OCULAR ASPECTS OF ELECTROMYOGRAPHY*†

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THE purpose of this paper is to provide simplified concepts of the principles of electromyography, as applied to the extra-ocular muscles, in the hope of encouraging more frequent recourse to this diagnostic procedure. No apology is made for this simplification as it is felt that previous publications on electromyography of the extra-ocular muscles have been too technical and that this has discouraged their perusal and, thereby, their practical application. Electromyography is of real value to the practising ophthalmic surgeon. It is not suggested that all ophthalmologists should practise ocular electromyography, but that appropriate cases (as indicated below) should be referred to centres where diagnostic electromyography is performed under the surveillance of an ophthalmologist.

The technique of electromyography of the extra-ocular muscles (see Björk and Kugelberg, 1953; Breinin and Moldaver, 1955; Marg, Jampolsky, and Tamler, 1959) is not described, since this paper is concerned with the practical applications of ocular electromyography. However, it is felt that the value of percutaneous sampling of the levator palpebrae superioris and inferior oblique muscles should be emphasized. (This is done in the former muscle by inserting the needle obliquely upwards through the upper palpebral furrow when the eye is looking down; the inferior oblique muscle is sampled by pushing the needle through the skin at the medial edge of the lower lid just above the orbital margin.) It often suffices in apprehensive adults and children to examine these muscles.

Principles

The motor unit (Fig. 1, opposite) consists of the nerve cell, axon, and muscle fibres innervated by the branching of the axon (Sherrington, 1929). It should be noted that, in Fig. 1, one nerve fibre is innervating five muscle fibres, for in the extra-ocular muscles, the nerve muscle fibre ratio is approximately 1:5–10 (Tergast, 1873; Bors, 1926), compared with a ratio of 1:150 in skeletal muscle (Eccles and Sherrington, 1930). In electromyography, the electrical potentials created by the muscle fibres contracting in response to a nerve impulse arriving at the motor-end plates are recorded. An electrode,

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a modified hypodermic needle (Fig. 2), is used to detect these potentials; a fine wire runs down the lumen insulated from the rest of the needle which is then described as a concentric electrode (Adrian and Bronk, 1929).

When inserted in the muscle (see Fig. 1), this electrode detects the electrical potentials from the contracting muscle fibres.

Such electrical potentials have a very small voltage and have to be measured in micro-volts.* These measurements are amplified electronically and are then passed to a cathode-ray tube where they become visible to the observer as a series of varying waves (Fig. 3). Another cathode-ray tube (recording the same waves) is attached to a camera so that the action potentials may be permanently recorded to allow accurate assessment at leisure. It is advantageous to amplify the action potentials and render them audible by means of a loudspeaker, for, besides the insertion potentials provoked by pushing the needle into the muscle, various disorders of function are more readily recognized by the characteristic sound of their electrical activity, *e.g.* the sharp click of fibrillation.

FIG. 3.—E.M.G. of right levator palpebrae superioris on upward gaze. Normal.

The sine-wave represents time and voltage, the wavelength being equivalent to 10 m.sec. and the amplitude 100 μ v.

The Normal Motor Unit

Since the nerve-fibre/muscle-fibre ratio is large as compared with that of skeletal muscle, the action potentials are of low voltage and of short duration, but in other respects (mono- or biphasic contour) they resemble those obtained from limb muscles (Fig. 4, opposite). Björk and Kugelberg (1953) found an average amplitude of 20–150 μ v. and a duration of 1–2 m.sec.† for normal action potentials, and observations at this centre concur with these findings. However, on occasion, the rather large potentials of 300–400 μ v. obtained by Breinin and Moldaver (1955) and recorded by them as normal have been obtained.

* 1 millionth of a volt = 1 μ V.

† One-thousandth of a second = 1 m.sec.

FIG. 4.—E.M.G. of levator palpebrae superioris in a case of Marcus Gunn phenomenon.

A No action potentials on attempted elevation of the lid.

B Single motor unit firing on movement of the jaw.

C Increased frequency of firing as jaw movements become more rapid.

In skeletal muscle no action potentials are recorded at rest, but in the extra-ocular muscles there is a constant production of action potentials in the primary position which disappear in deep sleep, anaesthesia (Breinin, 1957a), and on severance of the muscle tendon from the globe (Breinin, 1957b). This latter finding is attributed to the relaxation of the muscle spindles described by Cooper, Daniel, and Whitteridge (1955).

Lower Motor Neurone Lesions

Mild paresis of the extra-ocular muscles, caused by lower motor neurone lesions, cannot be recognized electromyographically, for there is no apparent reduction in the number of action potentials (Fig. 5).

FIG. 5.—E.M.G. during upward gaze of left levator palpebrae superioris of same patient as in Fig. 3. This patient had developed an incomplete left ptosis and paresis of left medial rectus of unknown aetiology after an operation for retinal detachment in the left eye. An apparently normal trace.

In more severe ocular nerve lesions, a reduction in the number of action potentials is caused by the loss of motor units, the duration remaining unaltered and the amplitude being usually decreased.

Lower motor neurone palsy, resulting in denervation, is characterized electromyographically by fibrillation potentials (Fig. 6), which result from the irregular and spontaneous contraction of single muscle fibres, attributed

to the sensitivity of the denervated muscle fibre to minute amounts of circulating acetylcholine (Denny-Brown and Pennybacker, 1938).

FIG. 6.—Fibrillation potentials recorded from orbicularis oris in a patient with a facial palsy following sacrifice of the seventh nerve during removal of an acoustic neuroma.

The extra-ocular muscles also give rise to fibrillation potentials on denervation, but these are difficult to recognize since they may easily be confused with single motor unit firing.

A paralysis which clinically appears total often shows action potentials on the electromyogram (Fig. 7).

FIG. 7.—E.M.G. of levator palpebrae superioris of a patient with a clinically total third nerve palsy from a posterior communicating aneurysm. Action potentials are present, though abnormal.

The course of re-innervation of a muscle may be followed by electromyography, for recovery of function can often be anticipated by the gradual re-appearance of action potentials which are evident long before any movement is clinically apparent.

Attention has been drawn to the infrequency with which long-duration, high-voltage polyphasic potentials are obtained from the extra-ocular muscles in patients recovering from denervation affecting these muscles (Björk, 1954; Breinin and Moldaver, 1955; Marg and others, 1959). This is in marked contrast with the findings in patients recovering from comparable lesions affecting the limb muscles. An explanation of this discrepancy may be found in the morphological basis for these polyphasic potentials suggested by Coërs and Woolf (1959). These authors draw attention to the increasingly high voltage and temporal dispersion of the potentials encountered in the more proximally situated lesions, *e.g.* in poliomyelitis and motor neurone disease. The increased distance between lesion and muscle gives correspondingly increased opportunities for collateral branching within the nerve trunk, and consequent enlargement of the motor unit pool. At the same time differential delays in the arrival of impulses at the end plates, consequent on variation in the maturity of the axonic sprouts, will be correspondingly magnified (Fig. 8, overleaf). In the eye muscles, however, the motor nerves are so short and the normal nerve-fibre/muscle-fibre ratio so large that there is far less scope either for development of greatly enlarged motor units or for differential delays in conduction within those units.

Breinin (1957c) has made the interesting observation that diabetic neuropathy, producing an extra-ocular muscle palsy, results in minimal electrical activity in the affected muscle, and that this electrical silence often persists for more than 2 weeks in contradistinction to the appearance of fibrillation potentials after 2 weeks when denervation follows pressure from aneurysms or trauma. This may be a reflection of the tendency for the disease to remain halted at the end plate as shown by Coërs and Woolf (1959) in vitally-stained muscle biopsies from the limbs.

It is clear that electromyography has a limited application in the assessment of extra-ocular muscle paresis and cannot help in the important sub-group of mild paresis.

Diseases of Muscle

Electromyography is of undoubted value in the diagnosis of ocular myopathies. Only muscle biopsy can rival electromyography in providing the answer to difficult clinical problems, but it is easier, and more desirable, to obtain an electromyogram.

Fig. 9 (overleaf) illustrates the motor unit in a case of ocular myopathy—the number of muscle fibres in the unit is reduced and the muscle fibres

FIG. 8.—Denervation—skeletal muscle.

FIG. 9.—Myopathy.

that remain are abnormal. This causes the action potentials to be of low voltage and short duration. Fig. 10 shows the potentials of normal, myopathic, and fibrillating muscle to the same scale.

FIG. 10.—Action potentials at the same scale.

Electromyography has provided final evidence for the contention that chronic progressive nuclear ophthalmoplegia is due to a myopathy of the

extra-ocular muscles rather than to a nuclear degeneration (Breinin, 1957d; Esslen, Mertens, and Papst, 1958; Papst, Esslen, and Mertens, 1958) as suggested by Kiloh and Nevin (1951). All suspected cases should have the diagnosis confirmed by electromyography (Fig. 11).

FIG. 11.—E.M.G. of lateral rectus during attempted abduction by a patient with chronic progressive nuclear ophthalmoplegia. Eyes practically immobile. Trace shows low-voltage short-duration potentials.

Myasthenia gravis is a condition which often presents great difficulty in diagnosis both to the ophthalmologist and the neurologist. Even after the intravenous injection of "Tensilon" (3-hydroxyphenyl, dimethyl-ethyl ammonium chloride), which temporarily rectifies the curare-like block at the myo-neural junction, the normal motility of the globe may not return. However, an electromyograph taken within 2 minutes of the intravenous injection of "Tensilon" will show an undoubted increase in the size and frequency of the action potentials, although the functional improvement may be minimal. Fig. 12 (opposite) is an electromyogram of the levator palpebrae superioris of a myasthenic patient before and after intravenous "Tensilon" injection.

Disturbances of Motility

Breinin and Moldaver (1955) have shown, in two cases of intermittent exophoria, that the divergence occurred as a result of increased amplitude and frequency of the action potentials in the lateral rectus, *i.e.* divergence is an active process. However, this finding has been criticized by Jampolsky, Tamler, and Marg (1959) on the grounds that recordings should have been taken from the horizontal recti of both eyes simultaneously, and not from the horizontal recti of the diverging eye only.

Vertically incomitant horizontal strabismus, the A and V syndrome of Urist (1951), has been investigated by Breinin (1957e). His studies appear to support the suggested importance of the lateral recti in upward gaze and

FIG. 12.—E.M.G. of levator palpebrae superioris.

A Attempted elevation of lid before "Tensilon".

B, C, D Traces taken at 20-sec. intervals after "Tensilon". Note early fatiguability manifest in C and D.

of the medial recti in downward gaze in the production of these phorias. In the few cases in which the horizontal recti were shown by electromyography to be unaffected during vertical movements, the vertical muscles might be considered to be at fault. In this way, an indication may be provided whether the vertical incomitance should first be corrected by surgery of the horizontal or vertical muscles.

Electromyography is also of value in deciding whether defects of motility are due to simple mechanical faults (Björk, 1954). For example, in cases of trauma (resulting in fracture of the orbital floor or haemorrhage) or tumour (producing a displacement of the globe), the electromyogram will appear normal, indicating a mechanical rather than a neurogenic origin for apparent muscle paresis.

Congenital affections of the extra-ocular muscles may give rise to difficulties in diagnosis and in these conditions electromyography can be useful. In the superior oblique tendon sheath syndrome, even a struggling infant can be examined, for insertion of the electrode percutaneously will find the inferior oblique muscle. This will induce a Bell's phenomenon and thus ensure good firing of the action potentials, which should be normal if the diagnosis is correct, thus confirming the mechanical aetiology of the failure of elevation.

Cases of Duane's retraction syndrome are amenable to investigation with electromyography; it has been confirmed in such cases that the lateral rectus does not function normally, for almost no action potentials can be recorded on attempted abduction. Moreover, widening of the palpebral fissure is accompanied by an increase of action potentials in the levator palpebrae superioris (Breinin, 1957f).

Although the practical applications of electromyography of the extra-ocular muscles have obvious limitations, the following remarks on the electromyography of skeletal muscle (Gilliatt, 1957) can now be applied to the extra-ocular muscles:

"It is dangerous to prophesy, but one future trend is clear. In the past, clinical application of electromyography has been hindered by the complexity of apparatus required, and the investigation has usually been carried out by electrophysiologists in specially equipped departments. A steady simplification of recording apparatus is taking place, and commercial instruments are already being designed for those who have no special knowledge of electronics. As this process continues and suitable apparatus becomes available, it is likely that diagnostic electromyography will be performed increasingly by clinical neurologists themselves".

Summary

Simplified principles of electromyography of the extra-ocular muscles are presented. The information that can be gleaned by electromyography in lower motor neurone lesions, diseases of muscle, and disturbances of motility is outlined.

An explanation is offered for the rarity with which polyphasic potentials are recorded in the extra-ocular muscles during recovery from denervation.

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