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Impairments in the Face-Processing Network in Developmental Prosopagnosia and Semantic Dementia

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Abstract

Background—Developmental prosopagnosia (DP) and semantic dementia (SD) may be the two most common neurologic disorders of face processing, but their main clinical and pathophysiologic differences have not been established. To identify those features, we compared patients with DP and SD.

Methods—Five patients with DP, five with right temporal-predominant SD, and ten normal controls underwent cognitive, visual perceptual, and face-processing tasks.

Results—Although the patients with SD were more cognitively impaired than those with DP, the two groups did not differ statistically on the visual perceptual tests. On the face-processing tasks, the DP group had difficulty with configural analysis and they reported relying on serial, feature-by-feature analysis or awareness of salient features to recognize faces. By contrast, the SD group had problems with person knowledge and made semantically related errors. The SD group had better face familiarity scores, suggesting a potentially useful clinical test for distinguishing SD from DP.

Conclusions—These two disorders of face processing represent clinically distinguishable disturbances along a right hemisphere face-processing network: DP, characterized by early configural agnosia for faces, and SD, characterized primarily by a multimodal person knowledge disorder. We discuss these preliminary findings in the context of the current literature on the face-processing network; recent studies suggest an additional right anterior temporal, unimodal face familiarity-memory deficit consistent with an “associative prosopagnosia.”

Keywords

prosopagnosia; face recognition; visual perception; semantic dementia; primary progressive aphasia

Disorders of face processing have a number of causes. Developmental prosopagnosia (DP), also called congenital prosopagnosia, is a disorder of face recognition from impaired development in the brain’s face-processing network (Dalrymple et al, 2012; Duchaine and

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Nakayama, 2006; Gruter et al, 2008). Prosopagnosia can also be acquired from strokes and other focal lesions, particularly those affecting the right hemisphere (Barton, 2008; Gainotti, 2014). The most common neurodegenerative disorder of face processing may be right anterior temporal lobe (ATL) atrophy from semantic dementia (SD), also known as the semantic variant of primary progressive aphasia (Busigny et al, 2009; Evans et al, 1995; Gainotti et al, 2003; Joubert et al, 2006; Thompson et al, 2003; Tyrrell et al, 1990).

Evidence is converging that these disorders affect parts of a right hemisphere face-processing network. The initial core face-perceptual components of this network are the occipital face area in the lateral inferior occipital cortex (Dricot et al, 2008; Gauthier et al, 2000; Jonas et al, 2012; Rossion et al, 2003) and the fusiform face area in the lateral medial fusiform gyrus (Kanwisher et al, 1997). Subsequent face-processing components are the superior temporal sulcus (Haxby et al, 2000) and the right inferior ATL (Davies-Thompson and Andrews, 2012; Fox et al, 2009; Haxby et al, 2000; Pyles et al, 2013; Rossion et al, 2012; Weiner and Grill-Spector, 2010).

The occipital face area may preferentially represent the parts of a face, primarily the eyes, nose, and mouth, at an early non-configural stage (Haxby et al, 2000; Pitcher et al, 2011a, 2011b). By contrast, the fusiform face area may optimally represent the invariant configuration of the geometric features that distinguish unique human faces (Atkinson and Adolphs, 2011; Barton, 2008; Barton and Cherkasova, 2003; Kanwisher, 2000; Kanwisher et al, 1999; Nakamura et al, 2000; Pinker, 1999), such as the arrangement and spacing of the eyes, nose, and mouth (Barton, 2009; Dalrymple et al, 2014; Kanwisher, 2000; Tanaka and Sengco, 1997).

The superior temporal sulcus responds to variant aspects of faces, such as expressions, eye gaze, and mouth movements, while emotional expression and context are handled in the amygdala, insula, and elsewhere (Haxby et al, 2002).

Finally, in the right ATL, intact structural percepts of faces are coupled with facial familiarity memories in “face recognition units” that, in turn, access identity-semantic information in the form of “personal identity nodes” (Young and Bruce, 2011). This face-processing model includes separate recognition pathways for faces (face recognition units), voices (voice recognition units), and names (name recognition units) that converge in personal identity nodes for semantic information about a person (Bruce and Young, 1986).

DP could affect face processing at the perceptual level, before the level of face recognition units, or else at the level of the face recognition units themselves. SD could affect face processing at the linkages from face recognition units to personal identity nodes, or else at the personal identity nodes themselves.

Although DP and SD appear to disrupt different stages of face processing, controversy continues over the precise mechanisms and possible overlap between the two disorders. For example, investigators suggest that the prosopagnosias fall into the agnosia dichotomy between *apperceptive*, caused by disturbed facial perceptions insufficient for recognition, and *associative*, caused by impaired linkage of facial perceptions to memories and other associations (De Renzi et al, 1991). DP may result from specific facial apperceptive

disturbances, such as encoding deficits for the spatial arrangement of facial elements (Barton et al, 2003). SD, which may extend to core face perceptual areas (Josephs et al, 2008; Omar et al, 2011), could result in an apperceptive prosopagnosia (Joubert et al, 2003), a progressive associative prosopagnosia (McNeil and Warrington, 1991; Tyrrell et al, 1990), or even face-processing difficulty owing to a multimodal person recognition disorder with inability to retrieve semantic associations (Gainotti, 2010, 2013).

In this preliminary study to clarify aspects of the face-processing network, we compared 10 individuals with face-processing difficulty, five with a lifelong difficulty in recognizing faces and the other five with right temporal-predominant SD. We further compared the DP and SD groups with ten normal controls on measures of visual perception, facial familiarity, person knowledge for famous people, and semantic aspects of face processing, as well as the identification of facial emotions. We predicted that the participants with DP would show an early configural problem in face perception and those with SD would reveal a later semantic problem in person knowledge, reflecting opposite ends of a right hemisphere face-processing network.

METHODS

Participants

We recruited ten people with a history of face-processing difficulty for this study, along with 10 normal controls. Of the participants with face recognition difficulty, five had a history of DP and five had SD.

We recruited four of the participants with DP from Internet websites for face-blind persons, and the fifth participant through the UCLA Behavioral Neurology Program. We focused on recruiting people with DP who were at least 40 years old in order to match them to the patients with SD. When the people with DP enrolled in the study, none of them had a history of autism. After the study, they scored in the non-autism spectrum (scores = 8, 13, 19, 20, 25) on the Adult Autism Spectrum Quotient questionnaire, on which the autism spectrum threshold is 26 (Baron-Cohen et al, 2001; Woodbury-Smith et al, 2005).

We recruited the five patients with right temporal-predominant SD through the UCLA Behavioral Neurology Program, where they had sought help primarily for face-processing difficulty. These five community-based patients had undergone a clinical evaluation and been diagnosed with mild to moderate cognitive impairment by their Mini-Mental State Examination (Folstein et al, 1975) scores. Magnetic resonance imaging had shown the patients to have right-predominant (ie, right greater than left) ATL atrophy. Figure 1 shows the imaging for one of the patients.

Ten age- and sex-matched normal controls from our registry of normal controls volunteered to undergo testing. None of the controls had any vision impairments or face-processing difficulty.

We matched the DP, SD, and control groups by age and education.

All 20 study participants were long-time (> 15 years) residents of California, Caucasian, and similar in their socioeconomic and educational backgrounds. We further screened all the participants to exclude any other neurologic, psychiatric, and medical conditions that required medication management or active intervention.

The UCLA Institutional Review Board approved the study. We obtained written informed consent from all participants.

Tests

We selected tests to assess general cognition, general visual perceptual skills, and multiple aspects of face processing. The screening task for visual object agnosia and most of the face-processing tasks were experimental assessments that we had developed and that were not standardized or normed tests. We interpreted all of the visual perceptual and face-processing tasks in a “process approach,” ie, by observing and noting the participants’ actual strategy during performance.

General Cognition and Visual Perception—A behavioral neurologist (M.F.M.) administered the tests of cognitive (Table 1) and visual perceptual functions (Table 2).

To evaluate a range of perceptual and spatial abilities, we gave the Hooper Visual Organization Test (Hooper, 1983), consisting of divided and rotated parts of objects requiring mental assembly; the Gestalt Completion Test (Ekstrom et al, 1976); several tests from the Visual Object and Space Perception Battery (Warrington and James, 1991); the Benton Judgment of Line Orientation Test, a matching task of lines arranged at different angles (Benton, 1983); the Rey-Osterrieth Complex Figure Copy (Osterrieth, 1944); and screening for visual object agnosia with our own test of 20 common, real objects presented for visual identification and naming.

Face Processing—To characterize the 20 participants’ face-processing abilities, we gave them seven facial tasks:

1. **Benton Facial Recognition Test, Short Form** (Benton, 1983). In each trial, we showed the participants a front-view photograph of a face and asked them to match it with one of the faces in a set of six photographs presented in front view, in three-quarters view, or in different lighting. During the test, we observed the participants, asked them about their strategy in matching the faces, and noted how long they took to complete the matching. The 54 test items had a published normative performance of 45.4 (standard deviation = 3.96) (Benton, 1983).
2. **Face Configuration.** In a modification of the method reported by Barton et al (2007a), we showed participants front-view photographs of 20 anonymous (non-famous) individuals taken from public sources. We presented the photographs one at a time. Along with each, we presented a display of four slightly altered faces, one of which was an altered version of the original face. We had distorted the faces with WinImage 7 software (www.winimage.com), minimally changing features and their spatial relationships (10% F/x program distortion at six key control points: eyes, ears, mouth, and nose, with an emphasis on slight alterations in spatial

position). We presented all of the facial stimuli as 4-inch $\times$ 4-inch photos. We asked the participants to match the master photograph with the correct altered face. The participants had unlimited time to decide. We tallied each participant's number and types of errors.

3. **Face Familiarity.** We showed the participants photographs of 24 familiar and 24 unfamiliar faces, one at a time. We had established the 24 familiar faces as being generally well known to most people in the participants' socioeconomic, educational, and geographical cohort (Mendez and Ghajarnia, 2001). We asked the participants to say whether each face looked familiar. We followed all "yes" answers by asking the participants what they knew about the person in the photograph, to see if they continued to feel that the face was familiar. We scored them with a "yes" for sense of familiarity if they persisted in saying that the face was familiar rather than changing their mind after being probed, regardless of whether they came up with any actual knowledge of the person. Our prior use of this protocol had yielded a normal performance of 43.49 (standard deviation = 4.5) (Mendez and Ghajarnia, 2001).
4. **Face Identification.** We showed the participants 24 photographs of famous people, one at a time, for naming or other identification. The famous people were either politicians or entertainers well known to most residents of southern California. The photographs were cropped 2.5-inch-square front-view portraits. We did not tell the participants anything about the famous people or their categories, but simply asked the participants to name the person. To exclude impairments in name recall, if participants could not name the person, we asked them to describe at least one of the person's major identifying features, eg, "movie star," "president," or "world leader." For each person, we had determined a minimum of three features that were specific, ie, unlikely to reflect random guesses. We scored participants as failing to identify a person if they could not either name or describe the person. Our prior use of a similar version of this Face Identification Task had yielded a normal performance of 21.36 (standard deviation = 2.4) (Mendez and Ghajarnia, 2001).
5. **Name-to-Face Matching.** In another modification of a reported method (De Renzi et al, 1991), we showed the participants 32 displays, each with four faces of people of the same sex. In each display, three of the four people were famous. One of the four faces was the index face (eg, former President Bill Clinton). The others were a semantically related famous face (eg, former President Jimmy Carter), a semantically unrelated famous face (eg, comedian Jay Leno), and an unfamiliar and not famous but visually similar face. (Figure 2 shows another example.) For each display, the examiner named the index person and asked the participant to point to that person's photograph. We tallied the number and types of responses, especially semantically related (pointing to the person in same semantic category) and visually related (pointing to the person with the greatest physical resemblance).
6. **Person Knowledge from Name.** We tested the participants' ability to identify semantic and visual facial aspects of 25 famous people. We told the participants the name of a famous person and asked them to describe him or her. For each famous

person, we had predetermined three salient semantic characteristics (general occupation, a prominent associated feature, and a specific accomplishment) and three salient visual characteristics (general appearance, hair, and eyes or some other facial feature). For example, for Elizabeth Taylor the three semantic characteristics were: famous actress, marriage to Richard Burton, and “Cleopatra” or some other famous movie. The three visual characteristics were: beautiful face, black or dark hair, and blue eyes or dark eyebrows. We asked the participants to keep describing characteristics until they named all six of our predetermined semantic and visual items. If they did not spontaneously describe a characteristic, we cued them with related questions. Their total score included the cued score.

7. **Ekman 60 Faces Test of Facial Emotions** (Ekman et al, 1972). This test presents 60 photographs from the original Ekman and Friesen series of ten actors (four men and six women) expressing six emotions (happiness, sadness, anger, surprise, fear, and disgust). The Ekman 60 Faces Test assesses recognition of facial expressions of these emotions. We asked the participants to identify the emotions expressed in the photographs, and we recorded their oral responses. One prior study reports a performance of 49.8 (standard deviation = 3.0) for normal participants (Diehl-Schmid et al, 2007).

Statistical Analysis

For the demographic and visual perceptual variables, we used one-way analysis of variance with Bonferroni-corrected post-tests, as well as two-tailed *t* tests and the Fisher exact test. For these analyses we used SPSS Statistics for Windows, Version 22.0 (IBM Corporation, Armonk, New York).

For the face tasks, whose results were not all normally distributed, we used nonparametric Kruskal-Wallis tests with Mann-Whitney post-tests with Bonferroni correction ($P < 0.017$) reported as the standardized test statistic (*z*). We also analyzed the Face Familiarity Task with signal detection theory methods to determine participants' true sensitivity to facial familiarity (d') and potential inclination or bias for endorsing faces as familiar or unfamiliar (*c*, which stands for *criterion*). For d' and *c*, we standardized the participant responses and used analysis of variance for group comparisons.

RESULTS

Baseline Characteristics

As shown in Table 1, the three study groups did not differ in age or education. As expected, however, they differed on basic cognitive measures. The patients with SD had significantly lower scores than the DP group and controls on the Mini-Mental State Examination, category fluency, naming and word recognition, and memory testing.

Visual Perceptual Tests

As shown in Table 2, the overall results did not reveal deficits in either the DP or SD group; however, we observed that the groups used different strategies on two of the tests. The participants with DP were slow and precise on the Benton Judgment of Line Orientation

Test and the Rey-Osterrieth Complex Figure Copy, both untimed tests. On the Rey-Osterrieth Complex Figure Copy test, the DP group copied in an unusual way: Rather than starting with the overall rectangle and the large items, as most people do, they drew feature by feature. By contrast, the participants with SD performed the two tasks at normal speed and copied the figure using the expected patterns.

Neither the DP nor the SD group had statistically significant problems on the Hooper Test, the Gestalt Test, Visual Object and Space Perception Battery, or visual object recognition, despite both groups trending toward doing worse than the controls.

Face-Processing Tests

Table 3 shows the results of the face-processing tests.

1. Benton Facial Recognition Test—We found no significant group differences on this test. Despite their normal scores, however, the participants with DP displayed a strategy similar to the one that they used for the Rey-Osterrieth Complex Figure Copy, of serial matching by salient features. When debriefed, all five participants with DP, but none of those with SD, said that they had tried to match the faces on the basis of eyebrows, hairline, nose, or some other specific feature.

2. Face Configuration—The three groups differed significantly on the Kruskal-Wallis analysis, primarily because of worse performance by the DP group. The post hoc analyses with Bonferroni correction showed that the participants with DP had done significantly worse than both the SD group ($z = 2.53$, $P < 0.017$) and the controls ($z = 2.98$, $P < 0.017$). The SD group and controls did not differ.

3. Face Familiarity—The three groups had significant differences on both d' and c . The DP group had worse d' scores than the SD group and the controls, despite both the DP and SD groups' having a conservative bias in not endorsing face familiarity if they were uncertain (c scores). The post hoc analyses revealed that the DP group did significantly worse than the controls on both d' (Tukey honest significant difference $P < 0.05$) and c (Tukey $P < 0.001$), and significantly worse than the SD group on c (Tukey $P < 0.001$). On d' , the differences between the DP and SD groups approached but did not attain significance.

4. Face Identification—The DP group performed significantly worse than the controls ($z = 3.08$, $P < 0.017$), as did the SD group ($z = 3.10$, $P < 0.017$). The SD group did worse than the DP group on this task, but, after Bonferroni correction, the difference did not reach the $P < 0.017$ level. Notably, all five participants with DP reported having tried to identify the famous people by their hair (even though we had cropped as much hair as possible out of the pictures, and some showed only a minimal amount), eyebrows, or another salient feature.

5. Name-to-Face Matching—The DP group performed significantly worse than the controls on total score, semantically related errors, and visually related errors ($z = 3.08$, 2.98 , and 3.24 , respectively; $P < 0.001$ for all); so did the SD group ($z = 3.08$, 3.18 , and 3.26 , respectively; $P < 0.001$ for all). Comparing the DP and SD groups, the only difference that

reached significance was that the SD group had more semantically related errors ($z = 2.60$, $P < 0.017$).

6. Person Knowledge from Name—The SD group performed significantly worse than the controls on total score, semantic items, and visual items ($z = 3.07$, 3.10 , and 3.10 , respectively; $P < 0.017$ for all). Likewise, the SD group did significantly worse than the DP group on all three scores ($z = 2.62$, 2.61 , and 2.64 , respectively; $P < 0.017$ for all). The DP group did not differ from the controls.

7. Ekman 60 Faces Test of Facial Emotions—We found no significant group differences.

DISCUSSION

Our study showed that two face recognition disorders, DP and SD, have different mechanisms for impaired face processing. Our participants with DP had impaired configural analysis; they relied on feature-by-feature processing and salient features to recognize faces. Our participants with SD had lost person knowledge and semantically related responses. This finding is consistent with the theory of distinguishable perceptual and semantic face-processing disorders along a right hemisphere face-processing network (Gobbini and Haxby, 2007; Young and Bruce, 2011).

In DP, face recognition deficits appear in early childhood. DP is heterogeneous and may be associated with visual perceptual difficulties and an elevated family risk of similar complex visual difficulties (Johnen et al, 2014; Wilson et al, 2010). Clinicians may confuse DP with the face-processing impairment seen in individuals with autism spectrum disorder (Wilson et al, 2010). Although two of our five participants with DP had a history of social isolation, language delay, or hyperlexia, none had been diagnosed with autism or had elevated Autism Quotient scores, and all were independent adults with normal relationships and successful jobs or careers (Barton et al, 2004a; Duchaine and Nakayama, 2004, 2006a).

Our findings and the literature indicate that people with DP have selective impairment in face perception from early childhood, with sparing of facial emotion recognition and covert (eg, physiological responsiveness) facial recognition (Chatterjee and Nakayama, 2012; Duchaine et al, 2003; Nemeth et al, 2014; Palermo et al, 2011a, 2011b; Rivolta et al, 2010, 2012, 2013; Towler and Eimer, 2012; Tree and Wilkie, 2010). When tested, patients with DP may have normal structural magnetic resonance imaging scans and may respond to faces with functional magnetic resonance activation in the fusiform face area (Avidan et al, 2005); however, there is much evidence that DP is associated with impaired configural processing of faces very similar to fusiform face area dysfunction (Barton et al, 2003, 2007a, 2007b; Huis in 't Veld et al, 2012). Because of this configural impairment, people with DP may revert, as did our participants, to sequential feature-by-feature inspection, particularly of salient features such as clothing, hairstyle, and eyebrows (Duchaine, 2000; Duchaine and Nakayama, 2004; Michelon and Biederman, 2003; Stollhoff et al, 2010).

In contrast to lifelong DP, investigators report the emergence of “progressive prosopagnosia” from right ATL atrophy associated with SD (Evans et al, 1995; George and Jose, 2009; Tyrrell et al, 1990). SD manifests as a loss of conceptual knowledge in word comprehension with left temporal involvement and in face processing with right temporal involvement (Thompson et al, 2003). Although the right temporal lobe atrophy may extend into the anterior fusiform gyrus and affect core face perception (Josephs et al, 2008; Joubert et al, 2003; Omar et al, 2011), most patients with SD have preserved face configural ability. Their impaired person knowledge is multimodal, affecting their perception of voices and names as well as faces (Busigny et al, 2009; Evans et al, 1995; Gainotti, 2007b, 2013; Gainotti et al, 2003; Hailstone et al, 2010; Joubert et al, 2006). Our patients with SD had loss of person knowledge for faces and made semantically related responses consistent with differential impairment of person-specific knowledge.

Since prosopagnosia is impaired familiarity with previously seen faces, the intact face familiarity among our patients with SD further suggests that they did not have prosopagnosia but, rather, a loss of person-specific knowledge (Gainotti, 2010). Feelings of familiarity appear to be generated at the recognition level, before explicit person identification and semantic information at the personal identity nodes level (Burton et al, 1990; Gainotti, 2014). Most patients with SD whose disease has not advanced into the right fusiform gyrus (Joubert et al, 2003) have a problem with person-specific knowledge at the personal identity nodes level rather than a true prosopagnosia (Busigny et al, 2009; Evans et al, 1995; Gainotti, 2010; Gainotti et al, 2003; Gobbini and Haxby, 2007; Tyrrell et al, 1990; Young and Bruce, 2011).

An important caveat to this conclusion is that the face-processing impairment in patients with SD has a potential contribution from impaired processing of names. In our current study, a naming impairment could have affected the SD group’s performance, particularly on the Name-to-Face Matching and Person Knowledge from Name tasks.

Despite our findings in patients with SD, some studies of patients with right ATL lesions indicate that unimodal face memory can be impaired independent of person knowledge (Drane et al, 2008; Gorno-Tempini and Price, 2001; Olson et al, 2007; Rogers et al, 2006; Simmons et al, 2010; Snowden et al, 2004; Tsukiura et al, 2008). Areas in the ventral polar ATL are sensitive to face memory (Von Der Heide et al, 2013). In addition, ATL damage can impair familiarity feelings from faces (and related person knowledge) while sparing familiarity feelings from names (and related person knowledge) (Gainotti, 2007a, 2007b; Gainotti et al, 2010). Although our patients with SD had impaired face identification, other patients with SD and right ATL lesions may have dissociable memory and familiarity difficulty at the recognition level for faces, voices, or names (Hailstone et al, 2011; Liu et al, 2014; Neuner and Schweinberger, 2000). Together, and in the absence of documented difficulty with voice or name recognition, these findings indicate that ATL lesions and early SD can also result in damaged “face recognition units” and a unimodal facial agnosia that could be described as a modality-specific “associative agnosia” (Barton, 2008; Barton et al, 2004a; Gainotti et al, 2008; Joubert et al, 2004; Liu et al, 2014; Snowden et al, 2004, 2012).

Our study had several possible shortcomings. First was the small number of participants, only five with DP, five with SD, and ten controls. For this reason, the study remains preliminary, as a larger number of participants might have revealed additional significant differences in performance. Nevertheless, we found differences sufficient to illustrate the two face-processing disorders and their effects on the face-processing network.

A second possible shortcoming is that DP is so heterogeneous that the prosopagnosia can take different forms and our results may not be generalizable to all people with DP. However, our DP group's impairments reflected a wide enough spectrum to be compared with patients with SD. In addition, patients with right side-predominant SD are relatively rare, thus allowing our comparison.

Finally, because many of our tasks have not been standardized or normed, they permit only an experimental comparison between groups.

In conclusion, there are at least two distinct face-processing disorders, DP and AD. Most, but not all, people with DP have apperceptive difficulty in face configuration. In contrast, most, but not all, patients with right temporal-predominant SD have a multimodal person knowledge disorder with a potential contribution from impaired semantic knowledge for names. Our findings in this study and a review of the literature support a right hemisphere face-processing network (Busigny et al, 2009; Haxby et al, 2000; Hudson and Grace, 2000; Takahashi et al, 1995) that begins in the occipital face area and fusiform face area with feature detection and configural binding into a face, as shown by our patients with DP. The network ends in the right ATL at the personal identity nodes level with the linking of perceptual representations to semantic memories or person knowledge, as shown by our patients with SD (Gobbini and Haxby, 2006, 2007; Grossi et al, 2014; Nakamura et al, 2000; Shah et al, 2001). The literature further suggests an intermediate disturbance of face memory familiarity in the right ATL or its connections. Clinicians can distinguish DP from AD using clinical tests like the ones we described here. Future studies may clarify our findings in larger cohorts of individuals with DP and SD.

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Glossary

ATL	anterior temporal lobe
DP	developmental prosopagnosia
SD	semantic dementia

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FIGURE 1.

Representative brain magnetic resonance images (coronal views) from our study participants. Panel 1: This image, from the only member of the developmental prosopagnosia group who had an available scan, shows a normal anterior temporal region. Panels 2–6: One image is shown from each of the patients with semantic dementia. All have atrophy involving the inferior lateral anterior temporal lobes bilaterally, but more pronounced on the right, consistent with semantic dementia.

FIGURE 2.

Example of a display for Face Recognition by Name. The examiner asks the participant, “Which one is Adolph Hitler?” The three faces other than Hitler’s (top right) are a semantically related familiar face (Joseph Stalin, top left), a physically similar but generally unfamiliar face (bottom left), and a familiar but semantically unrelated and only partially similar-looking face (Charlie Chaplin, bottom right). We kept the large details of all the faces at least somewhat similar to avoid presenting a salient difference of cue.

TABLE 1
Baseline Characteristics of Patients with Developmental Prosopagnosia (DP), Patients with Semantic Dementia (SD), and Controls

	DP (n = 5)	SD (n = 5)	Control (n = 10)	Significance (df = 2,17)
Age (years)	45.6 (16.68)	60.2 (6.22)	54.2 (7.31)	NS
Sex	3 men, 2 women	4 men, 1 woman	6 men, 4 women	NS
Education (years)	15.3 (2.59)	15.2 (2.17)	14.8 (3.01)	NS
Digit span	7 (1.1)	5.8 (1.1)	7.2 (1.3)	NS
Verbal fluency: "F" words/minute	15.8 (3.77)	10.2 (6.8)	14.6 (2.3)	NS
Category fluency: animals/minute	18.0 (3.74)**	8.0 (4.42)**	19.1 (2.71)	$F = 18.51, P < 0.0001$
Mini-Mental State Examination ¹ (maximum = 30)	28.6 (1.14)*	25.2 (2.77)*	28.4 (1.3)	$F = 6.68, P < 0.01$
Mini-Boston Naming ² (maximum = 15) (% error recognition)	14.4 (0.89)*** (100%)	4.2 (2.17)*** (42.8%)	14.2 (1.1) (100%)	$F = 98.23, P < 0.0001$
CERAD ³ : delayed recall (maximum = 10)	8.8 (0.84)**	2.2 (3.19)**	8.6 (1.02)	$F = 26.25, P < 0.0001$
CERAD ³ : recognition (maximum = 20)	19.0 (1.0)*	14.6 (3.65)*	18.7 (1.2)	$F = 8.04, P < 0.01$

Data are shown as numbers or mean (standard deviation).

Significant differences from controls are shown in **bold type**.

Significant differences for DP versus SD:

* $P < 0.05$.

** $P < 0.01$.

*** $P < 0.0001$.

¹ Folstein et al, 1975.

² Kaplan et al, 1983.

³ Welsh et al, 1991.

NS = not significant. CERAD = Consortium to Establish a Registry in Alzheimer's Disease.

Visual Perceptual Test Results for Patients with Developmental Prosopagnosia (DP), Patients with Semantic Dementia (SD), and Controls

TABLE 2

	DP (n = 5)	SD (n = 5)	Control (n = 10)	Significance (df = 2,17)
Hooper Visual Organization ¹ (maximum = 30)	26.0 (1.58)	23.8 (2.68)	26.4 (2.05)	NS
Gestalt Completion Test ² (maximum = 20)	16.4 (1.48)	15.6 (1.52)	16.8 (2.01)	NS
Visual Object and Space Perception Battery ³				
Incomplete Letters (maximum = 20)*	18.1 (0.89)	17.2 (1.1)	18.4 (1.08)	NS
Silhouettes (maximum = 30)*	25.61 (1.3)	24.4 (1.95)	25.8 (1.87)	NS
Number Location (maximum = 10)*	9.6 (0.55)	9.8 (0.45)	9.8 (0.45)	NS
Benton Judgment of Line Orientation Test ⁴ (maximum = 30)	23.4 (2.7)	25.0 (0.71)	25.4 (1.7)	NS
Rey-Osterrieth Complex Figure Copy ⁵ (maximum = 36)	34.0 (2.35)	34.2 (1.3)	34.54 (1.8)	NS
Visual object recognition (maximum = 20)	18.6 (0.71)	18.2 (2.05)	19.0 (1.11)	NS

Data are shown as mean (standard deviation).

¹ Hooper, 1983.

² Ekstrom et al, 1976.

³ Warrington and James, 1991.

⁴ Benton, 1983.

⁵ Osterrieth, 1944.

NS = not significant.

TABLE 3

Face-Processing Test Results for Patients with Developmental Prosopagnosia (DP), Patients with Semantic Dementia (SD), and Controls

	DP (n = 5)	SD (n = 5)	Controls (n = 10)	Significance (df = 2,17) [†]
Benton Facial Recognition ¹ (maximum = 54)	39.0 (7.48)	44.0 (1.22)	43.22 (5.74)	NS
Face Configuration ² (maximum = 20)	13.0 (2.24)*	16.2 (0.84)*	17.52 (1.7)	11.83, $P < 0.017$
Face Familiarity ³ (maximum = 48)				
d'	1.44 (0.52)	2.36 (0.62)	2.41 (0.77)	3.64, $P < 0.05$
c	0.62 (0.18)**	0.164 (0.09)**	0.16 (0.12)	23.0, $P < 0.001$
Face Identification ³ (maximum = 24)	3.6 (2.07)	1.6 (0.89)	20.8 (1.81)	15.05, $P < 0.001$
Name-to-Face Matching ⁴ (maximum = 32)				
Total score	14.6 (2.79)	17.0 (1.22)	30.09 (1.81)	14.83, $P < 0.001$
Semantically related errors	3.6 (1.42)*	5.6 (1.3)*	1.5 (0.71)	15.14, $P < 0.001$
Visually related errors	6.8 (2.59)	5.4 (0.55)	0.3 (0.48)	15.58, $P < 0.001$
Person knowledge from name (maximum = 150)				
Total score	125.0 (3.61)*	61.0 (7.71)*	120.22 (17.73)	10.82, $P < 0.017$
Semantic items	47.0 (8.15)*	24.2 (2.59)*	45.2 (6.07)	11.20, $P < 0.017$
Visual items	73.6 (8.65)*	36.8 (6.42)*	73.8 (12.93)	10.78, $P < 0.017$
Ekman Faces ⁵ (maximum = 60)	50.0 (4.3)	45.4 (2.07)	49.6 (4.06)	NS

Data are shown as mean (standard deviation).

Significant differences from controls are shown in **bold**.

[†] All values are Kruskal-Wallis χ^2 except for the F statistic for Face Familiarity.

Significant differences for DP versus SD:

* $P < 0.017$.

** $P < 0.001$.

¹ Benton, 1983.

² Modified from Barton et al, 2007a.

³ Mendez and Ghajarnia, 2001.

⁴Modified from De Renzi et al, 1991.

⁵Ekman et al, 1972.

NS = not significant.