

# The Delusions of Capgras and Intermetamorphosis in a Patient with Right-Hemisphere White-Matter Pathology

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## Key Words

Capgras delusion · Right hemisphere · White-matter pathology · Face processing

## Abstract

Previous neuropsychological studies have demonstrated an association between person misidentification and right-hemisphere dysfunction. In the study reported here, we explore the contribution of facial and visual recognition impairments in a patient with right-hemisphere subcortical white-matter pathology in the frontal and parietal lobes and a diagnosis of vascular cognitive impairment. The patient displayed false recognition of unfamiliar faces and deficient retrieval of key biographic detail for famous faces. These results are discussed in the context of the contribution of deficiencies in the visual system and subcortical white-matter lesions to the development of Capgras delusion.

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## Introduction

Disorders of face memory and face perception have been the focus of much neuropsychological investigation over the past 50 years. The most extensively studied of

these is prosopagnosia – a condition characterised by the inability to identify all familiar faces, including famous persons, friends, family and even the prosopagnosic's own face [1]. The critical lesion sites in prosopagnosia involve right ventromedial occipitotemporal regions which include the inferior temporal cortex and medial temporal lobe limbic areas [2].

Although less well documented, areas of the prefrontal cortex are also involved in facial processing: projections from the face-processing areas in the right ventromedial occipitotemporal regions to the ventromedial prefrontal cortex via the uncinate fasciculus as well as limbic pathways are well established [3]; single cell recordings have identified face-sensitive neurons in the inferior frontal convexity region of non-human primates [4], and neuropsychological studies have documented an association between false facial recognition and right frontal lobe dysfunction [5]. These frontal patients, in striking contrast to the lack of conscious facial recognition described in prosopagnosics, mistakenly believe that unfamiliar faces are familiar [6, 7].

In a small subset of psychotic patients, false recognition can lead to frank misidentification which can reach delusional proportions [5, 8–10] – as in the case of Frégoli delusion (FD) [11]. The hallmark of FD is the recognition of a familiar other in a stranger. The patient believes that the familiar other is able to take on different physical

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forms and adopt another's appearance. It has been suggested that FD develops when spurious feelings of familiarity evoked by novel faces are combined with the retrieval of incorrect biographic detail [5]. A related phenomenon – the delusion of intermetamorphosis (ID) [12] – is marked by the false belief that a person has been transformed, physically and psychologically, into another. Patients report the presence of physical similarities between the original and misidentified individuals, and a dynamic transformation occurring between the two. Finally, the Capgras delusion (CD) [13], a subtly different form of misidentification from FD and ID, is characterised by the delusional belief that a significant other has been replaced by an impostor who bears a close physical resemblance to the original [14]. Again, small misperceived differences, for instance in physical appearance and behaviour, are frequently used to distinguish the impostor from the loved one.

Neuroanatomical evidence suggests that CD is commonly associated with right-hemisphere abnormalities, particularly in the region of the temporal and/or parietal lobes coupled with right or bilateral frontal pathology [15, 16]. It has been suggested that CD results from a disconnection between the face-processing areas in the inferior temporal lobe, responsible for the conscious recognition of the face and recall of associated biographic information, and key limbic structures which mediate the feelings of familiarity which accompany overt recognition [17, 18]. A loss of the normal skin conductance response to familiar faces has been demonstrated in a small number of CD patients [18, 19].

In this study, we explore facial and visual recognition in a patient with CD and ID arising in the context of cognitive impairment. This case is of interest on two counts: firstly, the neuro-anatomical evidence of combined white-matter lesions in right prefrontal and parietal regions – previous reports of pathology have been largely restricted to the grey matter. Secondly, the transient co-existence of ID and CD raises questions as to the role played by transient (functional changes in terms of neurotransmission and/or neurophysiology) as well as stable (i.e. organic) factors in the development of these symptoms of delusional misidentification.

## Case Report

### Background

V.J. was a 64-year-old right-handed man, with a 20-year history of hypertension. He was married with 1 grown-up son. V.J. worked as an instrument maker before retiring, aged 59. His hobbies included

renovating classic motor bikes, building computers and following the stock market. There was no personal or family history of psychiatric disorder, alcohol or drug abuse. For the preceding 12 months, V.J. had experienced a gradual deterioration in his memory, ability to work previously familiar gadgets, problems with writing and understanding written material. Medication included propranolol (80 mg daily), trazodone (50 mg nocte) and oxypertine (20 mg daily).

An MRI scan showed abnormal areas of high T<sub>2</sub>-weighted signal in the brain stem and basal ganglia bilaterally, primarily involving the globus pallidus. In the white matter of the right hemisphere, 3 small ischaemic lesions were identified in the frontal (postfrontal cortex and just below the superior frontal gyrus) and anterior parietal lobes (fig. 1: upper, middle and lower images, respectively). The reported abnormalities were consistent with a diagnosis of cerebrovascular disease. Corrected visual acuity was 6/6 for both eyes, near vision was N5.

The reported study was carried out over a 7-month period. Informed consent was obtained, and patient initials have been changed. During mental state examination, V.J. was well oriented for time and place, although concentration (attention and calculation 0/5) and immediate recall (0/3) were poor. There were no apparent perceptual problems and he was able to follow verbal commands. A summary of V.J.'s cognitive neuropsychological assessment follows – a full report is available on request.

### Episodes of Misidentification

Over the 7-month period of contact, V.J. exhibited episodes of CD and ID. Episodes of CD for his wife were experienced on a recurrent basis. He claimed that there were several 'Jeans' in the house, the 'Jean in the kitchen' was different from the 'Jean' he had been chatting to in the living room. Occasionally, these episodes took a more ominous turn – V.J. believed that the 'Jean-impostor' was a spy sent by the Inland Revenue who was out to trap him. He described the impostor as looking like Jean but that the 'real Jean' had gone away: 'I care a lot about Jean, but I don't like you at all.'

Instances of ID occurred less frequently, one centred around the delusional belief that his wife had been transformed (physically and psychologically) into his (deceased) mother-in-law. Recognition of his grown-up son and elderly mother, whom he saw on a weekly basis, was preserved.

The periods of misidentification were selective (focused on his wife), occurred on average a couple of times a day, were transient (seconds to minutes) and appeared to be exacerbated by feelings of anxiety (e.g. an out-patient visit), changes in the appearance of his wife (e.g. leaving the room and returning in a coat), fatigue and disorientation (episodes occurred more frequently in the evening, or when waking from a nap). These episodes were frequently accompanied by verbally aggressive behaviour, which has been quite a shock for his wife, who described him as a quiet, easygoing man who rarely raised his voice.

V.J. did not always regard his wife as an impostor, and during these lucid periods he would acknowledge her as 'the real Jean'. No other psychotic features were evident, and he was not depressed.

### Cognitive Neuropsychological Assessment

His current levels of functioning (table 1) were placed in the mentally deficient range [20], which was well below his premorbid levels of functioning based on his former employment and hobbies.

There was clear evidence of both ideomotor and constructional apraxia, which is consistent with his reported problems in using gad-



**Fig. 1.** An MRI scan showing abnormal areas of high T<sub>2</sub>-weighted signal in the white matter of the right hemisphere. Three small ischaemic lesions were identified in the post-frontal cortex, just below the superior frontal gyrus and the anterior parietal lobe (arrows in upper, middle and lower images, respectively).

**Table 1.** V.J.'s profile on the Wechsler Adult Intelligence Scales – Revised

Verbal		Performance	
Information	6	Picture completion	5
Digit span	2	Picture arrangement	1
Vocabulary	7	Block design	4
Arithmetic	3	Object assembly	2
Comprehension	1	Digit symbol	2
Similarities	3		
Verbal IQ	68	Performance IQ	68
Full-scale IQ	67		

**Table 2.** Facial and visual processing

Test	Score	Normal range
Face sorting (max. 25)		
Familiar face hits	25	23–25
Novel face rejections	17	21–25
Familiar face recognition (max. 25)		
Profession	22	22–25
Name	10	17–25
Minimal features (max. 25)	22	19–25
Object decision (max. 32)	21	22–32
Odd-one-out (max. 10)		
Visual	1	10
Spoken	3	10
Written	6	10

gets around the home. Assessment of language function revealed preservation of repetition, ability to name pictures, read high-frequency and high-imageability words. However, difficulties were evident on tests of comprehension, writing and reading nonwords, low-frequency and low-imageability words. Both apraxia and comprehension deficits have been reported following damage to the dominant basal ganglia [21].

Assessment of facial recognition revealed abnormalities in familiarity judgements and accessing stored knowledge from vision (table 2). A face familiarity task, which involved sorting photographs of famous from unfamiliar faces, revealed an elevated false recognition rate for unfamiliar faces but a normal hit rate for famous faces. However, retrieval of stored information (profession and name) was poor. This was confirmed in a second task which required identification of the odd-one-out from a set of three photographs on the basis of profession (e.g. John Major, Margaret Thatcher, *Michael Aspel*). Interestingly, when this odd-one-out task was repeated using spoken or written names (matched for profession and notoriety), V.J.'s performance showed improvement – particularly in the written version. His poor performance on the visual version of the odd-one-out task was not attributable to a generalised naming problem as evidenced by his ability to correctly name a set of 30 line drawings of animate and inanimate items.

Deficiencies in ability to access stored knowledge from vision were also evident using animate and inanimate stimuli (table 2). V.J. performed poorly on an object decision task [22] which involved discriminating real (animate or inanimate exemplars) from unreal items (i.e. constructed by replacing one feature of a real item with a feature from another item, e.g. a tortoise's head and snake's body). A visual test of object constancy [22] which involved matching line drawings of the objects presented from different viewpoints was performed within normal limits. Ability to identify 10 inanimate objects taken from V.J.'s home (work tools, clothing, household items) from a set of matched unfamiliar ones was flawless.

A group of 11 age- and education-matched normal controls is reported for the sorting and facial recognition tasks. V.J.'s wife was the control for the 3 odd-one-out tasks. All of the controls were right-handed, and none reported a neurological or psychiatric history.

## Discussion

CD, FD and ID are highly correlated with facial processing impairments. Abnormalities are evident at the level of discriminating between pairs of familiar and unfamiliar faces on the basis of same/different judgements [23], recognition memory for unfamiliar faces [24] and retrieval of biographic data from photographs of famous faces [23]. In this study, we have demonstrated a discrepancy in V.J.'s ability to access stored biographic knowledge – increased levels of preservation are apparent when this is done verbally, i.e. spoken or written name as compared to a visual image. These findings extend work which documented a discrepancy in recognition memory for words (which was good) and faces (which was poor) by showing that the verbal/visual disparity is also evident at the level of *access* to stored knowledge – i.e. retrieval of biographic data is facilitated tapped from the verbal domain. Studies of recognition memory in psychotic patients who do not exhibit any evidence of CD, FD or ID have reported poor recognition memory for faces *and* words, which suggests that delusional misidentification syndromes like CD, FD and ID emerge when right-hemisphere dysfunction is substantial relative to left-hemisphere preservation [24].

Recently, Ramachandran and Blakeslee [25] described a patient with CD who misidentified his father when in his company, but not when speaking to him on the phone. This report seems to indicate the important role of accessing stored knowledge from vision in the aetiology of CD. However, CD has also been reported in a number of blind patients which suggests that it cannot have an exclusively visual basis [26, 27]. In the study of Reid et al. [26], their patient believed her cat had been replaced by an impostor because its miaow was different. Testing of her auditory

recognition revealed abnormalities in her ability to recognise familiar voices.

Posterior coupled with anterior right-hemisphere dysfunction has been implicated in the emergence of CD [16]. The neuro-imaging evidence reported for V.J. is consistent with this – however, his pathology is at the level of white rather than grey matter. The authors are aware of only one other case which reports delusional person misidentification arising in the context of white-matter pathology, which – as in the present case – was also located mainly in the frontal and parietal lobes [28].

The rich reciprocal afferent-efferent connections between the prefrontal cortex with limbic structures and cortical association areas make it a highly complex site for the integration of perceptual, cognitive and affective data [29, 30]. Pathological lesions in subfrontal white matter have been reported in the context of metachromatic leukodystrophy, a rare inherited demyelinating disorder of the central and peripheral nervous systems, which is accompanied by psychiatric symptoms including complex auditory hallucinations and bizarre delusions. The presence of subfrontal white-matter lesions suggest that psychosis [31, 32] and CD in particular [28, 33] may result from disruption of corticocortical and corticosubcortical connections especially involving the frontal lobes. It has been suggested that CD results from the disconnection of the face-processing areas in the inferior temporal lobe from structures in the limbic system, particularly the amygdala, which is crucial in assigning an emotional valence to familiar faces [17, 19]. Although V.J.'s sMRI failed to reveal any evidence of an amygdala lesion, the presence of functional abnormalities arising from the

remote effects of lesions on corticolimbic networks cannot be ruled out.

However, anatomical disconnection models fail to adequately account for the transient nature of the misidentification episodes. It has been suggested that CD may be associated with the 'kindling of subcortical structures' [33]. Kindling refers to repeated subthreshold stimuli which may result in psychomotor outbursts or overt seizure activity [34]. According to Kim [33], the effects of psychomotor outbursts may interfere with the integration of perceptual, cognitive and affective processing in the prefrontal cortex, by altering interlobar signal transmission through subcortical tracts. Psychomotor outbursts of limited duration may be responsible for the transient nature of the delusional experiences, whilst subtle changes in foci may account for impaired processing at different stages in the facial recognition system which, in turn, would have an impact on the particular form of misidentification experienced. On this basis, the cognitive impairments represent stable or mediating vulnerability factors, rather than transient indicators of dysfunction. Inherent weaknesses within the face-processing system remain hidden during non-psychotic periods but may be artificially exposed during formal cognitive neuropsychological testing by challenging the processing system with the particular stimuli and tasks found most difficult. Such deficits in visual processing, when combined with other factors such as changes in mental state (paranoia or anxiety) and impaired cognitive reasoning, operate in a complex interaction to produce symptoms of delusional misidentification [23].

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