SHORT REPORT

Surface EMG and muscle fibre conduction during attacks of hypokalaemic periodic paralysis

T P Links, J H van der Hoeven, M J Zwarts

Abstract

Surface EMG, muscle fibre conduction velocity (MFCV), muscle force, and biochemical variables were investigated in a 13-year-old boy with familial hypokalaemic periodic paralysis during and after three attacks of paralysis. After normalisation of the serum potassium values, strength rapidly returned to interictal values, but the integrated EMG and to a lesser extent the MFCV recovered more slowly. These findings suggest that a complete electrophysiological recovery is not necessary for a restoration of muscle force and that the pathogenetic defect is localised in the muscle membrane.

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Hypokalaemic periodic paralysis is characterised by episodes of paralysis combined with hypokalaemia. During attacks muscle excitability after nerve stimulation is severely reduced or absent and concentric needle EMG shows complete electrical silence.¹ Troni *et al*² found a reduced muscle fibre conduction velocity (MFCV) by an invasive method. Surface EMG recording also showed a reduced MFCV in these patients.³ The cause of paralysis is supposed to be a depolarisation block of the muscle fibre membrane. During an attack, a partial membrane depolarisation has been found.⁴ The time course of recovery is not known. Troni *et al*² only mentioned "a long period of recovery". We report surface EMG and MFCV measurements and muscle force in a patient with hypokalaemic periodic paralysis, during and after three episodes of paralysis.

Methods

The methods for the surface and invasive EMG measurements have been previously described.⁵ Muscle strength was measured by a hand-held dynamometer according to a standardised procedure.

Patient history

A 13-year-old boy with a family history of hypokalaemic periodic paralysis⁶ had had

attacks of muscle weakness since he was 11 years old.⁶ During the first described attack he had woken up with complete paralysis at about 0500. No drugs were used. At 1100 he was admitted to our hospital with a total flaccid paralysis of his limbs. Serum potassium was 2.3 mmol.l⁻¹ and he was treated with intravenous potassium. After three hours strength returned to his arms, and slight movements were possible in his legs (table). After six hours strength returned to his legs. After 12 hours he was able to walk without help and potassium infusion was stopped. A total of 75 mmol potassium had been infused. The next morning MFCV measurements were made while his strength was normal.

During the second attack the serum potassium was 2.6 mmol.l⁻¹ on admission. The same therapeutic regime was started and the muscle strength recovered to normal in about four hours. Serum potassium was 4.4 mmol.l⁻¹. Surface EMG measurements were obtained four hours after complete recovery. The serum potassium during the third attack was 2.4 mmol.l⁻¹. Strength returned to normal within three hours and surface EMG was performed five hours later, when serum potassium was 4.9 mmol.l⁻¹.

Results**EMG FINDINGS**

During the early recovery after the first attack, the surface EMG showed very low voltages, which prevented a reliable estimation of MFCV. After complete recovery of the muscle force the next day, the integrated value of the EMG (IEMG) at maximal voluntary contraction, the MFCV, and median frequency had not yet returned to interictal values. After the second and third attacks only the IEMG was low (379 and 231 μ V, respectively) when muscle strength was normal. The interictal measurements were performed after an attack-free interval of three weeks (figure).

Invasive measurements were made during the first attack. In the paralysis phase a severely reduced MFCV was found. During late recovery the MFCV was clearly increased. The changes between the fastest and slowest measured fibres were roughly equal (table).

Department of
Internal Medicine
T P Links

Department of
Neurology, University
Hospital, Groningen,
The Netherlands
J H van der Hoeven
M J Zwarts

Correspondence to:
Dr TP Links, University
Hospital, Department of
Internal Medicine,
PO Box 30.001,
9700 RB, Groningen,
The Netherlands.

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Muscle strength, laboratory measurements and EMG values in the recovery phase during the first attack of hypokalaemic periodic paralysis

	Time during recovery (h)					
	Interictal	11 00	14 00	17 00	22 00	09 00
Force (Newtons):						
Shoulder abduction	220	0	0	160	220	220
Elbow flexion	> 250	0	95	> 250	> 250	> 250
Elbow extension	230	0	70	220	230	230
3 point grip	140	0	40	125	140	130
Hip flexion	240	0	0	220	240	230
Hip abduction	240	0	0	220	240	230
Knee extension	> 200	0	0	> 200	> 200	> 200
Laboratory values:						
K ⁺ (normal 3.6–4.8 mmol.l ⁻¹)		2.3	3.3	4.6	4.6	4.7
Creatine kinase (normal 0–50 u.l ⁻¹)		114	87	82	85	82
Myoglobin (normal 0–80 µg.l ⁻¹)		30	36	62	46	72
Total urinary K ⁺ excretion (mmol)			5	8	10	25
Surface EMG:						
Mean (SD) S-MFCV (m.s ⁻¹)	3.31(0.20)					2.68(0.50)
Mean (SD) Fmed (Hz)	81.3(12.6)					55.6(8.0)
IEMG (µV)	672					58
Invasive MFCV:						
Mean I-MFCV (m.s ⁻¹)		2.07				2.53
Fastest I-MFCV (m.s ⁻¹)		3.06				3.31
Slowest I-MFCV (m.s ⁻¹)		0.93				1.30

Measurements were performed interictally and successively during the first attack. Force was measured with a hand-held dynamometer, (mean values of both sides). Laboratory values were obtained during recovery the next day. S-MFCV = Mean muscle fibre conduction velocity, surface method. Fmed = mean median frequency; IEMG = integrated value of EMG at maximal voluntary contraction; mean I-MFCV = mean muscle fibre conduction velocity, invasive method.

OTHER MEASUREMENTS

The table shows the changes in muscle force and some laboratory values during the first attack. Serum glutamic pyruvic transaminase, serum glutamic oxaloacetic transaminase, and lactate dehydrogenase remained unchanged during the attacks.

Discussion

We measured muscle force, MFCV, and surface EMG during early and late recovery in a patient after a severe attack of hypokalaemic periodic paralysis, and repeated the measurements after two less severe attacks. During the recovery stage of all three attacks the IEMG was low, despite a normalisation of the muscle force. Invasive measurement during the parietic phase showed very low MFCV values, which had partially recovered by the next day. It showed the possibility of depolarisation of at least a part of the muscle fibres, despite the serious paresis. This indicates a selective depolarization or "post-synaptic" block, a mechanism that has already been suggested in a single fibre study, by means of a fibre density decrease during an attack.⁷ Also, the roughly equal decrease of the fastest and slowest MFCV results, suggests that both type I and type II muscle fibres are affected

(and possibly blocked) to the same degree (table).

A puzzling phenomenon is the discrepancy between the IEMG and the muscle force during recovery, which are usually linearly related. Model studies indicate that low values of the IEMG are a direct consequence of a reduced MFCV.⁸ Nevertheless, this does not explain the discrepancy, because after restoration of the MFCV the IEMG values were still low.

Several reasons can be suggested for this temporary discrepancy: (1) A partial depolarisation of the muscle membrane results in action potentials of lower amplitude,⁹ thus disturbing the normal relation between force and IEMG. (2) The frequency change of the signal could also alter this relation. Because the surface electrodes act as a high-pass filter depending on the electrode separation¹⁰ the change of the signal to lower frequencies will result in the transfer of less energy.

In vitro investigations of muscle fibres of patients have shown a slight depolarisation of the muscle cell membrane interictally to -75mV (normal -87 mV), increasing to -50 mV during paralysis.⁴ A partial depolarisation is accompanied by a slowing down in conduction velocity,¹¹ as was found in this study during the severest attack. To account for the discrepancy between the functional and electrophysiological recovery we suggest the following hypothesis. During paralysis potassium moves to the intracellular space. Although muscle strength recovers quickly after intravenous potassium treatment, the ratio between the intracellular and extracellular ion concentrations and the membrane potential is not restored with the same rapidity. Nevertheless, the action potential is sufficient to initiate calcium release, which is necessary for normal force generation. The return of the membrane potential to interictal values occurs only after complete recovery of the external and internal ion concentrations.

Relation between muscle force and IEMG.

This is supported by the time course of potassium excretion, which is also a long lasting process (table).

By contrast with earlier reports,¹² no rise in serum myoglobin concentration and creatine kinase was found in our patient after recovery. Interictally serum myoglobin concentration and creatine kinase activity are raised to varying degrees,^{6,12} which suggests a fluctuating, and during the attacks, temporarily increased membrane leakage.

Force recovered quickly during potassium infusion (table). The initial recovery was in the arms during all three attacks. Recovery of the legs took longer. There are possible explanations for this difference. Vacuolar myopathy is most pronounced in the leg and pelvic girdle muscles⁶ and this suggests a more pronounced membrane disturbance in the legs, possibly because of the local, relatively larger muscle fibre diameters. This could result in a longer recovery time for the membrane potential in terms of transport capacity, between membrane surface and intracellular volume, and maybe enhanced local membrane damage. Another explanation could be that in a lying position the arms are used more than the legs; this could result in a less severe membrane disturbance as slight movements result in a faster recovery,¹³ or even prevent an attack.

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