THE EYE IN DYSTROPHIA MYOTONICA* WITH A REPORT ON ELECTROMYOGRAPHY OF THE EXTRA-OCULAR MUSCLES

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THOMSEN'S disease (myotonia congenita), dystrophia myotonica (myotonia atrophica), and paramyotonia are characterized by the presence of myotonia. As an isolated symptom, myotonia had already been described by Benedikt (1874) and Leyden (1874) when Thomsen (1876) published his paper (following the refusal by the Prussian army medical officers of a certificate of the disease in one of his sons), tracing the occurrence of myotonia in his family back to 1742. Erb (1886) established the clinical, electrical, and histological characteristics of the combination of congenital myotonia and muscular hypertrophy which became known as Thomsen's disease. It soon became apparent that in many cases the myotonia of Thomsen's disease might be associated with atrophy of the facial muscles, the muscles of mastication, the sternomastoids, the forearm muscles, and, less often, the vasti and peronei (Jolly, 1896; Hoffmann, 1900; Rossolimo, 1902; Pelz, 1907; Hunt, 1908; and others), and this led to the suggestion that such cases were distinct from those of Thomsen's disease (Batten and Gibb, 1909; Steinert, 1909) and were in fact examples of a specific condition, dystrophia myotonica. Steinert (1909) also emphasized the non-muscular dystrophic aspects of the disease—baldness, testicular atrophy with reduced potency, and acrocyanosis—and these were later elaborated by Curschmann (1912). The very rare disease of paramyotonia, in which the myotonia becomes apparent only in the cold, was described by Eulenburg (1886).

Dystrophia myotonica occurs more frequently than the other diseases in the myotonic group and has a far worse prognosis on account of the dystrophic features. Indeed, a tendency to early death in families carrying dystrophia myotonica has repeatedly been noted (Bell, 1947).

Ocular Signs in Dystrophia Myotonica

In dystrophia myotonica, characteristic muscle groups are affected by the dystrophic process, *viz.* the facial muscles including the muscles of mastication, the sternomastoid muscles, the muscles of the forearm and hand, and, later, the extensors of the knee and dorsiflexors of the foot. These are not, however, the only muscles to atrophy, for as the disease progresses most of the skeletal muscle becomes dystrophic. The orbicularis oculi muscles in a

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large proportion of cases are seriously affected and this results in infrequent blinking. This lagophthalmos may well explain the chronic conjunctival injection, often described as "blepharo-conjunctivitis", from which many of these patients suffer (*e.g.*, see Waring, Ravin, and Walker, 1940) and the occasional occurrence of keratitis (Birnbacher, 1927; Walsh, 1957). This chronic conjunctival irritation was present in eight of the ten patients examined electromyographically (*vide infra*). Moreover, the "increase in lacrimal secretion" described in patients with dystrophia myotonica (Ortleb, 1912; Hauptmann, 1916) may in fact be an epiphora resulting from the loss of tone in the dystrophic orbicularis oculi muscles which are unable to maintain the lower lid in contact with the globe. As is well known, the levator palpebrae superioris muscles are frequently affected by the dystrophic process, and this results in ptosis (enhanced by the concomitant enophthalmos), the lids becoming paper thin. Electromyography of the levators (*vide infra*) has confirmed the dystrophic nature of the wasting.

There is no doubt that enophthalmos is a common finding in dystrophia myotonica and deserves greater attention. Indeed, enophthalmos was found in nine out of ten patients personally observed (*vide infra*). Thomasen (1948) recorded enophthalmos as being "an almost invariable symptom" and suggested that it might be related to endocrine dysfunction. In a paper on the involvement of the heart in dystrophia myotonica, Fisch (1951) described five patients with abnormal electrocardiograms, all of whom had enophthalmos. That the sign is not related to muscle dystrophy or state of nutrition is borne out by its presence in patients with minimal muscle dystrophy and good nutrition. The commonly-found ptosis, together with the subsequent wasting of the dystrophic levator palpebrae superioris muscles, is made worse by the enophthalmos.

E. Brand (1950) reported the occurrence of low intra-ocular pressure in a case of dystrophia myotonica. This observation was followed by the examination of twelve cases of dystrophia myotonica, three cases of Thomsen's disease, and four cases of myotonia of doubtful classification (I. Brand, 1955). The cases of dystrophia myotonica all showed a lowered intra-ocular pressure, whereas the tension in the cases of Thomsen's disease was normal or near normal. The low intra-ocular pressure in the cases of dystrophia myotonica was attributed to "a functional disturbance of the centre in the hypothalamus that controls intra-ocular pressure". It was suggested that intra-ocular hypotonia was an early diagnostic sign by which dystrophia myotonica might be differentiated from Thomsen's disease when muscular atrophy had not yet developed and myotonia was the only common symptom.

A case of dystrophia myotonica with bilateral cataracts where the lower part of the corneae showed a few delicate opacities was reported by Maillard (1926); five weeks after extraction of the cataracts, bullae appeared in the epithelium of the cornea of the right eye together with an increase of the

grey-white opacities. Corneal sensation was diminished. Similar grey opacities and epithelial damage were found in the left cornea. After 3 months there was no further change, the condition being symptomless. Maillard thought this to be a trophic lesion, localized to the cornea, and made worse by operation. Birnbacher (1927) described a case of dystrophia myotonica in which irregularity of the corneal epithelium was found, the stroma being uninvolved. The endothelium was not seen because of the epithelial changes. There was infrequent blinking and diminished corneal sensation, the epithelial disturbance being most marked in the palpebral fissure. Birnbacher considered this to be an epithelial dystrophy arising from a trophic disturbance due to involvement of the fifth nerve in dystrophia myotonica. It would, however, seem that the condition described could be better explained on the basis of xerosis resulting from the lagophthalmos.

The occurrence of abnormal pupillary reactions in dystrophia myotonica has been reviewed by Maas and Paterson (1947), and would appear to be more than coincidental. The most interesting of the abnormalities, which varied greatly, was a myotonic reaction. If this is due to involvement of the sphincter iridis in the disease process, it is the only example of smooth muscle being affected.

Bartels (1906) had described the coincident occurrence of cataract with myotonia and muscular dystrophy in a family, thinking the lens changes were somehow related to tetany, but it was Greenfield (1911) who showed that the association of cataract with dystrophia myotonica was not fortuitous. In the family he described, cataract appeared at an earlier age in successive generations until finally dystrophia myotonica became manifest—that is to say anticipation occurred. This progressive inheritance is also accompanied by an increase in the severity of the disease in succeeding generations, *i.e.* potentiation (Adie and Greenfield, 1923; Ravin and Waring, 1939). Fleischer (1918, 1922), Vogt (1921), and Adie (1924), amongst others, soon confirmed Greenfield's findings. The characteristic lenticular opacities, as seen with the slit-lamp, are white dust-like subcapsular opacities associated with refractile iridescent particles that appear red, green, and blue. Cortical changes, starting at the posterior pole, appear later, and result in a stellate cataract which may not progress and is symptomless. Later the lens may become opaque, resulting in a senile cataract (Goulden, 1928; Gifford, Bennett, and Fairchild, 1929; Caughey, 1933). All of the ten patients examined electromyographically (*vide infra*) showed subcapsular iridescent particles.

Optic atrophy occurring in dystrophia myotonica was first noted by Löhlein (1914), the right disc in his case being pale yellow and atrophic with indistinct edges. This was associated with a nasal depression of the peripheral isopters and an arcuate scotoma of nerve-fibre bundle type extending downwards from the blind spot. The left field of vision after

cataract extraction showed a concentric contraction. Hauptmann (1918) reported a further case in which he found pallor of the disc but the visual fields were not recorded. Three of the fifteen cases of dystrophia myotonica detailed by Heine (1925) had abnormally pale discs, and one of these three patients, in whom there was temporal pallor, had a central scotoma.

The concurrence of dystrophia myotonica and retinitis pigmentosa was observed in a single case by Godtfredsen (1949), who thought that this might be an entity linked to the heredo-degenerative abiotrophic disorders of the central nervous system. Bilateral macular degeneration has been described in a patient with dystrophia myotonica (Verrey, 1947), but the two conditions were probably unrelated. Disturbances of development, such as microphthalmos, coloboma of the choroid and retina, and "congenital amaurosis", have been reported (Babellis Gorina and de Gispert Cruz, 1948).

Electromyography of the Extra-Ocular Muscles in Dystrophia Myotonica

Methods.—Ten patients with dystrophia myotonica, confirmed by muscle biopsy and electromyography of limb muscles, were examined. After topical anaesthesia, concentric needle electrodes (No. 30 gauge) were inserted into the levator palpebrae superioris through the skin of the upper lid in all cases, and transconjunctivally into the muscle bellies of the horizontal recti in seven cases. Insertion and action potentials were recorded at rest and on attempted movement on a "Medelec" single channel electromyograph recorder, and were photographed with a "Cossor" oscillograph camera. Some potentials were recorded on a magnetic memory and subsequently analysed.

Findings.—As was to be expected, a direct correlation was found between the degree of ptosis and the amplitude of the action potentials (Figs 1 and 2).

FIG. 1.—Electromyogram of levator palpebrae superioris on upward gaze. Minimal ptosis. In this and the following figures calibration equals $100 \mu\text{v}$ and time is measured in milliseconds.

FIG. 2.—Electromyogram of levator palpebrae superioris on upward gaze. Father of patient shown in Fig. 1. Gross ptosis.

Compare Trace A which is to the same scale as Fig. 1. As the gains are increased (Traces B and C), abnormally short-duration low-voltage potentials become apparent.

It was also noted that the characteristic high-frequency discharge (so called “dive-bomber” sound) of dystrophia myotonica was easily provoked in seven cases by moving the electrode in the levators (Fig. 3).

FIG. 3.—Insertion potentials obtained from levator palpebrae superioris—“dive-bomber” effect.

A curious feature of the traces obtained from the levator palpebrae superioris muscles was the notched appearance of many of the action potentials (Fig. 4, overleaf). That this may be an early stage in the disintegration of the action potentials is suggested by its occurrence in early stages of metabolically determined and presumably reversible myopathies, such as that accompanying thyrotoxicosis (Hed, Kirstein, and Lundmark, 1958; Woolf, 1960).

FIG. 4.—Electromyogram of levator palpebrae superioris. Note notched appearance of action potentials.

It is difficult to recognize polyphasic potentials in ocular electromyograms because of the fullness of the interference pattern on minimal effort. However, in one case, such potentials were unusually prominent both because of the degree of polyphasicity and because of the long duration (Fig. 5).

FIG. 5.—Trace A shows several polyphasic potentials. Trace B, faster scan, analyses one polyphasic potential of 6 msec. duration.

The horizontal recti were examined in seven patients and in five of them abnormal electromyograms were obtained. This is of particular interest as these muscles showed no abnormality on clinical examination. The “dive bomber” effect was obtained from four patients (Fig. 6) and analysis of the potentials showed them to be of less than one millisecond duration (Fig. 7), the normal being one to two milliseconds, and of an amplitude which not infrequently exceeded that of the action potentials. The amplitude of the latter, as obtained from the medial and lateral recti, were usually within normal limits in contrast to the low potentials obtained from the levators (Fig. 8*a, b*, overleaf). In some cases the frequency of the potentials, as well

FIG. 6.—Electromyogram of lateral rectus, showing characteristic high-frequency discharges.

FIG. 7.—Faster scan of potentials obtained in Fig. 6.

as their short duration, suggested that there had been a fragmentation of the motor unit action potentials, such as is characteristically seen in traces from the limb muscles in cases of muscular dystrophy (Fig. 8*b*, overleaf).

One of the cases examined warrants a detailed description:

A boy aged 15 years, of poor intellect, complained that his muscles had been weak and wasted "for a long time". Drooping of his lids had been present since age 13 and his eyes had been immobile during the last 18 months. Examination revealed a youth with marked wasting of all the skeletal muscles (poor power and tone difficult to evaluate), Hutchinsonian facies, and perspiring profusely. There was a small testis in the scrotum, the other being undescended and impalpable in the inguinal canal. Bilateral ptosis was associated with an external ophthalmoplegia, the pupillary reactions being normal. The fundi were normal and there was some weakness of the orbicularis oculi muscles. Slit-lamp examination showed slight posterior cortical changes with a few subcapsular iridescent particles. Electromyography of the horizontal recti yielded high-frequency potentials (Fig. 9*A*, overleaf), the characteristic "dive-bomber" sound effect being obtained from no other muscles in the body. Biopsy of a forearm muscle gave histological confirmation of the diagnosis of dystrophia myotonica.

FIG. 8a.—Levator palpebrae superioris.

FIG. 8b.—Lateral rectus.

While the ocular muscles in all the cases of dystrophia myotonica mentioned above yielded electromyographic changes similar to those encountered in the limb muscles in this disease, it was only possible in this one case to demonstrate in the ocular muscles the myotonia, *i.e.* continued activity after cessation of volitional effort. This was in fact the only case showing myotonia clinically—the patient was unable to relax the recti on attempted lateral deviation. This failure of relaxation was associated with electromyographic evidence of continued firing of motor units instead of the electrical silence usually present under these circumstances (Fig. 9C, opposite). The firing, however, was not very marked.

Our only case of Thomsen's disease differed strikingly from the cases of dystrophia myotonica, not only in the absence of ptosis, but also in the prominence of the motor unit discharge observed electromyographically in the levator palpebrae superioris on looking down (Fig. 10, opposite). This phenomenon was not accompanied by definite lid-lag.

FIG. 9.—Electromyogram of medial rectus in a male patient aged 15 years.

- (A) Medial gaze.
- (B) Gaze straight ahead.
- (C) Lateral gaze.

FIG. 10.—Electromyogram of levator palpebrae superioris in a case of Thomsen's disease.

Trace A (left to right). Normal flat base line of electrical silence, action potentials on gaze up, calibration 100 μ V.

Trace B. Continued activity on gaze down when there should be a flat base line as in first part of Trace A.

Histology of the Extra-Ocular Muscles in Dystrophia Myotonica

In spite of the frequency of ptosis as a sign in dystrophia myotonica, a survey of most of the autopsies in the literature (Steinert, 1909; Hoffmann, 1919; Fischer, 1920; Hitzemberger, 1920; Bramwell, 1922; Adie and Greenfield, 1923; Weil and Keschner, 1927; Guillain, Bertrand, and Rouquès, 1932; Keschner and Davison, 1933; Gonzalez Segura and Lanari, 1941; Aranovich, 1946; Black and Ravin, 1947; Benda and Bixby, 1947; Nadler, Steiger, Troncelletti, and Durant, 1950; den Hartog Jager, 1951; Fisch and Evans, 1954) reveals only a single histological study of an extra-ocular muscle, namely that of Wohlfart (1951), who examined the levator palpebrae

superioris. He noted "accumulations and rows of nuclei in some fibres and a few scattered hypertrophic fibres with large vacuoles or a central hollow". On the only occasion that the globe was examined (Vos, 1936), atrophy of the ciliary body was found, and this may be the explanation of the low intra-ocular pressure observed by Brand (1955).

The following histological description of muscle obtained *post mortem* from a patient who had suffered from dystrophia myotonica with myotonia of the forearm and leg, together with atrophy of the sternomastoids, forearm muscles, vasti, and peronei, was given by Dr. A. L. Woolf.

HISTOLOGY

Sternomastoid.—Very few muscle fibres remain and these show great variation in diameter without any grouping according to size. There is some slight increase in interstitial, fibrous, and fatty tissue.

Vastus internus.—A minority of the muscle fibres show prominent trains of rectangular, centrally-placed nuclei. There are occasional slender muscle fibres but these are quite exceptional.

Peroneus.—Chains of nuclei are very conspicuous and some of these show fusion of the nuclei. Some of the laterally placed sarcolemmal nuclei are also fused. Atrophy of muscle fibres is more pronounced though not severe. There are also some *Ringbinden*.

Levator palpebrae superioris.—Occasional muscle fibres show centrally-placed lines of nuclei which have fused into one another. There are also slightly more frequent lines of fused nuclei within very slender muscle fibres, the small size of which makes it impossible to determine whether they originally had a central or peripheral position (Fig. 11, opposite).

Superior rectus.—There are many muscle fibres with centrally-placed, but often quite widely-separated, nuclei, which are elongated and approximately rectangular. Some of the nuclei are large and almost devoid of chromatin. Some of these muscles have very little striation and considerable quantities of lipo-fuchsin. There are also areas of proliferation of the interstitial cells, probably fibroblasts, with an occasional myophage.

External rectus.—No abnormal features noted.

Internal rectus.—Occasional muscle fibres show centrally-placed elongated nuclei.

Superior oblique.—Again occasional muscle fibres show centrally-placed elongated nuclei and very occasionally there are groups of fused sarcolemmal nuclei. There is one focus of lymphocytic infiltration (Fig. 12, opposite).

It will be seen that the histological changes in the extra-ocular muscles are very slight. In view of the fact that striking electromyographic abnormalities were observed in the traces from the extra-ocular muscles in all the cases studied, it would seem reasonable to suggest tentatively that the changes demonstrable electrically precede histological evidence of dystrophy.

Discussion

In dystrophia myotonica, the localization of the myotonia often differs from that of the dystrophy. For example, myotonia is most commonly apparent in the forearm muscles whereas dystrophy is most pronounced in the facial and sternomastoid muscles. Myotonia is rarely present in the facial muscles in dystrophia myotonica but is commonly found there in

FIG. 11.—Levator palpebrae superioris. Haematoxylin and eosin. $\times 140$.

FIG. 12.—Superior oblique. Haematoxylin and eosin. $\times 168$.

Thomsen's disease, especially in the orbicularis oculi, when it results in an inability to open the eyes after forcible closure of the lids. The extra-ocular muscles are often involved in the myotonic process in Thomsen's disease and this results in lid-lag (see Sedgwick, 1910, who traced this sign through five generations in a family with Thomsen's disease), squint, and diplopia, especially on moving the eyes suddenly (Jacoby, 1887; Raymond, 1891;

Meara, 1905; Toomey, 1916; Thomasen, 1948). It is of interest that we were able to demonstrate this myotonic failure of relaxation in the levator palpebrae superioris (see Fig. 10) in the absence of obvious lid-lag.

There have been very few reports of extra-ocular muscle involvement in dystrophia myotonica. Verbiest (1937) reported a case of dystrophia myotonica in which convergence resulted in a myotonic reaction in the medial recti together with pupillary contraction, citing this as evidence of a convergence centre and of a central origin for the myotonia. In support of their contention that Thomsen's disease represents an early stage of dystrophia myotonica, Maas and Paterson (1939) reported two patients who, besides showing myotonic and dystrophic signs in the limbs, had difficulty in moving their eyes quickly, which in one of them resulted in double vision and squint. Verrey's case (*vide supra*) showed limitation of extra-ocular muscle movement, most marked at the extremes of age. Walton and Nattrass (1954), in an important paper, cited a further patient who, at the age of 12 years, showed generalized muscular hypertrophy and myotonia typical of Thomsen's disease. By age 24, she still showed muscular hypertrophy and severe myotonia in the limbs and trunk with no wasting of the limb muscles. There was, however, some wasting of both sternomastoids, slight facial weakness, impairment of ocular movements in all directions (in addition to a concomitant strabismus), and bilateral early cataracts; that is to say that she had developed features of dystrophia myotonica. These authors thought that their case supported the suggestion of Maas and Paterson (1939, 1950) that dystrophia myotonica and Thomsen's disease are different manifestations of the same disease. Walton and Nattrass (1954) had also examined fifteen patients with dystrophia myotonica, fourteen of whom had ptosis. Six of their patients had symmetrical involvement of the extra-ocular muscles; three of them had only a few degrees of ocular movement in any direction, while similar but less severe impairment was apparent in the others.

It has been shown that the electrical phenomena accompanying myotonia are the same in both dystrophia myotonica and Thomsen's disease (Buchthal and Clemmesen, 1941), and in dystrophia myotonica Thomasen (1948) demonstrated these phenomena in muscles without active or mechanical myotonia, thus indicating a more extensive lesion than was apparent clinically. Our electromyographic studies of the extra-ocular muscles in dystrophia myotonica have confirmed the involvement of the levator palpebrae superioris and have shown an undoubted abnormality of the horizontal recti. It is well known that slight movements do not usually elicit myotonia, so that blinking, and the movements of facial expression, etc., are not impeded, whereas strong closure of the eyes or clenching of the fist will be followed by a long delay in relaxation (Adams, Denny-Brown, and Pearson, 1953). Moreover, relaxation of the myotonia becomes more rapid with repetition of the movement. This may be the explanation of the compara-

tive rarity with which myotonia has been clinically demonstrated in the extra-ocular muscles in dystrophia myotonica, since their continual fine, saccadic, and searching movements presumably prevent the myotonia from disturbing their smooth action. (This is borne out by the patient reported by Hirschfeld (1911) who, after waking in the morning, saw everything double for 3 to 5 minutes). The relatively frequent occurrence of extra-ocular muscle involvement in Thomsen's disease provides a contrast of great interest but still lacks an explanation.

In dystrophia myotonica, the myotonia is regularly followed within a few years by atrophy, and in cases in which limitation of ocular movement develops after the disease has been in progress for several years and in which atrophy is already present in the limb muscles (see Maas and Paterson, 1939; Walton and Nattrass, 1954), it is possible that the basis of the limited ocular movement is dystrophic rather than myotonic, as is certainly the case with the lids.

The report of Walton and Nattrass (1954) shows that involvement of the ocular muscles in dystrophia myotonica is much more frequent than has formerly been observed, and our own findings go further in suggesting that, if electromyography is carried out, the extra-ocular muscles will be found to be involved in the majority of cases of this disease.

Summary

The ocular signs in dystrophia myotonica are reviewed.

Electromyography of the extra-ocular muscles yielded abnormal and characteristic traces in each of ten patients suffering from this disease.

The histological findings in the extra-ocular muscles in a case of dystrophia myotonica are reported.

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