Awareness of Involuntary Movements in Huntington Disease

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Objective: To determine why patients with Huntington disease are apparently unaware of their involuntary movements.

Design: Correlative study using a subjective report questionnaire of physical symptoms and objective measures of neurologic and cognitive dysfunction.

Patients: Forty patients with Huntington disease attending a regional Huntington disease clinic.

Results: Patients were poor at reporting experiential symptoms of involuntary movements. There was no relationship between self-report of these symptoms and objective indices of motor dysfunction or severity of cognitive impairment. Patients could, however, report secondary consequences of their movement disorder, which correlated highly with nonchoreic indices of motor dysfunction.

Conclusions: Patients with Huntington disease have impaired subjective experience of chorea. Denial of symptoms is likely to have a physiological basis and is not a secondary consequence of patients' cognitive impairment or a psychological defense against a debilitating disease.

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RESULTS

SUBJECTIVE REPORT OF PHYSICAL SYMPTOMS

All but 2 of the 40 patients reported some physical symptoms. Symptoms relating to the consequence of movement disorder were reported more frequently than those relating to the direct experience of involuntary movements (Wilcoxon matched pairs signed rank test, z = 4.6; P < .01). Thirteen patients (33%) reported no direct experience symptoms, whereas in only 4 patients (10%) consequence-type symptoms were absent. There was no significant relationship between subjective indices of movement disorder and age or duration of illness. There was no relationship between direct experience-type symptoms and illness severity as measured by the Functional Capacity Scale, whereas consequence-type symptoms showed a significant relationship (Spearman rank correlation, rs = −.46; P < .01): fewer symptoms were associated with milder levels of disability.

RELATIONSHIP TO OBJECTIVE MEASURES OF MOVEMENT DISORDER

All patients exhibited a significant movement disorder on the QNE, defined by a QNE cutoff score greater than 3. The movement disorder varied in severity from mild (QNE total score = 8) to severe (QNE total score = 82), reflecting the wide range of illness severity within the patient group (Table 2). Similarly, the degree of chorea and nonchoreic motor impairment covered a wide range of severity. There was a positive correlation (Spearman rank correlation) between subjective reporting of physical symptoms and objective indices of movement disorder as measured by the QNE (Table 2). More subjective symptoms were associated with higher QNE scores (ie, more severe physical disability). However, this positive relationship was present only for consequence-type symptoms. Direct experience measures were not significantly related to any QNE measure, including the Chorea Scale.

PATIENTS AND METHODS

PATIENTS

Forty patients, 20 men and 20 women, with clinically diagnosed HD, confirmed by genetic testing and participated in the study. Their median age was 53 years (range, 18-76 years). The study group encompassed a spectrum of disease severity as measured by illness duration and ratings on the Functional Capacity Scale. The median duration of illness was 6 years (range, 1-20 years); mean Functional Capacity Scale rating was 6 (range, 2-13). In all, patients' involuntary movements were evident both to the clinician and to the patient's spouse or caregiver. Patients were consecutively referred to a regional HD clinic and are considered to be a representative sample of individuals with HD.

SUBJECTIVE REPORT QUESTIONNAIRE

A subjective report questionnaire of physical symptoms was developed for use with patients with HD (Table 1). Questions about physical symptoms relating to the direct experience of involuntary movements were interspersed with those relating to the consequences of abnormal movements. Patients were informed that the purpose of the questionnaire was to learn more about how patients with HD experience the disease. Patients were advised to report what they felt themselves and not what they had been told by other people. Consent was obtained from the patient after the procedure had been explained.

NEUROLOGIC EXAMINATION

Patients' neurologic status was assessed using the Quantitated Neurologic Examination (QNE), which yields a measure of abnormal movement on a scale from 0 (no abnormality) to 125 (maximum abnormality). The mean score for healthy control subjects is 1. From the QNE data, a series of cognitive tests was administered to assess patients' memory (object recall and story recall), mental manipulative skills (reversal of months of the year, backward digit span and road map test of left-right orientation), abstraction and mental set shifting (Modified Wisconsin Card Sorting Test), generational skills (category and word fluency), organization and planning (picture sequencing), attention (Stroop), and mental speed (reciting months forward). We have found these tests to distinguish patients with HD from healthy controls and from asymptomatic carriers of the HD gene and are therefore sensitive to the presence of HD. Comparable findings have been reported by other authors. The card sorting, verbal fluency, picture sequencing, and Stroop tasks are known to be sensitive to frontal lobe dysfunction. The National Adult Reading Test was used to provide a measure of patients' premorbid intellectual ability.

STATISTICAL ANALYSIS

Nonparametric analyses applicable to ordinal data were performed using the SPSS-PC statistical package (SPSS Inc, Chicago, Ill). The Wilcoxon matched pairs signed rank test was used to compare frequency of reported direct experience and consequence-type symptoms. The Spearman rank correlation coefficient, which provides a measure of association between 2 variables and has high power efficiency, was used to examine the relationship between subjective reported data and objective measures of disability.
scores, even though it was chorea that the experiential symptom questions were specifically designed to tap. Moreover, the positive relationship between consequence measures and objective scores was present only for nonchoreic aspects of movement disorder, such as bradykinesia, measured by the Motor Impairment Scale, and not for chorea, measured by the Chorea Scale.

RELATIONSHIP TO COGNITIVE IMPAIRMENT

The extent of cognitive impairment shown by patients differed markedly across the group, commensurate with differing levels of functional disability and duration of illness. Test scores ranged from normal or mild impairment to floor level performance. Despite this wide spectrum of cognitive disability, there were relatively few significant associations between subjective reports of physical symptoms and objective measures of cognitive function (Table 3). Where significant relationships were present, these were invariably associated with consequence measures. There was no relationship between direct experience measures and objective measures of cognitive function, and in particular, no association between direct experience measures and frontal lobe test performance. The significant associations with consequence measures were all in the same direction: fewer subjective symptoms was associated with better test performance. There was no evidence that report symptoms was associated with more severe cognitive impairment, as predicted by the cognitive impairment hypothesis. The strongest associations with consequence-type symptoms occurred for tests with a high motor speed component (Table 3): recitation of the months of the year and word reading on the Stroop test. There was no relationship between the report of physical symptoms and premorbid intellectual attainments, predicted from scores on the National Adult Reading Test, suggesting that patients’ capacity to

Table 1. Subjective Report Questionnaire

Direct experience questions
Do you ever feel fidgety, as if your arms and legs are on the go?
Do you ever feel your face twitch?
Do you ever feel that your mouth moves against your will?
Do you ever have the feeling that your fingers twitch?
Do you ever feel your arms jerking or moving against your will?
Do you experience odd sensations in your arms?
Have you noticed any odd sensations in your legs?
Do you ever feel your legs moving involuntarily?
Do you ever have the feeling that your toes twitch?

Consequence questions
Have you noticed any change in your physical abilities?
Do you choke or splutter when eating or drinking?
Do you think there has been any change in the clarity of your speech?
Do you drop things?
Do you have difficulty doing up buttons?
Do you spill drinks?
Do you trip or fall over?
Do you lose your balance?
Do you bump into things?

Table 2. QNE Scores and Their Relationship to Subjective Report Measures*

<table>
<thead>
<tr>
<th>Instrument (Maximum Score)</th>
<th>Score</th>
<th>Consequences of Movement Disorder, r.s</th>
<th>Direct Experience of Movement Disorder, r.d</th>
</tr>
</thead>
<tbody>
<tr>
<td>QNE total (125)</td>
<td>35</td>
<td>8-62</td>
<td>0.41†</td>
</tr>
<tr>
<td>Chorea Scale (25)</td>
<td>9</td>
<td>4-15</td>
<td>-0.12</td>
</tr>
<tr>
<td>MIS (28)</td>
<td>8</td>
<td>1-21</td>
<td>0.80‡</td>
</tr>
</tbody>
</table>

*QNE indicates Quantitated Neurologic Examination; MIS, motor impairment scale; and r.s, Spearman rank correlation coefficient.
†P<.05.
‡P<.001.

Most patients with HD in the study reported some physical symptoms on specific questioning. However, patients were more likely to report symptoms pertaining to the consequences of movement disorder, such as dropping objects, than the direct experience of involuntary movements, such as feelings of twitching in the fingers. Statistical analyses, using a measure of association, revealed no relationship between direct experience measures and objective indices of movement disorder, in particular of chorea, which the direct experience questions were designed to tap. Involuntary movements that were detected on neurologic examination were poorly reported by patients themselves. This finding cannot be attributed to a general loss of insight into the movement disorder, since reports of the consequences of abnormal movements were significantly correlated with objective measures of movement disorder: the more subjective the symptoms, the poorer the QNE scores. These findings suggest that patients fail to experience involuntary movements, yet are aware of the consequences of their movement disorder.

What do the consequence measures actually measure? Do they represent the secondary consequences of the involuntary movements per se, or alternatively, do they reflect impairment in the voluntary aspects of the movement disorder? The finding that consequence-type symptoms were strongly related to nonchoreic aspects of the movement disorder but were unrelated to measures of chorea is relevant in this regard. It suggests that the consequence-type symptoms represent the effects of nonchoreic aspects of the movement disorder, such as bradykinesia, and not of chorea. Such a view is supported, moreover, by the cognitive test data. Significant relationships with cognitive test performance occurred only for consequence-type symptoms, and the strongest relationships were with tests that have high demands on motor speed. Patients who performed more slowly on these tasks reported more consequence-type symptoms. The finding of a significant correlation between consequence-type symptoms and functional capacity would imply that it is nonchoreic aspects of movement disorder and not chorea that lead to functional disability. Indeed, chorea has been shown to be unrelated to functional decline.12
Since subjective reports accurately reflect components of the movement disorder such as motor slowing, they suggest that patients may experience those aspects of their disorder. It is specifically the choreiform movements of which patients with HD are unaware.

These findings rule out a psychodynamic explanation of patients’ unawareness of chorea: it is not that patients are denying physical symptoms. Indeed, some symptoms would appear to be reported accurately because of their association with objective measures of movement disorder. The cognitive hypothesis also can be excluded. There was no significant relationship between failure to report physical symptoms and severity of cognitive impairment, in particular, frontal lobe dysfunction. Indeed, the few significant relationships were in the reverse direction: having fewer symptoms was associated with superior cognitive test performance compatible with milder illness. That the strongest association was with timed tasks (recitation and reversal of the months of the year and word reading) suggests that the physical symptoms that patients with HD accurately report relate to their physical slowing (bradykinesia).

Our findings are consistent with the physiological hypothesis: patients with HD fail to complain of their involuntary movements because they do not have a subjective experience of chorea. They appear to be aware only of the repercussions of their movement disorder, or more precisely those aspects of their movement disorder, such as physical slowing, that are distinct from chorea. This apparent dissociation in awareness for different components of the movement disorder is not unique to HD. A parallel phenomenon is commonly observed in patients with Parkinson disease. Patients who are distressed with their bradykinesia may nonetheless appear oblivious to their drug-induced dyskinesias. The physiological hypothesis requires critical scrutiny by future studies. The precise mechanisms that might underlie the experience or lack of experience of movement remain to be discerned. Nevertheless, the physiological hypothesis is important in highlighting a viable alternative to the traditional interpretations of why patients with HD fail to report chorea.

These findings have implications for the treatment and management of HD. In the past the use of dopamine-depleting drugs, such as tetrabenazine, for the treatment of chorea was widespread. In recent years their use in HD has become more restricted on the grounds that such pharmacological agents may exacerbate motor disability and increase patients’ predisposition to functionally disabling symptoms, such as depression. Our finding that patients with HD are unaware of their choreiform movements reinforces the validity of that conservative position. It would seem inappropriate to treat an aspect of motor disorder of which the patient is unaware with agents that may worsen those aspects of motor dysfunction for which the patient does have awareness and that are associated with greatest functional disability.

<table>
<thead>
<tr>
<th>Test</th>
<th>n</th>
<th>Task Condition</th>
<th>Score</th>
<th>Consequence Symptoms, rs</th>
<th>Direct Experience Symptoms, rs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Object recall</td>
<td>39</td>
<td>Immediate (maximum score, 20)</td>
<td>5</td>
<td>3-11</td>
<td>-0.33†</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Delayed (maximum score, 20)</td>
<td>4</td>
<td>0-9</td>
<td>-0.36†</td>
</tr>
<tr>
<td>Story recall</td>
<td>39</td>
<td>Immediate (maximum score, 14)</td>
<td>3</td>
<td>0-10</td>
<td>-0.20</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Delayed (maximum score, 14)</td>
<td>2</td>
<td>0-8</td>
<td>-0.24</td>
</tr>
<tr>
<td>Months</td>
<td>39</td>
<td>Forward RT</td>
<td>10</td>
<td>4-45</td>
<td>0.42†</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Backward RT</td>
<td>38</td>
<td>14-105</td>
<td>0.41†</td>
</tr>
<tr>
<td>Road map§</td>
<td>31</td>
<td>% Correct</td>
<td>66</td>
<td>41-100</td>
<td>-0.31</td>
</tr>
<tr>
<td></td>
<td></td>
<td>RT</td>
<td>170</td>
<td>75-840</td>
<td>0.38†</td>
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<tr>
<td>Digit span</td>
<td>39</td>
<td>Forward</td>
<td>5</td>
<td>3-7</td>
<td>-0.19</td>
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<tr>
<td></td>
<td></td>
<td>Backward</td>
<td>3</td>
<td>1-4</td>
<td>0.09</td>
</tr>
<tr>
<td>Card sorting</td>
<td>39</td>
<td>Categories (maximum, 8)</td>
<td>2</td>
<td>0-7</td>
<td>-0.33†</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Total errors (maximum, 48)</td>
<td>23</td>
<td>1-42</td>
<td>-0.07</td>
</tr>
<tr>
<td>Word fluency</td>
<td>39</td>
<td>% Perseverations</td>
<td>37</td>
<td>0-100</td>
<td>-0.09</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Total errors (maximum, 48)</td>
<td>23</td>
<td>1-42</td>
<td>-0.07</td>
</tr>
<tr>
<td>Picture sequencing</td>
<td>36</td>
<td>Errors (maximum, 24)</td>
<td>4</td>
<td>0-12</td>
<td>0.11</td>
</tr>
<tr>
<td></td>
<td></td>
<td>RT</td>
<td>253</td>
<td>100-709</td>
<td>0.09</td>
</tr>
<tr>
<td>Stroop†</td>
<td>26</td>
<td>Black/white reading RT</td>
<td>80</td>
<td>48-288</td>
<td>0.47†</td>
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<td></td>
<td></td>
<td>Color reading RT</td>
<td>86</td>
<td>45-280</td>
<td>0.50†</td>
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<td></td>
<td>Color naming RT</td>
<td>134</td>
<td>72-450</td>
<td>0.34</td>
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<tr>
<td></td>
<td></td>
<td>Interference RT</td>
<td>230</td>
<td>107-950</td>
<td>0.28</td>
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<tr>
<td></td>
<td></td>
<td>Interference errors</td>
<td>4</td>
<td>0-45</td>
<td>-0.03</td>
</tr>
</tbody>
</table>

*rs indicates Spearman rank correlation coefficient; RT, response time in seconds.
†P < .05.
‡P < .01.
§The road map test requires left/right judgments. The chance level of performance is 50% correct.
| The Stroop task includes (a) black/white reading, involving reading names of colors printed in black and white; (b) color reading involving reading names of colors printed in colored ink; (c) color naming involving naming blocks of colors; and (d) interference involving naming the ink color of incongruous color words (eg, red written in green ink).
Our findings also have relevance to the understanding of HD. Patients who fail to report symptoms should not be dismissed as demented or in denial. Their failure to report symptoms reflects their subjective experience. Clinicians who inform patients newly diagnosed as having HD that they have the disease should bear in mind that the chorea that is so immediately apparent to the external observer may not be within the subjective experience of the patients themselves.

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REFERENCES


