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Disgust in pre-clinical Huntington's disease: A longitudinal study

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Abstract

Emotion recognition from both face and voice and experience of emotions were investigated in a group of non-symptomatic people at risk of carrying the Huntington's disease gene who presented for genetic testing. Based on the results of the DNA test, a group of people carrying the Huntington's disease gene (HD+), and a group of non-carriers (HD−) were formed. Since we were especially interested in the time course of possible deficits in emotion recognition, all people at risk were reassessed 6 and 12 months after the initial assessment. Recognising facial expressions of disgust was significantly impaired on all three assessments in the HD+ group, while recognition of vocal emotions and the experience of emotions were largely unaffected, confirming that deficits in recognition of facial expressions of disgust are an early correlate of carrying the gene for Huntington's disease. The inclusion of a healthy control group ($n = 37$) further allowed an estimate of the genetic and environmental contribution to deficits in facial emotion recognition.
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1. Introduction

Recognition of fearful and disgusted facial expressions can be differentially impaired in people with amygdala damage and people with insular-striatal dysfunction, respectively.

Adolphs and co-workers (Adolphs, Tranel, Damasio, & Damasio, 1994; Adolphs, Tranel, Damasio, & Damasio, 1995) first described deficits in recognising fear in people with bilateral amygdala damage. These initial findings were substantiated by several studies looking at people with lesions or functional deficits to the amygdala (Broks et al., 1998; Calder et al., 1996; Edwards, Pattison, Jackson, & Wales, 2001; Meletti et al., 2003; Rosen et al., 2002; Sato et al., 2002; Sprengelmeyer et al., 1999; Thomas et al., 2001) and backed up by demonstrations of activation of the human amygdala in response to facial expressions of fear in a number of studies using both PET and fMRI (e.g. Breiter et al., 1996; Fischer et al., 2003; Morris et al., 1996). Further investigation of one of the people (DR) for whom a deficit in recognising facial expressions of fear had already been established (Calder et al., 1996) showed that recognition of auditorily presented fear was also impaired (Scott et al., 1997). This case was comparable to a later report of NM, a person with bilateral gliosis of the amygdala (Sprengelmeyer et al., 1999) who showed selective deficits in recognising facially, vocally and also gesturally displayed signals of fear. Both single cases suggest that lesions to the amygdala may not only affect recognition of fearful facial expressions, but also other social signs indicating fear, such as vocal intonation, and body postures. This view forms the 'cross-modal' hypothesis of fear recognition (Calder, Lawrence, & Young, 2001). Further support comes from Dolan, Morris, and de Gelder (2001), who reported that hearing fearful voices can modulate amygdala activation to fearful facial expressions. However, the 'cross-modal' hypothesis has been challenged by Anderson and Phelps (1998) and Adolphs, Tranel, and Damasio (2001) who reported that their patients showed deficits for facial expres-

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sions only, and therefore favoured a 'uni-modal' account involving a face-specific deficit.

In contrast to these reports of selectively impaired recognition of fear, an interestingly different pattern of findings comes from studies looking at recognition of facial and vocal expressions of emotion in people with Huntington's disease (Sprengelmeyer et al., 1996; Sprengelmeyer, Young, & Sprengelmeyer, 1997; Wang, Hoosain, Yang, Meng, & Wang, 2003) who were found to show an overall deficit in recognising emotion, with particularly severe problems for recognition of expressions of disgust from both face and voice (Sprengelmeyer et al., 1996). In subsequent studies, Gray, Young, Barker, Curtis, and Gibson (1997) as well as Hennenlotter et al. (2004) found evidence for a selective deficit in facial disgust recognition even in pre-symptomatic Huntington's disease gene carriers, pointing to the basal ganglia and insular cortex as a possible site for disgust processing. However, one study (Milders, Crawford, Lamb, & Simpson, 2003) reported a more generalised deficit of emotion recognition in people with manifest Huntington's disease, and no emotion recognition deficit in people at risk of carrying the Huntington's disease gene.

People with other disorders involving the basal ganglia have also been reported to be poor at recognising disgust faces. People with Tourette syndrome with co-morbid obsessive compulsive behaviour, people with obsessive compulsive disorder (Sprengelmeyer, Young, & Pundt, 1997), and people with Wilson's disease (Wang et al., 2003) all show selective deficits in facial disgust recognition. Involvement of the basal ganglia, but also the insular cortex, in facial disgust recognition has been confirmed by functional imaging studies (Hennenlotter et al., 2004; Phillips et al., 1997; Sprengelmeyer, Rausch, Eysel, & Przuntek, 1998) and a study looking at evoked responses to facial expressions using implanted depth electrodes in people suffering from temporal lobe epilepsy (Krolak-Salmon et al., 2003). An interesting patient (NK), who in some respects parallels the single case reports of fear deficits in patients DR and NM (Calder et al., 1996; Sprengelmeyer et al., 1999), was reported by Calder, Keane, Manes, Antoun, and Young (2000). NK, with a lesion affecting the left putamen and the insular cortex, showed selective deficits in facial and vocal disgust recognition and also a reduced sensitivity for disgust as measured by a questionnaire. Based on the findings of the Huntington study conducted by Sprengelmeyer et al. (1996), in which a facial and auditory disgust recognition deficit was reported, and on single case NK, a 'cross-modal' hypothesis for disgust recognition was suggested (Calder et al., 2001).

To date, only facial expression recognition has been studied in the early stages of Huntington's disease; auditory recognition and experience of emotion have not been investigated. The present study addresses the question of whether or not there is a selective disgust processing deficit in people in the pre-clinical stage of Huntington's disease by using established tests assessing facially and vocally displayed emotions as well as questionnaires for measuring self-assessed emotions. We were especially interested in the time course of the development of possible emotion processing deficits; therefore people at risk of carrying the Huntington's disease gene were assessed at three different points in time, each investigation separated by a 6-month interval.

2. Participants

Twenty-two people at risk of carrying the Huntington's disease gene gave written informed consent to take part in the study. The Huntington gene (or huntingtin gene) is defined as the section of the DNA that codes the huntingtin protein. The Huntington's disease gene is a mutated version of the Huntington gene, containing at least one allele with a higher than normal number of CAG repeats (>34). The risk of carrying the Huntington's disease gene was defined as having at least one parent suffering from manifest Huntington's disease or having a parent diagnosed as a mutation carrier. All participants were taken from a consecutive series of people presenting for genetic testing at the Department of Human Genetics at the Ruhr University of Bochum.

People at risk of carrying the Huntington's disease gene were assessed neuropsychologically at three different points in time. The interval between the first and second assessments was 6.9 months (S.D. 2.3), and between the second and third assessments 6.2 months (S.D. 0.8). Four people declined to take part in the second and third neuropsychological assessments. Therefore, 18 people were included in the analysis of the second and third assessments.

Genetic testing was performed using standard procedures (Goldberg, Andrew, Clarke, & Hayden, 1993). The participants were informed about their genetic status after the first and before the second neuropsychological assessment. For statistical analysis, two groups were formed from the results of the DNA tests, with one group consisting of people carrying the Huntington's disease gene (HD+) and another group whose members were found to carry the normal huntingtin alleles (HD−). In the HD+ group, the mean number of CAG repeats of the affected allele was 44.0 (S.D. 3.0), in the HD− group the mean number of CAG repeats of the longest allele was 21.3 (S.D. 3.9). A description of the individual participants can be found in Table 1.

The mean age of the participants in the HD+ group was 31.0 years (S.D. 8. 5), mean duration of schooling was 11.3 years (S.D. 1.8), and the average IQ was 113.2 (S.D. 8.1) as assessed by a short version of the WAIS (WIP). The mean age of the participants in the HD− group was 38.3 years (S.D. 14.5) and mean duration of schooling was 10.6 years (S.D. 1.4). Their average IQ was 108.8 (S.D. 9.7). The *t*-tests showed no significant differences between the HD+ and HD− groups with respect to age ($t = 1.49$, $P = 15$), years of formal education ($t = -0.89$, $P = 0.39$), or intelligence ($t = -1.16$, $P = 0.26$).

Basic visual processing was assessed using the Vistech VCTS 6000 contrast sensitivity chart. This measures the

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