Dysphagia IN HUNTINGTON’S DISEASE: A STUDY WITH BEDSIDE SWALLOWING ASSESSMENT SCALE.

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BACKGROUND

Huntington’s disease (HD) is an inherited neurodegenerative disorder characterized by motor impairment, cognitive decline and psychiatric disorders. Dysphagia is a pathologic condition that increases morbidity and mortality of the affected people because of its complications: malnutrition, dehydration, airways obstruction and aspiration pneumonia. Patients with HD may develop dysphagia with the progression of the disease. Bedside swallowing assessment scale (BSAS) of Smithard modified by Masotti [1][2] is a simple method to apply with patients affected by neurological diseases as a dysphagia screening tool, by considering the total score with a cut-off ≥ 24 indicating inadequate and unsafe oral alimentation or hydration. A validated dysphagia scale, the Dysphagia Outcome and Severity scale (DOSS) [3], rates the functional severity of dysphagia and recommends a dietary and independence levels, thus conveying information about dysphagia severity and related disability (scores range from 1 to 7, with 7 representing normal swallowing and 1 representing severe dysphagia).

OBJECTIVE

To evaluate, in a cohort of HD patients, the prevalence and the gravity of dysphagia, its correlation with motor, cognitive and functional decline.

METHODS

Dysphagia symptoms were assessed in 38 genetically confirmed HD patients by means of BSAS of Smithard modified by Masotti, and DOSS. All patients were also evaluated by the Unified Huntington’s disease rating scale (UHDRS) [4]. A group of 39 sex and age matched controls were recruited for BSAS scores normalization.

• BSAS: (preliminary screening of swallowing abilities). The examination covered clinical items that evaluate the patient’s alertness and ability to participate, posture and respiratory status as well as the oral motor and sensory function. The protocol included a water test which consists of two 10 ml bolus of water provided in step 1 and a 60ml bolus in step 2, without time limit. Patients were observed throughout the test for signs of aspiration (reflexive cough, change in respiratory rate, or alteration in voice quality). Each item in the scale is scored according to severity. All scores from the different subskills were tallied in order to obtain a total score.

• DOSS: the scale based on clinical assessment and instrumental evaluation was employed to complete information coming from the BSAS results. Additional swallowing evaluations were conducted using different food consistencies representing the range of food consistencies consumed in real life. Other information regarding home environment, mealtime routines and use of adaptive eating aids, were collected through caregiver and family interviews.

• UHDRS: patients underwent the UHDRS for motor (Motor scale), cognitive (Cognitive assessment) and functional assessment (Total functional Capacity). To evaluate motor assessment we considered both total score and partial scores; we particularly considered some relevant items (Systemic Chorea, Chorea in the Oral Lingual district, Bradiakinesia and Dystonia).

We divided HD patients in three groups, according to BSAS total score, corrected for age: irrelevant alterations (total score ≤ 19), average alterations (total score 20-23), severe alterations (total score ≥ 24). All clinical features were compared among groups, by means of one way ANOVA and post-hoc Bonferroni test. We further attributed to each patient a DOSS score from 7 (normal level) to 1 (severe dysphagia), which was correlated with main clinical symptoms by means of Spearman correlation test.

RESULTS

The BSAS scores indicated that in our HD cohort 32.4% presented with relevant or serious dysphagia. Patients with severe dysphagia presented with more severe motor impairment compared to other patients. Cognitive and functional capacity were not significantly reduced in patients with average and severe dysphagia. Hyperkinetic movements in the oral-lingual district were significantly more severe in patients with dysphagia, compared to the rest of the patients. The DOSS levels were significantly correlated with main clinical features, such as age, disease duration and motor impairment, with special regard to chorea in the oral-buccal district and bradiakinesia. The total functional capacity scale did not show significant correlation with DOSS levels.

CONCLUSION

Dysphagia is a frequent and potentially invalidating symptom in HD patients. The DOSS scale, preceded by BSAS, indicated that dysphagia is a prevalent motor symptom of HD, specially caused by chorea in the lingual-mouth district, so patients with severe chorea in the inferior facial zone may represent a clinical phenotype with more severe swallowing disturbances. This may be relevant in order to improve early diet prescription and modification with a therapeutic approach able to modify facial chorea.

REFERENCE: