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Associative (prosop)agnosia without (apparent) perceptual deficits: A case-study

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Abstract

In associative agnosia early perceptual processing of faces or objects are considered to be intact, while the ability to access stored semantic information about the individual face or object is impaired. Recent claims, however, have asserted that associative agnosia is also characterized by deficits at the perceptual level, which are too subtle to be detected by current neuropsychological tests. Thus, the impaired identification of famous faces or common objects in associative agnosia stems from difficulties in extracting the minute perceptual details required to identify a face or an object. In the present study, we report the case of a patient DBO with a left occipital infarct, who shows impaired object and famous face recognition. Despite his disability, he exhibits a face inversion effect, and is able to select a famous face from among non-famous distractors. In addition, his performance is normal in an immediate and delayed recognition memory for faces, whose external features were deleted. His deficits in face recognition are apparent only when he is required to name a famous face, or select two faces from among a triad of famous figures based on their semantic relationships (a task which does not require access to names). The nature of his deficits in object perception and recognition are similar to his impairments in the face domain. This pattern of behavior supports the notion that apperceptive and associative agnosia reflect distinct and dissociated deficits, which result from damage to different stages of the face and object recognition process.

Keywords: Face recognition; Object recognition; Apperceptive/associative; Visual agnosia; Prosopagnosia

1. Introduction

Prosopagnosia is a neurologicical deficit which is characterized by a severely reduced ability to recognize faces (Bodamer, 1947). This deficit cannot be attributed to a general loss of semantic memory or knowledge, as prosopagnosic patients can identify familiar people from their voice, gait, or salient facial features, such as a mustache. Moreover, they are able to supply ample biographical information when provided with a name, or conversely, name a person based on his or her verbal description. Thus, the face-recognition impairment associated with prosopagnosia is limited to the visual modality.

Following the classical distinction proposed by Lissauer (1890) between types of impairments in visual object recognition, the different manifestations of prosopagnosia are also traditionally classified into two broad subclasses: *apperceptive prosopagnosia* involves deficits during early (pre-categorical) stages of visual processing, prior to the formation of a facial representation. In a*ssociative prosopagnosia*, however, the patient has great difficulty in accessing semantic information of a facial percept which he or she was able to construct. One conclusion arising from this typology is that although both forms of prosopagnosic patients will be impaired in recognizing famous faces, only those with the apperceptive form of prosopagnosia will encounter difficulties in recognizing non-famous faces.

Although this dissociation has been reported in several studies (e.g., De Renzi & di Pellegrino, 1998; De Renzi, Faglioni,

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Grossi, & Nichelli, 1991; Henke, Schweinberger, Grigo, Klos, & Sommer, 1998; McNeil & Warrington, 1991; Temple, 1992), and has been articulated theoretically in several models of face recognition (Bruce & Young, 1986; Burton, Bruce, & Johnston, 1990; Gobbini & Haxby, 2007; Haxby, Hoffman, & Gobbini, 2000; Hay, Young, & Ellis, 1991), recent reports have questioned its validity (e.g., Delvenne, Seron, Coyette, & Rossion, 2004; Duchaine & Weidenfeld, 2003; Farah, 1990). For example, Farah (1990) concluded, after reviewing a large corpus of associative agnosia and prosopagnosia cases, that none of them shows clear evidence of intact early visual analysis. More specific claims have undermined the validity of the neuropsychological assessment tools which commonly have been used to determine that high-order visual processes are preserved in associative prosopagnosia (Duchaine & Nakayama, 2004; Duchaine & Weidenfeld, 2003). Finally, while some traditional tests of face recognition may not have been sensitive enough to detect perceptual deficits in people with associative prosopagnosia, others have been (Delvenne et al., 2004; Farah, 1990). Findings based on these more sensitive tests lead to the conclusion that the underlying deficit in associative agnosia, for both faces and objects, is at the perceptual level, and that the dissociation between apperceptive and associative types of the disorder is artifactual (Bay, 1953; Farah, 1990).

Such a conclusion would undermine models of face and object recognition that honor this distinction. A more conservative (and maybe more warranted) approach, however, leaves open the possibility that associative (prosop)agnosia does exist, while acknowledging that the past literature may have overestimated its frequency of occurrence. Finding such a case, therefore, has important implications for theories and models of face and object perception and recognition.

In the present study, we describe a new case of acquired associative (prosop)agnosia in patient DBO, a 72-year-old male, who presented with deficits in visual object and face recognition. His object recognition in the tactile and auditory modalities is normal, and he does not seem to have any low-level visual deficits. Although he cannot identify pictures of famous figures, he is able to supply biographical information about them when presented with their names. Using a combination of traditional tests, and new ones we devised to address issues raised by critics regarding higher order face-processing deficits, we believe we can show that DBO is indeed a case which exemplifies a (prosop)agnosia of the associative type.

2. Case history

DBO is a 72-year-old right-handed male who was born in Latvia and arrived in Canada at an early age. He earned a Ph.D. degree in Chemistry and specialized as a criminologist. He was admitted to hospital on March 9, 2003, following a sudden onset of confusion and tachycardia. A CT scan showed a left occipital lobe infarct extending from the cortex into the periventricular white matter, with some parietal involvement. Areas with periventricular white matter hypodensity were observed bilaterally, some compatible with lacunar infarcts. He was diagnosed as having suffered multiple strokes secondary to emboli, related to atrial fibrillation (Fig. 1).

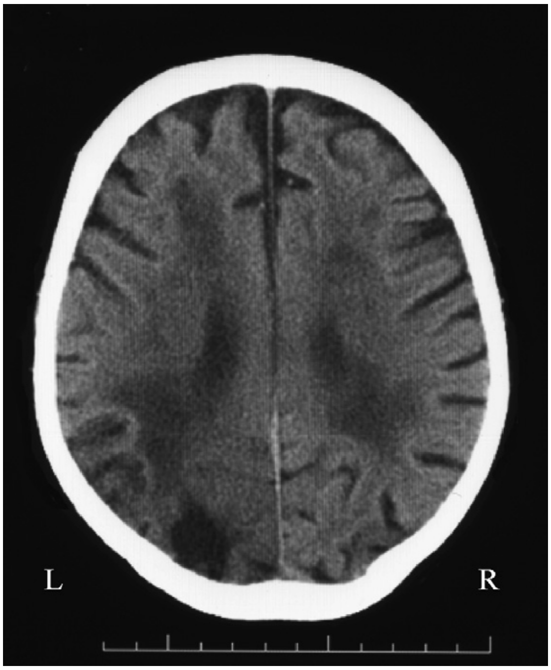

Fig. 1. A CT transversal slice showing the extent of lesion in DBO's occipital area.

Following his stroke he had memory impairments, word-finding difficulties, impaired language comprehension, impaired object, letter, word, and face recognition and a right homonymous hemianopia. There were also mild hand tremors, noted especially when he attempted to perform purposeful fine motor tasks.

He was admitted for neuro-rehabilitation at Baycrest Centre for Geriatric Care on June 2003. His full scale intellectual score in the Wechsler Abbreviated Scale of Intelligence (Wechsler, 1999) was in the average range (55th percentile). He yielded high average scores in verbal IQ subtest (84th percentile), but low average scores at performance IQ subtest (23rd percentile). His performance in the Kaplan Baycrest Neurocognitive Assessment (KBNA; Leach, Kaplan, Rewilak, Richards, & Proulx, 2000) and Dementia Rating Scale-2 (DRS-2; Jurica, Leitten, & Mattis, 2001) showed deficits in several cognitive abilities which may be attributed to his global visual agnosia. He encountered difficulties in short- and long-term verbal and visual recall, yet exhibited improved recognition capacities, verbal and visual alike. His verbal fluency and practical reasoning were average, although impairments were found in conceptual shifting (assessed also by the Wisconsin Card Sorting Task; Kongs, Thompson, Iverson, & Heaton, 2000). Problems in concentration and selective attention were also observed. DBO exhibited throughout the assessment considerable difficulties in letter and word reading, and was greatly impaired in identifying complex form and visual objects, attesting to his alexia and object agnosia. We describe his deficits in object and face recognition in more detail below. In all the tests reported henceforth (carried during July–August 2003) DBO's performance was compared to that of five healthy controls matched in age, education, and sex.

3. Object processing

DBO was severely limited in visual object recognition as illustrated by a simple example: while in the dining room he was asked to take a cup and fill it with water from the sink. Although he was able to find a cup, he fumbled through several objects, such as microwave, water pitcher, garbage can, and roll of paper towels, while saying repeatedly "This is a sink . . . Oh! This one could be a sink . . .. This is also a sink", before he finally identified the sink and obtained water. A few minutes later, when asked to pour the water from the cup into the sink, the laborious procedure was repeated.

3.1. Object identification in vision and other modalities

DBO's visual recognition of three-dimensional objects was severely impaired. He named only 1 of 20 real, common objects presented to him for an unlimited exposure. A similar impairment was seen with line drawings, as he was able to name only eight pictures in the Boston Naming Test (BNT; Kaplan, Goodglass, & Weintraub, 1983; controls 58.4, S.D. = 1.52, $Z = -33.16$). His errors were classified as unrelated (30%), perseverative (27%), semantic (23%), or omissions (20%). He was then administered the responsive naming form of the BNT, where participants are required to give a name in response to a definition (e.g., "What musical instrument do angels play?" response: Harp). DBO was able to provide correctly 52 names (controls 54.8, S.D. = 3.11, $Z = -.90$). His ability to identify the function of the objects by gesturing was impaired, scoring only 1/14 for real objects and 6/14 for drawings. The control subjects, in contrast, scored perfectly. No impairments were observed when the object was named to him and he was asked to demonstrate by gesture how the object is used.

His object recognition difficulties were apparent only when visual modality was required. He had no problem identifying objects by palpation (100% accuracy, $n = 20$). In addition, when presented with auditory sounds of animate (e.g., rooster) and inanimate (e.g., train) objects, he performed normally, naming 12 animate objects ($n = 17$, controls 11.80, S.D. = 2.59, $Z = .08$), and 14 inanimate objects ($n = 17$, controls 12.8, S.D. = 2.49, $Z = .48$).

3.2. Object perception and recognition: performance on the Birmingham Object Recognition Battery (BORB)

Table 1
DBO and control group performance in subtests of the BORB

BORB subtests	DBO	Controls ($n = 5$)	
	Mean	Mean	S.D.
Length match task (test 2; $n = 30$)	26	25.80	2.28
Size match task (test 3; $n = 30$)	30	26.80	1.48
Orientation match task (test 4; $n = 30$)	24	26.40	2.19
Position of gap match task (test 5; $n = 40$)	34	35.80	1.30
Minimal feature view task (test 7; $n = 25$)	23	24.80	.45
Foreshortened match task (test 8; $n = 25$)	22	24.60	.55
Object decision task A (test 10; $n = 32$)	31	30.40	1.14
Object decision task B (test 10; $n = 32$)	27	25.40	3.71
Item match task (test 11; $n = 32$)	27	31.80	.45
Association match task (test 12; $n = 30$)	6	29.4	.55
Picture naming (test 13; $n = 76$)	20	72.8	1.92

To pinpoint further his difficulties in object recognition, DBO was tested with several subtests of the Birmingham Object Recognition Battery (Riddoch & Humphreys, 1993). His visual perceptual pre-categorical capacities seemed intact. His performance was normal, sometimes exceeding the controls' average, at the *length match task* (test 2), *size match task* (test 3), *orientation match task* (test 4), and the *position of gap match task* (test 5). His performance at the *minimal feature view task* (test 7), and *foreshortened match task* (test 8; see Table 1) was somewhat low compared to the age- and education-matched controls in this study, but average in comparison to the control norms reported in the battery. These tests probe object constancy and require matching two objects which are depicted from different viewpoints. Although they are not regarded as tasks that involve access to stored knowledge, nonetheless, the identification of the object presented in the standard viewpoint may assist in matching its counterpart. The inability of DBO to use this semantic information may account for his minor decrement in performing these tasks at the same level as that of his matched controls.

The following tests in the BORB were designated to assess the ability to access semantic information based on visual information. An interesting dissociation was found in DBO in performing these tasks. He showed intact capacity at the *object recognition task* (test 10) in both the easy (A) and hard (B) versions. In contrast, he was severely impaired at the *item match task* (test 11). Although his performance at this task was not at floor level, he was still more than 8 S.D.s below the controls' average. In addition, it should be noted that many items could be matched in the *item match task* on the basis of visual similarity (G. Humphreys, personal communication, September 2003). Thus, we think it likely that DBO was relying to a great degree on his intact perceptual capacities in performing the task.

DBO encountered great difficulty in the *association match task* (test 12), in which an object target is to be matched to the most closely associated item. Interestingly, when we modified this task to a verbal version, in which the target and the two to-be-matched items were named, DBO scored 100%. A similar picture was revealed in a complementary and more comprehensive task (The Pyramids and Palm Trees Test; Howard & Patterson, 1992). While visual matching was greatly impaired (29/52, controls 51.2, S.D. = 1.10, $Z = -20.18$), verbal matching was preserved. Finally, poor performance was seen at the *picture naming task* (test 13), reflecting the findings seen previously in the BNT.

3.3. Visual imagery for objects and letters

Despite his impaired visual perception, DBO's visual imagery was intact (see Table 2). He had no difficulty in judging object size or color (Behrmann, Moscovitch, & Winocur,

Table 2
DBO and control group performance in visual imagery tests

Visual imagery tests	DBO	Controls ($n=5$)	
	Mean	Mean	S.D.
Imagery for letter shape A ($n=26$)	26	25.80	.45
Imagery for letter shape B ($n=26$)	24	24.20	2.68
Imagery for object color ($n=32$)	30	30.80	1.30
Imagery for object size ($n=5$)	5	4.80	.45
Imagery for object shape A ($n=20$)	18	17.4	.55
Imagery for object shape B ($n=20$)	18	17.33	.57
Verification of high- and low-imagery sentences ($n=32$)	31	30	1.00
Clock test ($n=24$)	12	22.2	2.68

1994). DBO also verified correctly 31 out of 32 low- and high-imagery sentences (Eddy & Glass, 1981). He also demonstrated intact knowledge of animals body parts (tails and ears; Farah, Hammond, Mehta, & Ratcliff, 1989). These tasks are especially sensitive measures of mental imagery as information about body parts is usually stored visually and not verbally. In spite of his alexia, he was able to judge the form of letters in two tasks. In the first task, he was asked to imagine whether lowercase letters had lines extending from their main body (Kosslyn, 1987). In the second task, he had to determine whether uppercase letters had curved parts. He performed both tasks easily.

The only imagery test in which DBO was impaired was the clock test (Craik & Dirkx, 1992; Paivio, 1978). In this task the participant is asked to imagine a clock face and then told different times of the day. He then has to decide whether the two hands on the clock face, when representing these times, are at an angle of greater or less than 90°. DBO's performance at this task was at chance level. The deficit may be related to the mental rotation or constructive component of the task which often is impaired in many neurological disorders (Freedman, Leach, Winocur, Shulman, & Kaplan, 1994).

3.4. Summary and discussion of DBO's object agnosia

DBO is impaired in identifying both real and line-drawn objects from vision. In contrast, his identification is normal when objects are presented tactually, or when he is supplied with their characteristic sounds, arguing against a general semantic deterioration, such as semantic dementia. The integrity of his semantic knowledge is also reflected in his ability to image mentally different characteristics of animate and inanimate objects and in his ability to name an object in response to a definition.

His poor performance in visual identification and recognition is also not dependent on response mode as he is poor at gesturing the use of objects, and in matching related items by pointing. This finding is not compatible with a modality-specific deficit of accessing names of objects, such as in optic aphasia, where gesturing to visually presented objects is intact (e.g., Coslett & Saffran, 1992; Lhermitte & Beauvois, 1973; Luzzatti, Rumiati, & Ghirardi, 1998). Even where pantomime ability is not demonstrated (e.g., Goldenberg & Karlbauer, 1998; Hillis & Caramazza, 1995), these patients perform well on other tasks that require preserved semantic access, such as category sorting. Moreover, optic aphasia patients do not encounter difficulties in interacting with the visual world in everyday life and they manage their daily chores with ease (see Luzzatti et al., 1998). DBO, in contrast, is greatly handicapped in his daily life functions. One illustrative example was his disability, during his hospitalization period, to find his clothes and other personal belongings. This trivial deficit was the source for several emotional bouts of agitation and unrest, in the course of which DBO complained that his personal possessions were stolen from him. The emotional distress incurred by the impairment in identifying visual objects is hardly seen in optic aphasia patients. Thus, the pattern of performance displayed by DBO is characteristic of visual agnosia.

Probing the locus of DBO's impairment revealed that his basic perceptual processes are probably unimpaired. His performance equaled that of controls in tasks that required length, size, orientation, and location matching, attesting to his preserved ability to encode and manipulate basic dimensions of visual stimuli. Moreover, he was able to recognize identical items presented across different viewpoints, indicating that a viewpoint-independent representation was formed. Although, he was somewhat impaired in these latter tasks compared to his controls, his performance was at 90% accuracy, much higher than the batteries' norms. Thus, it is reasonable to conjecture that DBO could not benefit from the identification of the object in the standard view, in contrast to his controls. Following Marr's terminology (1982), DBO was able to extract both the 2.5-D sketch and the 3-D modal representation, and as such he cannot be considered to be a classic case of apperceptive agnosia.

Neuropsychological evidence suggests that access to semantic information is hierarchical and can be fractionated into several modular subsystems (e.g., Hillis & Caramazza, 1995; Riddoch & Humphreys, 1987, 2001). Structural encoding involves the mapping of the object's parts and the spatial relationship among them. A successful matching between the visual input and its stored structural representation would lead to a feeling of familiarity. At a higher level of processing, the unique functional and associative information about a particular object, which distinguishes it from other objects, will be extracted.

DBO's ability to perform the object decision task normally, while failing to execute any task which required access to semantic information about the object, indicates that his impairment can lie at three possible loci: access to any semantic information about objects through the visual modality, access to fine and detailed semantic information (Dixon, Bub, & Arguin, 1997), or a degraded semantic store. Although our results do not allow us to endorse fully any of these three alternatives, the first one receives most support. DBO was able to provide detailed information about objects and name them in other modalities. His visual imagery was also intact. These results argue against a degradation of semantic knowledge. His errors in the various tasks, which were mostly unrelated or perseverative, do not support the claim that he was able to access the related general domain but was unable to pinpoint the specific details. Thus, the evidence favors the claim that the source of DBO's difficulties lies in his inability to access a semantic store through the visual modality. Thus, his disorder qualifies as associative visual agnosia.

Several reports have appeared in the literature which describe similar patients who are unable to name objects but, nonetheless, can access semantic information from other modalities (DJ, Fery & Morais, 2003; DHY, Hillis & Caramazza, 1995; AB, Luzzatti et al., 1998; GV, Miozzo & Caramazza, 1998; JB, Riddoch & Humphreys, 1987; MD, Jankowiak, Kinsbourne, Shalev, & Bachman, 1992; RC, Carlesimo, Casadio, Sabbadini, & Caltagirone, 1998). Likewise, they are characterized by intact perceptual abilities and their deficits seem to be localized to more advanced stages of object processing. Thus, for example, DJ, GV, DHY, MD, and RC were able to demonstrate unimpaired visual–perceptual abilities equivalent to those of controls and preserved access to stored structural description. Difficulties emerged in these patients, similar to DBO's, when access to semantic information was required. Yet, the semantic access deficits exhibited by DBO are apparently more drastic than those shown by other patients. For some of these reported patients (GV and DHY), more detailed and refined tasks were required to uncover the impairments in accessing semantic information through the visual modality. Moreover, the pattern of the errors in naming and in identifying line drawings suggest that partial semantic access was available for those patients as the majority of their errors was semantic in nature (GV—82%, DHI—75%, DJ—65%, MD—87%). The errors generated by DBO, on the other hand, were perseverative, unrelated, or omissions. Only a minority (23%) of his errors could fit the category of semantic errors, consistent with his diagnosis of associative visual agnosia. In this regard DBO is similar to RC (Carlesimo et al., 1998) whose perceptual abilities were intact as was his object/non-object discrimination. RC was severely impaired in object identification with errors mainly of the perseverative type. We now turn to his face-recognition capacities to determine whether this condition also extends to faces.

4. Face recognition and identification

DBO was severely impaired in identifying famous people from photographs. In a preliminary test he could not identify any of the pictures of famous people presented to him ($n = 20$, controls = 15.8, S.D. = 4.14, $Z = -3.81$). In contrast, DBO was able to provide detailed biographic information for 18 people when given their names (controls 18.4, S.D. = 1.82, $Z = -.22$). A series of tests, traditional and newly devised, were administered to DBO in order to assess the locus of his face-processing deficits, and to consider whether they stem from perceptual or semantic locus of impairment.

4.1. Unfamiliar face matching

The Benton Facial Recognition Test (BFRT; Benton, Sivan, Hamsher, Varney, & Spreen, 1983) was used to assess DBO's capacities to match unknown faces. In this test participants are required to match a frontal view photograph with three other photographs of the same face taken from different angles or under different lighting conditions. DBO's total corrected score was 42, classifying his performance as normal, although at the lower range (41–54). More importantly, his score was not significantly different from that of his matched controls (47.60, S.D. = 3.86, $Z = -1.46$).

Although DBO was able to perform well in the BFRT, these results should be interpreted with caution. Since the target face and the faces to be matched were presented simultaneously, DBO may have used a strategy in which he matched individual features rather than the entire face as a whole (Duchaine & Weidenfeld, 2003). Our impression was that DBO performed the task with ease equal to that of controls. His performance could not be described as laborious and slavish as apparent in reports of patients who depend on feature-to-feature matching strategy. Yet, the possibility that DBO used a strategy which does not require holistic processing cannot be ruled out since his responses were not time. Thus, we supplemented our assessment of DBOs' perceptual facial abilities with additional tests in which matching is not possible.

4.2. Memory for unfamiliar faces

DBO's ability to recognize newly learned faces was assessed using the Warrington Recognition Memory for Faces Test (WRMT; Warrington, 1984). DBO scored 40/50 which was not different from the published norms of his age-matched group, and from the scores of the controls in our study (41.6, S.D. = 4.16, $Z = -.38$).

Although the WRMT is a widely used test to assess potential deficits in face perception, some criticism was raised lately claiming that it may not tap unique face-recognition processes which are dependent on the processing of internal features (Duchaine & Weidenfeld, 2003). It was noted that the photos in this test contain rich external features (such as hairstyle) and other contextual information (such as body positions, paraphernalia) which may assist participants in differentiating between target and distractor photos. We believe that this possibility is improbable in the case of DBO, due to his visual object agnosia, which prevents him from identifying specific external features (e.g., tie) and using them as a cue. However, in order to examine this option more closely, we devised a modified memory test for faces which consisted only of internal features of unfamiliar faces.

4.2.1. Memory for unfamiliar faces without external features

The modified test consists of 40 faces (20 males, 20 females, 5.6 cm × 5.6 cm; Fig. 2), 20 of which were randomly designated as target faces and 20 as distractors. Hair and contour were deleted from the photos using the Adobe Photoshop 6 software package.

4.2.1.1. Procedure. Stimuli were displayed on a Dell color monitor controlled by E-Prime software (Psychological Software Tools, Inc., 2000) implemented in a Dell PC compatible computer. DBO was required to study 20 faces (half male, half female), which he was asked to recognize later. Each face appeared on the screen for 10 s. Immediately following the study

Fig. 2. Examples of unfamiliar faces used in the memory for unfamiliar test without external features (4.1.2).

phase, a forced-choice recognition test was administered. As in the WRMT the target face was presented with a distractor of the same sex and age, and DBO was required to select the face which appeared recently. One hour after the immediate recognition test, a delayed recognition test, similar to the first, was administered. DBO was not informed in advance of the delayed recognition test.

4.2.1.2. Results and discussion. In the immediate recognition test, DBO was correct in recognizing 17/20 faces as old, and was not different from controls (16.4, S.D. = 2.61, $Z = .23$). His median response time (RT) for the accurate trials were also within normal range (4443 ms, controls 3753 ms, S.D. = 902 ms, $Z = -.76$). In the delayed recognition test, DBO was again highly accurate and identified 15/20 faces correctly (controls 14.6, S.D. = 2.19, $Z = .19$). His RT performance was also within the normal range (5302 ms, controls 4025 ms, S.D. = 932 ms, $Z = -1.37$).

The results DBO obtained in the modified recognition test show that his facial perception capacities are intact, and converge with the results found both in the WRMT and BFRT. In contrast to objections that can be directed at the former tests, whose administration format enable either feature matching or reliance on external features, the latter test does not suffer from these weaknesses. Interestingly, DBO's accuracy was slightly better than that of controls in the modified version of the WRMT than in the original test. This may result from the ability of the healthy participants to utilize external non-facial cues in the WRMT, but not in the modified version of the test. DBO, however, due to his visual agnosia, was at a disadvantage, which resulted in slightly lower scores in the WRMT.

4.3. Face inversion and misalignment effects

The inversion effect, namely, the difficulty in recognizing an inverted face compared to an upright face, is considered to be a hallmark of holistic processing (Valentine, 1988; Yin, 1969). This effect was traditionally interpreted as indicating the existence of a face-specific processor which specializes in processing faces holistically with little part decomposition, and whose function is disrupted by inversion (e.g., Maurer, Le Grand, & Mondloch, 2002; Moscovitch, Winocour, & Behrmann, 1997). Neuropsychological studies have found, accordingly, that the majority of prosopagnosic patients show a reduced inversion effect (Boutsen & Humphreys, 2002; Delvenne et al., 2004; Marotta, McKeeff, & Behrmann, 2002), with some even exhibiting an opposite trend, and performing better at matching inverted than upright faces (*inversion superiority effect*; de Gelder, Bachoud-Levi, & Degos, 1998; de Gelder & Rouw, 2000; Farah, Wilson, Drain, & Tanaka, 1995). In light of DBO's normal ability to perform the previous facial tasks, we sought to investigate whether he would demonstrate a different inversion effect than normal. A smaller inversion effect or an inversion superiority effect will indicate that the locus of DBO face-recognition problems lies at the perceptual level(s). In contrast, normal inversion effect will be in accord with the previous tests showing that his difficulties arise at a later, semantic processing stage.

In addition, we also examined DBO's ability to recognize misaligned faces. Inversion may disrupt the processing of the features themselves (e.g., Moscovitch & Moscovitch, 2000). Misalignment effects, however, are known to affect the configural aspects of a face, but not its facial features (Gauthier, Williams, Tarr, & Tanaka, 1998; Moscovitch et al., 1997).

4.3.1. Method

4.3.1.1. Stimuli. The critical stimuli consisted of 80 pictures of Caucasian faces (half male, half female) taken from the Max Planck Institute for Biological Cybernetics (Tuebingen, Germany). The faces were in frontal-view position, with a neutral expression and without makeup, accessories or facial hair. The original color pictures were converted to a 256 gray-level format (74 dpi) and extended 8.79 cm × 8.79 cm.

4.3.1.2. Procedure. DBO was seated approximately 50 cm from a computer screen (see previous task for apparatus details). Each trial began with a 1000 ms centrally presented fixation mark (+). Following its offset an upright whole face appeared for 400 ms followed by a black screen interval for 250 ms. To eliminate effects of afterimages or other types of visual persistence, a mask appeared for 500 ms occupying the area in which the faces were presented. The mask was created using minute pieces of facial features taken from different faces (Fig. 3). Following the mask, a display of three faces was presented, consisting of the previously presented face as target and two other faces as distractors (selected equally often from among the study's critical stimuli). The faces in the display were presented randomly in either an upright, inverted or misaligned orientation. The misaligned faces were constructed by dividing each face into two parts by slicing it under the eyes. The nose in the bottom segment was positioned under the left ear of the upper segment.

DBO was asked to select the face in the display that was identical to the target face and to respond by pressing one of three keys corresponding to left, middle, and right faces on the display. The experiment consisted of 240 trials. Each of the 80 critical stimuli was presented once in each of the orientation conditions of the recognition array (upright, inverted, misaligned).

A set of 15 practice trials was administered prior to the commencement of the experiment. The results of these trials were not included in the analysis.

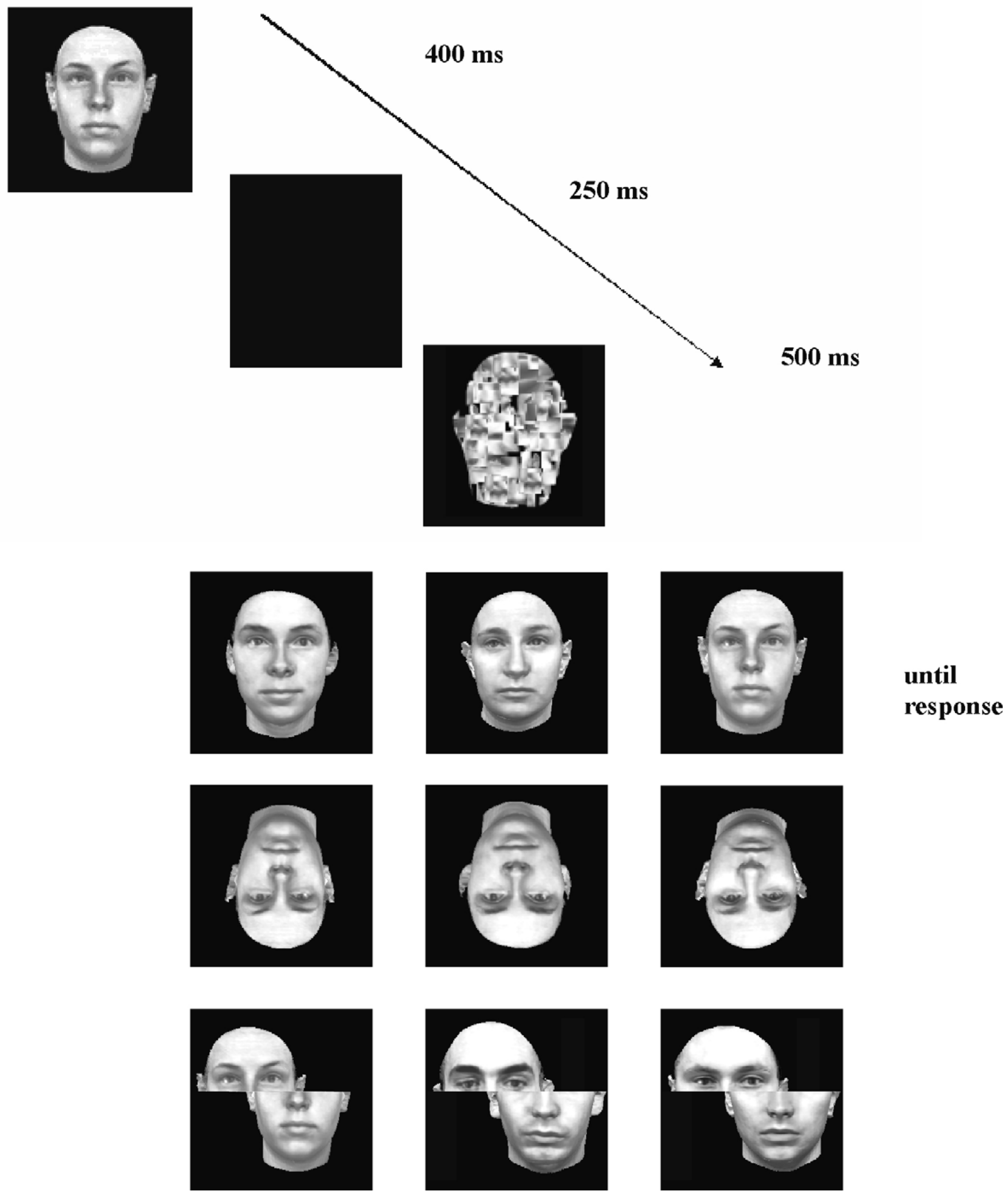

Fig. 3. Sequence of events in a typical trial in the face inversion and misalignment effects task (4.2).

4.3.2. Results and discussion

DBO's overall accuracy was .62 and he was impaired relative to controls (.81, S.D. = .07, $Z = -2.69$). However, he correctly identified .75 of the upright presented targets, .60 of the misaligned faces, and .50 of the inverted faces, revealing significant inversion (25%, $\chi^2(1) = 10.66$, $p < .005$; controls 17%, S.D. = 14) and misalignment effects (15%, $\chi^2(1) = 4.1$, $p < .05$; controls 10%, S.D. = 9; Fig. 4). His lower than average overall accuracy may have been caused by his hemianopia, or by the typical drop in performance associated with brain damage of any sort on difficult tests with rapidly presented stimuli.

Analysis of DBO's correct RTs revealed that he was not significantly slower than controls overall (3665 ms, controls 2842 ms, S.D. = 660 ms, $Z = 1.24$). More specifically, his RTs to upright, inverted, and misaligned faces did not differ significantly from those of controls. Finally, the inversion effect of DBO was comparable to that of controls, though his misalignment effect was greater than that of controls (Fig. 4).

In sum, although DBO's overall performance is less accurate (but not necessarily slower) than that of his matched controls, he shows patterns of normal perceptual facial processes in both indices of accuracy and RT, suggesting that he is processing upright faces in a configural fashion which is disrupted when the face is misaligned or inverted. In this, DBO differs from other prosopagnosic patients, who either show reduced or enhanced performance for inverted over upright faces. We know only of a single study (Gauthier, Behrmann, & Tarr, 1999) which demonstrated normal inversion effects in apperceptive (prosop)agnosia patients (BFRT score 36/54), but there are a number of peculiarities in that study which suggests that the findings should be interpreted cautiously. Close inspection of the performance of the participants in the inverted faces task shows that both patients performed far worse than controls, with one patient performing at or below chance on many conditions. Even the patient who performed above chance on upright faces is two to four times slower than controls, which may mean that even upright face recognition is piecemeal, though somewhat easier for the patient than recognizing inverted faces. For these reasons, it is difficult to know whether their inversion effect arises from impaired configural processing, from deficits in discrimination, or from both.

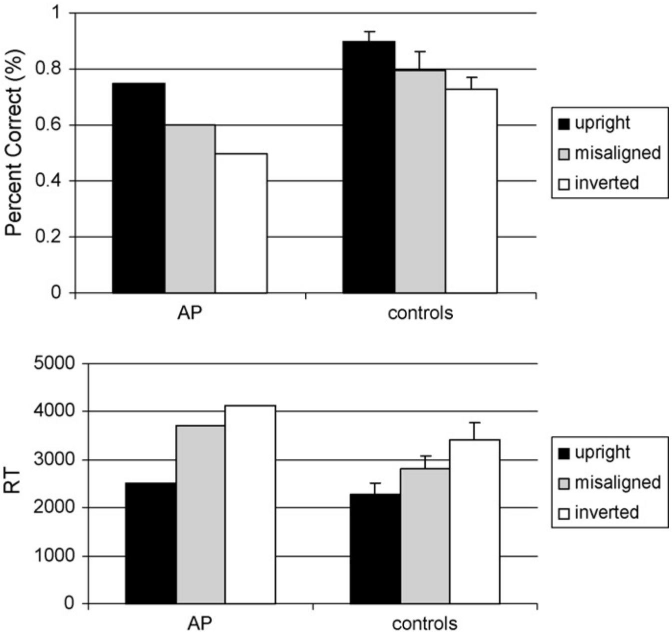

Fig. 4. Mean accuracy and RT (SE in parentheses) of DBO in matching upright, misaligned, and inverted faces (4.2).

4.4. Familiarity task

The previous experiments indicate that DBO's deficits cannot be easily attributed to deficits in basic perceptual stages of face recognition which appears to be intact. The next set of tests was administered to examine more advanced levels of face recognition. The first was a test of familiarity designed to explore whether famous faces could be distinguished from non-famous faces even if DBO cannot name them or supply any biographical information about them.

4.4.1. Method

4.4.1.1. Stimuli. The critical stimuli consisted of 24 quadruplets. Each quadruplet included one target picture of a famous face and three non-famous distractors matched as closely as possible to the famous face in age, gender, and general external appearance (Fig. 5). Pictures were converted to a 256 gray-level format (74 dpi) and extended 8 cm × 11 cm and did not contain any external cues of the identity of the figure.

4.4.1.2. Procedure. In each trial the four faces were presented together horizontally in the center of a computer screen. The famous face appeared equally, across trials, in all of the four locations. DBO was asked to point to the picture which he had seen before and was familiar to him. If he pointed to the famous picture, he was asked to name the face or supply any identifying information.

Following the presentation of all 24 quadruplets, the 24 famous faces were presented to DBO alone, without the distractors, and he was asked to name the face or supply identifying information. The aim of this test was to see whether DBO's performance was enhanced when visual distractors were removed and only the famous face was presented. Finally, in the last section of the task, the experimenter read the 24 names of the famous faces and asked DBO to supply semantic information about them.

4.4.2. Results and discussion

DBO was able to identify 19/24 famous faces as familiar, a score which cannot be attributed to chance performance ($\chi^2(1) = 8.17$, $p < .01$), and was also not different from the controls' score (controls 21.6, S.D. = 2.07, $Z = -1.25$). Yet, despite his ability to select the familiar face from the distractors, he was able to supply identifying information only to 9 of the 19 faces that were familiar to him (controls 21.6, S.D. = 2.07, $Z = -6.08$), including two faces which he was able to name (*John F. Kennedy* and *Groucho Marx*; controls 21, S.D. = 1.73, $Z = -10.96$).

Fig. 5. Examples of stimuli material used in the familiarity task (4.3).

The naming section of the famous faces of the familiarity task did not show any improvement. The only faces which he was able to name were the two previous ones and he was able to give some information to the same nine faces.

In the last section, DBO was able to supply semantic information for 22/24 names when they were read to him (he did not know *Mel Gibson*, an actor, and *Joe Clark*, a well-known Canadian politician). His ability to supply semantic information in response to a name cue was greater than to a face cue ($\chi^2(1) = 15.39$, $p < .001$).

It is evident from the results of the familiarity task that DBO has a sense of familiarity when encountering a famous face. He is unable, though, to derive more identifying information regarding the figure he recognizes as familiar. From a theoretical perspective, DBO is able to match an intact perceptual representation with an existing structural representation in memory. He is limited, however, in accessing from the visual modality specific semantic knowledge that is associated with a particular person. This deficit disappears when the names are read to DBO as he is able to supply enough information that differentiates one person from the others.

One possible objection to this interpretation of the current findings may be that DBO's high performance in the familiarity task does not stem from recognizing the faces but rather from some external attributes that facilitates identifying a figure as famous. For example, the quality of the famous figures' photographs may have cued DBO to select the picture as familiar. We think that this possibility is unlikely since we attempted to match the target face and the distractor as closely as possible. In addition, we have recently collected in our lab data from a congenital prosopagnosic person (Anaki, Itier, O'Craven, & Moscovitch, in preparation) whose performance in this task was at chance level. If the target face did differ from the distractors on any dimension, this person should have also been able to detect it, but that was not the case.

4.5. Recognizing caricatures of famous people

Caricatures are formed by exaggerating the distinctive features of a face to produce a grotesque or a comic effect. A well-known finding in the face-recognition literature is that distinctive faces are recognized better than faces judged to be less distinct (Bartlett, Hurry, & Thorley, 1984; Benson & Perrett, 1994; Going & Read, 1974; Valentine & Bruce, 1986a, 1986b). It is, therefore, of no surprise that caricatures are recognized as accurately as veridical depictions (Calder, Young, Benson, & Perrett, 1996; Chang, Levine, & Benson, 2002; Rhodes & Tremewan, 1994), and sometimes even better (Calder et al., 1996; Rhodes, Brennan, & Carey, 1987). Thus, despite the fact that DBO is unable to identify photographs of famous people, he might display some intact capacities when caricatures of famous people are presented.

4.5.1. Method

4.5.1.1. Stimuli and procedure. The critical stimuli consisted of 15 caricatures of famous people taken from the Moscovitch et al. (1997) study. DBO was shown each caricature separately, and if he failed to recognize the caricature, a target name with three distractor names (semantically related, phonemically related, unrelated) were shown, and he was asked to select the correct name. Finally, if an accurate response was not supplied on the multiple-choice task, the name was read by the experimenter and the participant was required to supply semantic information about the figure.

4.5.2. Results and discussion

The presentation of a famous figure in caricature form did not help DBO recognize faces. He was unable to recognize any of the caricatures or to supply any identifying information about them (0/15, controls 12.74, S.D. = .89, $Z = -13.86$). Interestingly, when presented with the target face and the distractors, he was able to select the correct name for 10/15 caricatures. Thus, he was able to use the name of the figure as a guiding cue to process the visual information of the caricature. One explanation on how this was achieved may be through the activation of the mental image of the face by the name. Once this mental image was aroused, DBO was able to match it with the visual input. Although mental imagery of faces was not assessed systematically with DBO, informal observational data indicate that he can visualize faces of known figures (e.g., *Joseph Stalin*, *John F. Kennedy*, *Marilyne Monroe*, etc.). Alternatively, DBO may possess partly preserved access to semantic information, which, by itself, does not allow complete identification (e.g., Dixon et al., 1997). However, with an additional cue, such as a name, this semantic activation may reach a threshold which would allow identification. It is important to bear in mind, however, that his performance, though improved, was not excellent, and although all the figures were known to him (as confirmed by the last phase of the task where he provided information to the names of the faces he did not recognize), he was able to implement these presumed strategies for only two-third of the faces.

4.6. Famous people association task

In the present task we further tested DBOs' (dis)abilities to access semantic information about famous people by presenting three photographs of famous people, two of whom were closely related to each other. The verbal requirements in this task were eliminated as DBO did not have to supply the names of the people, but only to point to the two most closely related items. Moreover, intact performance of this task supposedly does not require explicit retrieval of semantic information and may depend on implicit associative activation of the related photographs.

4.6.1. Method

4.6.1.1. Stimuli and procedure. The critical stimuli consisted of 21 triads of famous figures (Fig. 6). The target picture (e.g., *Boris Yeltsin*) was situated on top of a page and the participant was required to match this target to one of the two faces shown on the lower part of the page (e.g., *Gerald Ford* and *Mikhail Gorbachev*). DBO was told that one of the two faces is most closely related to the target face. Following the presentation of the triads, each face was presented separately and DBO was asked to identify the face. Finally, the triads were presented orally and DBO was asked to select the most related pairs.

4.6.2. Results and discussion

DBO was able to match the target to the correct picture in only 12 triads, a result that does not differ from chance

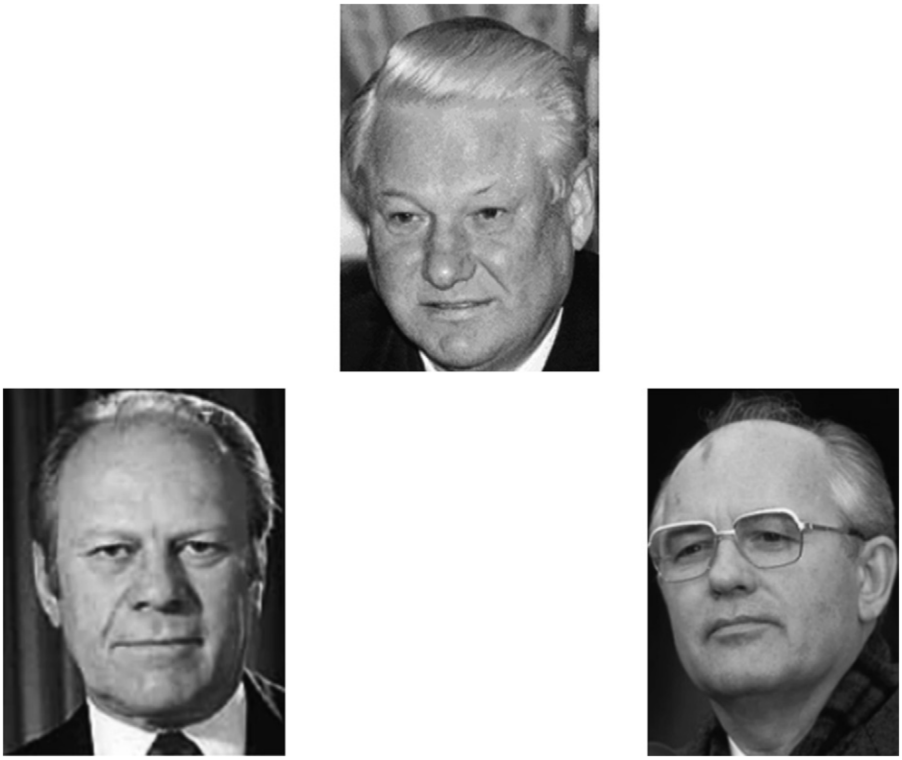

Fig. 6. Examples of familiar triads used in the famous faces association task (4.5; the faces depict *Boris Yeltsin*, *Gerald Ford*, and *Mikhail Gorbachev*).

performance ($\chi^2(1) = .04$, $p > .05$, controls 19.2, S.D. = 1.78, $Z = -4.02$). DBO was able to name only 2 out of the 59 faces presented (4 faces were presented twice across triads, controls 53, S.D. = 3.74, $Z = -13.63$). In contrast, he was able to perform the matching task correctly when given the names (19, controls 20.2, S.D. = .84, $Z = -1.43$). Thus, even in a task which does not require name generation or self-initiated retrieval of semantic information, DBO is unable to perform well when matching pictures of faces semantically, further strengthening the notion that his deficit lies at accessing the semantic knowledge via the visual modality.

4.7. Summary and discussion of DBO's prosopagnosia

DBO seems to have intact abilities to form a structural representation of a face. He was able to perform the standard tests (BFRT, RMF) within the normal range of both the published norms and his matched controls. Moreover, the shortcomings of some of these tests were taken into account and a modified test of recognition memory for faces was administered, in which only the internal features were presented. DBO again displayed normal performance, in accuracy *and* in RT, in both the immediate and delayed recognition tests. Finally, DBO showed inversion and misalignment effects which were similar in their magnitude to those of controls. Taken together, these findings attest to DBO's relatively preserved perceptual processing of faces.

Assessment of his higher order face identification capacities, however, revealed a unique dissociation between recognition of a face and its identification. DBO can recognize a famous face as familiar, and select it from an array containing similar distractors. Yet, he is unable to provide any identifying information about individuals from their faces even when a verbal response is not required. The inability to identify familiar faces is observed also for faces encoded prior to his injury, ruling out prosopamnesia as a possible cause for his deficits (Tippett, Miller, & Farah, 2000). When probed with the spoken name, however, he can supply the needed information indicating that his semantic knowledge is preserved but cannot be accessed via vision.

The pattern of deficits exhibited by DBO is consistent with the Bruce and Young's (1986) model of face recognition, according to which all faces, familiar and non-familiar alike, are first encoded as structural representations, which contain context-independent records of the face (structural encoding). From this stage the model distinguishes between a processing level in which a perceived face is compared to stored representations (FRU), and a level at which specific semantic information about the perceived face is activated and becomes accessible (PINs). DBO is able to compare the facial percept with its stored image and establish its familiarity, but he cannot access visually the specific semantic information associated with this person.

Several patients were described in the literature, who purportedly can be classified as associative prosopagnosics, as they do not show the classical low-level visual deficits characterizing apperceptive prosopagnosia (Carlesimo et al., 1998; de Haan, Young & Newcombe, 1991; Delvenne et al., 2004; De Renzi & di Pellegrino, 1998; Dixon et al., 1997; Farah et al., 1995; Henke et al., 1998; Nunn, Postma, & Pearson, 2001; Temple, 1992; Van der Linden, Bredart, & Schweich, 1995). The case of DBO is substantially different from the majority of these patients as his impairment appears to arise from deficits in higher level processes. Specifically, DBO is able to recognize famous faces as familiar whereas in most studies where this was examined, patients failed to achieve normal level of performance (e.g., Delvenne et al., 2004; Temple, 1992). Some patients, however, did show intact recognition. For example, ME (de Haan et al., 1991) performed as well as controls in judging the relative familiarity of highly familiar, low familiar and unfamiliar faces. Yet, she was extremely poor when asked to supply identity-specific semantic information or to name the person whose face it was. However, in contrast to DBO, she was equally impaired when names were also presented, indicating that her impairment is not domain or modality specific and may stem from a more general semantic memory degradation. Likewise, patient GB (Van der Linden et al., 1995) did not differ from controls when presented with famous faces and asked to judge their familiarity. Yet, only famous faces were presented to him in the familiarity task, thus biasing his response and probably yielding an overestimation of his recognition. Finally, RC (Carlesimo et al., 1998) was able to make familiarity judgments when presented with a famous face among distractors. Although this performance is highly suggestive of associative prosopagnosia, the lack of additional supporting evidence in that report prevent us from fully endorsing him as such. Note, however, that such familiarity/identification dissociation is predicted by theoretical models of face recognition (e.g., Bruce & Young, 1986) and is mirrored in the object recognition domain as well. It is, therefore, of no surprise that evidence for intact familiarity and impaired identification is apparent in DBO and possibly in other patients as well.

5. General discussion

This study reports a detailed examination of a patient DBO who presents with severe deficits in identifying famous faces and common objects in the visual modality. His semantic knowledge, however, is relatively preserved as demonstrated when stimuli are presented to him in modalities (e.g., auditory, tactile) other than vision, where his performance is compromised. Using different tasks, we attempted to assess whether his difficulties with both faces and objects in the visual modality stem from faulty processes at the early perceptual, structural encoding level, or at a later, semantic level. Converging evidence points to a similar locus of impairment in both domains. While his ability to form a structural representation of both faces and objects appears to be unimpaired, his ability to access visual semantics of both objects and familiar faces is severely damaged. Moreover, for both types of stimuli, we believe that the locus of the visual semantic impairment can be more precisely pinpointed. DBO is unable to access semantic information pertaining to the specific (or even general) identity of the face or the object. Nonetheless, he is able to discern whether he encountered the stimulus before, demonstrated by his ability to make familiarity and objects decision judgments to famous faces and objects, respectively. In the following discussion we will address the theoretical implications of these results to the ongoing debate concerning the underlying deficits of associative visual agnosia for faces and objects.

5.1. Can visual associative agnosias be attributed to perceptual deficits?

The classical distinction proposed by Lissauer (1890) between apperceptive and associative types of agnosia has served as an effective, albeit coarse, framework for understanding visual cognition of faces and objects for more than a century. Challenges to this dichotomy were voiced during the years (Bay, 1953; Farah, 1990), claiming that perceptual deficits are responsible for the emergence of associative visual agnosia. Yet, opposing views (Ettlinger, 1956; de Haan, Heywood, Young, & Edelsteyn, 1995) have reiterated the basic dichotomy by demonstrating that deficits in sensory processing are not sufficient to account for associative agnosia.

The debate has been recently re-opened both by considerations of the adequacy of traditional tests to detect perceptual deficits (Delvenne et al., 2004; Duchaine & Weidenfeld, 2003), and by new cases of associative prosopagnosia whose perceptual deficits are only revealed by more sensitive tests. A particularly illuminating example is patient NS described recently by Delvenne et al. NS has normal drawing abilities, and performs well on different object and face matching tasks used in various neuropsychological tests, suggesting that his deficit is of the associative type. When probed with more sensitive tests of higher order visual perception, however, such as recognizing objects from different viewpoints, and when his response times, as well as his accuracy, are taken into account, his performance is impaired. For these reasons, Delvenne et al. concluded that he cannot be regarded as a pure case of associative (prosop)agnosia, and is better classified as an integrative type (Humphreys & Riddoch, 1987).

Though we agree with their interpretation regarding this particular case, we take exception with their more far-reaching statement that "the present case and the previous literature supports the idea that 'associative' prosopagnosia refers actually to a deficit at the perceptual level" (Delvenne et al., 2004, p. 611). As these authors correctly remarked, a visual (prosop)agnosic without perceptual deficits, or one in whom the perceptual deficits are not sufficient to account for the associative (prosop)agnosia, may be described in the future. DBO appears to be such a case. The difference between his and NS's performance on various tests further reinforces the claim that DBO's primary deficit is at the semantic level. Despite his advanced age and hemianopia, DBO was not impaired, as NS was, in the BFRT and WRMF. His short- and long-term memory of non-familiar faces, presented without external features, was also comparable to that of healthy adults. In addition, he was able to perform well on a familiarity task while NS could not. Moreover, DBO's inversion and misalignment effects were normal and comparable in magnitudes to that of controls, while NS did not show an inversion effect. Finally, while DBO was able to perform normally on the *object decision task* and the *item match task* in the BORB, NS was severely impaired. Taking into account: (a) that more sensitive tests may reveal perceptual impairments in DBO, (b) that probing other domains (e.g., facial expression, age, and gender discriminations) may reveal perceptual deficits, and (c) that our conclusions are based on null findings, the present disparities between the two patients are consistent with the classic distinction between apperceptive and associative types of (prosop)agnosia, with NC being an integrative (prosop)agnosic and DBO, an associative one. Like Delvenne and colleagues, we leave open the possibility that tests more sensitive than ours may reveal perceptual deficits in patients like DBO, but it would be incumbent also to show how those perceptual deficits account for the associative (proso)agnosia, not just that they can co-occur.

5.2. The anatomical basis of associative prosop(agnosia)

Previous studies have localized the lesions underlying apperceptive prosopagnosia to right-posterior areas such as the lingual, fusiform, and parahippocampal gyri (for current review, see Mayer & Rossion, 2007). The generally consensual view is that the right hemisphere lesion is *necessary* to cause prosopagnosia, as to date only one case of prosopagnosia has been reported where the lesion was restricted to the left hemisphere (Mattson, Levin, & Grafman, 2000). The question whether this lesion is *sufficient* is still debated (e.g., Barton, Press, Keenan, & O'Connor, 2002; Delvenne et al., 2004; De Renzi & di Pellegrino, 1998).

In contrast, the lesion contributing to the emergence of associative prosopagnosia was claimed to be localized to bilateral anterior temporal lobes (Damasio, Tranel, & Damasio, 1990; De Renzi et al., 1991). For example, in patient ELM (Dixon et al., 1997) lesions in the right and left mesiotemporal lobes were apparent. Other studies point to different roles of the left and right temporal lobes in recognition of famous faces. Patients who underwent left anterior temporal lobectomy were impaired in naming faces of famous people but not in identifying characteristics related to these figures. In contrast, patients who underwent right anterior temporal lobectomy were also impaired in providing semantic information about these famous individuals (Glosser, Salvucci, & Chiaravalotti, 2003). A study with unilateral left or right epilepsy patients has revealed a similar trend (Seidenberg et al., 2002; see also Tranel, Damasio, & Damasio, 1997). Together, these studies emphasize the role of the temporal lobes in accessing semantic knowledge of familiar people by using visual facial knowledge (see also Gainotti, 2007, for recent review of single case and group studies which arrives at similar conclusions).

The primary lesion evident in DBO was in left occipital cortex, extending into the periventricular white matter, consistent with an infarct in the left posterior cerebral artery. This lesion accounted for his homonymous hemianopia. Whether this lesion alone could account for his associative agnosias, or whether other minor and distributed infarcts contributed to his deficits, remains unknown. However, his visual object agnosia resembles that of patient DJ reported by Fery and Morais (2003) whose lesion is in the same location, but who does not have multi-infarct diagnosis, suggesting that such a lesion on its own can produce the pattern of deficits we observed. Another patient who is similar to DBO in his functional impairments both in face and object domains is RC (Carlesimo et al., 1998). His lesions were also concentrated in the left hemisphere and involve the occipital pole and the mesial surfaces of the occipital and temporal lobes. One possible reason why such associative prosp(agnosia) deficits appear after these types of lesions is that the occipital lesion accompanied by periventricular white matter damage may prevent input from both occipital cortices from being conveyed to more anterior temporal regions, implicated in semantic processing and memory. This neuroanatomical account is consistent with our observations that structural processing of faces, presumably mediated by the right lingual and fusiform gyri, are spared in DBO, whereas associative, semantic aspects of face and object recognition are impaired, because structural information is disconnected from the neural substrate mediating semantics. If our hypothesis is correct, these latter cases represent an additional lesion locus from which associative visual agnosias could arise.

5.3. Summary

In the present study, we provide a detailed description of patient DBO who presented with difficulties in face and object identification. The pattern of his deficits is not consistent with impairments in the perceptual level, and cannot be fully accounted also by other pathological manifestations such as optic aphasia, semantic dementia, or prosopamnesia. We are therefore inclined to conclude that DBO demonstrate a classic case of associative agnosia for faces and objects, arising, most probably, from impaired visual access to semantic representations.

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