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Hyperfamiliarity for unknown faces after left lateral temporo-occipital venous infarction: a double dissociation with prosopagnosia

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Summary

Right hemisphere dominance in face processing is well established and unilateral right inferior temporo-occipital damage can result in prosopagnosia. Here, we describe a 21-year-old right-handed woman with acute impairment in face recognition that selectively concerned unfamiliar faces, following a focal left lateral temporo-occipital venous infarct. She was severely impaired in discerning that unknown people seen in everyday life were unfamiliar, although she had no difficulty recognizing familiar people. Thus, she had no prosopagnosia, but abnormal 'hyperfamiliarity' for unknown faces. Her difficulty was not accompanied by delusions or deficits in discrimination, identification or memory for faces. Standard neuropsychological testing showed that her recognition of familiar faces was entirely normal. By contrast, her sense of personally knowing faces was severely impaired when unknown faces evoked weak signals of familiarity based on spurious cues, to the extent that she would misattribute fame to faces that were unknown but to which she had been incidentally exposed on a prior occasion. Priming experiments also revealed that, unlike normal subjects, she made familiarity judgements without accessing semantic identity representations. Moreover, in face recognition tests, she generally showed bias in that she relied more on right-hemisphere strategies to identify global traits and less on left-hemisphere processes compared with healthy subjects. This case provides novel evidence for a differential contribution of the two hemispheres to face recognition. Hyperfamiliarity for unknown faces might arise from an imbalance between reciprocal hemispheric functions in face recognition, with relative hypoactivation of left hemisphere processes but hyperactivation of right-hemisphere processes for retrieving stored associations about people, linking seen faces to representations of affective and personal relevance. Hence, abnormal bias in attributing some personal meaning to unknown faces could be evoked by spurious signals of familiarity based on irrelevant affective associations in the right hemisphere.

Keywords: face recognition; familiarity; prosopagnosia; misidentification; hemispheric laterality

Introduction

Different disorders of face recognition may follow brain damage. Prosopagnosia, although rare, is the best-known syndrome, and is defined as the inability to recognize familiar persons by their face alone (Bodamer, 1947; Damasio *et al*., 1982; Grüsser and Landis, 1991). Different forms have been distinguished on the basis of performance in visual tests (Bruyer and Schweich, 1991; Grüsser and Landis, 1991; McNeil and Warrington, 1991), such as apperceptive cases thought to suffer from a perceptual deficit in encoding facial traits, and associative cases thought to suffer from the destruction of stored representations of known faces or a deficit in accessing these representations (Bruce and Young, 1986; Burton *et al*., 1990). Remarkably, some patients with associative prosopagnosia may show evidence of implicit visual recognition of known faces, despite a complete lack of familiarity with such faces (Bauer, 1984). This suggests that the subjective experience of recognizing a face may involve not only adequate perceptual processing and activation of internal representations of face traits, but access to more complex associations stored in memory that pertain to the identity of a known person (Damasio *et al*., 1990; Schreiber *et al*., 1991). Accordingly, the perception of a familiar face

Table 1 *General neuropsychological assessment*

Test	Patient J. R.	Maximum or minimum score	*
Language			
Object naming			
Boston Naming 1	31	/44**	Centile 10–50
Boston Naming 2	43	/44	
ExaDE Bachy–Langedock test	89	/90	
Word reading	198	/200	
Oral spelling	12	/12	
Writing	4	/4	
Calculation	4	/4	
Ideomotor and constructional praxis			
Right hand	8	/8	
Left hand	8	/8	
Orofacial	8	/8	
Drawing	6	/6	
Body schema			
Culver right–left orientation test	20	/20	
Benton finger agnosia test	Right hand, 15	/15	
	Left hand, 15	/15	
Visual object recognition			
Poppelreuter overlapping shapes	5	/5	
Montreal–Toulouse Agnosia Test	20	/20	
Object parts	20	/20	
Colour test	18	/18	
Executive functions			
Verbal letter fluency (2 min)	32	Min. = 25	
Verbal category fluency (2 min)	39	Min. = 27	
Visual design fluency (3 min)	32	Min. = 27	
Trail Making Test A	25 s	Max. = 34	
Trail Making Test B	77 s	Max. = 85	
Luria graphic sequences	15	/15	
Luria gesture sequences	5	/5	
Colour-Word Stroop	48	/49	
D2 Test of sustained attention			
Correct targets cancelled	404	/700	
Errors	4		
Memory			
Digit verbal span	**5**	**	Centile 10
Corsi visuospatial span	7		
Rey word list learning			
Immediate recall 1	12	/15	
Immediate recall 2	14	/15	
Immediate recall 3	15	/15	
Immediate recall 4	15	/15	
Immediate recall 5	15	/15	
Delayed recall	14	/15	
Delayed recognition	15	/15	
Rey visual shape learning			
Immediate recall 1	5	/15	
Immediate recall 2	7	/15	
Immediate recall 3	13	/15	
Immediate recall 4	13	/15	
Immediate recall 5	15	/15	
Delayed recall	15	/15	
Delayed recognition	15	/15	

*Norms are based on the Standardized Geneva Hospital Neuropsychology Battery (unpublished) or published data (Spreen and Strauss, 1991; Benton *et al*., 1994). Abnormal performance in patient J. R. is indicated by ** (centile 10); otherwise, performance was within the normal range (centile 50).

generally triggers the activation of a rich assemblage of information concerning unique biographical details, past contextual episodes, affective relevance, and so on, which all participate in the subjective recollective experience associated with face recognition (Damasio *et al*., 1990; Van Lancker, 1991). However, such associative components of face recognition, beyond purely visual perceptual processing, remain largely unknown.

While prosopagnosia is most often associated with bilateral ischaemic lesions in the territory of the posterior cerebral arteries, unilateral damage to the inferior temporo-occipital cortex (areas 20, 21 and 37) in the right hemisphere appears sufficient to cause associative prosopagnosia, with complete loss of familiarity for known faces despite relatively intact perception and discrimination abilities (Landis *et al*., 1986, 1988; De Renzi *et al*., 1994). However, some authors have suggested that bilateral damage may be mandatory (Damasio *et al*., 1982) or lead to more profound deficits (Rhodes, 1985). It remains uncertain (i) whether right hemisphere function is crucial for recognizing familiarity, besides its superior role in visual facial analysis; and (ii) whether the left hemisphere makes any useful contribution to face recognition in humans. No case of prosopagnosia or any other selective disorder of face recognition subsequent to a unilateral left hemisphere lesion in a right-handed subject has yet been reported.

Here we report a young woman who presented with a peculiar disorder of face recognition following a lesion restricted to the lateral temporo-occipital junction of the left hemisphere due to a venous infarction. The patient exhibited a selective deficit in judging the familiarity of unknown faces, but she had no difficulty discriminating and recognizing known faces, and showed no delusion, confabulation or any other general cognitive impairment. Such a deficit has not been described previously as an isolated disturbance, and constitutes double dissociation with respect to defective recognition of familiarity for known faces in associative prosopagnosia. This patient provides unique insights into the left hemisphere's contribution to face processing and the data suggest a reciprocal function of the two hemispheres in associative recognition processes that bestow an experience of personal familiarity on seen faces. It also exemplifies the unique value of lesions situated outside common vascular territories in revealing novel brain–behaviour relationships, as historically shown for other rare deficits due to tumours (Trescher and Ford, 1937) or venous infarctions (Zihl *et al*., 1983).

The false familiarity with faces presented by our patient was very different from disorders of familiarity associated with delusional misidentifications, such as Capgras syndrome, in which the patient believes that a familiar person has been replaced by a disguised impostor, or Frégoli syndrome, in which the patient believes that a known person has disguised himself as another person (Ellis and Young, 1990; Young *et al*., 1990). These psychiatric disorders may occasionally follow brain damage (Young *et al*., 1990; Signer, 1994). It has been suggested that a critical aspect of such disorders is that they involve a disturbance in the activation of affective and semantic associations about known people, triggered by inappropriate faces and/or other irrelevant cues (Ellis and Young, 1990). Thus, subjective appraisal of familiarity is specifically altered but identification of known faces can be preserved (Ellis and Young, 1990; Sno and Linszen, 1990), although memory and perceptual processing of faces are often reported to be also impaired (Young *et al*., 1990). A wide variety of lesions has been reported in such patients, including unilateral right (Young *et al*., 1990), bilateral (Burgess *et al*., 1996) and diffuse (Förstl *et al*., 1991) cerebral damage.

Unlike these cases, our patient had a very focal cortical lesion and she showed no delusion and no deficit in face perception or face memory, even during the acute post-onset stage. Detailed neuropsychological tests were conducted to document her performance at various levels in the face recognition system and to investigate the possible factors associated with her hyperfamiliarity for unknown faces.

Case history

J. R. was a 21-year-old right-handed (Oldfield's laterality index 100%) female student without previous neurological or psychiatric history. She was born in Switzerland and spent her childhood in Geneva. Following normal education in primary school, she obtained her high school diploma and started studying the history of art and linguistics at university. She was very keen on theatre plays and artistic paintings. She had never experienced any difficulty in person recognition or memory in the past.

J. R. was admitted to hospital because of sudden headaches, nausea and a generalized tonic–clonic seizure. She had no neurological deficit, in particular no visual field defect, and no language disturbance. Detailed neuropsychological assessment was performed in the first and third weeks after onset. There was only mild word-finding difficulty in the first testing session that resolved on the second session, and slightly impaired short-term verbal memory (Table 1). Verbal and visual learning, frontal executive and basic visuoperceptive functions were intact. Brain CT revealed a haemorrhagic infarct in the left lateral temporo-occipital region in the territory of the vein of Labbé, caused by thrombosis of the left transverse and sigmoid venous sinuses, and it was confirmed by conventional arteriography and MRI (Fig. 1). There was no other brain lesion. A hereditary defect in antithrombin III was diagnosed. After local intravenous thrombolysis, oral warfarin and phenytoin were started; the medical course was favourable, with partial sinus recanalization on magnetic resonance angiography 10 days later. EEG showed slow waves and occasional sharp waves in the left posterior regions during the first week, with rapid improvement and only moderate slowing during the second week. Repeated EEGs were normal afterwards.

In hospital, J. R. had no difficulty recognizing known people and learning new faces but spontaneously complained

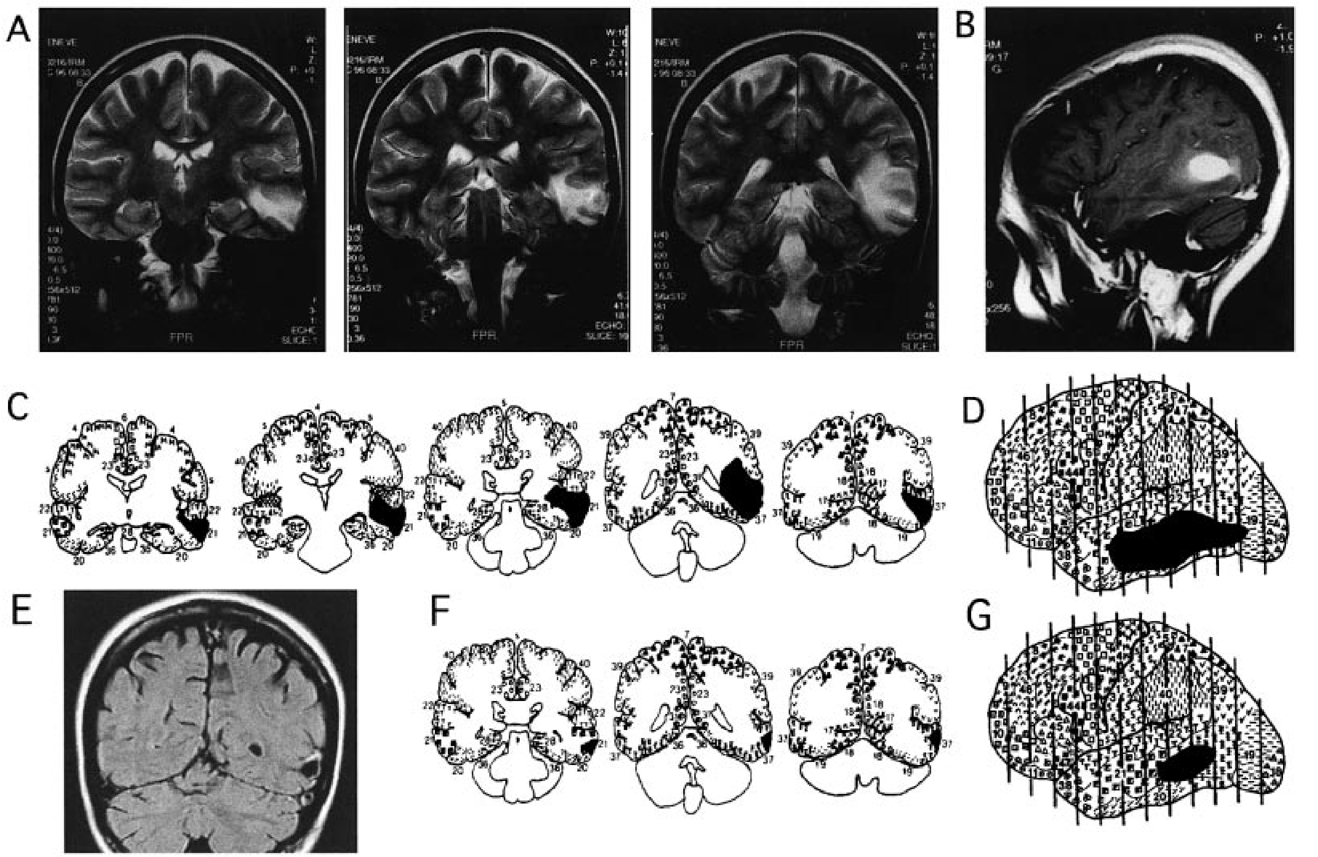

Fig. 1 Brain MRI of the patient 10 days after stroke (**A–D**) and 1 year later (**E–F**). (**A**) T_2-weighted coronal and (**B**) T_1-weighted sagittal images showing haemorrhagic infarction in the posterior middle and inferior temporal gyri of the left hemisphere, close to the temporo-occipital junction, and in the paraventricular occipital white matter. (**C**, **D**) Corresponding anatomical templates using the method of Damasio and Damasio (1989). The acute lesion was centred on Brodmann areas 37, 21 and 20. (**E**) In the chronic stage, T_1-weighted coronal images show a focal cystic lesion in the same area. (**F**) Corresponding anatomical templates, showing that persistent damage is centred on the middle temporal gyrus.

about continuously experiencing erroneous familiarity for unknown people's faces. Her errors concerned other patients, visitors and hospital staff, and greatly embarrassed her. She often engaged in effortful memory searching to decide that she was mistaken, or had to ask the person whether they knew each other. Such errors were very frequent during the first 3 weeks (almost all encounters during the first week and more than half of new encounters up to 3 weeks, according to the patient) then gradually decreased. These errors were still frequent during the time of our investigations, and persisted to a lesser degree after more than 1 year of follow-up. Initially every person who entered her hospital room was familiar to her, she thought she knew him or her, and she would smile at him or her, expecting the person to sit next to her bed and chat with her. It was quite frightening to her that many visitors did not notice her and continued towards her roommate, an elderly woman she knew she had never met. At these moments she tried to figure out where she knew these people from, and would engage in lengthy memory searches. Was it the father of a school friend she had met once? A person seen in the news? A writer seen in a newspaper? A man met at a recent party? She rapidly became suspicious about her own perception of intimate familiarity with these people, but was still often initially convinced that she knew a face at first sight despite clear evidence to the contrary. On questioning, she admitted that during the first 48 h she might also have experienced on a few occasions a similar feeling that the voices of these unknown persons sounded familiar. However, this feeling of false voice familiarity was much weaker than that of face familiarity; it was always accompanied by simultaneous false familiarity for the face and then faded rapidly. She never experienced any difficulty recognizing voices on the telephone or radio.

During the first grand round on the ward (4 days after admission), J. R. greeted one of the authors (T. L.) by using the familiar form of personal pronoun (*tu*, unusual in French with an unknown physician), smiling at him as if he was somebody known to her, though she could not retrieve his name right away. Realizing her mistake, she excused herself and said: 'Sorry, you must be the professor, it got me again, I cannot trust my own perceptions. When you entered this door I thought I knew you well, well enough so that you would

Table 2 *General face perception abilities*

	Patient J. R.: raw score	Controls: cut-off (mean ± SD)
Benton matching test (Benton *et al*., 1994)		
Total correct	49/54	39 (45.4 ±3.9)
Total time (min : s)	6 : 45	8 : 00 (5 : 33 ± 74 s)
Mooney face closure test (Lansdell, 1968)		
Male/female discrimination	29/30	
Young/old discrimination	30/30	
Bruyer and Schweich battery (Bruyer and Schweich, 1991) subtests		
1. Face/non-face decision	24/24	24 (24.0)
2. Matching isolated facial features	9/9	9 (9.0)
3. Matching identity across different view	10/10	10 (10.0)
4. Matching across different expressions	12/12	12 (12.0)
5. Gender classification	20/20	18 (19.6)
6. Age classification	30/30	26 (29.1)
7. Recognition of facial expressions	12/12	11 (11.7)
8. Familiarity decision		
Total correct	43/48	39 (46.2)
Famous faces correctly recognized	23/24	NA
Unknown faces correctly rejected	20/24	NA
9. Recognition of famous faces		
Semantic information	23/24	15 (20.5)
Naming: spontaneous	20/24	
Naming: cued	23/24	19 (23.0)
Eye-gaze direction (Campbell *et al*., 1990)	42/42	40 (40.3 ± 0.8)
Recognition of caricatures	18/24	9 (12.5 ± 3.2)
Disguise Task (Young *et al*., 1990)	19/24	19 (20.8 ±1.5)
Actors Task		
Total correct	100/110	97 (101 ± 4.0)
False alarms on distractors	1/50	3 (2.0 ±1.0)
Mean reaction time (correct responses)	564 ms	604 (± 225) ms

In all tests, the patient's performance was compared with that of normal subjects of similar age and education level, as available from published norms (Bruyer and Schweich, 1991; Benton *et al*., 1994) or from our own data.

embrace me and call me by my first name. Apparently we do not know each other, but I still have that feeling of having met you many times'. She also explained: 'My eyes are caught by someone's face; I have the strong feeling of knowing him or her without being able to place him or her … I have the feeling we have met in some place or talked together, but I cannot figure out where or when, or what we talked about. This happened to me occasionally before, as to everyone I suppose, but now it is present all the time and with everybody. This occurred with many nurses, I often asked them if we were together at school, or if we were living in the same neighbourhood. Also, I thought I had already met with some of the doctors, long before my illness, yet I don't know when'.

By contrast, correct recognition was confident and associated with immediate retrieval of a specific identity. 'When I truly recognize someone, I have no doubt: I just know who the person is. It is more automatic, more natural. When I am not sure, I give a smile or say hello, I wait and see'.

In the initial phase of her illness she said that the inability to recognize unfamiliarity was practically ubiquitous, concerned most newly encountered people, and was independent of the surroundings within the hospital, whereas her ability to recognize truly familiar persons was never impaired. However, after discharge a few weeks later, it appeared that her errors with unfamiliar faces were influenced by the situational context. Familiarity errors were less common when she was walking in the street or using public transport, but increased specifically when she was on the university campus, in her neighbourhood or visiting the hospital during follow-up. J. R. never misidentified a person for another and never thought people were disguised. Her familiarity errors with unknown faces were quite dramatic during the first 2 weeks and then decreased gradually over a period of months, but were still present after 1 year of follow-up (10% of new faces, in the patient's estimation). Subsequent MRI scans 6 and 12 months after onset showed considerable shrinking of the lesion, with a chronic infarct centred on the posterior middle temporal gyrus (Fig. 1E and F).

In summary, this striking disturbance in which unknown faces are appraised as familiarity from unknown faces constitutes a novel clinical phenomenon, hitherto never reported, caused by an unusual lesion outside the common

Table 3 *General memory abilities*

	Patient J. R.: raw score	Controls: cut-off (mean ± SD)
Recognition memory test (Bindschaedler *et al*., 1996)		
1. Faces		
Hits (H)	34/40	(34.3 ± 5.1)
False alarms (FA)	4/60	(3.1 ± 2.8)
Total correct (H – FA)	30	22 (31.1 ± 5.5)
Discrimination (d′)	4.19	(2.8 ± 0.6)
Response bias (C)	0.43	(0.31 ± 0.2)
2. Words		
Hits (H)	35/40	(37.8 ± 2.8)
False alarms (FA)	0/60	(0.8 ± 0.8)
Total correct (H – FA)	35	28 (37.0 ± 2.9)
Discrimination (d′)	6.66	(3.7 ± 0.4)
Response bias (C)	1.47	(0.34 ± 0.32)
Doors and People Test (Baddeley *et al*., 1994)		
People	30/36; percentile 60	18 (28)
Doors	24/24; percentile >99	14 (19)
Names	16/24; percentile 10	15 (19)
Semantic knowledge of people (Kapur *et al*., 1992; Bindschaedler *et al*., 1995)		
Dead–Alive People test: total score	87/115	80 (99)
1. Dead/alive decision	43	39 (43)
2. Date of death	16	20 (25)
3. Mode of death	28	26 (30)
4. Profession	44	NA; max. 45
5. Achievement	42	NA; max. 45
6. Nationality	41	NA; max. 45

In all tests, the patient's performance was compared with that of normal subjects of similar age and education level, as available from published norms (Baddeley *et al*., 1994; Bindschaedler *et al*., 1995) or our own data.

arterial territories involved in ischaemic strokes. It provides a unique piece of evidence for an important function of the left cerebral hemisphere in the recognition of faces and in mechanisms of familiarity. Below, we report our examination of various aspects of face processing in J. R. using a large battery of standard tasks that are commonly used in prosopagnosia, and a number of *ad hoc* experiments.

General face processing and memory abilities

All the following experiments were performed during the third week after stroke to test for the presence of prosopagnosia, i.e. an impairment in the ability to discriminate and remember familiar faces. Several standardized tests were given to probe (i) basic face perception abilities (Table 2) and (ii) general memory abilities for faces, people and other complex visual material (Table 3). As indicated in Table 2, testing for general face perception abilities comprised a battery of nine subtests assessing separate recognition processes, including a familiarity decision task that required classification of a series of faces as famous or unknown [the Bruyer and Schweich battery (Bruyer and Schweich, 1991)], a test of configurational facial organization (Lansdell, 1968), the discrimination of individual faces in different views (Benton *et al*., 1994), and the perception of gaze direction (Campbell *et al*., 1990).

A few other special tests assessed the identification of faces with an unusual appearance. Recognition of caricatures (e.g. of politicians) was examined because it has been suggested that memory representations of familiar faces may exaggerate distinctive physiognomic features, as do caricatures (Rhodes *et al*., 1987), and a defect in extracting such distinctive features might conceivably induce abnormal familiarity with unknown people. Also, we used a disguise task (Young *et al*., 1990) that required matching unfamiliar faces masked by various disguises (e.g. glasses or a false beard), and a similar task, the Actor Task, that required recognition of famous actors in different roles (e.g. with different make-up or age). Patients with delusional misidentification are particularly impaired on such tasks, whereas they have no difficulty recognizing undisguised faces (Young *et al*., 1990).

General memory abilities were assessed with a recognition memory test for faces and words (Bindschaedler *et al*., 1996), the Doors and People Test (Baddeley *et al*., 1994), and structured questionnaires assessing semantic knowledge about familiar people and celebrities (Ellis *et al*., 1989; Kapur *et al*., 1992), adapted and standardized for the Swiss population (Bindschaedler *et al*., 1995).

Results

The patient's performance in these standardized tests was well within the normal range (Table 2). She had no impairment in matching unfamiliar faces or identifying familiar faces (i.e. no prosopagnosia). She made no misidentification of known or unknown faces. Caricatures, faces in disguise and actors were all recognized correctly. Similarly, her scores in the visual recognition memory test were in the normal range (Table 3). False alarms were not increased in the face memory task (Table 3), and signal detection measures (Bindschaedler *et al.*, 1996) showed excellent discrimination ability (high value of d′, corresponding to efficient discrimination of signal from noise), without a significant response bias in making recognition judgements compared with the normal range (loose criteria for familiarity decisions correspond to small or even negative values of C; see below). Note, however, that her response criteria appeared to be higher for words than faces, unlike normal controls (Table 3). Finally, retrieval of biographical information about well-known people in the Dead–Alive test (Kapur *et al.*, 1992) did not differ from results for normal subjects (Table 3); her slightly poorer performance on dates of death was probably due to the patient's young age (Bindschaedler *et al.*, 1995). Structured interviews with the patient and her family (Ellis *et al.*, 1989) revealed no loss in semantic knowledge about close relatives.

From these tests and the clinical picture of the patient, who never mistook for a stranger somebody she knew or failed to name a person really familiar to her, it can be concluded that she was not prosopagnosic and had no basic perceptual difficulties with faces. Therefore, her difficulties with face familiarity must have had an origin other than a deficit in the basic visual analysis of faces or memory.

Global and featural visual processing of faces

Several additional tests were designed to examine face recognition beyond the basic perceptual stages. All these tests were administered between the fourth and eighth weeks after onset. During this period, the patient was still making frequent familiarity errors with unknown faces in everyday life, and control MRI scans demonstrated that a chronic lesion restricted to the middle temporal gyrus persisted up to 1 year after onset (Fig. 1E and F).

The following experiments probed specifically for an impairment in encoding local compared with global traits in faces. It is known that the left hemisphere is preferentially involved in parsing local features, while the right hemisphere is more concerned with global configurational traits (e.g. Hillger and Koenig, 1991; Tanaka and Farah, 1993; Rossion *et al.*, 2000). As shown by Yarbus (1967), visual exploration of faces is driven strongly by internal parts, such as the eyes, nose and mouth. These internal parts are more important than external features or whole contours in the recognition of known faces, whereas both internal and external features are equally important for unfamiliar faces (Ellis *et al.*, 1979; Young *et al.*, 1985*b*). Left hemisphere damage might have disrupted the processing of local features, leading to false recognition based on the predominance of right-hemispheric processing of global or external traits. The three tasks described below (Inverted Face, Face Parts, and Altered Face) were used to compare the roles of parts and wholes in face recognition and familiarity judgements.

Whenever possible, performance was measured not only by accuracy and error rates, but also by signal detection measures of discrimination and response bias. Indeed, J. R.'s problem involved a false feeling of familiarity for persons without real personal meaning, i.e. she made false alarms and accepted new faces as already known, but did not fail to recognize previously known or famous faces (i.e. she was not prosopagnosic). Signal detection measures allowed us to take the whole of her decision behaviour into account, by calculating d′ values for her visual discrimination sensitivity and C values for her personal response criteria strategy (Green and Swets, 1966). At a d′ value of zero it can be assumed that the observer cannot distinguish between a signal and background noise. Increasing d′ values indicate that the signal is more salient and more easily recognized. On the other hand, a low C value indicates that the observer accepts less salient signals as possible hits, and thus makes errors but also achieves a high rate of correct answers. High C values indicate a more strict decision threshold, with fewer mistakes but also a lower hit rate. To compare J. R. with normal subjects, we calculated the confidence intervals for d′ and C values from the means and standard deviations obtained for the normal subjects. As in the method of Hirsig (1998), we used t values instead of Z values because of the small control sample (e.g. six or eight subjects). Values of t corresponding to the appropriate number of degrees of freedom (d.f.) were taken from tables provided by Hirsig (1998), with the 95% level set at $t = 2.57$ for six subjects and $t = 2.37$ for eight subjects. We calculated first the estimated standard deviation ($\sigma = s/\sqrt{\text{d.f.}}$, where σ is the standard deviation of the control sample), and then the confidence interval for the mean ($CI = \mu \pm t_{95\%} \times \sigma$, where μ is the mean value of the control sample). This confidence interval is the range of values within which 95% of the general population would be likely to lie, given the sample. We assumed that the performance of our patient was abnormal if it was outside this interval.

Inverted face task

This test compared recognition of upright and upside-down faces. Extraction of salient local features is more important for upside-down than for upright presentation (Yin, 1969). Inversion can remove the right hemisphere superiority normally found with upright faces (Hillger and Koenig, 1991). Thus, defective encoding of local features should disturb recognition of inverted faces disproportionately. In this task, black-and-white photographs of 20 unknown upright faces were shown sequentially for study (1 s each).

Table 4 *Recognition of inverted and upright faces*

Faces	No. of 'old' responses J. R. (controls) Correct	False alarms	d′ value J. R	Controls 95% CI	C value J. R.	Controls 95% CI
Inverted	11 (12.8 ± 1.8)	8 (6.5 ± 1.2)	**0.4**	0.6 to 1.0	0.1	–0.1 to 0.2
Upright	10 (12.5 ± 1.3)	1 (3.5 ± 1.9)	1.6	0.9 to 1.7	**0.8**	0.1 to 0.5

Controls were eight age- and education-matched normal subjects (five females and three males, mean age 27.5 years). Significant differences between patient and controls are indicated in bold. CI = confidence interval.

Then, the same (old) faces and an equivalent number of new faces were briefly presented upside-down (500 ms) in random order. Familiarity (old/new) was indicated for each face by a key press. In a third phase, the old faces and another set of new distractors were presented in the normal upright position using the same procedure. Response time and accuracy were recorded.

Results

The number of correct 'old' responses made by J. R. was within the controls' range for both inverted and upright faces, but she tended to produce a greater proportion of incorrect 'old' responses for inverted faces (Table 4). Inversion decreased face recognition sensitivity (d′) in J. R. to a greater degree than expected from the decrease in control subjects. Her response criterion (C) did not differ from that of controls for inverted faces, but was more conservative for upright faces. Her mean reaction time for correct responses (1493 ms) was comparable to that of controls (1331 ± 574 ms). This pattern would be consistent with a mild difficulty arising when recognition must rely more on the left hemisphere for processing internal facial features.

Face parts task

This task compared recognition of facial parts and wholes in separate phases. We used two versions (given a few days apart), which used either famous or unknown faces. Each version employed 24 black-and-white photographs of faces that were cut out into four parts (hair, eyes, nose and mouth) (Fig. 2A), yielding a total of 96 stimuli. In the study phase, 12 full target faces were presented on a computer screen (5 s each). Two test phases immediately followed. First, all 48 parts from the target faces were presented in random order mixed with 48 distractor parts from the other 12 faces. Subjects made familiarity decisions (old/new) with a key press. Stimuli remained on the screen until there was a response. Immediately afterwards, a second test phase assessed the recognition of whole faces. All target and distractor faces were presented once again, but now in wholes. Subjects made familiarity decisions (old/new) as in the first test. Response time and accuracy were recorded.

Results

J. R. showed no obvious deficit in recognizing single facial features from either famous or unknown people (Table 5). Signal detection values confirmed that her discrimination sensitivity for facial parts and wholes was within or even above the normal controls' range in all conditions. However, she made more incorrect 'old' judgements on 'new' nose and mouth parts [$\chi^2(1) = 4.0$, $P = 0.04$] and used looser response criteria for these parts compared with normal subjects. Her mean reaction time for correct responses (1684 ms) was similar to that of controls (1651 ± 813 ms). These data suggest that, despite her well-preserved visual discrimination abilities, J. R. might have experienced some uncertainty when using only local feature information as the basis for the recognition of face familiarity, especially when such facial features were new.

Altered face task

This task pitted recognition of parts and wholes against each other within the same stimulus. It consisted of a continuous recognition memory test in which 60 faces recurred once each, intermingled in random order with 60 distractors (180 trials in total). Four additional faces served as a buffer at the beginning of the test. All faces were from unknown persons. Intervals from first presentation to repetition varied between 10 and 60 intervening stimuli (mean 34). On repetition trials, the target faces were either unchanged or altered at the internal (eyes, mouth), external (hair) or global (whole face) level (12 trials in each condition; Fig. 2B). All stimuli were shown sequentially on a computer screen (1 s). Subjects were required to indicate whether the face of any given person was repeated or seen for the first time by pressing one of two keys (old/new).

Results

Table 6 shows that J. R. recognized the recurrence of unchanged faces as well as normal subjects did [75 versus 83%; $\chi^2(1) = 0.09$, $P = 0.75$] and made fewer false alarms to new faces (19 versus 29%). The rate of 'old' response to faces that were altered on repetition trials was significantly affected by the type of change, both in J. R. [$\chi^2(3) = 15.6$, $P = 0.001$] and in normal subjects [$\chi^2(3) = 8.0$, $P = 0.050$]. Such 'old'

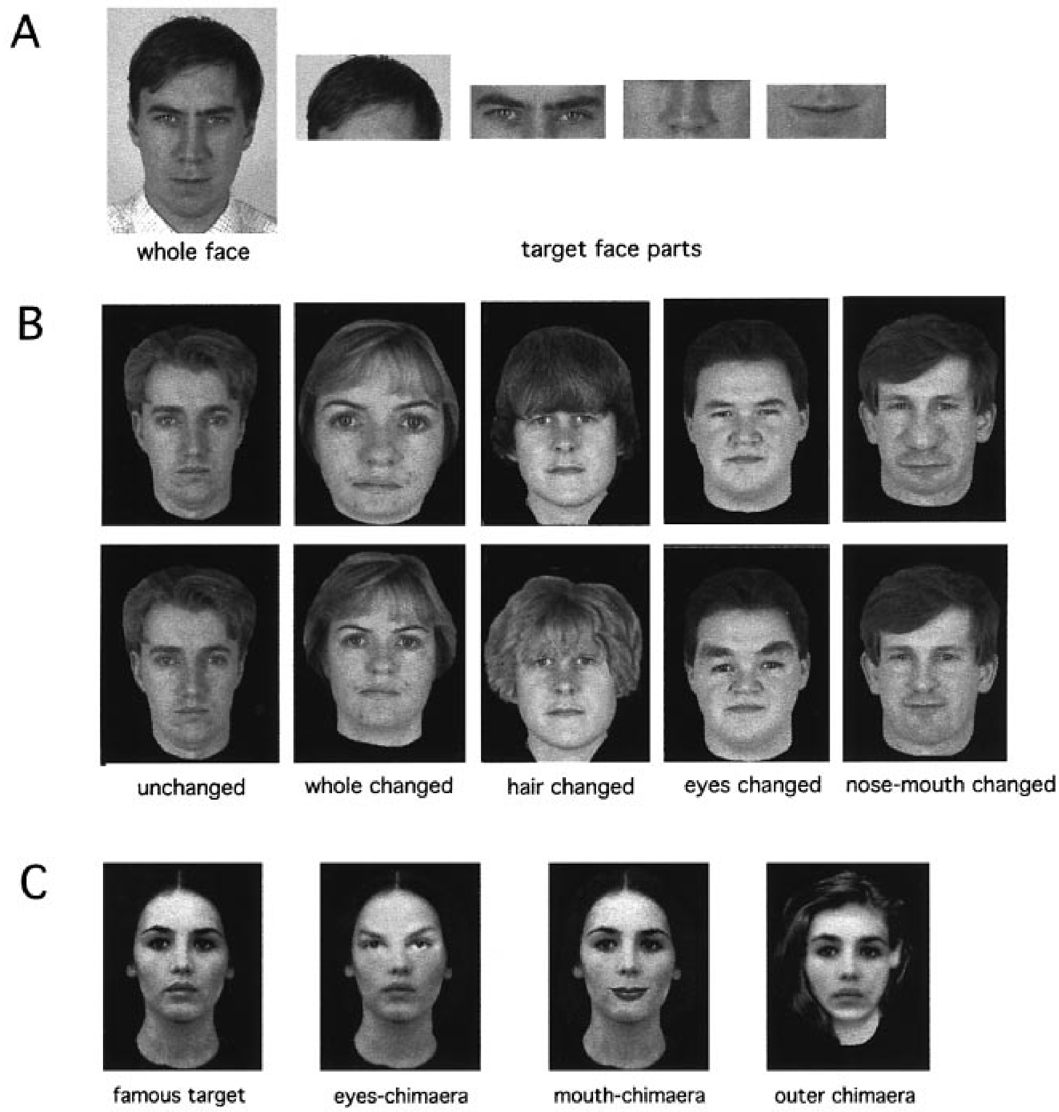

Fig. 2 Examples of stimuli used to test global and local visual processing. (**A**) Face parts. Unfamiliar whole faces were presented in the study phase and their parts (hair, eyes, nose, mouth) were presented in a subsequent recognition test. A similar task employed famous faces. (**B**) Altered faces. From first presentation (*upper row*) to repetition (*lower row*), target faces were either unchanged or altered at the global (whole face), external (hair) or internal (eyes, mouth) level. (**C**) Chimaeric faces. Three kinds of chimaera were constructed (the French actress Isabelle Adjani is shown as an example), using the global contour and hair from a known face but changing the eyes or mouth (inner chimaeras), or vice versa (outer chimaeras). The original face itself was not presented, and a given individual appeared only once. For the patient, similar chimaeras were constructed using both famous faces and faces of relatives.

responses to altered faces were lower in J. R. than in normal subjects when the hair was changed (25 versus 47%), but higher when the eyes or mouth were changed (75–83 versus 54–67%), and identical when the whole face was changed globally (58%). This pattern would be expected if J. R. recognized familiarity by relying more on external and global components of faces, and less on internal features (eyes and mouth) compared with normal subjects. The total difference between external and internal changes was significant in J. R. (10 versus 19 faces recognized out of 24, $P = 0.002$, Fisher's test) but not in controls (mean 12.6 versus 14.5 recognized, $P = 0.15$). Her mean reaction time for correct responses (1382 ms) was also similar to that of controls (1472 ± 667 ms).

Chimaeric face task

The above results are partly consistent with an impairment of face processing involving the left hemisphere, possibly resulting in defective encoding of internal features and abnormal reliance on global components. However, all the preceding tests examined judgements of familiarity in conditions in which the patient was required

Table 5 *Recognition of facial parts and wholes*

	No. of old responses J. R. (controls)		d′ value		C value	
	Correct	False alarms	J. R.	Controls 95% CI	J. R.	Controls 95% CI
Unknown faces						
Hair	10 (8.8 ± 1.3)	1 (3.3 ± 2.1)	**2.4**	0.8 to 2.0	0.2	–0.4 to 0.5
Eyes	7 (7.3 ± 1.3)	2 (2.3 ± 1.6)	1.2	0.7 to 2.1	0.4	0.0 to 0.8
Nose	9 (5.3 ± 1.9)	**6** (3 ± 1.5)	0.7	0.2 to 1.1	**–0.3**	0.0 to 0.9
Mouth	8 (6 ± 1.7)	**4** (2.5 ± 1.6)	0.9	0.4 to 1.5	**0.0**	0.1 to 0.9
Whole	12 (11.5 ± 0.5)	0 (0.5 ± 0.8)	**4.7**	3.4 to 4.1	0.0	–0.4 to 0.5
Famous faces						
Hair	11 (10.5 ± 1.2)	0 (0.5 ± 0.5)	3.7	2.3 to 4.1	0.5	0.0 to 0.5
Eyes	12 (9.8 ± 1.4)	0 (0.8 ± 0.5)	**4.7**	2.0 to 3.1	0.0	0.0 to 0.6
Nose	9 (6.4 ± 1.8)	1 (2.4 ± 1.2)	**2.1**	0.2 to 1.8	0.4	0.2 to 0.6
Mouth	12 (10.0 ± 1.3)	0 (2.5 ± 1.8)	**4.7**	1.1 to 2.9	0.0	–0.3 to 0.2
Whole	12 (12)	0 (0)	4.7	4.7	0.0	0.0

Controls were eight age- and education-matched normal subjects (five females and three males, mean age 27.5 years). Significant differences between patient and controls are indicated in bold. CI = confidence interval.

Table 6 *Recognition of altered faces*

	No. of 'old' responses	
	J. R.	Controls (mean ± SD)
Repeated faces		
Unchanged	9/12	10 ± 0.8
Global change	7/12	7 ± 2.7
Hair change	3/12	5.6 ± 1.8
Eyes change	10/12	8 ± 1.7
Mouth change	9/12	6.5 ± 2.6
New faces	23/120	34.5 ± 15.4

Controls were eight age- and education-matched normal subjects (five females and three males, mean age 27.5 years).

to recognize whether she had already seen a given face rather than judge whether any particular face was from a person known to her. Critically, her disturbing feeling of abnormal familiarity with strangers in real life was associated not only with an impression of past occurrence, but also with some aspect of personal relevance and more intimate knowledge. Therefore, if preferential encoding of global and external components at the expense of internal features were sufficient to induce such a false familiarity, our patient would be expected to judge mistakenly that she knew chimaeric faces who have familiar external features and incorrect internal features, but not faces with the reverse chimaeric manipulation (i.e. familiar internal and unfamiliar external features).

In this task, we constructed chimaeric faces from photographs of 14 relatives of the patient, 14 famous actors and 21 unknown people. Seven faces in each category served as unchanged stimuli, while the others were employed for chimaeric stimuli. There were three possible chimaeras for both relatives and famous people (Fig. 2C): outer contour chimaeras (global contour and hair from an unknown face with eyes and mouth from a known face); inner eyes chimaeras (unknown eyes in known face contours); and inner mouth chimaeras (unknown mouth in known face). Each individual face and parts were used only once, resulting in 63 stimuli. All faces were projected briefly (800 ms) on a computer screen and familiarity judgements (known/unknown) were made by key press as quickly as possible. Controls were tested with unknown and famous faces only.

Results

J. R. recognized real non-chimaeric faces normally, and showed no increase in false alarms or response times to chimaeric faces with familiar global contours (Table 7). There was no difference for famous faces known through the mass media and faces of personally known relatives. This indicates that J. R. could still correctly assess the familiarity of faces using both internal and global features within a single face, and did not mistakenly recognize a face with familiar global features as a known face.

Therefore, although the preceding experiments suggest that the patient tended to rely more on face-processing abilities subserved by the right than the left hemisphere, with the patient showing better encoding of external than internal features when the recognition of familiarity or a previous encounter with a face was required (e.g. in the inverted face and altered face tasks), this visual bias did not seem sufficient to induce a false feeling of personal acquaintance with unknown faces, as otherwise exhibited by J. R. in her everyday life.

Associative and semantic processes in face recognition

The previous series of experiments showed that visual facial processing was well preserved in J. R. overall. Preferential

Table 7 *Recognition of chimaeric faces*

	No. of 'known' responses	
	J. R.	Controls (mean ± SD)
Unknown people	2	0 ± 1
Famous people		
Real	6	6 ± 1
Outer contour chimaeras	1	1 ± 1
Inner eyes chimaeras	1	1 ± 1
Inner mouth chimaeras	0	0
Relatives		
Real	7	(max. = 7)*
Outer contour chimaeras	0	(max. = 7)*
Inner eyes chimaeras	0	(max. = 7)*
Inner mouth chimaeras	0	(max. = 7)*
Mean reaction time (correct)	1029 ms	1131 ± 226 ms

Controls were four age- and education-matched normal subjects (four females, mean age 27.4 years). *Controls were tested only with unknown and famous faces, whereas J. R. was also tested with faces of family relatives.

Table 8 *Misattribution of fame*

	No. of 'fame' responses	
	J. R.	Controls (mean ± SD)
Faces		
Real fame	11/20	12.7 ± 2.4
New lures	3/20	2.2 ± 3.3
Old lures	**16**/20	6.3 ± 0.5
Names		
Real fame	11/20	16.2 ± 0.8
New lures	1/20	1.7 ± 0.5
Old lures	6/20	5.3 ± 1.2

Controls were six age- and education-matched normal subjects (five females and one male, mean age 25.0 ± 3.7 years). The bold figure indicates a significant difference between patient and controls.

engagement of the right hemisphere for global visual analysis of faces, with relatively deficient encoding of local features in the left hemisphere, could not fully account for her abnormal feeling of personally knowing new faces. We therefore designed the next series of experiments (performed 7 and 8 weeks after onset) to investigate whether impaired processing of faces in the left hemisphere might affect a stage of stimulus analysis beyond purely visual perceptual encoding, at the level where associations with representations stored in memory are formed and used to access specific information about personal identity.

False fame and prior face exposure

Failure to activate appropriate semantic representations can lead to false recognition and misattribution of familiarity when retrieval of such information is necessary in order to recognize (or reject) the source of subjective familiarity induced by erroneous cues (Jacoby *et al*., 1989*a*; Whittlesea and Williams, 1998). In normal people, previous exposure to unknown names (Jacoby *et al*., 1989*b*) or unknown faces (Bartlett *et al*., 1991) can generate such a feeling of familiarity and induce erroneous judgements of celebrity, especially when the individual fails to recollect the context associated with prior exposure. We examined whether J. R. would be abnormally prone to false familiarity effects under such conditions.

In this task, a series of 60 faces were shown sequentially and had to be judged as famous or unknown. No specific biographical information or name was required. The stimuli included 20 people with real but modest fame (e.g. minor local politicians, second-rank artists etc.), together with 20 unknown people who had been presented only once as distractors in a previous test (old lures), and 20 other unknown people who had not been seen before (new lures). Stimuli were presented in random order without time constraint.

Results

J. R. and normal controls made correct fame judgements for 55 and 63% of faces with real, though modest celebrity, respectively (Table 8). For new lures (i.e. faces that had never been seen before), J. R. made false fame judgements as often as the controls (15 and 11%, respectively). Critically, when confronted with the old lures (i.e. unknown faces that had already been seen in a prior experiment), both J. R. and normal controls made more false fame judgements compared with the new lure faces [control mean: 32 versus 11%, $\chi^2(1) = 15.6$, $P = 0.0001$; patient mean 80 versus 15%, $P = 0.0001$, Fisher's test] (Table 6). However, this increase in false fame judgements for old lures was much higher in J. R. than in normal subjects [$\chi^2(1) = 14.9$, $P = 0.0001$], whereas her ability to reject new lure faces and to recognize real famous faces did not differ from that of normal subjects [$\chi^2(1) = 0.03$ and 0.21, respectively]. These data reveal that J. R. was severely impaired in attributing a correct source to false signals of familiarity with unknown faces, and showed a marked bias towards considering unknown faces as known by fame based on coarse signals of subjective familiarity.

False fame and prior name exposure

To determine whether our patient's abnormal increase in false familiarity judgements was specific to faces or reflected a more general bias in assessing the source of subjective familiarity, a second task using names was given to her and the same controls in a different session. As in the previous face task, 60 proper names (e.g. Paul Martin) were presented singly and had to be judged as famous or unknown. The names included those of 20 people with real but modest fame (e.g. minor politicians or artists), 20 unknown people already seen as distractors in a previous test (old lures), and 20 unknown people never seen before (new lures). Stimuli were presented in random order without time constraint.

Results

J. R. correctly recognized real fame and correctly rejected new lures and controls did also (Table 8). Again, a significant increase in false fame judgements occurred for old lures (i.e. previously exposed unknown names) compared with new lures, both in normal controls [mean 27 versus 8%, $\chi^2(1) = 14.7$, $P = 0.0001$] and in the patient (30 versus 5%, $P = 0.050$, Fisher's test). However, the rate of such errors was not different between J. R. and controls [$\chi^2(1) = 0.06$, $P = 0.80$]. This indicates that the patient's deficit in associating subjective familiarity with a correct source was not present with names, but concerned faces selectively.

Associative processing and priming

In the prior exposure experiments, our patient showed an abnormally high rate of familiarity misattribution for faces that were unknown but previously seen in a different context (but no such errors for names). This result reveals a marked bias towards considering unknown faces as famous when coarse signals of familiarity were elicited but failed to activate more specific information in memory (e.g. to identify the source of familiarity). This led us to consider whether J. R. had a particular deficit in activating stored representations that give access to identity-specific knowledge about a face. This was examined in two different tests of face priming, as used previously in normal subjects (Bruce *et al*., 1993; Young *et al*., 1994) and prosopagnosics (De Haan *et al*., 1992): (i) visual repetition priming, which refers to facilitated recognition of a face that has been seen previously and reflects activation of the visual representation of this face (Bruce *et al*., 1993), possibly involving the right hemisphere (Grüsser and Landis, 1991); and (ii) associative semantic priming, which involves facilitation of recognition based on some information linked to a given person (such as the name of the person or someone related), reflecting an activation of specific identity knowledge, is maximal when the prime and target concern the same individual (Young *et al*., 1994), and possibly implicating more the left hemisphere (Rhodes, 1985).

Each test was given in a separate session. For visual priming (face–face task), a *face* was first shown for 2 s (to be name aloud), and was immediately replaced by a second face (target) that remained on the screen until a familiarity decision (known/unknown) was made as quickly as possible on the latter by pressing one of two keys (Fig. 3A). For associative priming (name–face task), a *name* was first shown for 2 s (to be read aloud), again it was replaced by a face (target) that remained on the screen until a speeded familiarity decision (Fig. 3B). Thus, in both tasks, familiarity decisions were always made on a face, but the face was preceded either by another face (face–face visual priming) or a written name (name–face semantic priming). Faces (and names) of 30 celebrities from three categories (politicians, actors, musicians) were selected to create 15 pairs of associated people, while 30 unknown faces (and 30 unknown names) served as unfamiliar stimuli. In each task, a famous face could be preceded by four possible primes (30 trials per condition): (i) the same person (but different pictures in the face–face task to prevent same-stimulus repetition); (ii) a related celebrity from the same category; (iii) a celebrity from another category; or (iv) a neutral unknown person. Unknown faces were also presented as fillers, preceded by either a known celebrity or an unknown person (30 trials per condition). All trial types occurred in random order. The critical conditions concerned the effect of identity-specific priming for known faces, i.e. familiarity decisions that were made on a known face preceded by either the face or the name of the same person.

Results

Error rates were low in J. R. (4%) and healthy controls (8%) in both tasks. Median correct reaction times (RTs) were computed for each subject in each condition. Overall RTs were similar in J. R. (mean 691 ms) and controls (mean 677 ms) in each task (unpaired t test, $P > 0.45$). Priming effects were assessed by repeated-measures ANOVA (analysis of variance) on correct RTs to known faces, with the different prime types as a factor, first across and then within individual controls to allow comparison with the patient.

In the face–face task, healthy controls showed reliable visual priming (Table 9). There was a significant effect of the preceding face type on RTs [$F(3,5) = 3.79$, $P = 0.03$], due to faster responses in the same-person condition compared with others [$t(5) > 2.94$, $P < 0.05$]. There was no significant effect of same-category faces. J. R. showed the same pattern of significant priming effects (Table 8) ($F = 2.34$, $P = 0.045$ in two different sessions), with faster responses after same-person faces than after other face types ($P < 0.05$). The same results were obtained in a second session 2 weeks later, in which faces from close relatives (family and friends) were added to those of celebrities, using the same four priming conditions for the relatives and famous people as above (Table 8). Healthy subjects from the same family showed similar performance for both relatives and celebrities. These results demonstrate intact visual identity-priming in J. R. (Bruce *et al*., 1993).

In the name–face task, healthy controls again showed significant priming effects (Table 9) [$F(3,7) = 7.36$, $P = 0.002$]. Their RTs were faster after same-person names than in other conditions [$t(7) > 3.1$, $P < 0.02$], consistent with facilitation of face recognition by the activation of identity-specific associations (Young *et al*., 1994). There was no significant effect of same-category names. These effects were found in each individual

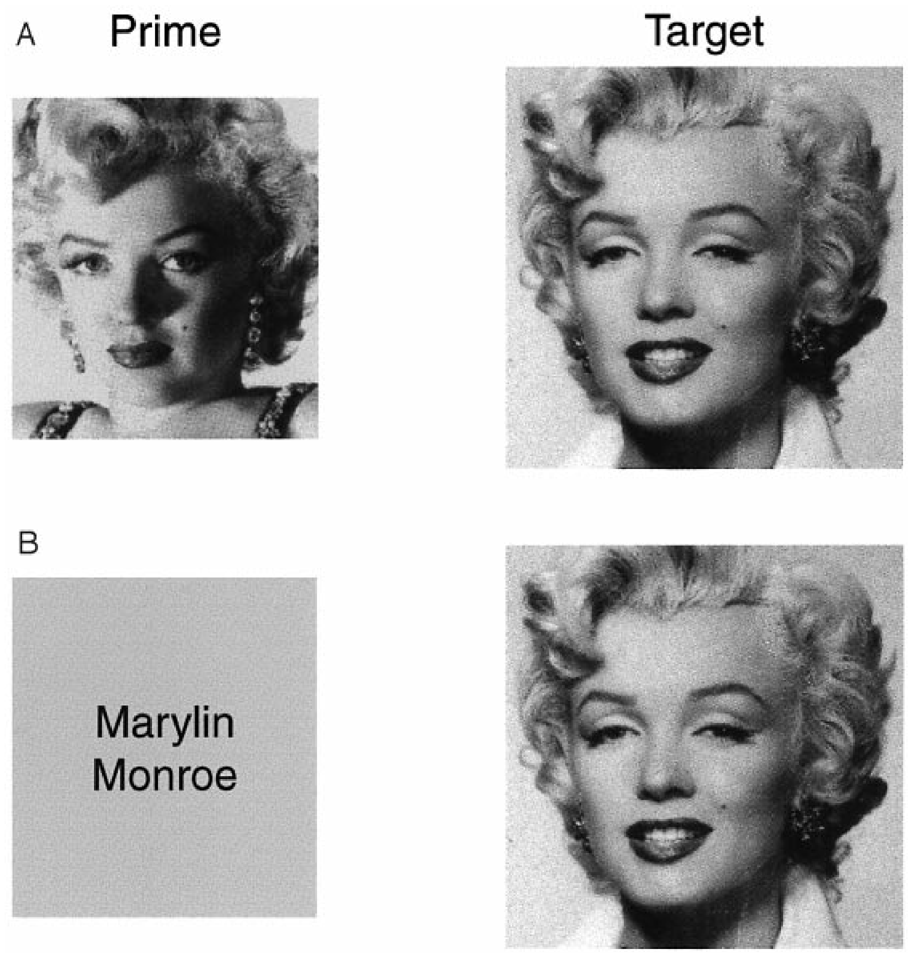

Fig. 3 Example of stimuli used to test face recognition priming. (**A**) Visual repetition priming (face–face task). On each trial, a target face was preceded by the face of the same or another person. (**B**) Associative semantic priming (name–face task). On each trial, a target face was preceded by the name corresponding to the same or another person. In both tasks, the subject had to make a speeded familiarity judgement (known/unknown) on the target face.

control. By contrast, J. R. showed a very abnormal pattern (Table 8). Whereas different name types had an effect in the first session ($F = 2.72$, $P = 0.022$) but not in the second session ($F = 1.23$, $P = 0.28$), in both instances the same-person names tended to slow rather than facilitate familiarity decisions compared with neutral, unknown names ($P < 0.05$). Impaired identity-priming was replicated in both sessions for celebrities, and was similar for faces of relatives and celebrities in the second session (Table 8). Altogether, these priming results suggest a disturbance in activating identity-specific semantic associations in the patient when she made judgements of familiarity, despite normal activation of the visual representation of known faces (Bruce *et al*., 1993).

Discussion

We describe a young woman who, subsequent to a left temporal venous infarction, exhibited a disturbing feeling of familiarity, even intimacy, towards faces of people unknown to her. Although she had no difficulty recognizing people who were already known, she mistook many new faces as belonging to people with whom she was well acquainted. Later, this phenomenon partly abated but remained more frequent in familiar surroundings, even 1 year after onset. In many respects, this disorder presented as the reverse of prosopagnosia. Whereas prosopagnosics typically have right inferior temporal lesions and no longer recognize familiar faces (Landis *et al*., 1986, 1988; De Renzi *et al*., 1994), our patient suffered left lateral temporal damage and experienced exaggerated familiarity with unknown faces, even though she could still identify known faces and learn new ones. Occasional feelings of false familiarity with unknown faces are not uncommon in normal people, though they are usually faint and are rapidly acknowledged as a trivial error (Young *et al*., 1985*a*). These impressions rarely lead to the conviction of intimately knowing a stranger, to the point of searching at length for any possible links, as in our patient. As an acute

Table 9 *Visual and semantic priming of face recognition*

Type of prime condition	Median reaction time (ms)			
	Face–face task (visual priming)		Name–face task (semantic priming)	
	Patient J. R.	Controls*	Patient J. R.	Controls*
Session 1: faces of celebrities				
Same person	605	624 ± 111	**817**	576 ± 39
Related person	672	744 ± 193	798	683 ± 71
Unrelated person	673	751 ± 200	662	646 ± 75
Unknown person	754	689 ± 188	704	679 ± 100
Session 2: faces of celebrities and relatives				
Celebrities				
Same person	634	468 ± 91	**741**	454 ± 45
Related person	708	610 ± 88	619	600 ± 64
Unrelated person	801	684 ± 82	717	669 ± 75
Unknown person	860	702 ± 99	595	561 ± 138
Relatives				
Same person	676	425 ± 29	**654**	435 ± 72
Related person	788	526 ± 55	607	484 ± 29
Unrelated person	721	585 ± 36	751	559 ± 56
Unknown person	787	592 ± 66	656	553 ± 109

*Mean ± SD of median reaction time. Controls were six age- and education-matched normal subjects (five females and one male, mean age 26.2 ± 4.0 years) in the face–face task of session 1; eight matched normal subjects (six females and two males, mean age 26.3 ± 4.5 years) in the name–face task of session 1; and four normal subjects of the same family (two females and two males, mean age 31.7 ± 4.6 years) in both tasks of session 2. Significant deficits between the patient and controls are indicated in bold.

neuropsychological impairment, in direct relation to a focal cortical lesion, this phenomenon has never been reported previously.

Using standardized and *ad hoc* tests, we tried to clarify the nature of this disorder. Tests designed to uncover various forms of prosopagnosia showed that our patient's face recognition abilities were in the upper part of the range of normal subjects. The same was true for memory, basic visual perception and a comprehensive neuropsychological examination. Thus, J. R. was clearly not prosopagnosic and her false familiarity could not be explained by a basic recognition deficit or general cognitive disturbances. Furthermore, her errors did not involve the misidentification of specific individuals and delusional elaboration, as in Frégoli syndrome (Ellis and Young, 1990). False face recognition has rarely been observed without prosopagnosia in patients with right brain damage and piecemeal visual processing, in whom misidentification may result from resemblance to the local facial features of a known person (Young *et al*., 1993; Rapcsak *et al*., 1994). Misidentification can also occur without visual deficit in patients with frontal damage and confabulation, due to impaired monitoring (Rapcsak *et al*., 1998, 1999) or inappropriate semantic associations (Ward *et al*., 1999). These conditions all differ clearly from those of our case. J. R. showed no frontal dysfunction, confabulation or delusion. She performed normally on a variety of discrimination, identification and memory tasks with faces and showed normal visual-repetition priming for known faces, suggesting preserved perceptual input and preserved formation of structural facial representations.

Among the variety of tests tapping different levels of face processing, three significant deficits were found in J. R.: (i) deficient use of internal face parts to guide recognition; (ii) lack of face-identity priming by known names during familiarity judgements; and (iii) abnormal misattribution of fame to unknown faces after incidental prior exposure. We suggest that all three deficits may similarly point to a deterioration of face processing involving the left hemisphere and greater reliance on right hemisphere function.

First, J. R. showed difficulty recognizing upside-down faces (inverted face task), with a tendency to rely more on external and global features and less on internal parts (altered face task) compared with normal subjects. It is known that inversion suppresses the right-hemispheric advantage for face recognition and requires the greater featural analysis mediated by the left hemisphere (e.g. Hillger and Koenig, 1991). Moreover, although J. R. exhibited normal discrimination sensitivity (d′) for recognizing isolated features (face part task), her response criterion (i.e. her subjective certainty) appeared looser than that of normal subjects. Overall, this would be consistent with relative failure to encode internal facial parts by visual processes that depend on the left hemisphere, and greater reliance on right-hemisphere processes encoding global features instead (Tanaka and Farah, 1993). Since internal features are more important for identifying known faces (Ellis *et al*., 1979; Young *et al*., 1985*b*), erroneous familiarity might have arisen from resemblance to known faces on the basis of global or external traits, in contrast to the piecemeal visual errors of right brain-damaged patients (Rapcsak *et al*., 1994). However, deficient processing of internal facial parts with false familiarity based on global traits is not sufficient to explain J. R.'s disorder because she made no false recognitions of chimaeric faces that contained familiar external features with unknown internal parts, and she could easily recognize caricatures in which distinctive features were exaggerated and configurational cues were distorted (Rhodes *et al*., 1987). Therefore, whereas an abnormal bias towards basing recognition on global facial traits processed in the right hemisphere could expose J. R. to spurious familiarity signals, some additional factor must be necessary for such signals to be accepted subjectively by the patient as compelling feelings of personally knowing a face.

Note that, in all of the above tests assessing the recognition of altered faces or features, judgements of familiarity required a decision as to whether a facial stimulus had already been seen or not, and in such tests J. R. showed excellent discrimination and memory abilities. However, her abnormal feeling of familiarity with strangers in real life involved more than a false judgement of past occurrence: she had a compelling sentiment of *knowing* a person, with a strong feeling of personal closeness and intimacy. The subjective recollective experience triggered by seeing an individual face is inherently dependent on the activation of a complex network of associations between the visual representation of that face and other stored knowledge, such as specific biographical details, relevant episodes, particular contextual situations, and affective links (Rhodes, 1985; Damasio *et al*., 1990). A disorder within this network in our patient might have altered the pattern of associations evoked by unknown faces and distorted her subjective experience of familiarity.

In line with this, two other significant deficits in J. R. were observed in tests that required a face to be associated with distinctive information in memory, unique to the identity of a known or previously seen person. Thus, she showed a selective loss of identity-specific facilitation in the name–face priming task (despite intact visual priming in the face–face task), and a highly pathological rate of false fame attributions to unknown faces after incidental prior exposure. Both deficits suggest a problem in the associative processes involved in the recognition of a person (Burton *et al*., 1990; Young *et al*., 1994), at the level where specific knowledge pertaining to identity (e.g. a proper name in the semantic priming task, or a unique contextual trace in the false fame task) must be accessed from visual facial cues. In fact, the most striking deficit of J. R. in experimental tests of familiarity was observed in the false fame task, in which she had to judge not whether a face had been seen before, but whether it was known by fame. In this task, she mistook as famous ~80% of the faces that were basically not known but had been seen just once before in earlier experiments. This false feeling of having personal knowledge about an unknown face was elicited almost three times more often in J. R. than in normal subjects. Such a deficit was not found with names, demonstrating that it was specific to faces. Since other tests revealed that J. R. was unimpaired in judging whether a face had already been presented (e.g. in memory tasks), it must be concluded that her problem arose at the stage where a personal association is evoked by seen faces, leading to a subjective meaning, such as fame or personal acquaintance (Damasio *et al*., 1990). This pathological rate of false fame judgements in J. R. suggests an exaggerated bias towards attributing personal meaning to unknown faces in the presence of only minimal signals of familiarity (such as those induced by incidental prior exposure in this task). This misattribution of familiarity was specific to faces and unlikely to reflect more general problems of memory source attribution, confabulation or dysexecutive impairment, given her good performance in other general neuropsychological tests. Semantic memory also appeared intact, including knowledge about people (e.g. the Dead–Alive test) and naming, although more formal testing of semantic memory was not conducted.

In normal subjects, a misattribution of subjective familiarity can also be induced by spurious signals based on unconscious inferences and expectations, when more precise information about the source of familiarity cannot be accessed (Bartlett *et al*., 1991; Whittlesea and Williams, 1998; Yonelinas *et al*., 1999). Impaired access to person-specific associations was evidenced in J. R. by her lack of identity priming in the name–face task, indicating that she judged face familiarity without accessing semantic representations pertaining to identity-specific knowledge (such as names), unlike normal subjects (Young *et al*., 1994). The fact that she was much more prone to falsely recognizing fame in unknown faces, even without accessing more precise information in memory, indicates that unconscious signals of familiarity induced by the non-remembered prior exposure with these faces was sufficient to produce a much stronger feeling of knowing these faces in her than in normal subjects (Whittlesea and Williams, 1998). These findings therefore suggest not only decreased activation of specific semantic associations about seen faces, but also a greater tendency to activate associations of personal and affective meaning (Van Lancker, 1991). This would be compatible with an imbalance of face recognition mechanisms caused by impaired processing in the left hemisphere and the release of right hemisphere processes, in the context of interactive callosal inhibition between hemispheric functions (Regard *et al*., 1994; Cook *et al*., 1995). Broad and unconsciously generated associations about people might be more readily activated in the right hemisphere, even by occasional unknown faces, and underlie a subjective feeling of personal relevance and affective relationship (Van Lancker, 1991; Seeck *et al*., 1993), while the concomitant activation of more precise associations in the left hemisphere would be necessary in order to focus recognition on specific semantic knowledge related to a single individual (Rhodes, 1985; Damasio *et al*., 1990; Verstichel, 2001).

More generally, loss of such left-hemisphere semantic components in associative networks might contribute to the severe recognition deficit seen in prosopagnosics with bilateral brain lesions (Damasio *et al*., 1982), while right-hemisphere associations might be especially critical for the subjective feelings of familiarity that seem to be lost in prosopagnosics with unilateral right lesions, even in the presence of implicit recognition (Landis *et al*., 1986; Tiberghien and Clerc, 1986). Like false familiarity induced in normal subjects by unconscious associations based on the situational context (Thompson *et al*., 1982; Debruille *et al*., 1996; Sinha and Poggio, 1996), many errors made by J. R. in the chronic stage occurred in familiar places (e.g. university) in which many seen faces had no identity-specific representation other than a link with this familiar context (Davies, 1988). In such conditions, contextual cues can influence the perceived familiarity of faces (Young *et al*., 1985*a*; Davies, 1988). Greater reliance on right-hemisphere associative networks in J. R. might therefore facilitate the misattribution of personal meaning to unknown faces (as shown experimentally in the false fame task), especially in the presence of spurious signals of familiarity evoked by non-specific cues (Thompson *et al*., 1982; Tiberghien, 1986; Bartlett *et al*., 1991; Whittlesea and Williams, 1998).

An imbalance between reciprocal hemispheric functions to account for J. R.'s disorder converges with recent findings from functional neuroimaging in healthy humans showing that face processing implicates a bilaterally distributed network beyond the visual areas in the fusiform cortex (Haxby *et al*., 2002). In this network, the left lateral temporal cortex may store unique semantic representations associated with identity (Rhodes, 1985; Gorno Tempini *et al*., 1998; Leveroni *et al*., 2000), whereas homologous right temporal areas may constitute a system evaluating social and affective meaning (Landis *et al*., 1990; Allison *et al*., 2000; Mendez and Ghajarnia, 2001; Winston *et al*., 2002), including the appraisal of personal familiarity (Ellis *et al*., 1989; Leveroni *et al*., 2000; Nakamura *et al*., 2000; Shah *et al*., 2001). Our normal subjective experience of knowing a seen face may depend critically on coordinated activity within this bilateral network, linking the visual representation of faces and other contextual cues to stored associations pertaining to person identity and affective relevance (Damasio *et al*., 1990; Van Lancker, 1991). While face recognition disorders are rare after left brain damage (Warrington and James, 1967), J. R. had a very unusual lesion due to cortical venous thrombosis, centred on the posterior middle temporal gyrus. Such venous lesions may affect different brain regions and cause only partial cortical damage rather than complete destruction of brain tissue, as do arterial infarcts. Moreover, episodes of false familiarity may be missed if the patient does not complain spontaneously or has larger lesions, resulting in aphasic and naming disorders. We note that Damasio and colleagues briefly described a disorder they termed 'deep prosopagnosia' in two patients who had left temporo-occipital damage and mistook famous faces for other, semantically related faces (Damasio *et al*., 1988), which is consistent with a role of the left temporal cortex in retrieving information about known people. However, the contribution of perceptual or naming difficulties may be difficult to establish in such cases because of the larger extent of cortical and subcortical damage caused by arterial occlusion.

In summary, this case extends other reports suggesting that abnormal face familiarity can arise from deficits at several stages of processing (Young *et al*., 1993; Rapcsak *et al*., 1999; Ward *et al*., 1999). This implicates the activation of associative networks integrating facial cues with more complex representations of personal relevance (Burton *et al*., 1990; Damasio *et al*., 1990; Schreiber *et al*., 1991; Hanley *et al*., 1998) rather than only the abnormal activation of the visual representation of known faces, as proposed by earlier models (Bruce and Young, 1986). To our knowledge, this case constitutes a new neuropsychological disorder that demonstrates that both hemispheres have a significant role in face processing.

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