The Ekman 60 Faces Test as a diagnostic instrument in frontotemporal dementia

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Abstract

Frontotemporal dementia (FTD) is characterized by dramatic changes of personality and behaviour. Impaired ability of emotional processing could contribute to these symptoms, as it may lead to misinterpretation of emotional cues that would normally guide behaviour. The aim of the present study was to investigate if the Ekman 60 Faces Test, an instrument to test the recognition of basic facial emotions, enables the differentiation between patients with mild FTD and cognitively healthy subjects (HC). We found that compared to 33 cognitively healthy subjects, 25 patients with mild FTD were impaired in the recognition of basic emotions. At a cut-off score from 46 out of 60 points, the Ekman 60 Faces Test discriminated between patients with mild FTD and HC with 97% diagnostic accuracy (sensitivity: 94%; specificity: 100%). The results of the present study were consistent with the findings of prior studies on smaller patient samples.

Keywords: Frontotemporal dementia; Behavioural disturbances; Recognition of emotions; Ekman 60 Faces Test

1. Introduction

Frontotemporal lobar neurodegeneration (FTLD) causes 30–50% of presenile dementias (Hodges, Davies, Xuereb, Kril, & Halliday, 2003; Ratnavalli, Brayne, Dawson, & Hodges, 2002). Recent consensus criteria (Neary et al., 1998) divide FTLD into three major subtypes: (1) frontotemporal dementia (FTD), denoting a behavioural syndrome with selective involvement of the frontal and/or temporal cortices; (2) semantic dementia defined as a disorder of language, semantics and recognition of visual percepts caused by predominant anterior temporal pathology; and (3) progressive non-fluent aphasia, a syndrome associated with asymmetric degeneration of the frontotemporal cortex in the language-dominant hemisphere.

Behavioural disturbances in FTD often occur before cognitive deficits become obvious. Impairment of modulating behaviour results in inappropriate responses or activities. One of the first conspicuous symptoms is the dissolution of social attachment. Patients are indifferent and unconcerned about their spouses, relatives or friends. Social behaviour often becomes superficial, tactless, rude and sometimes foolish. Disinhibition may be observed in terms of hyperorality or inappropriate sexual behaviour. Patients are inflexible, rigid and unable to adjust to their environment. Affect is...
typically shallow and monotonous. Some patients show euphoric or even hypomanic mood. Patients with FTD may be overactive and show motor restlessness or pressure of speech. Apathetic behaviour, however, is more frequent. Typically, there is a lack of insight into the disease (Diehl & Kurz, 2002; Greck & Kurz, 2000; Neary et al., 1998). In contrast, however, the disease has little impact on the patients’ cognitive performance at the early stage. Basic memory and spatial abilities are relatively preserved. Patients typically achieve normal scores on simple cognitive screening tests, which makes these instruments insensitive for the detection of FTD (Hodges & Miller, 2001). Even the results of more demanding neuropsychological tests including tests for frontal lobe functions can be within a normal range at the early stage (Gregory, Serra-Mestres, & Hodges, 1999).

It is not clear which mechanisms give rise to the dramatic changes of personality and behaviour that occur in patients with FTD. Impaired ability of emotional processing could contribute to the behavioural disturbances in FTD, as it may lead to misinterpretation of emotional cues that would normally guide behaviour.

Recognition of affective states, particularly the basic emotions such as sadness, disgust, anger, fear and surprise, is a prerequisite of intact social behaviour. A great deal of interpersonally relevant information is gathered from the observation of faces. Deficits in processing these perceptions could make appropriate reactions impossible. Most brain structures that participate in the recognition of basic emotions involve perceptual processing (i.e., identifying the geometric configuration of facial features in order to discriminate between different stimuli on the basis of their appearance) and recognition of the emotional meaning of a stimulus (Adolphs, 2002).

Several small studies have demonstrated that the ability of identifying facial expressions of emotions is impaired in patients with FTD (according to the revised Lund–Manchester criteria) as compared to healthy persons. Keane, Calder, Hodges, and Young (2002) administered photographic representations of six basic emotions from the Ekman and Friesen series (Ekman & Friesen, 1976) to six patients with FTD and 12 healthy control subjects (HC). They found that the ability to recognize happiness, sadness and anger—but not fear or disgust—was significantly impaired in patients compared to HC (Keane et al.). Fernandez-Duque and Black (2005) examined six patients with FTD with the Ekman 60 Faces Test and found that the recognition of negative emotions was impaired. Rosen et al. (2004) compared the performance of 13 patients with FTD and 16 HC with the facial identity discrimination subtest of the Florida Affect Battery (Bowers, Blonder, & Heilman, 1992) that consists of photographs of faces depicting one of five expressions: happiness, sadness, anger, fear and neutral. Patients were impaired on all emotion subtests (Rosen et al.). Lough et al. (2006) used quite an experimental design: morphed images of faces were shown, that ranged in sequence from neutral to full expression of one of six basic emotions. This study found that emotion recognition was globally impaired in patients with fvFTD, particularly for anger and disgust (Lough et al.).

In the present study, the Ekman 60 Faces Test was chosen to assess the ability to identify facial expressions of emotions. The objective of the study was to determine how well the Ekman 60 Faces Test differentiates between patients with mild FTD and cognitively healthy persons. We hypothesized that the Ekman 60 Faces Test should be able to discriminate patients with mild FTD and healthy persons.

2. Patients and methods

2.1. Patients and healthy control subjects

Twenty-five outpatients (17 male, 8 female) diagnosed with FTD according to the revised Lund–Manchester criteria (Neary et al., 1998) were randomly recruited from a sample of 34 patients with mild FTD, diagnosed between 2003 and 2005 in our memory clinic.

The diagnostic process consisted of a psychiatric and neurological examination as well as laboratory screening. History was taken from an informant, and behavioural disturbances were assessed using a standardized interview, the Frontal Behavioural Inventory (FBI) (Kertesz, Davidson, & Munoz, 1997). Patients underwent neuropsychological testing including the German versions of the Mini-Mental-State-Examination (MMSE) (Folstein, Folstein, & McHugh, 1975), the Consortium to Establish a Registry of Alzheimer’s Disease-Neuropsychological battery (CERAD-NAB) (Monsch, 1997) and frontal-executive tests (Frontal Assessment Battery) (Dubois, Slachevsky, Litvan, & Pillon, 2000), Color-Word-Test (Fleischmann & Oswald, 1994), Trail Making Test (Reitan, 1958). Disease severity was assessed using the Clinical Dementia Rating (CDR) (Hughes, Berg, Danziger, Coben, & Martin, 1982). Patients with moderate or severe FTD (CDR = 2 or 3) were excluded from the study. Every patient underwent cranial computed tomography or magnetic resonance imaging to identify and exclude focal lesions. Furthermore, cranial positron emission tomography
was performed, demonstrating frontal or frontotemporal glucose hypometabolism typical for FTD (Diehl et al., 2004) in all patients. Patients with semantic dementia or non-fluent progressive aphasia were excluded from the study as well as patients with other forms of dementia.

Ethical approval was obtained from the local Ethics Committee. Thirty-three healthy control subjects were spouses of unrelated patients who had been examined at the same unit as the FTD patients. HC were matched by age and gender. HC had no history of neurological or psychiatric disorders and did not complain about cognitive deterioration. An extensive interview was performed to rule out dementia. The MMSE was administered to all control subjects. Individuals who scored less than 27 out of 30 points were excluded.

2.2. The Ekman 60 Faces Test

The Ekman 60 Faces Test uses a range of photographs from the Ekman and Friesen series of Pictures of Facial Affect (Ekman & Friesen, 1976), which has been the most widely used and validated series of photographs in facial expression research. From this series, the faces of 10 actors (6 female, 4 male) were chosen, each displaying six basic emotions (happiness, sadness, disgust, fear, surprise and anger). The Ekman 60 Faces Test can be used to assess recognition of facial expressions of basic emotions. The maximum test score indicating best performance is 60 for all six emotions and 10 for each basic emotion.

The computer software for the test was available on CD-ROM. Patients were allowed unlimited time for the response. Immediately prior to testing, we verified that patients and HC semantically understood the words anger, disgust, fear, happiness, sadness and surprise. Patients and HC were asked to provide an example for each emotion by answering the questions: “Name a situation when you feel happiness, fear, etc…”. Any incorrect answer would have led to exclusion from this study; however, this was not the case.

2.3. Statistical analysis

Comparisons between FTD patients and controls regarding demographic variables and scores on the MMSE and the Ekman 60 faces tests were performed using the Chi²-test and the Mann–Whitney U-test, respectively. The association between MMSE and Ekman 60 Faces Test scores of the patients were examined by calculating Pearson’s correlation.

Receiver Operating Characteristic (ROC) curve analysis was used to plot the hit rate (proportion of patients correctly classified as impaired) against the false alarm rate (controls incorrectly classified as impaired) for every possible threshold on each test. The optimal cut-off score of the Ekman 60 Faces Test for the discrimination between patients and HC was determined using Youden’s index. The area under the ROC curve (AUC) was also calculated as a measure of test accuracy.

3. Results

There were no significant differences between patients and HC regarding demographic variables (Table 1).

Patients scored significantly lower on the MMSE (25.3 ± 3.1) than HC (29.0 ± 1.2). Significant differences were detected between patients and HC on the Ekman 60 Faces Test. The total score was lower in patients (mean: 29.8 ± 7.4 out of 60 points) than in HC (mean: 49.8 ± 3.0). Patients with FTD were impaired in the recognition of all six basic emotions relative to healthy control subjects. Results are shown in Table 2.

Table 1

<table>
<thead>
<tr>
<th>N</th>
<th>Female/male</th>
<th>Age</th>
<th>Education (years)</th>
<th>MMSE</th>
<th>FBI</th>
</tr>
</thead>
<tbody>
<tr>
<td>HC</td>
<td>33</td>
<td>15/18</td>
<td>60.9 (10.8)</td>
<td>13.3 (2.6)</td>
<td>29.0 (1.2)</td>
</tr>
<tr>
<td>FTD</td>
<td>25</td>
<td>8/17</td>
<td>63.2 (10.6)</td>
<td>12.8 (4.1)</td>
<td>25.3 (3.1)</td>
</tr>
</tbody>
</table>

N: number of patients; n.a.: not applicable.

* Significant difference HC > FTD (p < 0.001).
Table 2
Performance (mean scores with standard deviation in parentheses) on the Ekman 60 Faces Test

<table>
<thead>
<tr>
<th></th>
<th>HC</th>
<th>FTD</th>
<th>Difference</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total</td>
<td>49.8 (3.0)</td>
<td>29.8 (7.4)</td>
<td>HC &gt; FTD *</td>
</tr>
<tr>
<td>Total minimum–maximum</td>
<td>41–54</td>
<td>18–45</td>
<td></td>
</tr>
<tr>
<td>Happiness</td>
<td>10.0 (0.2)</td>
<td>9.5 (0.5)</td>
<td>HC &gt; FTD *</td>
</tr>
<tr>
<td>Sadness</td>
<td>7.0 (1.6)</td>
<td>3.8 (1.9)</td>
<td>HC &gt; FTD *</td>
</tr>
<tr>
<td>Disgust</td>
<td>8.7 (1.3)</td>
<td>3.7 (3.0)</td>
<td>HC &gt; FTD *</td>
</tr>
<tr>
<td>Fear</td>
<td>6.8 (1.9)</td>
<td>3.2 (1.9)</td>
<td>HC &gt; FTD *</td>
</tr>
<tr>
<td>Surprise</td>
<td>8.5 (1.2)</td>
<td>5.0 (3.0)</td>
<td>HC &gt; FTD *</td>
</tr>
<tr>
<td>Anger</td>
<td>9.2 (1.0)</td>
<td>4.4 (2.1)</td>
<td>HC &gt; FTD *</td>
</tr>
</tbody>
</table>

* p < 0.001.

Fig. 1. ROC curve for the Ekman 60 Faces Test: discrimination between patients with FTD and HC.

(Fig. 1). The area under the curve was 0.97 at a cut-off of 46 out of 60 points. Sensitivity and specificity were 94% and 100%, respectively. None of the patients scored higher than 45 out of 60 points, whereas only two healthy control subjects scored less than 45 points.

4. Discussion

The study demonstrates that patients with FTD are severely impaired on the task of identifying the facial expression of basic emotions including happiness, sadness, disgust, fear, surprise and anger. These results are consistent with the findings of a recent study of Rosen et al. (2004).

As expected, patients scored significantly lower on the MMSE than HC. However, poor performance on the Ekman 60 Faces Test cannot probably be explained by crude cognitive deficits captured by the MMSE. Patients’ results on the MMSE were relatively high and indicate that cognitive impairment was mild in all patients. Furthermore, we did not find a correlation between the patients’ results on the Ekman 60 Faces Test and on the MMSE. In addition, previous studies also have shown that impaired ability to recognize the facial expression of emotion is not accounted for by other domains of cognitive impairment, even when assessed with appropriate neuropsychological measures (Keane et al., 2002; Rosen et al., 2002, 2006).

We found that the Ekman 60 Faces Test is able to discriminate between patients with FTD and HC with excellent diagnostic accuracy.
While the differential diagnosis between FTD and AD has been subject of various studies including neuropsychological testing and brain imaging (Diehl et al., 2005; Jenner, Reali, Puopolo, & Silveri, 2006; Mathuranath, Nestor, Berrios, Rakowicz, & Hodges, 2000; Whitwell & Jack, 2006), research about the discrimination between FTD and healthy persons has been neglected so far. A recent study has shown that in 3 of 17 patients with FTD, no medical explanation was given at the first presentation (Pijnenburg, Gillissen, Jonker, & Scheltens, 2004). This goes along with our clinical experience: several patients with early FTD are diagnosed as healthy by neurologists or psychiatrists. When subtle behavioural disturbances are not obvious during a consultation, in many cases, neither neuropsychological testing nor imaging is performed. A study of our own group showed that the diagnostic latency between the onset of first symptoms and exact diagnosis was 4 years (Diehl-Schmid et al., 2006), in part reflecting the problems of early diagnosis of FTD.

This points to the fact that there is probably quite a long preclinical stage, sometimes referred to as “mild behavioural impairment (MBI)” (Schoelzel-Dorenbos, 2006). In contrast to “mild cognitive impairment”, that can be easily diagnosed, there are no tests for the detection of MBI. If further studies are able to proof the validity of the Ekman 60 Faces Test as a diagnostic tool in early FTD, the test should be used in prospective follow-up studies of patients suspected of MBI to find out if the Ekman 60 Faces Test is able to detect FTD in preclinical stages.

However, it has to be kept in mind that recognition of facial expressions can be impaired in several neurological and psychiatric disorders such as Parkinson’s disease (Sprengelmeyer et al., 2003) or schizophrenia (Hall et al., 2004). On the contrary, recognition of facial emotions seems to be unimpaired in patients with mild Alzheimer’s disease (Burnham & Hogevorst, 2004; Keane et al., 2002). Further studies have to investigate the ability of the Ekman 60 Faces Test to discriminate between FTD and Alzheimer’s disease and other neurological or psychiatric disorders.

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