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Social cognition in frontotemporal dementia and Huntington's disease

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Abstract

Frontotemporal dementia (FTD) and Huntington's disease (HD) are degenerative disorders, with predominant involvement, respectively of frontal neocortex and striatum. Both conditions give rise to altered social conduct and breakdown in interpersonal relationships, although the factors underlying these changes remain poorly defined. The study used tests of theory of mind (interpretation of cartoons and stories and judgement of preference based on eye gaze) to explore the ability of patients with FTD and HD to interpret social situations and ascribe mental states to others. Performance in the FTD group was severely impaired on all tasks, regardless of whether the test condition required attribution of a mental state. The HD group showed a milder impairment in cartoon and story interpretation, and normal preference judgements. Qualitative differences in performance were demonstrated between groups. FTD patients made more concrete, literal interpretations, whereas HD patients were more likely to misconstrue situations. The findings are interpreted as demonstrating impaired theory of mind in FTD, as one component of widespread executive deficits. In HD the evidence does not suggest a fundamental loss of theory of mind, but rather a tendency to draw faulty inferences from social situations. It is concluded that social breakdown in FTD and HD may have a different underlying basis and that the frontal neocortex and striatum have distinct contributions to social behaviour.

Keywords: Frontotemporal dementia; Huntington's disease; Social cognition, theory of mind; Behaviour

1. Introduction

Frontotemporal dementia (FTD) and Huntington's disease (HD) are degenerative brain disorders that affect frontostriatal systems. FTD is a predominantly neocortical disorder, characterised by radical alterations in personality, emotions, and social, interpersonal conduct [12,25,35,44,46,47,56]. Behavioural changes include disinhibition, tactlessness, and loss of social proprieties [12,37,44,45,47]. Cognitive assessment typically shows deficits predominantly in frontal executive functions [47,57], indicating deficits in abstraction, problem solving, attention, mental set shifting, sequencing, and mental generation of information. Patients are not clinically amnesic, although formal memory test performance is often inefficient, attributed to executive impairments.

Neuroimaging [48,62] and pathological studies [41] of FTD demonstrate severe frontal and anterior temporal neocortical atrophy, which may be largely confined to orbital regions, or (particularly with progression of disease) more widespread extending into anterior cingulate and dorsolateral frontal cortex. Modest pathological changes in the striatum reflect the emergence of striatal neurological signs usually relatively late in the disease course.

Huntington's disease (HD) is a predominantly subcortical disorder, distinguished clinically by its characteristic involuntary movements [30]. Patients' social conduct is altered, albeit less profoundly than in FTD, and there is frequently severe breakdown in interpersonal relationships. Patients are often described as self-centred, lacking in sympathy and empathy, and mentally inflexible, sometimes with fixed ideas, which may not be consistent with the prevailing view or available evidence. As in FTD deficits have been reported in the processing of emotions [23,32,59]. Cognitive changes are predominantly in the realm of frontal executive function [14], although generally less marked in degree than in FTD, and memory impairment is ascribed to inefficient encoding and retrieval strategies rather than a primary failure of retention.

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Pathological [40,63] and structural neuroimaging [6,38] studies of HD have demonstrated marked atrophy of caudate and putamen, which form the dorsal part of the striatum or neostriatum. This is present even in the early stages of disease [4], and has been reported in some studies in pre-symptomatic individuals who carry the HD mutation [5]. Some frontal neocortical atrophy may also occur later in the disease course [7], assumed to be at least partly (although not necessarily exclusively) secondary to striatal differentiation [40].

Thus, FTD and HD represent complementary disorders in which there is a virtual, although not exclusive, double dissociation with respect to the distribution of degenerative change within the frontal neocortex and striatum. FTD and HD thus provide ideal models for the study of frontal-striatal function. The striatum has traditionally been recognised for its importance in the domain of motor functioning, in the execution of learned motor plans [42]. Conditions such as HD attest to its crucial role also in cognition. The identification of parallel and segregated frontal-subcortical circuits, distinguished by their areas of origin in the frontal cortex [3,43], has led to the notion that the striatum is intimately linked functionally to the cerebral cortex. The assumption is that analogous cognitive deficits may arise from disruption at different levels (i.e. frontal cortical or striatal) of the circuit.

Commonalities between FTD and HD with respect to the prominence of behavioural changes and pattern of cognitive deficits are thus unsurprising. Nevertheless, it cannot be inferred that deficits underlying FTD and HD are identical. Executive tasks make multiple demands, so that test scores may mask fundamental differences in the reason for failure. Similarly, disordered social behaviour might have different underlying substrates. Comparative studies of FTD and HD ought to clarify the nature of change in each condition. Moreover, in view of the predominance of frontal neocortical changes in FTD and of striatal changes in HD, such studies provide the potential for improving knowledge of the relative contributions of the frontal lobes and striatum in behaviour and cognition.

Traditional executive tasks do not capture the full range of abnormalities in FTD and HD and may be a relatively poor predictor of the patient's functioning in daily life. Indeed, some patients with FTD, in whom the pathology is confined to the orbital regions of the frontal lobes, perform relatively well on conventional executive tasks, despite impaired judgement and gross breakdown in their social conduct in daily life [39,57]. Such a finding is consistent with reports that lesions of the orbital frontal lobes may give rise to severe breakdown in social behaviour in the context of normal executive functioning [15,16,54]. In HD, disorganised behaviour and breakdown in interpersonal relationships in daily life are often prominent clinical features, outweighing changes in neuropsychological test performance. There are at least two factors that are likely to contribute to the relative insensitivity of traditional tests to some of the changes in FTD and HD. Traditional neuropsychological tests are structured and typically require a constrained set of responses. By contrast, everyday life situations are open-ended, and require self-generated structure and organisation. Secondly, traditional tests are impersonal, whereas everyday life involves social interaction. Neuropsychological tasks that are both open-ended and involve interpretation of social scenarios are likely to be particularly informative in FTD and HD because they mirror the daily life situations in which FTD and HD patients so dramatically fail. They may also have the potential to reveal fundamental differences between FTD and HD.

Recent years have seen an accumulation of literature on social cognition [1,2]. A core component of social functioning is the capacity to attribute independent mental states to others and to predict other people's behaviour on the basis of their mental states, a capacity known as "theory of mind" [9,36,51]. There is a growing body of evidence from both neuroimaging [10,18,20,21,27] and brain lesion studies [24,29,53,60,61] that the frontal lobes have a pivotal role in theory of mind. However, to date there have been no direct comparisons in performance on tests of social cognition between patients with predominantly frontal neocortical and predominantly striatal pathology.

Clinical observation of patients with FTD and HD leads to the prediction that performance on tests that require interpretation of event scenarios is likely to differ. FTD patients typically lack insight into the change in their own behaviour and appear oblivious of the effects that their behaviour has on others, leading to the prediction that such patients show a genuine loss of theory of mind. By contrast, at clinical interview HD patients may make pertinent and insightful remarks about the effects of their illness on a close relative (e.g. "It is hard on my husband having to do everything for me. He must get very fed up"). Such apparent cognisance of others' mental states leads to the prediction that social breakdown in HD arises for reasons other than a primary inability to ascribe mental states to others. In FTD, a purported problem in theory of mind is unlikely to be exclusive. FTD patients commonly show concreteness of thought. A concrete interpretation of events would be expected to be manifest in a general difficulty in the interpretation of social scenarios, even when they do not depend on attribution of mental states.

The present study investigates the ability of FTD and HD patients to interpret social situations and explores by means of error analysis possible differences between the two groups. The study involved four tasks drawn from the literature on social cognition that have been used to address theory of mind. The tasks differ with respect to their level of difficulty. The cartoon and story tasks (tasks 1–3) make relatively great mental demands on the patient raising the possibility that they may exceed the capabilities of some patients for reasons that have little to do with social cognition per se. The judgement of preference task (task 4) examines the capacity for mental state attribution while minimising