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Acute and Latent Effect of Poliomyelitis on the Motor Unit as Revealed by Electromyography

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ABSTRACT: When polio virus attacks the motor neuron it may be completely destroyed, damaged, or unaffected. Muscle fibers of a destroyed motor neuron are orphaned or reinnervated. Nearby functioning motor units will then send terminal axon sprouts to reinnervate the orphaned muscle fibers. If there are too many orphaned muscle fibers and not enough surviving motor units to reinnervate them, the orphaned muscle fibers will continue to fibrillate until they atrophy and die. The resultant effect of poliomyelitis upon the affected muscle is an overall loss of motor units with the remaining units innervating many more muscle fibers than they originally did.

There appears to be a late effect of polio upon these larger reinnervated motor units. After approximately 20 to 30 years, impulse transmission to the muscle fibers of the large reinnervated motor unit begins to fail. These transmission difficulties increase with age and time from recovery. These late onset transmission abnormalities may be factors in patient complaints of fatigue and progressive weakness.

The Normal Motor Unit

The motor unit is the functional component of the neuromuscular system.[1] Motor units, which vary in size, are composed of a motor neuron, its axon, and all the muscle fibers innervated by that axon. Smaller motor neurons have smaller diameter axons and innervate fewer muscle fibers. They also have lower thresholds of excitation and are the first motor units recruited in a contraction. Muscle fibers of these first recruited or low threshold motor units are high in oxidative enzyme activity, have a slow twitch in their response to electrical stimulation and are referred to as Type I. Conversely, larger diameter motor neurons have larger axons and innervate larger numbers of muscle fibers. They are high in anaerobic enzyme activity, have a fast twitch response, and are classified as Type II. Type II motor units tend to be recruited at higher strengths of contraction. All muscle fibers belonging to one motor unit are of the same histochemical fiber type.

The Normal EMG

Electromyography (EMG) is the electrophysiological study of the motor unit performed by inserting a small wire or needle electrode into a muscle.[2] While the muscle is at rest there is no electrical activity in the normal state. With very mild activation of a muscle, the electrical depolarization of the individual muscle fiber membranes prior to contraction is recorded, amplified and displayed on an oscilloscope. Each single muscle fiber depolarization is recorded as a biphasic spike with an initial positive deflection. The recording surface of the standard monopolar electrode is approximately 0.1mm^2 and records from a sphere with a radius of about 1mm. Between four and seven single muscle fibers' depolarizations (biphasic spikes) belonging to one anatomical motor unit are recorded and algebraically summated and are recorded as triphasic wave with an initial positive deflection. This summated depolarization of only a few fibers that comprise a whole motor unit is referred to as the motor unit action potential (MUAP). MUAPs have specific shapes and sizes that vary from muscle to muscle and change with different strengths of contractions. There is an orderly recruitment of larger sized motor units with increasing strength of contraction. At least 20 MUAPs are recorded from each muscle to indirectly sample the anatomic architecture of that muscle.

Acute Poliomyelitis and the EMG

With acute poliomyelitis, motor neurons are directly attacked. Some are destroyed, others are not affected and some are affected but recover with residual scars. In the acute phase of poliomyelitis there is a loss of motor units as a result of motor neuron death. The muscle fibers that are orphaned or denervated spontaneously depolarize in a regular fashion that is called *fibrillation*. These spontaneous single muscle fiber depolarizations are recorded by EMG with the muscle at rest and are referred to as fibrillation potentials.[3]

If a motor unit survives the attack of polio and its muscle fibers are near orphaned or denervated muscle fibers, the motor neuron will send out terminal axon sprouts to reinnervate orphaned muscle fibers. This process is called reinnervation by terminal axon sprouting.[3]

The end result of poliomyelitis is a loss of motor units for a muscle with the surviving motor units having an abnormally increased number of muscle fibers. After acute poliomyelitis, EMG recordings with the muscle at rest demonstrate the presence of free fibrillating muscle fibers. Motor units recorded with mild voluntary contraction show the reinnervated motor units to be of larger sizes and irregular shape, due to the increased fiber numbers per motor unit. As the strength of voluntary contraction increases, a reduction or loss of motor units recruited per strength of contraction is observed. Occasionally, a motor unit or

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a portion of a motor unit will spontaneously discharge while the muscle is at rest.[3] This spontaneous discharge of a motor unit or portion of a motor unit is called *fasciculation*. Fasciculation potentials are commonly recorded in polio patients with the muscle at rest. They are usually felt by the patient and can often be seen as an indentation of the skin. Fasciculations are a phenomenon of normal individuals occurring with fatigue or too much caffeine or stimulants. They are very common in polio patients in the acute phase but are less commonly seen after recovery except when a weakened muscle is overworked to fatigue.

There is a limit to the number of muscle fibers a motor unit can reinnervate. If only a few of the motor neurons of a muscle survive, the surviving motor neurons cannot possibly reinnervate all the deinnervated muscle fibers. As a result, the deinnervated muscle fibers not rescued will continue to fibrillate until they become totally atrophic and die. How long a deinnervated fiber will continue to fibrillate is unknown.

In this process, which can start in the surviving motor units as early as three to four weeks, unmyelinated axon sprouts grow and contact orphaned or reinnervated muscle fibers.[3,4] This site of contact will become the new neuromuscular junction. It takes about four to six months for these sprouts to become myelinated and for the immature neuromuscular junctions to transmit impulses without delays.

Single Fiber EMG

These steps in maturation of the reinnervated motor unit can be followed by a special EMG technique called Single Fiber EMG (SFEMG).[5,6] SFEMG allows the study of impulse transmission within one motor unit. With this technique an electrode with a recording surface of only 25 μm is placed within a muscle such that it can record the depolarizations of two single muscle fibers belonging to the same motor unit. The time that occurs between the two single fiber depolarizations is referred to as the Inter Potential Interval (IPI). There is a normal variation in the IPI as the normal motor unit fires repetitively. This normal variation is called *jitter* and is primarily related to the time variations in the chemically mediated neuromuscular transmission. Jitter can become abnormally increased with disorders affecting three areas of the motor unit. The three determinants of jitter are:

1. Transmission in the terminal axon branch to the fiber;
2. Neuromuscular transmission;
3. Impulse propagation down the muscle fiber to the recording site of the electrode.

In abnormal conditions the transmission can be severely affected and actually fail at any of these three sites. This condition is called "blocking."

In the early stages of reinnervation by terminal axon sprouting, there are transmission abnormalities that can be detected by SFEMG. Transmission frequently fails at the site of the axon sprout budding from the main axon. The axon sprout is initially unmyelinated and conduction is slow and variable. The newly formed neuromuscular junction is immature and is also a frequent site of transmission variability and failure. SFEMG in early reinnervation shows an increase in jitter and frequent blocking. With maturation of these reinnervated structures, the blocking stops and the jitter slowly reverts toward normal over a four to six month period.[4] Therefore, about a year after reinnervation there should be no evidence of blocking and essentially normal jitter. If the disease process or cause of deinnervation is continuous or ongoing, as in ALS or progressive spinomuscular atrophy, some motor units will be constantly undergoing reinnervation. As a result SFEMG will continuously record blocking and increased jitter of varying degrees. Poliomyelitis or the resultant viral infection of the spinal motor neurons is a one-time event. Therefore, the abnormalities of increased jitter and blocking are not expected since terminal axon sprouting should be completed within the first year post-polio.

Late Changes in the Motor Unit

In recent years, patients who recovered from polio 30 to 60 years previously have become frequent visitors to physiatrists and orthopedists.[7] Most of these patients were victims of the epidemics of the 1940s and 1950s. Most patients have similar complaints: increasing fatigue and muscle weakness not only in the previously weakened muscles, but also in those muscles that had recovered to normal or near-normal levels in the "good" leg or arm. Many of these patients also complain of muscle pain. In some cases, this muscle pain is actually the result of early degenerative arthritis in biomechanically overstressed hips, knees, and ankles. However, in many other cases the pain is related to the more common "tension and stress" muscle soreness, fibrositis syndrome or tension myalgia. While further study into etiology and treatment are certainly needed, two things seem to be certain: these patients are getting older and many of them are becoming weaker.

Routine EMG studies of these patients demonstrate the continuing presence of large, polyphasic reinnervated motor units in varying degrees. As the strength of contraction increases, the loss of motor units or the absence of motor units to provide an orderly stepwise recruitment of increasing size motor units remains obvious. Even in many normal strength muscles, a mild to moderate loss of motor units can be recorded, demonstrating subclinical abnormalities.

The continued presence of free fibrillating muscle fibers in some muscles studied raises several questions. Some post-polio muscle fibers appear fibrotic, and the EMG needle electrode is difficult to insert. These muscle fibers have obviously long since stopped fibrillating. Thus, the question arises: why do some muscle fibers survive and continue to fibrillate while others die and become fibrotic? It is well known that, with normal aging, motor neurons are lost. Are these fibrillating muscle fibers the result of a motor neuron's recent demise, or have they simply managed to survive and fibrillate for 40 to 50 years? Another possibility could be that individual muscle fibers are falling off the large reinnervated motor units over time and could be the reason that fibrillation continues to be recorded many years post-polio.

At first glance routine EMG studies in old polio patients do not demonstrate any new or different abnormalities compared to recordings made in previous years. In the late 1970s this author began to apply the techniques of SFEMG to study impulse transmission in the reinnervated motor units of older polio patients.[8] We began by studying previously compromised muscles that were now of normal strength or Grade G (4/5 MRC) in polio patients who were not complaining of new problems. In this initial study, we discovered that the impulse transmission within the old reinnervated motor unit was grossly abnormal and, in most cases, demonstrated not only an increase in jitter but also

prominent blocking. By studying patients of various ages and length of time since recovery from polio, we found that an almost linear relationship existed between the number of motor units demonstrating impulse transmission abnormalities and the time since recovery from active poliomyelitis. In continuing studies by the author of those muscles that are of antigravity strength and less, it has been seen that these weakened muscles tend to evidence transmission abnormalities of increased jitter and blocking in essentially all of the functioning motor units.

Routine EMG can be used to record the impulse transmission abnormalities of old reinnervated motor units. By using a trigger and delay device, which is now a standard feature of most EMG instruments, a single MUAP can be displayed as it fires repetitively with voluntary contraction. In normal motor units, the size and shape of the motor unit remains constant with repetitive firing. In those post-polio motor units that demonstrate marked increases in jitter and blocking on SFEMG, the size and shape of the MUAP is unstable and changes with repetitive discharge. This technique of determining motor unit stability with routine EMG is discussed further in a recently published paper.[9]

Muscle Weakness and the EMG

If impulse transmission is intermittently blocking to a large number of individual muscle fibers, varying degrees of weakness or fatigue could be noted in that muscle. Like the myasthenia gravis patient who complains of fatigue and weakness, some of the fatigue and weakness of the post-polio patient may be the result of the variability of impulse transmission within the motor unit. The normal motor unit was designed to nourish and drive 200 to 500 individual muscle fibers for 70 to 80 years. Studies with SFEMG in normal subjects over 65 years of age demonstrate the early development of mild transmission abnormalities. The reinnervated motor neuron must nourish and drive 1000 to 5000 or more individual muscle fibers, and abnormalities in its functioning have been detected within a 25 to 30 year period post recovery from polio. It is also possible that many of the surviving motor neurons carry permanent scars from the polio virus infections and are just not able to keep pace with the metabolic demands of innervating all of their muscle fibers.

Another factor in the late onset of weakness in post-polio patients may be the natural loss of motor neurons with aging. In polio each reinnervated motor unit contains a greater than normal proportion of functioning muscle fibers. The subsequent loss of a few motor units with the normal aging process can then have a significant effect on the individual muscle strength.

There are obviously many factors involved in the post-polio patients' complaints of fatigue and progressive weakness. No doubt, abnormalities of impulse transmission within the reinnervated motor units seen by the astute electromyographer on routine EMG are a significant contributing factor. Certainly further study is warranted to provide diagnostic insight and therapeutic support.

References

1. Peachey L, Adrian R, Geiger S: *Skeletal Muscle Handbook of Physiology.* Bethesda, American Physiology Society, 1983, Section 10.
2. Johnson E, Wiechers D: Electrodiagnosis, in Kottke F, Stillwell G, Lehmann J (eds): *Krusen's Handbook of Physical Medicine and Rehabilitation.* Philadelphia, WB Saunders Co, 1982.
3. Wiechers D: Anterior horn cell disease, in Johnson E (ed): *Practical Electromyography.* Baltimore, William & Wilkins, 1980.
4. Halkelivs L, Stalberg E: Electromyographical studies of free autogenous muscle transplants. *Man Scand J Plast Peconstr Surg* 1974; 8:211.
5. Stalberg E, Trontelj J: *Single Fiber Electromyography.* Surrey UK, Mirvalle Press, 1979.
6. Wiechers D: Single fiber EMG, in Johnson E, (ed): *Practical Electromyography.* Baltimore, Williams & Wilkins, 1980.
7. Halstead L, Wiechers D: *Effects of Poliomyelitis.* Miami, Symposia Foundation, 1984.
8. Wiechers D, Hubbell S: Late changes in the motor unit after acute poliomyelitis. *Muscle and Nerve* 1981; 4:524-528.
9. Wiechers D: Single fiber EMG with a standard monoplar electrode. *Arch Phys Med Rehab* 1985; 66:47-78.