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Case JP: Twenty experiments on a person with Congenital Prosopagnosia and inability to experience visual imagery from birth

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Abstract

Congenital Prosopagnosia and inability to experience visual imagery was investigated through administration of neuropsychological, face processing and visual imagery tests on a single patient, JP. JP had no general intellectual impairment, no problems affecting visual and spatial abilities and had little problems affecting overall memory. She was impaired on unfamiliar face matching, perception of eye gaze and famous face recognition, and was in the normal range for facial expression recognition; thus suggesting that JP is of the “prosopagnosic” type. Subjective vividness and spontaneous use of imagery was rated as extremely poor and performance of standard tests of visual imagery yielded mixed results. These findings were interpreted as suggesting that JP adopted different cognitive strategies when performing imagery tasks

Project Declaration

The project was conducted in line with ethical guidelines set by the University of Plymouth. This was achieved through submission of details of the project to the ethics committee, which resulted in ethical clearance. To be specific, ethical guidelines were adhered to in the following way: informed consent was gained by briefing participants of the aims of the study and by explaining the tasks that they would be required to complete. Participants were informed that they had the right to withdraw at any time and were given the opportunity to ask any questions before proceeding. Following experimentation, participants were fully debriefed as to the nature of the investigation and given both the experimenter's and supervisor's contact details should they have any remaining questions, wished to find out more about the study or withdraw their data.

All the data presented in the report was collected by the researcher, unless otherwise stated.

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JP and Control Participants

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Introduction

Face Processing

"Faces are among the most important visual stimuli we perceive, informing us not only about a person's identity, but also about their mood, sex, age and direction of gaze" (Kanwisher & Yovel, 2006, p. 2109). Whether the multiple cognitive components involved in the perception and processing of faces rely on domain-specific or domain-general mechanisms has been the cause of great debate throughout the history of cognitive neuroscience. On one hand, is the view that that the mechanisms engaged by faces are not specific for a particular stimulus class (i.e. faces), but for a particular process that may run on multiple stimulus classes (Kanwisher & Yovel, 2006). On the other hand, the face-specificity hypothesis suggests that humans have specialized cognitive and neural mechanisms dedicated to the perception of faces that are not recruited for the perception of other stimuli.

There is considerable behavioural and neuropsychological evidence in favour of the face-specificity hypothesis. Firstly, Farah (2000) argues that given that face and object recognition are mutually independent systems localised in different brain regions, it stands to reason that they are two separate systems. This statement is further supported by findings that the two systems can be individuated according to their information processing functions (Farah, 2000). Furthermore, Duchaine, Yovel, Butterworth and Nakayama, (2006) found support for the notion that face recognition is carried out by face-specific mechanisms by assessing the recognition abilities of an individual with lifelong face recognition impairments (Edward). Edward was found to have impairments with many types of face processing ability (identity, emotions, gender, attractiveness), yet performance of non-face recognition tests was normal. Edward's face specific impairment indicated to the researchers that he had failed to develop face-specific mechanisms yet was able to develop normal object recognition mechanisms. Thus it was concluded that "face and non-face mechanisms are created, at least in part, by different developmental processes" (Duchaine et al., 2006, p.30).

A functional model of face processing was proposed by Bruce and Young (1986) to account for the multiple cognitive components involved in the perception and processing of faces. The model suggests that there are separate pathways for the processing of facial expressions, the directed visual analysis required to match unfamiliar faces, facial speech, and the recognition of familiar faces. According to the model, structural encoding processes provide descriptions suitable for the analysis of expression and facial speech, directed visual processing and for face recognition units. Recognition of familiar faces involves a match between the products of structural encoding and previously stored structural codes describing the appearance of familiar faces (held in face recognition units); identity-specific semantic codes are then accessed from person identity nodes, and subsequently name codes are retrieved. It is also proposed that the cognitive system plays an active role in deciding whether or not the initial match is sufficiently close to indicate true recognition or merely a 'resemblance'; several other factors are seen as influencing these decisions (Bruce & Young, 1986).

According to Haxby, Hoffman and Ida Gobbini, (2000) face processing is carried out by a domain-specific circuit; Haxby et al. (2000) suggest that face perception is mediated by a distributed neural system that is comprised of multiple, bilateral regions.

A model for the organization of this system was put forward that emphasises a distinction between the representation of invariant and changeable aspects of faces; according to the model the representation of invariant aspects of faces underlies the recognition of individuals, whereas the representation of changeable aspects of faces, such as eye gaze, expression, and lip movement, underlies the perception of information that facilitates social communication. Thus the model is divided into a core system and an extended system. The core system consists of three bilateral regions in the occipitotemporal visual extrastriate cortex, specifically in the inferior occipital gyri, the lateral fusiform gyrus, and the superior temporal sulcus (the fusiform gyrus, specifically the fusiform face area (FFA) is the most robust of the three face-selective regions (Kanwisher et al. 1997; Yovel & Kanwisher 2004)). The model proposes that these regions perform the visual analysis of faces and participate differentially according to the type of face perception, for example, the region in the lateral fusiform gyrus is thought to be involved more in the representation of identity, whereas the region in the superior temporal sulcus is thought to be involved more in the representation of changeable aspects of faces (Haxby et al., 2000). The extended system (in anterior temporal and frontal cortices) is comprised of regions from neural systems for other cognitive functions that can be recruited to work with the regions in the core system to extract meaning from faces. The network model of face perception supports the cognitive model of face processing as it also emphasises a distinction between processes involved in the recognition of identity and those involved in the recognition of expression and speech-related movements of the mouth. Further support for the network model comes from the fact that a variety of anatomical lesions have been described in Prosopagnosia, a “neurological deficit characterized by an inability to recognise faces despite intact intellectual and cognitive function and spared visual processing” (Steeves, Culhama, Duchaine, Pratesi, Valyear, Schindler, Humphrey, Milner & Goodale, 2006, p. 594). For example, although Prosopagnosia is normally associated with lesions in ventral occipitotemporal cortex that are usually bilateral (Damasio, Damasio & Vanhoesen, 1982) there have been cases reported that suggest that Prosopagnosia may result from right occipito-temporal damage (De Renzi, 1986).

Visual Mental Imagery

Visual mental imagery, that is, “the faculty whereby we can revisualise a visual item from memory” (Bartolomeo, 2002, p. 357) is thought to have shared underlying mechanisms with visual perception. There is wide support for this notion of representational commonality, despite there being some evidence that the two processes are dissociated.

Indeed, neuroimaging studies have revealed that the specific pattern of activation during visual imagery is very similar to that found during vision (Gazzangia, 2004). For example, a portion of the FFA, which has been found to be more active during viewing of faces than viewing of scenes, is also more active during imagining faces than imagining scenes; and conversely, a portion of the parahippocampal place area (PPA), which has been found to be more active during viewing of scenes than viewing of faces, has been found to be more active during imagining scenes than imagining faces (O’Craven & Kanwisher, 2000). Further evidence for this representational commonality comes from patients whose imagery and perception deficits affect the same domain (Behrmann, Moscovitch & Winocur, 1994). Farah (1984) reviewed the literature on visual agnosias and visual mental imagery and found that of 28 cases of visual agnosia, half of the patients had similar visual and visual imagery deficits. It is important to note that of the remaining patients, six were not tested for visual imagery and three that had seemingly intact visual imagery, underwent no objective tests (e.g. drawing from memory) (Gazzainga, 2004). Thus there is considerable neuropsychological and behavioural evidence to suggest that the parts of the occipitotemporal areas engaged by the different visual attributes and object classes are shared by visual imagery and vision.

To account for cases that do not evince a parallelism between visual imagery and vision, a shared-systems framework has been proposed (Gazzaniga, 2004). This framework assumes that visual processes such as object recognition and identification rely (in part) on low and mid level visual processes that are not required by visual imagery, and that visual imagery involves a separate, attention-demanding process, needed for imagery, but not for visual perception and object recognition. "It is these intact, higher level processes that…access the stored representations" (Behrmann et al., 1994, p.1084). The dissociation between the two processes is explained as damage to either the low and mid-level processes or the higher-level process, depending on whether the patient evinces a visual or imagery deficit.

Prosopagnosia

Prosopagnosia, i.e. inability to recognise faces, is not a single functional entity but a family of disorders (Fox, Iaria & Barton, 2008). The disorder can be divided into two broad subtypes: Apperceptive and Associative (De Renzi, Faglioni, Grossi, & Nichelli, 1991). In Apperceptive Prosopagnosia, the ability to *perceive* faces is manifestly impaired (this would suggest a problem at the structural encoding stage of Bruce & Young's (1986) model); "the patient is unable to form an accurate perceptual representation of the structure of a viewed face" (Fox et al, 2008, p. 996). Whereas in Associative Prosopagnosia, face perception appears to function normally, but there is a deficit in matching the input to facial memories in order to facilitate recognition. Thus Associative Prosopagnosia represents a loss between face perception process and semantic information. There are three forms of Prosopagnosia: Acquired, Developmental and Congenital. Acquired Prosopagnosia typically results from a neurological insult to the ventral visual cortex in premorbidly normal individuals (Thomas et al., 2009). Developmental Prosopagnosia is a broad term to describe individuals who have sustained brain damage either before birth or in early childhood (Duchaine & Nakayama, 2004; Barton et al., 2003; Farah et al., 2000; de Gelder & Rouw, 2000 as cited in Behrmann & Avidan, 2005). "There appear to be a number of possible routes to Developmental Prosopagnosia; these include genetic conditions, early brain damage, and possibly early visual problems such as infantile cataracts or severe myopia" (Duchaine et al., 2006, p. 6). Finally, Congenital Prosopagnosia refers to the impairment in face processing that is apparent from birth in the absence of any brain damage (Behrmann & Avidan, 2005). There are several alternative explanations of Prosopagnosia: the face-specific explanation, the individuation explanation, the holistic explanation, the configural processing explanation, the curvature explanation and the expertise explanation (see Duchaine et al., 1996 for a full review). For the present purposes however, the focus will be on Congenital Prosopagnosia.

Congenital Prosopagnosia (CP)

Congenital Prosopagnosia is a severely debilitating disorder in which individuals are disproportionately impaired at face processing in the absence of any brain damage or cognitive deficit, and in the absence of a low-level visual processing disorder (Behrmann, Avidan, Marotta & Kimchi, 2005). This impairment is apparent from birth. The disorder can affect the recognition of the very familiar, for example famous faces, friends and family members, and can also affect ability to discriminate between unknown faces. This suggests a perceptual, rather than a memorial basis for the deficit (Behrmann & Avidan, 2005). Across the literature, CP individuals often report using other cues to recognise people, for example voice, clothing, gait, hair, facial hair and mannerisms, however, these are often unreliable and can make facial perception worse (Tiberghien & Clerc, 1986, as cited in Ellis & Young, 1996).

According to Duchaine (2000), CP "is caused by a domain-specific inability to match novel views of faces with previously derived representations" (Duchaine, 2000, p.79). Duchaine (2000) tested this hypothesis i.e. that CP is the result of a domain-general impairment in configural processing, in a man with developmental Prosopagnosia (BC), and found that despite BC's Prosopagnosia, he could perform normally on configural processing tasks. This led to the suggestion that Prosopagnosia can exist without configural processing deficits. Therefore, Duchaine (2000) suggested that "the cause of BC's Prosopagnosia is an impairment in mechanisms specialised for face recognition" (Duchaine, 2000, p. 82). Support for Duchaine (2000) comes from neurological studies, for example those that have investigated the multiple, bilateral regions described in the network model of face perception by Haxby et al. (2000).

Earlier neurological studies revealed that CP is not the result of decreased activation or lack of selectivity in ventral occipito-temporal areas (Hasson, Avidan, Deouell, Bentin & Malach, 2003; Avidan, Hasson, Malach & Behrmann, 2005). Thus the proposition that CP emerges from a disruption in the structural connectivity between the nodes of the distributed face-processing network was presented (Behrmann, Avidan, Gao & Black, 2007; Thomas, Avidan, Humphreys, Jung, Gao & Behrmann, 2009).

Behrmann et al. (2007) conducted detailed morphometric and volumetric analyses of the occipitotemporal cortex in a group of CP individuals and control participants, and found that the CP individuals evinced a larger anterior and posterior middle temporal gyrus and a significantly smaller anterior fusiform gyrus. This lead to the proposition that reduced cortical volume in the anterior regions of the fusiform gyrus may result in failure to compute more fine-grained representations, which can result in poor propagation of signal to other areas of the distributed face-processing network, causing a deficit in face processing such as that evinced in CP. As well as this, the researchers examined the structural connectivity of white matter tracts passing through the fusiform gyrus. They found that the volume and fractional anisotropy (FA) of the two major tracts, namely the inferior fronto-occipital fasciculus (IFOF) and the inferior longitudinal fasciculus (ILF), was smaller and reduced in FA in the CP individuals compared to the control participants. (Whether the reduction in the volume of the anterior fusiform gyrus is a consequence of this and/or of decreased gray matter volume is unclear at this stage (Behrmann et al. (2007)). Taken together, these findings support the hypothesis that CP emerges from a disruption in the structural connectivity between the nodes of the distributed face-processing network. Further support for the structural connectivity disruption hypothesis comes from Thomas et al. (2009). A group of CP individuals and control participants were tested using diffusion tensor imaging and tractography, and the researchers found that the CP individuals evinced “a marked reduction in the structural integrity of the ILF and IFOF bilaterally across all measures” (Thomas et al., 2009, p.29). This lead to the proposition that CP is the result of “a reduction in microstructural integrity of the white-matter tracts along the ventral occipito-temporal cortex” i.e. that impaired face processing is the result of impaired connectivity linking the core regions with the extended regions outlined in the distributed neural system for face perception by Haxby et al., (2000). Not only does this study provide an explanation of the neural basis of CP, it marks a dramatic breakthrough in our understanding of face perception, that is, that white matter fibers in ventral occipto-temporal cortex support the integrated function of the distributed cortical network that subserves normal face processing (Thomas et al., 2009).

An alternative explanation for CP is that it is hereditary (Behrmann & Avidan, 2005; Greuter, Grueter, Bell, Horst, Laskowski, Sperling, Halligan, Ellis & Kennerknecht, 2007; Kennerknecht, Grueter, Welling, Wentzek, Horst, Edwards & Grueter, 2006). According to Kennerknecht et al. (2006), “the segregation pattern of…Prosopagnosia is fully compatible with autosomal dominant inheritance” (p.1617) and as such is best explained by “the mutation of a single gene” (Greuter et al., 2007, p. 734). There are no molecular DNA and linkage studies reported in the literature as of yet, thus, establishing the potential candidate genes and their role in the development of neuropsychological face perception mechanisms remains as the next step in this line of research (Greuter et al., 2007). Such findings would represent a dramatic breakthrough in the way we understand human cognition (Behrmann & Avidan, 2005).

Prosopagnosia and visual imagery

Levine, Warach and Farah (1985) reported a patient (LH) who, following bilateral inferior temporal damage, became prosopagnosic and evinced some degree of object agnosia. Levine et al. (1985) found that the preserved and impaired aspects of visual imagery paralleled LH’s visual abilities; LH was impaired at perceiving object identity from appearance and describing object appearance from memory (especially faces and animals, and colours of objects), but was good at localizing visual stimuli and at describing their locations from memory. Thus patient LH supports the claim that visual mental imagery activates the same stored representations that are engaged by an external stimulus during visual perception. Having said this, there have been reports in which face perception and visual imagery for faces have been found to be dissociated. For example, Madame D (Bartolomeo, Bachoud-Lévi, De Gelder, Denes, Dalla Barba, Brugières & Degos, 1998), a patient with bilateral regions in the extrastriate visual areas, was found to be profoundly impaired in both overt and covert recognition of familiar faces yet had intact ability to consult the configural and computational aspects of faces in mental imagery. According to the shared-systems framework, Madame D’s visual deficit and preserved imagery ability can be explained as damage to the low and mid-level processes and that the processes required by visual imagery are in tact.

In the current study, an experimental battery composed of various tests assessing neuropsychological, face processing and imagery abilities was administered in order to assess the face processing and visual imagery abilities of a single patient, JP, who reported difficulties with facial recognition and visual imagery. First, a battery of tests intended to investigate intellectual, memory and visual perception abilities was administered in order to provide a background context from which to consider any impairments affecting face recognition/inability to use visual mental imagery. Second, a battery of tests intended to investigate visual and verbal memory, memory and recognition of faces and facial expressions and eye gaze perception was carried out to investigate the nature and specificity of JPs Prosopagnosia. Finally, a battery of tests intended to investigate subjective vividness of visual imagery and imagery abilities was administered to investigate JPs reported inability to experience visual imagery. Twenty tests were carried out in total. Given JP's symptoms, it was predicted that JP would perform poorly on tests of face processing and on reports and tests of visual imagery. The report will follow a classical neuropsychological model used to report research with neurological patients/single-cases to allow for the work carried out with JP to be best reflected and reported.

Method

Design

Congenital Prosopagnosia and inability to experience visual imagery was investigated using a single-case experimental design, thus the research focused on a single case, JP, rather than a group of participants.

Participants

Case JP

JP (not actual initials) is a healthy, 45-year-old, right-handed woman. JP contacted Senior Clinical Lecturer in Psychology at the University of Plymouth, Dr. Paul Broks, reporting difficulties with the recognition of faces and with the experience of visual imagery. JP is intelligent, articulate and keen about detail; she claims to have no difficulty reading facial expressions of emotion and feels that she is 'rather good at intuiting other people's states of mind' (Broks, 2008). JP uses specific features to identify people including hairstyle, shape of eyebrows, facial features such as spots and whether one has particularly distinctive features such as "a squashed up nose". Furthermore, clothing, the way people walk and context is also regarded as crucial for identification of familiar people.

Control participants

The performance of JP was compared on relevant measures to that of 20 control participants who were matched for sex, age, handedness and IQ. In the instances where it was not possible to compare the performance of JP with this control group, control data is taken from test norms (see test references for origins of normative data). For each test, the nature of control data is explicitly stated.

Materials and Procedure

The experimental battery was composed of various tests assessing neuropsychological, face processing and imagery abilities. All of the tests administered are described below;

Investigation of neuropsychological abilities

Handedness based on the Edinburgh Handedness Inventory (Oldfield, 1971); WAIS-R IQs (full scale, verbal and performance) predicted from the National Adult Reading Test (NART) (Nelson, 1982); performance of the Wechsler Memory Scale (Third Edition) (Wechsler, 1998) and BIRT Memory and Information Processing Battery (Coughlan, Oddy & Crawford, 2007) and assessments of object perception and space perception made with the Visual and Object Spatial Perception Battery (VOSP) (Warrington, 1991) were investigated. Each of these tests will be described in turn.

<u>Edinburgh Handedness Inventory</u>

An inventory of 22 items with was given. Participants were instructed to indicate their preference in the use of their hands in each of the activities by putting a '+' in the appropriate column (Left or Right).

<u>NART</u>

Participants were required to read aloud down a list of 50 words printed in order of increasing difficulty and the number of errors made was recorded. The words were relatively short in order to avoid the possible adverse effects of stimulus complexity on the reading of participants, and they were all 'irregular' with respect to the common rules of pronunciation in order to minimise the possibility of reading by phonemic decoding rather than word recognition (Nelson, 1991). WAIS-R full scale, verbal and performance IQs were predicted based on the number of errors made.

<u>The Wechsler Memory Scale III</u>

To assess working memory, a series of spatial patterns was visually presented on a three-dimensional board. For the first task, the examiner pointed to a series of blocks at a rate of one block per second, and JP was instructed to point to the same blocks in the same order. For the second task, the examiner pointed to a series of blocks, and JP was required to point to the same blocks but in the reverse order. Responses were recorded. The examiner then read a series of digits aloud and asked JP to say the digits in the same order. Next, the examiner read a series of digits aloud and asked JP to say them in the reverse order. Responses were recorded.

<u>BIRT Memory and Information Processing Battery</u>

A short story comprising 30 ideas was read to JP, who was then required to recall it immediately and after a 40-minute delay. Correctly recalled, vaguely or partially recalled and incorrect/additional ideas were scored accordingly in both the immediate and delayed conditions. JP was then asked to copy a complex a complex two-dimensional figure and to reproduce it from memory immediately afterwards and after a 40-minute delay. Correct, clearly recognisable but rotated or misplaced or misaligned or disproportionate or incomplete or embellished lines and features were scored accordingly in both the immediate and delayed conditions.

<u>Visual and Object Spatial Perception</u>

To establish adequate shape discrimination the Shape Detection Screening Test was administered. The test stimuli were random patterns, on half of which a degraded X was superimposed. JP was required to judge whether the X was present or absent. The

20 stimulus items were preceded by two practice items, which were used to explain the task. A total correct score was recorded, summing the false positive and false negative responses. Secondly, the Silhouettes and Progressive Silhouettes tests were administered to assess object perception. 15 silhouette drawings of animals and 15 silhouette drawings of inanimate objects drawn from the outline of each object rotated through varying degrees around the lateral axis was presented to JP. The silhouettes in each set were arranged in their order of difficulty, ranging from very easy silhouettes to difficult silhouettes. The examiner presented the first animal silhouette and informed JP that it was a drawing of an animal and that the task was to name it. This procedure was repeated for each silhouette. JP was then informed that the next set was all common objects and that the task is the same. The total number of silhouettes named (maximum = 30) was recorded as the score. JP was then presented with a series of ten silhouettes constructed by varying the angle of view from 90 rotation to 0 rotation around the lateral axis. The Progressive Silhouettes test consists of two series, a gun and a trumpet. These particular objects were selected because no normal subject was able to identify the first silhouette in either series, and all normal subjects were able to identify the tenth silhouette in both series (Warrington, 1991). The first silhouette was presented and used to explain the task: ‘This is a silhouette drawing of an object and as I turn the pages it will get progressively easier to identify’. Each page was presented in turn and JP was asked if she was able to identify the object. The number of trials required to identify each object was summed and recorded as the score (maximum trials = 10 + 10). To assess space perception, the Position Discrimination and Number Location tests were administered. Each test stimulus in the Position Discrimination test consisted of two adjacent horizontal squares, one with a black dot (5mm) printed exactly in the centre and one with a black dot just ‘off’ centre. In each of the 20 stimuli the ‘off’ centre dot is in a different position within the square, in ten stimuli the centre dot is in the left square and in ten in the right square. JP was instructed that the task is to point to the dot that is in the centre and the number of correct choices was recorded. The ten test stimuli in the Number Location test consisted of two squares (62mm · 62mm), one above the other with a small gap between them. The top square contains randomly placed numbers (1–9) and the bottom square a single black dot corresponding to the position of one of the numbers. The position of the dot is different in each of the stimulus cards and there are four different number arrays. JP was informed that the task is to identify the number that corresponds with the position of the dot on each trial. The ten test cards were presented and the number selected was noted and the total number of correct responses was recorded (maximum = 10) and used as a score.

Investigation of face processing abilities

Total, visual and verbal memory, forgetting, and visual-verbal and recall-recognition discrepancies based on the Doors and People Test (Baddeley, Emslie, & Nimmo-Smith, 1994); unfamiliar face matching based on the Benton Face Discrimination Test (Benton, Hamsher, Varney & Spreen, 1983); perception of eye gaze direction based on Perrett’s Gaze Direction Test (Young, Aggleton, Hellawell, Johnson, Broks, & Hanley, 1995); ability to recognise, name, and provide identifying information from photographs of faces based on the Famous Faces Test (Greene & Hodges, 1996); and facial expression recognition based on the Facial Expression and Emotion Stimulus Test

(FEEST) (Young, Perrett, Calder, Sprengelmeyer & Ekman, 2002) was investigated. Each of these tests will be described in turn.

Doors and People Test

The full battery of tests were administered in the following order: the People Test (to assess immediate verbal recall); the Doors Test (to assess visual recognition); the People Test (to assess delayed verbal recall); the Shapes Test (to assess immediate visual recall); the Names Test (to assess verbal recognition); and the Shapes Test (to assess delayed visual recall). The stimuli in the People Test comprised three high-frequency and one low-frequency forename/surname pairs of differing syllabic length. Each name was paired with an occupation and presented as a caption to a coloured photograph. JP was instructed to learn the names of the four people and informed that she should concentrate on remembering the forename and surname. The stimuli in the Doors Test were coloured photographs of doors, from different types of buildings (e.g. houses, garages, sheds, barns, churches, public buildings), varying in country of origin, age and condition. There were 27 ‘target’ doors and 81 ‘distractors’. The target doors were mounted singly on a grey background. For the recognition set, each target was presented with three distractors in a 2 x 2 matrix, with the position of the target door in the array counterbalanced. Three of the target items were for practice and the remaining targets were in two sets of twelve, an easy set (A) and a harder set (B). The targets in each set were presented in random order and tested in a different order. The recognition test for the easier set was given before the harder test began. The examiner presented the 12 photographs from set A at 3-second intervals, giving a verbal label with each. JP was then asked to pick the target door from the recognition set (selection of four doors; the target and three distractors). The same procedure was followed for Set B. JP was then asked to recall the names of the four people presented previously in order to obtain a forgetting score. The stimuli for the Shape test were four line drawings of crosses. Each drawing was presented in black ink on a rectangular white card. The four cross shapes were potent, pommee, quadrate and Celtic, thus were varied systematically on three dimensions: overall shape (elongated or square), presence of features at the end of the arms, and presence of a feature at the intersection of the arms. The examiner presented the four drawings and informed JP that the task is to copy it. Three recall trials then followed. Scores for the three trials were taken and combined. The stimuli in the Names Test were forename/surname pairs. There were 27 ‘target’ names and 81 distractors. The target names were mounted singly on white card. For the recognition test the targets were each presented with three distractors (all of which shared the same forename) in a vertical list with the position of the target in the list counterbalanced. Three of the target items were for practice and had male forenames; the remaining targets comprised two sets of twelve, an easier set (A) which had female forenames, and a harder set (B) which had male forenames. The recognition test for the easier set was given prior to the harder set. Presentation order of targets was randomised and a different random order was used in the recognition test. Set A was presented to JP at 3-second intervals and JP was instructed to read each one as it was presented. The same procedure for presentation and testing was administered for Set B. Correct responses were recorded to provide a score. Finally, JP was instructed to draw as many of the shapes as she could remember from the previous Shapes Test, to assess delayed visual recall.

Benton Face Discrimination Test

The Benton Face Discrimination Test was given to assess unfamiliar face matching. In this test, JP was required to choose which of six photographs of unfamiliar faces are pictures of the same person as a simultaneously presented target face photograph. 22 items were given. The test included items involving choice of identical photographs, as well as transformations of orientation or lighting. Responses were recorded and pooled to give an overall total. Advantages of the Benton test are that it is well-standardized and quite demanding (Broks, et al., 1998).

Perrett's Gaze Direction Test

A forced-choice task was used to assess JP's ability to determine eye gaze direction. Pairs of photographs of the same person's face were presented alongside each other. For one-third of the pairs (six trials) both pictures were full-face photographs, for one-third (six trials) they were both facing 20 ° to the left, and for the remaining third of pairs (six trials) both faces were facing 20 ° to the right. In each pair, the eyes of the target face were oriented directly toward JP, and the non-target face was looking away to the left or right by 5 °, 10 ° or 20 °. The combination of six directions of gaze for the non-target faces (left and right directions of gaze at 5 °, 10 and 20 °) and three possible head orientations for both members of each pair (full-face, 20 ° left or 20 ° right) produced a total of 18 trials. On each of these, JP was asked to choose the photograph of the face that was looking directly toward her.

Famous Faces Test

Identification of familiar faces was assessed using 70 highly familiar faces (famous people). For each face, JP was asked to recognize, name and provide identifying information from photographs of faces. The measures of performance involved the number of highly familiar faces recognized as familiar, the number named correctly and the level of correct detail given (1 point was given for vague but correct detail; 2 points were given when a detailed account of the famous person was provided).

Facial Expression and Emotion Stimulus Test (FEEST)

Recognition of facial expressions of emotion was examined using the Ekman 60 Faces test and the Emotion Hexagon test. Both tests were run from software on FEEST CDROM. In the Ekman 60 Faces test, JP viewed 60 photographs faces taken from the Ekman and Friesen (1976) series (10 faces were taken from this series; 6 female and 4 male) one at a time. The face displayed one of the six basic emotions – anger, disgust, fear, happiness, sadness, and surprise, and was presented for 5 seconds. This was followed by a blank screen. JP was then required to decide which of the emotion names (anger, disgust, fear, happiness, sadness, and surprise) best described the facial expression shown by clicking on the appropriate on-screen button with the mouse. There were 6 practice trials followed by 60 randomised test trials (10 for each of the six emotions), leading to an accuracy score out of a possible maximum of 60 correct choices. The Emotion Hexagon test was administered in order to test facial expression recognition with stimuli of graded difficulty. 30 computer-manipulated images of faces from the Ekman and Friesen (1976) series were used to test recognition of basic emotions (anger, disgust, fear, happiness, sadness, and surprise). The morphed images created examples of emotions that lie close to or more distant from the prototype expression. The computer software on the CDROM presented the morphed images in random order for 5 seconds each across one practice and 5 test blocks of 30 trials each. A blank screen followed the image and JP was asked to decide which of the emotion names (happiness, sadness, surprise, disgust, anger, and fear) best described the facial expression shown. Responses were recorded based on mouse clicks to on-screen buttons, and converted to scores out of a maximum of 120 for overall performance and scores out of 20 for recognition of the six emotions.

Investigation of imagery abilities
Subjective vividness of visual imagery based on the Vividness of Visual Imagery Questionnaire (VVIQ) (Marks, 1973); spontaneous use of imagery based on the Spontaneous Use of Imagery Scale (SUIS) (Reisberg, Pearson & Kosslyn, 2003); subjective vividness of auditory imagery based on the Bucknell Auditory Imagery Vividness Scale *(*Halpern, Zatorre, Bouffard & Johnson, 2004); and auditory imagery control based on the Bucknell Auditory Imagery Control Scale *(*Halpern et al., 2004) was investigated. Moreover, imagery abilities were studied through assessment of responses to high and low imagery questions (Eddy & Glass, 1981); and through use of the Animals Tails Test (Behrmann, Moscovitch & Winocur, 1994); Letter Form Tests *(*Weber & Castleman, 1970); Manikin Test (Ratcliff, 1979); Famous Face Feature Test (Zeman, Della Sala, Torrens, Gountouna, McGonigle & Logie, unpublished manuscript) and Facial Expression Feature Test (Zeman et al., unpublished manuscript). Each of these tests will be described in turn.

Vividness of Visual Imagery Questionnaire (VVIQ)

Participants completed the VVIQ. This is a brief 16-item questionnaire with a test-retest reliability coefficient of 0.74 *(n = 68)*, and a split-half reliability coefficient of 0.85 *(n = 150)* (Marks, 1973). Participants were required to summon an image for each item and rate its vividness along a five-point scale (with a 1 indicating ‘perfectly clear and as vivid as normal vision’, a 2 ‘clear and reasonably vivid’, a 3 ‘moderately clear and vivid’ a 4 ‘vague and dim’ and a 5 ‘no image at all, you only "know" that you are thinking of an object’). Participants completed this once with their eyes open, and once with the eyes closed. Scores were calculated for eyes open, eyes closed and a total score was calculated. A high score on the questionnaire is indicative of little/no use of imagery (maximum score = 160) and a low score is indicative of good use of imagery (minimum score = 32).

Spontaneous Use of Imagery Scale (SUIS)

The SUIS is a brief 12-item questionnaire. The items were designed to measure (self-reported) spontaneous use of imagery, sample items include ‘If I am looking for new furniture in a store, I always visualize what the furniture would look like in particular places in my home’ and ‘When I first hear a friend’s voice, a visual image of him or her almost always come to mind’. Participants were asked to read the items and to ‘indicate the degree to which each is appropriate for them’, using a 1-5 scale (with a 5 indicating ‘completely appropriate’, and a 1 ‘never appropriate’). Thus a low score on this questionnaire is indicative of little/no use of imagery (minimum score = 12) and a high score is indicative of high use of imagery (maximum score = 60).

Bucknell Auditory Imagery Vividness Scale *(*Halpern, Zatorre, Bouffard & Johnson,*2004)*

Participants read 14 items involving imagined auditory stimuli (voices, music, environmental sounds), and rated the vividness of the resulting imagery experience on a scale of one (no image present at all) to seven (as vivid as actual sound). Thus a low score on this questionnaire is indicative of little/no use of imagery (minimum score = 14) and a high score is indicative of good use of imagery (maximum score = 98).

Bucknell Auditory Imagery Control Scale *(*Halpern, Zatorre, Bouffard & Johnson, *2004)*

Participants were required to read 14 items involving imagined auditory stimuli (voices, music, environmental sounds), for example, 'consider driving in a car'. There were 2 statements for each item ('a' and 'b') and participants were required to consider how easily they could change their image of the first sound to that of the second sound. For example, for the sample item: 'consider driving in a car', part 'a' involves considering 'the sound of an upbeat rock song on the radio' and part 'b' involves considering that 'the song is now masked by the sound of the car coming to a screeching halt'. Participants were instructed to "rate how easily you could make this change" on a scale of one (no image present at all) to seven (extremely easy to change the image). A low score indicated little/no use of imagery (minimum score = 14) and a high score indicated good use of imagery (maximum score = 98).

High and Low Imagery Questions

Thirty-six critical sentences were used in a sentence verification task, selected from Eddy and Glass (1981). Eighteen sentences were high-imagery and eighteen were low-imagery. A sentence was rated high in imagery if an image was required to determine whether the sentence was true or false, and a sentence was rated as low in imagery if the sentence did not require an image to determine its truth value (Eddy & Glass, 1981). Nine sentences were high-imagery and true; nine were low-imagery and true; nine were high-imagery and false; and nine were low-imagery and false. The items were block randomised so that the 36 sentences were in 9 blocks of 4 items each. Each block had one high-imagery true sentence, one low-imagery true sentence, one high-imagery false sentence, and one low-imagery false sentence. The sentences appeared consecutively on a screen and a timer started simultaneously with each sentence. JP was instructed to hit the key on the left of the keyboard marked" false" with a finger from her left hand when she thought the sentence to be false, and to hit the key on the right of the keyboard marked "true" with a finger from her right hand when she thought the sentence to be truthful. Pressing the keys terminated the timer. Responses and reaction times were recorded.

Animals Tails Test

JP was required to make judgements about whether animals have a long or short tail relative to its overall body length. A list of 40 animals was read aloud by the examiner and JP was asked to rate whether the animal in question has a long or short tail relative to their body size. Responses were recorded and used as a score and JP was timed.

Letter Form Tests

JP completed two forms of this test; the first requiring judgements about whether letters have curved parts, and the second, requiring judgements about whether letters extend ‘below an imaginary line’. The test stimuli in the first task were the 26 letters of the alphabet. The letters appeared in lower case, consecutively, and in random order. JP was required to rate whether “the upper case letter corresponding to each of the lower case letters has curved line segments”. Two practise items were used to explain the task. JP was asked to perform the task as accurately and as quickly as possible using mental images of the letters. The stimuli in the second task were the 26 letters of the alphabet. The letters appeared in upper case, consecutively, and in random order. JP was required to rate whether “the lower case letter corresponding to each of the upper case letters extends above or below imaginary lines”. Two practise items were used to explain the task. JP was asked to perform the task as accurately and as quickly as possible using mental images of the letters.

Manikin Test

This task required mental rotation in space. The test stimuli was 32 pictures of manikins in four different orientations: A, B, C and D (A = facing forward in the anatomical position; B = facing away in the anatomical position; C = facing forward and upside down; D = facing away and upside down). For each orientation, a black disc marked the right hand in half of the stimuli and the left hand in the other half. Stimuli were presented in a random order. JP was required to say, on each trail, which of the manikin’s hands was marked with the black disc. At the beginning of the test, JP was shown a manikin in orientation A (but without either hand being marked) and her attention was drawn to the hair, ears and face. JP was informed that she would see a series of manikins, and that the manikin might be facing her (in which case the face will be visible), or might have his back to her (in which case the hair and ears will be visible); and that the manikin might be the right way up or upside down. JP was then informed that one of the hands of each figure would be marked with a black disc and that she was to say “right” or “left” as soon as she had decided which hand was marked. Two practice trials were given, any errors corrected, and the task explained again if necessary. To ensure that JP used the words “right” and “left” appropriately, 24 additional trials were given in which the stimuli were simply pairs of circles, one white and one black, with a vertical line between them. JP was required to say “right” or “left” depending on the side on which the black circle appeared. Twelve of these trials were given before the main test and twelve afterwards. All responses were recorded.

Famous Face Feature Test

This task required JP to make judgments about visual details of 12 familiar faces, firstly, using imagery, and secondly, with a photograph in view. To begin with, JP was asked for some identifying information for each of the 12 familiar faces to establish familiarity. After asking for this information, the examiner asked six questions about each of the familiar faces, all of which required a yes/no response. Responses and the total length of time taken to answer the questions were recorded. Subsequently, the same questions were asked but with a photograph of the famous person in view**.** Responses and the total length of time taken to answer the questions were recorded.

Facial Expression Feature Test

This task required JP to make judgments about visual features associated with six facial expressions of basic emotions (sadness, surprise, happiness, anger, disgust and fear). The examiner asked JP six questions about the visual features associated with each of the six facial expressions, all of which required a yes/no response. Responses and the total length of time taken to answer the questions were recorded.

Results

Background Neuropsychological Information

Background neuropsychological information for JP is given in Table 1. This shows handedness, estimated premorbid IQs, performance of the Wechsler Memory Scale (Third Edition) and BIRT Memory and Information Processing Battery and assessments of shape, object and space perception. The performance of JP was compared on relevant measures with the performance of 20 control participants aged 38–55 years. This control group was well matched to JP on sex (female), handedness (JP = 22/22 right-handed; control mean = 21.25 right-handed, SD = 1.59), age (JP = 45 years; control mean = 46.25 years, SD = 5.08), and on predicted IQ (based on the NART) (JP = 115; control mean = 109.90, SD = 5.18). Control data for handedness and IQ is taken from this group. Control data for the other tests are taken from the test's norms (see test references for origins of normative data).

Table 1 shows that JP is right handed and of above-average intelligence. Moreover, JP's scores on the two subtests of the Wechsler Memory Scale – III reveal that she has some problems with visually presented information compared to orally presented information. Specifically, although JP had more correct (53.1%) than incorrect (46.9%) responses on the spatial span task of the Wechsler Memory Scale – III (17 out of 32 correct), a binomial test failed to reject the hypothesis that this was due to chance: Exact p = .860 (two tailed). Moreover, although JP had more correct (83.3%) than incorrect (16.7%) responses on the digit span task of the Wechsler Memory Scale – III (25 out of 30 correct), a binomial test rejected the hypothesis that this was due to chance: Exact p = .001 (two tailed). The probability that JP scored 25 out of 30 correct on the Digit Span Task simply due to chance is less than .01%. Further, it can be seen that JP's performance of the BIRT Memory and Information Processing Battery and Visual and Object Space Perception Battery was in the normal range.

Table 1 *Background neuropsychological information for JP, and means and SDs for control subjects of comparable sex, handedness age and IQ.*

	JP	Controls	
		Mean	SD
Handedness test			
Edinburgh Handedness Inventory (/22)	22 (right handed)	21.25 (right handed)	1.59
WAIS-R Full Scale IQ	115	109.90	5.18
WAIS-R Verbal IQ	113	108.25	4.82
WAIS-R Performance IQ	114	109.35	4.70
Wechsler Memory Scale – III			
Spatial Span (/32)	17	NT	NT
Digit Span (/30)	25	NT	NT
BIRT Memory and Information Processing Battery			
Story recall:			
Immediate (/60)	25	29.2	9.1
Delayed (/60)	19	25.3	9.0
Retained %	76	86.5	15.9
Figure recall			
Copy %	100	96.1	5.8
Immediate %	73.75	77.2	15.0
Delayed %	56.25	64.2	25.2
Retained %	76.27	82.1	28.4
VOSP: Shape Detection Screening Test (/20)	20	19.92	0.33
VOSP: Object perception			
Silhouettes (/30)	23	23.1	4.1
Progressive Silhouettes (/20)	13	9.8	2.4
VOSP: Space perception			
Position Discrimination (/20)	20	19.7	0.8
Number Location (/10)	10	9.4	1.1

NT indicates not tested with this task

Face processing abilities

JP’s face processing abilities are summarized in Table 2. This shows total, visual and verbal memory, forgetting, and visual-verbal and recall-recognition discrepancies, unfamiliar face matching, perception of eye gaze direction, ability to recognise, name, and provide identifying information from photographs of famous faces and facial expression recognition. Control data for unfamiliar face matching and ability to recognise, name, and provide identifying information from photographs of faces is taken from the group of 20 control participants. Control data for total, visual and verbal memory, forgetting, and visual-verbal and recall-recognition discrepancies, perception of eye gaze direction and facial expression recognition is taken from test norms (see test references for origins of normative data).

Table 2 shows that JP was within the normal range in terms of overall episodic memory performance. However, when this score is broken down, it can be see that JP is below the range of normal control scores on visual memory tests ($z = -2.18$, $P < 0.05$) and slightly above-average on verbal memory tests (0.39 SDs above control mean). Further, it can be seen that JP’s visual-verbal discrepancy is below the normal range ($z = -2.24$, $P < 0.05$). Moreover, Table 2 shows that JP’s accuracy score on the Benton Face Discrimination Test was severely impaired (23 out of 54 correct, $z = -2.56$, $P < 0.01$) and that JP was impaired on the Perrett’s Gaze Direction Test (15 out of 18 correct, $z = -2.19$, $P < 0.05$). JP made no errors when the gaze direction of the non-target face deviated by 10° (six out of six correct) and 20° (six out of six correct), but she performed less well with 5° deviations (three out of six correct). Table 2 further shows that JP’s score on the Famous Faces Test was significantly impaired (45 out of a possible 280, $z = -3.83$, $P < 0.001$). JP was significantly impaired at famous face recognition (19 out of 70 identified, $z = -4.49$, $P < 0.001$), at naming them (12 out of 70 named, $z = -2.91$, $P < 0.01$) and at giving identifying information for them (14 out of 140, $z = -3.21$, $P < 0.001$). Finally, it can be seen that JP’s performance of the Ekman 60 Faces test and Emotion Hexagon Test was within the normal range.

Table 2 *JP's performance of face processing tasks, and means and SDs for control subjects of comparable sex, handedness age and IQ.*

	JP	Controls	
		Mean	SD
Doors and People			
Total Memory	6	9.2	3.2
Combined Visual	4*	10.1	2.8
Combined Verbal	10	8.7	3.3
Overall Forgetting	4	9.4	3.3
Visual-Verbal Discrepancy	7*	10.8	1.7
Recall-Recognition Discrepancy	10	9.0	1.7
Benton Face Discrimination Test (/54)	23**	39.20	6.32
Perrett's Gaze Direction Test (/18)	15*	16.95	0.89
Famous Faces Test			
Recognized as "famous" (/70)	19***	48.65	6.60
Named (/70)	12**	31.70	6.78
Identifying Information (/140)	14***	61.05	14.65
Total (/280)	45***	141.30	25.13
Facial Expression of Emotions			
Ekman 60 Faces Test			
Anger (/10)	8	7.86	1.90
Disgust (/10)	10	8.59	1.62
Fear (/10)	6	7.19	2.03
Happiness (/10)	10	9.87	0.42
Sadness (/10)	9	8.33	1.66
Surprise (/10)	10	8.55	1.44
Total (/60)	53	50.64	5.04
Emotion Hexagon Test			
Anger (/20)	19	17.84	2.80
Disgust (/20)	20	18.01	3.65
Fear (/20)	20	16.56	3.76
Happiness (/20)	20	19.74	0.80
Sadness (/20)	20	18.38	3.42
Surprise (/20)	17	17.69	2.16
Total (/120)	116	107.97	9.51

Asterisked scores are significantly impaired in comparison to the performance of controls: *z> 1.65, P<0.05; **z> 2.33, P<0.01; ***z> 3.10, P<0.001.

Imagery abilities

JP's imagery abilities are summarized in Table 3. This shows subjective vividness of visual imagery, spontaneous use of imagery, subjective vividness of auditory imagery, auditory imagery control and imagery abilities, based on responses to High and Low Imagery Questions and performance of the Animals Tails Test, Letter Form Tests, Manikin Test, Famous Face Feature Test and Facial Expression Feature Test. Control data for subjective vividness of visual imagery, spontaneous use of imagery, subjective vividness of auditory imagery and auditory imagery control is taken from the group of 20 control participants.

Table 3 *Visual Imagery Abilities of JP, and means and SDs for control subjects of comparable sex, handedness age and IQ.*

	JP	Controls	
		Mean	SD
Subjective vividness of Visual Imagery (VVIQ)	134**	72.90	21.28
Spontaneous Use of Imagery (SUIS)	14**	41.00	9.50
Subjective vividness of Auditory Imagery	55	59.95	12.76
Auditory Imagery Control	61	70.55	15.34
Imagery Tests			
High and Low Imagery Questions			
High Imagery (/18)	8	NT	NT
Low Imagery (/18)	8	NT	NT
Animals Tails Test (/40)	32	NT	NT
Letter Form Test			
Curved line segments (/26)	26	NT	NT
Extend above/below imaginary lines (/26)	25	NT	NT
Manikin Test (/32)	19	NT	NT
Famous face feature Test I (/48)	29	NT	NT
With photograph in view II (/48)	34	NT	NT
Facial Expression feature Test (/36)	17	NT	NT

Asterisked scores are significantly impaired in comparison to the performance of controls: *z> 1.65, P<0.05; **z> 2.33, P<0.01; ***z> 3.10, P<0.001.
NT indicates not tested with this task

Table 3 shows that JP's performance of the VVIQ was significantly below that of controls (z = 2.87, P<0.01) and that her performance of the SUIS was also significantly below that of controls (z = 2.87, P<0.01). Further, it can be seen that JP's performance of the Bucknell Auditory Imagery Vividness Scale and Bucknell Auditory Imagery Control Scale was in the normal range.

With regard to imagery abilities, table 3 shows that JP had more incorrect (55.6%) than correct (44.4%) responses to high imagery questions (8 out of 18 correct), a binomial test failed to reject the hypothesis that this was due to chance: Exact p = .815 (two tailed). JP took 1:54 minutes in total to answer all high imagery questions. Further, table 3 shows that JP had more incorrect (55.6%) than correct (44.4%) responses to low imagery questions, again, a binomial test failed to reject the hypothesis that this was due to chance: Exact p = .815 (two tailed). JP took 2:06 minutes in total to answer all low imagery questions. Moreover, table 3 shows that JP had more correct (80%) than incorrect (20%) responses on the Animals Tails Test (32 out of 40 correct), a binomial test rejected the hypothesis that this was due to chance: Exact p = .001 (two tailed). Thus the probability that this result was simply due to chance is less than .01%. JP took 2:45 minutes in total to complete the Animals Tails Test. Table 3 further reveals that JP was unimpaired at the two Letter Form Tests; she preformed at ceiling on the test requiring judgements about whether letters have curved parts (26 out of 26 correct, 100%), a binomial test rejected the hypothesis that this was due to chance: Exact p = .001 (two tailed), and close to ceiling on the test requiring judgements about whether letters extend ‘below an imaginary line’ (25 out of 26 correct, 96.2%), a binomial test rejected the hypothesis that this was due to chance: Exact p = .001 (two tailed). Indeed, the probability that these results were simply due to chance is less than .01%. With regard to the Manikin Test, although JP had more correct (59.4%) than incorrect (40.6%) responses (19 out of 32 correct), a binomial test failed to reject the hypothesis that this was due to chance: Exact p = .377 (two tailed). JP made no errors on the left/right identification task prior to the Manikin Test (12 out of 12 correct, 100%) and after (12 out of 12 correct, 100%). Of the four different orientations (A, B, C and D), JP achieved 5 out of 8 correct in orientation A (manikin facing forward in the anatomical position); 6 out of 8 correct in orientation B (manikin facing away in the anatomical position); 2 out of 8 correct in orientation C (manikin facing forward and upside down); and 6 out of 8 correct in orientation D (manikin facing away and upside down). Finally, with regard to the Famous Face Feature Test, although JP had more correct (60.4%) than incorrect (39.6%) responses (29 out of 48 correct), a binomial test failed to reject the hypothesis that this was due to chance: Exact p = .193 (two tailed). JP took 4:36 minutes in total to complete the Famous Face Feature Test. When a photograph of the famous face was in view, JP had more correct (70.8%) than incorrect (29.2%) responses (34 out of 48 correct), a binomial test rejected the hypothesis that that this was due to chance: Exact p = .006 (two tailed). Thus the probability that this result was simply due to chance is less than 1%. JP took 3:44 minutes in total to complete the Famous Face Feature Test with a photograph in view. Finally, table 3 shows that JP had more incorrect (52.8%) than correct (47.2%) responses on the Facial Expression Feature Test (17 out of 36 correct), a binomial test failed to reject the hypothesis that this was due to chance: Exact p = .868 (two tailed). JP took 3:30 minutes in total to complete the Facial Expression Feature Test. Thus, to summarize, binomial testing rejected the hypotheses that JP’s performance of the Animals Tails Test, Letter Form Tests and Famous Feature test when a photograph of the famous person was in view was due to chance; moreover, it failed to reject the hypotheses that JP’s performance of the sentence verification, Manikin, Famous Face Feature and Facial Expression Feature tests were due to chance.

Discussion

JP is of above-average intelligence, thus there is no ground for thinking that she has any general intellectual impairment. Moreover, she performed within the normal range on the Visual and Object Space Perception Battery, thus indicating that she has no problems affecting visual and spatial abilities. Her performance of the BIRT Memory and Information Processing Battery and overall episodic memory performance on the Doors and People Test was in the normal range, indicating that she has little problems affecting memory. Having said that, JP's performance of the Wechsler Memory Scale – III does indicates that she has a greater capacity to remember and manipulate orally presented information in short-term memory storage compared to visually presented information. Moreover, her performance of the Doors and People Test supports this conclusion, as JP performed significantly below the normal range on visual memory tests compared to verbal memory tests, in which she scored slightly above average. Taken together, this indicates that JP has a minor generalized difficulty with visual recognition. Indeed, JP's above average performance on the verbal memory tests of the Doors and People Test may be explained as overcompensation for poor visual memory.

With regards to specific face processing tests, JP was significantly impaired on unfamiliar face matching, perception of eye gaze and famous face recognition, and was in the normal range for facial expression recognition. Her impaired performance of the Benton Face Discrimination Test indicates that JP has poor perception of the face's physical structure. Moreover, her interpretation of eye gaze direction was defective; taken together, these findings are indicative that JP has a generalized deficit in face perception. Moreover, JP performed very poorly on all aspects of the Famous Faces Test, which is indicative of a selective deficit of face memory. JP's normal performance on the verbal tests of the Doors and People Test (Names and People Tests) confirm that her difficulty in recognizing faces is not due to a generalized loss of semantic information about people, but rather to a problem in accessing knowledge via face perception. Interestingly, although JP performed significantly below the normal range on the Perrett's Gaze Direction Test, she only made errors with 5° deviations. This is similar to the performance pattern of controls, who similarly only made errors with 5° deviations (Young et al., 1995). This similar response pattern suggests that JP may possess some ability to understand gaze. Moreover, JP reported that she had no difficulty reading facial expressions of emotion; indeed her performance of the FEEST supported this claim as JP performed within the normal range on both the Ekman 60 Faces Test and the Emotion Hexagon Test. This suggests that JP is able to read social signals from the face.

Thus, with regard to face processing, the investigation has confirmed the hypothesis that JP would perform poorly on tests of face processing; specifically it has highlighted JP's problems in matching unfamiliar faces, understanding gaze, and in recognition of famous faces. In contrast, ability to understand facial expression was found to be well preserved. It can be said that JP's difficulties are of the "prosopagnosic" type: she has no general intellectual impairment, no problems affecting visual and spatial abilities, has little problems affecting overall memory and could recognize people from their names. However, her face perception and recognition deficits were severe. JP's problems did not affect all face processing tasks, but were selective to certain aspects of face processing; as such, the findings are consistent with the cognitive and network models of face processing as a distinction was found between processes involved in the recognition of identity and those involved in the recognition of expression. JP's Prosopagnosia is of the apperceptive rather than the associative type as face perception did not function normally; her impaired performance of the Benton Face Discrimination Test shows that she has poor perception of the face's physical structure;

her impaired performance of the Perrett's Gaze Direction Test shows that she has poor perception of eye gaze; and her impaired performance of the Famous Faces Test shows that JP is unable to form a percept of famous faces thus is unable to form facial memories.

Furthermore, with regard to visual mental imagery, the investigation has confirmed the hypothesis that JP would perform poorly on reports and tests of visual imagery. Specifically, JP rated her subjective vividness of visual imagery and spontaneous use of imagery as extremely poor; indeed her results of the VVIQ and SUIS were significantly different to control participants. However, JP rated subjective vividness of auditory imagery and auditory imagery control within the normal range. Given the finding that JP has a greater capacity to remember and manipulate orally presented information compared to visually presented information, this finding is consistent with the notion of representational commonality, as a parallelism is evident in JP's deficits.

With regard to the standard tests of visual imagery, it was confirmed that JP's scores on the Animals Tails and Letter Form Tests were not due to chance; in contrast, chance remained as a possible explanation for JP's performance of the mental rotation in space, sentence verification and face imagery tasks. The reasons for JP's seemingly intact ability to judge in her mind's eye a previously encoded animal or letter remain unclear; it may be due to image generation, or it may be that performance was based on propositional processes and tacit knowledge (Pylyshyn, 1973; 2002) of animal body parts and shapes of upper and lower case letters. Moreover, JP's performance of the Manikin Test is indicative of poor use of mental rotation (the left/right identification tasks ensured that poor performance was not due to misuse of the words "right" and "left"); of the four different orientations (A, B, C and D), orientation C (manikin facing forward and upside down) required the largest mental rotation to identify the location of the black disc and orientation B (manikin facing away in the anatomical position) should have required no mental rotation. Indeed, JP correctly identified the location of the black disc in orientation B considerably more than in orientation C, thus suggesting that she was not capable of performing mental rotation. What's more, JP's scores on the sentence verification test are indicative of problems both rating sentences that require imagery to determine its truth value, and at rating sentences that do not require imagery to determine its truth value. Moreover, JP took longer in total to complete the low imagery questions compared to the high imagery questions. High imagery true sentences, high imagery false sentences, low imagery true sentences and low imagery false sentences were randomly intermixed throughout the task, thus represented constant switches of use of and no use of imagery. It is possible that this fact could account for JP's poor accuracy across all conditions of this experiment, because she may have adopted a strategy of attempting mental imagery on every trial, even when this was unnecessary. Alternative explanations of JP's performance of the sentence verification test include that she was fatigued and therefore completing the task as quickly as possible, that she did not understand the nature of the task, or that she found the task discomforting thus was paying little attention; these explanations seem unlikely however, as JP reported that she was keen to establish the nature and specificity of her problems. Rather than risk skewing the results, it seems likely that her performance was the consequence of confused, maladaptive or dysfunctional imagery. Finally, JP performed poorly when making judgments about visual details of familiar faces and about visual features associated with facial expressions of basic emotions. In contrast to patient LH (Levine et al., 1985), who showed a similarity in the pattern of deficit between visual imagery and vision, JP's imagery problems seems to extend beyond imagery for faces. A further complication is that JP was able to recognize facial expressions of emotion when responding to stimulus in the here and now (as shown by her performance of the FEEST), but was unable to make judgments about the visual details of visual features associated with facial expressions of basic emotions when using visual imagery. Farah (1984) distinguished between 'visual imagery generation deficits', causing impairment on measures of visual imagery without impairment on tests of perception; deficits of long-term visual memory, causing parallel impairments of recognition and imagery; and 'inspection process deficits', reflecting a lower level visual disorder impairing description, recognition and imagery (Zeman et al., unpublished manuscript). Although both patient LH and JP provide examples of deficits of long-term visual memory, JP's deficit is rather more complex given her ability to recognize facial expressions of emotion and her apparent inability to perform mental rotation in space and her performance of the sentence verification task. Thus JP appears to have problems with the generation process as well as with long-term visual memory. Testing perception of non-face stimuli would provide a more detailed insight into JP's imagery problem. Testing JP using the famous buildings tests (i.e. the Famous Buildings Test and Graded Buildings Test) would assess whether JP's difficulty in recognizing famous faces is specific to faces. Moreover, this would provide a more detailed account of JP's recognition difficulties and provide expansion to the finding that JP has a minor generalized difficulty with visual recognition.

Thus, with regard to visual imagery, the investigation lends support to the notion that different strategies may be adopted when performing imagery tasks. Indeed Zeman et al., (unpublished manuscript) suggested that a mental imagery strategy or a propositional strategy may be adopted depending on the task and the effectiveness with which each strategy can be used. For example, mental imagery may be useful when tacit knowledge is insufficient and tacit knowledge may be useful when mental imagery is insufficient. Thus, it is likely that JP adopted different strategies to perform the visual imagery tests; when she attempted to use imagery (on the mental rotation in space, sentence verification and face imagery tasks) her task performance was poor, however, when she was able to rely on propositional processes and tacit knowledge her performance was much better. Obtaining control data for the standard tests of visual imagery would allow for clarification of whether JP's adoption of different cognitive strategies was normal, or whether it was the result of her problematic visual imagery.

Moreover, given that the current study represents a single-case experimental design, there is a general concern of external validity. Despite the fact that this problem is mitigated to some extent by the fact that Prosopagnosia and visual imagery deficits have been demonstrated in several other single-case experimental investigations, it remains as a concern; thus, in order to address this issue it would be useful to conduct the investigation on more than one participant reporting difficulties with face recognition and the experience of visual mental imagery.

It has been noted that the findings from the present are consistent with the cognitive and network models of face processing as a distinction was found between processes involved in the recognition of identity and those involved in the recognition of expression. Moreover, the findings have highlighted JP's problems with image generation and with long-term visual memory. To support the findings, it would be useful to investigate JP's face processing and visual imagery using functional magnetic resonance imaging (fMRI) as this would shed some light on whether JP's behavioural impairments can be traced to any possible alteration in neural substrate. Specifically, examining the pattern of fMRI activation in the fusiform gyrus (FFA) and in other ventral occipito-temporal areas, in response to faces, buildings, and other objects would show whether JP exhibits normal or differentiable face and object related fMRI activation patterns relative to control subjects. Moreover, given JP's problems with image generation and with long-term visual memory, examining her brain activation during attempted visual imagery would show whether she has a normal or abnormal pattern of brain activation when generating mental images. Also, to examine the structural connectivity disruption hypothesis, it would be useful to test JP using diffusion tensor imaging and tractography. This would shed some light on the structural integrity of her ILF and IFOF and thus whether her Congenital Prosopagnosia is the result of impaired connectivity linking the core regions with the extended regions of the face processing circuit.

Finally, JP reported that a family member (her Father) has also reported difficulties with the recognition of faces. Given that the familial component in Congenital Prosopagnosia is of great interest and that "studies of the genetic predisposition and mechanism will allow us to start building bridges between behaviour and cortical development" (Behrmann & Avidan, 2005, p. 186), conducting a genetic investigation has the potential to shed some light on Congenital Prosopagnosia; of specific interest is whether it is compatible with autosomal dominant inheritance.

The investigation has acknowledged the effects of impaired face processing and visual imagery on case JP, and has confirmed the hypothesis that JP would perform poorly on tests of face processing and on reports and tests of visual imagery. JP's problems did not affect all face processing tasks, but were selective to certain aspects of face processing; specifically, she impaired on unfamiliar face matching, perception of eye gaze and famous face recognition, and was in the normal range for facial expression recognition. This is of the "prosopagnosic" type given that JP has no general intellectual impairment, no problems affecting visual and spatial abilities, has little problems affecting overall memory and could recognize people from their names. Moreover, with regard to reports of visual imagery, JP rated her subjective vividness of visual imagery and spontaneous use of imagery as extremely poor and her subjective vividness of auditory imagery and auditory imagery control within the normal range. On the standard tests of visual imagery, it is likely that JP adopted different strategies; when she attempted to use imagery (on the mental rotation in space, sentence verification and face imagery tasks) her task performance was poor, however, when she was able to rely on propositional processes and tacit knowledge her performance was much better, as shown by her performance of the Animal Tails and Letter Forms tests. To support the behavioural findings, it has been suggested that JP undergoes fMRI to shed some light on whether her behavioural impairments can be traced to any possible alteration in neural substrate. It would be useful to know whether JP exhibits normal or differentiable face and object related fMRI activation patterns relative to control subjects; moreover, whether she has a normal or abnormal pattern of brain activation when generating mental images. This would shed some light on whether she adopts a different cognitive strategy from controls when performing imagery tasks. Also, to examine the structural connectivity disruption hypothesis, it would be useful to test JP using diffusion tensor imaging and tractography to investigate whether her Congenital Prosopagnosia is the result of impaired connectivity linking the core regions with the extended regions of the face processing circuit. This would not only expand our understanding of the mechanisms underlying Congenital Prosopagnosia, but would also mark a dramatic breakthrough in our understanding of face perception more generally.

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