ELECTRODIAGNOSTIC FINDINGS IN NEUROMUSCULAR DISORDERS

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EVEN the most astute clinician is at times faced with diagnostic problems in which the differentiation between myopathy, myelopathy and peripheral neuropathy is difficult. In such instances he will rightly seek help from electrodiagnostic tests. The purpose of this brief survey is to review methods in current use and describe their most suitable application.

The basic principles and techniques are now well known and full descriptions are available in texts by Licht (1961) and Walton (1964). The growth of electrodiagnosis has followed in a logical way the development of improved electrical stimulators and amplifying systems.

Strength-duration Curves

In the first tests used, visual comparison of the response of muscle to pulses of long (galvanic) or short (faradic) duration was made. From this have developed the more sophisticated chronaxie measurement and later, strength-duration curves. This last is still a valuable test and is in current use. In it, square wave pulses varying in duration from 100 milliseconds to 0.01 milliseconds are applied to the motor point of the muscle to be tested. The current or voltage (see M.R.C. Sub-Committee Report, 1958) required to produce a visible contraction is noted for each duration step. Normally innervated muscle requires no more current with short pulses than with long until the duration of pulse is 0.1 millisecond or less (Fig. 1 A). Denervated muscle has to be stimulated directly and requires much higher currents as the duration of pulse is shortened (Fig. 1 C). Partial denervation gives intermediate response (Fig. 1 B). This examination gives some quantitative evidence of the degree of denervation. Changes in the curve may develop as soon as four to five days after acute nerve injury and are therefore of value as an early indicator of developing denervation. In slowly developing lesions or in the recovery phase serial plotting of the curve can give valuable objective evidence of improvement or deterioration. In Bell's palsy, the presence of a normal strength-duration curve five days after the development of complete paralysis is a useful and favourable prognostic sign.

The test has several disadvantages. (a) It cannot differentiate between denervation resulting from disease of anterior horn cell, nerve root or peripheral nerve. (b) It may not disclose denervation when this occurs as a result of patchy involvement of anterior horn cells or from partial compression of a single root. (c) Deeply lying muscles may be impossible to test. (d) In testing denervated muscles, high stimulus intensities may be required and are sometimes unpleasant to the patient.

Electromyography

Electromyography helps overcome disadvantages (b) and (c) but (a) is still applicable. Repeated needle punctures offer a different form of un-

FIG. 1.— S.D. curves on normal (A), partially denervated (B) and denervated muscle (C). (Reproduced from "Disorders of Voluntary Muscle" edited by J. N. Walton by permission of the Editor and Messrs. J. & A. Churchill.)

pleasantness but on the whole are well tolerated. Normal muscle is electrically silent at rest but graded volitional contraction produces electrical activity which summates as more motor units are activated to produce a full "interference pattern". Abnormalities in electromyography are of three types—

(1) *The presence of spontaneous discharges at rest or on mechanical stimulation.*

Fasciculation potentials, although occurring extensively in denervation from some causes (e.g. progressive muscular atrophy) are rather variable and may occur in some non-specific conditions. Fibrillation potentials however provide more definite evidence of denervation but do not appear until 14-21 days after the acute lesion occurs. The presence of high frequency "dive-bomber" potentials is a comforting and intriguing finding when one is seeking confirmation of the diagnosis of myotonia.

(2) *Reduction of motor unit discharges.*

The combination of fibrillation with reduction of interference pattern is excellent evidence of partial denervation. A good interference pattern in the presence of clinical weakness is more suggestive of a primary muscle disease.

(3) *Alteration of shape, size and duration of individual motor unit discharges.*

The loss of individual fibres rather than of whole motor units contributes to the highly "spiky" interference pattern seen in myopathies. Individual motor unit potentials are of low amplitude, short or normal duration and polyphasic. Large, long duration polyphasic potentials may be seen in neuropathies, where attempts at regeneration have occurred.

Electromyography is perhaps of most value in giving clear-cut evidence for or against a myopathic disorder and also in detecting small areas of denervation such as occur from partial lesions of single nerve roots.

Nerve Conduction Studies

These can be considered in three groups—

(1) *The simple observation of a muscle's response to stimulation of its nerve trunk* constitutes a valuable rapid screening test. Where a segmental nerve conduction block is present, the absence of response from proximal stimulation and normal response from stimulation distal to the block gives confirmation of both the site and degree of injury. Instances of anomalous innervation can also be detected.

In myasthenia gravis, the drop in amplitude of muscle action potentials evoked by repetitive supra-maximal nerve stimulation may be a useful confirmatory test although it is positive only in clinically affected muscles. Similar testing after decamethonium and edrophonium (Tensilon) may be more conclusive (Churchill-Davidson and Richardson, 1953).

(2) *Estimation of latency from nerve stimulation to muscle response* gives a more quantitative

measure. Control values for such "terminal latencies" have been established (Gassel, 1963, 64) for several muscles as also have the results in abnormal situations. Some help is given by this in separating peripheral neuropathies from myelopathies and radiculopathies. Applied to the facial muscles, a terminal latency of more than 4 milliseconds suggests partial denervation in patients with Bell's palsy. Increased latency may also occur in some acute polyneuropathies before conduction rate is significantly slowed in proximal segments of the nerve and is also a useful sign of compression of the median nerve in the carpal tunnel, where the propagated response has to traverse the damaged segment.

(3) *True conduction rates in large motor fibres can be estimated* when a nerve can be stimulated at two or more points. Recording of evoked nerve potentials gives similar information in a more direct fashion and can also be applied to pure sensory nerves such as the digital nerves, or superficial branches of the radial (Downie and Scott, 1964). Evoked potential examination is rather more sensitive than motor conduction rate estimation to early acute nerve damage as the amplitude of the response may be reduced before significant slowing of rate appears. Slowing of conduction rate by either technique is most marked in chronic neuropathies in which the nerves are diffusely involved and indeed such slowing may antedate clinical abnormalities, e.g. in diabetic or uraemic neuropathy (Downie and Newell, 1961; Preswick and Jeremy, 1964). Slow conduction rates have been used to signal the early development of peroneal muscular atrophy in clinically unaffected members of an affected family (Dyck, Lambert and Mulder, 1963), and also to indicate peripheral nerve involvement in metachromatic leucodystrophy (Fullerton, 1964). In acute polyneuropathy of Guillain-Barré type changes in nerve conduction are variable. If this disorder is confined to nerve roots and degeneration does not occur, conduction may remain normal throughout the illness. More often slowing of rate and diminution or loss of evoked potentials is seen.

The ability to examine a nerve segmentally (Fig. 2) allows very precise delineation of local compression neuropathies such as occur in the ulnar nerve at the elbow or median nerve at the wrist. In patients with severe atrophy of the small hand muscles the presence of normal rates and amplitude of response in sensory fibres of the median and ulnar nerves supports a presumptive diagnosis of anterior horn cell disease. This is of particular help in young children with possible Werdnig-Hoffman disease where sensory examination clinically may not be as reliable. If motor conduction rates can be estimated in anterior horn cell disease they are usually normal. These tests therefore do help to differentiate myelopathies from neuropathies.

Extensive electrodiagnostic testing can be a

FIG. 2.—Normal evoked potentials in ulnar and median nerves. (Reproduced from "Disorders of Voluntary Muscle" edited by J. N. Walton by permission of the Editor and Messrs. J. & A. Churchill).

time-consuming process. Although strength-duration curves are usually carried out by physiotherapists, the other techniques described require the participation of a physician. Full knowledge of the clinical problem is essential therefore for selection of the most appropriate examinations and to allow proper evaluation of the results obtained. Normal results may often prove as useful as abnormal particularly where a diagnosis of conversion reaction is suspected. In children, and in adults unable to give a clear history, electrodiagnostic tests may give objective findings not otherwise obtainable.

While techniques continue to be developed and improved the availability of those already established is far from optimal. It is to be hoped that this situation will improve and keep pace with further technical advances in future years.

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