

Unawareness of dyskinesias in Parkinson's and Huntington's diseases

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Abstract We performed a clinical study to evaluate the unawareness of dyskinesias in patients affected by Parkinson's disease (PD) and Huntington's disease (HD). Thirteen PD patients with levodopa-induced dyskinesias and 9 HD patients were enrolled. Patients were asked to evaluate the presence of dyskinesias while performing specific motor tasks. The Abnormal Involuntary Movement Scale (AIMS) and Goetz dyskinesia rating scale were administered to determine the severity of dyskinesias. The Unified Parkinson's disease rating scale (UPDRS) and Unified Huntington's Disease Rating Scale (UHDRS) were used in PD and HD patients, respectively. In PD we found a significant negative relationship between unawareness score at standing and AIMS score and between unawareness score at hand pronation-supination and AIMS score for upper limbs. In HD we found a significant positive relationship between total unawareness score and UHDRS score and between total unawareness score and disease duration. In PD the unawareness seems to be inversely related with severity of dyskinesias, while in HD it is directly related to disease duration and severity.

Patients with Huntington's disease (HD) rarely complain of their dyskinesias, possibly due to an impaired subjective experience of chorea [1]. A similar phenomenon is commonly observed in patients with Parkinson's disease (PD), who may appear oblivious to their drug-induced dyskinesias. Although the phenomenon is well recognized clinically, its basis has not been systematically investigated. The aim of our study was to evaluate the unawareness of dyskinesias in PD as compared with HD patients, by means of a subjective questionnaire pertaining to the direct experience of involuntary movements while performing specific motor tasks. Subjective reports were examined in relation to objective measures of neurological status.

Thirteen PD patients (median age, 60.1 years; range, 49-77 years) with motor fluctuations and levodopa-induced dyskinesias and 9 patients with HD (median age, 58.5 years; range, 31-71) confirmed by molecular analysis participated in the study. The median duration of illness was 11.5 years (range, 6-20 years) in PD patients and 7.7 years (range, 5-10 years) in HD patients. Cognitive status was assessed by the Mini-Mental State Examination (MMSE) [2]. Only patients with an MMSE score equal to or higher than 23 were enrolled in the study. In all patients involuntary movements were evident both to the clinician and to the patient's caregiver. Patients were asked to subjectively evaluate the presence of dyskinesias while performing specific motor tasks (standing, gait, finger-tapping, hand pronation-supination). A score of unawareness ranging from 0 (full awareness) to 2 (full unawareness) was attributed during each motor task.

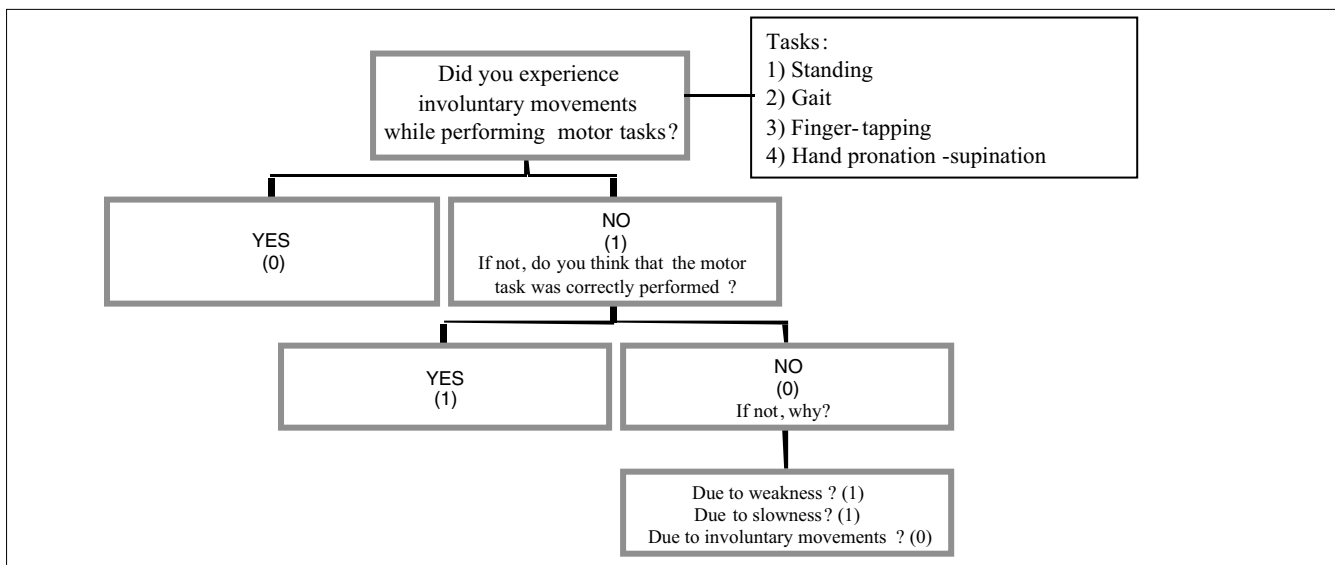


Fig. 1 Subjective questionnaire of dyskinesia awareness. Scores ranged from 0 (full awareness) to 2 (full unawareness)

Thus, unawareness total score ranged from 0 (full awareness) to 8 (full unawareness) (Fig. 1). Objective rating of dyskinesias was obtained using the Abnormal Involuntary Movement Scale (AIMS) [3] and the Goetz dyskinesia rating scale [4] in all patients. We used the Unified Parkinson's Disease Rating Scale (UPDRS) [5] and the Unified Huntington's Disease Rating Scale (UHDRS) [6] to assess illness severity. Spearman's r correlation coefficient was used to examine the relationship between subjective reported data and objective measures of disability.

Five PD patients were completely unaware of their dyskinesias, 4 were partially aware and 4 were totally aware of their dyskinesias. We found a significant negative relationship between unawareness score at standing and total AIMS score ($r=-0.7$; $p<0.01$), unawareness score at hand pronation-supination and AIMS score for upper limbs ($r=-0.6$; $p<0.05$), and unawareness score at standing and AIMS score for lower limbs ($r=-0.6$; $p<0.05$). We also found a trend to a significant negative relationship between total unawareness score and total AIMS score ($r=-0.5$; $p=0.07$). Unawareness total score was not related with UPDRS score, Goetz dyskinesia rating scale score and disease duration.

Seven HD patients were completely unaware of their dyskinesias, while 2 patients were totally aware. We found a significant positive relationship between total unawareness score and UHDRS score ($r=0.7$, $p<0.05$) and between total unawareness score and disease duration ($r=0.9$, $p=0.001$). There was no significant relationship between objective measures of dyskinesias (AIMS and Goetz dyskinesia rating scale) and unawareness total score.

Discussion

Lack of awareness of involuntary movements is commonly observed in clinical practice in both PD and HD patients. Snowden et al. [1] suggested that lack of awareness in HD patients was not a consequence of patients' cognitive impairment or a psychological defense against a debilitating disease, but reflected an impaired subjective experience of chorea. To our knowledge, failure to report dyskinesias has not been previously investigated in PD patients. Our results confirm that both PD and HD patients frequently fail to report their dyskinesias. We found that unawareness was directly related with disease severity and duration in HD patients, and inversely related with severity of dyskinesias in

PD patients. To explain these findings, different hypotheses may be suggested. According to a physiopathological hypothesis, we suggest that dopaminergic overstimulation in PD, while inducing dyskinesias by acting on basal ganglia, may affect the awareness of involuntary movements by stimulating mesocorticolimbic pathways. Instead, it is well established that disease duration and severity in HD are associated with frontal lobe lesions [7] that may explain the impaired awareness of dyskinesias in patients with long-term disease.

We cannot exclude a psychodynamic explanation of our results. According to this hypothesis, dyskinesias in PD are the expression of an overtreatment of bradykinesia, and patients may fail to report them for the simple reason that they do not experience moving abnormally. In fact, PD patients were more likely to report dyskinesias when interfering with a specific motor task. Instead, in HD dyskinesias are the main clinical manifestation, so a psychological defense mechanism of denial may explain the lack of awareness of involuntary movements in our patients.

Both physiopathological and psychodynamic hypotheses require further investigations to highlight the precise mechanisms underlying the lack of awareness of involuntary movements in PD and HD.

References

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