Short report

Brainstem reflexes and brainstem auditory evoked responses in Huntington's chorea

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SUMMARY Blink reflex, corneal reflex, jaw reflex, exteroceptive suppression in masseter muscles and brainstem auditory evoked potentials were measured in 20 patients with Huntington's chorea and 12 controls. A significantly increased latency of the second component of the homolateral and heterolateral blink reflex was found in the patient group as compared with the controls. The other investigations revealed no significant differences between patients and controls except for some facilitation of the jaw reflex in the patient group. Increase of second component latency of the blink reflex in the presence of normal corneal reflexes is suggestive of functional impulse conduction disturbance in the lower brainstem. It is discussed whether in Huntington's chorea this is to be attributed to alterations of cortical or striatal influence or to local brainstem abnormalities.

The morphological and functional extent of damage of the central nervous system in Huntington's chorea has still to be determined. This particularly pertains to the lower brainstem. Histological investigations have revealed various changes in the olives, the vestibular, vagal and hypoglossal nuclei.¹ Also functional abnormalities attributable to brainstem lesions have been found in patients with Huntington's chorea, for instance in oculomotor studies² and in blink reflex investigations.^{3,4} Nothing unequivocally abnormal has been noted in brainstem auditory evoked responses.⁵ In studies of somatosensory evoked potentials, conflicting results have been reported concerning the central brain conduction time, which is an indicator of lower as well as of higher brainstem derangement.⁵ To the best of our knowledge, electrophysiological measurements of the jaw reflex, the exteroceptive suppression of activity in masseter muscles and the corneal reflex have not been reported in Huntington's chorea. Accordingly, it seemed worthwhile to study the brainstem reflexes as well as to re-examine the blink reflex and the brainstem auditory evoked responses so as to try to determine the extent of brainstem integrity in Huntington's chorea.

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Patients

After informed consent was obtained, according to the Declaration of Helsinki of 1975, twenty patients with Huntington's chorea were studied (10 males, 10 females). Age ranged from 29 to 54 years, (mean: 47.1 years). The duration of illness varied between 1 and 9 years (mean: 4.1 years). All patients showed characteristic choreic movements and had a positive family history for Huntington's chorea. Five patients showed moderate to severe cognitive impairment. The patients showed neither signs nor symptoms indicative for other neurological diseases. Fourteen patients were taking drugs. Medication had not been changed in the 3 months prior to the study. Four patients were taking dopamine-antagonists (tetraabenazine, tiapride, haloperidol or thiopropazate). Two patients took benzodiazepines (diazepam or chlordiazepoxide) and two patients tricyclic antidepressants (amitriptyline or imipramine). The spouses, if available, were used as controls (12 controls, 6 males, 6 females, mean age: 48.8 years, age range 26 to 51 years). The controls showed neither signs nor symptoms of neurological disease and were not on any kind of medication.

Methods

The methods used for recording reflexes running along the trigeminal system have been described by Ongerboer de Visser *et al.*^{6–8} Surface electrodes were connected with a Medelec MS 6 electromyograph. Stimuli were delivered through the Medelec surface stimulator. Latencies of blink reflex, corneal reflex and jaw reflex were defined as the mean of at least three measurements. Duration of the exteroceptive suppression of activity in masseter muscles was

Table Mean values and standard deviation of blink reflex, jaw reflex and corneal reflex in 20 patients with Huntington's chorea and 12 controls

		Huntington's chorea mean SD	Controls mean SD	p
BR latency (ms)	R1	10.7 ± 2.4	10.6 ± 1.8	NS
Stimulation on right side	R2 right	41.6 ± 10.8	36.2 ± 11.4	0.001
	R2 left	44.6 ± 16.2	36.1 ± 9.8	0.001
BR latency (ms)	R1	10.9 ± 2.8	11.0 ± 2.6	NS
Stimulation on left side	R2 right	40.5 ± 9.4	34.7 ± 7.8	0.003
	R2 left	38.5 ± 9.0	33.5 ± 8.6	0.001
JR latency (ms)	Right side	5.9 ± 3.2	7.0 ± 1.0	0.02
	Left side	6.0 ± 3.3	6.8 ± 1.9	NS (0.07)
CR latency (ms)	R right	43.7 ± 22.8	41.2 ± 15.2	NS
Stimulation on right side	R left	47.8 ± 23.6	44.7 ± 16.4	NS
CR latency (ms)	R right	51.5 ± 22.6	46.6 ± 24.0	NS
Stimulation on left side	R left	46.3 ± 20.4	43.2 ± 22.2	NS

The p values pertain to Wilcoxon's two sample test. NS: no significant differences. BR: blink reflex. CR: corneal reflex. JR: jaw reflex.

defined as the mean of at least three periods free of continuous electromyographic activity. To prevent habituation effects on the blink reflex one per 60 second stimuli were delivered to the supra-orbital nerves.

Auditory evoked potentials were obtained by delivering rarefaction clicks (duration 0.1 ms, rate 16.3 Hz, intensity 70 dB) at one ear, masking the contralateral ear by a 30 dB lower level white noise. Recordings were taken from homo- and heterolateral mastoid bone, reference to the vertex (Cz region, international 10–20 system) over a period of 12 ms (including a 10% prestimulus interval). Sampling rate was 42 KHz. Averaging was performed employing an automatic artefact rejection circuit at a 95% amplifier sensitivity level (50 μ V). Amplifier bandpass was 150–3000 Hz. Positive peaks were labelled (positive meaning positivity at the vertex relative to the mastoid electrode) according to the Jewett nomenclature.⁹ The apparatus used was a Nicolet Pathfinder II. The persons studied were kept alert by verbal contact during the whole procedure.

Statistical evaluation was performed according to the Wilcoxon's two sample test.

Results

The latency of the homo- and heterolateral second response of the blink reflex on stimulation of either side was significantly prolonged (table) when compared with the controls. The right jaw reflex latency was significantly shorter in the patients than in the controls (table). The latencies of the corneal reflex on either side were longer in the patients than in the controls. However, differences in corneal reflex latencies were not statistically significant (table). The duration of the exteroceptive suppression of activity in masseter muscles and the amplitudes and latencies of the brainstem auditory evoked responses did not differ significantly between patients and controls (results not shown).

Discussion

In this study no statistically significant differences were found between patients and controls with respect to the exteroceptive suppression of activity in masseter muscles and the corneal reflex.

In the only report so far on auditory responses with latencies less than 8 ms in Huntington's chorea, normal latencies and amplitudes were reported in 10 patients.⁵ Our results confirm these latter findings in a larger patient group.

A tendency to latency decrease of the jaw reflex was noticed in the patients as compared with controls, reaching a statistically significant level on the right side. Latency decrease of the jaw reflex indicates disinhibition of the reflex arc at brainstem level as a consequence of cortical dysfunction, as may be expected in patients with Huntington's chorea.

The blink reflex has been studied in lesions of the fifth nerve, the trigeminal complex and the lower brainstem.¹⁰ Esteban *et al*⁴ reported prolonged homolateral second component latencies on right, left, or both sides in eight patients with Huntington's chorea. Our patients as a group showed homo- as well as heterolateral increased second component latencies upon stimulation on either side.

A possible influence of medication¹⁴ appeared not to be of importance in our study since three out of the four patients with the most prolonged second component latencies were not on any medication.

Cortical lesions have been reported to prolong the latency of the blink reflex second component. However, the corneal reflex appears to be more sensitive to cortical lesions than the blink reflex^{8,11} and in our patients the corneal reflex latency did not differ significantly from control values, although it tended to be increased. Therefore, blink reflex abnormalities

in our patients possibly are not to be explained by assuming impairment of descending excitatory pathways although the latter kind of impairment may have caused some corneal reflex delay.

What may be the pathophysiological meaning of the blink reflex latency increase? Preponderance of descending inhibitory pathways as a consequence of increased dopaminergic striatal influence has been proposed to account for the habituation increase of the second component of the blink reflex in Huntington's chorea.^{3,12} Preponderance of descending inhibitory pathways may very well be the explanation for the latency increase of the blink reflex second component in our patients, since a descending inhibitory preponderance does not affect the corneal reflex which is known never to habituate.¹³ Of course the latency increase of the blink reflex second component may also be caused by medullary and pontine reticular formation abnormalities, differentially affecting the blink reflex and the corneal reflex tracts, but these are thought to run closely together.¹⁰ A discrepancy between corneal reflex and blink reflex latency has not been reported in Huntington's chorea before and possibly points to a striatal cause for these abnormalities.

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