



Facial Chorea in Huntington Disease

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ABSTRACT

OBJECTIVE: To characterize upper facial chorea in patients with Huntington disease (HD).

METHODS: Patient database at the Parkinson's Disease and Movement Disorders Center was queried for active patients with the diagnosis of HD whose examination was videotaped. Videos of 25 patients with confirmed diagnosis of HD and their medical records were reviewed and scored on the following scale, adopted from the Jankovic Rating Scale.¹ In addition, the UHDRS scores for items 1, 2, 3, and UHDRS motor total scores were entered into the database.

RESULTS: Among 25 patients with HD, 75% exhibited enlarged palpebral fissures with frontalis contractions. Periods of continuous repetitive blinking was observed in 17% and 8% had spasms of the orbital portion of the orbicularis oculi muscle. Procerus and corrugator supercilii contractions were frequent in 24% and continuous in 16% of patients; absent in 48%, rare in 12%.

CONCLUSIONS: Based on rating of videos from 25 patients with HD, upper facial chorea was present in 88% and was manifested chiefly by frontalis contractions and enlargement of palpebral fissures. This phenomenological characterization may be used in the differential diagnosis of facial dyskinesias.

INTRODUCTION

Facial involuntary movements are encountered in a variety of disorders including tardive dyskinesia, cranial dystonia, and HD. Because of some overlapping clinical features there is a need to better characterize the phenomenology of the movements, particularly those involving the upper face, which may aid in the differential diagnosis of the facial dyskinesias.^{2,3,4}

METHODS

Patient database at the Parkinson's Disease and Movement Disorders Center was queried for active patients with HD. Out of 33 patients, 8 were excluded because the video recordings were not available (N = 5) or did not show the upper face with sufficient clarity (n = 3). Videos of the remaining 25 patients were carefully reviewed and the upper facial movements were rated according to modified Jankovic Rating Scale¹: Eye Closing (0–4); Eye Opening (0–4); and Procerus/Corrugator Contractions (0–3).

Table 1. Demographic Information

Mean Age ± SD (years)	53 ± 15
Age at Initial Evaluation ± SD (years)	50 ± 16
% Male	52
Mean # of CAG Repeats (range)	43 (40-47)

Table 2. Rating Scales

Eye Closing	
0	Rare or absent eyeblinks
1	Normal rate of blinking
2	Fast rate of eye blinks
3	Continuous repetitive blinking
4	Blepharospasm

Eye Opening	
0	Complete inability to open eyes
1	Normal eye opening
2	Fast rate of eye blinks
3	Enlarged palpebral fissure without frontalis contraction
4	Enlarged palpebral fissure with frontalis contraction

Procerus / Corrugator Supercilii Contractions	
0	Absent
1	Occasional
2	Frequent

RESULTS

- Of the 25 patients evaluated, 76% exhibited intermittently widened palpebral fissures associated with frontalis contractions.
- Periods of continuous repetitive blinking were observed in 16% and 8% had spasms of the orbital portion of the orbicularis oculi muscles.
- Procerus and corrugator supercilii contractions were noted in 52% of all patients and were rated as frequent in 24%, continuous in 16%, and rare in 12% of the patients.

CONCLUSIONS

- Based on rating of videos from 25 patients with HD, upper facial chorea was present in 88% and was manifested chiefly by frontalis contractions and widening of palpebral fissures.
- This phenomenological characterization may be used in the differential diagnosis of facial dyskinesias.

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