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## Letter to the Editor

# A strange face in the mirror. Face-selective self-misidentification in a patient with right lateralized occipito-temporal hypo-metabolism

Jan Van den Stock<sup>a,\*</sup>, Beatrice de Gelder<sup>a,b,c</sup>, François-Laurent De Winter<sup>a</sup>,  
Koen Van Laere<sup>d</sup> and Mathieu Vandenbulcke<sup>a</sup>

<sup>a</sup>Brain and Emotion Laboratory Leuven (BELL), Division of Psychiatry, Department of Neurosciences, KU Leuven, Leuven, Belgium

<sup>b</sup>Tilburg University, Cognitive and Affective Neuroscience Laboratory, Tilburg, The Netherlands

<sup>c</sup>Athinoula A. Martinos Centre for Biomedical Imaging, Massachusetts General Hospital, Harvard Medical School, Charlestown, MA, USA

<sup>d</sup>KU Leuven, Division of Nuclear Medicine, UZ Gasthuisberg, Leuven, Belgium

## 1. Introduction

Delusions are intriguing neuropsychiatric phenomena. Some types are associated with specific neurological conditions, particularly monothematic delusions, characterized by a single abnormal belief (Coltheart et al., 2011). Next to the well-known Capgras delusion (Capgras and Reboul-Lachaux, 1923) (the belief that people are replaced by doubles), another remarkable delusion is the mirrored-self misidentification or mirror sign (inability to recognize one's self-image in the mirror). Although delusions like the mirror sign can occur in multiple stages of syndromes like schizophrenia and dementia (Connors and Coltheart, 2011), a better understanding of monothematic delusions requires focusing on the symptom rather than the syndrome (Coltheart, 2011). The two-factor theory (TFT) proposed by Coltheart et al. (2011) states that identification of both the specific origin (what caused the particular delusion?) and persistence (why isn't the abnormal belief corrected?) is required to understand the delusional nature and genesis. Two mirror sign cases have been studied and interpreted from the TFT (Breen et al., 2000, 2001). For one patient, impaired face-processing was withheld as the originating underlying neuropsychological abnormality, whereas for the other patient it was mirror agnosia. The second factor (persistence) is common for all delusions and relates to impaired belief evaluation, hypothetically associated with right lateral prefrontal cortex pathology (Coltheart, 2007, 2010; Coltheart et al., 2007; Langdon

et al., 2008). Here, we describe neuropsychological and neuro-imaging investigations in patient MV displaying mirror sign.

## 2. Case report

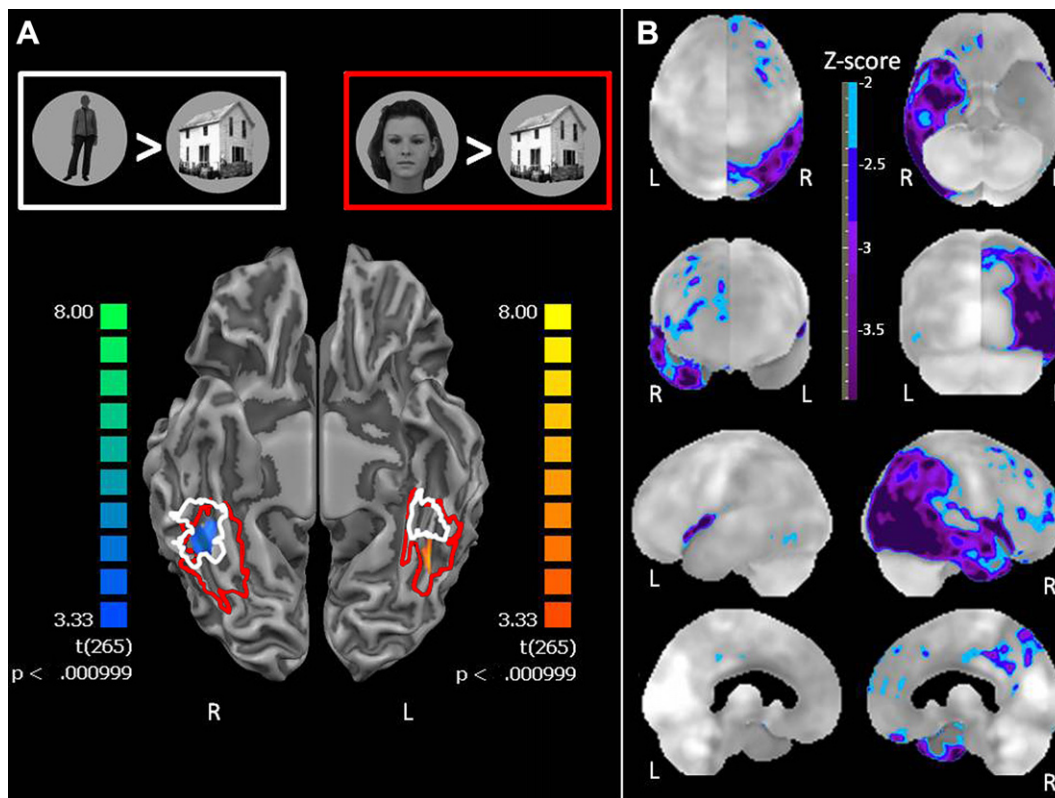
MV (85, female) was referred to our geriatric psychiatry unit after showing signs of hallucinations: caregivers heard her talking to another person when nobody was around. Additionally, MV displayed a phantom boarder delusion: she was convinced a woman was living uninvited in her house and later in the hospital at MV's expenses. MV had no neurological or psychiatric history and displayed no clinical features of parkinsonism. Ophthalmologic examination revealed no deficits. Neuropsychological evaluation showed mild deficits of memory, executive, language and visuo-spatial functions. The Mini Mental State Examination (MMSE-score) equaled 21/30. Magnetic Resonance Imaging (MRI) indicated cortico-subcortical atrophy and [18F] Fluorodeoxyglucose Positron Emission Tomography (FDG-PET) showed clear right lateralized lateral prefrontal, parietal and particularly occipito-temporal hypo-metabolism (Fig. 1). A diagnosis of probable mild Alzheimer's disease was withheld. During admission, it became clear that MV displayed a mirror sign. She identified her mirrored face as the "phantom boarder" (Supplementary Video). Confronted with her mirrored face image, MV explained she had no idea where the woman came from, but she always

\* Corresponding author. Department of Neurosciences, Division of Psychiatry, Brain and Emotion Laboratory Leuven (BELL), O&N II, Herestraat 49 bus 1022, 3000 Leuven, Belgium.

E-mail address: [jan.vandenstock@med.kuleuven.be](mailto:jan.vandenstock@med.kuleuven.be) (J. Van den Stock).

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doi:10.1016/j.cortex.2012.03.003



**Fig. 1 – Statistical map of brain activation of patient MV (A) in response to bodies compared to objects and faces compared to objects (stimulus examples shown on top). The regions defined by the white and red contours demark the areas containing the pooled individual activation maps in the left and right fusiform gyrus for bodies compared to objects (white contours) and faces compared to objects (red contours) in 31 control subjects ( $p < .001$ ). The activation maps are plotted on a ventral view of a cortical reconstruction of MV's brain, showing gyri in light gray and sulci in dark gray. B: Stereotactic surface projections of an FDG-PET brain scan in MV, with areas of significant hypo-metabolism superimposed (z-scores with respect to normal dataset of 30 control subjects).**

dressed just like her. We administered the mirror-mark test (Gallop, 1970) in which a mark was unnoticeably placed on MV's face. Subsequently, MV was positioned before a mirror. When subjects with intact self-face awareness discover the mark on their face after looking in the mirror, they typically spontaneously try to touch or remove it. Remarkably, MV did not react in any way to the mark. MV noticed the mark on the other woman's face when she was explicitly questioned about it. She had no difficulty accurately locating the mark when it was unnoticeably placed on the person next to her (Supplementary Video). MV was also able to recognize other persons in the mirror.

Supplementary video related to this article can be found at [doi:10.1016/j.cortex.2012.03.003](https://doi.org/10.1016/j.cortex.2012.03.003).

To investigate whether the self-misidentification comprised also her body, we positioned MV in front of a tilted mirror, reflecting her whole body except the head. MV then reported she saw herself in the mirror, based on her posture. However, as soon as she approached the mirror and also saw her face reflected, MV reported the other woman appeared (Supplementary Video).

We presented one by one face photographs of celebrities (20), close family (8), MV (10), and unfamiliar persons (20). We asked MV to indicate whether the face was familiar or

unfamiliar. Compared to an elderly control group ( $n = 23$ ; mean age = 63.7 years; standard deviation = 3.80), the familiarity judgments of MV were impaired for celebrity faces ( $p < .001$ ), but not for unfamiliar faces ( $p = .15$ ). She performed at chance for family and self faces. She categorized recent photographs of her own face as 'familiar'; however, she spontaneously identified her as the 'phantom boarder'. Subsequently, we presented the celebrity faces with three celebrity names printed underneath and instructed MV to match the face with a name. She performed within the normal range ( $p = .09$ ).

Using functional MRI (fMRI), we localized the brain areas that selectively respond to bodies and faces (Supplementary Method). Although face and body-selective brain areas are typically observed in the right hemisphere, we observed activation in the left but not right fusiform face area when comparing the perception of faces with objects. Contrarily, we observed activation of the right but not left fusiform body area when comparing bodies with objects.

### 3. Discussion

MV's face-processing abilities indicate that the originating factor in her delusion pertains to selectively impaired

face-familiarity recognition. This extends a previous report of abnormal face-processing in a patient with mirror sign, who contrarily showed over-familiarization (Breen et al., 2001). Our report additionally shows that famous face-name matching may remain relatively intact. Although the delusion only pertained to her own face, the face-familiarity deficit also applied to other faces, including close relatives. Possibly, MV relied on non-face cues like body or voice to recognize familiar persons, whereas self-identification in the mirror primarily relies on face-cues. In line with MV's familiarity-selective face-deficit, patients with Capgras delusion fail to exhibit the enhanced skin conductance response to familiar faces (Brighetti et al., 2007; Ellis et al., 1997; Hirstein and Ramachandran, 1997). The disconnection between MV's face-identity and face-familiarity processing is likely related to the right occipito-temporal hypo-metabolism. The self-face and body recognition shows an interesting analogy with the fMRI-results. Intact self-body recognition is associated with right lateralized body-selective activation, whereas deficient self-face recognition is associated with absence of face-selective activity in right occipito-temporal cortex. This extends previous findings that self-recognition involves the right hemisphere (Keenan et al., 2001). Concluding, our data suggest an impaired mapping of a perceived face on the stored memory representation (resulting in impaired face-familiarity), with relatively preserved person identification by name or body cues.

MV additionally shows right dorsal prefrontal hypo-metabolism, presumably underlying her impaired belief evaluation (Coltheart et al., 2011).

## Acknowledgments

J.V.d.S. is a post-doctoral researcher supported by FWO-Vlaanderen. B.d.G. is partly supported by European Commission (COBOL) Grant, FP 6 FET-Open, COBOL grant en FP7-FET-Open grants. KVL is Senior Clinical Investigator of FWO-Vlaanderen.

## Supplementary material

Supplementary data related to this article can be found online at [doi:10.1016/j.cortex.2012.03.003](https://doi.org/10.1016/j.cortex.2012.03.003).

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