Conduction velocity in human muscle
van der Hoeven, Johannes H.

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MUSCLE FIBER CONDUCTION VELOCITY IN THE DIAGNOSIS OF FAMILIAL HYPOKALEMIC PERIODIC PARALYSIS

Invasive vs surface determination


(Muscle Nerve 1994;17:898-905)

ABSTRACT

Muscle fiber conduction velocity (MFCV) in the brachial biceps muscle was determined in a large family of patients with hypokalemic periodic paralysis (HOPP) by both a surface and an invasive method. Other surface EMG parameters and the muscle force were also determined. Both the surface and the invasive method showed a significantly lower mean MFCV in the proven gene carriers but only the invasive method showed a lower MFCV in all proven carriers. It can be concluded that MFCV determination is a reliable method to detect the membrane defect in HOPP carriers and that the invasive method is not only easy to perform, but also more sensitive. The muscle force and the integrated EMG at maximal voluntary contraction were lower in the carrier group. A positive correlation between the surface MFCV and the neuromuscular efficiency (the quotient of force and integrated EMG) was found in the controls but not in the HOPP carriers. Since type II fibers have a higher neuromuscular efficiency, this suggests a preferential involvement of type II fibers in HOPP.
INTRODUCTION

The diagnosis of hypokalemic periodic paralysis (HOPP) is based on the occurrence of hypokalemia in combination with attacks of muscle weakness or paralysis. In a number of patients it is possible to induce attacks. Other indicators are a vacuolar myopathy in muscle biopsies from patients with a positive family history (De Fine Olivarius and Christensen, 1965; Links et al. 1990). Concentric needle EMG findings are slightly myopathic or not specific (Engel et al. 1965; Buruma and Bots, 1978). Despite these characteristics, the diagnosis remains difficult. Recently however it was shown that the muscle fiber conduction velocity (MFCV), measured with surface EMG, is reduced in HOPP. Asymptomatic relatives who have inherited the disease can probably be detected by this method (Zwarts et al. 1988). Unfortunately, the surface EMG method is unavailable in most clinical neurophysiology laboratories. Furthermore, the measurements are not always conclusive, sometimes because of poor signal quality and sometimes because of low normal MFCV results in (otherwise) proven carriers. Troni et al. (1983) found a reduced MFCV in HOPP interictally with needle electrodes. To compare the diagnostic values of these two methods, we performed surface MFCV measurements in a large family of HOPP patients, asymptomatic (first-degree) relatives, and controls. We also measured MFCV in a part of this group by means of needle electrodes, a relatively simple technique. Theoretical models (Stegeman and Linssen, 1992) indicate a direct relation between the MFCV and surface EMG parameters, i.e. the median frequency and the integrated EMG value. Since the reduced MFCV in HOPP patients could provide a model in vivo for studying this relation, the median frequency and the integrated EMG were measured simultaneously.

MATERIALS AND METHODS

We performed surface EMG on 33 proven carriers (mean age 42.8, range 16 - 75 years) and 56 asymptomatic first degree relatives (mean age 40.4, range 16 - 76 years). All were members of the same family known to suffer from HOPP, as described previously (Links et al. 1990). 22 carriers were determined on the basis of having typical attacks. 11 patients had never suffered from paralytic attacks but were shown to be carriers either because a muscle biopsy revealed a vacuolar myopathy, or because they had children with attacks (fig. 1). 12 proven carriers and 7 asymptomatic relatives out of the first group were also examined using an invasive MFCV method. The measurements were performed without acetazolamide medication. Normal values (both surface and invasive EMG) were derived from a group of 46 healthy individuals without complaints about their neuromuscular system (mean age 45.8, range 19 - 74 years). Subjects with other known causes for MFCV changes: such as neurogenic lesions or myogenic lesions due to other causes than HOPP (Gruener et al. 1979; Doriguzzi et al. 1987; Zwarts and van Weerden, 1989; Zwarts and van der Hoeven, 1990) or use of medication: such as steroids or anti-epileptics which can probably influence MFCV (Gruener and Stern, 1972; Hopf, 1973; Troni et al. 1990) were excluded. All subjects gave their informed consent prior to the measuring.
**Muscle Fiber Conduction Velocity in HOPP**

<table>
<thead>
<tr>
<th>TABLE I</th>
<th>Results of surface EMG measurements and standard deviation (SD) in controls, proven HOPP carriers, and asymptomatic relatives.</th>
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<table>
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<tr>
<th></th>
<th>Controls</th>
<th>HOPP</th>
<th>Relatives</th>
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<tbody>
<tr>
<td></td>
<td>m/f</td>
<td>mean</td>
<td>SD</td>
</tr>
<tr>
<td>S-MFCV (m.s⁻¹)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>m</td>
<td>4.25⁺</td>
<td>0.37</td>
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<tr>
<td>f</td>
<td>4.10⁺</td>
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<tr>
<td>all</td>
<td>4.20⁺</td>
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<td></td>
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<tr>
<td>Fmed (Hz)</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>m</td>
<td>88.1⁺</td>
<td>17.6</td>
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<tr>
<td>f</td>
<td>78.0⁺</td>
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<tr>
<td>all</td>
<td>84.9⁺</td>
<td>19.4</td>
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<tr>
<td>Max force (N)</td>
<td></td>
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<td></td>
</tr>
<tr>
<td>m</td>
<td>183⁺</td>
<td>51</td>
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<tr>
<td>f</td>
<td>86⁺</td>
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<tr>
<td>all</td>
<td>150⁺</td>
<td>62</td>
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<tr>
<td>IEMG at MVC (µV)</td>
<td></td>
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</tr>
<tr>
<td>m</td>
<td>579⁺</td>
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<td>f</td>
<td>379⁺</td>
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<tr>
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<tr>
<td>f</td>
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<td>19</td>
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</table>

Abbreviations: HOPP: proven HOPP carriers, relatives: asymptomatic relatives, m/f: gender, S-MFCV: mean muscle fiber conduction velocity, surface method, Fmed: mean median frequency, max force: force at maximum voluntary contraction (MVC), IEMG at MVC: integrated EMG value at MVC, number: number of subjects. Statistical analysis Students t-test, unpaired samples, 2-tailed. * significant difference between controls and proven carriers, NS not significant.

**Surface EMG recording**

The experiments were performed on the left biceps brachii muscle. The arm was fixed in a horizontal semiflexed position at an angle of 120 degrees and was supported at the elbow and the wrist. The isometric force of the elbow flexion was measured at the wrist with a strain gauge and then displayed in front of the subject on a voltmeter. Three silver electrodes (diameter 2 mm) were placed in a rigid bipolar array with a common centre electrode; interelectrode distance was 10 mm. The localization of the electrodes was parallel to the fiber direction, nearly half way between the innervation zone and the distal tendon. The two EMG signals were amplified differentially (Disa EMG amplifier type 14C13) and bandpass filtered (20 - 500 Hz). The EMG signals were digitized synchronously by a 12-bit A/D converter with two different sample rates: 6024 Hz (velocity estimation) and 1024 Hz (power spectra) over two connected signal periods of 0.34 and 2.05 seconds respectively. The high sampling rate and consequent interpolation of the signal used for MFCV estimation was necessary to obtain a sufficiently high time resolution. Data were analyzed by a microcomputer (PDP 11/23) off line. The surface MFCV (S-MFCV) was calculated with the
cross correlation method (Naeije and Zorn, 1983) after linear interpolation which raised the sample frequency to 12048 Hz. Only correlation coefficients higher than 0.85 were accepted. The power spectrum was computed over the frequency range of 5-250 Hz by applying the fast fourier transform to the digitized signal. The frequency resolution was 0.5 Hz. The median frequency and the integrated EMG were calculated. All measurements were performed at different force levels in duplicate at 20-30-50-75 and 100% of maximum voluntary contraction. On the basis of the summarized results, we calculated the mean S-MFCV and the mean median frequency.

**Invasive MFCV (I-MFCV) estimation**

Experiments were performed in the left brachial biceps (short head) with a Nicolet Viking I EMG apparatus using a modified version of the method of Troni et al. (1983) (van der Hoeven et al. 1993). A stimulation needle electrode (Dantec 13L64, area of uninsulated tip: 2 mm²) was placed in the distal part of the resting muscle. A silver surface electrode was used as anode 10 to 15 mm distally. The muscle was stimulated with gradually increasing strength (suprathreshold) until a clear twitch was palpable (1 - 2 mA, 0.2 msec, 1 Hz). Guided by the twitch, we inserted a concentric needle electrode (Dantec 13L58) 50 - 60 mm proximally and manipulated it until a reproducible polyphasic action potential was seen, amplitude 20 - 500 µV. The signals were amplified and bandpass filtered, 500 Hz - 10 kHz; the time base varied between 5 - 10 msec per division. Care was taken to place the electrodes perpendicular to the skin. A 4-trace storage was used to ensure the reproducibility of the action potentials. Only spikes larger than 20 µV were used for calculations. All latencies were measured at the positive turning points and resulting I-MFCV was calculated. The parameters used were the mean I-MFCV, fastest and slowest I-MFCV, and ratio fastest / slowest I-MFCV (F/S ratio), which indicates the scatter in conduction velocities.

**Figure 1. Diagram showing the carrier status of all family members tested with the surface EMG method.**
Surface EMG
The surface EMG results are summarized in table I. Since the mean S-MFCV and the mean median frequency of the male and female controls were not significantly different (p>0.05), these data were pooled. In 6 asymptomatic relatives it was not possible to get a reliable S-MFCV estimation, due to correlation coefficients lower than 0.85. In one proven carrier (on the basis of previous attacks) no surface or invasive measurement at all could be done because of a nearly total paralysis of the elbow flexors. The mean S-MFCV in the HOPP carriers was significantly lower than in the controls. The same was found for the median frequency but with a greater overlap between the two groups (fig 2). Four asymptomatic relatives showed S-MFCV values in the same range as the proven carriers. On the other hand, 7 out of the 22 carriers with attacks and 3 out of the 11 carriers

Figure 2. Relation between the muscle fiber conduction velocity, surface method (S-MFCV) and the median frequency (Fmed) of the proven carriers, asymptomatic relatives, and healthy controls. Abbreviations: HOPP/a: HOPP-attacks, HOPP/m: HOPP-myopathy, HOPP/c: HOPP-children with attacks, relatives: asymptomatic relatives. Dotted lines: 5th percentile of the control values of the mean Fmed (horizontal line) and the mean S-MFCV (vertical line). The control data are omitted to simplify the figure.

Statistics
Surface EMG results were analyzed by means of Students t-test, unpaired samples, 2-tailed. The Mann-Whitney non-parametric test, unpaired samples, 2-tailed, was used to test the difference of means in the invasive EMG groups. Statistical significance was accepted at a level of 5%. 

RESULTS

Surface EMG
The surface EMG results are summarized in table I. Since the mean S-MFCV and the mean median frequency of the male and female controls were not significantly different (p>0.05), these data were pooled. In 6 asymptomatic relatives it was not possible to get a reliable S-MFCV estimation, due to correlation coefficients lower than 0.85. In one proven carrier (on the basis of previous attacks) no surface or invasive measurement at all could be done because of a nearly total paralysis of the elbow flexors. The mean S-MFCV in the HOPP carriers was significantly lower than in the controls. The same was found for the median frequency but with a greater overlap between the two groups (fig 2). Four asymptomatic relatives showed S-MFCV values in the same range as the proven carriers. On the other hand, 7 out of the 22 carriers with attacks and 3 out of the 11 carriers
without attacks showed S-MFCV values in the low-normal range (above the 5th percentile value), which is a sensitivity of 70%. Table I shows the mean maximal force at elbow flexion for the different groups. The force in the male patient group was lower than in the controls, whereas in the females the differences were not significant. The integrated EMG data showed clearly lower values in the patient group, both for males and females. The force and integrated EMG values in the asymptomatic carriers and the controls were not significantly different. A positive correlation between max integrated EMG and max force was found in the control group as well as in the HOPP patients. When the S-MFCV is plotted against the neuromuscular efficiency, defined as the quotient of force and integrated EMG, a positive correlation is found in the controls and in the asymptomatic relatives. There was no such correlation in the HOPP patient group (fig. 3).

### Table 2

<table>
<thead>
<tr>
<th></th>
<th>Controls</th>
<th>HOPP</th>
<th>Relatives</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>mean (m.s⁻¹)</strong></td>
<td>3.17</td>
<td>2.35</td>
<td>3.15</td>
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<tr>
<td><strong>mean</strong></td>
<td>0.22</td>
<td>0.32</td>
<td>0.51</td>
</tr>
<tr>
<td><strong>slow (m.s⁻¹)</strong></td>
<td>2.75</td>
<td>1.59</td>
<td>2.69</td>
</tr>
<tr>
<td><strong>fast (m.s⁻¹)</strong></td>
<td>3.68</td>
<td>3.32</td>
<td>3.73</td>
</tr>
<tr>
<td><strong>mean</strong></td>
<td>0.23</td>
<td>0.33</td>
<td>0.63</td>
</tr>
<tr>
<td><strong>mean</strong></td>
<td>0.31</td>
<td>0.33</td>
<td>0.41</td>
</tr>
<tr>
<td><strong>F/S ratio</strong></td>
<td>1.35</td>
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<td>1.47</td>
</tr>
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<td>0.13</td>
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<td>0.38</td>
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<tr>
<td><strong>mean</strong></td>
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<td>0.47</td>
<td>0.38</td>
</tr>
<tr>
<td><strong>number</strong></td>
<td>46</td>
<td>12</td>
<td>9</td>
</tr>
</tbody>
</table>

Abbreviations: mean: mean I-MFCV, slow: mean of slowest measured fibers, fast: mean of fastest fibers measured, F/S ratio: mean ratio between fastest and slowest fibers measured, spikes pro ins: number of spikes pro insertion, n: number of measurements. Statistical analysis Mann-Whitney non-parametric test, unpaired samples, 2-tailed, *significant difference between controls and proven carriers.

Invasive MFCV

In table II a summary of the I-MFCV values is given. The mean I-MFCV in the HOPP carriers was significantly lower than in the controls and in the asymptomatic relatives: in the controls a mean I-MFCV value (data pooled for both sexes) of 3.17 m.s⁻¹ (SD 0.22), in the group of proven carriers a mean I-MFCV value of 2.36 m.s⁻¹ (SD 0.31), and in the asymptomatic relatives a mean I-MFCV value of 3.15 m.s⁻¹ (SD 0.51). A plot of I-MFCV vs S-MFCV (fig. 4) shows the division between the controls and proven carriers. This is mainly due to the much lower I-MFCV results. Both the slowest and fastest fibers showed lower values in the group of proven carriers (fig. 5 and 6). The relative decrease of the slowest fibers was more pronounced in the HOPP carriers giving them a higher F/S ratio. All proven carriers differed from the controls when the I-MFCV was plotted against the F/S ratio (fig. 7), which is a sensitivity of 100%. Two asymptomatic relatives showed similar disturbances with respect to the I-MFCV and S-MFCV, the slowest and fastest measured fibers, and the F/S ratio; this suggests that they belong to the group of carriers.
DISCUSSION

The surface EMG findings agree with those of our previous study on a smaller group of patients from this family (Zwarts et al. 1988). The S-MFCV levels were clearly lower in most proven carriers and there were no significant differences in the mean S-MFCV values between the asymptomatic relatives and the controls. Detecting carriers of the membrane defect with the surface MFCV method was not in all cases conclusive (sensitivity of 70%). It was, however, possible to show the MFCV disturbances in all proven carriers by the invasive MFCV determination technique (figs. 4, 5 and 7) but the specificity of neither methods is known. The low S-MFCV and median

Figure 3. Relation between the muscle fiber conduction velocity, surface method (S-MFCV) and the neuromuscular efficiency (NME): Force / integrated EMG (IEMG), both at maximal voluntary contraction in proven HOPP carriers and in healthy controls. Dotted line (HOPP): not significantly different from zero, Dashed line (controls): significantly different from zero, $r=0.53$, $p<0.0001$. Linear regression analysis.
frequency values found in four subjects (fig. 2) suggest that some gene carriers are included in the group of asymptomatic relatives. Two asymptomatic relatives with S-MFCV values in the 'carrier' range were also examined by means of the invasive method and showed low I-MFCV values as well (fig. 4). Additionally, some of the subjects who in our previous study belonged to the group of asymptomatic carriers are now classified as proven carriers because of the results of muscle biopsy performed later or because they now have children with attacks. The finding of a low MFCV in combination with a positive family history seems therefore highly suggestive for a HOPP gene carrier.

The discrepancy between the findings of these two methods can be explained by the drawbacks of the surface method. (1) Probably the most important problem is a less than optimal electrode location resulting in an overestimation of the S-MFCV (Sollie et al. 1985; Arendt-Nielsen and Zwarts, 1989). (2) Since the source of the electrical activity, the muscle fiber, has to be activated volitionally, variations in the central activation pattern are unknown. Motor units are recruited according to their size (Henneman, 1957) and the MUs with the highest force generating capacity (generally type 2 fibers) show mainly the highest MFCVs (Andreassen and Arendt-Nielsen, 1987). Additionally, rate-coding influences the MFCV. This has been related to the short period of hyperpolarization following repolarization (Nishizono et al. 1989; Mihelin et al. 1991). (3) The distance between the active fibers and the electrodes is variable. An increase in
radial observation distance results in a strong decrease in the signal/noise ratio (Stegeman and Linssen, 1992). Since noise has no phase shift, overestimation of MFCV will result in the extreme cases. (4) Electrical inhomogeneous tissue between the muscle fibers and the recording electrodes sometimes results in signals without clear time delay resulting in an overestimation of the MFCV (Broman et al. 1985; Schneider and Rau, 1991). (5) A finite fiber length results in the appearance of a positive, constant latency peak which will result in decrease or absence of latency. However, in bipolar recordings these non-moving potentials are largely cancelled (Gootzen et al. 1991).

The invasive method, on the other hand, is a much simpler and more rapid procedure requiring only minimal patient cooperation. Its advantage is the non-volitional, direct activation of the resting muscle, irrespective of the state of innervation (Troni et al. 1983). We were able to measure interictally nearly all subjects with the invasive method, except one carrier whose biceps muscle was almost totally paralyzed due to an advanced myopathy. The method can be performed with every modern EMG apparatus. The positioning of the electrodes is facilitated by the use of a concentric needle uptake electrode instead of the single fiber electrode proposed by Troni et al. (1983). Buchthal et al. and Meadows also pointed out the possibility of measuring I-MFCV with a concentric needle electrode, although in a different experimental set-up (Buchthal et al. 1955; Meadows, 1971). Use of the concentric needle electrode might cause some loss of selectivity, but most of the recording selectivity is due to the high-pass filtering (Payan, 1978). Despite the technical differences, our MFCV results in controls are well within the range found by those who
used different techniques (Arendt-Nielsen and Zwarts, 1989). A second possible drawback of the invasive method is that the occurrence of slower conducting fibers can vary locally within the muscle. Finding these "late" potentials is made much easier because of the greater uptake area of the concentric needle combined with a temporary lowering of the high-pass filter setting and slight needle repositions. In HOPP, however, almost all fibers have the same membrane disturbance, which makes a non-homogenous muscle involvement less likely.

The muscle force in the HOPP carriers is generally lower, especially in males. 80% of the HOPP patients in this family complained about fluctuations of muscle strength (Links, 1992) which suggests that the muscle fibers may be more or less permanently blocked. This phenomenon would also explain the increase in force after therapy with carbonic anhydrase inhibitors (Dalakas and Engel, 1983; Links et al. 1988). In such cases small variations in the membrane potential probably cause variation in the number of blocked fibers, resulting in changes in muscle force or a (partial) paralytic attack (De Grandis et al. 1978; Troni et al. 1983; Rudel et al. 1984). This would probably also explain the fact that the lowered I-MFCV values in the slowest fibers are clearly more pronounced than those of the fastest fibers (fig. 5).

The significantly lower integrated EMG values at maximal force in the patient group (table I) can be explained in various ways. (1) The partial depolarization of the muscle membrane results in action potentials of lower amplitude (Trontelj et al. 1987) causing lower integrated EMG levels. (2) The frequency change of the signal could also alter this relation. Since the surface electrodes act as a high pass filter, which depends on the electrode separation (Zipp, 1978), the change of the

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**Figure 6.** Examples of muscle fiber conduction velocity determination, invasive method, biceps brachii muscle. Two traces superimposed. A. Healthy control. B. Proven HOPP carrier.
signal to lower frequencies will result in the transfer of less energy. (3) Model studies indicate that low values of the integrated EMG could be a direct consequence of a reduced MFCV (Stegeman and Linssen, 1992). (4) Additionally, we hypothesize an effect of phase cancellation: the general decrease in conduction velocity will result in an increase in latency differences, based on the reciprocal relationship of conduction velocity and latency. A direct consequence of this relationship is an increase in the temporal dispersion of the depolarization zone within the motor unit. This effect will cause a phase cancellation within the summated potential of an activated motor unit, as recorded at some distance by surface electrodes.

A puzzling phenomenon in a generalized muscle disease like HOPP is the relatively unspecific findings at concentric needle EMG interictally. Myopathic changes are reported in only some of the cases, while often the absence of clear abnormalities is emphasized (De Fine Olivarius and Christensen, 1965; Engel et al. 1965; Buruma and Bots, 1978). We suggest the following explanations. (1) The low-pass filter properties of tissue are strongly dependent on the distance to the source. Close to the source the reduced MFCV does not influence the frequency content as much as in the case of surface electrodes (Lindstrom and Petersen, 1983; Lateva, 1988). (2) In HOPP a preferential atrophy of type II fibers is found (Brooke and Engel, 1969). So, if all fibers within a motor unit have a conduction slowing more or less to the same degree, a myopathic pattern based on the differences in MFCV will only be found at a larger distance from the end-plate zone. In that case small differences in conduction velocity will result in relatively larger differences
in latency. (3) A loss of excitable muscle fibers will have no influence on the MUAP waveform (Nandedkar and Sanders, 1989).

When the neuromuscular efficiency and the mean S-MFCV, both at maximum force, were plotted against each other (fig. 3), a positive correlation was found in the controls. We suggest that this is a reflection of differences in neuromuscular efficiency between type I and type II motor units. The type II motor units have the highest values with respect to muscle fiber diameters and, consequently, MFCV. This was also reflected in the positive correlation between force and MFCV (Andreassen and Arendt-Nielsen, 1987; Zwarts et al. 1988). Additionally, the maximal force of type II fibers may be about 4 times that of type I fibers (Linssen et al. 1991). On the other hand, there is no indication that the (extracellular) single fiber action potential, which forms the basis of the integrated EMG, differs to such an extent in the two fiber types (Wallinga-de Jonge et al. 1985). This implies that the highest neuromuscular efficiency values should be found in type II fibers. However, the positive correlation between the S-MFCV and the neuromuscular efficiency was lacking in the HOPP patient group as was the positive correlation between force and MFCV (Zwarts et al. 1988). This could be an effect of the preferential dysfunction (Zwarts et al. 1988) or the atrophy (Brooke and Engel, 1969) of type II fibers in HOPP, probably in combination with an altered relationship between the membrane properties and the muscle force. A comparable dissociation between force and integrated EMG was found during the phase of "transient paresis" in myotonica congenita (Zwarts and van Weerden, 1989).

In conclusion, we found clear changes in the MFCV in a large family of HOPP patients with the surface as well as with the invasive determination method. Since, however, only the invasive determination method showed MFCV disturbance in all proven carriers, we suggest that it is more sensitive than the surface method in detecting carriers of the membrane defect. Furthermore, the method can be easily performed without much discomfort for the patients. The reduced MFCV causes relatively low integrated EMG values. A significant positive correlation between the S-MFCV and the neuromuscular efficiency (the quotient of force and integrated EMG) was found in the control group but not in the HOPP patients. This suggests a preferential involvement of type II muscle fibers in combination with an altered relation between the membrane properties and the muscle force.

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Muscle Fiber Conduction Velocity in HOPP

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Chapter 6

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