Brain (1996), **119**, 1647–1665

Loss of disgust
Perception of faces and emotions in Huntington's disease

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Summary

Face perception and emotion recognition were investigated in a group of people with Huntington's disease and matched controls. In conventional tasks intended to explore the perception of age, sex, unfamiliar face identity (Benton test) and gaze direction from the face, the Huntington's disease group showed a borderline impairment of gaze direction perception and were significantly impaired on unfamiliar face matching. With a separate set of tasks using computerinterpolated ('morphed') facial images, people with Huntington's disease were markedly impaired at discriminating anger from fear, but experienced less difficulty with continua varying from male to female, between familiar identities, and from happiness to sadness. In a further test of recognition of facial expressions of basic emotions from the Ekman and Friesen (1976) series, interpolated images were created for six continua that lay around the perimeter of an emotion hexagon (happiness–surprise; surprise–fear; fear–sadness; sadness–disgust; disgust–anger; anger–happiness). In deciding which emotion these morphed images were most like, people with Huntington's disease again showed deficits in the recognition of anger and fear, and an especially severe problem with disgust, which was recognized only at chance level. A follow-up study with tests of facially and vocally expressed emotions confirmed that the recognition of disgust was markedly poor for the Huntington's disease group, still being no better than chance level. Questionnaires were also used to examine self-assessed emotion, but did not show such striking problems. Taken together, these data reveal severe impairments of emotion recognition in Huntington's disease, and show that the recognition of some emotions is more impaired than others. The possibility that certain basic emotions may have dedicated neural substrates needs to be seriously considered; among these, disgust is a prime candidate.

Keywords: Huntington's disease; facial expression; emotion recognition; disgust

Introduction

Huntington's disease is a hereditary neurodegenerative disorder caused by a single gene mutation and characterized by involuntary choreiform movements, intellectual deterioration (Butters *et al.*, 1978; Caine *et al.*, 1978; Brandt and Butters, 1986; Sprengelmeyer *et al.*, 1995*a*) and attentional deficits (Georgiou *et al.*, 1995; Sprengelmeyer *et al.*, 1995*b*). Affective disturbances, emotional problems and deficits in visual and auditory perception of social stimuli have also been noted, with impairments in comprehension of prosody even in the early stages of the disorder (Speedie *et al.*, 1990) and impaired recognition of emotional facial expressions and poor performance on the Benton Test of Facial Recognition (Jacobs *et al.*, 1995*b*).

As described to date, the reported deficits in facial expression recognition might easily be explained as consequences of more basic visual problems, since there is evidence from different sources that visuo-perceptual information processing is disturbed in people with Huntington's disease. Lange (1981) reported cortical atrophy of up to 30% of volume in areas associated with vision in the final stages of Huntington's disease, and a reduction in the amplitude of the early components of visual evoked potentials has been reported by several authors (Ellenberger *et al.*, 1978; Oepen *et al.*, 1981; Josiassen *et al.*, 1984; Hennerici *et al.*, 1985). People with Huntington's disease are also impaired on a range of neuropsychological tests assessing

Table 1 *Clinical characteristics of patients with Huntington's disease in this study*

Number	Sex	Age (years)	Disease (years)	Chorea (0–3)	Disability (0–3)	IQ[†]	Atrophy on CT scan (0–3):			CAG: % No. of repeats	Medication[‡]	Contrast sensitivity
							Caudate	Frontal	Posterior			
01	F	46	6	1.0	1.00	94	1	1.0	1.5	42	–	Normal
02*	M	42	8	0.5	1.00	95	3	1.5	1.0	44	S	Normal
03*	M	46	5	1.0	0.50	100	1	1.0	0.5	46	H	Normal
04	F	60	3	1.0	0.50	100	1	0.5	0.5	–	L	Normal
05	F	46	8	2.0	1.42	95	2	2.0	2.0	41	T	Normal
06	F	45	5	1.0	0.83	104	2	0.5	1.0	45	M	Normal
07	M	39	7	1.0	1.50	104	–	–	–	51	S	Normal
08	F	46	3	1.5	0.25	110[W]	1	0.5	1.0	42	M	Normal
09	M	30	10	1.0	1.00	124	1	0.5	1.0	57	–	Normal
10	F	58	5	2.0	1.33	100[W]	3	1.0	1.0	–	–	Normal
11	M	41	6	0.5	0.58	120[W]	1	1.0	0.0	43	H,F	Normal
12	M	44	10	2.0	0.92	124[W]	2	0.5	1.0	41	–	Normal
13	F	42	10	1.5	1.50	103	2	1.0	1.0	45	–	Normal

Disability was measured with the modified German version of the Shoulson and Fahn scale (Lange *et al.*, 1983). Contrast sensitivity was measured with the Vistech VCTS 6000 test; all subjects were performing within the normal range for all spatial frequencies. *Patients not included in the follow-up part of the study (Bogart–Grant continuum, 60 Ekman and Friesen series faces, auditory emotion recognition, emotion questionnaires); [†]IQ was measured by WAIS ([W]) or by the MWT-B vocabulary recognition test (Lehrl, 1977); [‡]medication: H = haloperidol, S = sulpiride, T = tiapride, M = memantine, F = flunitrazepam, L = Levomepromazin.

visuo-perceptual and visuo-spatial functioning (Moses *et al.*, 1981; Brouwers *et al.*, 1984; Jason *et al.*, 1988; Mohr *et al.*, 1991; Sprengelmeyer *et al.*, 1992).

In the light of these results, one might suppose that further research on facial expression recognition in people with Huntington's disease should reveal unspecific deficits compromising the recognition of all emotions.

There is, however, another possible line of argument. Recent studies of the consequences of bilateral amygdala damage have shown differentially severe impairment of the recognition of facial expressions of fear, leading to the hypothesis that there may be differences between the neural substrates of different emotions (Adolphs *et al.*, 1994, 1995; Calder *et al.*, 1966*b*). This is important because morphometrical post-mortem studies on Huntington brains (Lange and Aulich, 1986) revealed tissue loss of 25% in the amygdala and related structures. From this point of view, more specific deficits in facial expression recognition, focused on particular emotions, might be expected in Huntington's disease.

The present study therefore looks in detail at face perception in Huntington's disease and at the recognition of emotion. Because a persistent finding in the neuropsychological literature has been that brain injury can produce circumscribed deficits affecting only certain aspects of face perception (Parry *et al.*, 1991; Young and Bruce, 1991; Young, 1992), we used tasks intended to explore the perception of age, sex, unfamiliar face identity, familiar face identity, gaze direction and emotional expression from the face. These tasks were chosen to reflect abilities which might be differentially susceptible to impairment either on the basis of existing studies of the effects of lesions of the cerebral cortex (Campbell *et al.*, 1990; Young *et al.*, 1993) or in terms of theoretical models (Bruce and Young, 1986; Ellis, 1986). In addition, by using computer-interpolated (morphed) facial images in some tests, we were able to examine sensitivity to small changes in the cues conveying different types of social signal.

A particular focus of our interest was in the recognition of emotion. We sought to determine whether Huntington's disease compromises the recognition of all facial expressions of emotion, or has a particular impact on certain emotions. To this end, we used tasks which would be sufficiently sensitive to detect deficits affecting particular basic emotions identified by Ekman and his colleagues (Ekman, 1972, 1992; Ekman *et al.*, 1972). These emotions each have a distinctive facial expression, recognizable in most cultures of the world.

Since impairments in comprehension of prosody also occur in Huntington's disease (Speedie *et al.*, 1990), a test of auditory emotion recognition was included together with a further test of facial expression recognition in a follow-up study, to look for possible relations between deficits affecting visually and auditorily communicated emotion. In this way, we sought to determine whether any deficits in emotion recognition might reflect problems specific to the recognition of emotion from the face, or a more general deficit affecting the recognition of emotion from other sense modalities as well. In addition, we used questionnaires to examine self-assessed emotion.

Method

Subjects

Thirteen people (seven female, six male) with a clinically and genetically definite diagnosis of Huntington's disease gave their informed consent to take part in the study. The mean age of the participants in the Huntington's disease group was 45.0 years (SD 7.6 years), and mean duration of

schooling was 9.0 years (SD 1.7 years). Participants in the Huntington's disease group were not demented. Their mean IQ was 105.6 (SD 10.7) using the WAIS or the MWT-B German vocabulary recognition test (Lehrl, 1977). The mean degree of disability (range 0– 3) as measured by a modified German version of the Shoulson and Fahn (1979) scale (Lange *et al.*, 1983) was 0.95 (SD 0.41).

A clinical rating scale (range 0–3) was used to determine the severity of choreic movements (Lange *et al.*, 1983). The mean degree of motor disturbances in the Huntington's disease group was 1.23 (SD 0.53). The duration of motor disturbances estimated by study of case histories and by interviews with reliable informants varied between 3 and 10 years, with a mean of 6.6 years (SD 2.5 years). Descriptive data are given in Table 1.

Twelve of the 13 patients were given CT scans which were evaluated by two experienced neurologists not informed about the neuropsychological data. Cortical atrophy was determined by measuring the width of sulci in the frontal and posterior regions. The following scoring system was applied. A score of 0 was given for sulci barely visible, and a score of 1 for sulci smaller than 2 mm. Sulci from 2 to 5 mm width were scored as 2, and 3 was given for sulci >5 mm. The two raters worked independently and their scores were averaged, resulting in intermediate scores, when they diverged by 1. Atrophy of the caudate nucleus was defined as the CC:IT ratio multiplied by 100, where CC was the distance between the left and right heads of the caudate nuclei and IT the distance across the inner table of the skull, both measured at the level of the interventricular foramen. A ratio below 12 was scored as 0 (normal); a ratio from 12 to 16 as 1 (mild subcortical atrophy); a ratio from 16 to 20 was scored as 2 (moderate subcortical atrophy) and a ratio above 20 was scored as 3 (severe subcortical atrophy).

Genetic testing was performed on 11 of the 13 patients using standard procedures (Riess *et al.*, 1993). The CAG nucleotide sequence repetition rate was 45.2 (SD 4.9). Table 1 also gives details of medication, which varied from person to person.

The control group consisted of 17 healthy adults (eight female, nine male) free of neurological and psychiatric disorders. The mean age of the controls was 50.7 years (SD 14.3) and mean duration of schooling was 9.8 years (SD 2.4). The mean IQ of the control group was 107.5 (SD 10.0). Student's t tests showed no significant differences between the control and Huntington's disease groups with respect to age ($t = 1.29$, $P = 0.18$), years of formal education ($t = 1.02$, $P = 0.32$) or intelligence ($t = 0.50$, $P = 0.62$).

All testing was carried out by a native speaker of German, and German equivalents of English emotion names used were as follows: happiness–Glück; surprise–Erstaunen; fear–Angst; sadness–Trauer; disgust–Ekel; anger–Wut.

Because of the particular focus of our study on face perception and on the recognition of emotion, we were also anxious to establish that people with Huntington's disease who participated in the study would not fail such tasks because they were unable to understand the meanings of verbal emotion terms used for responses, or alternatively because of primary visual deficits. All were therefore tested for their comprehension of emotion terms, and proved able to understand the labels used. They could state what it means to say someone is happy, surprised, afraid, sad, disgusted, or angry, and could give plausible examples of circumstances under which people might experience such emotions.

To ensure that any differences between the Huntington's disease and control groups were not due to poor vision *per se*, all participants were given the Vistech VCTS 6000 contrast sensitivity chart. This measures the degree of contrast at which stationary sinusoidal gratings can be detected at each of five spatial frequencies (1.5, 3.0, 6.0, 12.0, 18.0 cycles per degree). It therefore gives useful measures of ability to see subtle changes in light and shade in static grey-scale displays; this is exactly appropriate to our needs, since our face perception tests were based on black and white photographic images.

As Table 1 shows, the performance of the VCTS 6000 test by all of the participants with Huntington's disease was within normal limits for all spatial frequencies; none had any measurable impairment of basic visual function. Furthermore, comparisons between the Huntington's disease and control groups revealed no significant differences in contrast sensitivity at 1.5 cycles per degree [$F(1,28) = 1.72$, $P > 0.1$]; 3.0 cycles per degree ($F < 1$); 6.0 cycles per degree ($F < 1$); or 12.0 cycles per degree ($F < 1$). There was a significant group difference at 18.0 cycles per degree [$F(1,28) = 4.25$, $P < 0.05$], but in the direction of the people with Huntington's disease out-performing controls! Since the more widely used (but less informative) acuity measure corresponds to the highest spatial frequency that can be seen at high contrast, we can infer that it is likely that the participants in the Huntington's disease group had, if anything, the more keen eyesight.

Face perception tests

For the first part of our study, we used a set of conventional tasks intended to explore the perception of a range of social information other than emotion from the face. These covered the perception of age, sex, unfamiliar face identity and gaze direction; they were considered to provide useful background information. These conventional tests were supplemented by a set of psychophysical tests using facial images which were morphed to different degrees. The tests with morphed images investigated the perception of sex, familiar face identity and emotion. Each test will be described in turn.

Conventional tasks

Separate tests examined perception of age, sex, unfamiliar face identity and gaze direction.

Age. Photographs of the faces of 10 young women, 10

young men, 10 old women and 10 old men were arranged into a fixed pseudo-random order. The faces were chosen to have a variety of hairstyles and hair-lengths, but to exclude bald people, and none wore beards, moustaches, glasses or earrings. Hence, all reliable cues to the person's age or sex had to come from the face's appearance. Eight additional faces were used for practice. The task was to work through these faces one by one, classifying each as of 'young' or 'old' appearance. The test was taken from a previous study by Young and Ellis (1989).

Sex. The photographs used for the age perception test were also used as a sex classification task. The person acting as subject was asked to work through the faces one by one, classifying each as of 'male' or 'female' appearance (Young and Ellis, 1989).

Unfamiliar face identity. The Benton Test of Facial Recognition (Benton et al., 1983) was given. In this test, subjects have to choose which of six photographs of unfamiliar faces are pictures of the same person as a simultaneously presented target face photograph. The test includes items involving choice of identical photographs, as well as transformations of orientation or lighting, which are pooled to give an overall total score.

Gaze direction. A forced-choice task was used to assess ability to determine eye gaze direction (Young et al., 1995). Pairs of photographs of the same person's face were presented alongside each other. For one-third of the pairs (six trials) both pictures were full-face photographs, for one-third (six trials) the heads were facing 20° to the left, and for the remaining third of pairs (six trials) both heads were facing 20° to the right. In each pair, the eyes of the target face were oriented directly toward the viewer, and the non-target face was looking away to the left or right by 5, 10 or 20°. The combination of six directions of gaze for the non-target faces (left and right direction of gaze at 5, 10 and 20°) and three possible head orientations for both members of each pair (full-face, 20° left or 20° right) produced a total of 18 trials. On each of these, the person acting as subject was asked to choose the photograph in which the face was looking directly toward them.

Perception of morphed facial images

Procedures in general. Morphing refers to computer graphics procedures used to interpolate images along a continuum between two prototypes (see Figs 1 and 2). These techniques were first used to investigate emotion recognition by Etcoff and Magee (1992) and to investigate the recognition of identity by Beale and Keil (1995). With appropriate programs, such interpolations can be made to photographic quality. The algorithms used here were initially developed by Benson and Perrett (1991), but have been extended and improved; more detail can be found elsewhere (Calder *et al.*, 1996). The basic technique involves locating the positions of a large number of specified feature points on each of two prototype images. The shape of one prototype image can then be moved toward the shape of the other prototype by calculating the vector difference for each landmark and using this to obtain positions for that point which take it along a straight line from its location in one prototype to its corresponding location in the other prototype. A continuous-tone (photographic quality) image can be created for each of these interpolated face shapes by 'stretching' both of the prototype images (as if they were printed on a rubber sheet) to this new shape, so that all points representing the same feature are aligned across images, and blending them with the appropriate weight.

As Figs 1 and 2 show, remarkably smooth transitions can be achieved with appropriate algorithms. The interest of the technique for present purposes is that it manipulates the information which the visual system is also using (otherwise morphing would not work at all) whilst simultaneously allowing us to introduce differences between images which can be as small as we wish, and therefore opening the possibility of creating tasks whose sensitivity can be adjusted to any required level.

For the present study, we used morphed images to examine perception of sex, familiar identity and emotional expression in face images.

The procedure for sex continuum, the familiar faces continuum, and the sadness–happiness and fear–anger emotion continua (*see* Fig. 1) is given below. The images shown in Fig 1 were presented on a computer screen, one at a time, for 5 s each in pseudo-random order. The subject's task was to decide whether each image was more male or more female in appearance (or whether each image was more like Humphrey Bogart or Cary Grant in appearance, whether the expression on each face was more like happiness or sadness, or whether it was more like anger or fear) with a verbal response which was recorded by the experimenter. There were six blocks of trials, with each of the five images used once in each block in different pseudo-random orders. The first block was discounted as practice and data from the remaining five blocks (25 trials) were used for analysis.

Sex continuum. Computer graphics techniques (Benson and Perrett, 1991; Rowland and Perrett, 1995) were used to create images of an average young male and an average young female. The manufacture of these and similar images has been described previously in studies of perception of facial gender (Brown and Perrett, 1993), attractiveness (Perrett *et al.*, 1994) and age (Burt and Perrett, 1995). Briefly, images were made by locating the positions of 224 specified feature points in 60 photographs of Caucasian males (aged 20–30 years) and 60 Caucasian females (aged 20–30 years). An average male face shape was calculated by averaging the corresponding feature positions across the set of 60 male faces, and an average female face shape was made similarly

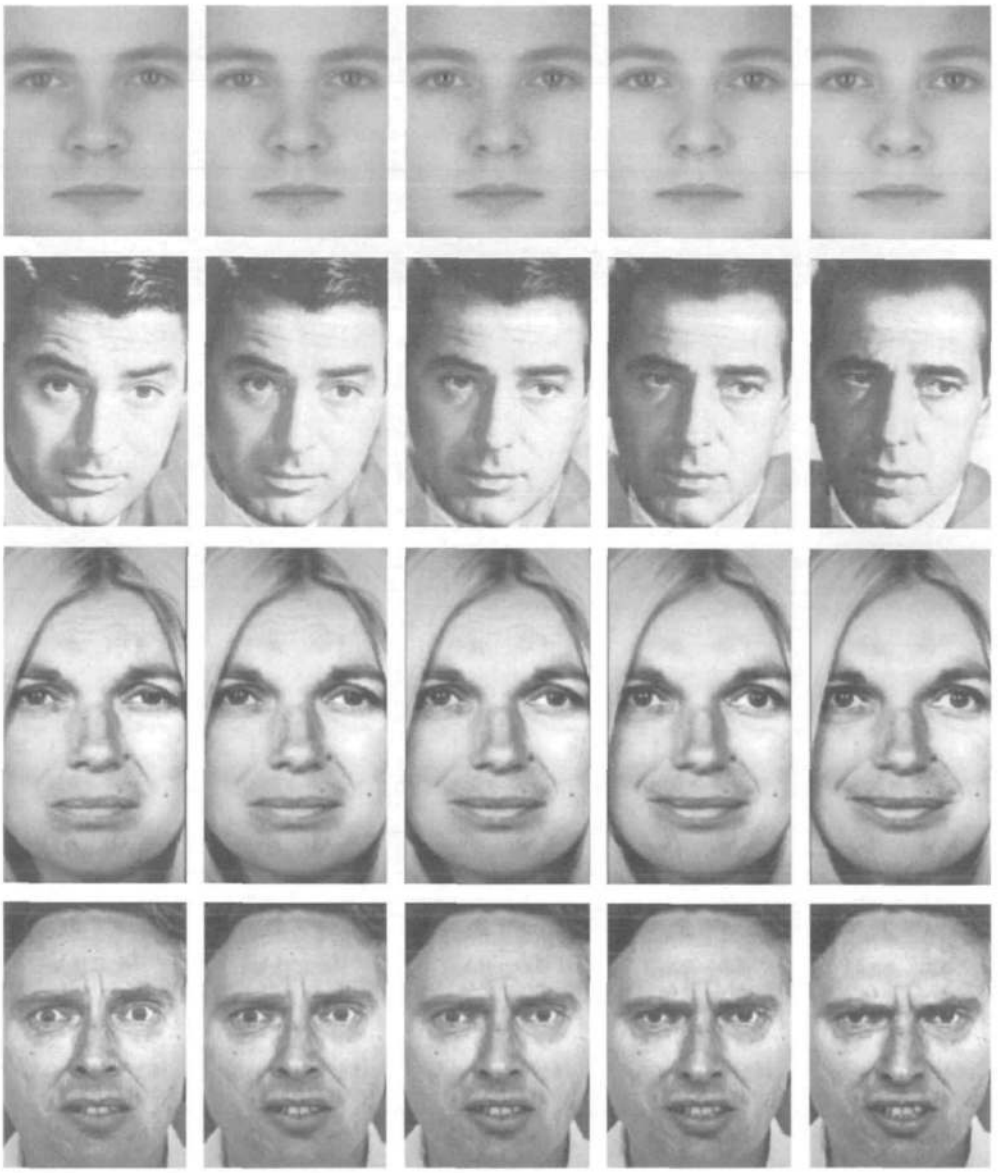

Fig. 1 Morphed images used to investigate perception of sex (male–female continuum), familiar face identity (Cary Grant–Humphrey Bogart continuum) and emotion (sadness–happiness and fear–anger continua).

from the set of female faces. Male and female prototype face images were created with even surface pigmentation by warping each of the 60 faces of the same gender to the appropriate average shape and then blending the 60 reshaped images. The shape of each prototype was then shifted 10, 30, 50, 70 and 90% toward the other prototype, to create a series of images running from one with predominantly male shape (90% of the average male shape) to one with predominantly female shape (90% of the average female shape). After being warped into the intermediate shapes, the two prototypes were then blended with the appropriate weight (e.g. 90% male pigmentation and 10% female pigmentation for the 90% male shape). The resulting images are shown in Fig. 1 (*see* first row).

Familiar faces continuum. Morphing techniques (Benson and Perrett, 1991) were used to create interpolated images between two different familiar faces—Humphrey Bogart and Cary Grant. This was done by locating the positions of 224 specified feature points in a photograph of Humphrey Bogart and a photograph of Cary Grant and using these feature points to divide each image into a mesh of triangular tessellations. These images were then blended by shifting the locations of each feature point and mixing the textures for each tessellation 10, 30, 50, 70 and 90% toward the other prototype, to create a series of images running from one which looked most like Humphrey Bogart (90% of his shape and surface pigmentation) to one which looked most like Cary Grant (90% of his shape and surface pigmentation).

This familiar face identity continuum (*see* second row in Fig. 1) was not ready when the initial study was carried out, and had to be included as part of the follow-up (*see* below).

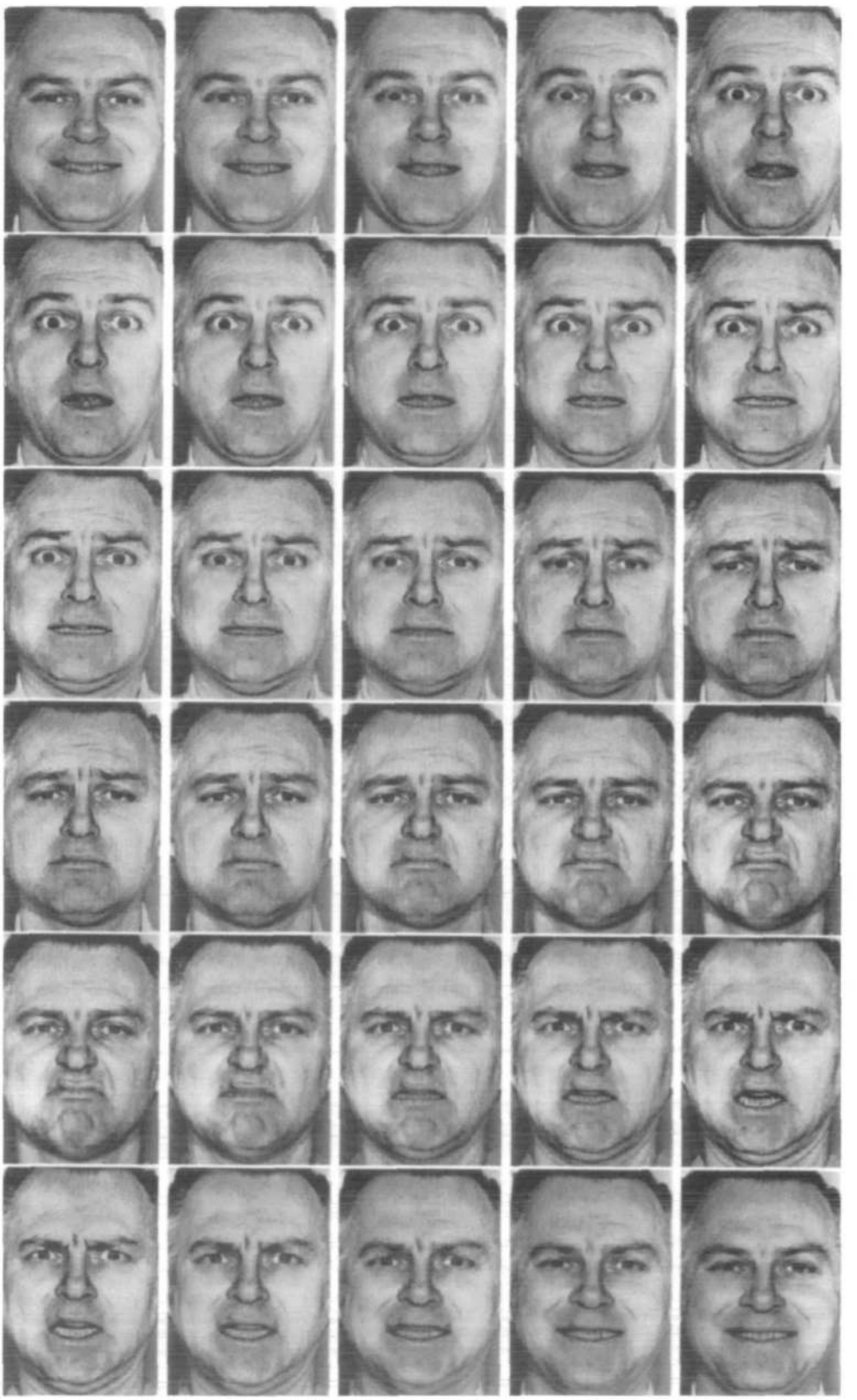

Fig. 2 Expression continua used in emotion hexagon experiment. Going from left to right, the columns show 90, 70, 50, 30 and 10% morphs along each continuum. From top to bottom, the continua shown in each row are happiness–surprise (top row); surprise–fear (second row); fear–sadness (third row); sadness–disgust (fourth row); disgust–anger (fifth row) and anger–happiness (bottom row).

Therefore, data are only available for 11 of the 13 people with Huntington's disease (*see* Table 1).

Emotion continua. To explore the perception of emotion with an analogous procedure to that for sex and identity, we used a continuum of morphed images ranging between sadness and happiness, and another continuum ranging between fear and anger. For each continuum, we selected prototype images where the two photographs of the poser were as similar as possible in general quality (i.e. same head positioning, same lighting, etc.). Furthermore, we opted for pairs of photographs which would not have gross changes in physical features, and especially from open to closed mouths. An additional constraint was that we avoided posers with pronounced upturning of the corners of the mouth in the prototype happiness image, or downturning in sadness, so that the discrimination of happiness from sadness would involve more than this cue alone. The sadness–happiness continuum was created using face PF from the Ekman and Friesen (1976) series and fear–anger with face EM. Mean emotion recognition scores for Ekman and Friesen's (1976) normal subjects were as follows: PF happiness 100%, PF sadness 100%, EM anger 83%, EM fear 92%. However, these scores relate to a much more difficult six-way choice as happiness, sadness, fear, anger, surprise or disgust, not to the simpler two-way choices used here.

As in the sex and identity tasks, the stimuli involved 10, 30, 50, 70 and 90% morphs along each continuum; these are shown in Fig 1. Trials with the sadness–happiness and fear–anger continua were given as separate tasks.

Identification of morphed facial expression: an emotion hexagon. As a further test of facial expression recognition with morphed images, which would enable us to examine all six basic emotions from the Ekman and Friesen series, we used what we will call an emotion hexagon. On the basis of Ekman and Friesen's (1976) norms, we ordered their six different emotional facial expressions by their maximum potential to confuse, i.e. placing each adjacent to the one it was most likely to be confused with. The result ran happiness–surprise–fear–sadness–disgust–anger, with mean percentage confusion, in normal subjects, for each pair of expressions in this sequence being happiness and surprise 0.8% (i.e. on 8% of trials with happy and surprised targets the subjects tested by Ekman and Friesen misidentified a happy face as surprised, or a surprised face as happy), surprise and fear 5.8%, fear and sadness 2.4%, sadness and disgust 2.7%, disgust and anger 6.4%. The ends of the sequence (anger and happiness) were then joined to create a hexagon, and five interpolated ('morphed') images were created for each of the six continua lying around the perimeter of this hexagon (happiness–surprise; surprise–fear; fear–sadness; sadness–disgust; disgust–anger; anger–happiness).

Face JJ (Ekman and Friesen, 1976) was chosen for morphing because the photographs of all six emotional facial expressions (happiness, surprise, fear, sadness, disgust and anger) were of consistent quality, with reasonably standardized pose and lighting. Photographic-quality continua were made, with five morphed images for each continuum; these were prepared by blending between two prototype expressions posed by JJ (e.g. happiness and surprise) in proportions 90:10 (i.e. 90% happy, 10% surprised for the happiness–surprise continuum), 70:30 (70% happy, 30% surprised), 50:50 (50% happy, 50% surprised), 30:70 (30% happy, 70% surprised) and 10:90 (10% happy, 90% surprised). The morphing technique was the same as that outlined above; it is described in detail elsewhere (Calder *et al.*, 1966*a*).

The resulting morphed faces are shown in Fig. 2. In total, there are 30 images (five from each of six continua). Moving from left to right in Fig. 2, the columns show 90, 70, 50, 30 and 10% morphs along each continuum. Note that a 90% morph on the happiness–surprise continuum would be the same as a 10% morph on a surprise–happiness continuum, and that the prototype expressions are not shown in Fig. 2. Moving from top to bottom of Fig. 2, the rows show the happiness–surprise continuum (top row); surprise–fear (second row); fear–sadness (third row); sadness–disgust (fourth row); disgust–anger (fifth row) and anger–happiness (bottom row).

The 30 morphed faces shown in Fig. 2 were used in an identification task. They were presented one at a time on a computer screen, in pseudo-random order. The subject's task was to decide whether the morphed image was most like happiness, surprise, fear, sadness, disgust or anger. Responses were made verbally, with the names of the six possible emotions being printed on a card which the subject was free to consult throughout the test. On each trial, the morphed image remained in view for 5 s, after which it was replaced by a question mark. No feedback was given as to the appropriateness of any responses.

There were six blocks of trials. In each of these blocks of trials, all of the 30 morphed faces were presented once. The first block was discounted as practice and data from the remaining five blocks (150 trials) were used for analysis. Note that the prototype face for each expression was never shown in the experiment; all the stimuli were morphs. Note too that the morphs were presented in pseudo-random order; they were not grouped into the underlying continua.

Follow-up study of emotion

Because of the interesting findings concerning the recognition of facial expressions of emotion which emerged from the study as described above, a follow-up study of emotion recognition was carried out some 3 months later. Eleven of the original 13 people with Huntington's disease and all controls were available for retesting (*see* Table 1).

In this follow-up, we presented two additional tests; one was designed to explore recognition of the six emotions from the Ekman and Friesen (1976) series of facial expressions, and the other one was an equivalent test of recognition of

emotion from the voice. Questionnaires relating to emotions of anger, fear and disgust were also given.

Recognition of specific emotions in facial expressions. Photographs of the faces of 10 people (six female, four male) were taken from the Ekman and Friesen (1976) series. For each face, there were poses corresponding to each of six emotions (happiness, surprise, fear, sadness, disgust and anger), giving a total of 60 photographs. These were shown one at a time in pseudo-random order, for 3 s each, and the person was asked to decide which of the emotion names (happiness, surprise, fear, sadness, disgust or anger) best described the facial expression shown. The names of the six emotions were printed on a card, and this was available throughout the test. There were 60 trials (one for each of the six emotions across the 10 posers), leading to an accuracy score out of a possible maximum of 10 for each of the six emotions.

Recognition of vocal expressions of emotion. In this test meaningless words (e.g. 'silzukankunkrei', 'hontraruru' or 'miromente') from Christian Morgenstern's poem 'Das groβe Lalula' were chosen to create a set of 10 different nonsense 'sentences'. The sentences were spoken by an actor with a happy, surprised, fearful, sad, disgusted or angry vocal intonation, to create a total of 60 stimuli. All sentences and expressions were recorded on tape and then arranged in a fixed pseudo-random order. Subjects had to listen to the taped sentences and decide which of the emotion names (happiness, surprise, fear, sadness, disgust or anger) best described the vocal intonation expressed by the actor. The names of the six emotions were printed on a card and this was available throughout the test. The use of 60 sentences allowed an accuracy score out of a possible maximum of 10 for each of the six emotions.

Emotion questionnaires. Standard questionnaires were used to examine self-assessed emotion for anger, fear and disgust. The anger scale (Novaco, 1975) asks the subject to rate situations on a five-point scale for the extent to which they would make you angry. We used the first 40 items from this 80-item questionnaire. The (Wolpe and Lang, 1964) fear schedule also uses a five-point scale for rating things and experiences that may cause fear or other unpleasant feelings; we used all 75 items and sub-items. The disgust scale (Haidt *et al.*, 1994) has 32 items, all of which were used. These are grouped into eight different domains, with four items for each domain. Two of these items require 'true' (disgusting) or 'false' (not disgusting) answers, the other two are rated on a three-point scale of disgustingness. Seven of the domains cover different forms of disgust provocation: food, animals, body products, sex, body envelope violations, death and hygiene. The eighth domain encompasses magical thinking via similarity and contagion.

Table 2 *Conventional face processing tasks: performances of Huntington's disease patients (HD) and control subjects*

	HD		Controls	
	Mean	(SD)	Mean	(SD)
Age perception (max. = 40)	39.1	(0.8)	39.3	(1.0)
Sex perception (max. = 40)	39.7	(0.5)	39.8	(0.4)
Unfamiliar face matching				
Benton test (max. = 54)	41.0*	(5.0)	47.9	(3.6)
Gaze direction (max. = 18)	14.3	(3.4)	16.9	(1.1)

*$P < 0.001$ (significantly different from control performance).

Results

We will describe analyses of the various tasks in the order they occur in the method section of this report. We have used entirely non-parametric analyses because of the non-homogeneity of the variances resulting from the deliberate use of tests with widely differing ceilings and floors.

Conventional tests of social information other than emotion

Table 2 summarizes results involving the performance of conventional face processing tasks by people with Huntington's disease and age-matched control subjects. We used Mann–Whitney U tests to compare performance across groups on each task.

For the age and sex perception tasks both the Huntington's disease and control groups performed at high levels, with no significant between-group differences (for age $U = 89$, $Z = 0.90$, $P > 0.1$; for sex $U = 102.5$, $Z = 0.34$, $P > 0.1$). Although the presence of a clear ceiling effect in these two tasks leaves us unable to be certain that the performance of the Huntington's disease group was entirely unimpaired, the tests are sufficiently sensitive to pick up impairments in cases of prosopagnosia (Young and Ellis, 1989). We can therefore be confident that gross impairments were not present in the people with Huntington's disease.

Perception of the identities of unfamiliar faces was assessed with the Benton Test of Facial Recognition (Benton *et al.*, 1983). Results showed that the Huntington's disease group performed less well than controls ($U = 28.5$, $Z = 3.43$, $P < 0.001$). Note, however, that the performance of the Huntington's disease group was well above the chance level of two out of 54 correct, and in fact the mean score of people with Huntington's disease fell at the lower end of the range regarded as normal performance by the originators of this test (Benton *et al.*, 1983).

Perception of gaze direction was tested with a forced choice procedure, involving picking the face which is looking directly at you. On this task, the performance of the Huntington's disease group fell in a borderline region ($U = 66$, $Z = 1.86$, $0.1 > P > 0.05$) which was well above the chance level of nine out of 18 correct.

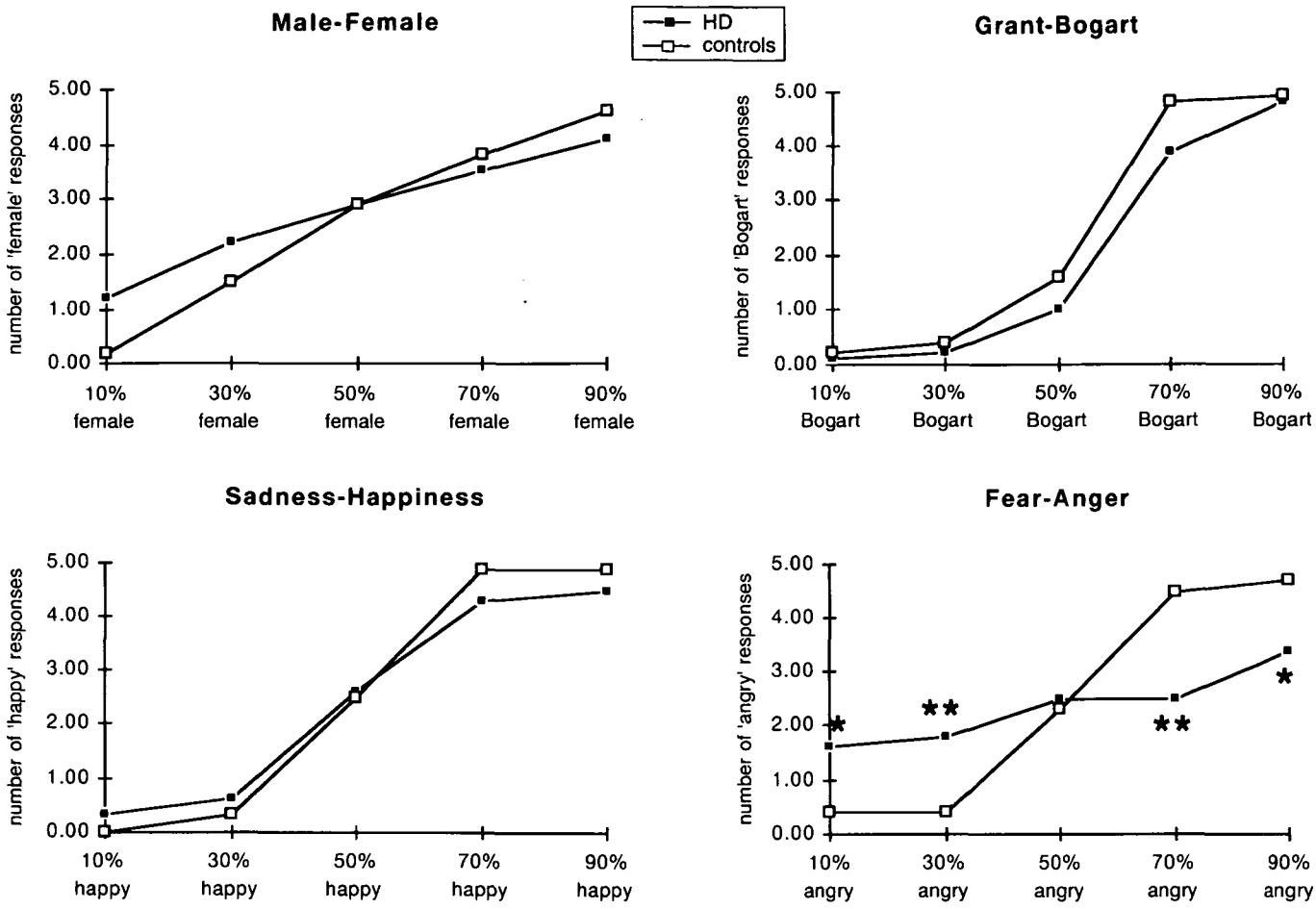

Fig. 3 Identification of morphed images in Fig. 1 by people with Huntington's disease (HD) compared with that by control subjects. The images fell along continua showing transformations of sex (male–female), familiar identity (Cary Grant–Humphrey Bogart) and emotion (happiness–sadness and anger–fear). $*P < 0.05$, $**P < 0.01$.

Perception of morphed facial images

Figure 3 summarizes data from the Huntington's disease and control groups for the male–female continuum, familiar identity continuum, and the sadness–happiness and fear–anger emotion continua.

In these tests with morphed continua, our aim was to examine perception of images interpolated between prototypes that are readily identifiable by normal controls. The consequence of this is that control performance is (intentionally) at ceiling and floor at the ends of each continuum. Hence, the variance of control scores tends always to increase toward the centre of each continuum, violating assumptions of homogeneity of variance which underlie techniques like ANOVA. For this reason, we have used non-parametric Mann–Whitney U tests to compare Huntington's disease and control scores at each point along each continuum.

For the morphed sex perception task, there were no significant differences between Huntington's disease and control performance (10% female, $U = 64.5$, $Z = 1.92$, $0.1 > P > 0.05$; 30% female, $U = 88.5$, $Z = 0.92$, $P > 0.1$; 50% female, $U = 104.5$, $Z = 0.25$, $P > 0.1$; 70% female, $U = 100$, $Z = 0.44$, $P > 0.1$; 90% female, $U = 90.5$, $Z = 0.84$, $P > 0.1$).

To investigate perception of the identities of familiar faces, we used another task with interpolated images. Again, there were no significant differences between Huntington's disease and control performance (10% Bogart, $U = 75.5$, $Z = 0.05$, $P > 0.1$; 30% Bogart, $U = 60.5$, $Z = 0.86$, $P > 0.1$; 50% Bogart, $U = 60$, $Z = 0.89$, $P > 0.1$; 70% Bogart, $U = 43$, $Z = 1.81$, $0.1 > P > 0.05$; 90% Bogart, $U = 72$, $Z = 0.00$, $P > 0.1$).

Two emotion continua were also used. For sadness–happiness, there were no significant differences between Huntington's disease and control performance (10% happy, $U = 85$, $Z = 1.01$, $P > 0.1$; 30% happy, $U = 70$, $Z = 1.70$, $0.1 > P > 0.05$; 50% happy, $U = 107.5$, $Z = 0.13$, $P > 0.1$; 70% happy, $U = 87.5$, $Z = 0.96$, $P > 0.1$; 90% happy, $U = 91$, $Z = 0.82$, $P > 0.1$). However, for the fear–anger continuum, the groups differed significantly at every point except the central 50% image, where the two functions cross (10% angry, $U = 62$, $Z = 2.03$, $P < 0.05$; 30% angry, $U = 33$, $Z = 3.24$, $P < 0.01$; 50% angry, $U = 103$, $Z = 0.31$, $P > 0.1$; 70% angry, $U = 42$, $Z = 2.87$, $P < 0.01$; 90% angry, $U = 60$, $Z = 2.11$, $P < 0.05$).

In summary, the only significant differences between

Huntington's disease and control performance for the continua shown in Fig. 3 were found in the anger–fear task, where people with Huntington's disease were much poorer at discriminating these emotions than were controls.

Although it is clear that the Huntington's disease group showed the most severe problems with the fear–anger continuum, we do not seek to claim that performance of the other continua was entirely normal. Inspection of Fig. 3 suggests that the slopes of the sex, identity and happiness–sadness functions were consistently slightly less steep in the Huntington's disease group, and our analyses showed occasional borderline differences for these continua which would have reached significance on one-tailed tests.

Our second way of exploring perception of morphed facial expressions involved the emotion hexagon (Fig. 2). Data for identification of morphed facial expressions from the emotion hexagon are presented in Fig. 4, which gives mean identification rates for the morphed images shown in Fig. 2 by people with Huntington's disease and control subjects. The morphed images shown in Fig. 2 (continuous sequence left to right and top to bottom) are placed along the horizontal axis of each chart in Fig. 4, i.e. running from 90% happiness, 10% surprise (the top left image in Fig. 2 labelled with the convention HA9-SU1 in Fig. 4) to 10% anger, 90% happiness (the bottom right image in Fig. 2 labelled AN1-HA9 in Fig. 4 – *see* the figure legend for details of code). Each chart in Fig. 4 represents a particular emotion; the vertical axis shows the mean number of times each image was identified as that emotion. The mean performance of people with Huntington's disease is compared with the appropriate control performance.

The extent to which the performance of people with Huntington's disease matched that of controls was assessed in two ways. First, the entire hexagonal continuum was divided into six sections corresponding to regions containing morphs that the controls consistently identified with one of the six emotion labels (happiness, surprise, fear, sadness, disgust and anger). We found that each of these regions comprised four morphs, two of these contained 90% of the target expression and the other two 70%. For example, the surprise section contained the morphs 70% surprised, 30% happy; 90% surprised, 10% happy; 90% surprised, 10% afraid and 70% surprised, 30% afraid. Secondly, we calculated the mean number of times, all of the morphs in each region were identified with the appropriate emotion label; the maximum possible score would be 20 for each region (four trials with five morphs), yielding a total out of 120 across all six regions. These values were compared for people with Huntington's disease and control subjects. As a rough guide, this measure of relative performance is shown in Fig. 4 (the patients' performance as a percentage of the mean number of times the controls gave the appropriate response for each of the six emotion regions). Values <100% indicate that people with Huntington's disease correctly identified fewer of the morphed expressions in this region than did the controls.

To deal with these data statistically, we used non-parametric Mann–Whitney U tests to compare Huntington's disease and control scores. Across the six defined regions taken together, there was a highly significant group difference; the controls' mean total number of correct responses was 106.0 (out of 120; SD = 14.1); Huntington's disease patients' mean was 72.1 (SD = 19.6, $U = 16$, $Z = 3.96$, $P < 0.001$). Therefore, the people with Huntington's disease performed poorly overall. Examining the six emotion regions separately, to find the principal sources of abnormality, gave the following result: no significant difference for happiness ($U = 89.5$, $Z = 0.88$, $P > 0.1$), but significantly poorer performance of the Huntington's disease group for all other emotions (for surprise $U = 61$, $Z = 2.07$, $P < 0.05$; for fear $U = 49$, $Z = 2.57$, $P < 0.05$; for sadness $U = 48$, $Z = 2.62$, $P < 0.01$; for disgust $U = 32.5$, $Z = 3.26$, $P < 0.01$ and for anger $U = 38.5$, $Z = 3.01$, $P < 0.01$).

Although the Huntington's disease group were found to be significantly impaired at perceiving all emotions except happiness, it seemed that the impairment of disgust was especially severe (*see* Fig. 4). In the range where disgust should have been perceived, seven people in the Huntington's disease group never reported it at all (scoring 0 out of 20 correct for this emotion) and three others showed gross impairments (scores of 4 out of 20, 2 out of 20 and 1 out of 20 correct). So, 10 of the 13 people in the Huntington's disease group were almost completely unable to see disgust in facial expressions. To confirm this differentially severe impairment of perception of disgust, we compared the performance of the Huntington's disease group at recognizing disgust with their performance with the next most badly affected emotion (fear). In doing this, though, it was important to allow for differences in control performance with these emotions; we therefore derived scores for each person with Huntington's disease which expressed their ability to see fear and to see disgust as a proportion of the appropriate control mean. A Wilcoxon matched-pairs, signed-ranks test applied to these data confirmed that the performance of the Huntington's disease group was worse with disgust than with fear ($Z = 4.25$, $P < 0.001$).

The performance of the Huntington's disease group was also evaluated for whether they were above chance level. If people responded entirely at random, we would expect an average of 3.3 out of 20 correct answers for each emotion. We therefore used the Wilcoxon matched-pairs signed-ranks test to determine whether performance was above this chance level for disgust (the most poorly recognized emotion) and for fear (the next most badly affected emotion). This showed above-chance performance for fear (average 8.5 out of 20 correct, $n = 13$, $T = 11$, $P < 0.05$) but not for disgust (average 4.3 out of 20 correct, $n = 13$, $T = 37$, $P > 0.1$).

As a second method of assessing their responses, we calculated the number of 'unusual' responses made by people with Huntington's disease and controls in the same six

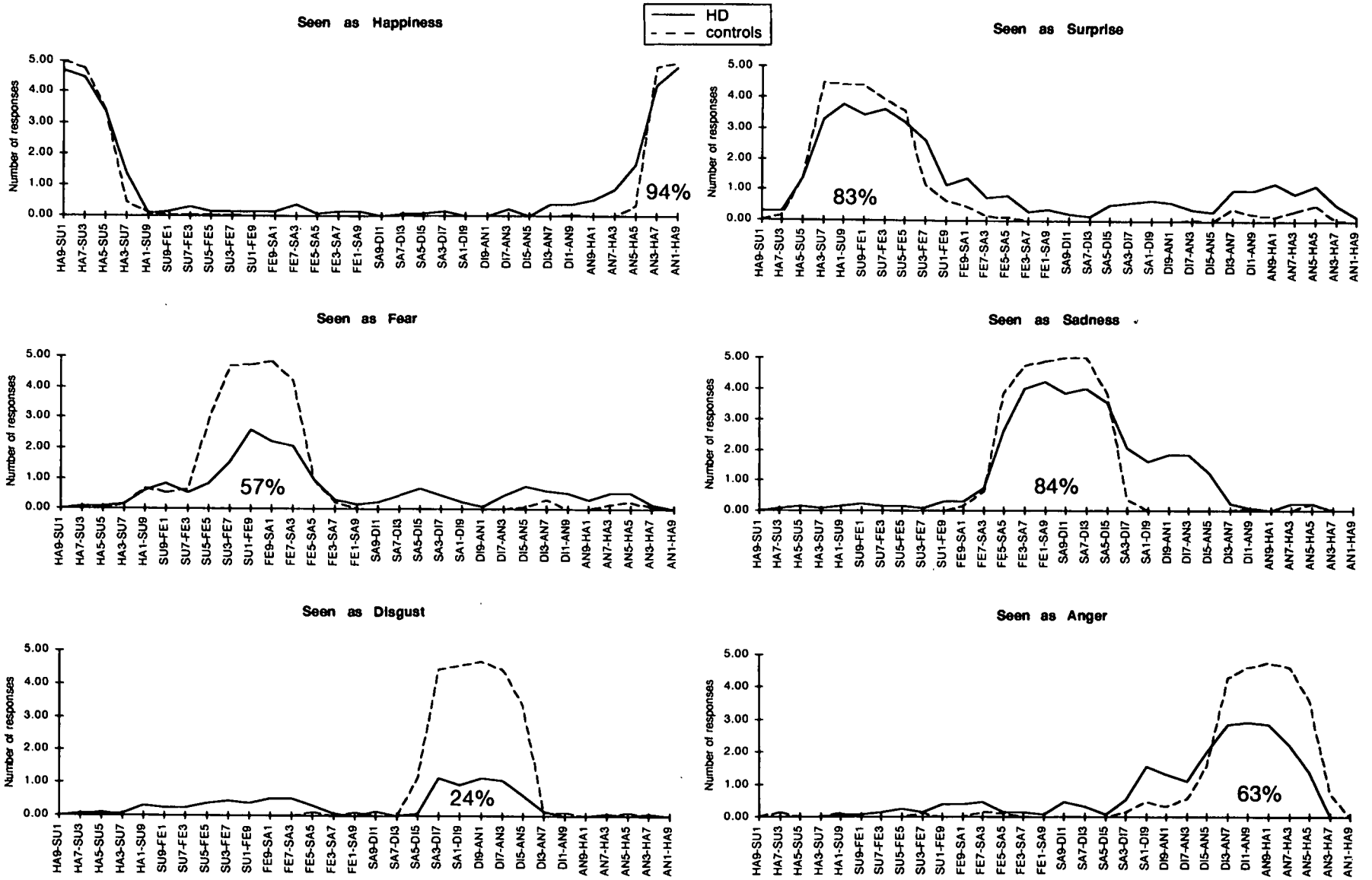

Fig. 4 Identification of the morphed images shown in Fig. 2 by people with Huntington's disease (HD) and control subjects. Along the horizontal axis HA, SU, FE, SA, DI and AN represent happiness, surprise, fear, sadness, disgust and anger respectively and 1, 3, 5, 7 and 9 represent 10, 30, 50, 70 and 90% respectively. Thus HA9-SU1 is a morphed image reflecting 90% happiness and 10% surprise.

Table 3 *Identification of emotion in facial and vocal expressions by Huntington's disease patients (HD) and control subjects*

Emotion	Facial expressions		Vocal expressions	
	HD	Controls	HD	Controls
Happiness	9.5 (0.9)	9.9 (0.2)	4.1* (2.2)	7.2 (2.8)
Surprise	6.0** (2.1)	8.9 (1.0)	5.1* (2.7)	8.1 (1.8)
Fear	2.9** (1.7)	7.3 (2.6)	3.4** (2.6)	7.6 (1.7)
Sadness	6.3* (2.5)	8.9 (1.2)	7.3 (2.9)	8.4 (2.1)
Disgust	1.9** (2.1)	8.1 (2.9)	0.5** (1.5)	5.9 (3.3)
Anger	4.1** (1.9)	8.6 (1.9)	7.7 (2.6)	9.4 (0.7)

*$P < 0.01$; **$P < 0.001$ (significantly different from control performance). Mean scores (out of 10) are given with standard deviations in parentheses.

emotion regions. It should be noted that the six expression continua had been prepared by interpolating images between maximally confusable prototype expressions. Given that normal subjects occasionally mistake confusable prototype expressions (e.g. surprise and fear) for one another, a response was only scored as 'unusual' if the morph was identified with an emotion label that was inappropriate for that particular region of the continuum and if it was inappropriate for the confusable regions to the immediate right and left of this region; we will call these 'remote prototype errors'. Hence, if a morph in the surprise region was labelled sadness, disgust or anger, that response was scored as a remote prototype error, whereas if the same morph was identified as fear or happiness, it was not scored as such since these are the more commonly confused adjacent prototypes. Remote prototype errors for each of the six emotion regions were calculated for people with Huntington's disease and compared with the numbers of remote prototype errors made by the age-matched controls. Across the six defined regions taken together, there was a highly significant group difference; the controls' mean total number of remote errors was 4.4 out of 120 (SD = 6.8); Huntington's disease mean was 19.5 (SD = 13.7, $U = 20.5$, $Z = 3.77$, $P < 0.001$). Therefore, the people with Huntington's disease again performed poorly overall. Examining the six emotion regions separately gave the following result: no significant difference for happiness ($U = 102.5$, $Z = 0.34$, $P > 0.1$), but significantly more remote prototype errors by the Huntington's disease group for all other emotions (for surprise $U = 51$, $Z = 2.49$, $P < 0.05$; for fear $U = 49$, $Z = 2.57$, $P < 0.05$; for sadness $U = 57.5$, $Z = 2.22$, $P < 0.05$; for disgust $U = 51$, $Z = 2.49$, $P < 0.05$; for anger, $U = 37.5$, $Z = 3.06$, $P < 0.01$).

In summary, the people with Huntington's disease were impaired at recognizing all emotions except happiness, both in terms of correct identifications and remote prototype errors. However, further analyses of correct responses showed that their problems were disproportionately severe with disgust, being at chance level overall and significantly below the next most badly affected emotion (fear).

Follow-up study of emotion

For our follow-up study of 11 people from the Huntington's disease group, we tested recognition of facial expressions of each of the six emotions from the Ekman and Friesen (1976) series and recognition of vocal expressions of the same emotions. We also gave the subjects questionnaires to examine self-assessed emotion.

Our follow-up test of perception of facial expressions used 10 examples of each of the six emotions in the Ekman and Friesen (1976) series. Results from this test of recognition of specific emotions are presented in Table 3, which shows mean performance by people with Huntington's disease and control subjects.

The analysis of correct identifications in this test followed the procedure for the emotion hexagon, using Mann–Whitney U tests to compare Huntington's disease and control scores. Across the six emotions taken together, there was a highly significant group difference; the controls' mean total number of correct responses was 51.8 out of 60 (SD = 6.4); Huntington's disease patients' mean was 30.6 (SD = 7.8, $U = 3.5$, $Z = 4.23$, $P < 0.001$). Examining the six emotions separately, to find the principal sources of abnormality, gave the following result: no significant difference for happiness ($U = 64.5$, $Z = 1.36$, $P > 0.1$), but significantly poorer performance by the Huntington's disease group for all other emotions (for surprise $U = 16$, $Z = 3.65$, $P < 0.001$; for fear $U = 20$, $Z = 3.46$, $P < 0.001$; for sadness $U = 33.5$, $Z = 2.82$, $P < 0.01$; for disgust $U = 13.5$, $Z = 3.76$, $P < 0.001$; for anger $U = 12$, $Z = 3.83$, $P < 0.001$).

Again, it seemed that the impairment of disgust was especially severe. None of the 11 people with Huntington's disease scored more than 5 out of 10 correct for recognizing disgust, and six people had scores of 1 or 0. To confirm this differentially severe impairment of disgust, we compared the performance of the Huntington's disease group in recognizing disgust with their performance with the next most badly affected emotion (fear), using derived scores for each person with Huntington's disease which expressed their ability to recognize fear and disgust as a proportion of the appropriate control mean. A Wilcoxon matched–pairs, signed–ranks test applied to these data confirmed that the Huntington's disease group made more errors with disgust than with fear ($Z = 4.11$, $P < 0.001$).

Mean scores for each emotion by the Huntington's disease group were also again evaluated for whether they were above the chance level of an average of 1.7 out of 10 correct answers for disgust and for fear. Wilcoxon matched-pairs signed-ranks tests showed above-chance performance for fear (average 2.9 out of 10 correct, $n = 11$, $T = 11$, $P < 0.05$) but not for disgust (average 1.9 out of 10 correct, $n = 11$, $T = 26$, $P > 0.1$).

The differentially severe problem of the Huntington's disease group in perceiving disgust does not merely reflect the relative difficulty normal subjects experience in recognizing this emotion in facial expressions. Our controls

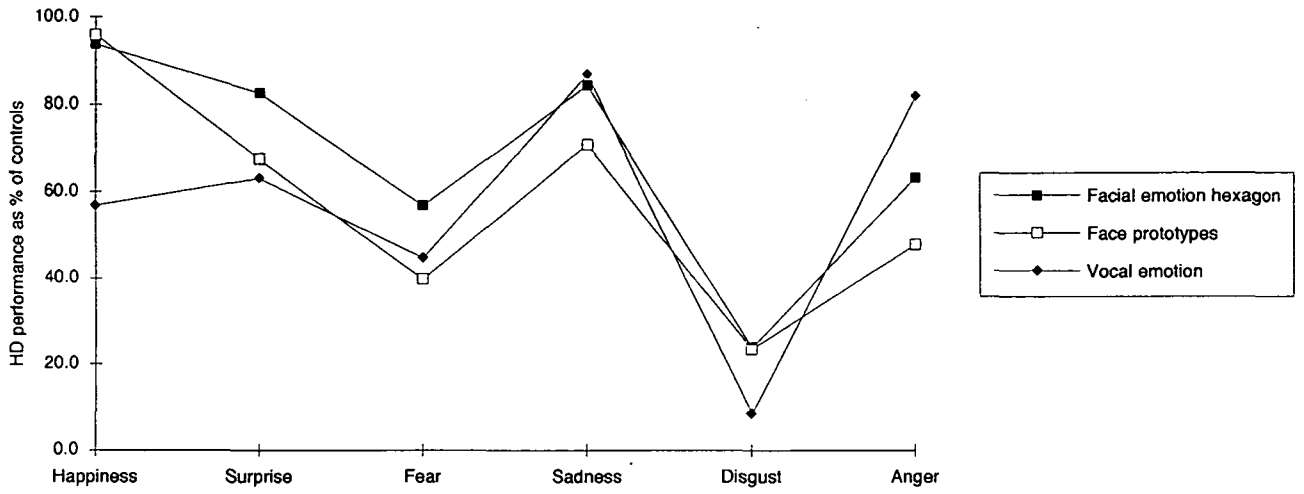

Fig. 5 Comparing three tests of recognition of basic emotions. Two tests are of facial expression recognition (morphed emotion hexagon, recognition of prototypes from the Ekman and Friesen series) and one test is of vocal emotion recognition. Performance of the Huntington's disease group is represented as a percentage of control performance for each emotion.

found fear the most difficult expression to recognize, also in the Ekman and Friesen (1976) norms for the recognizability of each of the photographs we used, to a much larger control group, disgust was second only to happiness in ease of recognition.

Results from the test of recognition of vocal expressions of emotion are also presented in Table 3 and were analysed in the same way. Across the six emotions taken together, there was a highly significant group difference; the controls' mean total number of correct responses was 46.6 out of 60 (SD = 9.3); the Huntington's disease patients' mean was 28.1 (SD = 11.7, $U = 17$, $Z = 3.60$, $P < 0.001$). Examining the six emotions separately gave the following result: no significant difference for sadness ($U = 69$, $Z = 1.15$, $P > 0.1$) or anger ($U = 57$, $Z = 1.72$, $0.1 > P > 0.05$), but significantly poorer performance by the Huntington's disease group for all other emotions (for happiness $U = 37$, $Z = 2.66$, $P < 0.01$; for surprise b$U = 34.5$, $Z = 2.78$, $P < 0.01$; for fear $U = 15$, $Z = 3.69$, $P < 0.001$ and for disgust $U = 11.5$, $Z = 3.86$, $P < 0.001$).

As for the facial emotion recognition tests, the impairment of disgust recognition was especially severe for vocal emotion. Nine of the 11 people with Huntington's disease scored 0 out of 10 for recognizing disgust, one person scored 1 out of 10 and one person (whose hobby had been acting, and who was trained by a professional actor in the past) scored 5 out of 10. To confirm this differentially severe impairment of disgust, we again compared the performance of the Huntington's disease group at recognizing disgust with their performance with the next most badly affected emotion (fear), using derived scores for each person which expressed their ability to recognise fear and disgust as a proportion of the appropriate control mean. A Wilcoxon matched-pairs signed-ranks test confirmed that the Huntington's disease group made significantly more errors with disgust than with fear ($Z = 4.11$, $P < 0.001$).

Mean scores for each emotion in the Huntington's disease group were again evaluated for whether they were above the chance level of an average of 1.7 out of 10 correct answers for disgust and for fear. Wilcoxon matched-pairs, signed-ranks tests showed that performance for fear was in a borderline region (average 3.4 out of 10 correct, $n = 11$, $T = 12$, $0.1 > P > 0.05$), whereas recognition of disgust was actually significantly *below* chance level (average 0.5 out of 10 correct, $n = 11$, $T = 11$, $P < 0.05$), reflecting consistent failure to report hearing this emotion at all.

In all, then, our original and follow-up studies used three tests of recognition of the six basic emotions in the Ekman and Friesen (1976) series; two tests of facial expression recognition (morphed emotion hexagon, recognition of prototype expressions) and one test of vocal emotion recognition. Figure 5 shows a comparison across the results of these tests, in which the performance of the Huntington's disease group is represented as a percentage of control performance. There is a consistent pattern of differentially severe impairment in recognition of disgust. As we have noted, mean recognition rates for disgust by the Huntington's disease group were always significantly below their rates for the next most badly affected emotion (for each test, this was fear) and did not rise above chance level in any of the three tests.

We also used questionnaires to examine self-assessed emotion. Results from the three emotion questionnaires are summarized in Table 4. For the anger scale (Novaco, 1975), anger ratings on a scale from 1 to 5 were summed across the 40 items we used, to give an overall score. For the 75-item fear scale (Wolpe and Lang, 1964), the ratings were converted into 1–5 scale equivalents and summed. The disgust

Table 4 *Self-assessed emotion on questionnaires involving anger, fear and disgust: results from Huntington's disease patients (HD) and control subjects*

Questionnaire	Descriptive statistics: mean (+SD)		Inferential statistics		
	HD	Controls	*U*	*z*	*P*
Anger (max. = 200)	133.7 (19.6)	140.8 (15.2)	37.0	1.33	0.09
Fear (max. = 375)	109.0 (47.3)	133.9 (37.6)	48.5	1.51	0.06
Disgust (max. = 100)	52.3 (17.5)	58.9 (18.5)	60.5	1.14	0.13
Disgust components (max. = 100):					
food	48.9 (18.9)	56.7 (23.1)	65.0	0.92	0.18
animals	73.9 (27.6)	74.2 (25.2)	80.5	0.11	0.46
body products	64.8 (20.8)	80.0 (25.4)	45.5	1.97	0.02
sex	42.1 (25.2)	60.8 (33.0)	53.0	1.58	0.06
envelope violations	51.1 (29.3)	49.2 (20.3)	79.0	0.18	0.43
death	47.7 (35.7)	42.5 (33.7)	76.0	0.34	0.37
hygiene	45.5 (29.2)	55.8 (24.9)	66.5	0.84	0.20
magical contagion	44.4 (28.7)	51.7 (29.1)	68.0	0.76	0.22

Inferential statistics show Mann–Whitney *U*, equivalent *Z* and 1-tailed probability. The questionnaires concerning anger, fear and disgust were from Novaco (1975), Wolpe and Lang (1964) and Haidt *et al.* (1994), respectively.

scale was scored slightly differently, using the procedure devised by the scale's originators (Haidt *et al.*, 1994). The two 'true or false' items scored 0 or 100, and the rating-scale items scored 0 (not disgusting), 50 (slightly disgusting), or 100 (very disgusting). An average score out of a possible maximum of 100 was then calculated for each of the eight domains, and for all domains pooled into an overall score.

As well as showing means and standard deviations for the Huntington's disease and control group scores on these emotion questionnaires, Table 4 gives inferential statistics (Mann–Whitney *U*, *Z* equivalent and one-tailed *P*) summarizing the differences between Huntington's disease and control performances. We used one-tailed probabilities for these comparisons because poorer performance by the Huntington's disease group was expected on the basis of our emotion recognition findings. As can be seen, the Huntington's disease group did generally report lower emotional responsiveness, but this only approached or reached statistical significance on the anger questionnaire ($0.1 > P > 0.05$), the fear questionnaire ($0.1 > P > 0.05$) and the body products ($P < 0.05$) and sex ($0.1 > P > 0.05$) sub-components of the disgust questionnaire.

Discussion

Our findings show severe impairments affecting the recognition of emotion by people with Huntington's disease, with a particularly striking loss of ability to perceive disgust. In two tests of facial emotion recognition and one test of vocal emotion recognition, the average rate at which disgust was detected by people with Huntington's disease was significantly below the next most badly affected emotion (in each case, fear) and did not rise above chance level. For all of these tests, the majority of individuals in the Huntington's disease group made no correct responses to expressions of disgust; they simply did not perceive disgust at all.

These problems in seeing emotion in facial expressions did not seem to reflect basic visual deficits. None of the people in the Huntington's disease group showed any impairment of contrast sensitivity on the VCTS 6000 test; this finding is particularly relevant because our face perception tests were based on black and white photographic images. Unimpaired spatial contrast sensitivity functions indicate that the people with Huntington's disease remained able to see subtle changes of light and shade in such static grey-scale images. This conclusion is reinforced by observations that their ability to pick up cues indicating sex from facial appearance was not significantly impaired, even when computer-graphics techniques were used to ensure that only subtle changes in the shapes of facial features were available for this decision (*see* Figs 1 and 3). Similarly, perception of physical cues differentiating two familiar identities was also not significantly impaired (Figs 1 and 3). Although inspection of Fig. 3 suggests that the slope of the sex and identity functions was slightly less steep in the Huntington's disease group, and our analyses showed occasional borderline differences for these continua, their problems in differentiating emotions were much more severe.

A consistent group difference in what might be considered higher-order perceptual abilities was found for the Benton unfamiliar face matching test (Table 2). In this case, although significantly below that of controls, performance of the Huntington's disease group was well above chance. In fact, the mean performance of the Huntington's disease group fell at the lower end of the range regarded as normal by Benton *et al.* (1983). Similarly, the substantial standard deviation of the scores of the Huntington's disease group on the Benton test indicated that, at the individual level, whilst some

people with Huntington's disease would show significant impairment, others were performing normally. A borderline deficit was also found for the eye gaze task.

Whilst we would not, therefore, wish to underplay the fact that the Huntington's disease group were showing impairments on tests of face perception, which is indeed consistent with other evidence of visual perceptual abnormalities in Huntington's disease (Moses *et al.*, 1981; Brouwers *et al.*, 1984; Mohr *et al.*, 1991; Sprengelmeyer *et al.*, 1992), we note that the deficits found in our emotion recognition tests were, in comparison, disproportionately severe.

A problem of interpretation which often affects investigations of higher-order perceptual deficits in neurodegenerative disorders concerns whether impairments on perceptual tests actually reflect consequences of cognitive deterioration; for example, failures to understand or remember task instructions properly, or to deploy effective strategies. The results with the morphed continua shown in Fig. 3 are particularly important here. All of the four tests summarized in Fig. 3 used an exactly equivalent general procedure; five computer-interpolated images were generated along a continuum between two prototypes which were recognizable to normal people, and subjects were asked to determine which of the prototypes each image was most like without seeing the prototypes themselves. All that varied across tests was the choice of physical continuum (sex, familiar identity, or emotion). People with Huntington's disease showed severe problems in differentiating fear from anger, but they could discriminate male from female, Humphrey Bogart from Cary Grant, or sadness from happiness much better. Since one continuum was disproportionately affected, this pattern cannot reflect generalized cognitive deterioration in the Huntington's disease group. Note also that the cues needed to tell the male from female faces in Fig. 1 are more subtle than those needed to tell facial expressions of anger from fear, yet only the latter was significantly impaired.

Problems in discriminating anger from fear clearly form only part of the deficit in social perception in Huntington's disease. The morphed expression hexagon showed that the recognition of other emotions was also compromised, with an especially severe impairment of disgust. This pattern was confirmed by follow-up tests of facial and vocal emotion recognition.

Investigators in previous studies of emotion recognition in Huntington's disease (Speedie *et al.*, 1990; Jacobs *et al.*, 1995*b*) have not used sufficiently sensitive tests to allow the assessment of different basic emotions and did not, in any case, include disgust. Taken together, however, our data not only reveal severe impairments of emotion recognition in Huntington's disease, but also show that the recognition of some emotions is more impaired than others.

We must now ask why this should happen and, especially, why the recognition of disgust should be so poor in people with Huntington's disease? The most obvious hypothesis would be that Huntington's disease compromises the recognition of all emotions to some extent, but has the most readily noticeable effects on the emotions which are the most difficult to decode. This would fit with our data for the auditory modality, where disgust was also the most difficult emotion for controls in the test we devised. However, the same argument does not work for the visual modality; for both visual tests, disgust was not the most difficult emotion for controls. In the morphed hexagon task, the rank order of control performance was happiness (best), sadness, disgust, surprise, anger, then fear (worst). For the recognition of prototype facial expressions of each of the six emotions from the Ekman and Friesen (1976) series, the rank ordering of difficulty for control subjects was happiness (best), sadness and surprise, anger, disgust, then fear (worst). In Ekman and Friesen's (1976) norms, which are based on a larger sample, the rank ordering for the faces we used was happiness (best), disgust, surprise, sadness, then fear and anger. So there are no grounds for thinking that disgust is an especially difficult emotion to perceive in the visual modality. On the contrary, it often involves a characteristic movement of the upper lip which can provide a highly distinctive cue (*see* Fig. 2).

Further grounds for thinking that perceptual difficulty *per se* does not give an adequate account of the problems experienced by the Huntington's disease group come from comparing their performance to that of people with bilateral amygdala damage. Figure 6 shows the average performance of the Huntington's disease group on tests of recognition of facial expressions of basic emotions (the morphed hexagon and the 60 prototypes from the Ekman and Friesen series) and the performance of the same tests by two people with bilateral amygdala damage (Calder *et al.*, 1966*b*); performance is represented as a percentage of control performance for each emotion. The two cases of bilateral amygdala damage involved surgery for intractable epilepsy (Young *et al.*, 1995) and encephalitis (Laws *et al.*, 1995). They showed less severe impairments of facial expression recognition than the Huntington's disease group overall, but experienced the greatest difficulties with fear. Problems in perceiving fear have also been reported in another case of bilateral amygdala damage due to Urbach–Wiethe disease (Adolphs *et al.*, 1994; Adolphs *et al.*, 1995). Figure 6 shows that people with amygdala damage are as impaired as our Huntington's disease group in perceiving fear, but they show only minor difficulties with disgust, i.e. there is a striking difference between the recognition of fear and the recognition of disgust (which is extremely poor in the Huntington's disease group and well preserved for the amygdala damage cases).

So it seems unlikely that the loss of disgust recognition in Huntington's disease reflects only perceptual difficulty; disgust is not generally the most difficult emotion to recognize. Instead, our data are consistent with the possibility already suggested by the effects of bilateral amygdala damage (Adolphs *et al.*, 1994; Adolphs *et al.*, 1995; Calder *et al.*, 1966), that different emotions may be compromised by different types of brain damage. On this view, bilateral

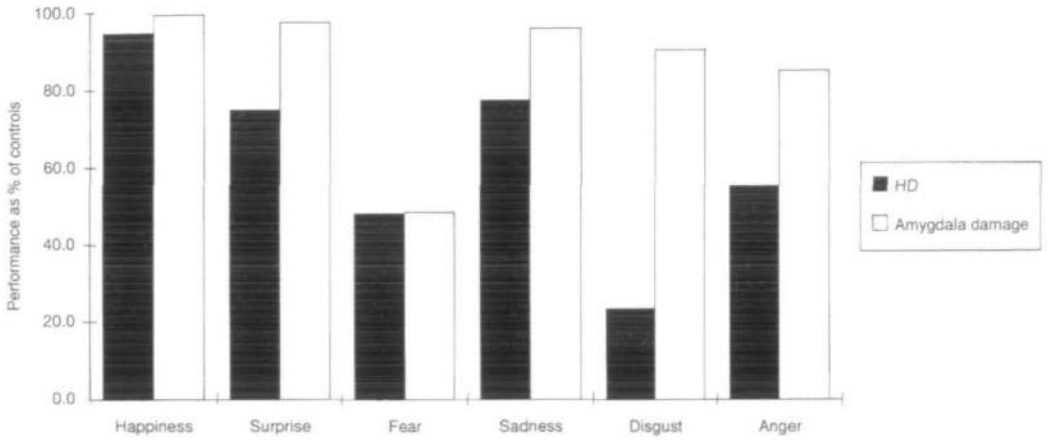

Fig. 6 Comparing the average performance of the Huntington's disease group on tests of recognition of facial expressions of basic emotions (morphed emotion hexagon, recognition of prototypes from Ekman and Friesen series) with that of two people with bilateral amygdala damage (Calder *et al.*,1996*b*). Performance is represented as a percentage of control performance for each emotion.

amygdala damage leads to problems in recognizing fear, whereas Huntington's disease especially affects the recognition of disgust. Of course, our data show that Huntington's disease affects the recognition of fear too, but this may also derive from neuronal loss in the amygdala, since this is known to occur in Huntington's disease (Lange and Aulich, 1986).

To explain differences between the patterns of impairment of emotion recognition found in Huntington's disease and those after bilateral amygdala damage, then, the possibility that certain basic emotions may have dedicated neural substrates needs to be seriously considered. Among these, disgust and fear are prime candidates. Indeed, it has often been noted that disgust can be traced back to a distinct evolutionary origin as a rejection response to bad tastes (Rozin *et al.*, 1993).

The position we have adopted here is, thus, that the almost complete loss of ability to perceive disgust shown by our Huntington's disease group may reflect a more central problem in experiencing this emotion. This would be consistent with our parallel findings of deficits in visual and auditory perceptual domains. We hypothesize that mechanisms for perceiving emotions in others may be closely linked to those involved in actually experiencing the equivalent emotions oneself because this has evolved as one of the main ways we learn our emotional reactions; many of the things which disgust us do so not because we have direct experience, but because we have learnt from the reactions of others. The same is true for fear; we can learn to fear things simply by seeing that they frighten other people. Learning from the reactions of others allows us to learn about different types of danger without being harmed ourselves. An interesting discussion of the biological importance of such a mechanism of emotional contagion can be found in Brothers (1989).

With this in mind, we used questionnaires to examine self-assessed emotion. However, although people with Huntington's disease generally rated their emotional reactions to be less marked than those of controls, which is in line with our account, they did not show such striking problems as were noted on the tests of emotion recognition. We suspect that this is because questionnaires do not tap directly into emotional experience; instead, they can be answered using some mixture of one's present emotional reactions and one's knowledge of what situations have provoked or were likely to provoke such reactions in the past. In other words, even if you had lost your ability to be disgusted by some stimulus, you might still rate this stimulus as disgusting because you recall having been disgusted by it (or thinking you would be) in the past. Looked at in this way, questionnaires are a rather blunt instrument for assessing what we wanted to get at, but a useful first step.

The issue of whether people with Huntington's disease have also lost the ability to experience (as well as to perceive) disgust thus merits further study; certainly, clinical observations of poor personal hygiene for people with Huntington's disease suggest it is a strong possibility.

We turn now to consider the neural substrate of disgust. Because neuronal degeneration is widespread in Huntington's disease, it does not provide an ideal starting point for finding this. However, we can identify some potentially useful pointers for further investigation in cases with different aetiologies. Morphometrical studies have found that atrophy is especially marked in three cerebral regions in Huntington's disease; the striatum, occipital and parietal cortex, and paleocortical structures (Lange, 1981). We can therefore consider the possibility that each of these regions might be involved in processing emotions, and disgust in particular.

The obvious candidates are the basal ganglia. The suggestion that these structures are involved in emotion processing has been put forward by Jacobs *et al.* (1995*a*), who found that people with Parkinson's disease showed deficits in comparing emotional facial expressions. However,

this needs to be interpreted cautiously because the nature of basal ganglia involvement is different in Parkinson's and Huntington's diseases, and there is also contradictory evidence; two other studies have reported that people with Parkinson's disease were not impaired at recognizing facial expressions (Dewick *et al.*, 1991; Madeley *et al.*, 1995).

Further studies of the possibility of basal ganglia involvement in the loss of disgust recognition we have documented in Huntington's disease are therefore needed, and a more detailed assessment of emotion recognition in Parkinson's disease could form a useful first step. The caudate nucleus is widely considered to form the core site of pathology in Huntington's disease, and is thought to be involved in stimulus–response habit learning. A plausible speculation is that integration and learning of behavioural responses may be more significant for disgust than other basic emotions.

Parts of the parietal and occipital lobes are also atrophied in Huntington's disease. It is well known that cerebral lesions in these regions can lead to disturbances in visual perception (McCarthy and Warrington, 1990). But such deficits have a broad impact on visual abilities and therefore seem unlikely to be able to explain satisfactorily the loss of particular emotions.

This brings us to the paleocortex. Given that what Rozin *et al.* (1993) have called 'core disgust' can be traced back to rejections of bad tastes and smells, it is possible that rhinencephalic structures are in some way involved in processing disgust-related social expressions. These could include the amygdala and periamygdaloid cortex, entorhinal cortex, piriform cortices, rostral insula cortex and caudal orbital frontal cortex.

Rozin *et al.* (1993, p. 588) made reference to the concept of preadaptation in evolutionary biology, arguing that other types of disgust can be added to core disgust by 'an opportunistic accretion of new domains of elicitors, and new motivations, to a rejection system that is already in place'. From this point of view, a neural structure has to be identified which is able to integrate olfactory, visual and auditory information. Candidates are amygdala, the medial dorsal nucleus of the thalamus, orbital frontal cortex, insula, those parts of rhinal cortex adjacent to the temporal pole and piriform cortex.

In Huntington's disease there is a 20% loss of volume in the prepiriform and periamygdalar regions and a 25% loss of volume in the amygdala (Lange, 1981; Lange and Aulich, 1986). The amygdala can be divided into a phylogenetically older part which, as well as many intrinsic connections, receives olfactory input (corticomedial nuclei) and a phylogenetically younger part with predominantly visual, acoustic and somatosensory input (basolateral nuclei). Given that it is the recognition of fear rather than disgust which is compromised by amygdala damage (Adolphs *et al.*, 1994; Adolphs *et al.*, 1995; Calder *et al.*, 1966*b*), the amygdala itself does not seem to be especially involved in mediating disgust, but other regions in close proximity, such as periamygdalar and piriform cortex, may be important to this emotion. The piriform region, in particular, has input from the lateral olfactory stria and is closely related to the visual and acoustic information processing basolateral nuclei of the amygdala.

Taken together, the paleocortical regions atrophied in Huntington's disease thus fulfil the basic requirements for processing disgust-related stimuli. Studies of the recognition of disgust by people with selective brain injuries affecting these regions are therefore warranted.

Since Papez (1937) put forward his theory of a neural circuit for emotion, there has been a tendency among many neuroscientists to assume by default that emotions depend on a common set of neural mechanisms, even though Papez's particular hypotheses concerning the specific localization of emotion have been discredited. However, we have demonstrated here that a degenerative neural disease can affect some emotions more than others, and that it has a differentially severe impact on one of the basic emotions; disgust. This merits further investigation because it will fundamentally change our understanding of emotion if basic emotions, to some extent, have different neural substrates.

Acknowledgements

We wish to thank Professor Paul Ekman for giving us permission to use and reproduce images derived from the Ekman and Friesen (1976) Pictures of Facial Affect series, and the Press Association for permission to use press photographs. We also wish to thank Professor John Aggleton for anatomical advice, Stefan Huschenbeck for technical support, Stephan Herms for help in preparing the auditory emotion test and two anonymous referees for helpful comments. The study was supported by grants from the Deutsche Forschungsgemeinschaft and the Royal Society to R.S.

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Received January 29, 1996. Revised May 13, 1996.
Accepted June 13, 1996