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Long Latency EMG Responses in Early Diagnosis of Huntington's Chorea

F. Leblhuber[1], E. Windhager[1], F. Reisecker[2], and H. Rittmannsberger[1]

[1]Department of Gerontology, Wagner-Jauregg-Krankenhaus, Wagner-Jauregg-Weg 15, A-4020 Linz, Austria
[2]Department of Neurology, Krankenhaus der Barmherzigen Brüder, Bergstrasse 27, A-8020 Graz-Eggenberg, Austria

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Summary. In healthy subjects, 2 EMG responses of the thenar musles can be distinguished, elicited by electrical stimulation of the median nerve during an isometric contraction: an early spinal response (M_1) and a long latency response (LLR) (M_2); earlier studies have shown that in patients with Huntington's chorea (HC) this late EMG response is missing. LLR were studied in nine subjects at risk out of three families with definite HC. In 6 of them, LLR was clearly asymmetrical or absent on one or both sides, while normal LLRs were seen in the rest of the subjects studied; LLR abnormalities found in clinically free members of HC families may assist in early diagnosis of individuals who may later develop symptoms and may help in genetic counselling.

Key words: Huntington's chorea – Early detection – Long loop reflexes

Introduction

Changes in evoked potentials and EMG parameters are described as electrophysiological markers for preclinical detection of Huntington's chorea (HC) [1, 5, 10, 11]. One of the EMG responses probably altered in the preclinical state are long latency responses (LLRs) of the thenar muscles evoked by electrical or mechanical stimulation of the median nerve [9, 10]. This M_2 response of instrinsic hand muscles is probably a transcortical long loop reflex [1, 9]. We studied the EMG responses of thenar muscles in nine subjects at risk looking for LLR abnormalities similar to those seen in HC patients, as an early sign of this inherited neurodegenerative disorder.

Material and methods

LLRs were recorded by surface electrodes over the belly of the thenar muscles under isometric conditions; the subjects pressed their thumb constantly against the investigator's thumb with about 50% of maximum force. The median nerve was stimulated by surface electrodes at the wrist with the cathode proximal (0,2 ms stimulus duration, constant current) and stimulus intensity just at the threshold of motor fibres. The rectified EMG activity of the thenar muscles was filtered (5 Hz–1 kHz); 64 trials were averaged. Measurements were performed with a Nicolet CA 2000, room temperature remained constant throughout. The series was repeated at least once and compared with measurements in ten HC patients and in ten age- and sex-matched normal controls.

Offprint requests to: Friedrich Leblhuber

Results

In all of the controls studied, the early spinal response M_1 was followed by a late reflex response M_2. No difference was found between left and right side latencies, the M_2 amplitude ratio between left and right side stimulation, mean values (MV) and standard deviations (SD) see Table 1. Of the subjects at risk, the late EMG response M_2 was missing completely in 2 and was found clearly asymmetrical or missing on one side in 4 of the risk subjects investigated (Table 2).

LLRs were also studied in ten patients with definite HC [2], 6 male, 4 female, aged 33–60 years. The M_2 response was lacking in all patients with HC, while the spinal response was always present.

Discussion

Early diagnosis of HC, a chronically progressive hereditary neurodegenerative disorder may be of great impor-

Table 1. Mean values (MV) and standard deviations (SD) of latencies (lat.) of early (M_1) and late (M_2) EMG responses in ms and M_2 amplitude (ampl.) ratio right (r)/left (l) side in 10 normal controls

$N = 10$	M_1 lat. ms		M_2 lat ms		M_2 ampl. r/l
	r	l	r	l	
MV	28,8	27,7	46,9	46,7	1,1
SD	1,7	1,4	4,3	4,4	0,1

Table 2. M_1 and M_2 latencies found in 9 subjects at risk, M_2 in 2 (pat. 1 and 8) absent on both sides, in 2 (pat. 5 and 9) M_2 absent on one side; in the rest the LLR was preserved, but was found clearly asymmetrical in 2 (pat. 4 and 7, M_2 amplitude ratio above 2,5 SD, marked with (). Subjects 1 to 7 belong to the same family, subjects 8 and 9 each belong to a separate family.

	Sex	Age	M_1 lat. (ms)		M_2 lat. (ms)		M_2 ampl. r/l
			r	l	r	l	
1	♀	37a	27,1	28,0	/	/	/
2	♀	35a	25,7	25,7	51,5	51,7	1,3
3	♀	30a	24,5	25,4	46,5	48,6	1,1
4	♀	27a	26,5	26,6	49,9	51,5	(1,5)
5	♂	25a	29,0	29,6	/	56,1	/
6	♂	22a	29,0	28,0	48,8	51,5	1,3
7	♀	17a	25,6	25,5	47,2	47,2	(2,1)
8	♂	14a	24,9	25,1	/	/	/
9	♀	14a	24,9	24,2	/	49,1	/

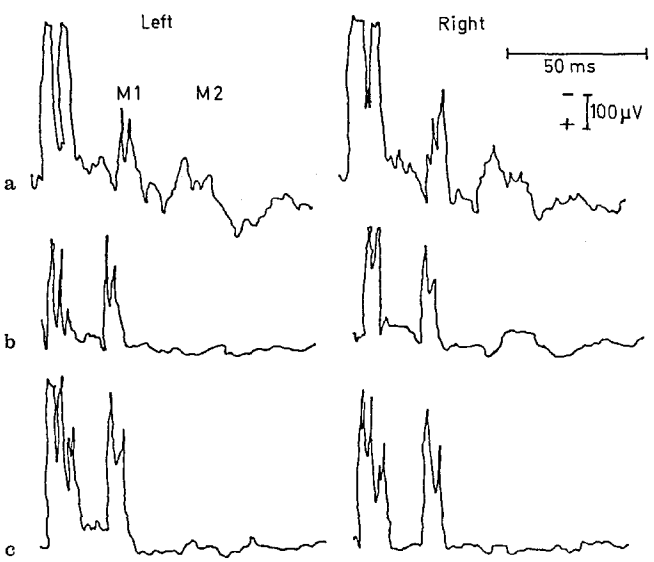

Fig. 1a–c. EMG reflex pattern in a (**a**) normal control with M_1 and M_2 responses on both sides clearly identifiable. (**b**) Subject at risk, lacking M_2 on the left side, low amplitude M_2 at right side median nerve stimulation. (**c**) Symptomatic HC patient with absence of M_2 on both sides

tance for unaffected off-spring [10], because clinical evidence of this autosomal dominant condition usually appears in the third or fourth decade, so that many patients at risk already have children or even may have completed their family. Gusella's [3] discovery of a genetic marker linked to HC offers a predictive test for people at risk, but besides technical limitations an ethical and psychological problems, this marker has so far rarely been used for clinical purposes. Thus, electrophysiological [10, 11] and metabolic [4, 6] markers are still important methods in identifying potential carriers of the HC gene. Earlier studies have shown that two reflex responses can be identified in the thenar muscles after electrical median nerve stimulation under isometric conditions and that patients with clinically manifest HC lack the LLR [1, 9]. LLR was also missing in all our patients suffering from definite HC. Like evoked potential abnormalities found in clinically affected as well as individuals at risk [11], LLR studies offer the possibility of early diagnosis of HC. In 6 out of 9 persons at risk we found LLR abnormalities probably indicating individuals who may later develop clinical symptoms of HC (Fig. 1). Analysis of these LLR studies suggests that like PET [4, 6] and SPECT [7, 8], identification of subjects at risk may be possible, as these abnormalities can be easily detected early and may therefore help in genetic councelling.

References

1. Claus D (1986) Long loop Reflexe – eine klinisch relevante Methode. Fortschr Neurol Psychiatr 54:35–41
2. Folstein SE, Leigh RJ, Parhard IM, Folstein MF (1986) The diagnosis of Huntington's disease. Neurology 36:1279–1283
3. Gusella JF, Wexler NS, Coneally PM, Naylor SL, Anderson MA, Tanzi RE, Watkins PC, Ottina K, Wallace MR, Sakaguchi AY, Young AB, Shoulson I, Bonilla E, Martin JB (1983) A polymorphic DNA marker genetically linked to Huntington's disease. Nature 306:234–238
4. Hayden MR, Martin WRW, Stoessel AJ, Clark C, Hollenberg S, Adam MJ, Ammann W, Harrop R, Rogers J, Ruth T, Sayre C, Pate BD (1986) Positron emission tomography in the early diagnosis of Huntington. Neurology 36:888–894
5. Josiassen RC, Shagass C, Mancall EL, Roemer RA (1982) Somatosensory evoked potentials in Huntington's disease. Electroencephalogr Clin Neurophysiol 54:483–493
6. Kuhl DE, Metter EJ, Riege WH, Markham CH (1984) Patterns of cerebral glucose utilisation in Parkinson's disease and Huntington'disease. Ann Neurol (Suppl) 15:119–125
7. Leblhuber F, Hoell K, Reisecker F, Gebetsberger B, Puehringer W, Trenkler J, Deisenhammer E (1989) Single photon emission computed tomography in Huntington's chorea. Psychiatr Res 29:337–339
8. Leblhuber F, Brucker B, Reisecker F, Trenkler J, Deisenhammer E (1989) Single photon emission computed tomography (SPECT) in probable Huntington's chorea (HC). J Neurol Transm [P-D Sect] 1:93–94
9. Noth J, Podoll K, Friedemann HH (1985) Long loop reflexes in small hand muscles studied in normal subjects and inpatients with Huntington's chorea. Brain 108:65–80
10. Oepen G (1986) Früherkennung der Huntington'schen Erkrankung durch neurophysiologische Untersuchungen, in: Die Huntington'sche Krankheit, H. Oepen (ed), Hippokrates Verlag, Stuttgart, pp 49–53
11. Oepen G, Doerr M, Thodon U (1981) Visual (VEP) and somatosensory (SSEP) evoked potentials in Huntington's chorea. Electroencephalogr Clin Neurophysiol 51:666–670