

JNNP

C Clinical characteristics

BMJ Publishing Group Ltd

J Neurol Neurosurg Psychiatry 2008 79: A9-A18

Updated information and services can be found at:

http://jnnp.bmj.com/content/79/Suppl_1/A9.full.html

These include:

**Email alerting
service**

Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To order reprints of this article go to:

<http://jnnp.bmj.com/cgi/reprintform>

To subscribe to *Journal of Neurology, Neurosurgery & Psychiatry* go to:

<http://jnnp.bmj.com/subscriptions>

EHDN Annual Meeting: abstracts

Conclusions: The results of this preliminary study suggest that HD patients are more prone to visual distractibility compared with controls when performing line bisection under illusory conditions.

C.11 PSEUDO-NEGLECT IN HUNTINGTON'S DISEASE: A RETROSPECTIVE ANALYSIS

M Couette, MF Boisse, A-C Bachoud-Levi. *Centre de Reference Maladie de Huntington, Faculte de Medecine, 8 rue du General Sarrail, 94010 Creteil cedex, France*

Early disorders in visuospatial perception in patients with Huntington's disease (HD) may correspond to a specific phenotype of the disease. Both pseudoneglect and neglect have been described in patients with HD, in a group study and in a single case study, respectively (Ho *et al*, 2003, 2004). Such hemi-attentional troubles in the right visual field are consistent with the time course of the neural degeneration of the disease, damaging first the left striatum evidenced by recent neuroimaging studies. Here, we aimed to confirm the existence of this pseudoneglect profile with a simple visuospatial paper-pencil task (Zazzo's cancellation task) in 20 consecutive unselected patients with 16 controls matched for age and educational level. Participants were instructed to cross out one, two or three small aligned signs covering a full page with a time constraint of 90 s for each page. We analysed their performance by separating the page into two equal parts. We rated the percentage of omissions on both sides (number of omissions/number of targets to cancel). We conducted a two-way analysis of variance with "target numbers" (one, two and three signs to cancel) and side (left, right) as within factors. Patients produced more omissions and the task elicited more omissions for the three than two targets condition and even more for the one target than the two targets condition. There was no effect of side but there was an interaction between side and group. The percentage of omissions was more important in the right part in patients only, suggesting a pseudoneglect profile on this rapid and simple paper-pencil task. Further studies are needed to determine what is the relevance of the pseudoneglect disorder as a marker of disease evolution and what is its prognosis value, if any.

C.12 THE POWER OF POSITIVES: EVIDENCE FOR AN OVERALL EMOTIONAL RECOGNITION DEFICIT IN HUNTINGTON'S DISEASE

L Robotham, I Trinkler, D Sauter. *INSERM U841, Creteil, France*

The recognition of emotions of disgust, anger and fear have been shown to be significantly impaired in Huntington's disease (eg, Sprengelmeyer *et al*, 1997, 2006; Gray *et al*, 1997; Milders *et al*, 2003, Montagne *et al*, 2006; Johnson *et al*, 2007; De Gelder *et al*, 2008). The relative impairment of these emotions might have implied a recognition impairment specific to negative emotions. Could the asymmetric recognition deficits be due not to the complexity of the emotion but rather reflect the complexity of the task? In the current study, 15 Huntington's patients and 16 control subjects were presented with negative and positive non-speech emotional vocalisations that were to be identified as anger, fear, sadness, disgust, achievement, pleasure and amusement in a forced-choice paradigm. This experiment more accurately matched the negative emotions with positive emotions in a homogeneous modality. The resulting dually impaired ability of Huntington's patients to identify negative and positive non-speech emotional vocalisations correctly provides evidence for an overall emotional recognition deficit in the disease. These results indicate that previous findings of a specificity in emotional recognition deficits might instead be due to the limitations of the visual modality. Previous experiments may have found an effect of emotional specificity due to the presence of a single positive emotion, happiness, in the midst of multiple negative emotions. In contrast with the previous literature, the study presented here points to a global deficit in the recognition of emotional sounds.

C.13 DISTURBED MOTOR RESONANCE AT THE BASIS OF THE EMOTION RECOGNITION DEFICIT IN HUNTINGTON'S DISEASE? AN EMG INVESTIGATION

¹I Trinkler, ^{1,2}S Devignevielle, ³A Achaibou, ^{1,2}A-C Bachoud-Levi. *Inserm U841, Equipe 1, Faculte de Medecine Paris XII, 8 rue du General Sarrail, 94010 Creteil, France;* ²Departement d'Etudes Cognitives, ENS, 29 rue d'Ulm, 75005 Paris, France; *laboratory for Neurology and Imaging of Cognition, Department of Neuroscience, University of Geneva, 1 rue Michel-Servet, 1211 Geneva, Switzerland*

Background: Patients with Huntington's disease (HD) have a deficit recognising emotional expressions. Recently, it has become clear that this deficit extends to other modalities and is not specific to disgust (Snowden *et al*, 2008; Johnson *et al*, 2007; Henley *et al*, 2008). In a recent study we established that this recognition deficit comes along with a production and that recognition and production are a highly correlated deficit (Trinkler and Bachoud-Levi, 2008). This points to a potential role of "motor resonance" in emotional recognition, ie, a mechanism for recognising emotional expression in somebody else through an internal motor simulation thereof, which might be impaired in HD.

Aims: We aimed to compare motor resonance and the production of emotional expressions between Huntington's patients and healthy controls using electromyography.

Methods/Techniques: 14 early HD and 14 matched healthy subjects were tested on three conditions of motor production of emotional expressions using electromyography: (1) spontaneous micro-mimicry of emotional expressions as is observed in healthy subjects (Dimberg, 1982); (2) overt imitation of facial expressions; (3) production of facial expressions from emotional words ("anger", "disgust", "joy"). Measurements were taken repeatedly from three facial regions: zygomatic (active in smiling), corrugator (active in frowning) and nasalis (active in wrinkling the nose in disgust).

Results: HD patients show less and less specific activation of the muscle active in the corresponding emotion in all three conditions. They thus lack automatic facial mimicry and show significantly less imitation and less ability to mime an emotional expression overtly compared with healthy subjects.

Conclusions: The absence of motor resonance for emotional facial expressions in HD might account for their difficulty in recognising emotions in others and may potentially be at the core of an empathy deficit.

C.14 A STUDY OF SOCIAL COGNITION IN HUNTINGTON'S DISEASE USING THEORY OF MIND TASKS

¹ V Havet-Thomassin, ³C Verry, ⁴D Bonneau, ³F Dubas, ¹ D Le Gall. *¹Neuropsychology Unit, Department of Neurology, University Hospital of Angers, Angers, France;* *²Laboratory of Psychology (JPRES EA 2646), University of Angers, Angers, France;* *³Department of Neurology, University Hospital of Angers, Angers, France;* *⁴Department of Genetic, University Hospital of Angers, Angers, France*

Background: Huntington's disease (HD) is a degenerative disorder with predominant involvement of the frontostriatal system. This condition gives rise to altered social and breakdown in interpersonal relationships, although the factors underlying these changes remain poorly defined.

Aims: This study used cognitive and affective theory of mind tasks, respectively, to explore the ability of patients with HD to interpret social situations and their ability to ascribe mental states to others. Methods: Ten HD patients and 10 healthy volunteers matched by age and educational level were given a non-verbal cognitive theory of mind task assessing the attribution of intentions to others (Brunet *et al*, 2003) and a revised version of the "Reading the Mind in the Eyes" Test (Baron-Cohen *et al*, 2001), which is an affective theory of mind task assessing the understanding of other people's mental states from their eyes.

Results: The two measures of theory of mind were indicative of a significant impairment in HD patients.