom it is very difficult to differentiate IBM both clinically and electromyographically from the progressive muscular atrophy form of motor neuron disease.

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