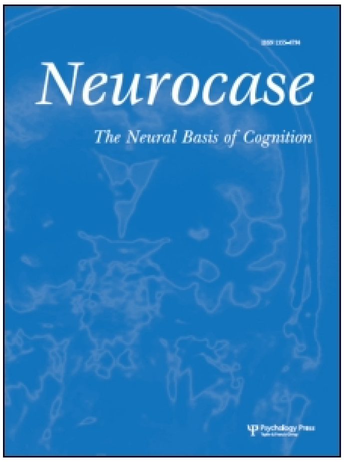

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Emotion, social functioning and activities of daily living in frontotemporal dementia

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Emotion, social functioning and activities of daily living in frontotemporal dementia

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Social functioning in FTD is profoundly affected, and forms the basis for the clinical diagnosis of the behavioural variant of the disease (bv-FTD). In particular, there are deficits in emotional processing, but the inter-relationship of such deficits to other aspects of social functioning remains unclear. We studied patients with bv-FTD ($n = 14$) and AD ($n = 14$), and compared their performance on a test of emotion recognition with their scores on two carer-based assessments: the Disability Assessment for Dementia (DAD) of activities in daily living (ADL), and the Cambridge Behavioural Inventory (CBI). The bv-FTD group had significantly greater impairments in ADLs, and had higher scores on the CBI, compared to the AD group. Despite a deficit in emotion recognition, particularly involving negative emotions, in the FTD group relative to AD and controls, performance on this task did not correlate with ADL ratings which instead, correlated highly with carer-rated apathy levels on the CBI. The study highlights the multifactorial nature of social dysfunction in FTD which is important in the management of these patients and in designing effective behavioural and therapeutic interventions. The relationship of emotional processing to other aspects of social cognition in FTD is reviewed.

Keywords: Frontotemporal dementia; Social cognition; Emotion processing; Activities of daily living.

INTRODUCTION

The ability to interact socially is dependent on several crucial elements. The capacity to appreciate emotion, both in self and others, an understanding of people's desires and intentions (Theory of Mind), the ability to regulate behaviour and be flexible in social interactions, and the competence to execute social behaviours and self-care skills (activities of daily living) are all important aspects of social cognition. In frontotemporal dementia (FTD), particularly the behavioural variant of the disorder (bvFTD), there is a marked deficit in social functioning (Neary et al., 1998; Gregory et al., 2002), yet many aspects of this deterioration are incompletely understood.

Deficits in emotion processing are prominent in FTD. While perception of faces is normal in patients with FTD, the identification of facial emotion (Keane, Calder, Hodges, & Young, 2002), is severely impaired (Fernandez-Duque & Black, 2005; Lavenu, Pasquier, Lebert, Petit, & Van der, 1999). These emotion recognition deficits are multimodal, with both facial recognition and vocal emotional prosody affected (Keane et al., 2002; Perry et al., 2001). Typically, negative emotions (anger, fear, sadness and disgust) are more affected than positive (happiness, sadness) or neutral expressions (Fernandez-Duque & Black, 2005; Lough et al., 2006; Rosen et al., 2004; Sturm, Rosen, Allison, Miller, & Levenson, 2006). Some of the observed behavioural changes in FTD could,

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in part, be explained by a deficit in emotion processing. Insensitivity to social cues, particularly those indicating disapproval such as anger or disgust, would make effective social interaction very difficult. Since profound lack of empathy is also characteristic of FTD (Lough et al., 2006; Neary et al., 1998; Rankin, Kramer, & Miller, 2005), perhaps failure to recognise sadness or disappointment in others might explain this associated deficit.

While emotion processing appears to be an important aspect of the social dysfunction in FTD, it seems unlikely that it explains all aspects. For instance, a prior study suggested that ability to perform theory of mind tasks was not associated with executive function (Torralva et al., 2007). Moreover, knowledge of social norms remains relatively intact, but the ability to process social norm violations or to be flexible about the context within which they operate is affected (Lough et al., 2006), and there is an inability to follow social rules despite the negative consequences (Mendez, Chen, Shapira, & Miller, 2005).

A relatively neglected area of study in frontotemporal dementia has been in the realm of self-care skills or activities of daily living (ADL). These abilities are vital aspects of social functioning as they allow the individual to partake in many of the other social activities mentioned earlier, and they are profoundly affected in FTD (Mioshi et al., 2007). Activities of daily living are generally sub-classified into basic and instrumental activities, and can be further fractionated into domains of initiation, planning and execution. Basic activities, or BADLs, relate to day-to-day core survival abilities such as eating, walking, dressing, and aspects of personal hygiene such as washing and using the toilet. Instrumental activities of daily living (IADLs), by contrast, are defined by their higher level of complexity, and reflect the ability to live independently in the community. They encompass use of the telephone, managing finances, medications, meal preparation, household chores, and leisure activities (Lawton & Brody, 1969; Trombly, 1993). Using a caregiver based rating scale, the Disability Assessment for Dementia (DAD) (Gelinas, Gauthier, McIntyre, & Gauthier, 1999), we recently established that bvFTD patients were more affected in their ADLs than those with Alzheimer's disease, or FTD patients with predominantly language dysfunction (Mioshi et al., 2007). Initiation of basic ADLs was far more affected than the ability to plan or execute them, which suggests that apathy or lack of motivation may play a significant role. It is possible that deterioration in ADLs may also relate, at least in part, to a decline in emotional processing secondary to an inability to detect disapproval, anger or disgust in caregivers.

The relationship between emotion, apathy and other neuropsychiatric symptoms and measures of social competence such as ADL performance has not been investigated in FTD. We were interested in whether the inability to appreciate emotion, particularly negative emotion (and thus disapproval), would impact upon ability to perform ADLs. We also investigated the relationship between perception of emotions, neuropsychiatric features which are prominent in FTD (disinhibition, challenging behaviours and apathy) and ADLs in FTD and a matched comparison group of patients with Alzheimer's Disease.

METHODS

Subjects

Twenty-eight patients and caregivers participated in the study, consisting of the following subgroups: behavioural variant frontotemporal dementia (N = 14) and early Alzheimer's disease (N = 14). We also included a group of age-matched controls (N = 16) for one of the clinical tests described below. Informed consent was obtained from patients and their care-givers. Patients were included if they (1) fulfilled clinical criteria for FTD (2) or AD (24); (2) had an accompanying person who could give a reliable account of the patient's routine, either due to living in the same house or due to close participation in the patient's everyday life; (3) did not have a physical disability that could confound assessment of ability to perform activities of daily living; (4) did not have major depression; AND (5) had undergone assessment using the Clinical Dementia Rating scale – CDR (Morris, 1997), Addenbrooke's Cognitive Examination Revised – ACE-R (Mioshi, Dawson, Mitchell, Arnold, & Hodges, 2006) and Mini Mental State Examination – MMSE (Folstein, Folstein, & McHugh, 1975) within 90 days of the functional assessment. Cognitive testing was given by a senior research nurse, and all patients were assessed by the senior author (JRH). Diagnoses were made on the basis of a multidisciplinary consensus (neurologist, neuropsychiatrist and neuropsychologist), and measures described in this study were not included in the diagnostic process. Patients with significant psychiatric illness were excluded.

Functional assessment

Activities of daily living were assessed by using the Disability Assessment for Dementia – DAD (Gelinas et al., 1999). The DAD is an informant-based scale, which includes 40 items – 17 related to basic self-care (BADLs: 'hygiene', 'dressing', 'continence', 'eating') and 23 related to instrumental activities of daily living (IADLs: 'meal preparation', 'telephoning', 'going on an outing', 'finance and correspondence', 'medications' and 'leisure and housework'). Lower scores on the DAD denote greater impairment. The scale is designed to have its total score corrected to 100, i.e., non-applicable questions are excluded from the total score to avoid gender bias towards activities (e.g., cooking; house chores; finances). IADL and BADL scores were combined for the analysis (total DAD score), and subscores for initiation, planning and execution derived. The DAD was consistently given by a senior occupational therapist (EM).

Behavioural assessment

Information on behavioural changes was available from the Cambridge Behavioural Inventory (CBI) (Bozeat, Gregory, Lambon Ralph, & Hodges, 2000; Nagahama, Okina, Suzuki, & Matsuda, 2006) which assesses behaviour in a number of different neuropsychiatric domains. Data were extracted to provide scores for four separate behavioural domains: mood, motivation, challenging behaviour and disinhibition. Each domain consists of several questions where caregivers are asked to rate behaviour on a four point scale (0 = never, 1 = a few times per month, 2 = a few times per week, 3 = daily and 4 = constantly). Scores were scaled for the total number of questions per domain which were not equivalent.

Emotion recognition

The recognition of facial expressions was assessed with the Emotion Hexagon (see Figure 1) (Calder, Young, Perrett, Etcoff, & Rowland, 1996a; Young, Perrett, Calder, Sprengelmeyer, & Ekman, 2002), which contains morphed (blended) facial expressions posed by model JJ from the Ekman and Friesen (1976) pictures of facial affect series. The test has been used in a number of previous studies (Calder et al., 1996b, 2003; Sprengelmeyer et al., 1996); a detailed description can be found in Calder et al. (1996a). In brief, the test comprises morphed (or blended) continua ranging between the following six facial expression pairs, happiness–surprise, surprise–fear, fear–sadness, sadness–disgust, disgust–anger, and anger–happiness. Each continuum consists of five morphed images moving from one end of the continuum to the other in 20% steps. For example, the images in the happiness–surprise continuum contained the following percentages of the happy and surprised expressions, 90% happiness–10% surprise, and then 70–30%, 50–50%, 30–70%, and 10–90% of the same two expressions. Data from healthy volunteers indicate that stimuli that contain 90 or 70% of an expression are consistently identified as the intended emotion (Calder et al., 1996a; Sprengelmeyer et al., 1996; Young et al., 1997). Images containing 50% of two emotions are categorized as each of the two contributing emotions with approximately equal frequency. The stimulus set consists of 30 images in total (6 continua × 5 morphed faces).

The 30 morphed images were presented individually on a computer monitor in random order (i.e., they were not grouped into the underlying continua). The task was to decide which of six emotion labels (happiness, sadness, anger, fear, disgust, and surprise) best described the facial expression displayed. The labels were visible throughout testing and participants were given as much time as they required to make their selection. No feedback was given regarding the appropriateness of any response. Participants undertook a total of 4 blocks of trials, 120 image

Figure 1. Morphed facial expressions on the emotion hexagon (disgust-anger continuum) showing disgust (90%)-anger (10%) on the left, through to disgust (10%)-anger (90%) on the right-hand side of the panel. Only one face at a time was displayed to subjects.

selections in all. Each block contained one presentation of each of the 30 morphed faces in random order. Performance was scored for each emotion label, and in addition, total scores for positive (happiness, surprise) and negative (anger, sadness, disgust and fear) were calculated. The scores for positive emotions were scaled appropriately to allow comparison with negative emotion totals.

Brief cognitive assessment

The ACE-R is the updated version of the ACE (Mioshi et al., 2006) designed to assess five cognitive domains: attention/orientation, memory, verbal fluency, language and visuospatial abilities. The total score is 100; higher scores reflect better ability. The ACE-R was designed to be sensitive to early stages of dementia, and incorporates the MMSE.

Dementia staging

The CDR (Morris, 1997) is a clinical staging instrument of dementia that combines six domains of cognitive and functional performance: memory, orientation, judgment and problem solving, community affairs, home and hobbies, and personal care. An algorithm allows the calculation of a total score; a score of zero reflects the performance of a healthy person and higher scores denote greater impairment.

Data analysis

Data were analysed using SPSS 12.0.1 (SPSS Inc, IL, USA). Demographic data such as age and education were analyzed using unpaired *t*-tests, as were the results for the MMSE, and CDR. Performance on the emotion recognition tests was analyzed using repeated measures ANOVAs followed by preplanned *post-hoc t*-tests to examine any interactions. Preliminary analysis suggested that scores on the ACE-R, ADL measures and CBI ratings were significantly skewed, and non-parametric Mann–Whitney *U*-tests were performed. The relationship between emotion recognition tests, CBI ratings and ADL measures was analyzed using *Kendall tau b* a non-parametric measure of association which corrects any observed correlation for ties.

Not all patients received every assessment: for the ADL assessment (N = 28, FTD = 14, AD = 14); Face emotion recognition (N = 20, FTD = 13, AD = 7); CBI (N = 21, FTD = 11, AD = 10).

RESULTS

Demographics (see Table 1)

Age was similar across the two patient groups (mean: FTD 63.2 years, AD 67.5 years; $t_{26} = 1.37$, $p > .05$), as was years of education (mean: FTD 12.7 years, AD 11.4 years; $t_{25} = 1.14$, $p > .05$). The ACE-R scores were similar across the two patient groups (FTD: $x = 79.6$, $SD = 14.4$; AD: $x = 77.9$, $SD = 7.6$; Mann–Whitney $U = 46$, $p > .05$), and while the clinical dementia rating (CDR) was slightly higher in the FTD group, the difference was not significant (mean: FTD 0.91, AD 0.75; $t_{19} = 0.91$, $p > .05$).

Activities of daily living (see Table 1)

On the DAD, both groups scored worse than predicted premorbid levels, but FTD patients were worse than those with AD on total DAD (Mann–Whitney $U = 48$, $p < .05$), and the execution subscores (Mann–Whitney $U = 48$, $p < .05$). Initiation and planning subscores were worse in FTD patients, but failed to reach conventional levels of significance (initiation, planning: Mann–Whitney $U = 61$ and 61.5, respectively, $p < .1$ for both).

Emotion recognition scores (see Figure 2)

In a repeated measures ANOVA (Groups: FTD, AD, controls) × (Levels: positive or negative emotion scores) there was an overall effect of

TABLE 1
Demographic details (mean; standard deviation in parentheses) and performance on cognitive (ACE-R), functional (CDR) and ADL assessment (DAD, including DAD subscores). Median; 25th and 75th percentiles are indicated in parentheses

Demographics and test scores (maximum scores)	*AD*	*FTD*
Age	68 (9)	63 (8)
Education in years	11 (2)	13 (14)
ACE-R (100)	74 (72 – 84)	83 (76 – 90)
CDR (3.0)	1.0 (0.5–1.0)	1.0 (0.5–1.0)
DAD total (100)	90 (81–98)	76* (42–92)
Initiation (100)	92 (80–100)	83 (39–94)
Planning (100)	88 (68–100)	73 (30–89)
Execution (100)	93 (82–96)	79* (60–89)

Mann–Whitney *U*-test, two-tailed. *$p < .05$.

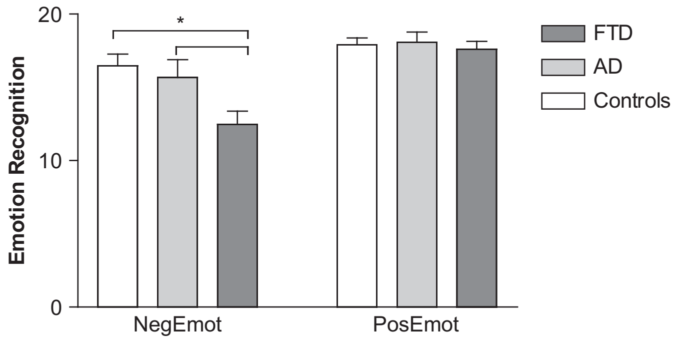

Figure 2. FTD patients are disproportionately bad at recognising negative emotions compared with both AD and controls. NegEmot, negative emotions (anger, fear, sadness, disgust scaled combined score), PosEmot (happiness, surprise scaled combined score). $*p < .01$ for interaction.

emotion level ($F(1, 33) = 33.5$, $p < .001$); negative emotions were harder to recognise than positive ones. The FTD group performed worse than the AD group and controls (mean: 15.0 vs. 16.9 and 17.2; ($F(2, 33) = 3.65$, $p < .05$) but there was no significant difference between controls and the AD group. There was also an interaction ($F(2, 33) = 5.94$, $p < .01$). *Posthoc* pairwise *t*-tests showed that the source of this interaction was that FTD patients were markedly worse at detecting negative emotions than both AD patients and controls.

Cambridge behavioural inventory (see Figure 3)

On the four subdomains of the CBI, there were marked differences, as expected, between groups: carers of FTD patients endorsed far higher levels of three of the four domains than carers of AD patients (disinhibition, challenging behaviour and motivation).

Correlation analyses

Emotion recognition and ADLs (see Figure 4A)

There was no relationship between emotion recognition performance and scores (or subscores) of the DAD for either FTD or AD patients.

ADLs and CBI subscores (see Figure 4B)

In contrast, when correlating scores on the CBI and the DAD, FTD patients showed a strong negative linear relationship between CBI-motivation subscores and total score on the DAD (*Kendall b tau* = –0.51, $p < .05$) i.e., the higher the endorsement of poor motivation, the worse the ability to perform ADLs. Similar results were obtained for the correlation with two DAD subscores (initiation $tau_b = -0.52$, $p < .05$; planning $tau_b = -0.61$, $p < .05$).

In AD patients, there was a strong correlation between the level of challenging behaviour as assessed by carers (CBI-behaviours), and the overall

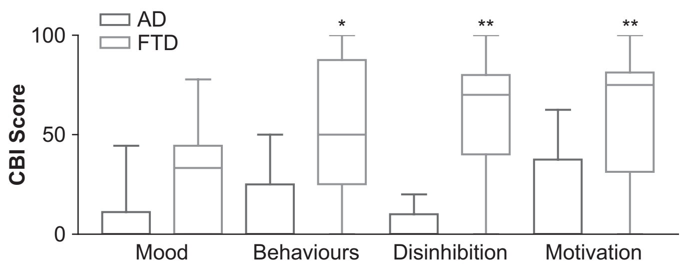

Figure 3. Graph showing CBI score in each domain for AD and FTD patients. The central bar indicates the median, the box the interquartile range and the error bars show the range of values. AD patients have far less behavioural disturbance than FTD patients as would be expected. Challenging behaviours, disinhibition and motivation are significantly more common in FTD. $*p < .05$, $**p < .01$.

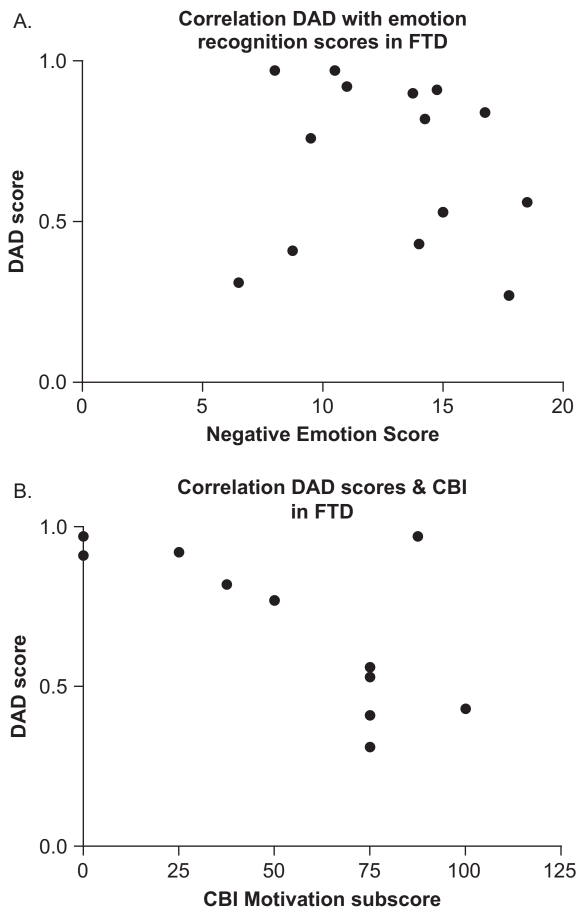

Figure 4. (A) No correlation is seen between DAD scores and performance on emotion recognition tests. (B) There is an inverse correlation in FTD patients between endorsements by carers on the CBI, and the level of impairment as assessed using the DAD (*Kendall b tau* = –0.51, $p < .05$), i.e., as the degree of apathy increases, the worse the performance becomes on ADLs.

ability to perform ADLs ($tau_b = -0.62$, $p < .05$). The initiation, planning and performance subscores on the DAD all correlated strongly with the CBI-behaviours rating (tau_b = –0.63, –0.59, –0.66, respectively, all $p < .05$). The DAD-performance subscore correlated strongly with the CBI-mood ratings ($tau_b = -0.64$, $p < .05$) and CBI-motivation ratings predicted DAD-initiation subscores ($tau_b = -0.64$, $p < .05$).

Emotion recognition and CBI

In FTD patients, CBI-mood subscores correlated strongly with emotion recognition performance ($N = 10$, positive emotions: *Kendall b tau* = 0.53, $p < .05$; negative emotions: $tau_b = 0.73$, $p < .01$). There were insufficient numbers of AD patients with data on both of these measures ($N = 4$) to perform any meaningful analysis.

DISCUSSION

This study highlights several interesting relationships. FTD patients were worse than AD patients and controls at recognizing emotion in others, particularly negative emotions. They were also markedly impaired in their ability to perform activities of daily living (ADLs) relative to the AD group, mainly with respect to the execution of these activities. Despite impaired performance on these aspects of social functioning, there was no clear relationship between the two. In fact, performance of ADLs in FTD was more strongly associated with motivation than emotion processing.

The finding of impaired emotional processing in FTD confirms prior reports (Fernandez-Duque and Black, 2005; Keane et al., 2002; Lavenu et al., 1999; Lough et al., 2006; Sturm et al., 2006) and contrasts with the normal performance of the AD subgroups. Studies of emotion processing in AD have reported variable results, finding that some have normal and others mildly impaired recognition of face emotions, perhaps related to the severity of the degeneration or the sensitivity of the tasks employed (Hargrave, Maddock, & Stone, 2002; Kohler et al., 2005; Lavenu & Pasquier, 2005; Lavenu et al., 1999; Luzzi, Piccirilli, & Provinciali, 2007).

The deficits in FTD patients were not simply related to severity of disease. While it is often difficult to match patients across neurodegenerative diagnoses, in this study the two patient groups had similar ACE and MMSE scores, and were judged by their carers to be as affected as each other on the CDR. Qualitatively, there were differences between the two groups: FTD patients had a lower level of performance of ADLs, which appeared to relate largely to their level of apathy or lack of motivation. In AD, the ability to perform ADLs was most compromised by the level of challenging behaviours present.

Another interesting finding was the correlation between the mood ratings on the CBI and emotion recognition performance. In other words, the presence of greater emotional reactivity in FTD patients predicted a better ability to judge emotional states of others. This is broadly supportive of the hypothesis that the same neural substrates are utilized for both recognition of emotion in others, and experience of emotion in oneself.

Our study has some limitations. The number of patients is relatively small considering the number of variables we considered. In view of the fact that this was a largely exploratory study, designed to generate hypotheses for the future, more detailed, study we did not correct our results for multiple comparisons.

Social dysfunction in FTD appears to be multifactorial. Impairments in emotion processing, particularly negative emotions, may cause FTD patients to be insensitive to social cues and unable to respond to signals of social disapproval. This deficit may engender a lack of consideration for the emotional concerns of others (lack of empathy), or situations that embarrass them. Decision-making may also be affected by the inability to use emotional cues to bias behaviour in social situations. Despite this, impaired emotion does not seem to explain the inability to perform ADLs in FTD, which seems to be more related to motivational factors. These motivational factors in turn were not linked to the ability to judge facial emotion.

The profound social disability of patients with FTD is highlighted by the significant carer burden and impairment in abilities of self-care compared with AD patients who have a similar level of cognitive performance. It is important to understand and appreciate the nature of the social dysfunction in these patients so that we can improve the advice we give to them and their families, and ensure that our interventions (therapeutic or behavioural) are effective.

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