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# Face shape and face identity processing in behavioral variant fronto-temporal dementia: A specific deficit for familiarity and name recognition of famous faces

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ABSTRACT

Deficits in face processing have been described in the behavioral variant of fronto-temporal dementia (bvFTD), primarily regarding the recognition of facial expressions. Less is known about face shape and face identity processing. Here we used a hierarchical strategy targeting face shape and face identity recognition in bvFTD and matched healthy controls. Participants performed 3 psychophysical experiments targeting face shape detection (Experiment 1), unfamiliar face identity matching (Experiment 2), familiarity categorization and famous face-name matching (Experiment 3). The results revealed group differences only in Experiment 3, with a deficit in the bvFTD group for both familiarity categorization and famous face-name matching. Voxel-based morphometry regression analyses in the bvFTD group revealed an association between grey matter volume of the left ventral anterior temporal lobe and familiarity recognition, while face-name matching correlated with grey matter volume of the bilateral ventral anterior temporal lobes. Subsequently, we quantified familiarity-specific and name-specific recognition deficits as the sum of the celebrities of which respectively only the name or only the familiarity was accurately recognized. Both indices were associated with grey matter volume of the bilateral anterior temporal cortices. These findings extent previous results by documenting the involvement of the left anterior temporal lobe (ATL) in familiarity detection and the right ATL in name recognition deficits in fronto-temporal lobar degeneration.



## 1. Introduction

Fronto-temporal lobar degeneration (Neary et al., 1998) is a neurodegenerative disorder associated with atrophy of the temporal and/or frontal lobes. The main regions of brain atrophy are often responsible for the corresponding symptoms. Patients with such brain atrophy can present with behavioral symptoms - designated behavioral variant fronto-temporal dementia (Rascovsky et al., 2011) - or language deficits - designated primary progressive aphasia (Gorno-Tempini et al., 2011). The former is characterized by progressive deterioration of personality, behavior and cognition, with atrophy situated in the anterior temporal, mesiofrontal and subcortical areas (Seeley et al., 2008; Whitwell et al., 2009). Neuropsychological deficits include the recognition of emotional expressions, which have primarily been consistently documented in the face domain (Baez et al., 2014; Bediou et al., 2009; Bertoux et al., 2014; Bertoux et al., 2012a; Bertoux et al., 2012b; Couto et al., 2013; Diehl-Schmid et al., 2007; Fernandez-Duque and Black, 2005; Hsieh et al., 2013; Keane et al., 2002; Kipps et al., 2009; Kumfor et al., 2014b; Kumfor et al., 2011; Kumfor and Piguet, 2012; Lavenu et al., 1999; Lough et al., 2006; Miller et al., 2012; Oliver et al., 2014; Omar et al., 2011a, 2011b; Rosen et al., 2004; Rosen et al., 2002; Snowden et al., 2008).

Patients with behavioral variant fronto-temporal dementia at times demonstrate decreased ability to detect the emotions conveyed by facial expression despite their ability to recognize the person as familiar (Couto et al., 2013; Keane et al., 2002; Rosen et al., 2004; Rosen et al.,

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2002; Snowden et al., 2008). However, studies also find decreased ability to recognize faces - so-called deficits in facial identity processing (Kumfor et al., 2015; Miller et al., 2012).

The study by Kumfor et al. (2015) further suggested that impaired face identity discrimination is associated with atrophy in the left temporal cortex, including the fusiform gyrus.

Recognition of famous faces can be considered a variant of face identity processing, combining processing of familiarity, an essential feature of famous face recognition (Bobes et al., 2013; Burton and Jenkins, 2011). There is evidence that the capacity for recognizing famous faces is impaired in fronto-temporal lobar degeneration, particularly in the semantic variant of primary progressive aphasia (Gefen et al., 2013; Gorno-Tempini et al., 2004; Snowden et al., 2004). The clinical phenotype of semantic variant primary progressive aphasia includes the loss of conceptual knowledge and is neuro-anatomically associated with anterior temporal lobe (ATL) atrophy. In line with this, patients with temporal variant fronto-temporal lobar degeneration display worse famous face identification compared to frontal variant fronto-temporal lobar degeneration (Omar, Rohrer, Hailstone, and Warren, 2011). In primary progressive aphasia, left anterior temporal grey matter volume (GMv) correlates with the ability to name faces and bilateral anterior temporal GMv correlates with recognition of famous faces (Gefen et al., 2013). These results complement earlier findings of famous person knowledge in semantic variant primary progressive aphasia, with primarily visuo-pictorial deficits in right lateralized semantic variant primary progressive aphasia and verbal deficits in left lateralized semantic variant primary progressive aphasia (Snowden et al., 2012).

The picture emerging from the findings reported above consists of a possible deficit in face identity processing in behavioral variant fronto-temporal dementia as well as a deficit in famous face recognition in semantic variant primary progressive aphasia, associated with anterior temporal grey matter volume. However, little is known about famous face recognition in behavioral variant fronto-temporal dementia, nor about the association between unfamiliar facial identity processing and famous face recognition. In the present study, we address these issues in a sample of behavioral variant fronto-temporal dementia patients. Furthermore, we investigate more basic face processing skills, i.e. the ability to detect a facial shape, which presumably precedes processing of identity and semantic associations according to influential face processing models (e.g. Haxby and Gobbini, 2011), as well as recognition of familiarity. We included the latter as our clinical observations supplemented with tailored neuropsychological investigations have indicated a specific degradation of familiarity processing in neurodegenerative disorders (Van den Stock, de Gelder, De Winter, Van Laere, and Vandenbulcke, 2012; Van den Stock et al., 2013).

Based on the documented involvement of the anterior temporal lobes in famous face recognition, in combination with the atrophic topography in behavioral variant fronto-temporal dementia, we expect a deficit in famous face recognition. On the other hand, considering the association between unfamiliar face recognition and face shape processing with more posterior temporal regions, we do not anticipate respective severe deficits.

## 2. Methods

### 2.1. Participants

All subjects were right-handed as assessed by the Edinburgh Handedness Inventory (Oldfield, 1971). A total of 29 consecutive behavioral variant fronto-temporal dementia patients were recruited. Six of these patients could not be included in the study since no experimental data could be acquired due to a lack of cooperation and/or agitation. The remaining 23 were recruited via the Memory Clinic ($N = 7$) and Old Age Psychiatry Department of University Hospitals Leuven ($N = 10$) and the Neurology Department of Onze-Lieve-Vrouwziekenhuis Aalst-Asse-Ninove ($N = 6$). All patients were evaluated via clinical assessment, neuropsychological testing and structural MRI. In addition, [$^{18}$F]-Fluorodeoxyglucose Positron Emission Tomography (FDG-PET) was performed in all but three patients. Two patients fulfilled the revised diagnostic criteria of 'behavioural variant FTD with definite FTLD Pathology', based on a C9orf72 pathogenic mutation, and 18 patients fulfilled the criteria for 'Probable behavioral variant fronto-temporal dementia' (Rascovsky et al., 2011). The remaining three patients were diagnosed as 'Possible behavioral variant fronto-temporal dementia' (Rascovsky et al., 2011). In none of the patients, language difficulty was the most prominent clinical feature. Furthermore, in none of the patients, aphasia was the most prominent deficit at symptom onset and during the initial phase of the disease. These phenotypes do not comply with the current diagnostic criteria for primary progressive aphasia (Gorno-Tempini et al., 2011). Patients were included after clinical judgment deemed them able to successfully undergo the experimental procedure.

The control group was recruited through advertisements in local newspapers. Twenty control participants took part in the behavioral experiments and underwent structural MRI and neuropsychological assessment. The exclusion criteria consisted of present or past neurological or psychiatric disorders. This included substance abuse as well as significant systemic comorbidities or use of medication susceptible to affect the central nervous system. Demographic data and neuropsychological test results of all participants are presented in Table 1. The individual demographic and neuropsychological data of the patients, including a detailed overview of the diagnostic criteria they fulfilled, are presented in Supplementary Table S1.

### 2.2. Experiment 1: face shape detection

Materials consisted of visual images that were validated regarding face-semblance, based on a computerized face-detection algorithm as well as on subjective ratings of face-semblance (Meng et al., 2012). The dataset consists of 5 categories of images showing increasing facial shape cues. A total of 40 images was selected, 20 images from the category with the highest face-semblance and 5 images of each of the 4 remaining categories.

The procedure differed from the one described by Meng et al. (2012). A trial consisted of simultaneous presentation of 2 images next to each other. One of the images always was from the category with the highest face-semblance, and the second image was from one of the 4 remaining categories. This resulted in 4 conditions of increasing

**Table 1**
Demographic and neuropsychological test results. MMSE = Mini-Mental-State Examination; RAVLT = Rey Auditory Verbal Learning Test; A1–A5 = the sum of scores on trials A1 to A5 of the RAVLT; Recognition = the recognition score constitutes the difference between the number of correct hits and false hits on the recognition trial; BNT = Boston Naming Test; AVF = Animal Verbal Fluency; TMT = Trail Making Test; BORB = Birmingham Object Recognition Battery; RCPMT = Raven Colored Progressive Matrices Test; AAT = Aachen Aphasia Test. [£] = ($N = 21$); [%] = ($N = 20$); [$] = ($N = 19$); [§] = ($N = 17$); [&] = ($N = 15$).

| | | bvFTD ($N = 23$) | Controls ($N = 20$) | | |
|---|---|---|---|---|---|
| | | | | t ($\chi^2$) | p |
| Age (SD) | | 64.5 (9.8) | 66.6 (6.1) | 0.854 | 0.398 |
| Sex (M/F) | | 13/10 | 12/8 | (0.000) | 1.000 |
| MMSE | | 26.7 (1.5)[£] | 29.2 (0.6) | 7.124 | 0.001 |
| RAVLT | A1–A5 | 29.0 (11.3)[%] | 50.8 (7.3) | 7.262 | 0.001 |
| | % recall | 56.1 (31.9)[%] | 80.9 (17.4) | 3.060 | 0.005 |
| | Recognition | 6.5 (7.5)[%] | 14.0 (1.3) | 2.135 | 0.043 |
| BNT | | 40.3 (12.7)[%] | 54.4 (2.9) | 4.861 | 0.001 |
| AVF | | 15.0 (5.5)[%] | 22.1 (5.8) | 4.016 | 0.001 |
| TMT | A (secs) | 63.5 (42.7)[$] | 32.5 (9.4) | 3.099 | 0.006 |
| | B (secs) | 193.1 (141.2)[&] | 89.8 (42.3) | 2.742 | 0.015 |
| BORB | Length | 87.6 (7.3)[§] | 90.1 (4.5) | 1.262 | 0.218 |
| | Size | 85.5 (6.9)[§] | 88.9 (6.3) | 1.569 | 0.126 |
| | Orientation | 81.4 (9.2)[§] | 86.1 (6.0) | 1.845 | 0.074 |
| RCPMT | | 16.4 (3.9)[%] | 20.8 (2.8) | 4.214 | 0.001 |
| AAT | Comprehension | 93. 9 (12.3)[$] | 109.5 (5.3) | 5.093 | 0.001 |