Computer method for the analysis of evoked motor unit potentials

2. Duchenne, limb-girdle, facioscapulohumeral and myotonic muscular dystrophies

JOHN P. BALLANTYNE and STIG HANSEN

From the University Department of Neurology, Institute of Neurological Sciences and the Department of Clinical Physics and Bio-engineering, Glasgow

SYNOPSIS Single motor unit potentials recorded from surface electrodes over the extensor digitorum brevis muscle and evoked by stimulation of the anterior tibial nerve at the ankle were obtained by a computer subtraction method. Their latencies, durations, amplitudes, and areas were measured in control subjects and patients with Duchenne, limb-girdle, facioscapulohumeral, and myotonic muscular dystrophy. Lateral popliteal motor nerve conduction velocities were also recorded. In the muscular dystrophies there was a significant increase in both the latencies and durations of motor unit potentials, the latter in notable contrast with the findings of conventional needle electromyography. Fastest motor conduction velocities were significantly reduced in the limb-girdle, facioscapulohumeral, and myotonic muscular dystrophy patients, while the shortest distal motor latencies were significantly prolonged in these patients and those with Duchenne muscular dystrophy. The results support the presence of a definitive neurogenic influence in the muscular dystrophies.

In an earlier study we described a computer subtraction technique for the isolation and measurement in man of motor unit action potentials (MUP) recorded from surface electrodes over the extensor digitorum brevis muscle (EDB) and evoked by stimulation of the anterior tibial nerve at the ankle (Ballantyne and Hansen, 1974b). The area, amplitude, duration, and latency of each MUP can be measured, providing indices of the electrophysiological activity in the motor unit and of the functional integrity of the innervating axon and/or neuromuscular junction. We have stated our preference for surface rather than concentric needle electrode recording in this situation (Ballantyne and Hansen, 1974b) as the action potential obtained more accurately reflects the number of muscle fibres in the motor unit and is relatively uninfluenced by muscle fibre density.

The purpose of this paper is to present the results obtained from the application of this technique to patients with Duchenne, limb-girdle, and facioscapulohumeral myotonic muscular dystrophies.

METHODS

All values are expressed as mean ± one standard deviation (SD). Mean values were compared using Student's t test.

SUBJECTS 1. Controls Thirty-three subjects, members and relatives of the staff of the Institute of Neurological Sciences, were investigated. None had evidence of neurological disease. Twelve of these subjects aged $10.7 \pm 2.2$ years were used as controls for the Duchenne study, while the other 21 normal subjects aged $38 \pm 14$ years formed a control group for the limb-girdle, facioscapulohumeral, and myotonic dystrophies.

2. Duchenne muscular dystrophy Twelve boys aged $10 \pm 2.9$ years were studied. In all cases, the primary
established the initial computer. The computer nerve and rate distal phases. of individual MUPs have properties to ankle measured tibial TE

3. Limb-girdle and facioscapulohumeral muscular dystrophies Twelve patients aged 37 ± 17.4 years were studied, of whom three had facioscapulohumeral muscular dystrophy. The mean duration of symptoms was 14 ± 8.2 years. The diagnosis was reached on the basis of clinical examination, muscle biopsy (seven patients), and electromyography (EMG) (12 patients). Serum enzyme studies, performed on all patients, were normal to slightly elevated. Only those patients in whom the EMG appearances were considered myopathic were included in the study.

4. Myotonic muscular dystrophy Twelve patients aged 45 ± 14.0 years were studied. The duration of the symptoms was 16 ± 12.2 years. In all patients the clinical and electromyographic features were characteristic of myotonic muscular dystrophy. In only one patient was a muscle biopsy performed and this was reported to show myopathic changes.

Motor unit counts from the EDB muscles of these patients have been reported (Ballantyne and Hansen, 1974c).

TECHNIQUES The composition and placement of the surface recording electrode over the EDB muscle, the properties of the stimulating electrodes over the anterior tibial nerve at the ankle, and details of the rate and strength of stimulation used to evoke MUPs have been described previously (McComas et al., 1971b; Ballantyne and Hansen, 1974a). The description of the amplification and display systems and of the computer handling of data for the isolation of individual MUPs has been presented (Ballantyne and Hansen, 1974b). The values for area and peak to peak amplitudes of MUPs were provided by the computer. The latency and duration of each MUP were measured manually from the computer printout, the former from the time interval between the onset of the stimulus artefact and the initial deflection of the potential in the print-out, the latter from the initial deflection to the final return of the baseline to zero potential including positive and negative phases.

Fastest motor conduction velocities in the lateral popliteal nerve (knee to ankle segment) and shortest distal motor latencies (anterior tibial nerve at the ankle to the EDB muscle) were measured using an established technique (Hodes et al., 1948). The evoked muscle action potential was recorded from surface electrodes over the EDB muscle as above. The limb temperatures were maintained at 33°C ± 1°C.

RESULTS

1. MOTOR UNIT PARAMETERS Control subjects

Both the mean amplitude and area of the MUPs of adult controls are significantly greater than those of the young control group (Tables 1 and 2). The mean duration and mean latency are similar in the two groups (Tables 3 and 4).

<table>
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<tr>
<th>Table 1</th>
<th>MOTOR UNIT POTENTIAL AMPLITUDES</th>
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<tbody>
<tr>
<td>Number of potentials</td>
<td>Amplitude of potentials* (mV)</td>
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<tr>
<td>Controls</td>
<td>160</td>
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<tr>
<td>Limb-girdle dystrophy</td>
<td>92</td>
</tr>
<tr>
<td>Myotonic dystrophy</td>
<td>107</td>
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<tr>
<td>Controls</td>
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<td>Duchenne dystrophy</td>
<td>97</td>
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* Mean ± SD.

<table>
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<tr>
<th>Table 2</th>
<th>MOTOR UNIT POTENTIAL AREAS</th>
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<td>160</td>
</tr>
<tr>
<td>Limb-girdle dystrophy</td>
<td>92</td>
</tr>
<tr>
<td>Myotonic dystrophy</td>
<td>107</td>
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<tr>
<td>Controls</td>
<td>111</td>
</tr>
<tr>
<td>Duchenne dystrophy</td>
<td>97</td>
</tr>
</tbody>
</table>

* Mean ± SD.

Duchenne muscular dystrophy In these patients there is a significant reduction in the mean amplitude of MUPs (Fig. 1, Table 1) with an increase in the percentage of low amplitude MUPs but none with amplitudes less than the lower limit of the normal range. The mean area of the MUPs (Fig. 1, Table 2) is slightly but not significantly reduced in these patients but mean
duration (Fig. 2, Table 3) is significantly prolonged. The mean latency of MUPs is considerably prolonged (Fig. 2, Table 4) and many units have latencies greater than the upper limit of the control range.

**Limb-girdle and facioscapulohumeral muscular dystrophies** The mean value for MUP amplitudes is not significantly greater than that of the control group (Fig. 3, Table 1) but mean area is significantly increased with a fall in the percentage of small area MUPs (Fig. 3, Table 2) and a rise in the percentage of large area MUPs. The mean duration of the MUPs (Fig. 4, Table 3) is significantly prolonged as is mean latency (Fig. 4, Table 4), with a considerable increase in the

### TABLE 3

<table>
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<tr>
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<th>Number of potentials</th>
<th>Duration of potentials* (ms)</th>
<th>P</th>
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<tr>
<td>Controls</td>
<td>160</td>
<td>9.50 ± 1.70</td>
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<td>Limb-girdle dystrophy</td>
<td>92</td>
<td>10.42 ± 1.63</td>
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<td>Myotonic dystrophy</td>
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<td>11.29 ± 2.23</td>
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<td>Controls</td>
<td>111</td>
<td>9.32 ± 1.63</td>
<td>—</td>
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<td>Duchenne dystrophy</td>
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<td>10.32 ± 2.10</td>
<td>&lt;0.001</td>
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* Mean ± SD.

### TABLE 4

<table>
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<td>5.65 ± 1.46</td>
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<td>Duchenne dystrophy</td>
<td>97</td>
<td>5.36 ± 1.34</td>
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* Mean ± SD.

**FIG. 1** Histograms of MUP amplitudes and areas. Young control subjects and patients with Duchenne muscular dystrophy.
FIG. 2  Histograms of MUP durations and latencies. Young control subjects and patients with Duchenne muscular dystrophy.

FIG. 3  Histograms of MUP amplitudes and areas. Adult control subjects and patients with limb-girdle and facioscapulohumeral muscular dystrophy.
percentage of long latency MUPs. Some of the latter are considerably longer than the upper limit of the normal range.

**Myotonic muscular dystrophy**  The mean amplitude of MUPs is significantly increased (Fig. 5, Table 1) as is mean area (Fig. 1, Table 2) with an increase in the percentage of large area MUPs. Mean MUP duration is also significantly increased, many have durations greater than the upper limit of the normal range (Fig. 6, Table 3) and the mean latency of MUPs is significantly increased.

### Table 5

<table>
<thead>
<tr>
<th>No. of patients</th>
<th>Mean age (yr)</th>
<th>Fastest motor conduction velocity* (m/s)</th>
<th>P</th>
<th>Shortest distal latency* (ms)</th>
<th>P</th>
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<tbody>
<tr>
<td>Controls</td>
<td>20</td>
<td>38</td>
<td>—</td>
<td>3.43 ± 0.46</td>
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<tr>
<td>Limb-girdle dystrophy</td>
<td>12</td>
<td>37</td>
<td>&lt;0.01</td>
<td>4.22 ± 0.71</td>
<td>&lt;0.001</td>
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<tr>
<td>Myotonic dystrophy</td>
<td>10</td>
<td>45</td>
<td>&lt;0.001</td>
<td>4.99 ± 0.87</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Controls</td>
<td>12</td>
<td>11</td>
<td>—</td>
<td>3.27 ± 0.61</td>
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<tr>
<td>Duchenne dystrophy</td>
<td>12</td>
<td>10</td>
<td>NS</td>
<td>4.53 ± 0.74</td>
<td>&lt;0.001</td>
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* Mean ± SD.
FIG. 5 Histograms of MUP amplitudes and areas. Adult control subjects and patients with myotonic muscular dystrophy.

FIG. 6 Histograms of MUP durations and latencies. Adult control subjects and patients with myotonic muscular dystrophy.
prolonged, in some cases, to almost twice that of the upper limit of the normal range (Fig. 6, Table 4).

2. MOTOR CONDUCTION VELOCITIES AND DISTAL MOTOR LATENCIES (Table 5) The fastest motor conduction velocities in the lateral popliteal nerves are within the normal range in the Duchenne dystrophy patients, but significantly reduced in the limb-girdle, facioscapulohumeral, and myotonic muscular dystrophies. The shortest distal motor latencies are significantly prolonged in all three groups of patients with muscular dystrophy.

DISCUSSION

Sacco et al. (1962) reported that both the durations and amplitudes of voluntary MUPs increase from infancy to maturity. In our young control subjects we, too, have found that the amplitudes of the evoked MUPs are reduced but their durations are comparable with values in adult subjects. Examination of the data of Sacco et al. (1962) indicates that this parameter may have reached adult values by the age of 11 or 12 years, the mean age of our young controls.

DUCHENNE MUSCULAR DYSTROPHY Needle electrode studies show a preponderance of short duration, low amplitude often polyphasic MUPs in Duchenne muscular dystrophy (Kugelberg, 1947; Buchthal and Rosenfalck, 1963). The reduction in duration of the potentials is a consequence of the reduced muscle fibre density in the motor units whereby the early and late components of the action potential are lost (Buchthal et al., 1960). In the normal individual these components are produced by volume conduction to the relatively small recording surface of the concentric needle electrode of action potentials of distant muscle fibres (Buchthal et al., 1960) and these are reduced in numbers in Duchenne muscular dystrophy. The potential recorded with the much larger surface electrode is more dependent on the number of muscle fibres in the unit than on their spatial distribution (Kaeser, 1970), so that the circumstances leading to the shortening of the MUPs found in needle studies do not occur.

We have noted a significant increase in the duration of the MUPs in these patients. Prolongation of the duration of the MUP may be attributed to a reduction in the propagation velocity of the action potential in individual muscle fibres which would cause an increase in dispersion of these potentials. This explanation is unlikely, as Buchthal et al. (1960) and Buchthal and Rosenfalck (1963) reported normal conduction velocities in dystrophic muscle fibres despite the known variation in muscle fibre diameter in this condition. In the normal individual the duration of the MUP is almost exclusively determined by the degree of dispersion of the innervation zone of the motor unit (Buchthal et al., 1955, 1957). In a primary myopathy general thinning out of muscle fibres in the muscle unit (Buchthal et al., 1960) cannot lead to an increase in the dispersion of the innervation zone. In the normal subject Buchthal et al. (1957) have reported that differences in the conduction time in the intramuscular nerve fibres contribute no more than 0.5 ms to the duration of the motor unit potential. However, in the presence of a slowing of conduction in these fibres this factor will become of increasing importance. We have found a significant increase in both the shortest distal motor latencies (Table 5) and in the latencies of individual MUPs (Fig. 2, Table 4) in Duchenne dystrophy patients. As the conduction velocity in the knee to ankle segment of the motor nerve was normal (Table 5), we submit that prolongation of the distal latency is a consequence of slowing of conduction in intramuscular nerve twigs and/or an increased delay at the neuromuscular junction. Decrementing muscle action potential responses at fast tetanization rates have been reported in Duchenne dystrophy (McComas et al., 1970). This need not indicate a disturbance in endplate function as Stålberg and Thiele (1972) have shown that decrementing responses may occur when there is blocking of transmission in intramuscular nerve fibres and that the degree of blocking increases with rising frequency of stimulation. They also reported the occurrence of 'paired blocking' of muscle fibre action potentials in cases of progressive muscular dystrophy indicating a disturbance of impulse conduction in the intramuscular nerve fibres. Furthermore, there is no evidence on light or electron microscopy of any anatomical abnormality of the motor endplate in Duchenne
muscular dystrophy (Jerusalem et al., 1974), unlike the known association between the neuromuscular transmission defect and the morphological changes at the endplate in myasthenia gravis (Bickerstaff and Woolf, 1960; MactDermot, 1960; Cöers et al., 1966; Desmedt, 1966; Simpson, 1971). While we cannot exclude the possibility that an increase in neuromuscular delay contributes to the prolongation both of MUP latencies and of distal motor latencies, the evidence would support the additional occurrence of slowing of conduction in the intramuscular nerve fibres. The differences in conduction time in these fibres will lead to a reduction in synchronization of individual muscle fibre potentials and an increase in the durations of MUPs. The small increase in duration of the muscle fibre action potential noted during intracellular recording in dystrophic muscles (Ludin, 1973) is insufficiently large to contribute significantly to the duration of the MUP.

The amplitude of the surface recorded MUP depends on the total number of muscle fibres in the unit and the temporal summation of their action potentials. Histological studies (Buchthal et al., 1960; Adams, 1974) indicate a reduction in the number of muscle fibres in the muscles in Duchenne dystrophy. We have reported normal numbers of motor units in the EDB muscles of these patients (Ballantyne and Hansen, 1974c). There is therefore a reduction in the number of muscle fibres in each motor unit. The amplitude of the muscle fibre action potential is reduced in dystrophies (Ludin, 1973). Both these factors contribute to the reduction in the amplitude of the MUP noted in this study.

While the mean area of the MUPs in Duchenne dystrophy is reduced compared with the control values, this is less marked than we would have anticipated in view of the reduction in the number of muscle fibres noted histologically in dystrophic muscles. The explanation for this finding is not clear but there are several possible interpretations. Examination of Ludin's data (1973) shows that, despite the reduction in amplitude of the spike potential in dystrophic muscle fibres, the area under the potential is moderately increased due to the prolongation of the potential duration. This will militate against a reduction in MUP area despite a loss of muscle fibres. Secondly, as the MUP area is composed of the summated biphasic or polyphasic potentials of individual muscle fibres, the increased temporal dispersion of these potentials which alters their phase relationships may lead to a disproportionate change in the area of the MUP. Finally, should polyneuronal innervation occur in dystrophic muscle such that some fibres are common to several motor units, the mean population of fibres in individual units would not fall in proportion to the total reduction in muscle fibres in the whole muscle. Woolf and Coërs (1974) have noted that multiterminal innervation may occur in muscular dystrophies and Hunt and Kuffler (1954) have claimed that polyneuronal innervation is present in normal muscles of the cat. Recently, Tate and Westerman (1973) reported multiple innervation of muscle fibres in the soleus muscle of the cat during reinnervation. Buller (1974) considers the question of polyneuronal innervation of skeletal muscle 'is not yet settled'. Which if any of these hypotheses are pertinent to the problem cannot be answered at this stage, but we hope to pursue this further.

**MYOTONIC MUSCULAR DYSTROPHY** In this group of patients there is significant increase in all parameters of the MUP and this is associated with reduction in the number of motor units in the muscle (McComas et al., 1971a; Ballantyne and Hansen, 1974c). These are not the expected findings in a primary myopathy. Larger than normal units should not occur. Our results support the conclusion of McComas et al. (1971a) that some of the surviving motor units in this condition have enlarged by collateral reinnervation of muscle fibres which have lost their primary innervation as the disease progresses. This will lead to an increase in both the amplitude and area of the MUP but without effect on its duration (Buchthal et al., 1960). Prolongation of the latencies of the MUPs and of the distal motor latencies may be in part due to a disturbance of neuromuscular transmission. Histological studies (MacDermot, 1961) have demonstrated abnormalities of the motor endplates and decrementing responses to tetanization are known to occur in myotonic dystrophy (McComas et al., 1971a). The site of this dysfunction could, however, lie in intramuscular nerve fibres rather than at the endplates. We
have already referred to the studies of Stålberg and Thiele (1972) in this context. Histological abnormalities of intramuscular nerve twigs have been reported (Allen et al., 1969; Kito et al., 1973). The reduction in conduction velocity in the proximal segment of the motor nerve and the prolonged distal latency which we have found in this condition (Table 5) support the presence of a disturbance of function in the motor axons and intramuscular nerves. As in the Duchenne dystrophy patients, the prolongation of the distal motor latencies and the latencies of individual motor unit potentials must in part be due to reduced conduction velocity in the intramuscular nerve fibres leading in turn to a prolongation of MUP durations.

**LIMB-GIRDLE AND FACIOSCAPULOHUMERAL MUSCULAR DYSTROPHIES** The changes in dimensions of the MUPs are intermediate between those noted in the Duchenne and myotonic muscular dystrophy groups. There is a rise in the percentage of MUPs of increased duration (Fig. 4) and area (Fig. 3) but the mean amplitude remains little changed. Figure 3 does, however, show that there has been a very small increase in the percentage of low and high amplitude MUPs. We have reported normal motor unit numbers in these patients (Ballantyne and Hansen, 1974c). This alteration in MUP parameters is probably the electrophysiological reflection of a pathological remodelling of the motor units. In this group of patients there is also a significant reduction in the fastest motor conduction velocities and an increase in distal motor latencies. We consider that the explanations for these changes are similar to those discussed above for Duchenne and myotonic muscular dystrophies.

If, as we have postulated above, slowing of conduction velocity in the intramuscular nerve fibres is mainly responsible for both the prolonged distal latencies and increased durations of the MUPs in these conditions, then we should expect to demonstrate a relationship between these two parameters. That such is indeed present is shown in Fig. 7 where there is a highly significant correlation (r = 0.992) between mean MUP durations and latencies.

It should be emphasized that MUP amplitudes quoted in this paper are (under our recording conditions) absolute and are not equivalent to the values quoted by McComas et al. (1971a, b, c), Sica and McComas (1971), and Panayiotopoulos et al. (1974). These authors measured the increments in amplitude of the muscle action potentials on the addition of each newly recruited MUP and assumed that these values were proportional to the absolute amplitudes of the contributing MUPs. This assumption is correct only if each MUP is of identical latency, duration, and shape with temporal coincidence of the potential maxima. Alternatively, the potential maxima of incrementing MUPs must always coincide irrespective of their latencies or durations. The results of the present study do not substantiate these assumptions. We have found a considerable variation in the latencies and durations of individual MUPs obtained by computer subtraction in both control subjects and patients with muscular dystrophy. Furthermore, during motor conduction velocity studies electromyographers are familiar with the increase in potential duration and the shortening of the latency to the initial deflection and peak amplitude of the muscle compound action potential that occurs as the intensity of stimulation to the

![Graph showing relationship between mean MUP durations and latencies.](http://jnnp.bmj.com/)

motor nerve is increased. These appearances are due to the recruitment of MUPs of different initial and peak amplitude latencies. The summation factor (Table 6) calculated from the mean amplitude of individual MUPs obtained by computer subtraction, and that obtained from MUP increments to the compound muscle action potential, is an index of the lack of coincidence of the potential maxima of the recruited MUPs. While this factor is comparable in the two control groups, it is increased in the muscular dystrophies (Table 6), indicating that the temporal relationships of the potential maxima of the MUPs do not conform in these pathological states to the pattern found in control subjects. The amplitudes of the MUP increments are not proportional to absolute MUP amplitudes when comparisons are made between control subjects and patients with neuromuscular disease.

CONDUCTION VELOCITY STUDIES (Table 5)
The responses in the EDB muscle evoked by stimulation of the anterior tibial nerve at the ankle were recorded in control subjects and patients from surface electrodes over that muscle. When concentric needle electrode recording is used the shortest latency motor units may lie outside the pick-up zone of the electrode (Simpson, 1964; Kaeser, 1970), producing spurious prolongation of the distal motor latencies by up to 1 ms (Simpson, 1964). In a group of our subjects in whom the distal motor latency was simultaneously recorded by both methods the latency to the needle electrode was never less than, and in most cases greater than, that to the surface electrode. Since the needle electrode may not record from the motor units of shortest latency, it may not record from the motor units whose axons have the fastest motor conduction velocities. This will lead to an underestimation of the fastest motor conduction velocities in the proximal segment of the nerve. For the measurement of shortest distal motor latencies and fastest motor conduction velocities surface recording electrodes must be used (Kaeser, 1970).

In the Duchenne patients and their controls the values we have obtained for fastest motor conduction velocities and shortest distal motor latencies (Table 5) are similar to those reported by McComas et al. (1971c). However Nakao et al. (1968) reported slowing of conduction in the proximal segments of median and tibial nerves but they did not measure distal motor latencies.

Kito et al. (1973) noted histological abnormalities in peripheral nerves in myotonic patients and stated that conduction velocities were depressed. Although McComas et al. (1971a) found a reduction in the mean value for motor conduction velocity in their myotonic dystrophy patients, it was not significantly different from control values. In the limb-girdle dystrophy group Sica

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**TABLE 6**

<table>
<thead>
<tr>
<th>No. of patients</th>
<th>Mean value of MUP amplitudes (μV)</th>
<th>Summation factor</th>
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<tr>
<td></td>
<td>From individual units* Mean I†</td>
<td>From increments to muscle compound action potential Mean P†</td>
</tr>
<tr>
<td>Controls</td>
<td>21</td>
<td>61.2 ± 13.6</td>
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<tr>
<td></td>
<td>Mean 1</td>
<td>42.1 ± 12.2</td>
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<td>11</td>
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<td>69.8 ± 27.2</td>
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<td>45.0 ± 14.1</td>
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<td>12</td>
<td>36.6 ± 10.8</td>
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* Individual MUPs obtained by computer subtraction of stored templates (see Method). I is the mean motor unit amplitude in each patient.
† P is the mean potential increment in each patient calculated from the formula P = (A^n/n) where n is the number of motor unit potentials summated to give a muscle compound action potential of amplitude A^n.
nerve endings in myasthenia gravis. Brain, 83,
10–23.

Buchthal, F., Guld, C., and Rosenfalck, P. (1955). Innerva-
tion zone and propagation velocity in human muscle. Acta

Buchthal, F., Guld, C., and Rosenfalck, P. (1957). Multi-
electrode study of the territory of a motor unit. Acta
Physiologica Scandinavica, 39, 83–104.

aspects of myopathy with particular reference to pro-
gressive muscular dystrophy. In Muscular Dystrophy in
Man and Animals, pp. 193–262. Edited by G. H. Bourne

unit territory and fiber density in myopathies. Neurology
(Minneapolis), 10, 398–408.

Diseases of Voluntary Muscle, 3rd edn, pp. 20–30. Edited

Coers, C. Joffroy, A., Hildebrand, J., and Malevez, R.
(1966). Les altérations du tissu musculaire et de son
innervation dans la myasthénie. In Progressive Muskel-
dystrophie, Myotonen, Myasthenie, pp. 325–339. Edited by

Desmedt, J. E. (1966). Presynaptic mechanisms in myasthenia
gravis. Annals of the New York Academy of Sciences, 135,
209–246.

Hodes, R., Larrabee, M. G., and German, W. J. (1948). The
human electromyogram in response to nerve stimulation
and the conduction velocity of motor axons. Studies on
normal and on injured peripheral nerves. Archives of
Neurology and Psychiatry (Chic.), 60, 340–365.

skeletal muscle: multiple innervation of individual muscle
fibres and motor unit function. Journal of Physiology,
126, 293–303.

Duchenne dystrophy. II. Morphometric study of motor
end-plate fine structure. Brain, 97, 123–130.

Kaeser, H. E. (1970). Nerve conduction velocity measure-
196. Edited by P. J. Vinken and G. W. Bwyn. North-
Holland: Amsterdam.

Kito, S., Yamamoto, M., Fujimori, N., Itoga, E., and
Ultrastructural lesions of the muscle and the nerve in
myotonic dystrophy. 2. Insulin and HGH responses in
myotonic dystrophy. In Basic Research in Myology, part 1,
Congress Series No. 294. Excerpta Medica: Amsterdam.

Kugelberg, E. (1947). Electromyograms in muscular dis-
orders. Journal of Neurology, Neurosurgery, and Psychiatry,
10, 122–136.

Ludin, H. P. (1973). Action potentials of normal and
dystrophic human muscle fibres. In New Developments in
Electromyography and Clinical Neurophysiology, vol. 1,

Electrophysiological study of dystrophy myotonica.
Journal of Neurology, Neurosurgery, and Psychiatry, 34,
132–139.

McComas, A. J., Fawcett, P. R. W., Campbell, M. J., and
Sica, R. E. P. (1971b). Electrophysiological estimation of
the number of motor units within a human muscle. Journal

electrophysiological study of Duchenne dystrophy.
MacDermot, V. (1961). The histology of the neuromuscular junction in dystrophia myotonica, Brain, 84, 75–84.
Computer method for the analysis of evoked motor unit potentials. 2. Duchenne, limb-girdle, facioscapulohumeral and myotonic muscular dystrophies.

J P Ballantyne and S Hansen

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doi: 10.1136/jnnp.38.5.417