

Famous faces and voices: differential profiles in early right and left Semantic Dementia and in Alzheimer's Disease

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ABSTRACT

Background: Famous face and voice recognition is reported to be impaired both in semantic dementia (SD) and in Alzheimer's Disease (AD), although more severely in the former. In AD a coexistence of perceptual impairment in face and voice processing has also been reported and this could contribute to the altered performance in complex semantic tasks. On the other hand, in SD both face and voice recognition disorders could be related to the prevalence of atrophy in the right temporal lobe (RTL).

Objective: The aim of the present study was twofold: (1) to investigate famous faces and voices recognition in SD and AD to verify the hypothesis that the two diseases show a differential pattern of impairment, resulting from disruption of different cognitive mechanisms; (2) to check if face and voice recognition disorders prevail in patients with atrophy mainly affecting the RTL.

Materials: To avoid the potential influence of primary perceptual problems in face and voice recognition, a pool of patients suffering from early SD and AD were administered a detailed set of tests exploring face and voice perception. Thirteen SD (8 with prevalence of right and 5 with prevalence of left temporal atrophy) and thirteen AD patients, who did not show visual and auditory perceptual impairment, were finally selected and were administered an experimental battery exploring famous face and voice naming and recognition.

Results: Results showed a differential performance profile in the two diseases, because AD patients were significantly impaired in the naming tests, but showed preserved recognition, whereas SD patients were profoundly impaired both in naming and in recognition of famous faces and voices. Furthermore, face and voice recognition disorders prevailed in SD patients with RTL atrophy, who also showed a conceptual impairment on the Pyramids and Palm Trees test more important in the pictorial than in the verbal modality.

Discussion: The data support the hypothesis of a different cognitive basis for impairment of face and voice recognition in the two dementias and suggest that the pattern of impairment in SD may be due to a loss of semantic representations while a defect of semantic control, with impaired naming and preserved recognition might be hypothesized in AD. Furthermore, the prevalence of face and voice recognition disorders in SD patients with atrophy mainly affecting the RTL and the presence in the same patients of a pictorial

conceptual defect are consistent with the hypothesis assuming that person-specific knowledge may be mainly based upon non-verbal representations in the RTL.

Key words: famous faces – famous voices–semantic dementia - Alzheimer’s disease – right temporal lobe atrophy

1. INTRODUCTION

Loss of knowledge for famous persons is one of the presenting features of Semantic Dementia (SD), a neurodegenerative disorder characterized by anterior temporal lobe (ATL) atrophy (Snowden, Neary & Mann, 1996; Neary, Snowden, Gustafson, Passant, Stuss, Black, Freedman, Kertesz, Robert, Albert, Boone, Miller, Cummings & Benson, 1998; Hodges, Patterson, Oxbury & Funnell, 1992). An alteration of this domain can be detected from the earliest stages of disease (Hailstone, Ridgway, Bartlett, Go, Buckley, Crutch & Warren, 2011), mainly affecting person recognition from face when the atrophy is prominent in the right ATL and from their name when it mainly affects the left ATL (Snowden, Thompson & Neary, 2004 and 2012; Gainotti, 2007 and 2015a). Results of Van Lanker & Canter's (1982) early investigations and of Gainotti's (2011) recent review of voice recognition disorders showed that these defects, just as face recognition disorders, are mainly due to right temporal lesions. Loss of knowledge of famous persons is one of the clinical features listed in the diagnostic criteria of Semantic Dementia (Neary et al., 1998).

In Alzheimer's Disease (AD) the earliest and most frequent sign is loss of episodic memory. Famous person knowledge is neither a presenting symptom nor is usually a prominent feature occurring during the course of the disease and is not mentioned as one of the clinical key features in the revised diagnostic criteria of AD (McKhann, Knopman, Chertkow et al., 2011).

It follows that to detect an impairment in this domain should help the clinician in the differential diagnosis since this disturbance is a prototypical feature of SD whereas it is presumably spared in AD. However, even if the prototypical clinical pictures of the two diseases seems to support this

assumption, neuropsychological investigations of this domain in SD and AD revealed that both SD and AD patients show problems on tests of famous person knowledge.

There are few studies exploring this issue in SD case groups. Most of literature is characterized by detailed studies performed on small groups of patients or on single cases, likely due to the rarity of this syndrome that makes it hard to recruit a high number of patients. The first study focusing on famous faces in a group of SD patients came from Snowden et al.(2004), who tested famous faces and famous names knowledge on 15 SD patients and 17 AD patients. Both patients groups were impaired compared to controls. However, SD patients were considerably more impaired than the AD group and only the SD patients were significantly impaired in forced choice familiarity judgement tasks. Furthermore, SD patients with predominantly left temporal lobe atrophy identified faces better than names, whereas patients with right temporal lobe atrophy showed the opposite pattern of performance. The existence of separate stores for faces and names was confirmed by results of a systematic survey made by Gainotti (2007) of all published cases of patients who showed a selective disorder of famous people recognition due to prevalent damage of the right or left ATLS. Results of this review documented a defective retrieval of person specific semantic information from face stimuli, when the right temporal lobe was damaged and a prevalent impairment in finding their names when the anterior parts of the left temporal lobe were affected. In 2011 Hailstone et al. performed a detailed examination of voice processing in 13 patients with temporal lobe atrophy and 22 AD patients. Both SD and AD patients showed deficits in voice recognition. Face and name recognition were impaired in both groups but deficits were more severe in SD patients. AD patients also showed impaired vocal gender perception and voice discrimination, consistent with the additional presence of perceptual impairments in voice processing. These authors also took into account the problem of asymmetric temporal lobe contributions to different components of person knowledge, but could not resolve this issue, because of small case numbers. In the present introduction we will mainly take into account the

problem of face and voice recognition disorders in AD and of the possible underlying semantic mechanisms, because this issue has been thoroughly investigated in the neuropsychological literature. For instance, famous face recognition has been widely studied in AD (Calabria, Sabio, Martin, Hernández, Juncadella, Gascón-Bayarri, Reñé, Ortiz-Gil, Ugas & Costa, 2012; Werheid & Clare, 2007, Clague, Dudas, Thompson, Graham & Hodges, 2005; Greene & Hodges, 1996, Hodges, Salmon & Butters 1993), showing that person knowledge deficits are present since the early stages of the disease (Clague et al., 2005). There is also a large agreement to acknowledge that the impairment in famous faces naming and identification in some degenerative diseases is not secondary to name access deficit, but to actual loss or impaired retrieval of stored knowledge of persons. More specifically, the nature of the poor performance in tasks involving semantic knowledge of famous persons is reasonably attributable to a primary semantic impairment in SD, because in these patients semantic knowledge is selectively impaired. On the other hand, in AD it is harder to explain the patients' performance in semantic tasks entirely as a consequence of a semantic impairment for a number of reasons. The extent of degradation of the semantic network in Alzheimer's Disease is questionable (Reilly, Peelle, Antonucci & Grossman, 2011). Some researcher (e.g. Hodges & Patterson 1995; Rogers, Ivanoiu, Patterson & Hodges, 2006) are in favour of a loss of semantic knowledge in AD, because semantic impairment is consistent across a variety of semantic tasks and across distinct modalities (verbal and visual channels). Other authors posit that the poor performance in semantic tasks could reflect an impaired retrieval from the semantic network (Nebes, Martin & Horn, 1984) or a deficit of strategic processing (Rich, Park, Dopkins & Brandt, 2002; Moreaud, David, Charnallet & Pellat, 2001). Still other authors (e.g. Reilly et al, 2011) maintain that the observation that language problems in AD usually consist of anomia in the absence of frank defects in single words comprehension points to an impairment of the lexical (rather than of the properly semantic) aspects of language in AD

These positions, questioning the existence of a loss of semantic knowledge in AD patients are supported by data (Rogers & Friedman, 2008) showing that when implicit lexical-decision priming

tasks were applied to AD and SD patients, different results were obtained by subjects belonging to these pathological entities. SD patients showed no priming effect for any semantic relationship, whereas AD patients showed an intact priming in superordinate and coordinate conditions but not an attribute priming. According to the authors, these data show that impairment in explicit semantic tasks in AD can be explained in terms of a deficient explicit retrieval in combination with a minimal disruption of the semantic network. Corbett, Jefferies, Burns & Lambon Ralph (2012) have also hypothesized that the semantic impairment of AD patients may be due a defect of semantic control similar to that described by Jefferies & Lambon Ralph (2006) in aphasic stroke patients. These authors have distinguished the semantic disorders of SD patients, which might be due to disruption of amodal representations, from the semantic disorders of aphasic stroke patients, which might result from a defect in the executive processes that help to control semantic activation in a task-appropriate fashion. This hypothesis is supported by the fact that the deficit of executive functions subsuming the defect of semantic control is present both in stroke and in AD patients (Corbett et al., 2012). In any case, the coexistence in AD of an impairment of several neuropsychological domains could influence performance in semantic tasks. In particular the potential presence of perceptual problems could play an important role in famous people recognition tasks, because an alteration of visual and auditory perception, often reported in AD (e.g. Golden, Augustus , Goll , Downey, Mummery, Schott, Crutch & Warren, 2015; Johnson & Chow, 2015; Kaeser Ghika J & Borruat, 2015; Moyse, Bastin , Salmon & Brédart, 2015) could influence the patient performance in semantic tasks involving face and voice domains.

In most studies of famous face and voice knowledge, tasks exploring face and voice perception were not performed and when this domain was investigated, such as in the Hailstone et al.'s (2011) work, it was found that the pattern of voice recognition impairment in AD was a sum of primary perceptual problems and of semantic association disorders. Obviously, when a perceptual impairment is present, it becomes hard to evaluate to what extent the perceptual deficit can

influence performance on semantic tasks which are dependent on the perceptual analysis of the shown stimuli.

The aim of the present study therefore consisted in investigating famous faces and voices in early AD and SD excluding the potential impact of perceptual impairment on patient profile. We administered to AD and SD patients a detailed set of neuropsychological tests exploring face and voice perception. Only patients who showed preserved perceptual abilities were included in the study and investigated with tasks exploring naming and recognition of famous faces and voices. We had two distinct predictions, concerning on one hand the differences between AD and SD patients in the recognition of famous faces and voices and, on the other hand, the prevalence within SD patients of face and voice recognition disorders when the atrophy mainly affected the right ATL. As for the first point, we expected to find in SD patients a failure both in naming and in recognition of famous faces and voices, whereas more problematic were the predictions concerning AD patients. To be sure, we expected to find a naming deficit in these patients, but we did not know if their capacity to recognize famous people through the visual (face) and auditory (voice) modalities would be impaired or spared. As for the second point, we expected to find a defect in naming and comprehension of famous faces and voices much more important when the right ATL was damaged than when the atrophy affected the left temporal lobe.

2. MATERIAL AND METHODS

2.1. Participants

The study group comprised 13 SD and 13 AD patients, all in the early stages of disease. Clinical diagnoses of SD and AD were made on the basis of clinical history, neuropsychological profile, structural neuroimaging (MRI or CT scan) and functional imaging (SPECT or PET). All patients fulfilled the respective international diagnostic criteria for AD or SD (Dubois, Feldman, Jacova et al., 2007; McKhann et al., 2011, Neary et al., 1998).

Twenty relatives of patient participants, who agreed to take part in the study, served as healthy controls (HC). All participants were native Italians who had lived in Italy from birth.

2.1.1. Evaluation of primary perceptual problems

Participants were excluded if there was a clinical history or objective evidence of hearing loss on audiometry. Similarly, they were excluded if they had visual deficits due to ocular disease, uncorrected by spectacles. A further exclusion criterion was the presence of primary visual and auditory perceptual problems, assessed with a set of tasks checking age, gender and same/different person discrimination through face and voice perception.

Face perception: a) age discrimination: 30 photographs of unfamiliar people, 15 “old” (>50 years) and 15 “young” (<50 years) were presented in turn. The participant was asked to report if the person was “old” or “young”; b) gender discrimination: 30 photographs of people (15 men and 15 women) were presented in a random order. Hairs were hidden and men had neither beard nor moustaches. The participant was asked to specify the gender; c) same/different face discrimination: a test similar to the Benton & van Allen (1972) face discrimination test, involving 30 face-pairs photographed from different perspectives (full face and $\frac{3}{4}$ view) was used. Hairs and other non-facial features were eliminated. Half the pairs were of the same person and half of different people. Participants were required to judge if the two photographs represented or not the same person.

Voice perception: very similar tests were used in the auditory modality: a) age discrimination, involving 20 voices from old (10) and a young (10) people; b) gender discrimination, involving 20 male (10) and female (10) voices; c) same /different voice: 20 voice-pairs were presented, 10 of the same person and 10 different. Voice-pairs with different identity were matched for gender and age. The stimuli consisted of 15 seconds sentences, in which the recorded person was talking about a specific topic (usually linked to personal experiences or feelings).

All tests were preceded by three practice items. Performance was compared with that of 46 healthy controls collected locally and patients were excluded if their performance fell more than 2 standard deviations outside the normal range for healthy subjects.

All participants gave their written informed consent and the study was carried out in accordance with the Helsinki declaration

2.1.2. Background neuropsychology

Background neuropsychological data were available for all patients enrolled. They included the MMSE (Folstein, Folstein & McHugh, 1975) as a measure of general cognitive status, the Bisyllabic Word Span and the Corsi Blocks (Spinnler & Tognoni, 1987) to assess short term memory, the Rey's auditory verbal learning test/Rey AVLT (Spinnler & Tognoni, 1987), and the delayed recall of the Rey-Osterrieth Complex Figure B (Luzzi, Pesallaccia, Fabi, Muti, Viticchi, Provinciali & Piccirilli, 2011) to evaluate long term memory. Furthermore, the Luria's Motor Sequences (Luzzi et al., 2011), the Stroop test (Luzzi et al., 2011) and the phonological fluency test (Caltagirone, Gainotti, Masullo & Miceli (1979) were used to assess executive functions and attention. The Visual Object and Space Perception Battery (Warrington & James, 1991) was used to evaluate visuo-perception and visuospatial abilities, whereas constructional praxis was assessed by means of the Rey-Osterrieth Complex Figure B copy version (Luzzi et al., 2011) and ideomotor apraxia using the test described by Luzzi, Piccirilli, Pesallaccia, Fabi & Provinciali (2010). Semantic-lexical functions were assessed using the easy picture naming test and the forced-choice word-picture matching test proposed by Snowden et al. (2004) and a test of categorical word fluency (Luzzi et al., 2011). Furthermore, conceptual-semantic associations were studied by means of the verbal and pictorial version of the Pyramids and Palm trees test/PPTT (Howard & Patterson, 1992).

All SD patients exhibited temporal lobe atrophy on structural magnetic resonance imaging (MRI), which was frankly asymmetrical. Eight showed a predominantly right and 5 a predominantly left sided atrophy. All patients with left-predominant atrophy had presented with word finding problems whereas in SD patients with right-predominantly atrophy the presenting symptom was a difficulty recognising famous and familiar faces.

The AD, SD and HC groups were well matched with respect to epidemiological variables. They did not significantly differ in age, education or sex distribution. The SD and AD groups did not differ in disease duration at assessment, or age at onset of disease. Furthermore, right and left SD patients did not differ in terms of demographics : age (U=15; ns), education (U=16; ns), sex ($X^2= 0.92$; ns), age at onset (U=16; ns), disease duration (U=18; ns), general cognitive impairment (MMSE: U=18.5; ns) or severity of semantic breakdown (naming: U=5.5; ns; word comprehension: U=9; ns).

2.2. Materials

A pool of sixty photos and 40 audio fragments of famous persons was initially selected. This initial pool of items was administered to a local group of 42 healthy controls (23 women and 19 males) aged > 50 years, who were requested to identify the person to whom the face or the voice belonged. The items were then rank-ordered on the basis of the frequency of correct answers and the first 49 photos and 30 voice fragments were selected to enter the final version of the test..

Famous face naming: the selected 49 photographs of famous people were presented consecutively, and patients were asked to name them.

Famous face recognition: recognition was explored through face-name matching test. The patient was shown the 49 famous faces in turn, and for each was asked to select the corresponding name from four alternatives, printed in the four quadrants of an A4 page. The four choices comprised: (1) the correct name (target); (2) the name of a famous person of similar age, sex and occupation as the

target (semantic distractor); (3) the name of an unrelated famous person (unrelated distractor) and (4) a non famous name, matched for sex and name characteristics and similar in sound (e.g. “Sofia Loren” → “Maria Polen”) (non-famous distractor). The positioning of the target was randomised.

Famous voice naming: patients listened to the selected 30 audio clips of famous people and were asked to identify the famous persons from their voice. In selecting voice stimuli care was taken to ensure that it was not possible to infer identity on the basis of sentence content (e.g. politician talking about politics; comedians producing a catch phrase with which they are associated).

Famous voice recognition: a 30-item cross modal, voice-to-name matching test was used. Distractor types and test methodology were the same used for the face-to-name matching test.

2.3. Statistical analysis

Because data were not normally distributed and the number of subjects was small, non-parametric tests were applied both in the comparisons between AD, SD and HC subjects and in the comparison between right and left variants of SD. Kruskal-Wallis and Mann Whitney tests were used for continuous variables and chi-squares for nominal variables.

Correlative analyses used Spearman’s statistic. McNemar’s test was used for the item-by-item analysis in recognition tasks.

3. RESULTS

3.1. Background neuropsychology

Background neuropsychology data of AD and SD patients are reported in Table 1

The SD and AD groups did not differ in term of general cognitive level. There were no significant differences on most tests. Predictably, AD patients performed significantly worse than SD patients on non-verbal long term memory (Rey’s figure B delayed recall) and spatial tasks (number location and cube analysis subtests of the VOSP), as well as on the number of errors II on the Stroop test. By

Famous face naming: both SD and AD groups were impaired compared to controls (SD vs. controls: $U=6.5$ $p<0.001$; AD vs. controls: $U= 28$; $p<0.001$). SD performance was also significantly poorer than AD performance (SD vs. AD: $U=19$; $p=0.001$).

Famous face-to-name matching: AD did not differ from healthy controls ($U=80.5$; n.s.). By contrast SD patients scored worse than AD and control subjects (SD vs. AD $U=20.5$; $p<0.001$; SD vs HC: $U= 20$; $p<0.001$).

Famous voice naming: both disease groups achieved significantly lower scores than healthy controls (AD vs HC: $U= 0$, $p<0.001$), SD vs HC: $U=0$, $p<0.001$). SD performance was significantly lower than AD performance ($U=1$, $p<0.001$).

Famous voice-to-name matching: AD patients did not differ from healthy controls ($U= 115.5$; ns). By contrast, SD patients performed significantly more poorly than both controls (SD vs HC: $U=0$; $p<0.001$) and AD patients (SD vs AD: $U=6.5$, $p<0.001$).

3.2.2. Individual subjects analysis within AD and SD patients

Looking to individual differences, when single subjects were compared to controls the number of subjects whose performance was at least two standard deviation lower than control means were the following: in face naming 6 AD and 11 SD; in face-to –name matching 3 AD and 11 SD; in voice naming 10 AD and 13 SD ; in voice-to-name matching 4 AD and 13 SD.

3.2.3. Modality / group interactions in AD and SD patients and in HC

The relationship between performance as a function of a) task type and b) input modality was also explored in AD and SD patients and in healthy controls .

Face naming vs. face matching: Both SD and control subjects showed a positive correlation between face naming and matching performance (SD: $r=0.62$; $p=0.03$; controls: $r=0.68$; $p= 0.001$). The correlation failed to reach statistical significance in the AD group ($r=0.5$; $p=0.09$).

Voice naming vs. voice matching: Both SD and controls showed a significant positive correlation between voice naming and matching performance (SD: $r=0.81$; $p=0.001$; controls: $r=0.58$; $p=0.007$). AD patients showed no significant correlation ($r=0.29$; $p=0.36$).

Face naming vs. voice naming and face matching vs voice matching: No significant correlations were found in any group between face and voice naming performance (SD: $r=0.06$; $p=0.84$; AD 0.08 ; $p=0.79$; controls: $r=0.02$; $p=0.93$). SD patients showed a significant correlation between face and voice matching performance (SD: 0.69 ; $p=0.02$). Performance in the AD and control groups was not correlated. (AD: -0.15 ; $p=0.61$; controls: -0.21 ; $p=0.36$).

3.2.4. Comparisons between left and right variants of SD

Figure 2 reports the mean values of scores obtained by right and left variants of SD on the naming faces and naming voices tasks and on the face-name and voice-name matching tasks.

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Figure 2
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Data reported in these figures suggest an effect of both side of atrophy and recognition modality, because scores obtained by SD patients with right-sided atrophy are systematically lower than those obtained by SD patients with left-sided atrophy and results obtained with voice stimuli are always worse than those obtained with face stimuli. The difference between patients with right- and left-sided atrophy was, however, not significant for the naming tasks (face naming: $U=11.5$; ns; voice naming: $U=12.5$; ns). On the contrary, on the recognition tasks, SD patients with right-sided atrophy scored significantly worse on the face-name matching task ($U=5$; $p=0.04$), and showed a trend toward significance ($U=5.5$; $p=0.07$) in famous voice recognition. The discrepancy between results obtained on naming and recognition tasks could be due to the fact that in the naming tasks a floor effect and the small number of subjects taken into account did not allow to reach significance.

As for the differences related to the recognition modality, they did not reach the level of statistical significance.

3.3. Results obtained on items in which the same famous people was present in both test modalities

In 19 items of the face and voice tasks the same famous people were present in both test modalities whereas in other items different celebrities were present only for one task-type. When the correlative analyses were restricted to the subset of 19 people common to both test modalities, the correlation for all groups were the following: face naming/face matching: $r=0.70$; $p<0.001$; voice naming/voice matching: $r=0.78$; $p<0.001$; voice naming/face naming: $r=0.79$; $p<0.001$; voice matching/face matching: $r=0.8$; $p<0.001$. Item-by-item analysis for individual SD patients, using the McNemar test always showed item consistent results except in one SD patient who gave discordant responses for 8/19 items. To be sure, in 12/13 SD patients when a celebrity had been correctly (or wrongly) identified in the '*famous voice-to-name matching*' task, there was a strong trend to obtain correct (or wrong) performances also in the in the corresponding '*famous face-to-name matching*' task. Only in one patient discordant responses were obtained for 8/19 items. For the naming tasks an item-by-item analysis was not carried out due to the low scores (approaching zero) obtained by most patients. The same item-by-item analysis could not be performed for individual AD patients, because scores on the matching tasks were at ceiling in this group.

Looking to the relations between kind of error (i.e. distractor type) in the matching tasks involving face and voice, it was found that in SD patients the error profile was similar in the two matching tasks: in the face-to-name matching task 57% of errors were semantic, 35% were unrelated distractors and in 8% were non famous names. In the voice-name matching test in 55% of cases SD patients chose a semantic distractor, in the 34% of cases they chose an unrelated distractor and in 11% a non-famous name.

4. Discussion

The two aims of our study consisted: (a) in evaluating if famous faces and voice recognition disorders observed in SD and AD patients result from disruption of different cognitive mechanisms and (b) in checking if face and voice recognition disorders detected in SD patients are mainly due to atrophies affecting the RTL.

As for the first aim of our study, both SD and AD patients showed impaired naming of famous faces and voices, even if the magnitude of the deficit was more pronounced in SD patients.

However, in cross modal matching tests involving face and voice recognition, the two disease groups performed very differently. AD patients achieved performances comparable to those of normal subjects whereas performance of SD patients was profoundly impaired.

These findings, which are very similar to those obtained by Snowden et al. (2004) studying famous faces and names in groups of SD and AD patients, could be interpreted as indicative of a different cognitive basis for the impairment in these two diseases. In SD the severe impairment in both naming and matching tasks points to a primary degradation of semantic knowledge. By contrast, in AD, poor naming in the context of preserved recognition would be more in keeping with a lexical problem in name retrieval or with a defect of semantic retrieval or of semantic control.

An apparent counter-argument to the hypothesis of a semantic loss in SD, contrasting with a lexical problem in name retrieval of AD patients, might be that the matching tests are not sensitive enough to detect the relatively subtle semantic impairment of AD patients. It is notable, however, that whereas naming and matching performance were correlated in both SD and controls, in AD they were not, again arguing for a distinct mechanism underlying the naming disorder in AD. On the other hand, the contrast in AD between poor naming and preserved recognition of faces and voices is not at variance with the defect of semantic control hypothesis (Jefferies & Lambon Ralph, 2006; Corbett et al., 2012) because the control defect is influenced by the ease with which relevant

semantic relationships could be identified and distracters rejected. Now, it is obvious that the amount of semantic control is much greater on a naming task (where the semantic/lexical retrieval involves many potential competitors) than on a matching task, where only one semantic competitor is present. The findings from our study appear at first sight at odds with those of Hailstone et al. (2011) which revealed deficits in voice recognition and cross-modal matching in AD as well as SD. These differences might, however, be due to methodological reasons. The aim of the Hailstone study was to characterise the profile of voice processing performance in an unselected AD sample. Thus, patients with elementary perceptual and discrimination deficits were not excluded. By contrast, the aim of the present study was to examine, through naming and matching tasks, the semantic contribution to performance when the potential confounding factor of perceptual impairment had been eliminated. Furthermore, Hailstone et al. (2011) used refined and complex matching tasks in which the subject had to match a stimulus (i.e voice or face) to an array of six to nine names (in the face to name matching test) and to 9 faces and names (in the voice to face-name matching test). The high number of distractors has the advantage of reducing the probability of ceiling effects and of correct responses elicited by chance. At the same time, it places demands on working memory and visual search skills which may be impaired in AD and hence contribute to performance errors. The four-choice version in the present study was designed to minimise such additional cognitive demands. Another potential source of difference between the two studies might lie in differences in overall disease severity, because Corbett et al. (2012) have shown that the nature of semantic impairment in AD changes with disease severity. According to these authors, characteristics of deregulated semantic cognition are exhibited by the mild AD cases, whereas, severe AD patients reproduce features of additional degradation of core semantic representations. Unfortunately, due to different tests used to explore the general cognitive level, namely IQ in the Hailstone et al. (2011) study and MMSE in the current study, this comparison cannot be made. Alternatively, the small numbers of AD patients recruited in the two studies cannot rule out an

intrinsic variability in the performance of AD patients, which are more heterogeneous than SD, in term of cognitive profile.

The second interesting result of our study is relate to the analysis of performance consistency across tasks, because in SD there is a positive relationship between naming and matching task performance whereas in AD this consistency across tasks is not found. This suggests that only in SD patients naming problems are due to a loss of semantic representations, because the same semantic knowledge is requested both for naming and for recognition. Furthermore, the fact that SD patients who failed in the matching tasks tended to choose in more than 50% of cases the semantic distractor reinforces this interpretation . It is interesting to note that a positive correlation between results obtained on the naming and recognition tests with faces and voices is detectable in normal subjects too, whereas in AD the lack of correlation between performance in naming and recognition may have a distinct underlying basis. It cannot be considered as an artefact, due to a ceiling effect on the matching tasks, because positive correlations were found on the same tasks in control subjects. This dissociation could rather reflect the fact that AD patients might have cognitive problems in other domains as well and these might influence performance differentially. The additional cognitive problems might decrease the correlations between naming and recognition because of their intrinsic heterogeneity. This does not necessarily rule out an underlying semantic disorder but allows us to conclude that in AD the cognitive basis of impaired performance in semantic tasks may be explainable in terms of multifaced and intercorrelated cognitive impairment in several domains, rather than as an exclusive semantic breakdown.

As for the second aim of the present study, our data are consistent with previous results showing that face (Snowden et al., 2004; Gainotti, 2007; Josephs, Whitwell, Vemuri, Senjem, Boeve, Knopman et al., 2008) and voice (Van Lanker & Canter, 1982; Gainotti, 2011) recognition disorders are usually due to lesions of the right temporal lobe, because scores obtained by SD patients with right-sided atrophy were systematically lower than those obtained by SD patients with left-sided atrophy. This asymmetry was, however, significant only in the matching tasks (and in

particular in the face-name matching task) because in the naming tasks (and to a lesser extent in tasks based on the voice modality) a floor effect, in association with the low number of SD patients included in our study, did not allow to reach significance. It must also be noticed that in SD patients with a right-sided atrophy the recognition disorder of famous people in the visual (face) and auditory (voice) modality was associated with a conceptual impairment on the PPTT more important in the non-verbal (pictorial) than in the verbal modality. On the contrary, SD patients with a left-sided atrophy were more impaired on the verbal than on the pictorial version of the PPTT. Similar results had been reported by Snowden et al. (2004 and 2012) and are consistent with the Gainotti's (2012 and 2015b) assumption that the format of conceptual and person-specific representations may be mainly non-verbal in the right ATL and verbal in the left ATL.

5. Concluding remarks

In conclusion, the present findings support the hypothesis that different cognitive mechanisms may subsume poor performance in semantic tasks involving famous persons in AD and SD. While a loss of conceptual knowledge for famous persons is responsible of the SD patient profile, characterized by impaired naming and recognition of famous characters, in AD the dissociation between poor naming and normal recognition suggests that conceptual knowledge is mostly spared and the naming problem could be due to a lexical access or to a semantic control defect. Our results also confirm the prevalence of face and voice recognition disorders in SD patients with atrophy mainly affecting the right ATL and show that in these patients face and voice recognition disorders are associated with a conceptual impairment on the PPTT more important in the non-verbal (pictorial) than in the verbal modality. Furthermore, since SD patients with a left-sided atrophy were more impaired on the verbal than on the pictorial version of the PPTT, our data support the hypothesis of a non-verbal format of conceptual and person-specific representations in the right ATL, contrasting with the verbal format of the same representations in the left ATL.

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CONFLICT OF INTEREST

The authors have no competing interests.

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Table 1 Background neuropsychology of patient participants

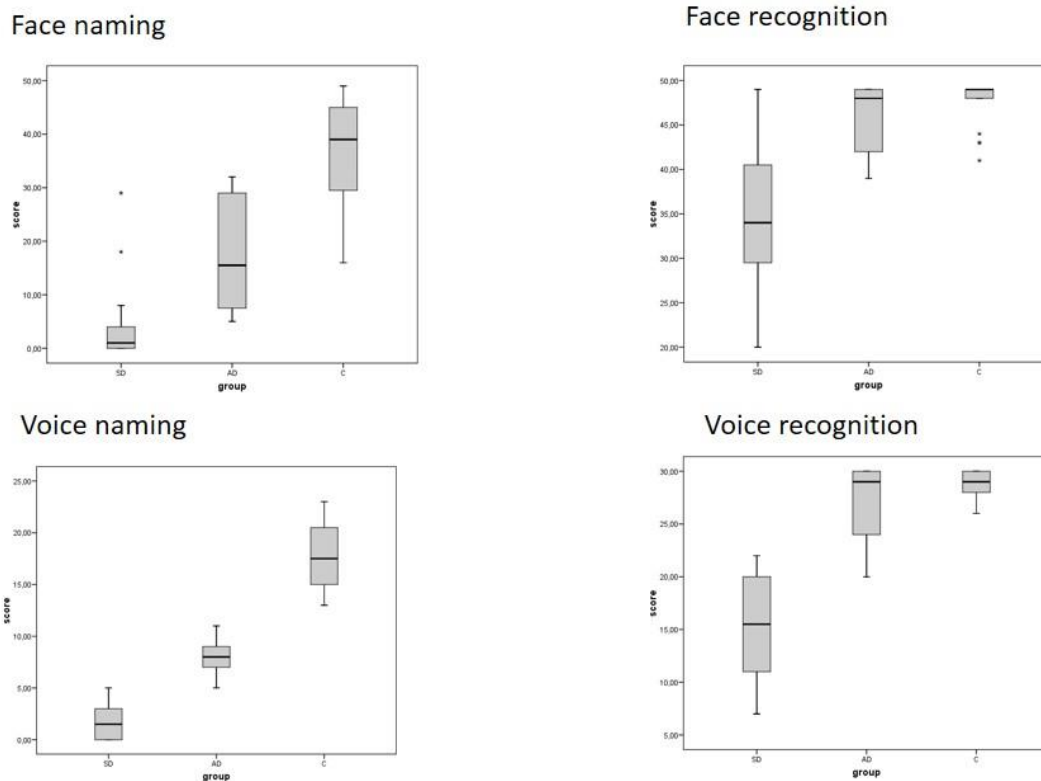
	SD	AD	T test values	Significant differences
<i>General abilities</i>				
MMSE (30)	22.7 (4.8)	23 (3.2)	t= 0.07	ns
Raven's Coloured Progressive Matrices (36)	28.7 (4.3)	22.3 (3.8)	t=1.4	ns
<i>Memory</i>				
Bi-syllabic word span	5 (0.7)	4.6 (0.7)	t=1.4	ns

Corsi blocks	4.9 (0.9)	4.4 (0.8)	t=1.6	ns
Rey AVTL long term(15)	4.1 (1.9)	1.26(1.3)	t=0.1	ns
Rey figure B delayed recall (31)	12.5 (8.4)	5.4 (3.1)	t=1.3	p=0.03
<i>Executive functions</i>				
Luria's Motor Sequences (30)	41.1 (11.9)	39.8 (6.2)	t=0.01	ns
FAS	15.9 (5.8)	18.7 (7.4)	t=1.3	ns
Stroop Test time I	43.3 (8.5)	49.1 (11.6)	t=1.6	ns
Stroop Test time II	117.7 (71.8)	127.3 (34.8)	t=1.6	ns
Stroop Test errors I	0 (0)	0 (0)		
Stroop Test errors II	5.1(4.1)	10 (5.7)	t=0.7	p=0.04
<i>Perceptual-spatial skills</i>				
VOSP				
- Shape detection test (20)	19.7 (0.6)	19.6(0.4)	t=0.08	ns
- Incomplete letters (20)	16.4 (3.8)	16.4 (3.5)	t=0.3	ns
- Silhouettes (30)	6.2 (3.2)	15.7 (3.7)	t=5.7	p<0.001

- Object decision (20)	12.8 (3.4)	13.2 (3.2)	t=0.8	ns
- Dot counting (10)	9.7 (0.6)	9.8 (0.4)	t=0.8	ns
- Number location (10)	7.8 (2.3)	4.9 (2)	t=2.1	p=0.005
- Position discrimination (20)	18.6 (2.2)	18.3 (1.8)	t=0.5	ns
- Cube analysis (10)	9 (1.5)	6.2 (2.4)	t=2.7	p=0,01
<i>Praxis</i>				
Rey figure B copy (31)	27.4 (5.7)	25.1 (3.91)	t=0.2	ns
Ideomotor praxis (right hand)	20 (0)	19.4 (2.1)	t=0.8	ns
Ideomotor praxis (left hand)	20 (0)	19.2 (2.6)	t=1	ns
<i>Language</i>				
Verbal fluency (3 categories 1 minute each)	22.5 (7.2)	28.5 (5.2)	t=2.3	p=0.02
Naming (40)	24 (8.1)	35.3 (2.7)	t=3.4	p=0.001
Reading (40)	39.7 (0.6)	39.8 (0.3)	t=0.2	ns

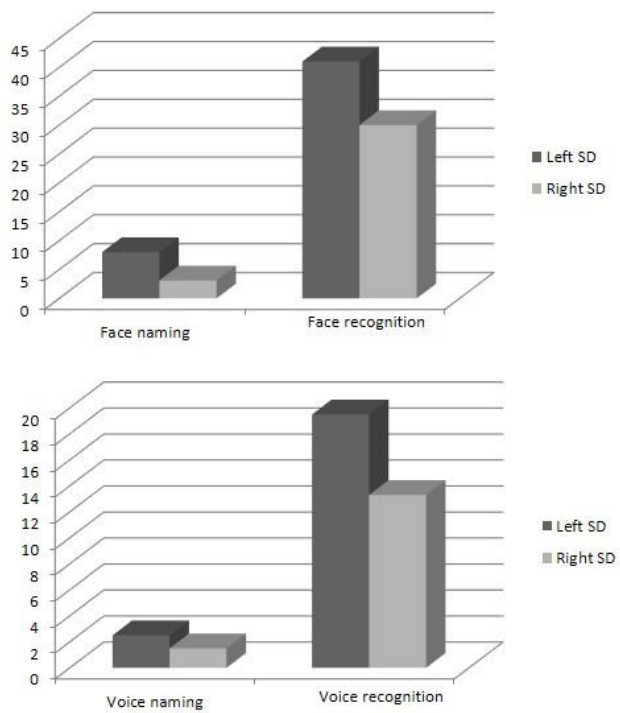
Single word comprehension (40)	36.6 (4.6)	40 (0)	t=4.6	p=0.03
Pyramids and Palm Tree Test (verbal version)	22.3 (2.9)	28.3 (1.5)	t=6.1	p<0.001
Pyramids and Palm Tree Test (visual version)	21.1 (7.2)	27.7 (1.9)	t=3.1	p=0.004

Figure 1: exploration of famous faces and voices: group comparison



SD: semantic dementia; AD: Alzheimer's Disease, C= controls

Figure 2 Comparison between left and right SD



SD: semantic dementia

