



Differential deficits in expression recognition in gene-carriers and patients with Huntington's disease

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Abstract

Previous studies in symptomatic patients and asymptomatic gene-carriers of Huntington's disease (HD) reported a differential deficit in the recognition of facial expressions of disgust. This impairment may point to involvement of the basal ganglia in the recognition of disgust. In this study, we compared the performance of 20 patients with symptoms of HD, 20 gene-carriers of HD and 20 healthy controls on two tests of facial expressions in order to further investigate the role of the basal ganglia in disgust recognition. Recognition of fear, rather than disgust, was most severely impaired in the patients, who were also impaired at recognising expressions of anger, disgust and sadness. Direct testing for a differential deficit in disgust at the group level (and at the level of individual HD cases) revealed that the patients were in fact significantly more impaired on the other negative expressions than on disgust. The gene-carriers were not impaired on any expression, although there was a trend for the gene-carriers to be poorer at recognising fearful faces than the controls. We argue that the expression recognition performance of the patients and gene-carriers simply reflects differences in task difficulty, rather than dysfunction of any mechanisms dedicated to specific emotions. In contrast to previous studies in patients or gene-carriers of HD, our findings provide no evidence for a role of the basal ganglia in the recognition of disgust and cast doubt on whether results from HD patients and gene-carriers can be used in support of a double dissociation between recognition of disgust and fear.

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1. Introduction

Selective or differential impairments recognising specific emotional expressions have attracted great interest as these may indicate that different (dissociable) mechanisms underlie recognition of different emotions. To date, most reports of selective impairments come from patients with lesions involving the amygdala who are particularly impaired at recognising expressions of fear (Adolphs, Tranel, Damasio, & Damasio, 1994; Adolphs et al., 1999; Broks et al., 1998; Calder et al., 1996; Sprengelmeyer et al., 1999). Together with neuroimaging studies (Morris et al., 1996; Phillips et al., 1998; Whalen et al., 1998) showing increased amygdala activation in healthy subjects on presentation of fearful faces, the patient data were seen as evidence for a role of the amygdala in processing expressions of fear. However, the claim that selective impairments in fear recognition may reflect damage to

a specialised neural mechanism has been criticised on the ground that this selective impairment may simply reflect differences in task difficulty. In healthy, normal subjects from various cultures, facial expressions of fear are typically recognised more poorly than any other expression (Biehl et al., 1997; Ekman & Friesen, 1976). In line with the argument of task difficulty, Rapcsak et al. (2000) found that recognition of facial expressions of fear was most severely impaired in a group of 63 patients with focal lesions in various brain areas. Furthermore, Rapcsak et al. (2000) showed that, although fear was the most poorly recognised emotion in patients with amygdala lesions, the same was true for patients whose neurological damage had spared the amygdala. Another recent group study in patients with focal lesions in various locations also reported that recognition of facial expressions of fear, together with sadness, was most severely impaired (Weniger & Irle, 2002). Differential deficits in fear recognition were also found in patients with diffuse brain damage following head injury (Braun, Baribeau, Ethier, Daigneault, & Proulx, 1989; Hopkins, Dywan, & Segalowitz, 2002).

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An important argument against the explanation of the selective impairment at fear recognition in terms of task difficulty came from findings in patients with Huntington's disease (HD), which suggested a double dissociation between impairments in recognising fear and disgust. Sprengelmeyer et al. (1996, 1997b), found that patients with HD were impaired compared to healthy controls at recognising facial expressions of surprise, fear, sadness, disgust and anger, but that the impairment at recognising disgust was more severe than any other emotion, including fear. Recently, Wang, Hoosain, Yang, Meng, and Wang (2003) reported a similar differential deficit in the recognition of disgust in six HD patients. From their findings Sprengelmeyer et al. (1996) suggested that recognising disgust may rely on a different neural substrate than recognising fear. However, determining the possible location of this substrate was difficult given the widespread neural degeneration in patients with symptomatic HD. Gray, Young, Barker, Curtis, and Gibson (1997) addressed this issue of localisation by examining recognition of facial expressions in gene-carriers of HD. Although emergence of the motor symptoms is regarded as the first symptom of HD, there is evidence for early signs of the disease in gene-carriers who are free of motor disturbances, such as atrophy and hypometabolism in the basal ganglia (Antonini et al., 1996; Aylward et al., 1994). Furthermore, asymptomatic gene-carriers can show patterns of cognitive impairments that are similar to those found in symptomatic patients (Hahn-Barma et al., 1998; Lawrence et al., 1998). Gray et al. (1997) argued that impaired recognition of disgust in the early stages of the disease would point at involvement of the basal ganglia, since the basal ganglia, and in particular the caudate nucleus is generally considered as the initial site of pathology in HD. Gray et al. (1997) reported that, when compared to persons who had also been genetically screened for HD but who did not carry the gene mutation, asymptomatic gene-carriers were only impaired at recognising disgusted faces. This is in line with a role of the basal ganglia in recognising disgust, and more importantly, provided further supporting evidence for the putative double dissociation between fear and disgust. However, in a very recent study, in persons at risk for HD Gray and Barker (2002) found that gene-carriers were impaired at recognising both facial expressions of fear and disgust relative to non-gene-carriers.

Another report of selectively impaired recognition of disgust came from patients with obsessive-compulsive disorder (OCD) (Sprengelmeyer et al., 1997a). Compared to normal controls, patients with OCD were severely impaired at recognising facial expressions of disgust but not other emotions. OCD is often associated with abnormal metabolism in the orbitofrontal cortex and the basal ganglia, including the caudate nucleus (Micallef & Blin, 2001). Therefore, the finding of a selective impairment for disgust in OCD patients could further support basal ganglia involvement

in disgust recognition. Finally, Wang et al. (2003) found recognition of disgust to be particularly impaired in patients with Wilson's disease, a hereditary neuro-degenerative disease which is associated with atrophy mainly in the basal ganglia.

Contrary to these neuropsychological studies, neuroimaging studies in healthy subjects showed activation in response to expressions of disgust in areas outside the basal ganglia, which question the importance of the basal ganglia in processing disgust. Phillips et al. (1997, 1998) found increased activation to disgusted faces in the putamen, but also in the anterior insula, extrastriate areas and areas in the frontal and temporal cortex. Similarly, Sprengelmeyer, Rausch, Eysel, and Przuntek (1998) found increased activation in the putamen, insula and the inferior frontal cortex to disgusted faces relative to neutral faces. Recently, Calder, Keane, Manes, Antoun, and Young (2000) reported a patient with lesions in the left insula and the basal ganglia (putamen, globus pallidus and part of the caudate nucleus), caused by an infarction, who showed a selective impairment at recognising facial as well as vocal expressions of disgust. On all other expressions (fear, anger, surprise, happiness, sadness and contempt) the patient scored within the normal range. This case study is in line with the findings from HD patients and gene-carriers and OCD patients in suggesting involvement of the basal ganglia in disgust recognition, but also with the neuroimaging studies that suggested involvement of other brain areas, such as the insula.

Given this uncertainty regarding the role of the basal ganglia in the recognition of disgust, it would be important to compare recognition of emotional expressions between people at a very early stage of HD, in whom the neuropathology would be restricted to the basal ganglia, and people at a later stage of the disease. Apart from the studies by Sprengelmeyer et al. (1996, 1997b), Gray et al. (1997), Gray and Barker (2002) and Wang et al. (2003), we are not aware of other studies that reported differential impairments in expression recognition in patients or gene-carriers of HD. Previous studies did find impaired recognition of emotional expressions in the face (Jacobs, Shuren, & Heilman, 1995) or the voice (Speedie, Brake, Folstein, Bowers, & Heilman, 1990) in HD, but none of these studies looked at impairments for specific emotions. In the study reported here, we assessed recognition of facial expressions in a group of patients with symptoms of HD and a group of asymptomatic gene-carriers who both came from families in the northeast of Scotland, using the same tests in both groups. This allowed us to examine whether already at a very early stage of HD there is a differential deficit for recognition of disgust, which continues into later stages of the disease. Such results would corroborate the double dissociation between disgust and fear recognition and provide further support for involvement of the basal ganglia in recognising disgust in facial expressions.

2. Materials and methods

2.1. Subjects

Twenty patients (12 male, 8 female) with a diagnosis of Huntington's disease agreed to participate in this study. Mean age of the patients was 47.6 (S.D. 8.45) and mean number of years in education was 11.6 (S.D. 2.2). The mean time since onset of the motor symptoms was estimated at 6.5 years (S.D. 3.2). In addition to the patients, 20 persons (7 male, 13 female) who carried the gene mutation but who had not yet developed symptoms of HD also participated in this study. Mean age in this group of gene-carriers was 38.4 years (S.D. 9.5), and mean number of years of education was 12.7 (S.D. 2.1). All gene-carriers had been screened by presymptomatic predictive testing of the CAG expansion in the Huntington gene and carried the typical pathological expansion in this gene. The third group of participants consisted of normal, healthy control subjects (12 male, 8 female). This control group was matched for age and years of education to the patient group; mean age was 47.9 (S.D. 9.3), mean number of years of education was 11.8 (S.D. 2.1). Mean premorbid IQ of the patients, as estimated with the National Adult Reading Test (NART), was 105.8 (S.D. 7.41), which did not differ significantly from the NART IQ of the gene-carriers (mean 110.1, S.D. 6.1) and the controls (mean 109.0, S.D. 6.0) ($P > 0.1$). The patients and the gene-carriers were recruited through the Department of Medical Genetics of Aberdeen Royal Infirmary. The healthy controls were recruited from the general public. All participants gave informed consent to participate in this study, which was approved by the Grampian Research Ethics Committee.

2.2. Tests

2.2.1. Recognising facial expressions

This test consists of 60 photographs from a standard set of facial expressions (Ekman & Friesen, 1976). The expressions depicted are fear, disgust, anger, happiness, sadness or surprise, which are displayed by 10 different individuals. The photographs are presented one by one and the names of the six emotions are printed below each photograph. The task is to choose the emotion name that best describes the facial expression shown. Sprengelmeyer et al. (1996) employed the same task in their study, while Gray et al. (1997) used a shorter version of this test.

2.2.2. Matching facial expression across identity

In this test, five photographs of facial expressions displayed by different individuals are fixed on a card. The faces come from the same set as those in the previous test. There are 18 cards, three for each of the six expressions: fear, disgust, anger, happiness, sadness and surprise. The task is to match the face at the top of the card with the face that has the same expression. The top face displays the same expres-

sions as one of the other faces, while the remaining faces all show different expressions.

2.2.3. Neuropsychological tests

All participants also performed a number of standard neuropsychological tests to establish the extent of the cognitive impairments in the patients and gene-carriers and to identify possible impairments in face perception that could interfere with facial expression recognition. The tests administered to every participant were the WAIS-III vocabulary subtest (Wechsler, Wycherley, Benjamin, Crawford, & Mockler, 1998) a semantic fluency test (animals), a phonemic fluency test (words beginning with F, A or S) and the short version of the Benton facial recognition test (Benton, Hamsner, Varney, & Spreen, 1983).

All participants were tested individually, either in the Department of Psychology or at home, by one of the authors (AL).

3. Results

3.1. Neuropsychological tests

Scores on the neuropsychological tests from the three subject groups are shown in Table 1. All scores are expressed as the mean number of correct responses. Performance of the three groups on each of the tests was compared with separate ANOVAs, which revealed significant group difference on all four tests ($F(2, 57) \geq 9.73$; $P < 0.0005$). Posthoc analyses with Tukey's HSD test revealed that the patients performed significantly poorer than both the healthy controls and the gene-carriers on all tests ($P < 0.01$). On the phonemic fluency test, the gene-carriers performed significantly poorer than the controls ($P < 0.01$), while performance on the Benton test, WAIS vocabulary and semantic fluency test did not differ between gene-carriers and controls. Although the patients' scores on the Benton test were below those of the controls, their overall performance was not severely impaired. In fact, the patients' mean Benton score of 40.15 (S.D. 5.02) is on the border of the normal range, and did not differ significantly from the mean Benton score ($M = 41.0$, S.D. = 5.0) of the patients in Sprengelmeyer et al.'s (1996) study ($t(31) = 0.48$; $P = 0.64$).

Table 1

Mean scores and standard deviations (S.D.) of the three participant groups on the neuropsychological tests

	Mean (S.D.)		
	Patients	Gene-carriers	Controls
WAIS-III vocabulary	33.70 (10.13) ^{a,b}	46.55 (9.37)	44 (9.75)
Phonemic fluency	16.15 (6.98) ^{a,b}	35.65 (12.17) ^a	44.8 (8.29)
Semantic fluency	12.85 (3.92) ^{a,b}	22.20 (4.29)	21.95 (3.86)
Benton	40.15 (5.02) ^{a,b}	47.25 (2.71)	49.10 (1.74)

^a Impaired relative to controls.

^b Impaired relative to gene-carriers, $P < 0.01$.

Table 2
Mean correct scores and standard deviations of the three participants group in the expression recognition and matching tests for the individual expressions

	Mean (S.D.)		
	Patients	Gene-carriers	Controls
Recognition (maximum = 10):			
Happy	9.60 (0.60)	9.90 (0.45)	9.90 (0.31)
Surprise	7.90 (2.22)	9.05 (1.15)	8.95 (1.05)
Fear	3.55 (1.67) ^{a,b}	5.70 (2.77)	7.65 (1.57)
Sadness	6.65 (1.31) ^{a,b}	8.75 (1.29)	8.45 (1.32)
Disgust	4.55 (2.82) ^{a,b}	8.40 (1.43)	8.45 (1.61)
Anger	4.45 (2.39) ^{a,b}	8.35 (1.39)	8.65 (1.27)
Matching (maximum = 3):			
Happy ^c	3.0 (0.0)	3.0 (0.0)	3.0 (0.0)
Surprise ^c	1.95 (0.83)	3.0 (0.0)	3.0 (0.0)
Fear	1.70 (0.86) ^{a,b}	2.90 (0.45)	2.85 (0.37)
Sadness	2.20 (0.77) ^a	2.65 (0.59)	2.85 (0.37)
Disgust	2.15 (0.59) ^{a,b}	2.90 (0.31)	2.90 (0.31)
Anger	1.70 (0.86) ^{a,b}	2.60 (0.50)	2.75 (0.44)

^a Impaired relative to controls.

^b Impaired relative to gene-carriers, $P < 0.01$.

^c Group performances on these expressions was not compared because of ceiling-effects.

3.2. Facial expressions

3.2.1. Recognition test

The mean number of correct responses from the three groups, separated by expression, are displayed in Table 2. Performance of the groups on the six different expressions was compared in a 3 (group) \times 6 (expression) ANOVA. This analysis revealed significant effects of group ($F(2, 57) = 48.45$; $P < 0.0005$) and type of expression ($F(5, 285) = 55.49$; $P < 0.0005$), as well as a significant interaction ($F(10, 285) = 8.62$; $P < 0.0005$). Because the gene-carriers were significantly younger than the other two groups ($P < 0.01$), we repeated this analysis in a 3 \times 6 ANCOVA using the subjects' age as covariate. This ANCOVA produced results very similar to those of the previous ANOVA; significant main effects of group ($P < 0.0005$) and expression ($P < 0.01$) and a significant interaction ($P < 0.0005$). To follow up on the results of the ANOVA, we compared the recognition scores for the six expressions in the three groups, using separate one-way ANOVAs. A Bonferroni correction was applied to control for the effect of multiple comparisons, i.e. the significance level was adjusted by the number of tests in this analysis. With six separate comparisons, the resulting alpha level was $0.05/6 = 0.008$.

These comparisons showed that the three groups did not differ in the recognition of happiness ($P = 0.07$) and surprise ($P = 0.04$), but on all other expressions there were significant differences between the groups ($F(2, 59) \geq 15.63$; $P < 0.0005$). Tukey HSD posthoc tests revealed that the patients scored significantly poorer than the controls on fear, sadness, disgust and anger ($P < 0.001$). The patients were also significantly poorer than the gene-carriers on recog-

nising fear ($P < 0.01$), sadness, disgust and anger ($P < 0.001$). Compared to the controls, gene-carriers were not impaired on any expression, although there was a trend for the gene-carriers to perform more poorly at recognising fearful faces; $P = 0.012$, critical adjusted P -value = 0.008. Pairwise comparisons within the patient group showed that recognition of fear was not significantly poorer than anger, the next most poorly recognised expression, but fear was recognised significantly poorer than disgust, $t(19) = 2.18$; $P < 0.05$. Within the gene-carriers, recognition of fear was significantly poorer than both anger and disgust ($t(19) \geq 4.15$; $P < 0.001$).

There was a high correlation between the mean correct responses on all six expressions in the gene-carriers and controls ($r = 0.90$; $P < 0.01$), and a similarly high correlation between the scores of the patients and controls ($r = 0.89$; $P < 0.01$). These correlations indicate that the patterns of relative difficulty in the patients and gene-carriers were comparable to the pattern of relative difficulty in the healthy controls; expressions that were relatively poorly recognised by the controls were also relatively poorly recognised by the patients and gene-carriers.

3.2.2. Matching expressions across identity

The mean numbers of correct responses in the matching test are also shown in Table 2. All three groups performed at ceiling for happy expressions, and both the controls and the gene-carriers were also at ceiling for surprise. Therefore, happy and surprise were not included in the analysis to compare performance of the three groups. The resulting 3 (group) \times 4 (expression) ANOVA revealed significant main effects of group ($F(2, 57) = 42.41$; $P < 0.0005$) and expression ($F(3, 171) = 3.57$; $P < 0.05$), but no group \times expression interaction. Repeating this analysis with an ANCOVA using age as covariate produced similar results, except that the main effect of expression was no longer significant. The group effect on the ANOVA was further analysed by comparing performance of the three groups on the four expressions (fear, anger, disgust and sadness) in separate one-way ANOVAs. These revealed significant group differences on all four expressions ($F(2, 59) \geq 6.22$; $P < 0.005$). Posthoc analysis with Tukey's HSD test showed that the patients performed significantly poorer than controls on all four expressions ($P < 0.004$), and significantly poorer than the gene-carriers on fear, disgust and anger ($P < 0.001$), but not sadness ($P = 0.053$). The matching scores of the gene-carriers and controls did not differ for any expression.

The results of the matching test are comparable to those in the recognition test, in that the patients were impaired compared to the healthy controls on all expressions, except happiness and surprise. These similarities in the patients' pattern of performance in the two tests suggest that their impairment in the recognition test can not simply be due to impaired understanding of the emotion labels, as the matching task requires no labelling of facial expressions.

3.2.3. A direct test for a differential deficit in disgust at the group and individual level

Although the expression recognition performance of the patients suggests no differential deficit in disgust, we directly tested this hypothesis by comparing the patients' score for disgust on the recognition test with their average score on the other emotions on which the HD sample showed a deficit (i.e. the other negative emotions: anger, fear and sadness). To perform this test we used a method described by [Strauss and Allred \(1987\)](#) and previously used to test for differential deficits in HD in another cognitive domain ([Crawford, Blackmore, Lamb, & Simpson, 2000](#)). An equally weighted composite, representing average performance on anger, fear and sadness, was formed by standardising scores on each of the three emotions using the means and S.D.s of the controls. The mean score on this composite was computed for each of the patients and controls. The point-biserial correlation between group membership (HD or control) and scores on this composite was then computed as was the point-biserial correlation between group membership and scores on disgust. These correlations are directly equivalent to running a *t*-test comparing the control and patient samples on each of these two measures. However, when group differences in performance are expressed as point-biserial correlations, it is possible to test whether the deficit on one measure significantly exceeds the deficit on another by testing whether their respective correlations are significantly different. The appropriate method is [Williams's test \(1959\)](#) for differences between non-independent correlations. The correlation between group membership and disgust scores (0.66) was lower than the correlation between group membership and the average score on the other emotions (0.84). This difference was statistically significant; $t(37) = 2.59$; $P < 0.05$. Thus the HD sample was significantly more impaired on the other emotions (anger, fear and sadness) than on disgust.

We also tested for a differential deficit in disgust recognition at the level of the individual patient. Patients with a differential deficit in disgust should exhibit significantly greater deficits on disgust compared to their average performance across the other three, impaired, emotions (the average referred to here is each individual patient's own average score). [Crawford and Garthwaite \(2002\)](#), [Crawford, Howell, and Garthwaite \(1998\)](#) developed a method to test whether there is a significant difference between an individual's scores on two tasks. In the present context, one of these tasks is the composite of the average performance on anger, fear and sadness. The method requires that the individual patient's scores are expressed as *z*-scores based on the mean and S.D. of controls. The difference between these *z*-scores is divided by the standard error of the difference in the control sample and this result is evaluated against the *t* distribution on $N - 1$ degrees of freedom ($N =$ the sample size of the control group). Application of this test for each of the 20 patients revealed that none exhibited a significantly greater deficit on disgust than on the other emotions. How-

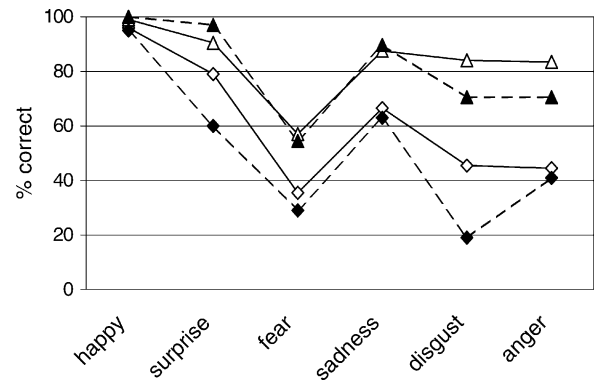


Fig. 1. Comparison between studies. Mean percentage of correct responses for each expression on the recognition test obtained by our sample of HD patients (open diamonds) and those reported by [Sprengelmeyer et al. \(1996\)](#) (filled diamonds), as well as by our sample of gene-carriers (open triangles) and [Gray et al.'s \(1997\)](#) sample of gene-carriers (filled triangles).

ever, three patients exhibited a significantly greater deficit on the other emotions ($t(19) \geq 2.22$; $P < 0.05$).¹ In sum, direct testing for a differential deficit in disgust, both at the group and individual level, failed to support the hypothesis of differentially impaired recognition of disgust in patients with HD; where significant effects were observed, their direction was the opposite to that predicted by the disgust hypothesis.

3.3. Comparison with other studies

In contrast to [Sprengelmeyer et al. \(1996, 1997b\)](#), [Gray et al. \(1997\)](#) and [Wang et al. \(2003\)](#), we found no indication of a differential deficit in the recognition of disgust in either the HD patients or the gene-carriers. To examine whether there were more differences in expression recognition between our samples and those of [Sprengelmeyer et al. \(1996\)](#) and [Gray et al. \(1997\)](#), we plotted the mean percentage of correct responses in the recognition test of the relevant groups in [Fig. 1](#). The recognition test was used in all three studies, although [Gray et al. \(1997\)](#) used a short version of this test. The HD patient groups in our study and in [Sprengelmeyer et al.'s](#) study performed very similar, except for disgust. This similarity was confirmed by the high correlation ($r = 0.93$; $P < 0.01$) between the mean correct responses to the six expressions in the two HD samples. Our sample and [Gray et al.'s](#) sample of gene-carriers also showed similar performance, except that our sample scored higher on disgust and anger. Still, the correlation between the mean correct responses on the six expression in the two gene-carrier groups was high ($r = 0.89$; $P < 0.05$). Note that [Gray et al. \(1997\)](#) originally presented their data

¹ Computer programs for PCs that carry out this test for a differential deficit for single-cases and the test for a differential deficit at the group level (diffllims.exe and diffdef.exe, respectively) can be downloaded from the following web page: <http://www.psyc.abdn.ac.uk/homedir/jcrawford/psychom.htm>.

as the proportion of individuals who obtained a particular score (ranging from 0 to 4) for each individual expression. However, when their data are presented as the mean total percentage correct for each expression, as shown in Fig. 1, recognition of fear appears poorer than recognition of disgust.

3.4. Comparison with other measures

In addition to impaired expression recognition, the patients also showed impairments on other neuropsychological tests (see Table 1). To investigate whether performance on these neuropsychological tests was related to the poor expression recognition, we performed four separate hierarchical regression analyses. The dependent variable in each case was a composite score comprised of the scores from the two facial expression tests. Individuals' scores on these two tests were converted to z -scores, using the means and standard deviations of the control group. These z -scores were subsequently summed to form the composite score. Summation was justified because the correlation between total scores on the recognition test and the matching test was high ($r = 0.88$; $P < 0.01$).

In the first of these regression analyses, scores on the Benton facial recognition test were entered into the regression model followed by group membership (i.e. patient or control). The remaining three regression analyses were identical except that the Benton was replaced, in turn, by phonemic fluency, semantic fluency and WAIS-III vocabulary. Results of the regression analysis with the Benton score as independent variable showed that performance on the Benton test did contribute to performance on the facial expression tests ($R^2 = 0.70$; $P < 0.01$), but addition of group membership significantly increased the proportion of variance explained ($R^2 = 0.78$; P for R^2 change < 0.01). Comparable results were found for the three other neuropsychological tests. Each of these variables contributed to the performance on the expression tests ($R^2 > 0.41$; $P < 0.01$), but addition of group membership always resulted in a significant increase in the amount of variance explained ($R^2 > 0.69$; P for R^2 change < 0.01). These results indicate that, although performance on the different neuropsychological tests did contribute to performance on the expression tests, impairments on these cognitive and perceptual tests could not fully account for the difference in expression recognition between patients and controls.

Finally, to test whether the expression recognition worsened with progression of the disease, we correlated the time since onset of the motor symptoms with the same expression composite score within the patient group. This correlation was not significant ($r = 0.26$; $P > 0.2$). Similarly, the correlations between time since symptoms onset and performance on the other neuropsychological measures were not significant, except for semantic fluency ($r = -0.47$; $P < 0.05$).

4. Discussion

Patients with symptoms of Huntington's disease (HD) were impaired at recognising and matching expressions of sadness, anger, disgust and fear when compared to matched healthy controls and to asymptomatic gene-carriers. There was no indication for a differential impairment at recognising disgust. Instead, the patients were more severely impaired at expressions of fear than disgust. Direct testing for a differential deficit in disgust at the group and the individual level confirmed that the patients were less impaired at disgust than on a composite score containing the other three impaired expressions (fear, anger, sadness). The performance of the asymptomatic gene-carriers did not differ significantly from that of healthy controls on any of the expressions, although there was a trend for poorer recognition of fear. Like the patients, the gene-carriers showed no sign of a differential impairment for disgust and, indeed, fear recognition was significantly poorer than disgust.

It should be noted that the controls were matched for age to the patients and not to the gene-carriers. However, the age difference appeared to have no major effect on the expression recognition performance. Ceiling effects in the matching test might have concealed differences between the gene-carriers and the controls, as both groups had maximum or near maximum scores for most expressions. Although ceiling effects may be a problem with the matching test, this was less in the case with the recognition test. On this test, all three groups had near ceiling scores only for happy expressions, but the gene-carriers still showed no significant impairments. The near-ceiling performance of all three groups for recognising happy expressions is in line with various reports of the high sensitivity for happy faces in healthy subjects (Ekman & Friesen, 1976; Feyereisen, Malet, & Martin, 1986; Kirita & Endo, 1995) and its relative invulnerability to the effects of brain damage (Adolphs et al., 1999; Broks et al., 1998; Rapsack et al., 2000; Weniger & Irle, 2002). In the light of these previous findings, we did not expect group differences for happy expressions. The HD patients and gene-carriers in Sprengelmeyer et al.'s (1996, 1997b), Gray et al.'s (1997) and Wang et al.'s (2003) studies also achieved near-perfect performance with happy faces.

Our results do not confirm the findings of Sprengelmeyer et al. (1996, 1997b), and Gray et al. (1997), concerning a differential deficit in recognising disgust, despite the fact that we used exactly the same expression recognition test as Sprengelmeyer et al. (1996), while Gray et al. (1997) used a shorter version of this test. Apart from the performance on disgust, expression recognition was very similar in our sample and Sprengelmeyer et al.'s sample of HD patients. Recognition of fear, sadness and anger was also severely impaired in the HD group studied by Sprengelmeyer et al. (1996). In other respects, the two samples of HD patients were also comparable. Sprengelmeyer et al.'s (1996) sample had about the same average age as our sample (mean 45 (S.D.

7.6) years and mean 47.6 (S.D. 8.4) years, respectively), the same estimated IQ (mean 105.8 (S.D. 7.41) and mean 105.6 (S.D. 10.7), respectively) and both groups were at roughly the same stage of the disease: mean time since onset the motor symptoms was estimated at 6.5 years (S.D. 3.2) in our sample and 6.6 years (S.D. 2.5) in the sample of [Sprenghelmeyer et al. \(1996\)](#). Direct comparison of the location and extent of the brain atrophy in the two patient groups was impossible, since we did not have atrophy data from all the HD patients in our sample. Although the prime localisation and progress of the neuropathology in HD is well documented, we can not rule out the possibility that slight variations in the areas most affected by the atrophy could account for the discrepancy in disgust recognition in the HD patients presented in this study and by [Sprenghelmeyer et al. \(1996\)](#).

With regard to the discrepancy between our study and that of [Gray et al. \(1997\)](#), an obvious difference was in the choice of control subjects. While [Gray et al. \(1997\)](#) used persons at risk of developing HD who turned out not to carry the gene mutation (AR), we compared performance of the gene-carriers to 'not at risk' healthy controls. However, our choice of control group should only have increased the likelihood of finding impairments in the gene-carriers, because [Gray et al. \(1997\)](#) found that their AR-controls scored poorer on fear, disgust and anger compared to age-matched 'not at risk', healthy subjects. [Gray et al. \(1997\)](#) suggested that the anxiety and stress associated with the genetic screening could have affected the expression recognition performance of both the gene-carriers and the non-gene-carrier controls. As previously noted, when [Gray et al.'s \(1997\)](#) data are presented as the mean percentage of correct responses across all their gene-carriers, recognition of fear is as poor, if not poorer, than recognition of disgust (see [Fig. 1](#)). Inspection of [Gray et al.'s \(1997\)](#) data in their [Fig. 1](#) reveals that there were in fact more gene-carriers who had a very low score for fear than for disgust. However, it was probably because of the relatively poor performance of the non-gene carrying controls on fear that [Gray et al. \(1997\)](#) failed to find a statistically reliable impairment in the gene-carriers. Indeed, the impairment in fear recognition in gene-carriers reported by [Gray and Barker \(2002\)](#), emerged because their non-gene carrying controls performed better on fear than the non-gene-carriers in [Gray et al. \(1997\)](#), and not because the gene-carriers in the two studies performed differently.

The results from our sample of HD patients are consistent with the findings reported by [Rapcsak et al. \(2000\)](#) and [Weniger and Irlle \(2002\)](#), who assessed recognition of facial expressions in patients with focal lesions in various locations. Both studies also found that recognition of fear was most severely impaired, while expressions of happiness were recognised best. [Rapcsak et al. \(2000\)](#) argued that the severe impairment in recognising fear was the result of differences in task difficulty, as the normal, healthy control subjects in their study also made most errors with fear. The healthy

controls in our study also had the lowest recognition scores for fear. The significant correlation between the recognition scores of the controls and the patients in our study ($r = 0.89$) further indicates that the pattern of relative difficulty and the patterns of impairment were very similar, and the impairments observed in the patients may simply represent an exaggeration of the performance pattern seen in the controls. In this context, it is interesting that there was a strong trend for the asymptomatic gene-carriers in our study to perform more poorly than the controls on fear (this result failed to achieve significance only after applying the Bonferroni correction). This may suggest that fear, being the most difficult expression to recognise, is the expression that is already affected at a very early stage of HD, and with progression of the disease the impairment in expression recognition generalises to other expressions but fear continues to be affected most severely.

Given that recognition of disgust was not impaired in the gene-carriers and not disproportionately impaired in the patients, our results provide no evidence for involvement of the basal ganglia in the recognition of disgust. More importantly, our results fail to corroborate evidence for a double dissociation between selective impairments for fear and disgust. This dissociation between poor recognition of disgust and relatively good recognition of fear has relied heavily on findings in patients and gene-carriers of HD ([Gray et al., 1997](#); [Sprenghelmeyer et al., 1996, 1997b](#)). Our results cast doubt on whether these findings can be generalised to other samples of patients or gene-carriers of HD, which means that one has to be cautious in taking patients with HD as evidence for a dissociation involving impaired recognition of disgust and spared, or at least less impaired, recognition of fear.

Finally, we do not deny that a double dissociation between fear and disgust recognition can be established. The studies in OCD patients ([Sprenghelmeyer et al., 1997a](#)), patients with Wilson's disease ([Wang et al., 2003](#)) and the patient reported by [Calder et al. \(2000\)](#) still provide evidence for selective impairments at disgust recognition in disorders other than HD. However, these impairments need not result from damage to or abnormal functioning of the basal ganglia. Obsessive-compulsive disorders are associated with metabolic abnormalities not only in the basal ganglia, but also in frontal and temporal areas ([Micallef & Blin, 2001](#)) and some studies have failed to find abnormalities in the caudate nucleus in OCD ([Aylward et al., 1996](#)). Most patients with Wilson's disease in [Wang et al.'s study \(2003\)](#) had cortical and subcortical atrophy in addition to basal ganglia abnormalities. The patient described by [Calder et al. \(2000\)](#) had lesions in the left insula as well as the basal ganglia, and it may be that the impairment in disgust recognition was related to the lesions in the insula. This would be in line with neuroimaging studies that demonstrated involvement of the anterior insula in processing facial expression of disgust ([Phillips et al., 1997, 1998](#); [Sprenghelmeyer et al., 1998](#)).

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