Short report

Measurement of motor conduction velocity with Hopf's technique in myotonic dystrophy

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SUMMARY Hopf's technique was used to measure maximal and minimal motor nerve conduction velocities, and the percentage of fibres with intermediate velocity, in the posterior tibial nerve in patients with myotonic dystrophy. A reduction of maximal and minimal conduction velocities was found. The distribution of fibres with intermediate velocity was nearly identical to that of the control group and the dispersion values were normal. These data do not support the hypothesis that a primary disturbance of the motor neurons is responsible for the muscle changes in myotonic dystrophy. The reduction of the motor nerve conduction velocity, which was an inconstant finding, should not be considered an indication of a neurogenic aetiology of myotonic dystrophy, but only one of the many disorders of a multisystem disease.

A neurogenic basis for the muscular disturbances in myotonic dystrophy has been suggested by neurophysiological evidence of motor unit loss in the atrophic distal muscles[1 2] and histopathological findings of selective atrophy of type 1 muscle fibres.[3 4] Other findings which seem to support this possibility include a reduction of motor conduction velocity (MCV)[5–9] and absence of H reflex.[10] Walton et al[11] however failed to find any significant abnormality of the lower motorneuron in subjects with myotonic dystrophy. In order to elucidate further the degree and significance of neural involvement in this disorder, the MCV has been determined using the Hopf technique.[12 13]

Patients and methods

We examined nine patients with myotonic dystrophy (eight males and one female) aged from 21 to 56 years. The data of this group have been compared with those obtained in a control group of seven normal volunteers (aged from 25 to 64 years). The following tests were performed: measurement of the MCV in the posterior tibial nerve by the standard technique and also by the Hopf technique. For the latter the nerve was stimulated at the popliteal fossa and behind the medial malleolus with paired rectangular supramaximal 0·2 ms stimuli, recording percutaneously from the abductor hallucis muscle. The intershock interval was varied by 0·1 ms at each step, starting from the minimum

delay at which the smallest potential following the proximal stimulation was elicited. At each time interval the increment in amplitude of the potential was recorded until the delay between the two stimuli was such that there was no further increase. The range between maximal and minimal MCV, terminal latencies and amplitude of motor responses also were evaluated. Analysis of the change in amplitude of the muscle potential for each time interval between the proximal and distal stimulus was made, a particular velocity value corresponding to each time interval. Histograms of conduction velocity against variation of amplitude of the potential were constructed for each group (myotonic dystrophy and controls), taking as reference point the median value of MCV for each subject. The axonal refractory period influences the measure of the motor conduction velocity when the Hopf technique is used.[14] However the eventual correction of the values of velocities would leave substantially unchanged the distribution of conduction values inside the histograms. After calculating the average change in amplitude of the median value of MCV in the group, the same measurement was then performed in steps each of 1 m/s to the left and right of the median value. In order to assess the difference between the myotonic dystrophy group and the control group, analysis of covariance was used. The analysis was carried out for each variable considered in the study, using the age as covariate.

Results

In the cases of myotonic dystrophy there was a statistically significant reduction of maximal and

minimal MCV values evaluated by Hopf's technique and also of the maximal MCV measured by the routine method. The other variables (range, terminal latency and amplitude of the responses) were not found to be significantly different between patients and controls subjects (Table).

Analysis of individual cases with respect to normal values ±2 SD shows that in cases 3 and 9 both the maximal and minimal MCV values (Hopf) were reduced; in cases 2 and 7 all these values were borderline and in case 8 only the minimal conduction was borderline, while the findings in the other cases were normal. Range between maximal and minimal MCV was at the lower limit in case 7 and normal in the other cases. The terminal latency values were slightly increased in cases 7 and 8, but normal in the remaining cases. The amplitude of the motor responses was reduced in case 8 and within normal limits in the other cases. The histograms of percentage of fibres with intermediate conduction show a Gaussian distribution both in the myotonic dystrophy and control groups (fig).
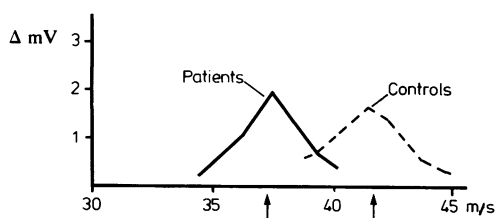

Fig *Histograms of myotonic dystrophy and control subjects. Abscissa: values of conduction velocity (m/s), MCV value indicated by the arrow represents the average median value of all subjects in that group. Ordinate: average amplitude variations (Δ mV) of the potentials by proximal stimulation, obtained at different time intervals between the proximal and distal stimulus.*

Discussion

In order to test the hypothesis of a neurogenic cause for the changes in the muscles in myotonic dystrophy, we measured the motor fibre conduction velocity using Hopf's technique. Although an abnormality detected with this technique may be due either to a slowing of conduction velocity or to a prolonged refractory period,[14] it has been demonstrated to be the most reliable and practical method for evaluating the MCV distribution of motor axons inside mixed peripheral nerves.[15] Other techniques used in myotonic dystrophy estimate the distribution of both sensory and motor fibres in the peripheral nerves and therefore are not suitable for the present investigation.[16] Hopf's technique allows one to evaluate not only the maximal and minimal MCV values, but also the percentage of fibres with intermediate conduction, thus making it possible to reveal selective involvement of a portion of the α motor neurons, as well as MCV reduction. The former characteristic seemed important in view of histopathological data which showed a selective atrophy of type 1 muscle fibres in myotonic dystrophy; these data could suggest involvement of tonic motor neurons which have relatively small diameter axons.[17] Some authors [18-21] by applying this technique in the study of cases with lower motor neuron pathology have found a sharp reduction of range between maximal and minimal MCV, attributed to degeneration of those nerve fibres originating from atrophic motor neurons, without any apparent reduction of conduction values. Moreover Caccia *et al*[13] starting from previously mentioned histological data and using the same technique, but limiting themselves to the measure of the MCV, found a reduction of maximal velocity in myotonic dystrophy cases. This finding suggested involvement of fast conducting axons, rather than the slower conducting

Table

Name	Age (Yr)	Max vel Hopf	Min vel Hopf	Range	Max vel classic method	Termin latency	Amplitude
1 BA	21	43·9	38·9	5·7	44·4	4·0	12·0
2 BN	30	39·0	32·8	6·2	41·8	5·2	18·0
3 BR	36	36·4	30·2	6·2	37·0	5·0	9·5
4 GA	37	45·4	39·5	5·9	49·3	4·0	7·7
5 MF	38	45·3	37·8	7·5	45·8	4·5	14·5
6 NG	38	42·8	37·1	5·7	45·0	4·0	20·0
7 SL	50	39·4	35·3	4·1	41·0	6·0	4·5
8 DP	54	41·7	33·3	8·4	47·0	6·0	1·2
9 BL	56	33·4	27·6	5·8	39·5	5·0	5·4
Mean	40	40·8	34·7	6·1	43·4	4·8	10·3
±1 SD	11·4	4·1	4·0	1·2	3·8	0·79	6·3
Controls							
Mean	42·8	45·1	38·1	6·9	47·5	4·1	9·9
±1 SD	13	2·2	1·9	1·2	3·7	0·8	3·2
		p = 0·014	p = 0·023	NS	p = 0·047	NS	NS

The SPSS (Statistical Package for the Social Sciences) Anova 601 program for the analysis of covariances was used.

axons supplying type 1 motor units, implied by the histochemical evidence of type 1 muscle fibre atrophy.

In our series of cases, a slight uniform reduction of MCV values has been found in some subjects, with involvement of both the faster and slower conducting fibres and with normal percentage of fibres with intermediate velocity, thus the values of the dispersion are normal. These data do not support the hypothesis of a selective involvement of the smaller tonic type 1 motor neurons in myotonic dystrophy. The specific atrophy of these motor neurons in fact should have caused the disappearance of the respective motor fibres and consequently a reduction of the dispersion values, as in motor neuron diseases.

Nevertheless we still have to evaluate the significance of the MCV reduction at times reported in myotonic dystrophy and also found in some of our patients. It is not a constant findings. Some authors have not observed it in their series of cases[22] and others have not found any correlation with the degree of muscle atrophy.[9] A certain connection with age has been reported by Roohi et al,[23] but we have been unable to substantiate this finding. Because of this inconsistency, these data cannot be considered to support a primary neurogenic cause for muscular atrophy in myotonic dystrophy.

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