Are you really angry? The effect of intensity on facial emotion recognition in frontotemporal dementia

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Are you really angry? The effect of intensity on facial emotion recognition in frontotemporal dementia

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Frontotemporal dementia (FTD) is a neurodegenerative brain disorder that affects the frontal and temporal lobes predominantly. Impaired emotion recognition has been reported in two FTD subtypes: behavioral-variant FTD (bvFTD) and semantic dementia (SD), but has not been investigated in the third subtype: progressive nonfluent aphasia (PNFA).

Methods: Recognition of six basic facial emotions (anger, disgust, fear, sadness, surprise, and happiness) was investigated in 41 FTD patients (bvFTD = 16; SD = 12; PNFA = 13) and 37 age- and education-matched controls, using two tests. In one task, intensity of emotional expression was increased to identify cognitive components contributing to emotion recognition performance.

Results: All patient groups demonstrated impaired overall facial emotion recognition compared to controls. Performance, however, improved with increased emotion intensity in bvFTD and PNFA groups, the effect of intensity on emotion recognition being particularly pronounced for negative emotions. In contrast, increased intensity of facial emotion did not change performance in SD.

Conclusions: Patients with SD demonstrate a primary emotion processing impairment, whereas PNFA and bvFTD patients’ emotional disturbance is in part mediated by attentional deficits. These findings indicate that a subset of FTD patients may benefit from enhanced emotional intensity that will facilitate facial emotion recognition.

Keywords: Caricatures; Behavioral-variant FTD; Semantic dementia; Progressive nonfluent aphasia.
CLINICAL PRESENTATION OF FTD

Frontotemporal dementia is the second most common neurodegenerative disorder affecting individuals under the age of 65 (Ratnavalli, Brayne, Dawson, & Hodges, 2002). Three clinical subtypes are generally described: behavioral-variant FTD (bvFTD), semantic dementia (SD), and progressive nonfluent aphasia (PNFA) (Hodges & Patterson, 2007; Neary et al., 1998; Piguet, Hornberger, Mioshi, & Hodges, 2011; Rascovsky et al., 2007). Diagnosis is based on the predominant clinical features at initial presentation, although the subtypes tend to merge with disease progression (Gorno-Tempini et al., 2011; Kertesz, McMonagle, Blair, Davidson, & Munoz, 2005). Early presentation in bvFTD is characterized by changes in personality and behavior, and executive dysfunction on cognitive testing. In contrast, the primary presenting features in SD and PNFA are language disturbances: a fluent speech output characterized by a lack of content words secondary to loss of semantic knowledge in SD (Hodges & Patterson, 2007), and nonfluent, effortful, and labored speech with relatively well-preserved comprehension in PNFA (Neary et al., 1998).

Cognitive and behavioral changes tend to reflect the pattern of underlying brain pathology. In bvFTD, greatest atrophy is observed in the frontal lobes bilaterally, most particularly affecting the orbitofrontal and medial frontal cortices (Seeley et al., 2008). In SD, marked atrophy is present in the anterior temporal lobe. This atrophy is typically asymmetrical, more commonly greater on the left than on the right (Hodges, Patterson, Oxbury, & Funnell, 1992). In PNFA, atrophy is less widespread and typically constrained to the inferior posterior frontal region (i.e., Broca’s area) and the insula (Nestor et al., 2003).

A range of deficits in social cognition, including emotion recognition, is present in FTD (Kipps & Hodges, 2006; Lavenu et al., 1999; Lough et al., 2006). Patients with FTD tend to show greater difficulty in recognizing facial emotions than both healthy controls and patients with Alzheimer’s disease. Findings are, however, somewhat inconsistent: Some studies report a recognition deficit that is specific to negative emotions (e.g., anger, disgust, fear, sadness) (Kessels et al., 2007), although others have found impaired recognition of both positive (e.g., happiness, surprise) and negative facial expressions (Bediou et al., 2009; Snowden et al., 2008). In these studies, however, FTD was investigated as a single disease entity, and FTD subtypes were not analyzed separately, and this may account for the disparity in results reported across studies.

NEURAL SUBSTRATES OF EMOTION RECOGNITION IN FTD

Ekman and colleagues proposed a small set of “basic” emotions (anger, disgust, fear, sadness, surprise, and happiness), each characterized by a distinct facial expression, physiology, and evolutionary history (Ekman, 1992; Ekman, Friesen, & Ellsworth, 1972). Subsequent lesion and functional neuroimaging studies have demonstrated specific neural substrates that underlie the recognition of these basic emotions (e.g., Calder, Keane, Lawrence, & Manes, 2004; Calder, Keane, Manes, Antoun, & Young, 2000a; Calder et al., 1996; Ekman, 1992; Kesler West et al., 2001; LaBar, Gatenby, Gore, LeDoux, & Phelps, 1998). These results led to the hypothesis that specific emotions are processed by discrete neural substrates, referred to as the multimodal system model of emotion recognition. To date, specific neural substrates have been identified for a number of basic emotions, with the amygdala associated with fear recognition (Adolphs, Tranel, Damasio, & Damasio, 1995; Allman & Brothers, 1994), the insula associated with disgust recognition (Calder et al., 2000a; Phillips et al., 1997, 1998; Sprengelmeyer et al., 1996), and the ventral striatum associated with anger recognition (Blair, Morris, Frith, Perrett, & Dolan, 1999; Calder et al., 2004).

The pattern of emotion recognition deficits in subtypes of FTD, however, matches only loosely what would be expected from predictions of the multimodal system model of emotion recognition. For example, the consistent impairment of anger recognition in bvFTD (Keane, Calder, Hodges, & Young, 2002; Kipps, Nestor, Acosta-Cabronero, Arnold, & Hodges, 2009b; Lough et al., 2006; Rosen et al., 2004) fits the predominant atrophy observed in the orbitofrontal and anterior cingulate cortices, which play an important role in processing this type of emotional stimulus. Nevertheless, these patients’ impairment is not emotion specific, as deficits in recognizing other emotions are also reported (Keane et al., 2002; Rosen et al., 2004). Similarly, despite the significant amygdala atrophy observed in SD, patients exhibit a broad deficit in recognizing negative emotions, rather than a recognition deficit limited to fear alone (Rosen et al., 2004; Rosen et al., 2002b). Emotion recognition in PNFA has been investigated in one study only (Rankin et al., 2009), and a specific or disproportionate impairment in recognition of disgust has not been reported.

The discrepancy between the multimodal system model predictions and current findings in FTD may be due to factors other than deficits in emotion processing. Patients with FTD may perform poorly because of
perceptual or attentional difficulties that interfere with emotion processing, resulting in pervasive rather than specific deficits. Despite an increased interest in this domain, understanding of the mechanisms underlying facial emotion recognition remains relatively limited.

The goal of this investigation was to identify the mechanisms contributing to emotion processing disturbance in FTD. In this study, we manipulated the perceptual and attentional demands by exaggerating the emotional expression of the stimuli in one of the facial emotion-recognition tasks. This process is achieved by digitally manipulating the position of numerous geographic points of difference, between an emotional and a neutral facial expression (e.g., corner of the mouth, eyebrow frown, eye opening) (Calder et al., 2000b). In this way, the intensity or salience of the emotional expression can be either enhanced (known as caricatures) or degraded (known as anti-caricatures). Previous studies have found that caricatured emotional expressions are more quickly identified (Calder, Young, Rowland, & Perrett, 1997), are rated as more intensely expressing the emotion (Benson, Campbell, Harris, Frank, & Tovée, 1999; Calder et al., 2000b), and are identified as well as, or better than, non-exaggerated “real-life” expressions. Increased intensity of facial expressions has also been associated with activation of the same specific neural regions associated with recognition of real-life emotions, and this activation amplifies with increasing intensity (Morris et al., 1998; Phillips et al., 1997). Thus, the main hypothesis of this study was that, in some instances, the use of caricatures would improve emotion recognition by reducing perceptual and attentional task demands.

In addition, how emotion recognition deficits vary across FTD subtypes is not well specified. The second aim of this study was to investigate facial emotion recognition in PNFA, which has not been extensively examined to date, and compare performance with bvFTD and SD profiles. It was hypothesized that the deficits observed in these groups would be consistent with their predominant regions of atrophy. Hence anger recognition was expected to be impaired in bvFTD, fear recognition impaired in SD, and disgust recognition impaired in PNFA.

METHODS

Participants
Forty-one patients meeting clinical diagnostic criteria for FTD (bvFTD = 16; SD = 12; PNFA = 13) were recruited from the Frontotemporal Dementia Research Group in Sydney. All patients were seen by the same experienced behavioral neurologist (J.R.H), and diagnosis was based on clinical assessment, comprehensive neuropsychological assessment, and presence of brain atrophy on structural magnetic resonance imaging (MRI). Patients presenting with behavioral features of bvFTD in the absence of brain atrophy or progression, also known as phenocopy bvFTD, were excluded (Davies et al., 2006). Three SD patients with predominant right-sided temporal lobe atrophy were also excluded, as this small number of cases prevented separate analyses for this group. Within the PNFA group, patients exhibiting impaired repetition and comprehension for sentences and reduced word span were excluded. This presentation, known as logopenic progressive aphasia, is strongly associated with Alzheimer’s disease (Gorno-Tempini et al., 2008). Other exclusion criteria included presence of other types of dementia or other neurological disease; diagnosis of major depression, schizophrenia, obsessive compulsive disorder or substance abuse; and Mini-Mental State Examination (MMSE) score below 19/30.

Thirty-seven healthy, age- and education-matched controls were recruited from the patients’ families and friends, or from the local area. Participants were excluded if any of the following criteria were present: significant history of psychiatric or neurological condition, history of substance abuse or medications with CNS side effects, and MMSE score below 27/30.

The Southeastern Sydney and Illawarra Area Health Service and the University of New South Wales ethics committees approved the study. Participants, or their person responsible, provided informed consent. All participants volunteered their time but were reimbursed for travel costs.

Materials and procedure

Ekman 60 Task

This task assesses recognition of 60 facial expressions across six basic emotions (anger, disgust, fear, happiness, sadness, and surprise), using stimuli from the Pictures of Facial Affect series (Ekman & Friesen, 1976). These images are not manipulated, and emotion is expressed at a normal or unchanged level of intensity, referred to here as 100% intensity. Images are pseudorandomly presented on a computer screen, and participants view each stimulus for 5 s and select the label that best describes the emotional expression.
The labels are present throughout testing, and selection is untimed. Participants respond either by using the mouse to click the appropriate label, pointing to the label, or saying their response (depending on their preference) for the researcher to record. No feedback on response accuracy is provided.

**Ekman Caricatures**

This task is similar to the Ekman 60 with the same procedure, but uses only two models (one male, one female). In this task, faces have been digitally manipulated to increase the intensity of emotional expression by altering critical facial features (see Appendix 1 for example stimuli) (Young, Perrett, Calder, Sprengelmeyer, & Ekman, 2002). In total, 48 images are presented across the same six basic emotions. These emotions vary across four levels of intensity, with emotional expression increased from 100% emotional expression by +15%, +30%, +50%, or +75%.

Hence, comparison of overall performance between the Ekman 60 and the Ekman Caricatures will identify the contribution of attentional and perceptual processes to emotion recognition. Investigations of each emotion in the Ekman Caricatures separately will identify the effect of manipulation of intensity on recognition of specific emotions.

**Neuropsychological and behavioral measures**

In addition to the MMSE (Folstein, Folstein, & McHugh, 1975) and the Addenbrooke’s Cognitive Examination-Revised (ACE-R) (Mioshi, Dawson, Mitchell, Arnold, & Hodges, 2006), all participants were given tests of attention and working memory (Digit Span subtest of the Wechsler Adult Intelligence Scale) (Wechsler, 1997), semantic knowledge (Boston Naming Test, 15-item version) (Mack, Freed, White Williams, & Henderson, 1992), processing speed (Trail Making Test) (Tombaugh, 2004), verbal fluency (Controlled Oral Word Association Test (COWAT) (Spreek & Strauss, 1998), and inhibition of prepotent response (Hayling Test) (Burgess & Shallice, 1997) (Table 2). These tests were administered as part of a larger evaluation of cognitive functions.

**Statistical analyses**

Data were analyzed by PASWS 19.0 (IBM, Inc., Chicago, IL, USA). First, variables were plotted and checked for normal distribution by Kolmogorov-Smirnov tests. Given the non-normal distribution of the individual emotion scores on the Ekman 60 and the Ekman Caricatures, these variables were analyzed by nonparametric tests.

Between-group comparisons for sociodemographic and neuropsychological tests were performed by univariate analysis of variance (ANOVA) followed by post-hoc tests where appropriate. The effect of intensity on overall emotion recognition was investigated with a $2 \times 4$ repeated-measures ANOVA with Intensity (Ekman 60, Ekman Caricatures) as the within-subjects variable and Diagnosis (control, bvFTD, SD, PNFA) as the between-subjects variable. Follow up $t$-tests comparing each group’s performance on the Ekman Caricatures with the Ekman 60 performance were then conducted. Between-group comparisons for the six basic emotions on the Ekman 60, and the Ekman Caricatures were investigated by Kruskal–Wallis analyses of variance, followed by post-hoc Mann–Whitney tests comparing each patient group with controls. Finally, to investigate the effect of emotion intensity, an index of change was calculated as based on the difference in performance between +15% and +75% for each basic emotion. Again, between-group differences were investigated by Kruskal–Wallis analyses of variance for each emotion, followed by post-hoc Mann–Whitney tests comparing each patient group with controls. Statistical significance was set at .05, and all analyses were corrected for multiple comparisons where appropriate.

**RESULTS**

**Demographic data**

Groups were well matched for age, $F(3, 72) = 2.26, p = .09$, and level of education, $F(3, 72) = .75, p = .53$. Within the FTD subgroups, disease duration differed significantly, $F(2, 38) = 4.74, p = .02$, with longer duration observed in the SD than PNFA group ($p = .01$). This difference probably reflects the reported longer time to diagnosis from disease onset in SD patients (Table 1).

**Neuropsychological performance**

All patient groups were significantly impaired on both cognitive screening measures (MMSE and ACE-R), compared to controls (Table 2). Across patient groups, PNFA performed significantly worse than bvFTD on the MMSE ($p = .046$), and SD performed significantly worse than bvFTD ($p < .001$) and PNFA ($p = .01$).
TABLE 1
Demographic characteristics of the study sample

<table>
<thead>
<tr>
<th></th>
<th>Controls</th>
<th>bvFTD</th>
<th>SD</th>
<th>PNFA</th>
<th>F</th>
<th>Patient vs. controls</th>
<th>Post-hoc</th>
<th>Between subtypes</th>
</tr>
</thead>
<tbody>
<tr>
<td>n</td>
<td>37</td>
<td>16</td>
<td>12</td>
<td>13</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male/female</td>
<td>24/13</td>
<td>12/4</td>
<td>9/3</td>
<td>9/4</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age</td>
<td>64.6 (4.5)</td>
<td>61.5 (9.7)</td>
<td>62.4 (8.8)</td>
<td>65.5 (11.4)</td>
<td>0.9</td>
<td>N/A</td>
<td>N/A</td>
<td></td>
</tr>
<tr>
<td>Education (years)</td>
<td>13.8 (2.5)</td>
<td>12.1 (3.3)</td>
<td>12.5 (3.4)</td>
<td>13.0 (3.3)</td>
<td>1.4</td>
<td>N/A</td>
<td>N/A</td>
<td></td>
</tr>
<tr>
<td>Disease duration (months)</td>
<td>–</td>
<td>42 (24.2)</td>
<td>58 (25.5)</td>
<td>30 (16.1)</td>
<td>4.7*</td>
<td>SD &gt; PNFA*</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Notes: Scores are means (SD); *p < .05. N/A = not applicable.

TABLE 2
Neuropsychological variables across healthy controls and three FTD subgroups

<table>
<thead>
<tr>
<th></th>
<th>Controls</th>
<th>bvFTD</th>
<th>SD</th>
<th>PNFA</th>
<th>F</th>
<th>Impaired compared to controls</th>
<th>Post-hoc</th>
<th>Between subtypes</th>
</tr>
</thead>
<tbody>
<tr>
<td>MMSE</td>
<td>29.4</td>
<td>26.8</td>
<td>24.0</td>
<td>23.5</td>
<td>22.4</td>
<td>bvFTD*</td>
<td>bvFTD &gt; PNFA*</td>
<td></td>
</tr>
<tr>
<td></td>
<td>(0.9)</td>
<td>(2.6)</td>
<td>(3.1)</td>
<td>(5.1)</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>ACE-R</td>
<td>95.6</td>
<td>82.8</td>
<td>59.7</td>
<td>72.1</td>
<td>52.6</td>
<td>bvFTD**</td>
<td>bvFTD &gt; SD**</td>
<td></td>
</tr>
<tr>
<td></td>
<td>(3.0)</td>
<td>(6.8)</td>
<td>(11.7)</td>
<td>(18.3)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Digit Span Forwards</td>
<td>7.0</td>
<td>5.6</td>
<td>6.3</td>
<td>4.4</td>
<td>14.3</td>
<td>bvFTD*</td>
<td>SD &gt; PNFA**</td>
<td></td>
</tr>
<tr>
<td></td>
<td>(1.3)</td>
<td>(0.7)</td>
<td>(1.3)</td>
<td>(1.3)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Digit Span Backwards</td>
<td>5.5</td>
<td>4.1</td>
<td>4.5</td>
<td>3.2</td>
<td>15.6</td>
<td>bvFTD**</td>
<td>SD &gt; PNFA*</td>
<td></td>
</tr>
<tr>
<td></td>
<td>(1.3)</td>
<td>(0.8)</td>
<td>(1.1)</td>
<td>(0.9)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Boston – 15</td>
<td>14.7</td>
<td>13.6</td>
<td>2.4</td>
<td>11.4</td>
<td>181.4</td>
<td>SD**</td>
<td>bvFTD &gt; SD**</td>
<td></td>
</tr>
<tr>
<td></td>
<td>(0.6)</td>
<td>(1.4)</td>
<td>(1.7)</td>
<td>(3.3)</td>
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<tr>
<td>Trails A</td>
<td>32.4</td>
<td>59.6</td>
<td>38.7</td>
<td>63.5</td>
<td>6.2</td>
<td>bvFTD**</td>
<td>ns</td>
<td></td>
</tr>
<tr>
<td></td>
<td>(12.7)</td>
<td>(38.9)</td>
<td>(18.0)</td>
<td>(45.6)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Trails B</td>
<td>81.9</td>
<td>145.1</td>
<td>107.3</td>
<td>142.7</td>
<td>6.3</td>
<td>bvFTD**</td>
<td>ns</td>
<td></td>
</tr>
<tr>
<td></td>
<td>(31.9)</td>
<td>(88.2)</td>
<td>(47.0)</td>
<td>(66.8)</td>
<td></td>
<td></td>
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<td></td>
</tr>
<tr>
<td>COWAT</td>
<td>44.2</td>
<td>24.5</td>
<td>25.2</td>
<td>16.8</td>
<td>21.7</td>
<td>bvFTD**</td>
<td>ns</td>
<td></td>
</tr>
<tr>
<td></td>
<td>(12.1)</td>
<td>(11.9)</td>
<td>(10.9)</td>
<td>(12.5)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hayling*</td>
<td>6.4</td>
<td>3.0</td>
<td>1.0</td>
<td>3.0</td>
<td>28.9</td>
<td>bvFTD**</td>
<td>ns</td>
<td></td>
</tr>
<tr>
<td></td>
<td>(.7)</td>
<td>(.3)</td>
<td>(.0)</td>
<td>(.25)</td>
<td></td>
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</tr>
</tbody>
</table>

Notes: Scores are mean (SD); *p < .05, **p < .01; ns: not significant. MMSE (Mini-Mental State Examination): total score /30; ACE-R (Addenbrooke’s Cognitive Examination-Revised): total score /100; Digit Span Forwards and Digit Span Backwards: maximum span raw score; Boston – 15 total: total score /15; Trails A and Trails B: time to complete (seconds); COWAT (Controlled Oral Word Association Test): total number of words produced (F, A, S); Hayling: overall scaled score /10 (average score = 6). * Only two SD and six PNFA patients were assessed on this task.

on the ACE-R, reflecting the relatively high language demands of this test. Significant differences between patient groups and controls were also observed on most cognitive tasks. Post-hoc tests showed that the cognitive profiles and deficits were consistent with the clinical presentation generally associated with each of the subtypes (Table 2). The bvFTD group was significantly impaired on tasks of attention (Digit Span Forwards), working memory (Digit Span Backwards), and executive functioning (COWAT and Hayling), whereas the SD group was significantly impaired on the semantic task (Boston Naming Test – 15) and two language-based executive functioning tasks (COWAT and Hayling) compared to controls, bvFTD and PNFA groups (all ps < .001). The PNFA group demonstrated impaired performance on all neuropsychological tests compared to controls and showed variable performance compared to the other FTD subgroups, performing poorer than bvFTD on the MMSE, ACE-R, and Boston Naming Test – 15, and worse than the SD group for Digit Span Forwards and Backwards. These deficits likely reflect the overall reduced verbal output in these patients compared to the other patient groups (Table 2).
Overall emotion recognition

A significant interaction between Diagnosis and Intensity was present, \( F(3, 73) = 3.52, p = .019 \), indicating that the effect of intensity differed across the various diagnostic groups. A main effect of Diagnosis \( F(3, 73) = 19.211, p < .001 \), was significant, with all patient groups performing worse than controls (all \( ps < .001 \)) and a trend for the SD group to perform worse than the bvFTD group (\( p = .058 \)). A significant main effect of Intensity was observed, with performance better on the Caricatures than on the Ekman 60 task \( F(1, 73) = 50.80, p < .001 \). Post-hoc \( t \)-tests indicated that increased intensity on the Caricatures task significantly improved emotion recognition for control (\( p < .001 \)), bvFTD (\( p < .001 \)), and PNFA (\( p = .006 \)), but not SD (\( p > .05 \)) groups (Figure 1).

Recognition of basic emotions: Ekman 60

An overall effect of group was present for each of the four negative emotions on the Ekman 60 task (anger: \( H(3) = 22.556, p < .001 \); disgust: \( H(3) = 24.497, p < .001 \); fear: \( H(3) = 14.937, p = .002 \); and sadness: \( H(3) = 14.713, p = .002 \)), but not for the positive emotions (Figure 2). Post-hoc tests indicated that bvFTD performed below controls for all negative emotions (anger: \( U = 123.0, z = -3.172, p = .002 \); disgust: \( U = 125.5, z = -3.152, p = .002 \); fear: \( U = 122.0, z = -3.161, p = .002 \); and sadness: \( U = 158.0, z = -2.466, p = .014 \)), as did the SD group (anger: \( U = 76.0, z = -3.448, p = .001 \); disgust: \( U = 29.0, z = -4.580, p < .001 \); fear: \( U = 113.0, z = -2.552, p = .011 \); and sadness: \( U = 108.0, z = -2.720, p = .007 \)). The PNFA group performed below controls for anger: \( U = 86.5, z = -3.458, p = .001 \); fear: \( U = 129.5, z = -2.473, p = .013 \); and sadness: \( U = 106.5, z = -3.043, p = .002 \), but not for disgust.

Recognition of basic emotions: Ekman Caricatures

On the Caricatures task, a similar profile of performance emerged with an overall effect of group observed for negative emotions only (anger: \( H(3) = 22.738, p < .001 \); disgust: \( H(3) = 31.242, p < .001 \); fear: \( H(3) = 23.582, p < .001 \); and sadness: \( H(3) = 23.595, p < .001 \)) (Figure 2). Post-hoc tests indicated that the bvFTD group performed worse than controls for recognition on all negative emotions, collapsed across all intensity levels (anger: \( U = 123.0, z = -3.172, p = .002 \); disgust: \( U = 183.0, z = -2.491, p = .013 \); fear: \( U = 134.5, z = -4.140, p < .001 \); and sadness: \( U = 137.5, z = -3.428, p = .001 \)). SD patients showed a similar pattern of performance (anger: \( U = 44.0, z = -4.553, p < .001 \); disgust: \( U = 33.0, z = -5.178, p < .001 \); fear: \( U = 65.0, z = -3.767, p < .001 \); and sadness: \( U = 51.5, z = -4.375, p < .001 \)). The PNFA group’s performance was worse than controls for anger: \( U = 141.5, z = -2.517, \)

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**Figure 1.** Percentage of correctly recognized emotions on the Ekman 60 and Ekman Caricatures tasks in healthy controls and FTD subtypes. Error bars represent SEM.

**Figure 2.** Percentage of correctly recognized emotions on the Ekman 60 (A) and Ekman Caricatures (B) for each of the six basic emotions (Ang: anger, Dis: disgust, Fea: fear, Sad: sadness, Sur: surprise, and Hap: happiness) in healthy controls and FTD subtypes. Errors bars represent SEM.
Figure 3. Percentage of correctly recognized emotions (anger, disgust, fear, sadness, surprise, and happiness) across the four levels of intensity in healthy controls and FTD subtypes.

\( p = .012; \) disgust: \( U = 147.5, z = -2.202; \) and sadness: \( U = 142.5, z = -2.497, p = .013, \) but not fear.

**The effect of intensity on emotion recognition**

Examination of change scores for each negative emotion, revealed an overall effect of group for disgust: \( H(3) = 11.013, p = .012; \) fear: \( H(3) = 15.345, p = .002; \) and sadness: \( H(3) = 10.348, p = .016, \) indicating that, for these three emotions, change in performance from low to high intensity differed across groups (Figure 3). In contrast, change in performance for anger stimuli did not differ across groups. Post-hoc tests showed that bvFTD performance was not significantly different from controls for these three basic emotions, indicating that bvFTD patients benefited from increasing intensity to a similar degree as controls. In contrast, the SD group’s change score was significantly less than controls for disgust (\( U = 108.0, z = -1.594, p = .001 \)), fear (\( U = 73, z = -3.731, p < .001 \)), and sadness (\( U = 113.5, z = -2.926, p = .003 \)).
these emotions in the previous analyses. Change scores for positive emotions are not reported given the absence of between-group differences for these emotions in the previous analyses.

DISCUSSION

This study sought to explore emotion recognition performance in FTD and to identify the mechanisms responsible for impaired performance in each FTD subtype by modulating the emotional intensity of the stimuli. This study reveals a number of novel findings regarding emotion processing in the three subtypes of FTD, findings that have clinical implications for the management of these patients. Using two tasks of facial emotion recognition, our study revealed significant impairment in emotion recognition in all three subtypes of FTD, including in PNFA, indicating that contrary to current assumptions, PNFA patients do have demonstrable deficits in emotion recognition. Results from this study further illustrated an effect of emotion intensity on performance accuracy. Importantly, the performance improvement observed with increased intensity of emotion was present in bvFTD and PNFA groups only and was absent in SD, indicating differences in the mechanisms underlying emotion-detection deficits across subtypes of FTD.

Profiles of emotion-recognition deficits

We predicted that deficits in emotion recognition would differ across FTD subtypes, the deficits reflecting the predominant locus of brain atrophy of each subtype (i.e., impaired anger recognition in bvFTD, impaired fear recognition in SD, and impaired disgust recognition in PNFA). Contrary to our predictions, deficits in emotion recognition were more widespread than expected.

Overall, bvFTD patients were impaired for recognition of all negative emotions, including anger, whether intensity of emotion was modulated or not, a finding in keeping with previous studies (Kipps, Mioshi, & Hodges, 2009a; Kipps et al., 2009b; Lough et al., 2006). Impaired recognition of anger in patients with bvFTD is consistent with the predictions of the model and reflects the orbitofrontal cortex atrophy reported in these patients (Krill, Macdonald, Patel, Png, & Halliday, 2005; Piguet et al., 2011; Seeley et al., 2008). Although impairment in the recognition of sadness, fear, and disgust, was not predicted, these emotion-recognition deficits can be accounted for by the multimodal system model, in that the discrete neural substrates necessary for these basic emotions undergo pathological changes in bvFTD. Preserved ability to recognize sadness has been associated with integrity of the subcallosal cingulate cortex (Phan, Wager, Taylor, & Liberzon, 2002). Atrophy in this region has been implicated in the disturbance of empathy (Rankin et al., 2006) and is the site of significant neuropathological changes in bvFTD (Schroeter, Raczka, Neumann, & Yves von Cramon, 2008; Seeley et al., 2008). Similarly, atrophy of the anterior insula, which is involved in disgust recognition, has been shown to be affected to some extent in bvFTD (Rosen et al., 2002a; Schroeter et al., 2008; Seeley, 2010; Seeley et al., 2008). Atrophy of the amygdala is also present in bvFTD (Barnes et al., 2006; Brambati et al., 2007) and most likely contributes to the reduced fear recognition in this group.

In SD, a severe impairment in fear recognition was observed with other negative emotions (anger, sadness, and disgust) affected to a lesser degree. This disproportionate impairment of fear is consistent with the multimodal system theory’s prediction of reduced fear recognition in the presence of anterior temporal lobe, particularly amygdala, atrophy (Adolphs et al., 1995; Ekman, 1992). Recognition of other negative emotions, however, was also impaired. This finding confirms prior reports of a global impairment for recognition of negative emotions in SD (Rosen et al., 2002b, 2004). Although it is possible that these patients performed poorly due to loss of semantic knowledge or difficulty in reading the label, this explanation does not fully account for their performance. These patients performed above chance for each of the emotions, indicating sufficient word knowledge to perform the task to some extent. Thus, the observed pattern of deficits more likely reflects a profound involvement of the amygdala in processing emotion. Theorists suggest the amygdala may be essential for processing ambiguous stimuli (Whalen, 1999), or for processing emotions related to behavioral withdrawal (Anderson, Spencer, Fulbright, & Phelps, 2000). With disease progression, atrophy in SD extends beyond the anterior temporal region to involve other brain regions necessary for recognition of other basic emotions (such as the insula, and striatal regions). Although the SD group included in this study was relatively mild, presence of atrophy in other regions cannot be discounted.

Contrary to the prediction of the model, a disproportionate impairment of recognition of disgust was not observed in PNFA at 100% intensity. In fact, recognition of disgust in PNFA was similar to controls. Although the insula has been implicated
in disgust recognition, only some regions of the insula are involved in emotion processing, with other areas specialized for other functions such as speech production (Ackermann & Riecker, 2004; Adolphs, 2002a). It is plausible that in PNFA, regions necessary for speech are disproportionately damaged compared to those areas necessary for recognition of disgust. Alternatively, some researchers have suggested that the insula may be necessary not only for processing of disgust but also for interoceptive awareness and the processing of “physical emotions” (Craig, 2009; Critchley, Wiens, Rotshtein, Öhman, & Dolan, 2004; Damasio et al., 2000). Consistent with this hypothesis, the PNFA group did demonstrate impaired recognition of anger, sadness, and fear at 100% intensity.

Emotion recognition impairment in PNFA has not been reported before (Rankin et al., 2009). The impairment observed in this study is unlikely to be due to greater disease severity in the selected PNFA patients, as these participants had a similar cognitive performance to the other patient groups. Rather, we suggest that using a larger group and a sensitive measure of emotion recognition has allowed us to uncover deficits not previously reported. The observed impaired recognition of sadness, anger, and fear is likely due to pathological changes in regions beyond the insula, such as the anterior cingulate and striatal regions, which may have resulted in the impaired sadness and anger recognition observed in this study (Fukui & Kertesz, 2000). Disconnection among brain regions involved in emotion recognition may also contribute to the observed deficits, with white matter changes of the superior longitudinal fasciculus and uncinate fasciculus previously reported in this FTD subtype (Whitwell et al., 2010).

Taken together, our results suggest that the multimodal system model of emotion accounts to some extent for emotion-recognition deficits in FTD, although its predictions tend to be more specific than the deficits reported in this study. The multimodal system model has evolved from studies involving patients with discrete lesions and studies using functional imaging in healthy controls. As such, the model appears less able to account for the deficits arising from diffuse brain changes and disconnections across brain regions, such as those seen in neurodegenerative disorders.

**Multiple processes of emotion recognition: The effect of intensity**

In this study, mechanisms underlying poor performance on emotion-recognition tasks were investigated by manipulating the intensity of facial emotional expressions. Both overall and individual emotion recognition significantly improved in bvFTD and PNFA with increased intensity, whereas the SD group showed limited response to that manipulation. Importantly, the bvFTD and PNFA groups were significantly impaired on neuropsychological measures of attention and working memory. Increasing the salience of the emotions, and therefore reducing the attentional and perceptual demands of the task, saw an improvement in performance in these two groups. These findings suggest that emotion-recognition disturbance in bvFTD and PNFA may be attributable in part to attentional deficits. In contrast, the lack of improvement in SD indicates a primary emotion-processing deficit in this group that cannot be overcome with changes in the attentional and perceptual demands of the task.

Emotional stimuli tend to be processed preferentially over nonemotional stimuli (Dolan, 2002; Öhman, Flykt, & Esteves, 2001). This preferential processing is mediated by two networks that are thought to work in parallel: Bottom-up, pre-attentive processing occurs in regions from the amygdala to cortical visual areas (Vuilleumier, Armony, Driver, & Dolan, 2001), whereas top-down, preferential allocation of spatial attention to emotional stimuli is mediated by prefrontal attentional networks (Hopfinger, Buonocore, & Mangun, 2005; LaBar et al., 1998; LeDoux, 1998; Palermo & Rhodes, 2007; Vuilleumier, 2005).

The top-down network relies on frontoparietal cortical regions, regions which overlap with the atrophy observed in bvFTD and to some extent PNFA (dorsolateral prefrontal cortex, anterior cingulate gyrus, ventromedial prefrontal cortex, superior temporal sulcus, and intraparietal sulcus) (Palermo & Rhodes, 2007). At low levels of intensity, emotional salience may be insufficient to attract preferential attentional resources in bvFTD and PNFA. With increasing intensity, however, as emotional salience becomes greater, attentional resources become preferentially allocated to the stimulus by the top-down network, resulting in accurate emotion detection. In contrast, in SD, the amygdala is compromised. This area is critical for emotion analysis and receives feedback both from the bottom-up and top-down pathways (Adolphs, 2002b). Hence, amygdala damage results in widespread emotion recognition impairment, which cannot be overcome by increasing emotional salience. This position is supported by a recent study that demonstrated that perceptual deficits contribute to impaired performance on emotion recognition tasks in bvFTD, but not SD (Miller et al., in press).

Notably, the effect of intensity was largest for negative emotions. From an evolutionary perspective, detection of some emotions is more critical than others.
to assist survival. As such, anger detection is important for identifying potential aggression in others and preparation for defense. Similarly, fear detection is important for avoidance of dangerous situations, and detection of disgust is important for avoiding potentially hazardous foods and environments (Darwin, 1872). From this perspective, processing of these emotions, which are critical for survival, is likely to be more robust and amenable to compensation than other emotions, such as surprise, that are relevant to social situations. Accordingly, a stronger response to modulation of intensity would also be anticipated in those emotions that are linked to survival than in those associated with social situations, as was generally observed in this study.

It is also possible that increasing salience has a specific effect on arousal, with increase in arousal sufficient for a critical threshold to be met, and recognition of the emotional stimulus to occur. In this study, highly arousing negative emotions responded to changes in intensity to a significant extent. Investigating whether there are circumstances under which manipulation of intensity is effective for low arousing emotions will help to distinguish these theoretical accounts.

Clinical implications

The emotion-recognition deficits observed in this study illustrate extensive disruption to emotion processing across all subtypes of FTD. The impact of these deficits on appropriate social interactions and the ability to form and maintain relationships is substantial. Thus, the potential for rehabilitation of emotion recognition is an important target for intervention. The current results suggest that increasing the emotional salience of environmental stimuli may be sufficient for attentional resources to be allocated to these stimuli and improve emotion recognition in PNFA and bvFTD, at least for negative emotions.

It can only be speculated whether increasing emotional salience might also offer an effective intervention in the later stages of the disease, and for other emotions. Investigating the effect of salience longitudinally may provide some insight into the effects of perceptual and attentional demands on emotion processing, and help determine whether increasing emotional salience is an effective intervention later in the disease course.

Summary

These results demonstrate significant impairment in recognition of emotions across all subtypes of FTD, including PNFA. Furthermore, they demonstrate that the underlying mechanism causing impaired performance on emotion-recognition tasks differs across subtypes. The results suggest that while SD patients perform poorly because of an underlying primary emotion-processing deficit, bvFTD and PNFA patients appear to be impaired because of inattention or perceptual difficulties, or due to partially degraded emotion-recognition structures that respond only to highly salient emotional material. Investigation of the neural correlates of emotion detection in these patients by structural or functional imaging will help understand what structures are necessary for recognition of specific emotions and help determine what effect manipulation of intensity is having in this population. Clinically, these results suggest that compensation of impaired emotion-recognition performance may be possible in the bvFTD and PNFA subtypes of FTD. Given the striking behavioral changes in these patients that alter their interpersonal conduct and ability to form and maintain relationships, simple interventions such as these are likely to have significant impact on these patients’ functioning.

REFERENCES


APPENDIX 1

Example stimuli from the Ekman 60 task (100%) and the Ekman Caricatures task for Model MO (A) for the emotion disgust and Model JJ (B) for the emotion sadness, across the four levels of intensity (+15%, +30%, +50%, +75%).