Electrodiagnostic Approach to the Patient with Suspected Myopathy

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KEYWORDS

• Electromyography • Myopathy • Neuromuscular disorders
• Electrodiagnostic testing

Electrodiagnostic testing is a key component of the evaluation of a patient with a suspected myopathy. Recognition of certain electrophysiologic patterns associated with acquired and inherited myopathies can help to guide appropriate use of laboratory studies, including genetic testing, and aid in determining the need for muscle biopsy and in selecting a biopsy site. This article emphasizes the electrodiagnostic approach to myopathies, but the clinical and laboratory features are also described because such knowledge is necessary for the electromyographer. This article is an update of a review published in *Neurologic Clinics*.[1]

CLINICAL FEATURES OF MYOPATHY

Whether a patient is being evaluated in the office or the electromyography (EMG) laboratory, the process begins with the history and neurologic examination. In the EMG laboratory, the findings such as the distribution of weakness and other associated clinical features obtained through a targeted history and examination help to direct the selection of the most appropriate nerves and muscles for study.

Weakness

The symptoms and signs of myopathy are listed in **Table 1**. The most typical symptoms are referable to proximal (limb-girdle) weakness, which is a manifestation of most myopathies and is usually symmetric. Patients may report difficulty arising from a chair (**Fig. 1**), climbing stairs, or performing tasks with their arms elevated. In most cases, the weakness is painless, but sometimes myalgias or cramps are present. Because of proximal as well as trunk muscle weakness, patients often experience

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Table 1
Symptoms and signs of myopathy

	Symptom	Sign
Most common	Proximal muscle weakness (eg, difficulty rising from a chair, climbing stairs, walking, or lifting and using the arms above the head)	Limb-girdle weakness Neck flexor weakness Waddling gait, hyperlordotic posture (chronic disorders) Trunk weakness (eg, difficulty with a sit-up maneuver)
Less common	Myalgias, cramps	Muscle tenderness
	Distal weakness (eg, foot-drop, hand weakness)	Foot-drop; forearm or intrinsic hand muscle weakness; pes cavus (chronic disorders)
	—	Scapular winging
	Diplopia	Extraocular muscle weakness
	Ptosis	Ptosis
	Dysphagia	Weak palate, tongue, or both
	Dysarthria	Dysarthria (eg, nasal speech)
	Fatigue	—
	Shortness of breath	Diaphragm weakness: tachypnea, use of accessory muscles, paradoxical respirations
	Impaired grip release	Grip or percussion myotonia
	Head drop	Neck extensor weakness Muscle atrophy (chronic) Muscle hypertrophy

Fig. 1. A man with myotonic dystrophy type 2 has difficulty arising from a chair because of proximal weakness. He ambulates with a walker.