PERGAMON

Neuropsychologia 41 (2003) 527–537

NEUROPSYCHOLOGIA

www.elsevier.com/locate/neuropsychologia

Impairment of recognition of disgust in Chinese with Huntington's or Wilson's disease

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Abstract

The selective involvement of the basal ganglia in recognition of the facial expression of disgust was investigated by examining a group of six symptomatic Huntington's disease patients and 32 Wilson's disease patients in China. Morphed photographs of facial expressions covering happiness–surprise–fear–sadness–disgust–anger were used and the patients were asked to label each photo. Other measures assessed basic cognitive functions and perception of non-emotion facial information, such as perception of gender, age, gaze direction, and recognition of unfamiliar as well as famous people. There was dissociation between the perception of emotions and other facial information, and between impairment of recognition of disgust and other emotions. The basal ganglia are the overlapping substrate involved in both Huntington's and Wilson's disease, although each has its own other lesions. The differentially severe impairment of recognition of disgust in the Chinese Huntington's disease and Wilson's disease patients strengthens the view that basal ganglia are selectively involved in processing the emotion of disgust.

Keywords: Basal ganglia; Huntington's disease; Wilson's disease; Emotion; Perception; Disgust

1. Introduction

The human face conveys much social information, and the ability to process such information is important for interpersonal interactions. Models of face recognition [8] tend to separate the processing of emotional expression from other kinds of face information, such as gender, age, and identity. Traditionally, Papez's circuit or the limbic system was thought of as responsible for emotion processing. Recently, more specific substrates for separate emotions have been explored.

One facial expression that has been investigated extensively is fear. Selective impairment in the recognition of fear has been found with bilateral amygdala damage [2,3,7,10], or bilateral cingulate gyrus damage [44]. Although cases reported in the literature showed the greatest impairment in fear recognition, they often showed impairment in recognizing other emotions too. For instance, SM also showed a slight problem with surprise and anger [2], DR had problems also with anger and disgust [10], and SE also had difficulty with anger [7,10]. They demonstrated a trend toward dissociation, with the alternative explanation being that fear is the most difficult emotion to recognize. It could mean that the other easier expressions remained relatively unaffected with milder general impairment. But this alternative explanation has been rejected on the grounds of data from normal subjects [2,10].

Stronger evidence that various basic emotions have distinct neural substrates comes from double dissociation, with a complementary finding that other lesions or disease groups show relatively intact fear recognition but impaired recognition of other emotional expressions. Disgust is such an example, involving the basal ganglia.

The significance of the basal ganglia-thalamocortical circuit for the processing of emotion has been suggested [4,6], and impairment of recognition of facial expression in stroke patients with basal ganglia lesions has been reported [11]. Because the basal ganglia deteriorate with Parkinson's disease (PD) and Huntington's disease (HD), two relatively common diseases, a number of studies have focused on patients with these diseases. Manifestations of PD include emotional dysprosody [21], as well as a deficit in identifying prosody [13,42]. Patients with PD were reported to be impaired in recognizing emotional expressions [24].

Jacobs et al. [23] reported that HD patients were impaired in recognizing facial expressions as well as identity

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0028-3932/02/$ – see front matter
PII: S0028-3932(02)00171-9

of faces. They tested five patients with HD on tasks of face discrimination, as well as emotional expression discrimination and matching. The emotions used in this study were happiness, sadness, anger, and fear. Four of the five patients were impaired in at least one task of face discrimination, and all demonstrated impairment in recognition of at least one emotional expression. Using morphed emotional continua produced from six basic emotions, Sprengelmeyer and coworkers [40,41] found disproportionately great impairment in the recognition of disgust in patients with HD, although there was also light impairment in the recognition of other emotions. These findings, together with those of impairment in the recognition of fear with bilateral amygdala or cingulate gyrus lesion [44] comprise a trend double dissociation. Although not classical dissociation [38], they suggest that the substrates for recognition of facial emotions are divisible, at least for fear and disgust.

HD is an inherited disease, initially affecting the basal ganglia. Patients characteristically manifest late onset involuntary chorea and eventually suffer general intellectual deterioration [15]. Pathological studies [26,27] have shown substantial atrophy of amygdala, periamygdala cortex and prepiriform cortex, besides obvious basal ganglia degeneration in HD. Thus, it can be argued that at least some of these other areas could also be involved in the recognition of disgust, although other studies have reported patients (e.g. SM, see [2]) with bilateral amygdala damage without impairment of recognition of disgust expressions, which indicate that amygdala have little role in recognizing disgust.

One method to rule out substrates other than the basal ganglia is to observe the manifestation of very early stage HD with less chance of damage to the other areas. Another method is to investigate other diseases with damage also to the basal ganglia in addition to areas different from those of Huntington's disease. Very early stage HD is supposed to only involve the basal ganglia, without showing the deterioration of general intelligence. Gray et al. [20] compared the performance of people in HD families, one group with and another without the HD gene. The gene carriers, including presymptomatic individuals, showed impairment in the recognition of disgust and relative sparing in the recognition of other emotions. As these very early stage patients likely had no apparent atrophy in periamygdala and prepiriform cortex, this result supported the notion that the basal ganglia is the substrate for the recognition of disgust.

The observation of HD in different cohorts also helps to clarify the picture. Because reports of selective deficit of disgust recognition are all based on cases in the west, it is appropriate to ask if, for example, Chinese patients have a similar deficit. We report here on six Chinese patients with HD. We also report on a group of Chinese patients with Wilson's disease (WD), disease which also involves the basal ganglia.

Wilson's disease, hepato-lenticular degeneration, is a hereditary neuro-degenerative disease. With neurological symptoms, WD patients often manifest behavior or emotional disorders (showing impulsive, instinctive behaviors, or depression), and mild cognitive deficit [28,30,43]. There is deficit in retrieving memory but learning and forgetting rates are normal [22]. Also, patients show lower scores on IQ, memory quotient, and Trail Making Test [29], the Mini-Mental Status Examination, and Benton's visuospatial tests [12].

2. Study 1

2.1. Method

2.1.1. Subjects

Six symptomatic Huntington's disease patients (four male and two female) were diagnosed by a group of neurologists and recruited from the First Hospital of Anhui Medical University, China. Their age ranged from 40 to 50, they had 5–10 (mean 8.5) years of education, and the onset age of disease was from 38 to 43 (mean 40.5). At the time of testing, all the subjects showed choreoathetic movements, with scores from 1 to 3 on a clinical rating scale (range 0–3) used to determine severity of choreic movement [25]. The duration after initial onset of motor disturbances, estimated from case history, ranged from 2 to 7 years (referred to as "Course" in Table 1). All patients could independently cope with everyday tasks but were not competent to return to work in a social environment.

The control group consisted of 16 normal people (10 male and 6 female) with similar cultural and educational background to the HD patients. They had normal vision and hearing, with no history or signs of neurological or psychiatric illness.

2.1.2. Background neuropsychological tests

Background neuropsychological tests included the Wechsler Adult Intelligence Scale-Revised Chinese Version (WAIS-RC, [19]), Wechsler Memory Scale-Chinese Version (WMS-C, [18]), Word Fluency Test (naming animals with four legs, within 1 min), Mini-Mental Status Examination-Chinese version (MMSE) [47], Benton Tests of Line Orientation and Visual Form Discrimination [5].

2.1.3. Face perception tests

These included perception of gender and age of various faces, gaze direction, and recognition of both unfamiliar and familiar faces. Perception of gender and age involved five photographs each of the faces of boys, girls, young men, young women, old men, and old women. Subjects were asked to identify the gender of each face and to classify their age level in terms of being child/young/old (with the maximum score in each measure being 30). Perception of gaze direction was tested with 15 photographs with different eye gaze directions, with three faces each for 20° left, 20° right, 20° up, 20° down, and straight ahead. Subjects had to report in which direction each face was looking (with a maximum