

A Primer on Electrophysiologic Studies in Myopathy

Matthew C. Lynch, MD^a, Jeffrey A. Cohen, MD^{a,b,*}

KEYWORDS

- Electromyography • Myopathy • Myositis
- Neurophysiology • Neuropathic disease

PURPOSE AND LIMITATIONS

The purpose of electromyography (EMG) is to localize a lesion within the peripheral nervous system. Peripheral causes of weakness can be divided into neuropathic processes, myopathic processes, and diseases affecting the neuromuscular junction. A neuropathic process is one which affects the anterior horn cell or its axon as it passes through the nerve root, plexus, or peripheral nerve. A myopathic process affects the muscle fibers. Specialized techniques can be performed, if necessary, to evaluate the neuromuscular junction. Weakness caused by disorders of the central nervous system such as multiple sclerosis or stroke is not usually diagnosed with EMG. This review focuses on physiologic principles involved in differentiating lesions of the lower motor neuron from primary muscle diseases.

Electrodiagnostic testing has some important limitations. A few muscle diseases do not cause abnormalities on EMG. Any muscle disease affecting the contractile apparatus of muscle fibers without affecting their electrical properties would not be detectable on EMG (eg, some congenital or endocrine myopathies).¹ A normal EMG can also be seen in the setting of steroid myopathy. This condition causes atrophy preferentially of type II muscle fibers,²⁻⁴ but EMG detects primarily type I fibers during the weaker levels of contraction at which individual motor units can be seen.⁵ Thus, a normal EMG would not rule out these conditions.

Even an abnormal EMG by itself does not often produce a specific diagnosis, because there are no findings that are specific to any given disease.¹ For example, electrical myotonia is almost always observed in myotonic dystrophy, but can also be seen in other conditions such as paramyotonia congenita, acid maltase deficiency,

The authors have nothing to disclose.

^a Department of Neurology, Dartmouth Hitchcock Medical Center, 1 Medical Center Drive, Lebanon, NH 03756, USA

^b Department of Neurology, Dartmouth Medical School, Hanover, NH, USA

* Corresponding author. Department of Neurology, Dartmouth Hitchcock Medical Center, 1 Medical Center Drive, Lebanon, NH 03756.

E-mail address: Jeffrey.A.Cohen@hitchcock.org

Rheum Dis Clin N Am 37 (2011) 253–268

doi:[10.1016/j.rdc.2011.01.008](https://doi.org/10.1016/j.rdc.2011.01.008)

rheumatic.theclinics.com

0889-857X/11/\$ – see front matter © 2011 Elsevier Inc. All rights reserved.

or rarely in neuropathic conditions. However, sometimes clinical history and examination findings supplement the electrophysiologic results to produce a specific diagnosis. Suppose EMG reveals myotonia in a 45-year-old man with weakness of the distal hand muscles, ankle dorsiflexors, and face. He also has frontal balding, cataracts, and diabetes mellitus. There is a family history of weakness in a similar pattern showing genetic anticipation. This patient can be diagnosed without further testing as having myotonic dystrophy because of the characteristic clinical history and EMG myotonia. At other times EMG aids diagnosis by localizing the pathology to muscle, directing further diagnostic testing such as muscle biopsy or genetic testing, which can be helpful in making a specific diagnosis. EMG can also be helpful in selecting an affected muscle for biopsy. Typically a biopsy site contralateral to the limb examined with EMG would be chosen, because damage from the EMG needle may affect biopsy results.⁶

PRINCIPLES OF MUSCLE PHYSIOLOGY

A brief review of basic nerve and muscle physiology serves as a foundation on which to build understanding of common EMG findings. Muscles are organized into motor units. A motor unit consists of a single alpha motor neuron and all of the muscle fibers it innervates, as shown in **Fig. 1A**. One motor neuron may innervate several muscle fibers (in muscles requiring high precision such as extraocular muscles) or hundreds of muscle fibers (in muscles requiring steady power such as the gastrocnemius). The muscle fibers of a single motor unit are not situated adjacent to each other.

When a motor neuron fires, an action potential propagates down its axon and all of the axon twigs. When the action potential reaches the nerve terminal, the depolarization triggers opening of voltage-gated calcium channels. The influx of calcium then triggers the release of acetylcholine into the extracellular space by fusion of the storage vesicles with the muscle membrane. Acetylcholine diffuses across the neuromuscular junction and activates nicotinic receptors, which are ligand-gated sodium channels. The influx of sodium into the muscle cell triggers an action potential in the muscle fiber similar to that in the nerve. However, muscle fibers contain a system of tunnels (called transverse or “T” tubules), which are invaginations of the cell membrane into the depths of the muscle cell, allowing the action potential to propagate rapidly throughout the muscle cell. The lumen of the T tubule is contiguous with the extracellular space. T tubules lie adjacent to an intracellular system of tunnels called the sarcoplasmic reticulum, which is a repository for stored calcium. The depolarization of the T tubules triggers voltage-gated calcium channels, leading to an initial calcium influx into the muscle fiber. This process causes the further release of calcium stores in the sarcoplasmic reticulum. The accumulation of calcium in the muscle fiber then drives the contractile apparatus. The flux of ions within muscle fiber creates an electric field that can be recorded from the EMG needle.

In neuropathic conditions the motor axon is injured, leaving muscle fibers of entire motor units without innervation. The muscle fibers become hyperexcitable, leading to spontaneous depolarization and contraction of a single muscle fiber, which is detected on EMG as fibrillations or positive sharp wave potentials. The denervated muscle fibers also produce signaling molecules that induce nearby motor neurons to sprout axon twigs to innervate the orphaned fiber (**Fig. 2**), which leads to enlarged motor units because then a single axon controls a greater number of motor fibers, as shown in **Fig. 1B**.

In myopathic disorders the pathology localizes to individual muscle fibers. Inflammatory or degenerative processes result in the loss of muscle fibers within the motor

Download English Version:

<https://daneshyari.com/en/article/3390694>

Download Persian Version:

<https://daneshyari.com/article/3390694>

[Daneshyari.com](https://daneshyari.com)