

Posterior Cortical Atrophy: The Role of Simultanagnosia in Deficits of Face Perception

by

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Abstract

When viewing a face, healthy individuals tend to fixate on upper regions, particularly the eyes, which provide important configural information about the spatial layout of the face. In contrast, individuals with face blindness (prosopagnosia) rely more on local features – particularly the mouth. Presented here is an examination of face perception deficits in individuals with Posterior Cortical Atrophy (PCA). PCA is a rare progressive neurodegenerative disorder that is characterized by atrophy in occipito-parietal and occipito-temporal areas. PCA primarily affects higher visual processing, while memory, reasoning, and insight remain relatively intact. Common among individuals with PCA is simultanagnosia, an inability to perceive more than one object or detail simultaneously. One might consider simultanagnosia the most extreme form of a feature-based approach. In a series of investigations, individuals with PCA and their healthy control participants completed a same/different discrimination task in which images of faces were presented as cue-target pairs. Eye-tracking equipment (Experiment 1) and the newly developed Viewing window paradigm (Experiment 2) were used to investigate scanning patterns when faces were presented in full view, and through a restricted viewing aperture, respectively. In contrast to previous prosopagnosia research, individuals with PCA each produced unique scan paths that focused on one aspect of the face. Individuals with PCA tended to focus on areas of high-contrast but many of these areas were not diagnostically useful, suggesting that they were having difficulty processing the face even at a featural level. These results suggest a role of simultanagnosia in the scan patterns of PCA patients that is not reflective of ‘typical’ prosopagnosia, and instead points to simultanagnosia, sometimes matched with basic perceptual impairments, as a significant contributor to the face perception deficits seen in PCA.

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CHAPTER 1

GENERAL INTRODUCTION

Preamble

Imagine looking through a family photo album, and realizing that you do not recognize the people in the photographs, or finding that the walls in your home change colour when you blink your eyes. These are some of the issues that RB, a 77-year-old female, began experiencing more than 5 years ago, despite an absence of health problems such as stroke or head injury. Preliminary investigations suggested that RB has prominent deficits in recognizing faces and line drawings of objects, along with significant problems seeing multiple objects in an array and a deficit in global processing (e.g., when shown a photo of a camera, RB mistook the lens of the camera for a tunnel). Also, despite being a talented artist, RB cannot copy even simple line drawings. Further, RB has reported that the colours of objects change drastically when she blinks her eyes. Despite these perceptual impairments, RB did not show any significant deficits in memory, reasoning, or visually guided action. This unusual pattern of deficits illustrates one example of a rare neurodegenerative disorder called Posterior Cortical Atrophy (PCA).

Posterior Cortical Atrophy (PCA)

PCA, also referred to as Benson's disease (Benson, Davis, & Snyder, 1988) or the visual variant of Alzheimer's disease (Bokde et al., 2001; Boxer et al. 2003; Rapoport, Schapiro & Horwitz, 2001), is a progressive neurodegenerative disorder that is associated with significant impairments in higher visual processing, while at early stages, memory, reasoning, and insight remain relatively intact (Chan, Crutch, & Warrington, 2001; Charles & Hillis, 2005; Crutch & Warrington, 2007). The initial symptoms of PCA often include problems such as achromatopsia, prosopagnosia, object agnosia, environmental agnosia, alexia, agraphia, left-right disorientation,

optic ataxia, oculomotor apraxia, dressing apraxia, visual neglect, and simultanagnosia (Chan et al. 2001; Charles & Hillis, 2005; Crutch & Warrington, 2007; Giovagnoli et al., 2009; Mendez, Shapira, & Clark, 2007). At later stages, individuals with PCA show impairments in memory, learning, language, and reasoning, and may appear similar to typical Alzheimer's disease (AD). However, it is important to mention that the symptoms of PCA tend to include problems in higher visual processing at earlier stages (e.g., visual agnosia), followed by problems in memory and reasoning at later stages, somewhat opposite to the typical course of AD symptoms (Whitwell et al., 2006).

PCA is characterized by progressive bilateral atrophy in the posterior areas of the brain (e.g., occipito-parietal and occipito-temporal areas), often with a predominance in the right hemisphere (Caine, 2004; Charles & Hillis, 2005; Goethals & Santens, 2001; Kaida, Takeda, Nagata, & Kamakura, 1998; Kirshner & Lavin, 2006; McMonagle, Deering, Berliner, & Kertesz, 2006; Nester, Caine, Fryer, Clarke, & Hodges, 2003; Mendez et al., 2007; Whitwell et al. 2006). Magnetic resonance imaging (MRI) investigations of PCA show evidence of atrophy in area MT (e.g., motion perception) of the inferior occipito-temporal junction (Caine, 2004; Nakachi et al., 2007), right fusiform gyrus, parahippocampal cortex (e.g., encoding and recognition of scenes) (Joubert et al., 2003), occipital poles (e.g., most posterior area of occipital lobes; central blindspot) (Chan et al., 2001), right occipital gyrus (e.g., occipital face area, involved in initial face perception; recognition of facial parts) (Boxer et al., 2003), Brodmann's area 17 (primary visual cortex), 18, and 19 (secondary visual association areas), area 7b and 7m of the posterior parietal cortex (primary sensory strip, somatosensory processing), and Brodmann's area 23 of the posterior cingulate (visuospatial processing, memorization of familiar routes or people, episodic memory) (Caine, 2004). The affected cortical areas largely involve vision, either primary or in

higher processing areas. In all, PCA appears to affect higher visual, and to a certain extent, other sensory association areas.

In addition, research with single-photon emission computed tomography (SPECT) and positron emission tomography (PET) have shown decreased blood flow (hypoperfusion) and reduced blood-glucose metabolism (hypometabolism), bilaterally, in occipito-parietal and occipito-temporal cortices (Mendez, 2001; Giovagnoli et al., 2009; Goethals & Santens, 2001; Mackenzie-Ross et al., 1996; Bokde et al., 2001; Boxer et al., 2003), which includes the lateral occipital cortex (Giovagnoli et al., 2009), primary visual cortex (V1), and sometimes the frontal eye fields (Bokde et al., 2001; Nester et al., 2003).

Despite significant visual impairments, many cases of PCA go undiagnosed, or are misdiagnosed as AD (Caine, 2004; Charles & Hillis, 2005). The earliest symptoms of PCA can masquerade as natural changes in vision due to aging. For example, initial symptoms typically include blurred vision, problems reading from a printed page, and trouble finding the correct key on a keyboard. Most people may take these signs to indicate that a new eyeglass prescription is necessary, which results in many individuals who have PCA going untreated for extended periods of time.

In some cases of PCA, cortical dysfunction eventually occurs in areas of the brain that are involved in memory, reasoning, and language (Chan, Crutch, & Warrington, 2001; Charles & Hillis, 2005; Crutch & Warrington, 2007), and post-mortem examination often reveals neuritic plaques and neurofibrillary tangles that are typical of AD (Renner et al. 2004). When PCA goes undiagnosed, problems in memory and reasoning might be the first symptoms presented to healthcare professionals, and may lead to a diagnosis of suspected AD. Further, most individuals who receive a diagnosis of PCA are considered to have a visual variant of AD, despite

differences in the overall patterns of atrophy between PCA and AD (Boxer et al. 2003; Renner, Burns, Hou, McKeel, Storandt, & Morris, 2004).

Using MRI, Boxer et al. (2003) found significantly less gray matter in the right inferior temporal gyrus of individuals with PCA, compared to individuals with AD. The AD group showed more atrophy in the left hippocampus, right and left perisylvian cortices, left angular gyrus, left middle temporal gyrus, left dorsomedial thalamus, and right hippocampus. In all, the AD group showed more atrophy in left hemisphere medial temporal structures, while the PCA group showed greater atrophy in right hemisphere occipito-temporal and occipito-parietal areas.

Using PET, Bokde et al. (2001) found more hypometabolism in posterior and visual association areas of a visual AD group, compared to a typical AD group. In contrast, the typical AD group showed greater hypometabolism in the frontal regions, where the visual AD group and controls appeared to be largely unaffected. In a similar study, Nester, Caine, Fryer, Clarke, and Hodges (2003) found that PCA cases showed significantly greater reductions in cerebral blood glucose metabolism in the right occipito-parietal area, specifically at the occipito-parietal junction when compared to AD. Hypometabolism in PCA differed most from AD in the right hemisphere, although PCA differed from controls bilaterally in posterior brain regions.

Renner et al. (2004) used MRI, PET, and neuropsychological tests, and found that the cortical atrophy that is associated with AD was also most predominantly associated with PCA. The authors examined 27 cases of atypical dementia where deficits in higher vision were most prominent. Following a battery of neuropsychological tests, and some scans, both post- and pre-mortem, the authors found that most of these patients presented with atrophy similar to that found in AD (e.g., senile plaques, neurofibrillary tangles).

Other causes PCA include corticobasal degeneration, Lewy body disease, Lewy body disease with progressive subcortical gliosis, fatal familial insomnia, and prion diseases (e.g., Cruetzfeld-Jakob's disease).

PCA Subtypes: Dorsal and Ventral

Some cases of PCA fit into one of two subtypes – dorsal-PCA and ventral-PCA. This distinction is used to more accurately describe differential patterns of impairment, along with atrophy that is more concentrated in either occipito-parietal (dorsal) or occipito-temporal (ventral) areas. To date, there are more reports in the literature of dorsal-PCA than ventral-PCA (Chan et al., 2001; Kaida et al., 1998; Nester et al., 2003).

Dorsal-PCA is used to describe cases of PCA where visual impairments such as optic ataxia, gaze apraxia, constructional apraxia, and simultanagnosia are most significant, with a relative absence of problems in visual perception (Goethals & Santens, 2001; Mizuno et al., 1996). These individuals may also show greater atrophy in occipito-parietal areas, compared to occipito-temporal areas. Ventral-PCA refers to individuals who experience predominantly perceptual visual problems such as object agnosia, prosopagnosia, achromatopsia, and alexia without agraphia, while visually guided action appears unaffected. In ventral-PCA atrophy is more likely to be found in occipito-temporal areas than in occipito-parietal areas.

Among 4 cases of predominantly dorsal-PCA, where atrophy occurred mainly within the occipito-parietal cortex, Mackenzie-Ross et al. (1996) found that the initial symptoms most often included optic ataxia, spatial disorientation, problems with tool use, agraphia, and dressing apraxia. Similarly, Goethals and Santens (2001) examined 2 cases of dorsal PCA, and found that symptoms were, at least initially, concentrated on problems such as agraphia, constructional

apraxia, and difficulty with daily tasks, including dressing, setting the table, and manipulating tools.

Cases of predominantly ventral-PCA have been described as characterized by visual problems in recognition of familiar faces (e.g., Joubert et al. 2003; Evans et al. 1995), apperceptive (i.e., inability to visually perceive an object or form) and associative agnosia (i.e., inability to retrieve the identity of an object or form upon visual presentation), finger agnosia, and prosopagnosia (e.g., Giovaglioni et al. 2009). Some cases show more accuracy for primary face perception (i.e., the ability to make discriminations based on faces vs. objects, facial expressions, gender, age), and most showed relative preservation of memory and intellect (e.g., Joubert et al.; Evans et al.; Giovaglioni et al.), as well as visuomotor abilities (e.g., Evans et al.). Also, these cases often showed a focus on individual features more than global attributes when viewing faces (Joubert et al.; Evans et al.), and some were more successful at detecting facial expressions than at identifying faces (e.g., Evans et al.). An MRI scan showed atrophy in the . Most commonly, atrophy was found in temporal, parietal, and lateral occipital lobes (Evans et al.), right fusiform gyrus, and parahippocampal cortex (Joubert et al.), while hypometabolism was found bilaterally in the parietal lobes, left temporoparietal and lateral occipital areas (Giovaglioni et al.).

Two Stream Theory of Vision

The differences in PCA symptoms attributed to different patterns of atrophy (ventral vs. dorsal) fits well with the two-stream theory of vision extended by Milner and Goodale (Milner & Goodale, 1995, 1998, 2006). The two-stream theory proposes that separate visual pathways, the ventral and dorsal streams, have evolved to process visual information into the representations needed for visual perception and visually guided action. Mechanisms in the ventral stream,

which project from primary visual cortex (V1) to the infero-temporal portions of the temporal lobe, are thought to be involved in allocentric, or object-centered, visual processing, which facilitates recognition of stimuli that include faces (Gauthier, 2001; Kanwisher, McDermott, & Chun, 1997; Milner & Goodale 1998, 2006; Tarr & Gauthier, 2000). The dorsal stream, which projects from V1 to the extrastriate cortex, and then the posterior parietal cortex (Milner & Goodale, 1995, 2006), deals with moment-to-moment information about the location and disposition of objects, with respect to the limb being used, in order to mediate the visual control of skilled actions directed at those objects (Crawford, Medendorp, & Marotta, 2004; Culham, 2004; Goodale, 1998; Goodale & Milner, 1992; Goodale & Westwood, 2004; Grill-Spector, 2003; James, Culham, Humphrey, Milner, & Goodale, 2003; James, Humphrey, Gati, Menon, & Goodale, 2002; Milner, 1998; Milner & Goodale, 1995, 2007; Ungerleider & Mishkin, 1982). It should be noted, however, that recent evidence suggests that the dorsal stream itself consists of 3 separate visuomotor pathways (Kravitz, Saleem, Baker, and Mishkin, 2011).

The strongest evidence for Milner and Goodale's (1995) two-stream theory of vision comes from lesion studies where individuals have shown a double dissociation between vision for perception and vision for action. Examples of this double dissociation have been observed through the investigation of two patients in particular, DF and RV, who have damage to the occipitotemporal, and occipitoparietal areas, respectively. DF, who incurred bilateral damage to her temporal lobes, shows deficits in identifying objects, with no impairments in visually guided action. In contrast, RV has bilateral damage to her occipitoparietal areas, and shows deficits opposite to those experienced by DF; RV can accurately name and identify objects, yet is unable to reach out and accurately grasp those very same objects (Milner & Goodale, 1995, 2006).

Face Processing

The processing of faces is thought to be subserved by an extensive neural network that encompasses many of the ventro-medial regions of the brain, particularly in the right hemisphere, from the occipital pole to the temporal pole, throughout the inferotemporal cortex and superior temporal sulcus (Barton, 2008; Fairhall & Ishai, 2007; Farah, Wilson, Drain, & Tanaka, 1998; Gauthier, Tarr, Moylan, Skudlarski, Gore & Anderson, 2000; Rossion et al., 2003). A number of neuroimaging studies of face perception have identified a discrete region in the middle fusiform gyrus, the fusiform face area (FFA), that responds preferentially to faces as compared to assorted common objects (Fairhall & Ishai, 2007; Gauthier et al., 2000b; Kanwisher et al., 1997; Roissson et al., 2003).

Beyond an increased relative activation for faces (see Gauthier et al., 2000b; Kanwisher et al., 1997; Rossion et al., 2003), the FFA is thought to be important for processing both the spatial relations between facial features (Gauthier et al., 2000b), as well as individual facial features (Yovel & Kanwisher, 2004). However, some researchers have suggested that activation in the FFA is not specific to faces and instead is an “expertise” area for recognizing fine distinctions between well-known objects (Gauthier et al., 2000a, 2000b). In other words, it may be that FFA responds more to faces than objects because humans are experts at discriminating between faces. In fact, Gauthier et al. (2000a) found a similar pattern of activation in the FFA when bird experts viewed birds, and when car experts viewed cars.

Located posterior to the FFA, the occipital face area (OFA) sends projections to both the FFA region and the superior temporal sulcus (STS), and is thought to be involved in initial face perception (Fairhall & Ishai, 2007), and discrimination between individual faces (Gauthier et al., 2000b; Yovel & Kanwisher, 2006). However, unlike the FFA, the OFA is thought to be involved

in discriminations that are based on face parts, rather than an overall face (Liu et al. 2003). The STS, an area of the temporal lobe that separates the superior temporal gyrus from the medial temporal gyrus, has been implicated in the detection of gaze direction, and changeable properties of faces, such as facial expression (Gauthier et al., 2000b; Haxby et al. 2000; Yovel & Kanwisher, 2006).

Dual-code View of Face Processing

The dual-code view claims that both featural and configural information are involved in face processing, and that these different types of information are stored separately (see Bartlett & Searcy 1993; Bruce, 1988; Cabeza & Kato, 2000; Farah et al., 1998; Sergent, 1984). The dual-code view is in contrast to the pure holistic view (see Farah, Tanaka, & Drain, 1995; Tanaka & Farah, 1993), which suggests that faces are processed based on only holistic information. Featural representations are those that include details of individual features, such as the shape, or colour, of the eyes. Configural representations relate to first-order organization (i.e., two eyes, above a nose, which is above a mouth), and second-order organization (i.e., distance between the nose and the eyes) (Chaby, Narme, & George, 2011; Diamond & Carey, 1986; Lobmaier, Klaver, Loenneker, Martin, & Mast, 2008; Lobmaier & Mast, 2008).

To study the relative contribution of each type of information in face processing, featural and configural representations are teased apart using Gaussian blur, or by changing the first-order relationships of a face (Lobmaier et al., 2008; Lobmaier & Mast, 2008). Blurring is thought to degrade featural information, and preserve configural information; the features are hidden, but first- and second-order relationships remain intact, and visible. Changing the first-order relationships is thought to remove configural information; features are broken apart and randomly rearranged.

Using methods to isolate featural and configural representations, evidence for the dual-code view exists in imaging, and behavioural research (see Collishaw & Hole, 2000; Lobmaier et al, 2008; Lobmaier & Mast, 2008; Schwaninger, Lobmaier & Collishaw, 2002). Featural representations are associated with left hemisphere temporal lobe structures (Lobmaier et al, 2008; Roisson et al., 2000), while configural representations are associated with right temporal structures (Lobmaier et al, 2008; Roisson et al., 2000). Roission et al (2000) used PET, and found greater activation in the right hemisphere FFA when participants were asked to pay attention to configural properties of faces, while the left FFA showed greater activation when participants were asked to attend to individual features of faces. Lobmaier et al (2008) also found a hemispheric differences using fMRI, where left hemisphere structures (e.g., left fusiform gyrus, parietal lobe, lingual gyrus, precuneus, and the right insula) were more activated for featural processing, and right hemisphere structures (e.g., right middle temporal gyrus) were more activated for configural processing.

In a behavioural study, Schwaninger et al. (2002) found that participants were highly accurate regardless of whether a face was blurred or scrambled, in a face-matching task. However, when faces were both blurred and scrambled, accuracy fell dramatically. Collishaw and Hole (2000) found similar results in a famous/novel face-matching task. Participants were accurate in their responses to upright/scrambled, and inverted/scrambled faces, with no differences found between upright and inverted faces for the scrambled stimuli. In contrast, errors increased significantly when faces were blurred/scrambled, and inverted/blurred. Further, Lobmaier et al. (2008) found that participants were most accurate, and fastest when responding to *scrambled novel* faces, and to *blurred familiar* faces, implying that featural and configural information may be more important for different aspects of face processing. Taken together,

these results implicate the existence of two types of representations of faces, and further suggest that although different, each can be utilized independently to recognize faces.

Prosopagnosia

As discussed above, healthy individuals are believed to process faces based on both featural and configural representations, and show visual scan paths that highlight the eyes, nose and mouth. However, individuals with prosopagnosia, an impairment in face processing, show a different pattern of results. Prosopagnosia is defined as the inability to recognize familiar faces, in the absence of other visual problems or cognitive deficits (Farah et al., 1998). Bilateral damage to occipito-temporal areas is most often associated with prosopagnosia; however, this deficit has also been shown to result from unilateral damage to the right hemisphere (Gauthier et al., 2000b; Le et al. 2003). Some researchers suggest that prosopagnosia is the result of a deficit in configural representations of faces, suggesting that these individuals are forced to rely solely on featural representations (Barton et al., 2006; Bukach et al., 2008; Caldara, Schyns, Mayer, Smith, Gosselin, & Rossion, 2005; de Xivry et al., 2008; Le et al., 2003). Evidence for a reliance on featural representations of faces comes from research with face inversion, manipulations of featural and configural information, eye tracking, and other methods for recording scan paths (e.g., Barton et al., 2006; Bukach et al., 2008; Caldara et al. 2005; de Xivry et al., 2008; Le et al., 2003).

The face inversion effect describes the finding that healthy individuals are slower, and less accurate at recognizing faces that are presented upside down (Yin, 1969), while individuals with prosopagnosia either show no difference, or show improved performance for inverted faces (Busigny, Thomas, & Roisson, 2010). Researchers suggest that inverting a face interferes with configural processing, and forces parts-based, or featural processing approach (Lobmaier et al.,

2008). This evidence helped develop the theory that the deficits seen in prosopagnosia are the result of a deficit in configural processing, and preserved featural processing, because individuals with prosopagnosia would use the same parts-based strategy for both upright, and inverted faces.

SB, an individual with prosopagnosia, was better at making upright-inverted discriminations than at making upright-scrambled discriminations (Le et al. 2003). Overall, compared to healthy participants, SB's response latencies were longer, and his responses were less accurate. SB is profoundly impaired in various aspects of face processing (e.g., discrimination of face vs. non-face, familiar vs. unfamiliar faces, facial expression recognition, and face identification), and has damage to the right fusiform gyrus, and right occipito-parietal cortex, while the left occipito-parietal areas were spared.

Simultanagnosia

Simultanagnosia is a problem experienced by all of the individuals with PCA who are involved with our lab, and this disorder is commonly associated with PCA. Often a component of Bálint syndrome (Rizzo, 1993), simultanagnosia is a rare neuropsychological disorder where patients are unable to attend to, or perceive more than a single object or detail at one time (Luria, 1959; Montoro, Luna & Humphreys, 2010; Moreaud, 2003; Rafal, 2003; Rizzo & Vecera, 2002). and is associated with bilateral damage to the parieto-occipital junction (Rizzo, 1993). Evidence for this problem in attention to, and/or perception of, multiple objects or details is seen in studies where individuals with simultanagnosia show a selective deficit in global perception, which is often seen in the Navon task (Clavagnier, Fruhmann-Berger, Klockgether, Moskau, & Karnath, 2006; Dalrymple, Kingstone & Barton, 2007; Dalrymple, Bischof, Cameron, Barton, & Kingstone, 2009; Dalrymple et al. 2010; Himmelbach, Erb, Klockgether, Moskau, & Karnath,

2009; Huberle, Driver, & Karnath, 2010; Jackson, G.M., Swainson, R., Mort, D., Husain, M., & Jackson, S.R., 2004; Karnath, Ferber, Rorden, & Driver, 2000; Navon, 1977).

The Navon task is thought to manipulate global vs. local processing by asking participants to identify stimuli that consist of large uppercase letters (e.g., H or S) that are composed of smaller letters which are either the same or different than the large letter (i.e., H made of tiny Hs, or H made of tiny Ss). Participants take part in two conditions for this task, where they are asked to identify each stimulus based either on the large letter (global condition) or the small letter (local condition). Individuals with simultanagnosia tend to perform poorly when asked to identify stimuli in the global condition, yet may perform similar to healthy individuals in the local condition.

Similar in name to the distinction of the two stream theory of vision, simultanagnosia is described within two subtypes; dorsal and ventral. Dorsal simultanagnosia, a component of Balint's syndrome, is associated with damage to occipito-parietal regions, bilaterally, and linked to visual impairments such as optic ataxia, gaze apraxia, agraphia, problems navigating the environment (e.g., bump into furniture), as well as perceiving multiple objects, or object components at one time (Duncan et al. 2003; Farah, 2004). Ventral simultanagnosia is associated with left occipito-temporal lesion, and is thought to be a problem of visual awareness of multiple components in an array (Duncan et al. 2003; Farah, 2004). Individuals with dorsal simultanagnosia will often report components of an image, and are unable to grasp the meaning of the overall image. Individuals with ventral simultanagnosia show alexia, on letter-by-letter reading, which is the inability to read a single word. Thus, ventral simultanagnosia is thought to be less obvious, and it is thought that only parts of an object are noticed (Farah, 2004). According to Farah (2004) dorsal simultanagnosia is characterized by piecemeal *perception* of

visual stimuli, while ventral simultanagnosia is characterized by piecemeal *recognition* of visual stimuli. That is, in ventral simultanagnosia, objects and their components may be perceived without awareness, leading to piecemeal processing.

Research has often focused on dorsal simultanagnosia, or refer only to simultanagnosia in a more general way, referring to problems in processing multiple visual stimuli, and problems in global processing. Some suggest that this disorder is caused by problems in the perception of spatial location and spatial relationships of objects or their elements (Dalrymple et al. 2009; Dalrymple et al. 2010; Karnath et al. 2000; Huberle et al. 2010; Montoro et al. 2010), a problem with disengagement from areas of high salience (Carota & Calabrese, 2011), while more recent accounts suggest a restricted window of visual attention as a possible explanation (e.g., Dalrymple et al. 2010; Dalrymple et al. 2011). Over the past decade, studies (e.g., Dalrymple et al. 2009; Dalrymple et al. 2010; Karnath et al. 2000; Huberle et al. 2010; Montoro et al. 2010) have shown that inter-element distance, and overall stimulus size affect response errors in individuals with simultanagnosia. Increased element size, and greater inter-element distance is thought to place greater demands to visual working memory. Montoro et al. (2010) found individuals with simultanagnosia and healthy participants were faster and more accurate when Navon stimuli were small, dense, or connected. Huberle et al. (2010) found similar results when stimuli or inter-element distance was physically smaller, or produced a smaller retinal image when ocular distance was increased.

Although a key characteristic of simultanagnosia is a selective deficit in the identification of global forms (e.g., Navon hierarchical letters), eye tracking studies have shown scan paths that closely trace the shape of these misidentified global forms (Clavagnier et al. 2006; Dalrymple, Kingstone & Barton, 2007; Dalrymple, Bischof, Cameron, Barton, & Kingstone, 2009;

Dalrymple et al. 2010). Clavagnier et al. found that scan paths of two individuals with simultanagnosia, RJ and GH, showed a pattern of fixations that closely matched the global shape of Navon stimuli, with no differences in scan paths for correct vs. incorrect responses. Dalrymple et al. (2007) found a similar result when eye movements were recorded for another individual with simultanagnosia, SL. This research has been taken to indicate that individuals with simultanagnosia lack awareness in global processing (Rizzo & Hurtig, 1987).

Since our initial meeting with RB, 4 additional individuals with PCA (SS, AP, MTB, and PLH) have become involved with our lab. All of these individuals show impairments in higher visual processing, with variability in the impairments experienced by each. For example, RB, in particular, shows a pronounced deficit in face perception, while others, such as AP, are less affected. Importantly, all of the individuals who have taken part in our research show signs of simultanagnosia. Prosopagnosia and simultanagnosia are explained by researchers in terms of local/featural, and global/configural processing (e.g., Farah, 1995; Lobmaier et al. 2008; Dalrymple et al. 2007), although there are differences in the behavioural results for each disorder.

The present study seeks to explore the nature of face perception deficits in 5 individuals with PCA, in an effort to determine the underlying cause of these impairments. Additionally, the present study is meant to further explore the symptoms associated with a rare, and likely under diagnosed disorder, PCA. Investigation of face perception deficits in 5 individuals with PCA may help to answer questions regarding the nature of the symptoms that characterize PCA, as well as those that relate to face processing. To meet these goals, a face-matching task was completed across two experiments. The first experiment utilized eye tracking, because previous research has used this technology to examine visual scanpaths that are associated with face

perception. The second experiment involved restricting vision to a small portion of an image at one time with the newly developed *Viewing Window* task. Restricting vision is thought encourage a serial, or part-by-part processing strategy, which may have different effects on scan paths in PCA and healthy populations.

CHAPTER 2

Eye tracking: Open-view task

Experiment 1 utilized an eye tracker to record participants' scan paths in a face-matching task. Eye movement studies have shown that when healthy individuals view faces, they tend to look at the area between the eyes, at the top of the nose, followed by the nose, the mouth, and external features (e.g., hairline, jawline, outer area of cheeks) (Barton, 2008; Caine, 2004; Le, Raufaste, & Demonet, 2003; Stephan & Caine, 2009). Importantly, ocular fixations that fall within the region at the top of the nose are thought to signify attention to the second-order configural relationships (e.g., distance between the eyes and the nose). Many researchers suggest that this pattern reflects an emphasis on configural representations in face identification, because second-order configural relationships differ between individual faces, and are thought to be important in face identification (Roisson et al., 2000). Within these regions, there are typically more fixations, and longer fixation durations (Barton, 2008; Barton, Radcliffe, Cherkasova, Edelman, & Intriligator, 2006; Le et al., 2003; Stephan & Caine, 2009).

In contrast, individuals with prosopagnosia are typically found to spend less time looking at the face regions that represent second-order configural information, and show a greater focus on the lower region (e.g., mouth), external features (e.g., jawline, hair), or individual features (e.g., make fixations to each eye, instead of top of nose) (Barton et al., 2006; Bukach et al., 2008; Caldara et al. 2005; de Xivry et al., 2008; Le et al., 2003). It has been suggested that the

observed differences between healthy individuals and individuals with prosopagnosia are due to a parts-based strategy, and reflect a deficit in configural processing. That is, problems in face perception are believed to be the result of an over reliance on featural representations.

de Xivry et al. (2008) found that patient PS, an individual with prosopagnosia, spent the most time looking at the mouth region of personally familiar faces, followed by the eyes, with the least amount of time spent looking at the top of the nose. PS's fixations suggested that she viewed each eye individually. In contrast, the healthy participant spent the most time looking at the area at the top of the nose, between the eyes, with the least amount of time spent looking at the mouth region. PS has damage to occipito-temporal areas, with a spared FFA. Overall, PS produced longer response latencies, more fixations, and was less accurate than a healthy participant. Le et al. (2003) found similar results with eye tracking in a study that compared scanpaths of healthy individuals to patient SB. SB's initial fixations were located along the hairline, and forehead, followed by fixations located mainly within the mouth region.

Stephan and Caine (2009) presented the case of a prosopagnosic patient, SC, who produced initial fixations to the eye region in a famous vs. novel face discrimination task, but went on to show abnormal patterns beyond the initial fixation. SC has damage to his left anterior temporal lobe, left parietal lobe, left occipital lobe, as well as the posterior area of the right posterior occipital lobe. The healthy control participants produced initial fixations to the eyes, followed by fixations to the nose, mouth, and, finally, the forehead. However, SC's fixations moved from the eye region almost directly to the mouth region, and he spent more time fixating external, compared to internal areas.

Experiment 1

Experiment 1 was designed to replicate previous findings in healthy individuals, and to determine whether or not individuals with PCA, who experience multiple perceptual and/or visuomotor impairments, produce scan paths that differ significantly from healthy control participants when viewing faces. More specifically, because some individuals in the PCA group show face perception deficits, and all are reported to experience simultanagnosia, Experiment 1 was meant to determine if the scanning patterns of the individuals with PCA show a pattern of differences from controls that are similar to that of prosopagnosia, or simultanagnosia. Previous research suggests that healthy individuals show a reliable, stereotypical pattern of fixations (e.g., eyes or nose, followed by the mouth) when looking at faces, while individuals with prosopagnosia show scanning patterns that are often very different than those seen in healthy individuals. In contrast, prosopagnosia is associated with scanning patterns that emphasize the mouth, or a featural approach, while simultanagnosia is associated with reduced fixations to the eye region and increased fixations to areas of high contrast. For Experiment 1 an attempt was made to replicate the findings of previous face perception eye-tracking studies in our own healthy sample, and to compare these results to those of the individuals with PCA.

Materials and Methods

Participants. Over the past 3 years, our lab has come into contact with 5 individuals who have been diagnosed with PCA by neurologist Dr. Paul Shelton. Three of these individuals, RB, SS, and AP, participated in Experiment 1, while two individuals, MTB and PLH, were unable to participate due to problems in maintaining fixation. The following section provides detailed case descriptions of each individual with PCA.

Case Descriptions.

RB. RB was a 76-year-old, right-handed female who is a retired travel agent, and an active artist. RB began to show perceptual symptoms approximately 4 years before being first introduced to our lab in September 2008. Upon our initial meeting, RB reported that she had been experiencing difficulty recognizing familiar faces (e.g., RB was unable to recognize the faces of her family and friends when looking through a photo album) and changes in colour perception over the previous two years. RB described an incident at her son's hockey game where she engaged in a detailed conversation with a stranger who she believed was her son.

RB scored very low on a famous faces task (1/50) in our lab (see Table 2), and, during a famous faces task with her neurologist, RB thought she was looking at photographs of her family members. RB also showed impairments in object perception when assessed by both her neurologist, and in our lab (see Table 2 and Table 3), and RB mentioned she had trouble recognizing logos and store signs. RB's neurologist suggested that RB experiences moderately severe apperceptive object agnosia; she was unable to accurately draw simple line drawings that she failed to recognize. In our lab, RB's performance on a simple shape-matching task (e.g., square) was less accurate compared to aged controls, however RB was much better (e.g., 83% correct) at this task compared to the face and object identification tasks (see Table 2).

According to RB's neurologist, RB shows signs of simultanagnosia. Specifically, when RB was asked to identify an object, RB's responses appeared to be based on a single feature. For example, RB when shown a photograph of a camera, RB pointed to the lens of the camera, and identified the image as a 'tunnel'. Similarly, when asked to copy simple images, RB appeared to focus her attention to a single detail of the image, which lead to a reproduction of only this single detail. RB's neurologist reported that RB would re-draw the same a feature a few centimeters

away from the original, seemingly without awareness of her initial drawing. RB's drawings often consisted of a series of disconnected lines, that she did not integrate. Further evidence for a perceptual problem such as simultanagnosia is noted where, according to the neurologist's report, RB's drawing was impaired, while her praxis appeared normal (see Case Descriptions).

RB's problems in colour perception were evident in the lab, when she could not correctly identify the colours presented in Ishihara colour plates. RB also reported that she experienced colour 'hallucinations' on a daily basis. Specifically, RB reported that the walls in the room changed colour when she blinked her eyes. Beyond colour perception, RB showed problems with reading, writing, and finding keys on a computer keyboard. According to RB's neurologist, RB experienced problems with spelling, sounding out words, and construction of letters. RB's neurologist reported that she was very slow at reading, and suggested that RB likely engages in a letter-by-letter reading. RB's neurologist reported that RB's verbal language, memory, and basic visual function appeared intact, and that RB did not show significant signs of hemispatial neglect or spatial disorientation.

According to her neurologist, RB's executive function was in a normal range when tasks did not use visual stimuli. Importantly, RB did not show any significant impairment in reaching and grasping, and her reasoning and memory were relatively intact (see Table 2; DRS-II and MMSE scores). RB did not report any problems managing personal finances. Importantly, RB was slower than aged controls on the Finger Tapping task (FTT), as well as a simple reaction time task, which suggests slowed motor speed, and possibly slowed processing speed (see Table 2).

RB's medical history indicated that RB had mild cataracts, and showed early signs of macular degeneration. Both conditions were considered to have little effect on RB's vision. RB

has sustained 2 head injuries in the past, with many years between each incident. One injury occurred in the 1960s, within the left temporal lobe. The second injury occurred in approximately 1971, within the right temporal lobe. There was no hospitalization, and no treatment necessary.

A recent MRI scan in our lab has shown atrophy in RB's right anterior temporal lobe (Figure 1). RB's neurologist reported that RB had likely experienced progressive visual agnosia for 2-3 years leading to initial assessment. In all, RB's neurologist suggested that her symptoms are part of a progressive disorder that have so far affected the inferior temporal cortex, and angular gyrus of the right hemisphere. RB's neurologist concluded that RB presented features that were consistent with PCA.

SS. SS was a 66-year-old right-handed male, and a retired accountant. SS's initial symptoms that included mainly spatial disorientation, were first noticed by his family in 2007. Initially, SS began to show difficulty finding his way around his house, and was unable to find objects that appear to be in view. At one point, SS could not find his way to a coffee shop to meet with a colleague for coffee. More recently, SS's spouse had to assume responsibility of household finances when SS became unable to complete simple math problems (i.e., acalculia).

According to a neurologist's report, SS has shown signs of optic ataxia: SS often misreached when reaching for, or pointing to an object, or while attempting to shake hands. SS's neurologist also indicated that SS displayed mild ideomotor apraxia; SS had mild to moderate problems in hand posture when asked to imitate familiar and novel hand movements, respectively. Also, according to SS's neurologist, SS showed significant constructional apraxia, and performed poorly at motor sequencing tasks. SS's neurologist interpreted SS's constructional apraxia as a problem related to impaired spatial perception.

SS's neurologist suggests that SS's most significant problems exist in spatial orientation and spatial perception. SS's responses were found to be most impaired when based on visual stimuli and/or spatial processing. In addition, SS's visual fields are limited; SS failed to identify visual targets that were presented in the left and right lower hemifields. SS's neurologist interpreted these limitations as a sign of severely impaired spatial attention. Importantly, SS has previously been diagnosed with a visual field defect, as well as macular degeneration that was identified by an ophthalmologist.

SS's neurologist suggested that SS experiences mild object agnosia, and mild alexia with agraphia, and indicated a diagnosis of Balint's syndrome. Tests in our lab also indicate that SS has problems such as mild object agnosia (see Table 2 and Table 3). SS's neurologist suggested that SS's object agnosia could be the result of simultanagnosia. Evidence of simultanagnosia was seen when SS often made errors after he attempted to identify an object based on only a single detail of the object, and failed to relate the single component to the whole object. For example, when presented with a photograph of a ball decorated with stripes and stars, SS identified the object as 'stars'. Also, SS produced drawing of misidentified objects that consisted of disconnected features.

Additional changes noted by SS's family include a slowed pace when walking, and difficulty sleeping. SS's family also reported that SS has become more 'emotional', showing an increase in anxiety and agitation. SS's neurologist also indicated that SS was slow at number cancellation, and in conversation, which was interpreted as an indicator of slowed processing speed. Additionally, SS performed below normal levels in a FTT, and produced longer reaction time (RT) in a simple reaction time task (see Table 2). SS shows problems finding the correct word during speech, and sometimes mixes up the meaning of words when reading aloud (e.g.,

says ‘dog’ when he means to say ‘cat’). SS’s primary language impairment is in reading; SS can repeat sentences that are presented auditorily, but is generally unable to read sentences. SS was also found to produce spelling errors, has problems with writing, and ignores words.

Based on both SS’s family and his neurologist’s report, SS shows poor memory for dates. According to his neurologist, SS’s has mild memory impairments that are thought to be primarily a retrieval deficit. SS’s neurologist reported that SS’s executive functions appeared less affected, while this was difficult to assess given SS’s significant problems in higher visual processing, and that many tests include visual stimuli.

SS received a diagnosis of PCA from his neurologist based on the predominance of problems in higher visual processing in the absence of brain trauma or stroke. Memory and reasoning were not the most predominant symptoms, while spatial perception, and problems such as optic ataxia appeared most prevalent for SS. An MRI in January 2009 showed mild diffuse atrophy, and enlargement of the ventricles (Figure 1).

AP. AP is a left-handed 78-year-old female who was first introduced to our lab in December 2009. AP is a retired journalist whose initial symptoms included unsteady gait, falling, and problems reading. Based on assessment by a neurologist, AP has been found to have impairments such as simultanagnosia, optic ataxia, mild hemispatial neglect, and some problems in speech production.

Based on an assessment by her neurologist, as well as in our lab, AP was found to have relatively unaffected face perception, and object recognition (see Table 2 and Table 3). AP’s performance was similar to aged controls on object naming, but AP was less accurate when asked to count objects (see Table 2). AP indicated that she had stopped reading due to increasing difficulty. AP’s neurologist interpreted AP’s reading problems as being the result of

simultanagnosia, as opposed to alexia. AP's reading was slow and hesitant, but improved when her neurologist provided a viewing window that allows for letter-by-letter reading.

According to AP's neurologist, AP has difficulty with balance while standing or walking, often bumps into objects while navigating the environment, and also experiences dressing apraxia. AP also shows mild bradykinesia of limb movement, increased muscle rigidity, and mild dysarthria. Coordination was determined to be relatively normal, but AP displayed signs of optic ataxia (e.g., misreaching) during neurological assessment. Further, AP was slower than aged controls on both a FTT and a simple reaction time task, suggesting slowed motor responses, and slowed processing speed (see Table 2).

AP's neurologist also found evidence of right hemisensory neglect, and AP also tended to misbisect lines to the left. Signs of hemispacial neglect were considered to be relatively mild. A SPECT scan conducted through her neurologist recently has shown moderate hypoperfusion in the left occipitoparietal area, and to a lesser degree in the right hemisphere. Further, a MRI completed through our lab suggests atrophy in similar areas (Figure 1). Due to lack of injury or stroke, in combination with impairments in higher visual processing, AP's neurologist suggested that AP's symptoms are consistent with PCA (e.g., components of Balint's syndrome), particularly the dorsal stream variant of PCA.

MTB. MTB is a right-handed, 67-year-old female and retired teacher. MTB first became involved with our lab in December 2008. MTB's initial symptoms, as measured by a neurologist, included problems in motor coordination, such as agraphia, unsteady gait, and dressing apraxia. MTB's memory function, attention, and executive function appeared less affected. However, tests that required responses based on visual stimuli were difficult to properly assess due to significant perceptual and visual-spatial problems.

According to her neurologist, MTB also experiences problems with left/right discrimination, spatial disorientation, often losing her way in familiar settings. Upon initially meeting with our lab, and her neurologist, MTB showed only mild problems in face and object recognition, normal object naming, normal verbal language, and experienced only slight memory loss. Over the past two years MTB has developed more significant deficits in object perception, face perception (Table 2), and she now reports that she is experiencing colour hallucinations, and problems with depth perception. For example, MTB reported that greyscale images of faces that were presented on a computer screen appeared to be ‘glowing purple’ when she looked at them.

An example of MTB’s depth perception problems are illustrated in an event where MTB was unable to walk across a patterned floor. MTB had been visiting her son at his university, and when she and her husband approached a large banquet room with a patterned floor, MTB reported that she was unable to cross the floor because the pattern appeared to ‘...move about on the floor...’ and looked as if each square was set at a different height than then others. MTB has also reported increasing difficulty with stairs because they appear uneven, or look like a large drop. Importantly, MTB reports that she is aware of the inaccuracies of her perception in these situations, and has indicated that she finds these experiences to be ‘terrifying’.

According to her neurologist, MTB appears to experience simultanagnosia, and this is thought to contribute most to MTB’s reading difficulty and problems in drawing. MTB could identify individual letters, but she had difficulty reading full words. Similar to AP, MTB reported that she stopped reading due to increasing difficulty, and reported that the letters ‘jump around’ on the page. MTB’s reading appears to improve markedly when she uses a viewing window to restrict her vision to single letters. Further, MTB’s neurologist suggested that, when writing a sentence, MTB produces poorly formed characters, while the sentence itself is grammatical.

MTB also displays a significant constructional problem when asked to copy simple figures. When attempting to copy a simple figure, MTB said that she could not complete the drawing because her ‘...eyes are flickering... it’s like it’s dancing’. Similarly, for the clock drawing MTB reported ‘I wish the paper would stay still... I can’t see it’. That is, reading appears to be impaired by problems in visual scanning, in that MTB is unable to follow a line of print.

MTB shows evidence of hemispatial neglect on line cancellation task (see Table 3), and slowed motor speed and slowed processing on tasks such as FTT and a simple reaction time task (see Table 2). A recent MRI scan suggests that MTB has atrophy in occipitoparietal areas, as well as some occipitotemporal atrophy (Figure 1). Based on MTB’s predominance of perceptual and visual-spatial symptoms, along with an absence of head injury or stroke, MTB’s neurologist suggested a diagnosis of PCA. Face recognition progressively worse. The significant of visual perceptual problems is consistent with PCA.

PLH. PLH is a 62-year-old, right-handed female, who had formerly worked in retail and secretarial positions. PLH first noticed problems approximately 4 years ago when, despite repeated attempts at obtaining the ideal prescription for her eyeglasses, PLH experienced significant problems in reading (e.g., blurry vision, eye fatigue). Upon assessment by a neurologist, PLH was found to experience multiple problems in perceptual and spatial vision.

Based on her neurologist’s assessment, as well as an assessment in our lab, PLH was found to be impaired in object recognition and face object recognition (see Table 2 and Table 3). According to PLH’s neurologist, PLH also shows significant impairments in word recognition. PLH was also unable to copy simple line drawings, including line drawings of the objects that she could not identify. PLH reported difficulty with finding items on her desk, and had to stop activities such as writing short stories and drawing cartoons due to increasing difficulty.

PLH also shows signs of simultanagnosia; when asked to describe images to her neurologist, PLH often described only a single component instead of describing an object as a whole. When shown a scene of a picnic, PLH thought the people in the picture were flying because she did not perceive the fact that they were seated on a blanket. Further, PLH was unable to describe the Boston Cookie Theft picture.

PLH showed signs of environmental agnosia; she has gotten lost on the way to the grocery store that is located across the street from her home. Based on her neurologist's assessment PLH also shows problems such as mild apraxia, and in visually guided action; PLH often misreached for a target, and she overshoots to left with both limbs when asked to touch her index finger to her nose. PLH has also complained that she often trips over furniture, which may be the result of dorsal simultanagnosia.

PLH was also found to experience problems in memory (see Table 2 and Table 3; DRS-II), and neurological assessment revealed that PLH's visual fields are impaired in all 4 quadrants. PLH experiences left visual extinction in the upper and lower quadrants, and her saccades were found to be slow to initiate in both vertical and horizontal directions.

An MRI scan performed through our lab showed diffuse atrophy (Figure 1). Due to a lack of brain injury, along with the prevalence of visuo-spatial symptoms, including components of Balint's syndrome, PLH's neurologist suggested a diagnosis of PCA. Further, PLH's diagnosis is described by her neurologist as involving both dorsal and ventral streams. Since this assessment, PLH has also received a diagnosis of AD.

Cognitive and Perceptual Tests. All of the individuals in the PCA group received their diagnosis from a local neurologist, based on a series of cognitive and perceptual tests (see Case Descriptions, Table 3) combined with imaging data. A set of tests was also completed in our lab

with individuals from the PCA and aged control group (see Table 2). Tests were run in our lab with both the PCA and aged control group to obtain information regarding each individual's basic functioning, which helped determine whether or not completion of a face-matching task was possible. If, for example, an individual performed very poorly in memory, completion of a face-matching task, where there is a delay between the cue and target stimuli, may not be possible. Also, neuropsychological testing was used to provide a more detailed comparison between the individuals with PCA and the aged controls. All tests completed in our lab are reported in Table 2, and all tests completed by Dr. Paul Shelton are reported in Table 3. The following section describes tests that were completed in our lab.

A famous faces task was included because individuals with prosopagnosia are often found to have a deficit in recognition of familiar faces. For this task, the participants were asked to identify famous individuals' faces as they were presented, one at a time, on a computer screen. If the participant was unsure of the individual's name, they were encouraged to describe any known attributes of the individual. For example, participants often produced responses such as "...the lady with all of the shoes" for Imelda Marcos, or "...he's an American comedian, in that movie called 'The Jerk'" when presented with an image of Steve Martin. Both responses would be scored as correct. There were not time limits on responses, and the experimenter recorded all responses. After the task was complete, for the incorrect responses, the experimenter read the names of the famous individuals out loud to the participant. The assumption was that a problem in face recognition accounted for incorrect responses when the participant reported that they were familiar with an individual based on their name. This task was included to assess recognition of familiar faces because individuals in the PCA group, such as RB reported problems recognizing family and friends.

The Benton Face Recognition Task, Short Form (BFRT-SF) was included to assess recognition of unfamiliar faces. Previous research suggests that differences sometimes exist in familiar versus unfamiliar face recognition (e.g., Benton et al. 1994). That is, problems in recognition of unfamiliar in a face matching task faces is thought to be more common, and dissociations may occur between familiar and unfamiliar face recognition abilities. For the first portion of this task, participants were asked to match a single, front-view, face to one of 6 target faces by pointing or verbal identification. The second portion of this task required the participants to identify the matching target face when the orientation of the target faces varied. The third portion included variations in light and shadow within the target faces. The stimulus items were presented in a booklet, with the response choices presented together on the page below the stimulus picture. Each trial was self-paced, and the participants were allowed to view both the stimulus and response faces for an unlimited duration.

An Object Identification task, developed in our lab, was utilized to examine object recognition, compared to aged controls, in each individual with PCA. This self-paced task consisted of 18 colour photographs, each of a single object (e.g., alarm clock, lamp, baseball). Participants were asked to identify a single object that was presented on a computer screen, while the experimenter recorded each response. An object counting task consisted of 14 objects from the identification task, where each image contained duplicates of the same object in an array (e.g., 4 alarm clocks). The participants were instructed to count the items presented on the computer screen, and to give a verbal response. Often, the individuals in the PCA group used their index finger to trace the location of each object while counting. This task was meant to compare perception of multiple objects in a visual array between the individuals in the PCA

group and the aged control group. These tasks were important to better determine the nature of the perceptual deficits experienced by the individuals in the PCA group.

The Boston Naming Task, Short Form (BNT-SF) was included to assess individuals' abilities to name visually presented line drawings, and is often used primarily as a test for aphasia, perceptual impairments, and problems in semantic retrieval. This task also provides insight into an individual's ability to identify objects. In typical AD, poor performance is thought to result from poor semantic retrieval. For the individuals in the PCA group, it was assumed that, based on poor performance on other perceptual tests, problems completing the BNT-SF would be due to impaired perception, rather than other cognitive impairments. In addition, this task was completed with healthy individuals to ensure normal functioning.

A shape-matching task, developed in our lab, was included to assess visual shape-matching and recognition ability. Each image consisted of a simple shape (e.g., triangle) that was presented at the centre of the computer screen. The participants viewed the initial cue stimulus for an unlimited amount of time, before pressing the Spacebar. Following the button press, the cue image was immediately replaced by a checkerboard mask for 250 milliseconds (ms), which was then followed by a target image. For each trial, the cue and target image were either identical or different. The participants pressed the Spacebar once more to terminate the target image, and bring up response screen. The participants either typed (e.g., 's') or verbally reported their responses. Individuals in the PCA group often experienced difficulty in finding keys on the keyboard beyond the Spacebar. In the cases where participants responded verbally, the participants made keys presses only to the Spacebar, while the experimenter typed in the participants response. This task was included to assess the individual's ability to match simple shapes that were presented with a slight delay (i.e., 250ms). The procedure for the shape-

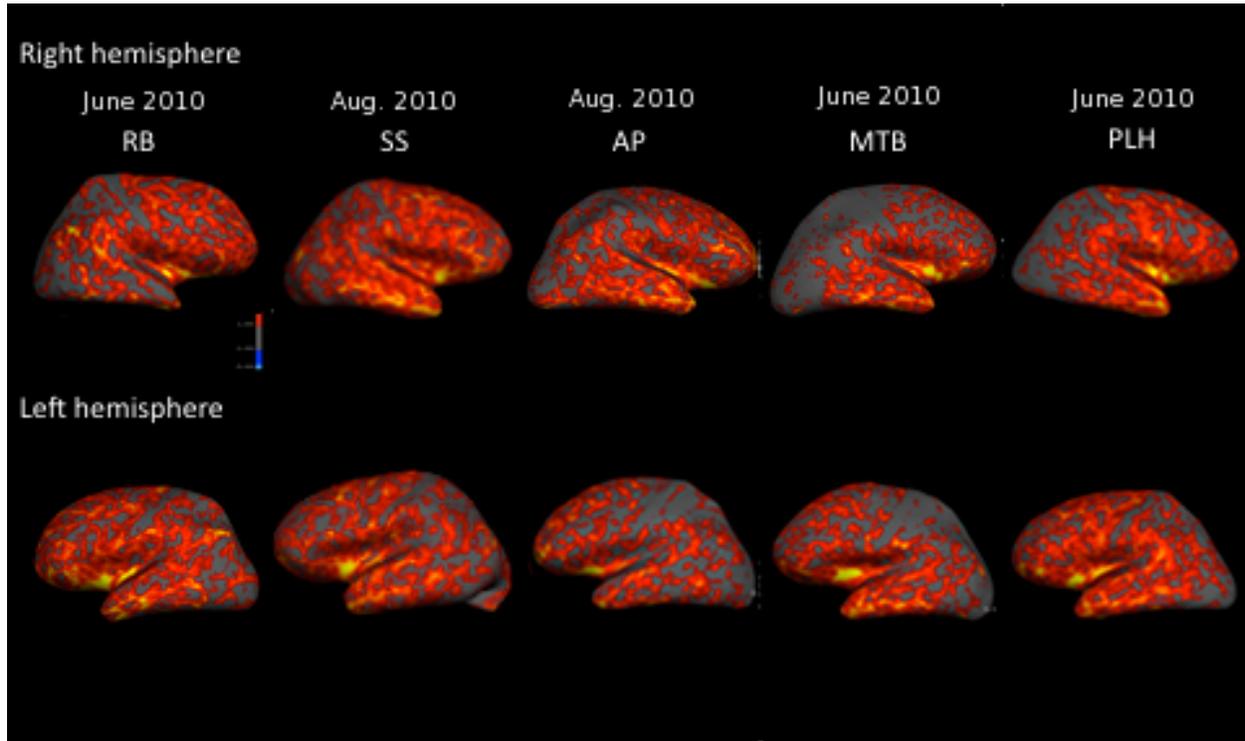
matching task was identical to that of the face-matching task of Experiment 1 and Experiment 2. In order to ensure that a delay between stimuli would not pose a problem for visual memory, this task was included.

A simple reaction time task and a finger-tapping task (FTT) were included to assess processing speed, and motor speed. The simple reaction time task was completed using a laptop computer. For each trial, the participants were asked to press the Spacebar as soon as they detected a green circle on the computer screen. Once the stimulus was detected, and the button press complete, a trial was terminated before the next trial began. The FTT was completed using a wooden board with a button at the top centre that was meant to record finger taps. At the beginning of the task, participants were instructed to place their dominant hand, palm down, on the wooden board with their index finger placed upon the button. Next, the participants were required to begin tapping as quickly as possible once the experimenter said 'go'. Tapping continued for 10 seconds (s) per trial, with three trials per hand. Dominant and non-dominant hands were alternated between trials. An average score per hand was then calculated.

A phonemic verbal fluency task, or controlled oral word association task (COWAT) was completed. The letter 'F', 'A', and 'S' were used, and participants were asked to generate as many words as they could for each individual letter. That is, for each letter, the participants were timed for one minute, and were instructed to avoid use of proper nouns as well as use of the same word with a different ending (e.g., 'run' and 'running'). This task is thought to be less affected by age, and more related to education level and vocabulary. In addition, this test is thought to be sensitive to cognitive changes; a lower than expected score given age and education may indicate cognitive decline in areas such as executive function and working memory. To obtain a score for each participant, the total number of words for each letter were added together for a grand total.

In addition to the above tests, we included the Mini Mental State Exam (MMSE), and the Dementia Rating Scale II (DRS-II) to assess cognitive function. The procedure for each test was completed in accordance to the instructions provided by the manufacturer except where individuals were asked to read words or sentences aloud, and where visual recognition of words was required. That is, many of the individuals in the PCA group had marked difficulty recognizing single letters within words, or even single letters when other letters were visible in the visual display. Within the Memory and Attention section of the DRS-II, participants are asked to read a sentence for later recall, and to read a list of words to be chosen later from visually-presented word pairs. Thus, for this section, the experimenter verbally presented the words to each individual in the PCA group (see Table 2 and Table 3).

Figure 1. RB, SS, AP, MTB, and PLH Cortical Thickness.



Cortical thickness maps presented on participants' "inflated" brains. Grey areas represent regions of cortical thinning (i.e. atrophy).

Control Groups. Four Aged-matched healthy participants (4 females, right-handed, Mean age=71 years old), and 12 Undergraduate participants (8 females, 10 right-handed, Mean age=23.5 years old) also participated in Experiment 1 as healthy controls (see Table 1 for participant demographics). All older participants completed tests of cognitive and perceptual functioning (see Table 2 and Table 3). Undergraduate students were recruited from the University of Manitoba's Introduction to Psychology Subject Pool, and all received course credit for their participation. All undergraduate participants were assessed for handedness, were fluent in English, and had normal or corrected-to-normal vision.

Table 1

Demographics

	RB	SS	AP	MTB	PLH	C1	C2	C3	C4	C5
Age (in years)	76	66	77	66	61	63	67	80	74	75
Gender	F	M	F	F	F	F	F	F	F	M
Handedness	R	R	L	R	R	R	R	R	R	R
Education (in years)	16	19	16	16	13	15	13	16	16	16

C1-C5 represent age-matched healthy control participants.

Table 2

Cognitive and Perceptual Tests - Completed in Lab

	RB	SS	AP	MTB	PLH	C1	C2	C3	C4	C5
BTFR-SF SF	<37 - severely impaired	<37 - severely impaired	47 - normal range	<37 - severely impaired	<37 - severely impaired	51- normal	51- normal	52- normal	54 - normal	54 - normal
Famous Faces Test	1	22	30	29	6	46	33		30	31
Object Identification	27.8%	72.2%	100.0%	100.0%	72.2%	100.0%	100.0%	100.0 %	94.4%	94.4%
Object Counting /14	85.7%	42.9%	78.6%	64.3%	28.6%	100.0%	100.0%	100.0 %	92.9%	100.0%
Shape- matching task	83.0%	100.0%	100.0%	94.4%	N/A	100.0%	N/A	97.2%	97.2%	97.2%
Boston Naming - SF	20.0%	46.7%	100.0%	73.0%	20.0%	100.0%	73.0%	100.0 %	100.0%	93.3%
Finger Tapping task										
Right Hand	19	26.43	11.75	15.3	27.83	36.33	31.33	30.17	29.5	50.17
Left Hand	18.5	19.07	14	11.83	28.16	36	26.65	25.33	31	36.5
Reaction Time task										
Mean	654	784	673	793	N/A	319	N/A	252	376	363
Median	559.5	717.0	641.5	672.0	N/A	286.0	N/A	234.0	375.5	301.0
Phonemic Fluency Task	F-17	F-19	F-16	F-12	N/A	F-8	F-19	F-13	F-17	F-18
	A-10	A-14	A-12	A-15	N/A	A-4	A-14	A-14	A-13	A-15
	S-16	S-20	S-17	S-16	N/A	S-11	S-14	S-15	S-12	S-13
MMSE	26	28	28	28	24	30	30	30	30	30
DRS	126, 6- 10th percentile	129, 6- 10th percentile	134, 29-40th percentile	125, 3- 5th percentile	108, <1st percentile	142, 82-89th percentile	136, 29-40th percentile		139, 60-71st percentile	142, 82-89th percentile

* Indicates scores below norms for control participants.

Table 3

Cognitive and Perceptual Tests - Completed by neurologist

	RB	SS	AP	MTB	PLH
Matching Face to name /10	--	10	--	10	6
Famous Face Naming /10	0	8	8	8	1
Object Naming /30	12	19	26	27	16
Category Fluency Task (Animals)	19 (50 th percentile)	6 (<10 th percentile)	--	--	9 (<10 th percentile)
COWAT	63 (>90 th percentile)	41 (50 th percentile)	--	--	57 (>90 th percentile)
Boston Naming - SF	3*	7*	15	11*	3*
Bells Cancellation /34	6*	10* (none in lower visual field)	10*	17*	--
Rey-Osterrieth Complex Figure Test /36	7* (copy) 2.5* (recall)	5* (copy) 1* (recall)	--	4* (copy) 0* (recall)	1* (copy) 0* (recall)
Digit Span Test:					
Forward	8 (25 th -50 th percentile)	--	--	6 (5 th percentile)	7 (25 th percentile)
Reverse	4 (5 th percentile)	--	--	2 (<5 th percentile)	4 (5 th percentile)
MMSE	27*	20*	24*	26*	25*
Trail Making Test	315 sec (<10 th percentile - A) --(B)	--	--	120 sec (A<10 th percentile), -- (B)	420 sec (A <10 th percentile), -- (B)
Clock Drawing Test /15	11	5*	12	10	4*

* Indicates scores below norms for control participants.

Stimuli. 76 digital pictures of faces taken from the Productive Aging Lab database (Minear & Park, 2004) were converted to a grayscale format and presented on a black background. A standard oval mask of 300 pixels across the centre was created with Corel Photopaint (Corel Corporation, 2008), and applied to all faces to remove external features such as hair, jewelry, and jaw line. Of the 76 faces, 38 were used as cue stimuli, and 38 were included as target stimuli. Cue-target pairs were presented in Matlab 2008a (MathWorks, El Segundo, CA). Each target face followed a cue face, and half of these cue-target pairs were comprised of the same face (matched pair), while half were comprised of two different faces (unmatched pair) half of the cue faces were matched to the target faces.

Materials. An Eye-link II (250 Hz sampling rate, spatial resolution < 0.5 degrees; SR Research Ltd., Osgoode, ON, Canada) was used to record eye-movements throughout the experiment. Pupil and CR-based tracking were used in monocular mode, and participants were tested individually at a station consisting of a 3.2 GHz computer, keyboard, and monitor. All stimuli were displayed on a 20.1" LCD monitor running at a resolution of 1600 × 1200 at 60 Hz. Participants were seated approximately 50 cm from the monitor, and a chin rest was used to prevent head movement related error.

Procedure. The individuals in the PCA group, as well as the aged control group were required to complete a series of cognitive and perceptual tests. For the individuals with PCA, these tests were in addition to tests that had been completed with a neurologist.

For the open-view (OV) task of Experiment 1, all participants received verbal and written instructions, followed by a demonstration of the proper use of the EyeLink II apparatus. To start, each participant's eye-movements were calibrated using a 9-point calibration screen and validated to less than 1-degree of error before beginning the experiment. Each trial began with a

fixation point to allow for the use of in-vivo drift correction, to compensate for headband slippage or other small movements in the head-mounted eye-tracker. Following the fixation marker, a checker-patterned mask appeared, followed by a single face that was presented at the centre of the computer screen.

There were no time limits on viewing cue or target stimuli, and the participants were instructed to look at each cue face until they memorized the image. The participants pressed the 'Spacebar' key once finished viewing each image, which terminated the cue face. Following the cue face, a mask was presented for 250 ms, followed by a central fixation point and the target face. Drift correction occurred immediately before each cue face. The participants were instructed to press the spacebar as soon as they made a same/different decision, and then required to press the '1' key for 'same' and the '2' key for 'different'. Due to the difficulty experienced pressing the response buttons within the PCA group, the patients were asked to simply press the spacebar key once they made their decision for the target, and to then lift their left hand slightly from the desk for a 'same' response, and to leave their left hand on the table top to respond 'different'. Verbal responses could not be used due to the chin rest. All of the patients completed enough training in this response procedure to demonstrate adequate understanding to the experimenter before the experimental trials began. For the PCA group, the experimenter pressed '1' for same, and '2' for different, using a response pad that was connected to the computer keyboard. Healthy participants used the standard key presses for same/different responses. The response key presses terminated the response screen, and initiated the next trial. No feedback was given with regard to accuracy. Stimulus presentation order was randomized for each subject, with four or more practice trials beginning the experiment.

Results

Both MTB and PLH did not participate in Experiment 1 due to problems maintaining fixation, which prevented calibration with the eye tracker.

Aged vs. Young Controls.

Errors. Comparisons between control groups were conducted using a two-tailed independent-samples t-test, with all results significant at $p < .05$. The results showed no significant differences in errors when aged ($M=83.04\%$, $SD=17.59$) and young ($M=86.31\%$, $SD=12.82$) controls were compared ($t(14)=.748$, $p < .05$)¹.

Viewing Time. An average median VT was computed in milliseconds (ms) for each control group. VT was computed for Cue and Target faces separately. VT for cue faces represents the amount of time an individual spent learning a face, and can be thought as the encoding portion of VT. VT for target faces is thought to represent the amount of time taken to retrieve the Cue face, and to decide whether or not the Cue-Target pair is a match. VT was measured separately for Cue and Target Stimuli because VT for Cue and Target stimuli would be affected by different variables; encoding and retrieval, respectively. Comparisons between the control groups were conducted using two-tailed independent-sample t-tests, with all significant results at $p < .05$.

Cue stimuli. There were no significant differences in viewing time when aged ($Mdn=5603$, $SD=3004.41$) and young ($Mdn=4429.43$, $SD=2854.60$) controls were compared ($t(14)=.598$, $p < .05$).

Target stimuli. There were significant differences were found between aged ($Mdn=3142.50$, $SD=1187.25$) and young ($Mdn=1553.57$, $SD=725.25$) controls when viewing

time was compared for target faces ($t(14)=3.254, p<.05$). The aged controls spent significantly more time viewing Target stimuli compared to young controls.

¹ A same-different signal detection paradigm was utilized, with both correct and incorrect responses included in the analysis (Macmillan & Creelman, 1991). The d' results are shown in Table 3. These results suggest bias did not play a large role in the participants' responses.

Table 4

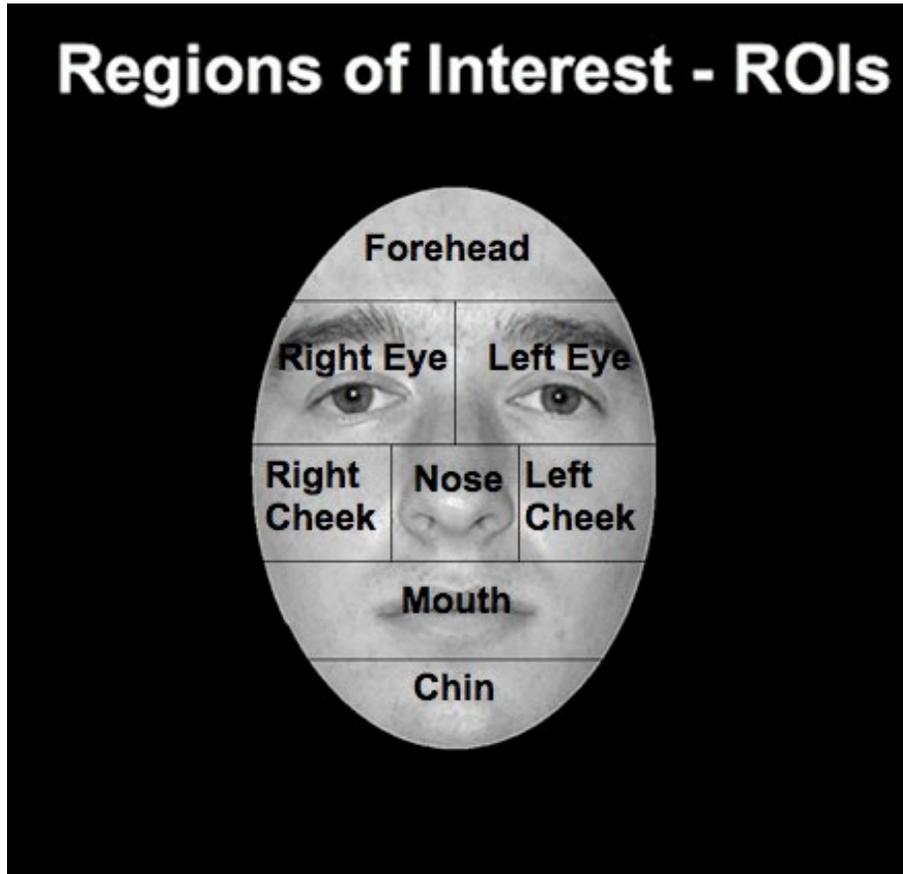
Same-different signal detection paradigm

	Stimuli=Different	Stimuli=Same	
AGED	d'	1.878 d'	1.886
	c	-0.097 c	0.101
YOUNG	d'	2.282 d'	2.256
	c	0.263 c	-0.25
RB	d'	-0.729 d'	0.714
	c	0.188 c	-0.193
SS	d'	0.182 d'	0.182
	c	0.267 c	-0.267
AP	d'	3.493 d'	3.493
	c	2.553 c	-2.553

Scanning pattern. Cue faces were divided into 8 regions of interest (ROI); Left Eye, Right Eye, Nose, Mouth, Chin, Forehead, Left Cheek, and Right Cheek (see Figure 2). ROIs were based on the stimulus, thus the left eye of the stimulus fell in the right visual field of the participant. To control for differences in the overall number of fixations generated, and the time spent viewing

stimuli, the proportion of fixation counts, and fixation duration were calculated. That is, for each stimulus, a total fixation count (FC), and fixation duration (FD) were calculated. Next, the total FC and FD were calculated for each ROI within the stimulus. A proportion FC (PFC) and FD (PFD) were then calculated by dividing each ROI total by the overall stimulus total. These numbers were then averaged for each participant, across stimuli. For the control groups, participant data was pooled for a grand mean of PFC and PFD, while means for the PCA group were calculated across stimuli only. 95% confidence limits were then obtained from the control mean PFC and mean PFD using SPSS for comparisons between controls and each individual with PCA. A series of paired-samples t-tests, corrected for multiple comparisons ($p < .0021$) using the Bonferroni procedure, were conducted within each control group to determine if there were any differences in scanning between ROIs. A series of independent-samples t-tests, corrected for multiple comparisons ($p < .0062$) using the Bonferroni procedure, were conducted to compare scanning patterns by ROI between the aged and young control groups. For within group (i.e., ROI analysis) and between group (i.e., Aged vs. Young controls) comparisons, only duration was included.

Figure 2. Defined Regions of Interest (ROIs).



Boundary lines and labels denote individually defined ROIs.

Count. There were no significant differences found between control groups for PFC, across ROIs (see Figure 3).

Duration. There were no significant differences found between control groups for PFC, across ROIs (see Figure 4). Significant differences in PFD between ROIs were found within both control groups. The aged control group showed significant differences only between the Right Eye and Right Cheek, while the young control group showed several differences in PFD, by ROI (see Table 5 and Table 6). Due to the overall lack of significant differences found between aged and young controls, further discussion, and comparison to individuals in PCA group will be limited to the aged control group (see Figure 3 and Figure 4).

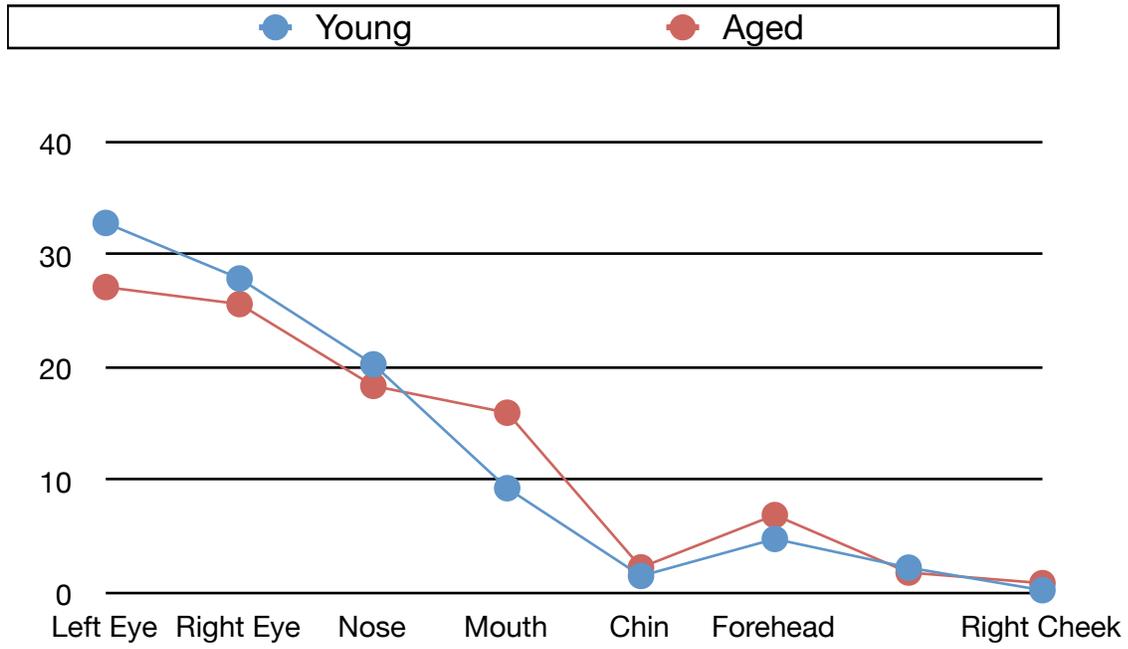
Figure 3. Aged and Young Controls - PFC across ROIs.

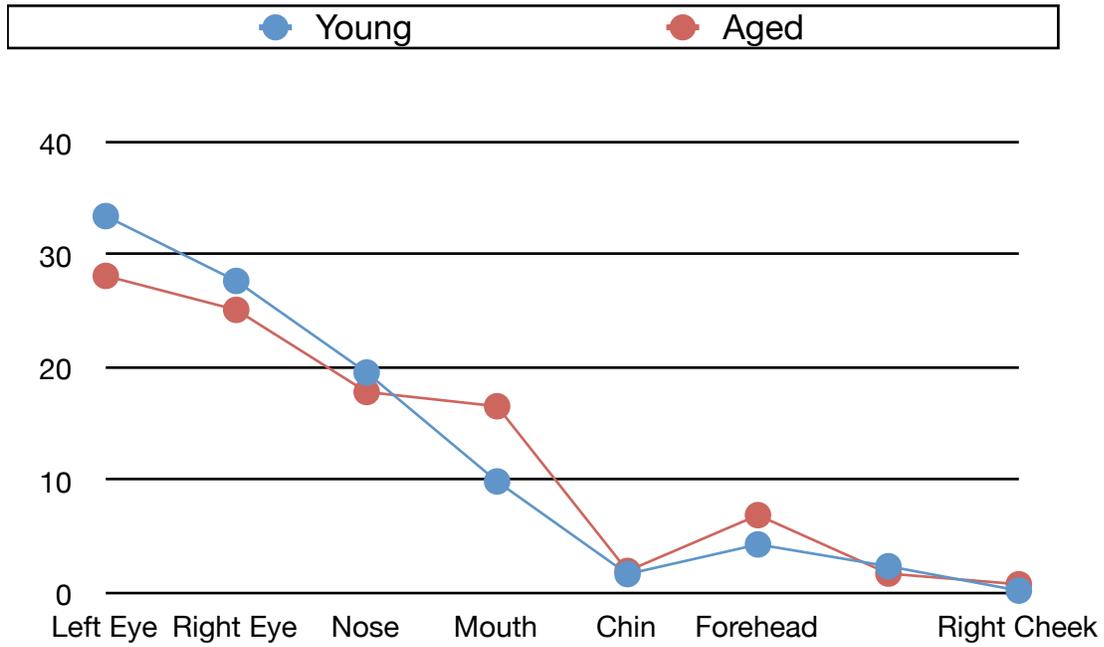
Figure 4. Aged and Young Controls - PFD across ROIs.

Table 5

Open-view task: Aged Controls Paired-samples t-tests for Differences Between ROIs

ROI	Left Eye	Right Eye	Nose	Mouth	Chin	Forehead	Left Cheek	Right Cheek
Left Eye	28.25	25.24	17.96	16.69	2.08	7.03	1.84	0.9
Right Eye	28.25	25.24	17.96	16.69	2.08	7.03	1.84	0.9*
Nose	28.25	25.24	17.96	16.69	2.08	7.03	1.84	0.9
Mouth	28.25	25.24	17.96	16.69	2.08	7.03	1.84	0.9
Chin	28.25	25.24	17.96	16.69	2.08	7.03	1.84	0.9
Forehead	28.25	25.24	17.96	16.69	2.08	7.03	1.84	0.9
Left Cheek	28.25	25.24	17.96	16.69	2.08	7.03	1.84	0.9
Right Cheek	28.25	25.24*	17.96	16.69	2.08	7.03	1.84	0.9

*All numeric values represent PFD. All significant differences are $p < .001^{**}$ and $p < .002^{*}$.*

Table 6

Open-view task: Young Controls Paired-samples t-tests for Differences Between ROIs

ROI	Left Eye	Right Eye	Nose	Mouth	Chin	Forehead	Left Cheek	Right Cheek
Left Eye	33.38	27.61	19.62	9.98**	1.67**	4.42**	2.49**	0.34**
Right Eye	33.38	27.61	19.62	9.98	1.67**	4.42**	2.49**	0.34**
Nose	33.38	27.61	19.62	9.98	1.67**	4.42**	2.49**	0.34**
Mouth	33.38**	27.61	19.62	9.98	1.67	4.42	2.49**	0.34**
Chin	33.38**	27.61**	19.62**	9.98	1.67	4.42	2.49	0.34
Forehead	33.38**	27.61**	19.62**	9.98	1.67	4.42	2.49	0.34
Left Cheek	33.38**	27.61**	19.62**	9.98**	1.67	4.42	2.49	0.34
Right Cheek	33.38**	27.61**	19.62**	9.98**	1.67	4.42	2.49	0.34

*All numeric values represent PFD. All significant differences are $p < .001^{**}$ and $p < .002^{*}$.*

Aged Controls. The aged control group performed similar to previous research on face processing in healthy individuals. That is, the aged control participants made few errors, and produced short VT. Errors and VT fit with assessment tests that were performed in our lab (see Table 2), in that all participants performed normally on perceptual tasks. It is important to note that one participant performed below the norm for their age for the phonemic fluency task, although this performance level has been interpreted as the result of participant anxiety. The individual reported feeling anxious about performing the task in the presence of the experimenter. Other tests of cognitive function for this individual were within the normal range.

The scanning patterns found within the aged controls also fit well with previous research with healthy individuals; the highest PFC and PFD occurred within the central ROIs; Eyes, Nose, and Mouth (see Figure 3 and Figure 4). More specifically, the healthy individuals tended to look at the areas that join these central features, such as the area between the nose and the mouth, as well as the areas between each eye and the nose. Initial fixations were often found in one of the Eye ROIs, or the Nose ROI, similar to previous research which suggests that healthy individuals often emphasize the eyes, and nose, followed by the mouth.

RB.

Errors. Comparisons between each individual in the PCA group and the aged controls were conducted using the 95% confidence limits around the mean for accuracy for the aged controls. Compared to healthy older participants ($M=83.04\%$ correct, $SD=17.59$, 95% CI [55.05%, 100%]), RB (35.71% correct) made significantly more errors.

Viewing Time.

Comparisons between each individual from the PCA group and the aged controls were made using the 95% confidence limits around the median, for the aged control group.

Cue stimuli. Compared to the age-matched controls ($N=4$, $Mdn=5603ms$, $SD=3004.41$, CI [822.31, 10383.69]), RB ($Mdn=12490ms$) produced significantly longer VT.

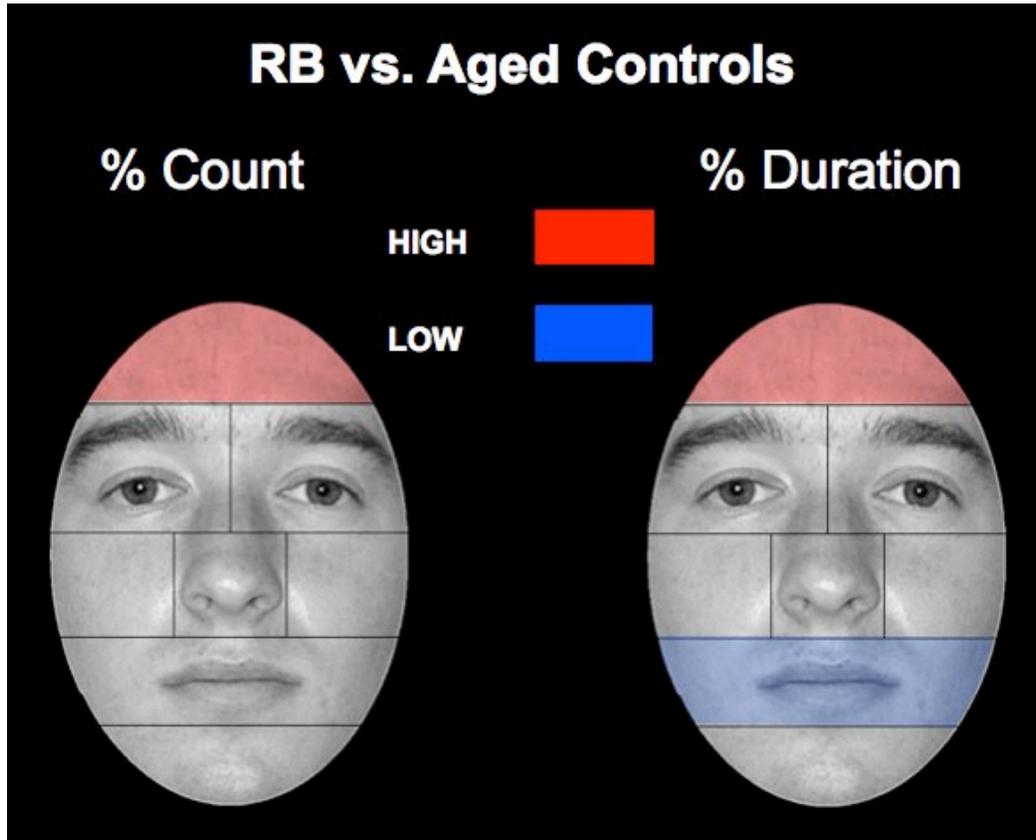
Target stimuli. Compared to age-matched controls ($N=4$, $Mdn=3142.3ms$, $SD=1187.25$, CI [1253.32, 5031.68]), RB ($Mdn=7324ms$) produced significantly longer VTs.

Scanning Pattern.

Count. Compared to age-matched controls, RB produced a significantly higher PFC in the Forehead ($M=22.59%$, CI [1.18%, 12.90%]) (see Figure 5).

Duration. Compared to age-matched controls, RB produced a significantly longer PFD in the Forehead ($M=22.37%$, CI [0.50%, 13.56%]), and significantly shorter PFD to the Mouth ($M=4.6%$, CI [5.59%, 27.79%]) (see Figure 5).

Figure 5. RB's PFC and PFD compared to aged controls.



Stimuli divided into ROIs. Red indicates ROIs where RB showed significantly higher percentages, while blue indicate where RB showed significantly lower percentages, compared to aged controls.

Compared to aged controls, RB produced more errors, and showed differences in her scanning pattern. RB's errors are not surprising, considering RB's problems in global processing (i.e., simultanagnosia) and face perception (see Case descriptions). For example, RB scored lowest on face recognition and face matching (see Table 2 and Table 3). RB does not appear to be using a feature-by-feature strategy, because a featural strategy is often found to lead to correct responses in a face-matching task with novel faces. Instead, RB's scanning pattern appears to reflect simultanagnosia.

RB's scanning pattern differed significantly compared to aged controls, and RB did not show obvious similarities to the scanning pattern that are associated with prosopagnosia (e.g., mouth region). RB spent more time looking at the forehead ROI, and less time looking at the mouth ROI. However, RB's scanning pattern was more focused on face regions that provide less second-order configural information (i.e., spatial relationships between features). By spending more time viewing the Forehead ROI, RB was obtaining information that is located outside of the 'T' shaped area that contains the internal features, where second-order configural information exists. This de-emphasis on second-order configural information fits with previous research on both prosopagnosia and simultanagnosia that suggests these disorders are characterized by impaired holistic/configural or global processing, respectively.

RB's scanning patterns fit more easily with previous research on simultanagnosia. Individuals with simultanagnosia have been found to make fewer fixations to the eye region, more fixations to areas of high contrast (e.g., Dalrymple et al. 2011), while some have suggested problems disengaging from salient areas (Carota & Calabrese, 2011). RB's scanning patterns are concentrated within the Forehead ROI, particularly along a high contrast border. Also, RB's initial fixation was often located within the forehead ROI, suggesting a problem disengaging

from a single detail, similar to her restricted focus seen with objects.

RB's assessment with her neurologist, and performance neuropsychological tests (see Table 2 and Table 3) suggest that RB experiences components of both ventral simultanagnosia (e.g., letter-by-letter reading), and dorsal simultanagnosia (e.g., identification of object details, rather than objects; see Case Descriptions). Both types of simultanagnosia may have affected RB's face processing. Ventral simultanagnosia may exist when RB is unaware of a given image beyond a single detail, while dorsal simultanagnosia may exist when RB is able to perceive simpler shapes, where multiple details are not present (e.g., simple shape matching task; see Table 2, see Case Descriptions). When viewing a face, RB often described a tiny facial detail, such as the iris, without an awareness of the rest of the face. In Experiment 1, RB spent the most time viewing a single detail; a high-contrast area of the Forehead ROI. The stimuli of Experiment 1 likely contained too many details, particularly within each feature (e.g., eyelashes, iris, pupil), to allow RB to be aware of multiple details, or to engage in perception beyond a single detail.

Previous research suggests that smaller stimulus size facilitates global perception in dorsal simultanagnosia (e.g., Dalrymple et al. 2007). Faces contain multiple features (e.g., eyes, eyebrows) which may be difficult for an individual with dorsal simultanagnosia, such as RB, because these facial features may be similar to processing multiple objects. Previous research on dorsal simultanagnosia suggests that individuals with this disorder are better at perceiving stimuli that are smaller. RB made fewer errors in perceptual tasks that contain smaller stimuli (e.g., object counting - 2-2.5in; shape-matching - 2in; BTFR-SF - 2in), and more errors in Experiment 1, where the faces contained multiple features, and they are large in overall size (i.e., 6x8in).

Neuropsychological assessment suggests that RB has a mild memory impairment, which

could partly account errors in Experiment 1. For example, RB produced fewer than normal responses for forward and reversed digits (see Table 3). RB made fewer errors in the simple shape-matching task (see Table 2) where items in cue-target pairs were repeated across trials, compared to Experiment 1, which forced participants to compare a new Cue-Target pair on every trial.

RB's increased VTs for both Cue and Target faces fit with findings from neuropsychological testing (see Table 2 and Table 3; see Case Descriptions) that suggests RB has slowed motor responses, and possibly slowed processing speed (e.g., RB was slow at reading, FTT, and simple reaction time task). RB's slowed VT could also be related to inadequate perception. That is, perception of faces is difficult for RB, while it is presumably easy for health participants, and may force RB to spend more time viewing each image to gain enough information to make a decision. RB performed normally in semantic and phonemic fluency (see Table 2 and Table 3), suggesting that problems in executive function, and language are unlikely to interfere with RB's ability to understand and complete the procedure of Experiment 1.

SS.

Errors. Compared to healthy older participants ($M=83.04\%$ correct, $SD=17.59$, 95% CI [55.05%, 100%]), SS (53.57% correct) made significantly more errors than aged controls.

Viewing Time.

Cue Stimuli. SS did not show any significant differences in VT to Cue stimuli ($Mdn=4064ms$) compared to the age-matched controls ($N=4$, $Mdn=5603ms$, $SD=3004.41$, CI [822.31, 10383.69]).

Target Stimuli. In contrast, SS showed significantly longer VT to Target stimuli ($Mdn=5886ms$), compared to age-matched controls ($N=4$, $Mdn=3142.3ms$, $SD=1187.25$, CI

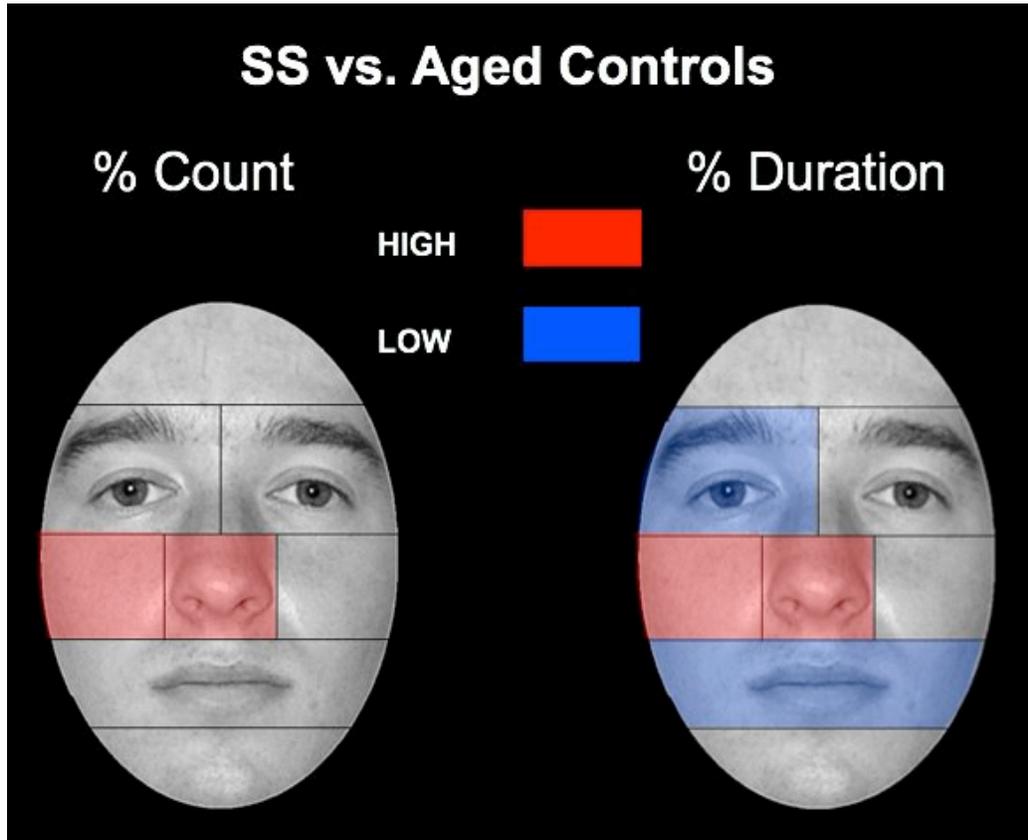
[1253.32, 5031.68]).

Scanning Pattern.

Count. Compared to age-matched controls, SS produced a significantly higher PFC to the Nose ($M=53.68\%$, CI [8.80%, 28.19%]), and the Right Cheek ($M=2.89\%$, CI [0.27%, 1.73%]) (see Figure 6).

Duration. Compared to age-matched controls, SS produced a significantly longer PFD in the Nose ($M=57.89\%$, CI [9.64%, 26.27%]) and Right Cheek ($M=2.56\%$, CI [0%, 1.95%]), and SS produced significantly shorter PFD in the Right Eye ($M=17.98\%$, CI [18.29%, 32.19%]) and Mouth ($M=4.26\%$, CI [5.59%, 27.79%]) (see Figure 6).

Figure 6. PFC and PFD for SS compared to aged controls.



Stimuli divided into ROIs. Red indicates ROIs where SS showed significantly higher percentages, while blue indicate where SS showed significantly lower percentages, compared to aged controls.

SS showed differences from aged controls in errors, VT to target faces, and scanning pattern. SS's errors and scanning pattern in Experiment 1 could be interpreted as the result of problems such as neglect, face perception, optic ataxia, and impaired global processing, from both ventral (e.g., SS's problems in awareness of multiple details in a single object) and dorsal (e.g., SS's problems bumping into furniture) simultanagnosia. Evidence for the effects of simultanagnosia are seen in that SS did not produce errors on a task that relies less on the integration of features (i.e., the shape-matching task) was similar to controls (i.e., no errors).

SS was slower than controls on tasks such as the FTT, simple reaction time task, and even at number cancellation (see Table 2, Table 3, and Case Descriptions). However, SS was not significantly slower than controls for VT to Cue faces. One explanation might be that, according to SS's spouse, SS showed signs of anxiety during testing with his neurologist, which may have hindered performance on some tasks. The fact that SS was slower to Target faces could be due to his retrieval deficit, which may have increased the time SS needed to compare stimuli. Also, SS may have produced increased viewing times compared to controls due to a general slowing in processing, including the speed with which button presses were made.

SS's focus on the Nose ROI could be related to SS's significant visual neglect; SS did not attend to stimuli in both lower visual quadrants (e.g., Bells Cancellation Task; see Table 2). If the Mouth ROI did not fall within SS's neglected fields, SS may have shown a focus within this ROI instead of the Nose ROI. SS made significantly more fixations to the Nose and Right Cheek, and made more fixations to the Nose, compared to the other ROIs. Compared to aged controls, SS spend more time viewing both the Nose and Right Cheek ROIs. SS's scanning pattern does not fit well with previous research on prosopagnosia, and is taken to reflect simultanagnosia, in addition to other perceptual issues (see Case Descriptions).

SS's emphasis on the Nose ROI could be interpreted as configural processing, however, unlike controls, SS's initial fixation was most frequently location within the Nose ROI, and SS showed a tendency to continue making fixations to the Nose ROI. In contrast, healthy individuals may fixate to the nose and eyes first, and will then fixate back and forth between the internal features. SS's greater fixation counts and duration within the Nose, and Right Cheek, combined with decreased a fixation count and duration within the Right Eye, suggest the effects of simultanagnosia. The Right Cheek does not hold important configural information, yet contains a high-contrast border area, while the Nose ROI only holds configural information when areas that connect the Nose with other internal features (e.g., eyes) are included. Also, the eye region is often fixated by healthy individuals. Previous research suggests that individuals with simultanagnosia make fewer fixations to the eye region, and more fixations to areas of high contrast (e.g., Dalrymple et al. 2011).

AP.

Errors. Compared to healthy older participants ($M=83.04\%$ correct, $SD=17.59$, 95% CI [55.05%, 100%]), AP (60.71% correct) did not make significantly more errors than aged controls.

Viewing Time.

Cue Stimuli. Compared to aged controls ($N=4$, $Mdn=5603ms$, $SD=3004.41$, CI [822.31, 10383.69]), AP ($Mdn=18368ms$) produced significantly longer VTs.

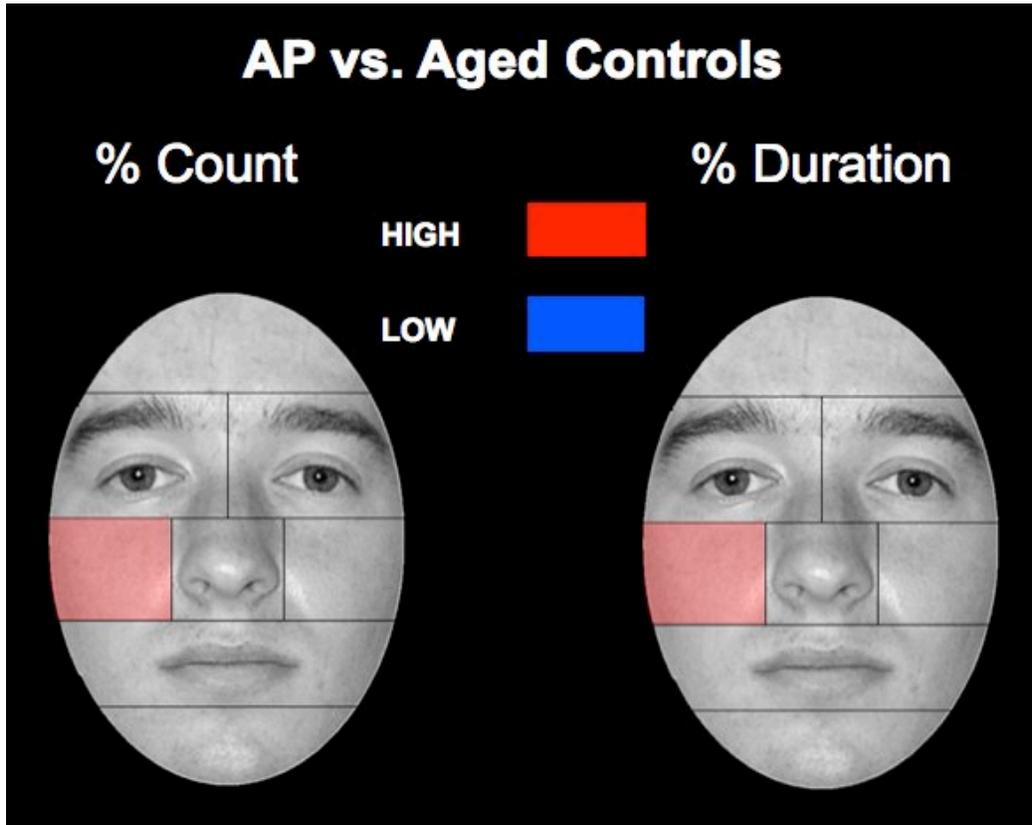
Target Stimuli. Similar results were seen for Target stimuli; compared to aged controls ($N=4$, $Mdn=3142.3ms$, $SD=1187.25$, CI [1253.32, 5031.68]) AP ($Mdn=6128ms$) produced significantly longer VTs.

Scanning Pattern.

Count. Compared to aged controls, AP produced a significantly higher PFC to the Right Cheek ($M=2.69\%$, CI [0.27%, 1.73%]) (see Figure 7).

Duration. Compared to age-matched controls, AP produced a significantly higher PFD in the Right Cheek ($M=3.47\%$, CI [0%, 1.95%]) (see Figure 7).

Figure 7. PFC and PFD for AP compared to aged controls.



Stimuli divided into ROIs. Red indicates ROIs where AP showed significantly higher percentages, while blue indicate where AP showed significantly lower percentages, compared to aged controls.

AP produced less errors than both SS and RB, and was not significantly different from aged controls. Importantly, AP does not display significant problems in face and object perception. However, a report by her neurologist suggested that AP experiences components of dorsal (e.g., AP bumps into objects in the environment) and ventral simultanagnosia (e.g., letter-by-letter reading; see Case Descriptions). Also, AP's accuracy may be affected by mild memory problems that prevent storage or retrieval that would be necessary to compare Cue-Target pairs. AP was slower than aged controls on both a FTT and a simple reaction time task, suggesting slowed motor responses, and slowed processing speed (see Table 2).

AP scored lower than aged controls on object counting, which suggests that AP experiences dorsal simultanagnosia; AP also showed problems counting objects compared to object identification. AP was the least impaired at object and face perception, compared to the other individuals with PCA (see Table 2 and Table 3), thus, it is no surprise that AP's scanning pattern is more similar to that of the aged controls. AP made more errors than controls in Experiment 1, but AP made fewer errors compared to RB and SS. These results are not surprising because AP does not display significant problems in face and object perception.

AP was significantly slower than aged controls for the amount of time spent viewing both Cue and Target faces. Due to the fact that AP is known to be less impaired in face processing, but slow at motor responses and reaction time, these results could be interpreted as a general slowing in processing. However, because AP experiences simultanagnosia, she may have difficulty perceiving each face as a whole, due to the size of the faces. That is, individuals with simultanagnosia are worse at global processing when stimuli are large; the stimuli of Experiment 1 were much larger than the stimuli used in the face perception and object identification tasks. However, AP did not report experiences of problems in face perception in her daily life.

AP was the least impaired at object and face perception, compared to the other individuals with PCA (see Table 2 and Table 3), thus, it is no surprise that AP's scanning pattern is more similar to that of the aged controls. AP showed a significant difference compared to aged controls only in the Right Cheek ROI, which may reflect a focus on high contrast areas, due to simultanagnosia. AP's increased viewing of the Right Cheek ROI may reflect a focus on the high contrast edge that is part of the Right Cheek. Individuals with simultanagnosia have been found to make fewer fixations to the eye region, and more fixations to areas of high contrast (e.g., Dalrymple et al. 2011).

Discussion

The results of Experiment 1 show that the healthy controls produced scanning patterns that fit with previous research on healthy individuals (e.g., Barton, 2008; Caine, 2004; Le, Raufaste, & Demonet, 2003; Stephan & Caine, 2009). That is, the aged controls tended to spend the most time viewing the eyes, nose, and mouth, with the least amount of time viewing peripheral areas. These results were not surprising, considering that the aged controls performed within the normal range for the neuropsychological tests. Further, the results did not reveal substantial differences between control groups, which allowed for comparisons to be limited to those of the aged controls and the individuals in the PCA group.

Each individual in the PCA group showed differences from the aged controls, with AP showing the least differences in scanning pattern. The differences seen between each individual in the PCA group, compared to aged controls were expected, considering the obvious perceptual difficulties experienced by each patient, combined with the results of cognitive and perceptual tests. The results showed that the individuals with PCA produced more errors, longer RTs, and scan paths that appear to fit best with accounts of simultanagnosia.

Interestingly, despite findings that the RB, SS, and AP were slower than controls on tasks such as FTT and simple reaction time, these individuals were often not significantly slower than controls when viewing Cue and Target faces. Further, a common theme in scanning patterns seen between individuals in the PCA group appears to be an emphasis on areas of higher contrast (e.g., border areas; Forehead, Right Cheek). Further, RB's and SS's focus on single features (e.g., Forehead and Nose, respectively), suggests a difficulty disengaging from these areas, which fits with recent accounts of simultanagnosia (e.g., Carota & Calabrese, 2011). Another common feature seen in the scanning patterns among the individuals with PCA was an emphasis on face regions that provide less second-order configural information (e.g., SS and AP - Right Cheek; RB - Forehead), or decreases in time spent viewing areas that do hold second-order configural information (e.g., SS - Eyes). Second-order configural information can be obtained from the relationship between features (e.g., area between the eyes and the nose). In studies that use eye tracking (e.g., Le et al., 2008), healthy populations often make fixations to second-order regions (Caldara et al. 2005).

CHAPTER 3

Viewing Window: Restricted-view task

Experiment 1 revealed novel information about how scan paths produced by PCA patients and healthy controls differ when unrestrictedly viewing a face. Experiment 2 was designed to extend these findings by utilizing the newly developed Viewing Window paradigm (Baugh & Marotta, 2007) to explore the differences in scan path that are produced when these groups have a restricted view of a face and are asked to move a small focus-window over a computer screen to complete a face perception task. By using a focus-window, which limits a viewer to a small portion of an image at a time, it was expected that participants would use a

parts-based, or piecemeal processing strategy. Both prosopagnosia and simultanagnosia are characterized by problems in global, but not parts-based processing. Further, previous research (e.g., Dalrymple et al. 2010; Dalrymple et al. 2011) have shown that healthy individuals' scanning patterns are altered in a way that resembles those of individuals with simultanagnosia when images are presented in a restricted viewing condition.

Past research has often used methods other than eye tracking to recording scanning patterns when faces are presented. Similar to eye tracking, this research has found evidence of atypical scanning patterns in individuals with prosopagnosia. Caldara et al. (2005) found results that are very similar to traditional eye tracking, using 'bubbles'; clear, circular regions that were randomly presented within images of blurred faces, so that a face became clear in certain areas, restricted by the size of the bubble. Healthy control participants were most accurate when bubbles were presented in the eye and nose region, while PS, an individual with prosopagnosia, was more accurate when the bubbles were presented in the mouth region, as well as external areas.

James et al. (2010) showed that even when healthy participants were thought to be limited to a sequential, or parts-based, processing strategy, the upper internal features of faces were still most often used in a face-matching task. For this experiment, faces were occluded by a solid mask and could be seen only through a small 24x24 pixel aperture. The aperture could be moved around on a computer screen using button-presses, allowing the participants to explore a face. Despite the fact that only a small portion of a face was visible at any given time, forcing a sequential integration of features, the scanning patterns were similar between the aperture view condition, and an unrestricted view condition.

However, when healthy participants viewed social scenes using both a mouse-contingent and a gaze-contingent window, Dalrymple et al. (2011) found that scanning strategies changed; fixations became less in the eye region, and increased in areas of high contrast. Mouse- and Gaze-contingent window paradigms limit participants' view of an image to a small window presented on a computer screen, which moves along with their the movement of the computer mouse, or the participants' eyes, respectively. Thus, vision is restricted to a small portion of an image at any one time. Using these paradigms, scanning patterns in healthy individuals were found to closely mimic those seen in individuals with simultanagnosia whose vision was not restricted with a window. Errors made by healthy individuals using the restricted window also became similar to those made by individuals with simultanagnosia. An earlier study by Dalrymple and colleagues (2010) found that restricting vision to the gaze-contingent window in a Navon hierarchical letters task (Navon, 1977) lead healthy individuals to make disproportionate errors to global forms. These results support the idea that simultanagnosia arises from a restricted window of visual attention.

Experiment 2

Experiment 2 utilized a paradigm, the newly developed *Viewing Window* task, which is similar to the mouse-contingent window used by Dalrymple et al. (2011). Previous research where participants used a restricted window paradigm for viewing faces (e.g., James et al. 2010) has shown that scanning patterns in healthy individuals follow a pattern similar to that found in eye tracking studies. In contrast, research with Navon letters and social scenes that contain many faces, has found that restricting visual information alters scanning patterns in healthy individuals in a way that resembles scanning patterns of simultanagnosia (e.g., Dalrymple et al. 2010, 2011). The individuals in the PCA group all experience simultanagnosia, and most experience problems

in face perception, which suggests a lack of configural, or global, processing. Experiment 2 sought to examine the effects of a restricted viewing condition on scanning patterns in PCA and healthy individuals. It was expected that, in healthy control, the the RV task would cause a switch from holistic processing to a more parts-based, serial processing approach. In contrast, each individual in the PCA group were not expected to show an obvious switch in processing, due to problems in global processing, or simultanagnosia. Thus, it was expected that, while healthy controls would show scanning patterns that indicate local processing, the individuals in the PCA group would show a different pattern.

The *Viewing Window* task, is software, developed in-house, that presents blurred images on a computer screen along with a small (1.3cm in diameter), user controlled, focus-window to display the underlying image with normal clarity. A touch-sensitive computer screen allows participants to move the ‘viewing window’ by moving a stylus on the screen. For this experiment, the *Viewing Window* task was meant to record information regarding the strategies that are used to identify faces (e.g., regions of the face that are viewed during identification), similar to the goals of eye tracking. Researchers have found that gaze patterns closely match scan paths of a focus-window, when each is recorded simultaneously (Dalrymple et al. 2007; Baugh & Marotta, 2008).

Materials and Methods

Participants. The entire PCA group (RB, SS, AP, MTB, and PLH) was recruited for this experiment, along with the same undergraduate control participants. One new participant was recruited for Experiment 2 after one participant withdrew from the study following Experiment 1. Thus, three of the four aged participants that participated in Experiment 2, had participated in Experiment 1 (3 females, all right handed, Mean age=73).

Stimuli. A separate set of stimuli, from the same database, was prepared in the same way as Experiment 1. In order to determine that the cue faces could not be recognized without the use of the focus-window, the cue face stimuli were first copied and then distorted using a Gaussian blur set at a level of 35 in Corel Photopaint (Corel Corporation, 2008). This procedure resulted in two distinct versions of each face; one clear and one blurred. The level of blur was determined in a pilot study, where individuals were asked to complete a 2 alternative forced choice task in which participants were asked to match a blurred cue face to one of two target faces. With a blur level of 35, participants produced correct responses that were no greater than chance and participants felt uncertain about the accuracy of their responses when asked to judge the confidence in their decisions following the experiment.

Importantly, the blur level for the present study was approximately double that of blur levels used in previous research that has utilized Gaussian blur to examine featural and configural processing (see Collishaw and Hole, 2000; Lobmaier et al., 2008; Lobmaier and Mast, 2008; Schwaninger et al., 2002). Those experiments required subjects to identify blurred faces without the use of a focus-window. Gaussian blur was chosen instead of a solid mask (e.g. James et al., 2010), because the blur allows for some information regarding the overall location and dimension of the face – which may assist participants in selecting a path for the focus-window.

Materials.

The Viewing Window. The outermost region of the focus-window displayed the image in full blur, while the innermost region displayed the image clearly, with a smooth transition between the two regions. The focus-window was presented on a touch-sensitive screen, and controlled via a stylus held in participants' right hand, placed under their index finger. The

touchscreen has the benefit of allowing for a 1-1 correspondence between the participant's physiological movement, and the resultant movement of the window over the presented face.

All participants were tested individually at a station consisting of a laptop PC, seated approximately 50 cm from the monitor. All stimuli were displayed on a 15" touch-sensitive monitor running at a resolution of 1280x800 at 60 Hz. To avoid any positional biases, the focus-window did not appear until the participant touched the stylus to the screen.

Procedure. The participants received both written and verbal instructions, followed by a demonstration by the experimenter as to the correct use of the touchscreen and focus-window. To begin the experiment, a single blurred face was presented at the centre of the computer screen. As soon as the blurred face was visible, the participants were required to touch the stylus to the screen, and to explore the underlying image with the focus-window. The remainder of the procedure was identical to Experiment 1, except that the PCA patients gave verbal responses that were entered by the experimenter.

Results

Aged vs. Young Controls

Errors. Comparisons between control groups were conducted using a two-tailed independent-samples t-test, with all results significant at $p < .05$. Comparisons between individuals in the PCA groups and control groups were conducted using a 95% confidence interval. There were no significant differences in accuracy found when aged ($M=83.04\%$, $SD=6.76$) and young ($M=88.69\%$, $SD=10.42$) controls were compared ($t(14)=-.246$, $p < .05$, 2-tailed)¹.

Viewing Time.

Cue stimuli. Significant differences in VT were found when aged ($Mdn=21630$, $SD=17786$) and young ($Mdn=7935$, $SD=6695$) controls were compared ($t(14)=2.337$, $p<.05$). Aged controls spent significant more time viewing cue faces.

Target stimuli. There were significant differences in VT when aged ($Mdn=8454$, $SD=7670$) and young ($Mdn=2527$, $SD=2962$) controls were compared ($t(14)=2.325$, $p<.05$). Aged controls spent significantly more time viewing target faces.

¹ The d' results presented in Table 4 again suggest bias did not play a role in the responses of the participants.

Table 7

Same-different signal detection paradigm

	Stimuli=Different	Stimuli=Same	
AGED	d'	2.12 d'	2.12
	c	0.065 c	-0.065
YOUNG	d'	2.421 d'	2.421
	c	-0.13 c	0.13
RB	d'	-0.553 d'	-0.553
	c	0.276 c	-0.276
SS	d'	0.176 d'	0.201
	c	-0.088 c	0.1
AP	d'	-0.729 d'	0.176
	c	0.188 c	0.088
MTB	d'	0.194 d'	0.194
	c	0.455 c	-0.455
PLH	d'	-0.447 d'	0
	c	0.582 c	-0.806

Scanning Pattern. Stimuli were divided in the same ROIs from Experiment 1. See Figure 8 and Figure 9 for descriptive statistics for both control groups. Both window movement and the duration that the window was in an ROI were included in the analysis. As in Experiment 1, the stimuli were divided into 8 ROIs and the percentages were calculated for each ROI. In order to better reflect the fact that manual movements, rather than eye-movements, control the Viewing Window, the term PWM (percentage window movement) was used to define the frequency the window was present in each ROI and PWD (percent window duration) was used to define the proportion of time the window was present in each ROI. Nevertheless, all percentages were calculated in the same way as Experiment 1 (see Figure).

Movement. There were no significant differences in PWM between control groups, across ROIs.

Duration. There were no significant differences in PWD between control groups, across ROIs. Both the aged and young controls showed significant differences across ROIs (see Table 8 and Table 9). Due to the overall lack of significant differences found between aged and young controls, further discussion, and comparison to individuals in PCA group will be limited to the aged control group (see Figure 8 and Figure 9).

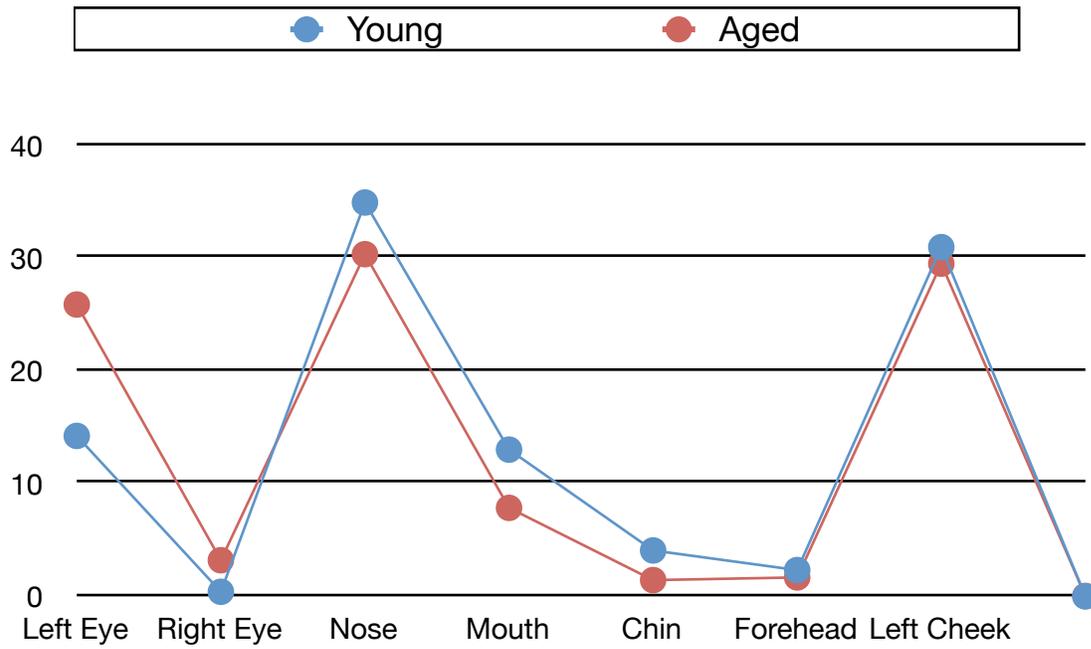
Figure 8. PWM for Aged vs. Young controls.

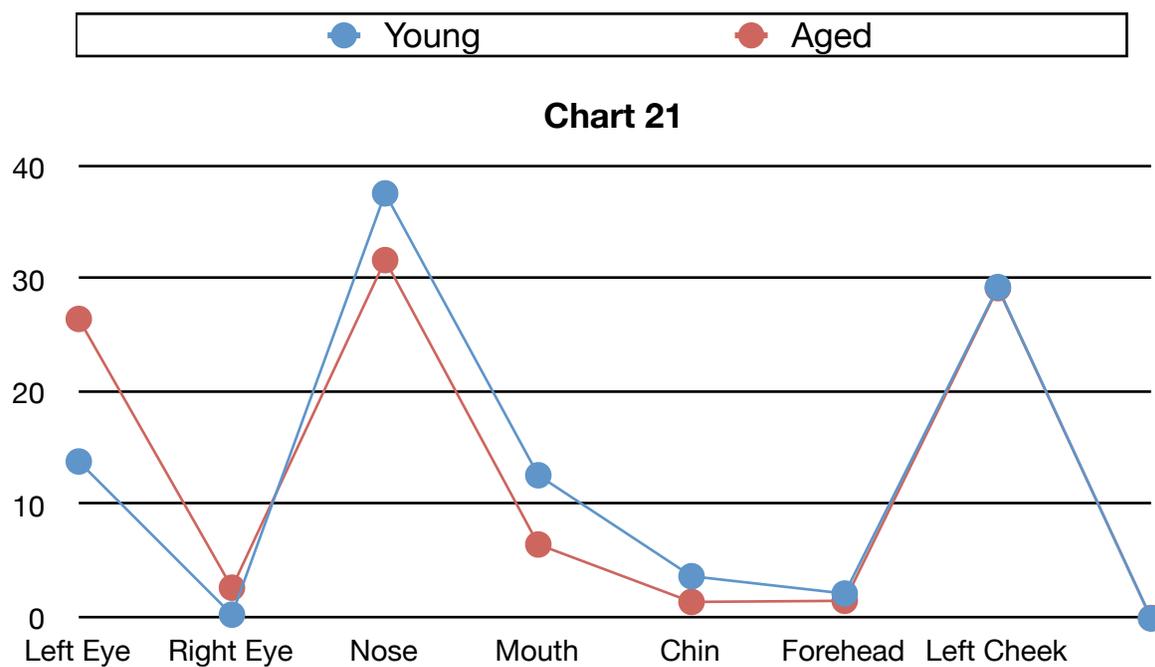
Figure 9. PWD for Aged vs. Young controls.

Table 8

Restricted-view task: Aged Controls Paired-samples t-tests for Differences Between ROIs

ROI	Left Eye	Right Eye	Nose	Mouth	Chin	Forehead	Left Cheek	Right Cheek
Left Eye	14.19	0.36**	39.41**	11.14	3.45**	1.92**	29.24**	0**
Right Eye	14.19**	0.36	39.41**	11.14**	3.45	1.92	29.24**	0
Nose	14.19	0.36	39.41	11.14**	3.45**	1.92**	29.24**	0**
Mouth	14.19	0.36	39.41	11.14	3.45**	1.92**	29.24**	0**
Chin	14.19**	0.36	39.41**	11.14	3.45	1.92	29.24**	0
Forehead	14.19**	0.36	39.41**	11.14**	3.45**	1.92	29.24**	0
Left Cheek	14.19**	0.36**	39.41**	11.14**	3.45**	1.92**	29.24	0**
Right Cheek	14.19**	0.36	39.41**	11.14**	3.45	1.92	29.24**	0

*All numeric values represent PWD. All significant differences are $p < .001$ ** and $p < .002$ **.

Table 9

Restricted-view task: Young Controls Paired-samples t-tests for Differences Between ROIs.

ROI	Left Eye	Right Eye	Nose	Mouth	Chin	Forehead	Left Cheek	Right Cheek
Left Eye	26.56	2.73	31.78	6.56*	1.47*	1.57*	29.3	0.04*
Right Eye	26.56	2.73	31.78**	6.56	1.47	1.57	29.3	0.04
Nose	26.56	2.73**	31.78	6.56	1.47**	1.57*	29.3	0.04**
Mouth	26.56*	2.73	31.78	6.56	1.47	1.57	29.3	0.04
Chin	26.56*	2.73	31.78**	6.56	1.47	1.57	29.3	0.04
Forehead	26.56*	2.73	31.78*	6.56	1.47	1.57	29.3	0.04
Left Cheek	26.56	2.73	31.78	6.56	1.47	1.57	29.3	0.04
Right Cheek	26.56*	2.73	31.78**	6.56	1.47	1.57	29.3	0.04

*All numeric values represent PWD. All significant differences are $p < .001$ ** and $p < .002$ *.*

The restricted-view (RV) task is meant to limit individuals to a serial parts-based processing strategy, a strategy that is associated with deficits in recognition of familiar faces. Research suggests that healthy individuals are highly accurate at matching novel faces, even when they are limited to either configural (e.g., blurred faces; Lobmaier et al. 2008) or featural (e.g., scrambled faces) information. The aged and young controls show similar performance, with few errors, and scanning patterns, where both groups spent a notable amount of time viewing the Left Eye, Left Cheek, and the Nose ROI. However, the aged controls spent significantly more time viewing both Cue and Target faces, while maintaining the same VT pattern as young controls; VT to Target stimuli was typically shorter than VT to Cue stimuli. The results suggest that the aged and young controls are using similar strategies when viewing faces, but that the aged controls process each face more slowly. Also, the RV task is thought to limit participants to featural information, and this limitation may have required more time for memorization and comparison in the aged control group, who tended to experience more visual problems (e.g, more likely to need glasses) and has less experience using a computer. Due to the overall similarity between control groups, only the aged controls were included in comparisons to the individuals in the PCA group, the same as in Experiment 1.

Importantly, the Left Eye, Left Cheek, and Nose ROIs are all located on the right side of the computer screen, or right visual field. One reason for the increase in scanning in the ROIs could be that the emphasis on parts-based, or local processing. The RV task is thought to limit participants to a parts-based approach because configural information cannot be acquired easily using the Viewing window. Parts-based processing is associated with the left hemisphere (refs), and the healthy participants show a bias toward the right visual field in Experiment 2. The bias toward the right visual field could be due to an increased reliance on parts-based processing from

the left hemisphere. Another possible explanation for the right visual field bias could be that the healthy participants were largely right-handed, which places the right visual field closer to the hand that is used to manipulate the stylus.

RB.

Errors. Compared to healthy older participants ($M=85.38\%$, $SD=7.76$, $CI [73.03\%, 97.73\%]$), RB (39.29% correct) made significantly more errors¹.

Viewing Time.

Cue Stimuli. Compared to the age-matched controls ($N=4$, $Mdn=21630ms$, $SD=17786$, $95\% CI [0, 49920]$), RB ($Mdn=13527ms$) did not produce significantly longer VT.

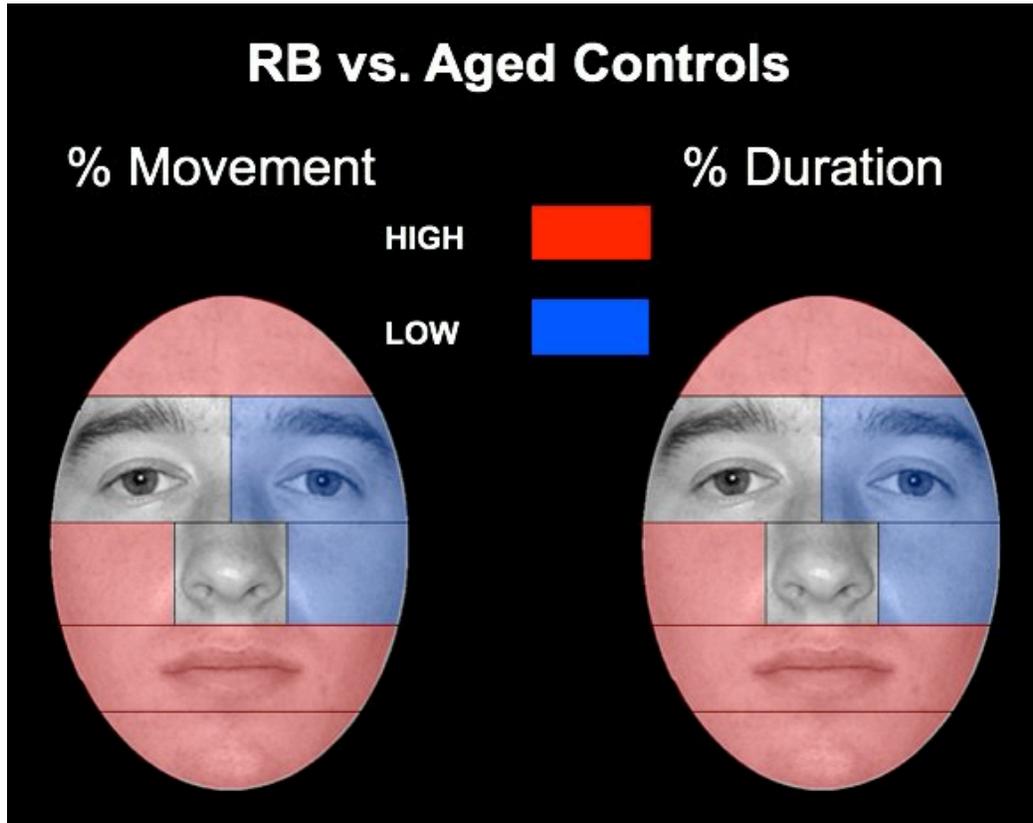
Target stimuli. Compared to the age-matched controls ($N=4$, $Mdn=8454ms$, $SD=7670$, $95\% CI [0, 20658]$), RB ($Mdn=6796ms$) did not produce significantly longer VT.

Scanning Pattern.

Movement. Compared to the age-matched controls, RB produced a significantly higher PWM in the Mouth ($M=21.60\%$, $95\% CI [1.43\%, 14.37\%]$), Chin ($M=7.03\%$, $95\% CI [0\%, 3.90\%]$), Forehead ($M=5.88\%$, $95\% CI [0\%, 5.22\%]$) and Right Cheek ($M=3.48\%$, $95\% CI [0\%, 0.16\%]$), and significantly lower PFC in the Left Eye ($M=15.36\%$, $95\% CI [20.01\%, 30.77\%]$) and Left Cheek ($M=13.47\%$, $95\% CI [16.82\%, 42.22\%]$) (See Figure 10).

Duration. Compared to the age-matched controls, RB produced a significantly longer PWD in the Mouth ($M=21.73\%$, $95\% CI [1.21\%, 11.92\%]$), Chin ($M=6.72\%$, $95\% CI [0\%, 3.78\%]$), Forehead ($M=5.75\%$, $95\% CI [0\%, 5.27\%]$) and Right Cheek ($M=3.29\%$, $95\% CI [0\%, 0.15\%]$), and significantly shorter PWD in the Left Eye ($M=15.14\%$, $95\% CI [18.78\%, 34.35\%]$) and Left Cheek ($M=14.96\%$, $95\% CI [15.61\%, 42.98\%]$) (See Figure 10).

Figure 10. PWM and PWD for RB compared to aged controls.



Stimuli divided into ROIs. Red indicates ROIs where RB showed significantly higher percentages, while blue indicate where RB showed significantly lower percentages, compared to aged controls.

Compared to aged controls, RB produced more errors, and showed differences in her scanning pattern. RB's errors are not surprising, considering RB's problems in global processing (i.e., simultanagnosia) and face perception (see Table 2 and Table 3, see Case descriptions). The fact that RB showed high error, and differences in scanning patterns suggests that RB is not using a feature-by-feature strategy. A feature-by-feature strategy is thought to lead to correct responses in a face-matching task with novel faces.

RB's scanning pattern appears to reflect simultanagnosia (e.g., RB focused on a single component when asked to copy simple figures, and would attempt to identify an object based on a single detail; see Case Descriptions). Signs of simultanagnosia are seen in RB's scanning patterns in that RB spent significantly more time viewing border areas (e.g., Chin, Right Cheek, and Forehead) that do not hold configural or featural information, and less time viewing areas that healthy individuals tend to view most (e.g., Left Eye). Previous research suggests that simultanagnosia is characterized by increased viewing of high-contrast areas, and a decrease in viewing the eye region when viewing faces in social scenes (e.g., Dalrymple et al. 2010).

If RB's impairments were limited to face perception, it would be expected that RB's scanning pattern reflect a feature-by-feature strategy. However, RB's scanning pattern does not show obvious similarities to scanning patterns that are associated with prosopagnosia (e.g., focus on Mouth, individual features, external features). Although RB did spend more time viewing the Mouth ROI, it is possible that RB was more focused on the edge of the ROI that also borders with the background, creating a high contrast area.

There were no differences in VT, and, similar to aged controls, RB's VT was shorter for Target faces compared to Cue faces. These results are interesting because Experiment 2 requires manipulation of a stylus, and RB shows slowed motor and/or processing speed (see Table 2).

The lack of differences in VT for the RV task lend support to the idea that RB's performance is affected differently than aged controls, due to simultanagnosia. That is, the results suggest that RB's VT was less affected by the viewing window of the RV task; RB's VT did not increase substantially (e.g., $Mdn=12,490$ and $Mdn=13,527$), as did the aged controls (e.g., $Mdn=5,603$ and $Mdn=21,630$). If RB already experiences a restricted window of visual attention due to simultanagnosia, while healthy individuals' strategies might be reduced to featural or parts-based processing, RB's strategy is likely less affected.

SS.

Errors. Compared to healthy older participants ($M=85.38\%$, $SD=7.76$, $CI [73.03\%, 97.73\%]$), SS (57.14% correct) made significantly more errors¹.

Viewing Time.

Cue stimuli. Compared to the age-matched controls ($N=4$, $Mdn=21630ms$, $SD=17786$, 95% $CI [0, 49920]$), SS, ($Mdn=13829ms$) did not produce significantly longer VT.

Target stimuli. Compared to the age-matched controls ($N=4$, $Mdn=8454ms$, $SD=7670$, 95% $CI [0, 20658]$), there were no significant differences in VT for SS ($Mdn=4787ms$).

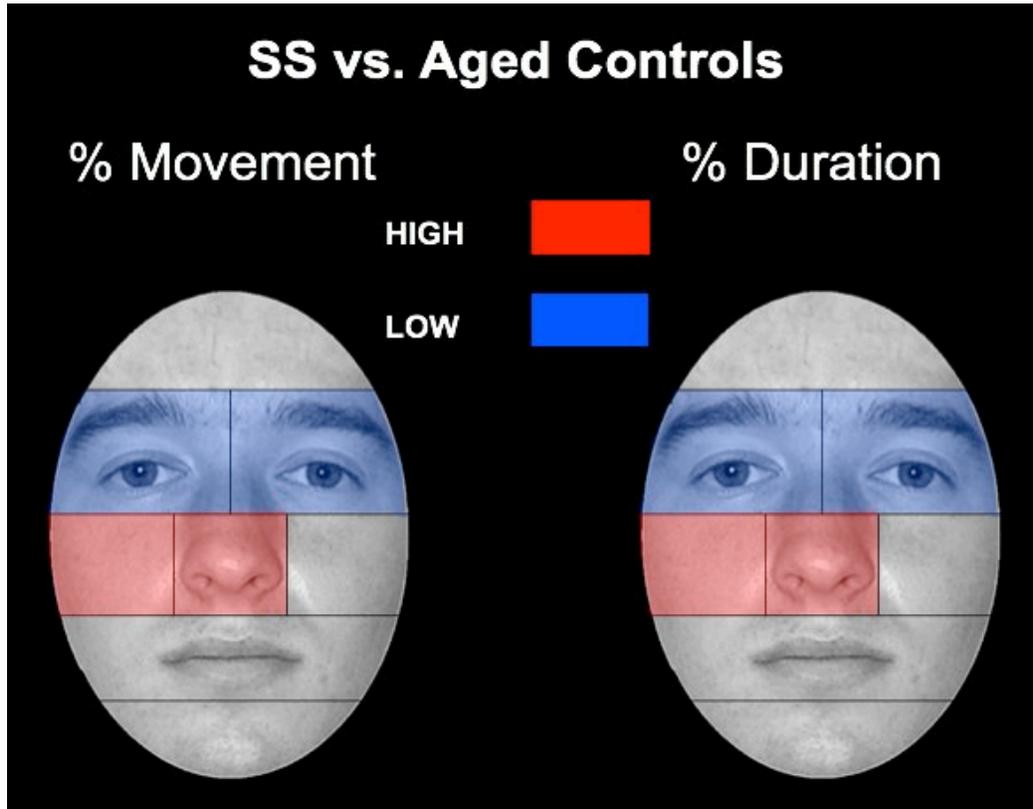
Scanning Pattern.

Movement. Compared to the age-matched controls, SS produced a significantly higher PWM in the Nose ($M=60.99\%$, 95% $CI [27.29\%, 33.43\%]$), and the Right Cheek ($M= 1.01\%$, 95% $CI [0\%, 0.16\%]$), and showed significantly lower PWM in for the Left Eye ($M= 11.90\%$, 95% $CI [20.01\%, 31.77\%]$), and Right Eye ($M=0.68\%$, 95% $CI [1.12\%, 5.30\%]$) (see Figure 11).

Duration. Compared to the age-matched controls, SS produced a significantly longer PWD in the Nose, ($M=63.49\%$, 95% $CI [27.66\%, 35.90\%]$), and the Right Cheek ($M= 0.94\%$, 95% $CI [0\%, 0.15\%]$), and showed significantly shorter PWD in for the Left Eye ($M= 11.92\%$,

95% CI [18.78%, 34.35%]), and Right Eye ($M=0.59%$, 95% CI [0.82%, 4.64%]) (see Figure 11).

Figure 11. PWM and PWD for SS compared to aged controls.



Stimuli divided into ROIs. Red indicates ROIs where SS showed significantly higher percentages, while blue indicate where SS showed significantly lower percentages, compared to aged controls.

Compared to aged controls, SS produced more errors, and showed differences in scanning pattern, yet there were no differences in VT. SS's errors are not surprising, considering that SS shows problems in global processing (i.e., simultanagnosia) and some difficulty in face perception (see Case descriptions). Also, SS experiences visual neglect of the lower quadrants (see Case Descriptions) which may interfere with SS's ability to view areas of faces that are helpful for matching. It is assumed that SS's scanning patterns are mainly the result of simultanagnosia, in addition to neglect.

SS's greater emphasis on the Right Cheek ROI is suggestive of the effects of simultanagnosia. SS's scanning pattern within the Right Cheek fell mainly along the border between the Right Cheek and the black background, where a high degree of contrast exists. Individuals with simultanagnosia have been found to make fewer fixations to the eye region, and more fixations to areas of high contrast (e.g., Dalrymple et al. 2011).

SS's performance is likely a combination of components of both dorsal and ventral simultanagnosia. SS was more impaired at object counting than object and face identification, SS has shown a focus on single details within objects, and appears unaware of the overall image. Both problems could affect SS's ability to perceive faces; SS might be unable to perceive more than one feature, and have difficulty perceiving more than a single detail within features.

SS's performance also shows signs of visual neglect, in that SS rarely views ROIs below the Nose ROI. In fact, SS spends the most time in the central area of the face, rarely viewing areas such as the Mouth or Chin, or even the the eye ROIs. SS's focus on the Nose and Right Cheek ROIs are in line with SS's neurologist's diagnosis of visual neglect, with a focus in the lower quadrants. The lower regions of the face may fall within these areas of neglect. Further, neglect of the lower regions of the face may interfere with SS's ability to engage in feature-by-

feature processing.

Overall, SS's performance appears to reflect a lack of configural or featural processing. SS's emphasis on the Nose ROI is not interpreted as configural processing because SS spent significantly less time looking at both eye ROIs. If SS was engaged in configural processing, SS would be expected to show a scanning pattern that emphasized the areas between the Nose and Eye ROIs, including the Eye ROIs. SS's emphasis in this single ROI shows that SS is not viewing each feature in a serial strategy. SS may not be able to use a feature-based strategy, and would have to based matching on a single feature or a single detail. The different viewing conditions that exist between the Cue (restricted) and Target (open) faces may interfere with any feature- or detail-matching strategies that SS may use. The differences between SS and controls in errors and scanning pattern are unlikely to be the result of memory problems because SS appears to be unable to obtain an adequate initial percept of faces.

AP.

Errors. Compared to healthy older participants ($M=85.38\%$, $SD=7.76$, CI [73.03%, 97.73%]), AP (35.71% correct) made significantly more errors¹.

VT.

Cue stimuli. Compared to the age-matched controls ($N=4$, $Mdn=21630ms$, $SD=17786$, 95% CI [0, 49920]), AP ($Mdn=29390ms$) did not produce significantly longer VT.

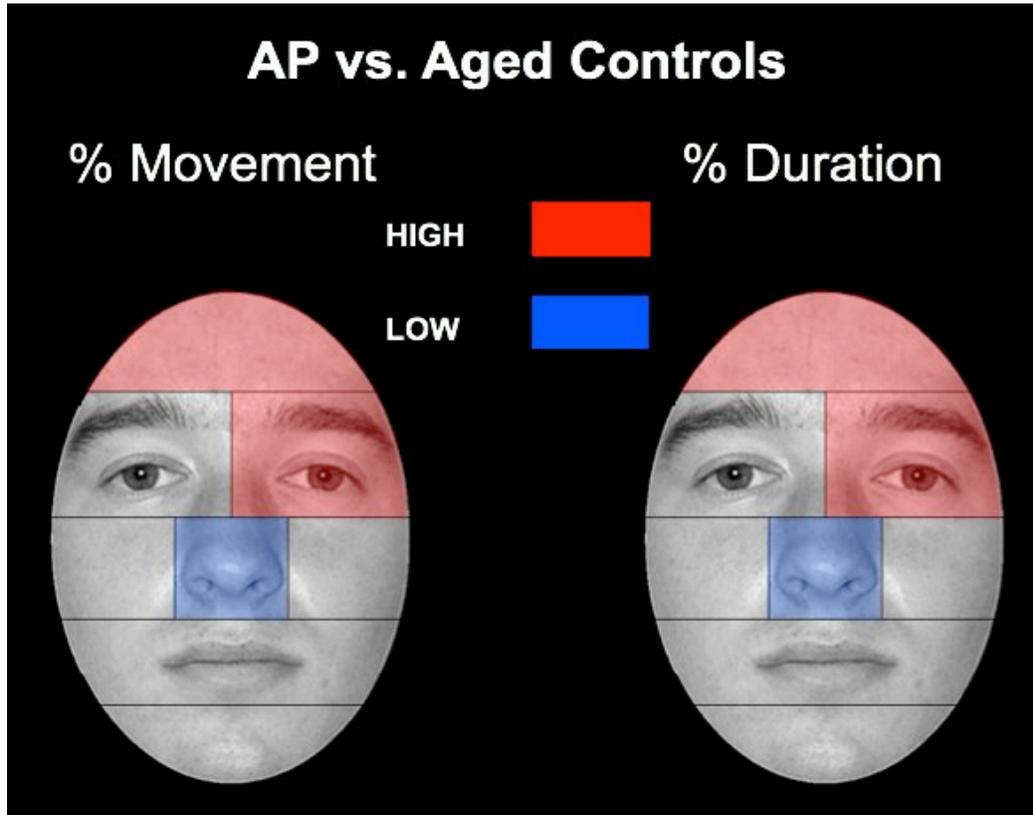
Target stimuli. Compared to the age-matched controls ($N=4$, $Mdn=8454ms$, $SD=7670$, 95% CI [0, 20658]), there were no significant differences in VT for AP ($Mdn=10438ms$).

Scanning Pattern.

Movement. Compared to the age-matched controls, AP showed significantly higher PWM in the Left Eye ($M=40.46\%$, 95% CI [20.01%, 31.77%]), Forehead ($M=14.03\%$, 95% CI [0%, 5.22%]), with significantly lower PWM to the Nose ($M=16.43\%$, 95% CI [27.29%, 33.43%]) (see Figure 12).

Duration. Compared to the age-matched controls, AP showed significantly longer PWD in the Left Eye ($M=40.75\%$, 95% CI [18.78%, 34.35%]), Forehead ($M=12.78\%$, 95% CI [0%, 5.27%]), with significantly lower PWM to the Nose ($M=17\%$, 95% CI [27.66%, 35.90%]) (see Figure 12).

Figure 12. PWM and PWD for AP compared to aged controls.



Stimuli divided into ROIs. Red indicates ROIs where AP showed significantly higher percentages, while blue indicate where AP showed significantly lower percentages, compared to aged controls.

AP showed significant differences in error rates, and scanning pattern, while there were no differences in VT. AP's higher rate of errors are not surprising, considering that AP experiences simultanagnosia, optic ataxia, and mild hemispatial neglect, all of which might interfere with perception of each face. AP was found to have relatively preserved face and object perception (see Table 2 and Table 3), so it is assumed that AP's error rates and differences in scanning patterns are the result of a combination of dorsal simultanagnosia, hemispatial neglect, optic ataxia, and other motor issues.

In addition to higher error rates, AP spent more time viewing areas such as the Left Eye and Forehead, and less time viewing the Nose ROI. These results are somewhat surprising when one considers AP's did not show impairments on tests of face perception (see Table 2 and Table 3). However, AP's scanning patterns are suggestive of simultanagnosia; AP shows an emphasis on high contrast areas, including the border along the Forehead.

AP's errors and scanning pattern in Experiment 2 were likely related to a combination of hemispatial neglect, optic ataxia, and motor issues in that AP would have been limited in the number of ROIs that she could perceive. That is, AP may not have been able to attend to certain areas of the faces that would have aided in accurate matching. For example, AP viewed the Nose ROI, an area associated with accurate matching and configural processing in healthy individuals, significantly less than controls. AP may have spent less time in the Nose ROI due to the Nose falling outside of the intact areas of visual attention.

AP's scanning would have been hindered by the visually guided action necessary to complete Experiment 2; participants were required to use a stylus to move the Viewing window on the computer screen. Importantly, AP chose to use her non-dominant (right) hand due an injury to her left arm. Thus, AP's errors and scanning patterns in Experiment 2 are interpreted as

the result of a combination of problems such as simultanagnosia, in addition to a significant effect of visual neglect, optic ataxia, and manual motor issues.

MTB.

Errors. Compared to healthy older participants ($M=85.38\%$, $SD=7.76$, $CI [73.03\%, 97.73\%]$), MTB (55.56%) made significantly more errors¹.

Viewing Time.

Cue stimuli. Compared to the age-matched controls ($N=4$, $Mdn=21630ms$, $SD=17786$, 95% $CI [0, 49920]$), MTB produced significantly longer VT ($Mdn=74510ms$).

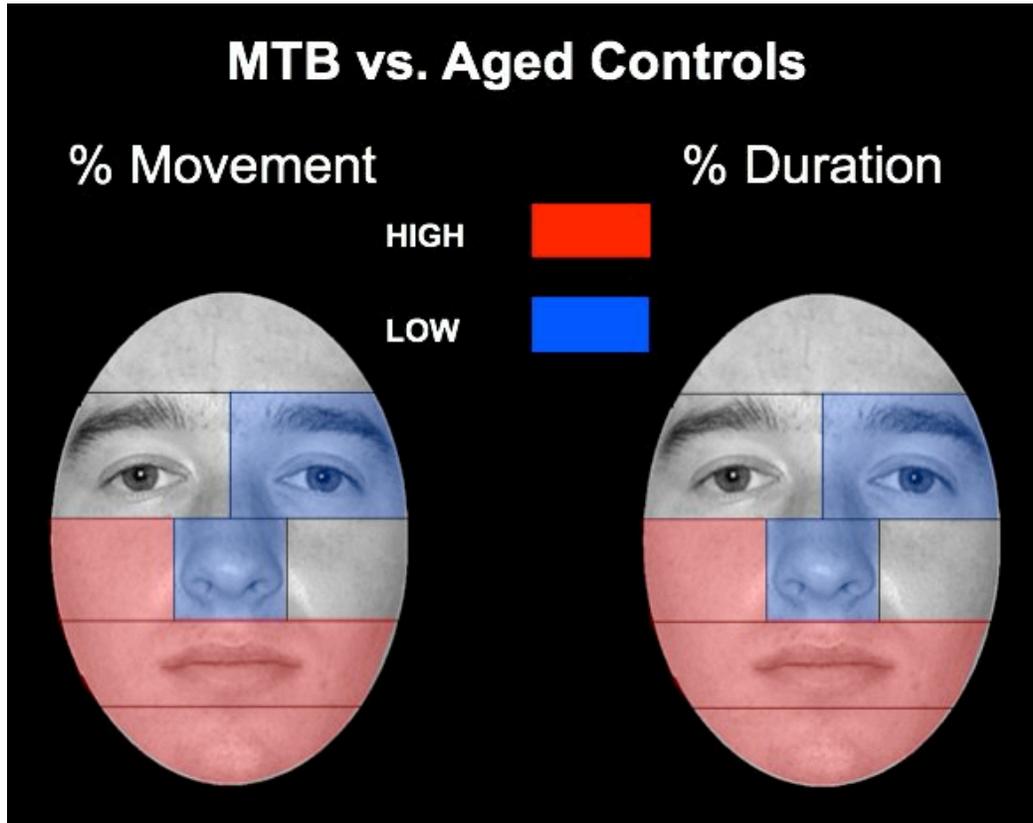
Target stimuli. Compared to the age-matched controls ($N=4$, $Mdn=8454ms$, $SD=7670$, 95% $CI [0, 20658]$), there were no significant differences in VT for MTB ($Mdn=11065ms$).

Scanning Pattern.

Movement. Compared to the age-matched controls, MTB showed significantly higher PWM to the Mouth ($M=27.60\%$, 95% $CI [1.43\%, 14.27\%]$), Chin ($M=15.03\%$, 95% $CI [0\%, 1.9\%]$), and Right Cheek ($M=2.76\%$, 95% $CI [0\%, 0.16\%]$), with significantly lower PWM to the Left Eye ($M=16.51\%$, 95% $CI [20.01\%, 31.77\%]$) and the Nose ($M=15.73\%$, 95% $CI [27.29\%, 33.43\%]$) (see Figure 13).

Duration. Compared to the age-matched controls, MTB showed significantly longer PWD to the Mouth ($M=28\%$, 95% $CI [1.29\%, 11.92\%]$), Chin ($M=14.99\%$, 95% $CI [0\%, 3.78\%]$), and Right Cheek ($M=2.11\%$, 95% $CI [0\%, 0.15\%]$), with significantly shorter PWD to the Left Eye ($M=16.56\%$, 95% $CI [18.78\%, 34.35\%]$) and the Nose ($M=15.52\%$, 95% $CI [27.66\%, 35.90\%]$) (see Figure 13).

Figure 13. PWM and PWD for MTB compared to aged controls.



Stimuli divided into ROIs. Red indicates ROIs where MTB showed significantly higher percentages, while blue indicate where MTB showed significantly lower percentages, compared to aged controls.

MTB, who is thought to experience the effects of both dorsal and ventral subtypes of PCA, was unable to participate in the OV task of Experiment 1 due to an inability to maintain fixation during the calibration phase. Compared to aged controls, MTB produced more errors, longer VT to Cue faces, and showed differences in scanning pattern. MTB's pattern of results are thought to be most affected by problems in motor coordination, neglect, as well as dorsal simultanagnosia. The results are interpreted as the effects of problems such as slowed motor speed, as well as impairments such as simultanagnosia, visual neglect, optic ataxia, and apraxia (see Case descriptions). Although MTB showed signs of simultanagnosia, the potential effects may not be displayed in MTB's scanning patterns due to the likely effects of visual neglect (see Case Descriptions), and problems such as optic ataxia. Thus, even in areas where MTB's visual neglect did not prevent attention, MTB's scanning pattern was probably affected by difficulty manipulating the stylus, and inaccurate reaching.

MTB is also known to experience simultanagnosia, which is associated with a focus on areas of high contrast, and reduced viewing of the eye region (e.g., Dalrymple et al. 2011). MTB's scanning pattern shows a focus on ROIs that contain the high-contrast border that lies between the face and a black background. MTB's performance appears to reflect a lack of configural processing, and very little featural processing. MTB spent more time looking at the Mouth, Chin, and Right Cheek; the Mouth contains featural information, while the Chin and Right Cheek are non-featural ROIs. MTB also spent less time viewing areas that are often viewed most by healthy individuals; the Left Eye and Nose.

Previous research suggests that novel faces are matched successfully when either featural or configural information is present. Therefore, if MTB was engaged in featural processing, MTB's error rates would be lower. The different viewing conditions that exist between the Cue

(restricted) and Target (open) faces may interfere with any feature- or detail-matching strategies that MTB may use. Also, the fact that only the Cue face is viewed The different viewing conditions that exist between the Cue (restricted) and Target (open) faces may interfere with any feature- or detail-matching strategies that MTB may use. Based on a neurologists assessment, MTB was determined to experience mild memory problems (see Case Descriptions), which may have increased errors, alongside MTB's obvious difficulty perceiving faces.

PLH.

Errors. Compared to healthy older participants ($M=85.38\%$, $SD=7.76$, $CI [73.03\%, 97.73\%]$), PLH (42.86%) made significantly more errors¹.

Viewing Time.

Cue stimuli. Compared to the age-matched controls ($N=4$, $Mdn=21630ms$, $SD=17786$, 95% $CI [0, 49920]$), PLH ($Mdn=21060ms$), did not produce significantly longer VT.

Target stimuli. Compared to the age-matched controls ($N=4$, $Mdn=8454ms$, $SD=7670$, 95% $CI [0, 20658]$), PLH ($Mdn=14850ms$), did not produce significantly longer VT.

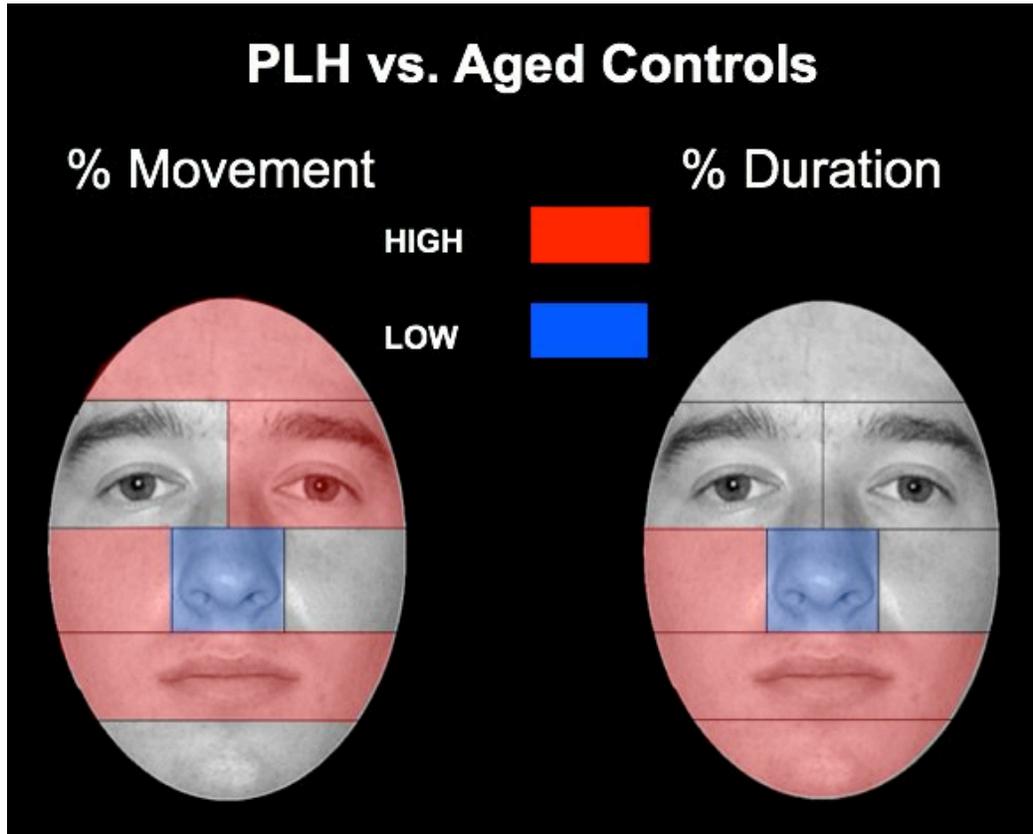
Scanning Pattern.

Movement. Compared to the age-matched controls, PLH showed significantly higher PWM to the Left Eye ($M=34.72\%$, 95% $CI [20.01\%, 31.77\%]$), Mouth ($M=20.93\%$, 95% $CI [1.43\%, 14.27\%]$), Forehead ($M=5.26\%$, 95% $CI [0\%, 5.22\%]$), and Right Cheek ($M=0.28\%$, 95% $CI [0\%, 0.16\%]$), with significantly lower PWM to the Nose ($M=8.62\%$, 95% $CI [27.29\%, 33.43\%]$) (see Figure 14).

Duration. Compared to the age-matched controls, PLH showed significantly longer PWD to the Mouth ($M=21.61\%$, 95% $CI [1.21\%, 11.92\%]$), Chin ($M=3.84\%$, 95% $CI [0\%, 3.78\%]$),

and Right Cheek ($M=0.23\%$, 95% CI [0%, 0.15%]), with significantly shorter PWD to the Nose ($M=12.22\%$, 95% CI [27.66%, 35.90%]) (see Figure 14).

Figure 14. PWM and PWD for PLH compared to aged controls.



Stimuli divided into ROIs. Red indicates ROIs where PLH showed significantly higher percentages, while blue indicate where PLH showed significantly lower percentages, compared to aged controls.

Compared to aged controls, PLH produced more errors, and showed differences in scanning pattern. PLH's scanning pattern is interpreted as the effects of problems such as dorsal simultanagnosia, mild apraxia, optic ataxia, and visual neglect. PLH spent more time viewing ROIs such as the Left Eye, Mouth, Chin, Forehead, and Right Cheek, and PLH spent less time viewing with the Nose ROI.

PLH's scanning patterns may be affected by visual neglect in all four quadrants (see Case Descriptions), and extinction in the left visual field. That is, scanning may be limited to the areas of attention, and PLH's increased viewing of the Left Eye may be the result of extinction. Further, PLH might be better able to perceive details or features on the Cue face, due to the restriction of the Viewing window, however, extinction within the left visual field may hinder matching when the Target face is presented without occlusion. That is, with the Target face, both sides of the image are visible, potentially causing a loss of information with if extinction of the left side occurs.

Issues such as mild ataxia and optic ataxia would have also affected PLH's performance in Experiment 2. PLH's manipulation of the stylus was affected, and her ability to move the stylus to areas she chose to view would have been impaired, in addition to problems attending to visual stimuli. Similar to MTB, PLH did not participate in Experiment 1 due to problems maintaining fixation during the calibration phase. Again, the extent of the effects of optic ataxia and ataxia on scanning is difficult to determine without a comparison from Experiment 1, where no stylus was necessary. PLH's scanning patterns and errors were subject to PLH's ability to attend to visual fields, as well as her ability to manipulate, and guide, the stylus.

In addition to problems in visual fields and movement, PLH's scanning patterns are suggestive of the effects of simultanagnosia. That is, PLH shows a focus on areas that are largely

without configural information, or even featural information (e.g., Mouth, Chin, Forehead, Right Cheek). Simultanagnosia, likely interferes with both configural and featural processing by introducing an extreme focus to salient areas (e.g., high contrast; Dalrymple et al. 2011), in addition to an inability in the perception or awareness of multiple details (Farah, 2004; Carota & Calabrese, 2011). Individuals with simultanagnosia have been found to make fewer fixations to the eye region, and more fixations to areas of high contrast .

Interestingly, despite being much slower than aged controls on both a FTT and a simple reaction time task, which suggest slowed motor responses and processing speed (see Table 2), PLH did not produce longer VT compared to controls. Similar to most of the other individuals in the PCA group, PLH's lack of difference in VT compared to aged controls, despite signs of slowed processing (e.g., slower than aged controls on both a FTT and a simple reaction time task, which suggest slowed motor responses and processing speed), is interpreted as a reduced effect of the RV task due to PLH's already significant perceptual and motor problems. That is, the RV task is thought to interfere with configural processing. However, PLH may not engage in configural processing due to simultanagnosia, which may remove some effect of the RV task on VT.

Discussion.

In the RV task of Experiment 2, all participants in the PCA group produced significantly more errors and showed significant differences in scanning patterns. However, most of the individuals in the PCA group did not show increased VT compared to aged controls. Although there were no clear similarities in scanning patterns between the individuals in the PCA group, a common theme was seen in relatively reduced viewing of both configural and featural information (e.g. chin, cheeks and forehead), and emphasis on the border areas of each face.

Open- vs. Restricted View. To better understand any effects of the RV condition, comparisons were made between measures of proportion of viewing duration from Experiment 1 (PFD) and Experiment 2 (PWD). These comparisons were made only for duration because fixation counts were not recorded in the RV task of Experiment 2. To examine any differences in viewing time between the OV and RV tasks, a difference score was computed for each ROI by subtracting PWD from PFD. Next, a series of one-sample t-tests were conducted to determine if any of these differences scores were significantly different from '0' (i.e., no change in percentage of duration between tasks). In order to make comparisons with the PCA group, 95% confidence intervals were generated from the control difference score data for each ROI. This analysis will compare how individuals with PCA differ from their controls under each of these viewing conditions. Does the Viewing Window influence the PCA group differently than the healthy controls?

Age-matched Controls. A series of one-sample t-tests, corrected for multiple comparisons using the Bonferroni procedure, were conducted to determine if there were any significant changes in percentage of duration for each ROI, between tasks. All significant observations had a corrected probability of $p < .0062$. There were significant differences in percentage of duration between the OV and RV tasks: Right Eye ($M1=25.24\%$; $M2=2.73\%$, $Mdiff=22.51\%$, $SD=5.38$; $t(3)=23.999$, $p < 0.001$), Nose ($M1=17.96\%$; $M2=31.78\%$, $Mdiff=-13.83\%$, $SD=3.75$; $t(3)=18.971$, $p < 0.001$), and the Left Cheek ($M1=1.84\%$; $M2=29.30\%$, $Mdiff=-27.45\%$, $SD=7.97$; $t(3)=16.755$, $p < 0.001$) (see Figure 15).

In the RV task, aged controls spent more time viewing the Nose and Left Cheek, and spent less time viewing the Right Eye, compared to the OV task (see Figure 15). The aged controls appear to be using a parts-based processing strategy in Experiment 2. That is, the aged

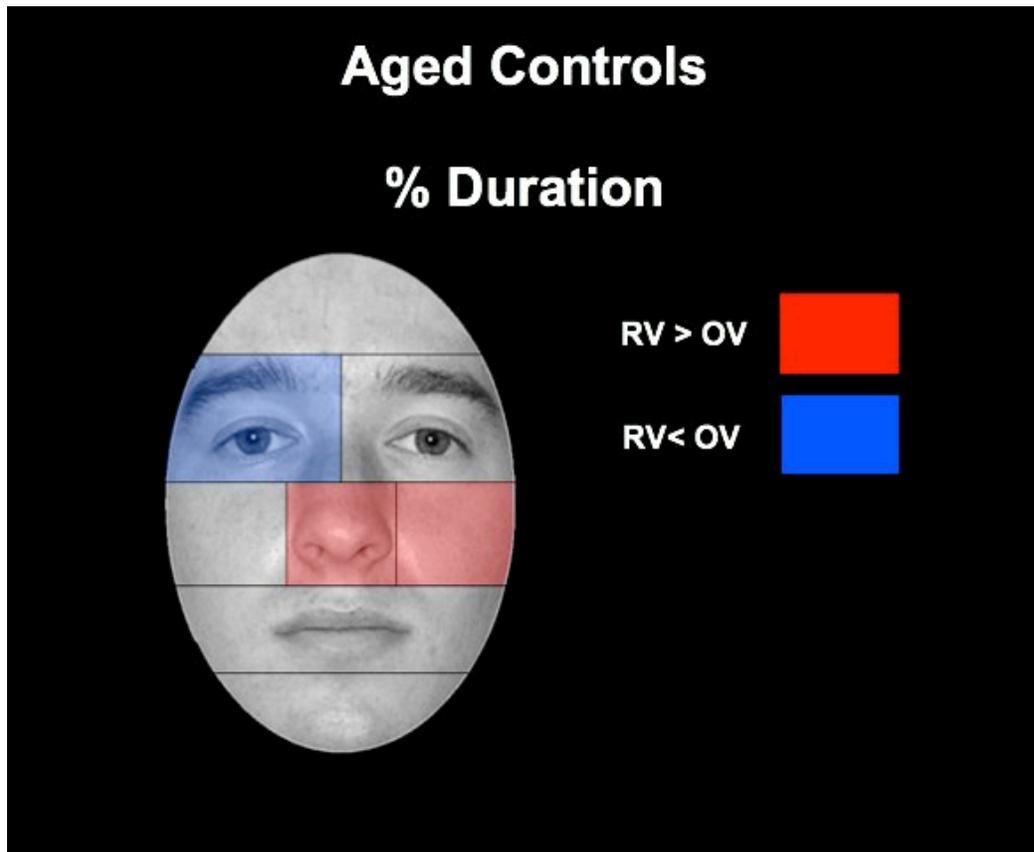
controls show a decrease in the LVF (i.e., Right Eye), and an increase in the RVF (Left Cheek), which is associated with local-processing. Part of this apparent RVF tendency could be due to the fact that the left portion of the face is located in closer proximity to participants' dominant right-hand.

Overall, the controls' scanning pattern in the OV task was characterized by scanning that was concentrated within the Eye, Nose, and Mouth, respectively. Scanning patterns seen in the RV task were characterized by scanning that was concentrated within the Nose, Left Cheek, and Left Eye, respectively. These results suggest that the change in task required a change in processing strategy from holistic to parts-based processing. However, the scanning pattern in the RV task still indicates the use of some configural information in that the aged controls appear to move between the Left Eye, Nose, and Left Cheek. That is, the relationship between the most viewed ROIs in the RV task likely provides second-order information. These results fit with Dalrymple and colleagues (2011) suggestion that healthy individual's processing strategies become more restricted when using a viewing window paradigm. Additionally, the evidence for some configural processing supports research by James et al. (2010) who found that scanning patterns did not change overall, between an open and restricted viewing task. Overall, the aged controls increased viewing within the Nose and Left Cheek, but decreased viewing in every other ROI, with the greatest decrease seen in the Right Eye.

Scanning patterns in the open- and RV conditions were compared within each control group to determine if there were any effects of viewing condition. However, the same statistical tests were could not be carried out for each individual in the PCA group because a standard error cannot be calculated for each individual. The following sections outline the descriptive analysis of each patient's scanning pattern, and compares changes to the degree of change seen in the

aged control group vs. individuals in the PCA group on a case-by-case basis. Tests of significance compare the degree of difference within each ROI to the differences found in the aged controls' scanning pattern. As with the control groups difference scores, values were calculated by subtracting RV durations from OV durations, for each ROI. Therefore, negative values indicate an increased duration in the RV task, and positive values indicate decreased duration in the RV task. For example, a difference score of -21.03% in the Nose ROI indicated that participants spent more time viewing the Nose in the RV task, compared to the OV task.

Figure 15. Aged Controls % Duration Differences in Change between Open- and Restricted-view tasks.

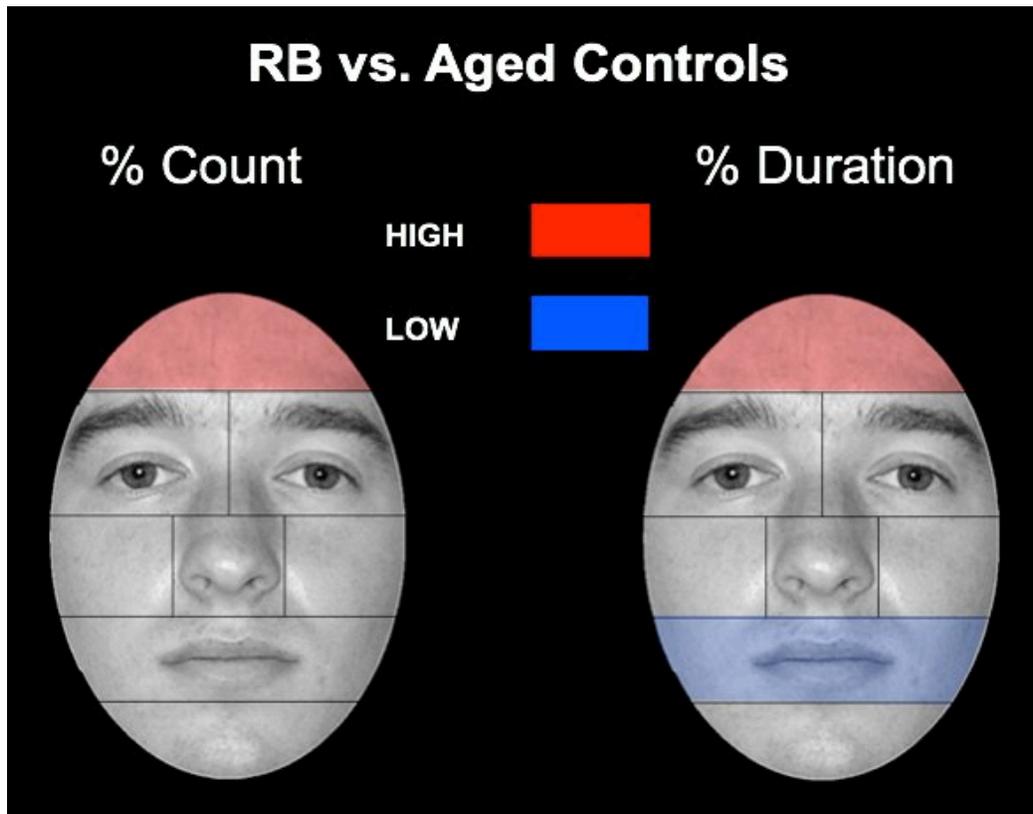


Red indicates ROIs where aged controls produced significant differences in % duration change that were increases in viewing. Blue indicates significant differences in % duration change that were decreases in viewing.

RB. Compared to aged controls, changes in RB's scanning pattern between the open- and RV tasks differed significantly in the following ROIs: Mouth (-17.13%), Chin (-6.36%), Forehead (16.62%), Left Cheek (-13.18%), and Right Cheek (-2.66%) (see Figure 16). Further, the descriptive statistics of RB's overall pattern of differences indicate that, in the RV task, RB spent more time viewing Nose, Mouth, Chin, Left Cheek, and Right Cheek, spent less time viewing the Left Eye, Right Eye, and Forehead. In the OV task RB's scanning pattern was concentrated within the eye ROIs, followed by the Forehead, Nose, Mouth. RB spent the least amount of time viewing the Chin and cheek ROIs.

RB showed similarities to the aged control in her relative emphasis in the Left Eye and Left Cheek, however, RB's emphasis on the Left Cheek was significantly less than aged controls. RB showed a greater focus on the Mouth ROI in Experiment 2, while the controls tended to spend the most time in the Left Eye, Left Cheek, and Nose. Overall, RB appears to switch to viewing along the lower portions of the face in the RV task.

Figure 16. RB % Count and Duration Differences in Change between Open- and Restricted-view tasks.

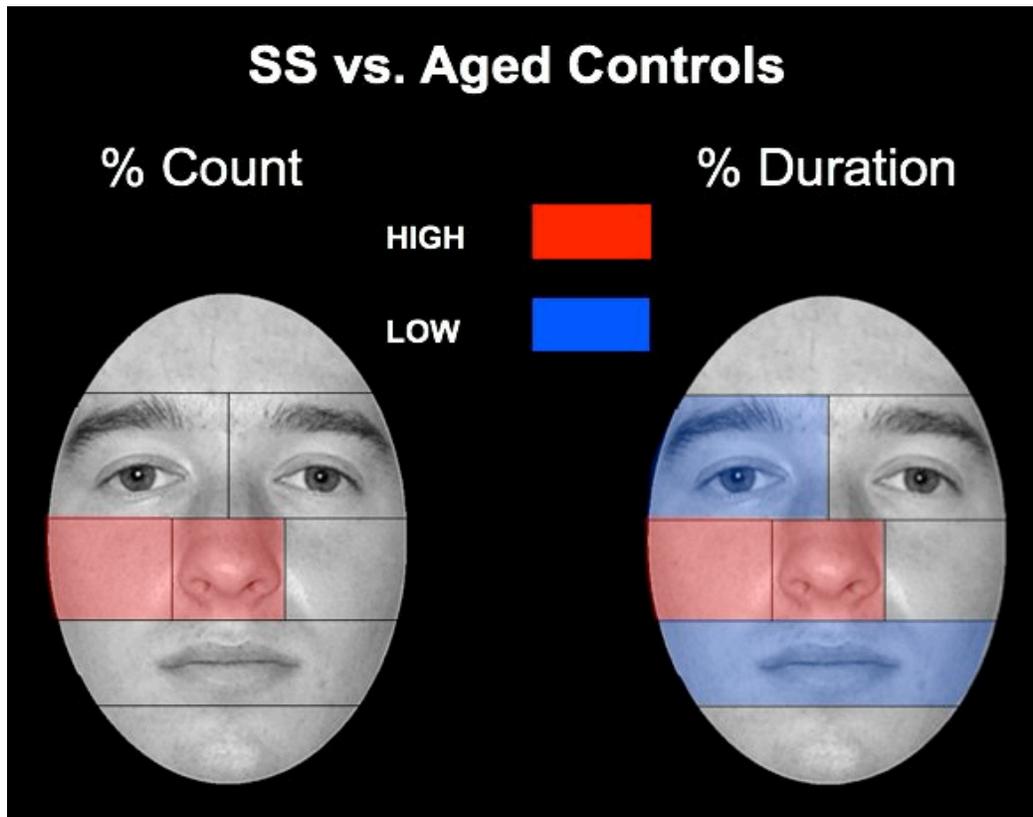


Red indicates ROIs where RB produced significant differences in % count and duration change that were increases in viewing. Blue indicates significant differences in % count and duration change that were decreases in viewing.

SS. Compared to the OV task, in the RV task, SS showed increased duration in the Nose and Left Cheek, and decreased duration in the Left Eye, Mouth, Forehead, and Right Cheek. There was no observed change seen in the Chin ROI; SS did not look at the Chin in either task. Changes in SS's scanning pattern between the open- and RV tasks showed significant differences in direction and/or degree compared to aged controls only within the Nose ROI (-5.60%) (see Figure 17), where SS's significant focus on the Nose increased.

SS showed the highest duration in the Nose across tasks, and increased duration to this ROI in the RV task. However, the aged controls showed higher increases to the Nose ROI in the RV task, compared to SS. SS viewed the eye ROIs less in the RV task, but followed the left-sided (RVF) emphasis that the aged controls employed in this task. However, SS's overall pattern in the RV task showed less viewing of central areas, aside from the Nose, which suggests that SS is using a different strategy than controls, and may be more focused on local details. It could be that SS is using an extreme form of local processing in that he becomes even more restricted to the Nose ROI in Experiment 2.

Figure 17. SS % Count and Duration Differences in Change between Open- and Restricted-view tasks.

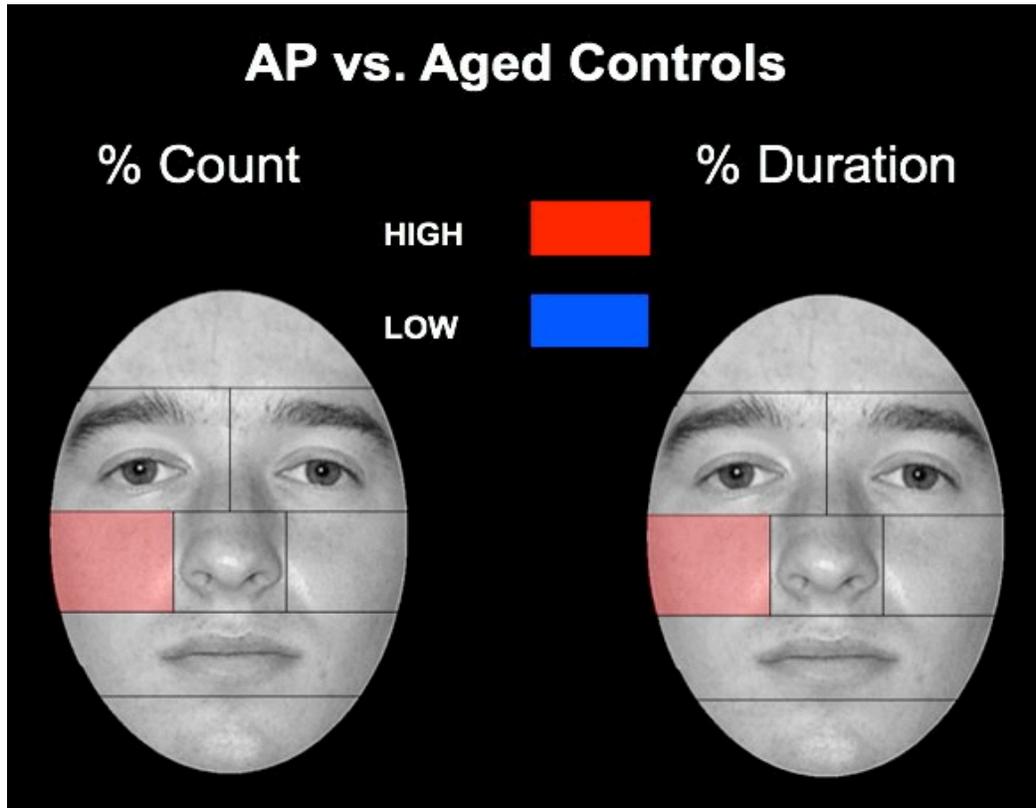


Red indicates ROIs where SS produced significant differences in % count and duration change that were increases in viewing. Blue indicates significant differences in % count and duration change that were decreases in viewing.

AP. Compared to the OV task, in the RV task, *AP* spent more time viewing the Left Eye, Chin, Forehead, and Left Cheek, and spent less time viewing the Right Eye, Nose, Mouth, and Right Cheek. Changes in *AP*'s scanning pattern between the open- and RV tasks showed significant differences in direction and/or degree compared to aged controls in the Nose (6.15%), Forehead (-4.99%), and Right Cheek (3.47%) (see Figure 18). These changes in *AP*'s scanning patterns may suggest a focus on peripheral areas (e.g., decrease on Nose, increase on Forehead),. Importantly, the changes between the OV and RV tasks, and the significant differences from aged controls are likely influenced by problems such as optic ataxia (e.g., *AP* showed misreaching), and *AP*'s injury and subsequent choice to use her non-dominant hand in the RV task.

In the OV task, *AP*'s overall scanning pattern was similar to aged controls, while *AP*'s scanning pattern in the RV task showed multiple differences compared to aged controls. *AP*'s lack of face perception deficits, or signs of simultanagnosia in scanning patterns of the OV task suggest that the differences seen in the RV task are related to optic ataxia, and manual motor problems.

Figure 18. AP % Count and Duration Differences in Change between Open- and Restricted-view tasks.



Red indicates ROIs where AP produced significant differences in % count and duration change that were increases in viewing. Blue indicates significant differences in % count and duration change that were decreases in viewing.

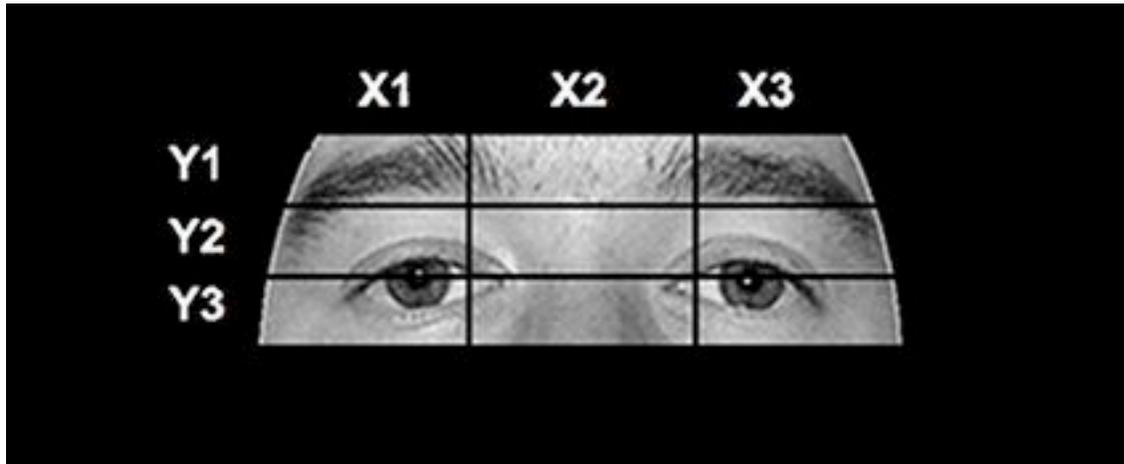
The differences in scan patterns between the open- and RVs suggest that the participants are processing faces in different ways under the two viewing conditions. Reduced time in the eye ROIs in the RV task may indicate that the control participants are relying less on holistic, or global, processing faces. Further, the reduction in the eye region is almost exclusively the result of reduced viewing within the Right Eye, which falls within the LVF, which is associated with local processing, as opposed to global processing. In addition, most of the participants were right-handed, resulting in closer proximity to the left portion of the faces. That is, if the stylus is held in the right hand, the left portion of the face is in closer range and presents an easy target for the stylus. In the RV task, the focus-window's aperture only allowed for a single feature to be viewed at one time, and differences in scanning patterns between the open- and RV tasks, across participants, suggest an effect of the type of task. The pattern of difference suggests that some individuals with PCA focused more on the lower regions of the face. For example, RB's decreases in duration for the eyes, and increases to the mouth and chin were significantly different than the changes seen in controls. The literature is mixed on these possibilities; some research suggests that scanning patterns would change, based on the change in available strategy (see Dalrymple et al. 2011). However, other researchers have found that scanning patterns were largely unaffected when visual scan paths, as opposed to manual visuomotor scan paths, were recorded using a gaze-contingent restricted viewing aperture (see James et al. 2010).

Eye ROIs. To gain a more detailed account of the scanning patterns of the individuals in the PCA group, we examined the eye region with a smaller set of ROIs. Previous research shows evidence of differences in fixation patterns to the eyes in cases of prosopagnosia (e.g., Barton et al., 2006; Bukach et al., 2008; Caldara et al. 2005; de Xivry et al., 2008; Le et al., 2003). More specifically, healthy individuals are known to view areas that provide second-order configural

information; fixations are typically made to the top of the nose, and scan paths move between the eyes and the nose. In contrast, prosopagnosia is associated with fixations to the eye region that fall on each eye, with very few fixations at the top of the nose, or scan paths that show movement between the nose and eyes. The scanning patterns in prosopagnosia are thought to reflect featural processing because fixations fall within each eye, rather than the spatial relationship of the eyes. In addition, simultanagnosia is associated with viewing of salient high-contrast areas (e.g., eyebrows, border areas) instead of looking at the eyes.

For the OV and RV tasks, the eye ROIs were recombined, and then parsed into three sections across the x- and y-axes. X ROIs were defined left to right, as x1, x2, and x3, respectively. Y ROIs were defined from lowest to highest y-value, as y1, y2, and y3, respectively. For example, x1 and y1 are the leftmost, and lowest ROIs, respectively (see Figure 19). All comparisons between control groups were made using independent-samples t-tests, corrected for multiple comparisons ($p=0.0042$) using the Bonferroni procedure. All comparisons between the aged controls and each individual with PCA were made using the 95% confidence intervals of the mean PFD and mean PWD for the aged controls.

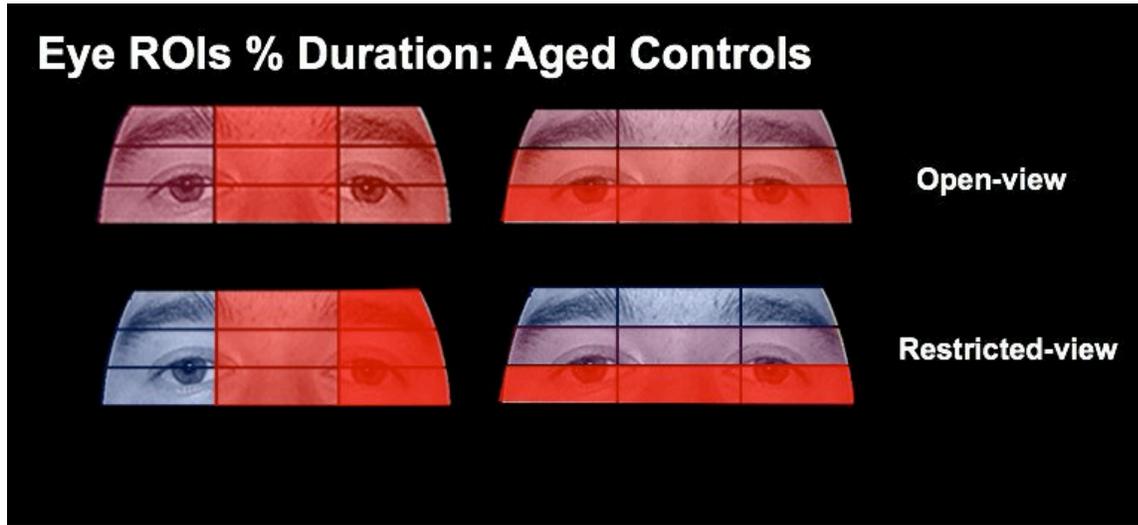
Figure 19. Eye ROIs - Divided across x- and y-axes.



Divisions between ROIs; x-axis ROIs are represented by the columns, and y-axis ROIs represented by the rows.

Aged vs. Young Controls. There were no significant differences within the x- and y-axis ROIs, across the open- and RV tasks. For the OV task. The aged controls spent the greatest amount of time viewing the central x ROI, as well as the y ROIs located closest to the eyes, y2 and y3 (see Figure 20). In contrast, in the RV task, the controls spent the most time viewing the x ROI located furthest in the RVF, x3, as well as the y ROI located closest to the eyes, y3 (see Figure 20). The overall pattern of differences in the open- and RV tasks suggests that healthy participants' scanning patterns may be more restricted when using the Viewing window; scanning patterns emphasis fewer ROIs, with a more concentrated count and durations in each. The remaining comparisons were conducted between the aged controls and each individual in the PCA group.

Figure 20. Mean PFD and PWD among Aged Controls in Open- and Restricted-view tasks.

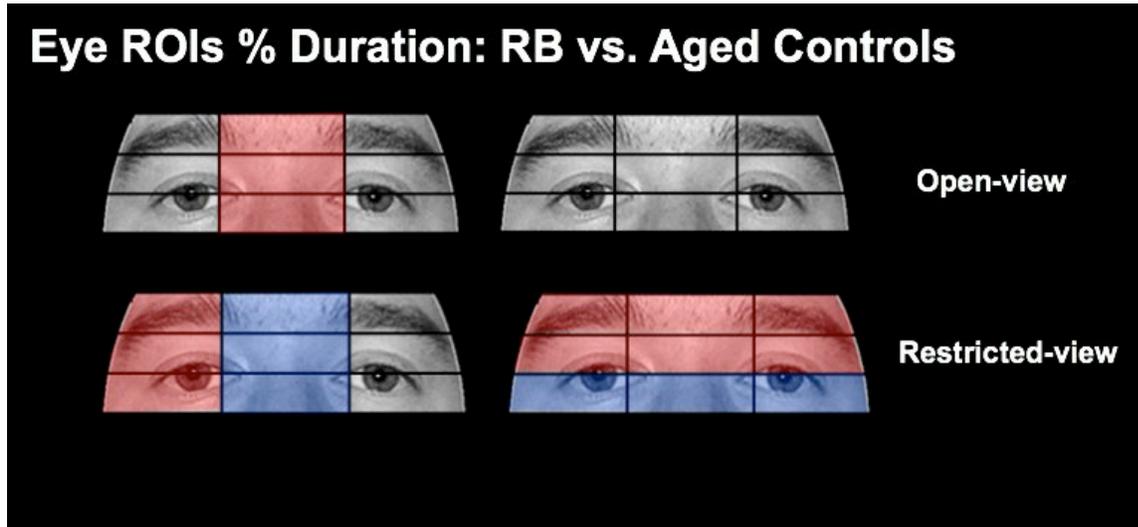


Red indicates higher % duration, and blue areas indicate lower % duration.

RB. Parsing of the eye ROIs revealed significant differences between *RB* and aged controls across both the x- and y-axes in Experiment 1 and Experiment 2. For the OV task, compared to aged controls, *RB* produced longer PFD in x2 ($M=46.43\%$, 95% CI [34.84%, 44.79%]), which shows that *RB* spent significantly more time viewing the centre of the eyes (see Figure 21). These results are interesting because *RB*, who shows significant problems in face perception, shows a scanning pattern that does not fit with previous research on impaired face perception. Previous research would suggest that *RB*'s scanning would be significantly reduced in the central region of the eyes.

For the RV task, *RB* produced significantly higher PWD in x1 ($M=0.81\%$, 95% CI [0%, 0.31%]), y1 ($M=18.71\%$, 95% CI [0%, 11.69%]), y2 ($M=35.80\%$, 95% CI [17.71%, 31.33%]). At the same time, *RB* produced significantly shorter PWD to x2 ($M=32.68\%$, 95% CI [33.34%, 44.68%]) and y3 ($M=45.5\%$, 95% CI [57.83%, 82.55%]). In all, *RB* spent significantly more time viewing the area that falls within the left visual field (right eye), as well as the mid-line, and top portion of the eye ROIs (see Figure 21). This pattern may indicate a parts-based approach compared to Experiment 1; in Experiment 2 *RB* shows increased viewing in areas of featural information and high contrast (e.g., right eye, border of right eye, eyebrows) and decreased viewing in areas of configural information (e.g., y ROIs at eye-level, central area).

Figure 21. Differences between RB and Aged controls in Open- and Restricted-view tasks.

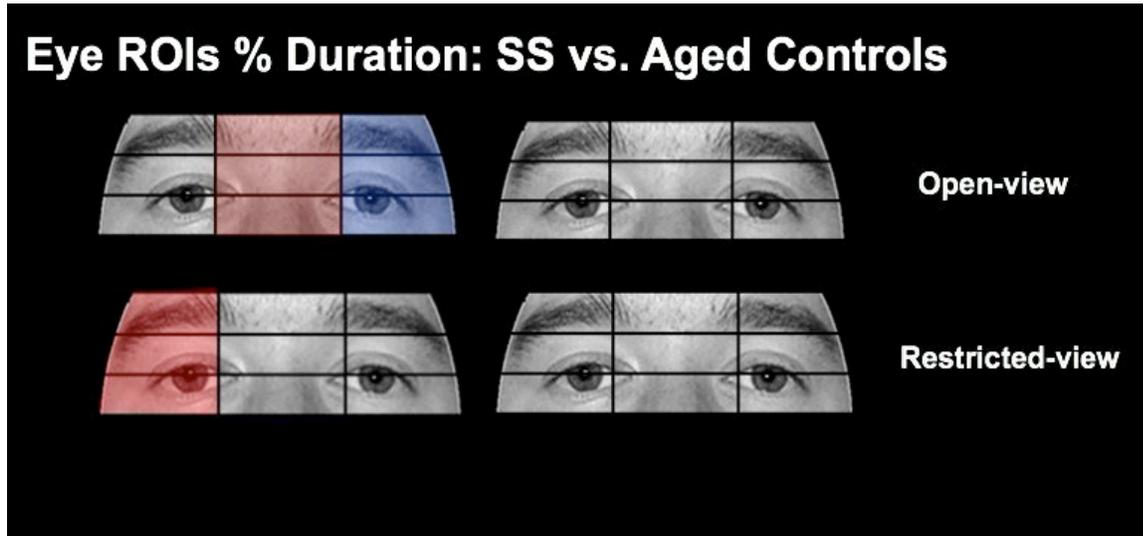


OV is displayed at the top, and RV is displayed at the bottom. The red area indicates areas that RB showed significantly longer PFD compared to aged controls.

SS. For the OV task, compared to aged controls, SS showed significantly longer PFD to x2 ($M=59.26\%$, 95% CI [34.84%, 44.79%]), and significantly shorter PFD to x3 ($M=16.3\%$, 95% CI [25.14%, 50.15%]) (see Figure 22). That is, SS spent more time viewing the area between the eyes, and less time viewing the portion of the eye ROIs that falls within the RVF. Similar to RB, SS may be engaged in holistic processing, however it is surprising that SS would view areas associated with holistic processing more than controls. Another explanation is that these results are influenced by SS's focus on the Nose ROI; SS's focus on the centrally located Nose ROI may have kept his gaze very close to the centre of each face, regardless of ROI.

For the RV task, SS showed significantly longer PWD x1 ($M=1.23\%$, 95% CI [0%, 0.31%]) compared to aged controls. SS did not show any other differences compared to aged controls, and SS's scanning pattern within the eyes may be interpreted as border-seeking. The Right Eye contains a border along the black background in each image, and provides a high degree of contrast. Further, the RV task is thought to encourage local processing, which is associated with the left hemisphere and scanning within the right visual field (RVF). SS does show more scanning within the RVF in the RV task, indicating that SS may be relying more on his left hemisphere when viewing faces in this task. Thus, the opposite pattern, relative to controls, within the parsed eye ROIs suggest a focus on contrast. Additionally, the increased LVF scanning, associated with global processing and right hemisphere structures, seen in SS is surprising considering that local processing is often associated with impaired face perception.

Figure 22. Differences between SS and Aged controls in Open- and Restricted-view tasks.



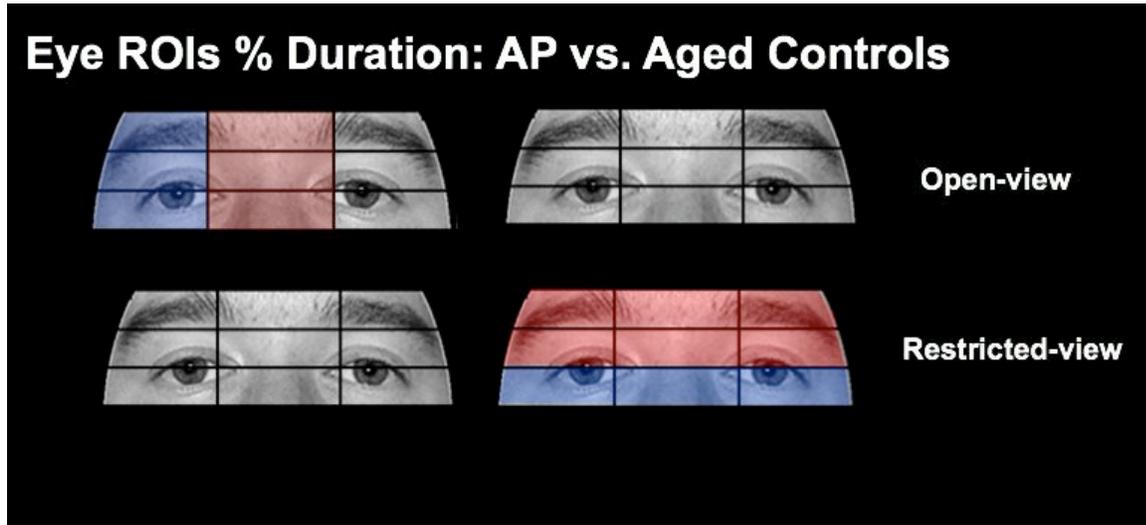
OV is displayed at the top, and RV is displayed at the bottom. The red area indicates areas that RB showed significantly longer PFD compared to aged controls.

AP. Compared to aged controls, in the OV task *AP* produced a significantly longer PFD to x2 ($M=53.81\%$, 95% CI [34.84%, 44.79%]), and shorter PFD to x1 ($M=10.12\%$, 95% CI [11.01%, 34.08%]). That is, *AP* spent less time viewing the right eye, and likely the border area near to the right eye, and more time looking at the area between the eyes. These results are not similar to the results of Experiment 1 overall; *AP* spent significantly more time viewing the Right Cheek. Together, these results might suggest that *AP* was viewing the border area along the right side of the face, but not obtaining important, featural or configural, information from the Right Eye. Importantly, *AP* shows more damage to left hemisphere structures (see Case Descriptions), and is left-handed, which may explain an emphasis on the x-axis ROI in the LVF. Despite showing the least problems in face perception (see Case Descriptions), *AP*'s scanning pattern suggests that *AP* was not viewing areas that would be helpful for processing configural information, or matching (e.g., border). However, in Experiment 1 *AP*'s overall scanning pattern showed the least differences compared to aged controls, and the differences seen in the eye ROIs are likely the result of an increased focus on high-contrast areas due to *AP*'s simultanagnosia.

For the RV task, compared to aged controls, *AP* produced a significantly longer PWD in y1 ($M=31.32\%$, 95% CI [0%, 11.69%]), and y2 ($M=33.69\%$, 95% CI [17.71%, 31.33%]), while *AP*'s PWD was significantly shorter in y3 ($M=35\%$, 95% CI [57.83%, 82.55%]) (see Figure 23). This pattern of differences suggest that *AP* showed a preference for the top and middle areas of the eye ROIs, which consist of the eyebrows, and area above the eyes. Importantly, *AP* spent less time viewing the y-axis ROI, y3, that contains the eyes. *AP*'s pattern in the RV task may suggest a focus on areas of higher contrast. *AP* may have also shown an increase in the uppermost, y1, ROI due to a focus on the Forehead ROI in Experiment 2, which was also interpreted as the result of the highly salient contrast along the border of the Forehead ROI. In all, based on the

emphasis on high-contrast as opposed to individual features, AP's scanning patterns within the parsed eye ROIs suggest the effects of simultanagnosia, across both experiments.

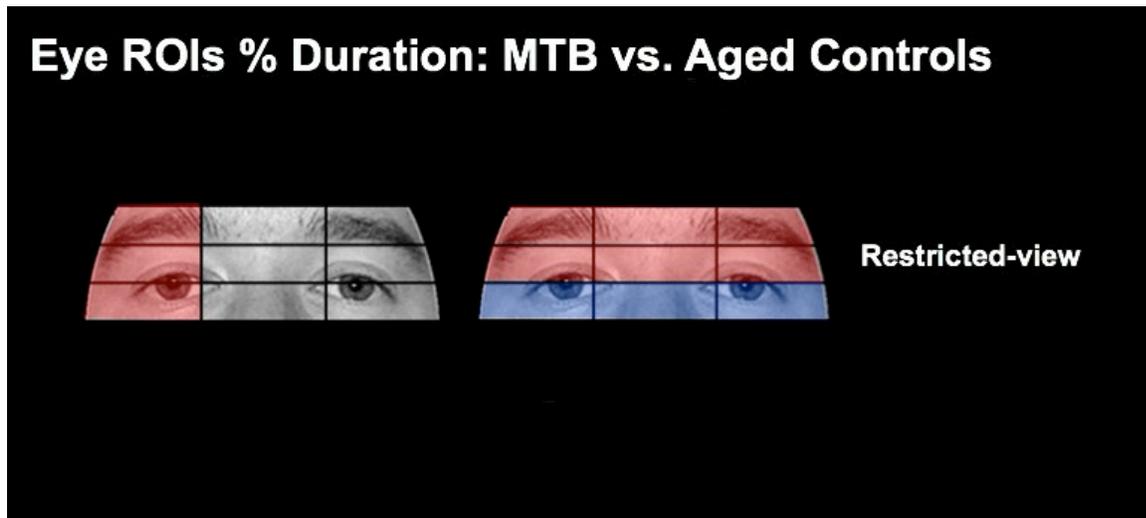
Figure 23. Differences between AP and Aged controls in Open- and Restricted-view tasks.



OV is displayed at the top, and RV is displayed at the bottom. The red area indicates areas that RB showed significantly longer PFD compared to aged controls.

MTB. Compared to aged controls, in the RV task, *MTB* produced significantly higher PWD in x1 ($M=3.77\%$, 95% CI [0%, 0.31%]), y1 ($M=17.55\%$, 95% CI [0%, 11.69%]), and y2 ($M=35.47\%$, 95% CI [17.71%, 31.33%]), and produced significantly shorter PWD in y3 ($M=46.98\%$, 95% CI [57.83%, 82.55%]) (see Figure 24). The fact that *MTB* spent more time in the outer x-axis ROIs could be due to a focus on high contrast border areas, indicating an effect of simultanagnosia. In addition to significantly longer PWD within the outer x-axis ROIs, *MTB* spent more time in the lowest y-axis ROI (closest to eye-level), which may suggest that *MTB* was viewing each eye separately, similar to previous research in impaired face perception. These results are not surprising considering *MTB*'s significant problems in face perception and simultanagnosia.

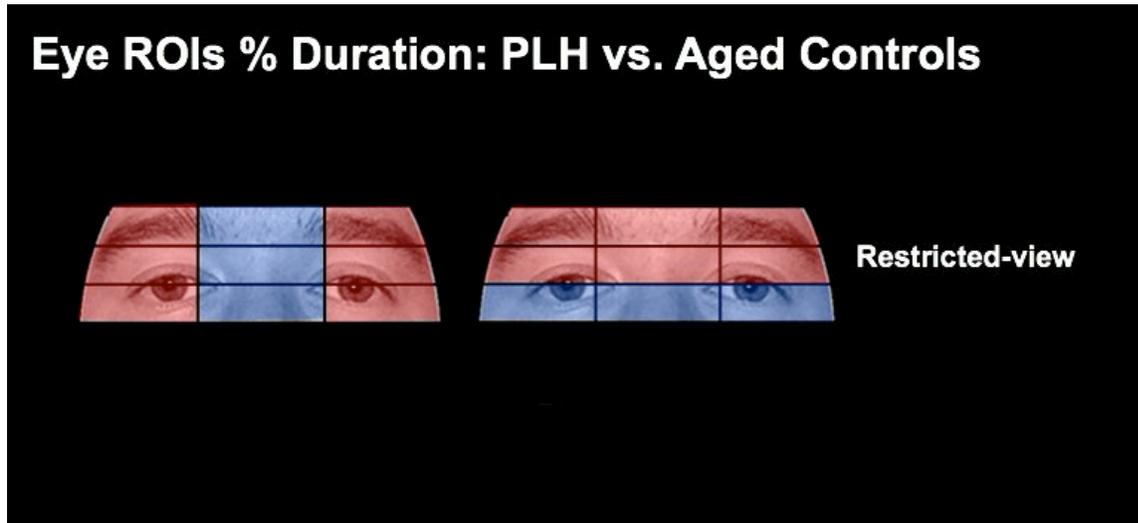
Figure 24. Differences between MTB and Aged controls in Open- and Restricted-view tasks.



OV is displayed at the top, and RV is displayed at the bottom. The red area indicates areas that MTB showed significantly longer PFD compared to aged controls.

PLH. Compared to aged controls, *PLH* showed significantly longer PWD in x1 ($M=0.93\%$, 95% CI [0%, 0.31%]), x3 ($M=73.9\%$, 95% CI [55.12%, 66.54%]), y1 ($M=23.16\%$, 95% CI [0%, 11.69%]) and y2 ($M=34.99\%$, 95% CI [17.71%, 31.33%]), and showed significantly shorter PWD for x2 ($M=25.17\%$, 95% CI [33.44%, 44.68%]) and y3 ($M=41.86\%$, 95% CI [57.83%, 82.55%]),(see Figure 25). These patterns suggest that *PLH* spent more time viewing areas of high contrast, and less time viewing areas that hold important configural information (e.g., area between the eyes). Further, *PLH* may not have obtained featural information from the x-axis ROIs, that contain each eye, considering that *PLH* focused on the upper areas near the eyebrows, as opposed to the eyes. In all, *PLH*'s scanning pattern within the eye ROIs appear to reflect the effects of simultanagnosia.

Figure 25. Differences between PLH and Aged controls in Open- and Restricted-view tasks.



OV is displayed at the top, and RV is displayed at the bottom. The red area indicates areas that PLH showed significantly longer PFD compared to aged controls.

The pattern of differences within the parsed eye ROIs suggest that the individuals in the PCA group processed the eye region of faces in a way that was different than aged controls across both the open- and RV tasks. In Experiment 1, there were significant differences found between each individual in the PCA group, compared to controls; RB and SS spent more time viewing the area at the centre of the eyes, while AP spent less time viewing this area. Both SS and AP spent more time viewing the x-axis ROI in the LVF, and is taken to indicate a focus on the high-contrast border area of the Right Eye. The eye region, and the area at the top of the nose are often cited as areas that are important for correct identification of faces. The fact that there were differences in the distribution of time between the parsed eye ROIs suggests that the individuals with PCA are missing on important configural information from this facial region.

CHAPTER 4

GENERAL DISCUSSION

Summary

This thesis was designed to more thoroughly investigate the perceptual deficits associated with PCA by recording scan paths in a face-matching task across two experiments: an open-view (OV) and a restricted-view (RV) task. In particular, we were interested in determining whether or not simultanagnosia contributes to impaired face perception in PCA. When looking at a face, healthy individuals spend more time looking at the eyes and nose, while individuals with prosopagnosia are often found to spend a disproportionate amounts of time looking at the mouth or on external features (Barton et al., 2006; Bukach et al., 2008; Caldara et al. 2005; de Xivry et al., 2008; Le et al., 2003). Individuals with simultanagnosia have been shown to spend less time looking at the eye-region and more time looking at areas of high contrast (Dalrymple et al. 2011).

Theories of prosopagnosia and simultanagnosia suggest that these visual disorders are characterized primarily by problems in configural, and global processing, respectively, with a reliance on parts-based or local processing. Global processing includes processing of visual information that encompasses both the parts, and the whole. For example, an image is identified as a face when the shapes inside (e.g., eyes, nose, mouth) are processed together as one overall image within and the larger shape (e.g., oval that makes up the face or head). Configural information is the information that represents the spatial relationships between the parts of an image (e.g., distance between the smaller parts) in a way that adds to the meaning of the image. The OV task recorded eye-movements with an eye-tracker during a face-matching task, and the RV task recorded the movement of a small manually controlled focus-window on a touch

sensitive screen. Both experiments revealed differences in scanning patterns between the individuals of the PCA group compared to their controls. An important question in the present study centered on the replicating previous findings healthy individuals in eye tracking tasks, and to determine whether or not the individuals with PCA who experience problems in face perception show similar scanning patterns to descriptions of prosopagnosia, simultanagnosia, and any other perceptual problems the individuals in the PCA group experience.

Aged Controls. Across tasks, the aged controls showed scanning patterns that fit with previous research in healthy individuals, as well as simultanagnosia (e.g., RV task). The results of each task are taken to reflect normal face processing, and an absence of significant cognitive or perceptual deficits (see Table 2 and Table 3). In the OV task, the aged control group produced results that fit well with previous research on face processing in this population. That is, the aged controls' responses were often correct, and VT were not more than a few seconds for each face.

Open-view task. Perhaps most importantly, the aged controls produced scanning patterns where fixations were concentrated in ROIs such as the eyes, nose, and mouth (see Figure 3) with the least fixations occurring in peripheral features (e.g., cheeks, chin, forehead). More specifically, the healthy individuals' fixations appeared occurred within areas that join these central features, such as the area between the nose and the mouth, as well as the areas between each eye and the nose. Initial fixations were often found in one of the Eye ROIs, or the Nose ROI, similar to previous research (e.g., Barton, 2008; Caine, 2004; Le, Raufaste, & Demonet, 2003; Stephan & Caine, 2009).

The scanning patterns that were observed in the aged control group are associated with second-order configural information, which is thought to be necessary, in addition to featural information, for normal face processing (e.g., Roisson et al.). In combination with performance

at normal levels on cognitive and perceptual tests that were completed in our lab (see Table 2), the performance of the aged controls are taken to be that of healthy individuals whose face processing is normal. Thus, the aged controls were assumed to be a suitable comparison group. It is important to note that one participant performed below the norm for their age for the phonemic fluency task, although this performance level has been interpreted as the result of participant anxiety. The individual reported feeling anxious about performing the task in the presence of the experimenter. Other tests of cognitive function for this individual were within the normal range.

When the scanning patterns of the aged controls were compared between the OV and RV tasks, the aged controls were found to spend significantly less time viewing the Right Eye, and significantly more time viewing the Nose and Left Cheek. These results indicate a change in processing strategy toward a more parts-based approach because of the increased viewing in the RVF, and decreases in areas that hold configural information. That is, the aged controls drastically reduced viewing of the Right Eye, and Nose.

In terms of the parsed eye ROIs, the aged controls' descriptive statistics indicated configural processing in the OV task, with less emphasis on configural processing in the RV task. That is, in Experiment 1, the aged controls viewed the area at the top of the nose, as well as the ROIs that were closest to eye level. In the RV task, the aged participants emphasized on the RVF, and the ROI closest to eye level. These results are interpreted as a switch to local, left-hemisphere-associated, processing; decrease in the area at the top of the nose, and an increase in RVF viewing.

The performance of the aged controls in Experiment 1 provide an adequate comparison for each individual in the PCA group. The results of the OV task in Experiment 1 show clear differences between the aged control group, and each individual in the PCA group. However, the

differences appear to vary between individuals in the PCA group, and can be interpreted based on the initial neuropsychological tests that were performed in our lab, and by a neurologist. The following section examines the results of the OV and RV tasks, as they apply to each individual in the PCA group.

Restricted-view task. In the RV task, the aged controls showed signs of a switch to a more parts-based approach, while still showing some signs of configural processing. For example, the aged controls spent the greatest amount of time looking at the Nose, followed by the Left Cheek, and Left Eye. The scanning patterns were taken to indicate more parts-based processing because the aged controls showed increased viewing of the Mouth, which is associated with prosopagnosia and a feature-matching strategy. Importantly, the aged controls' viewing in the left portion of the faces fall within the RVF, and increased viewing within the RVF could be due to a switch to local processing. That is, the left hemisphere is thought to be more involved in processing visual information that is located within the RVF, as well as local processing.

However, configural processing appears to occur in the aged controls' emphasis on the Nose, Left Eye, and Left Cheek. The emphasis on these ROIs suggest that the aged controls were moving the stylus between the Nose and Left Eye, which is an area of second-order configural information. However, part of the increased viewing of the Left Cheek and Left Eye could be due to the blurred nature of the faces; the participants may have used these border ROIs to localize the face more easily. Also, the majority of the aged controls were right-handed, which would place the left ROIs closer to the hand-held stylus.

Open-view vs. Restricted-view task. The comparison of the OV and RV tasks is thought to suggest a switch from configural-featural processing, which is associated with normal face processing, to a more local, or featural processing strategy. In the RV task, the aged controls

viewed the Mouth more, and the Right Eye less, which fits with previous research on a feature-based approach in prosopagnosia. However, participants appeared to still use configural processing strategies when scanning of the Nose, Left Eye, and Left Cheek are considered (i.e., areas that hold second-order configural information).

The results of the aged controls scanning patterns fits with accounts scanning patterns in studies that use gaze-contingent and mouse-contingent viewing window paradigms. That is, James et al. (2010) found evidence of configural processing in healthy individuals who viewed faces using a restricted viewing aperture; individuals scanned similar areas in open- and restricted-view conditions. However, Dalrymple et al. (2011) found that healthy individuals produced scanning patterns that more closely resembled those of individuals with simultanagnosia when a gaze- or mouse-contingent viewing window was used in viewing social scenes. That is, Dalrymple found that healthy individuals showed reduced viewing of the eye region, and increased viewing of high-contrast areas. These results were taken to indicate that simultanagnosia is the result of a restricted-window of visual attention.

RB. Despite RB's significant problems in face perception (e.g., BTFR-SF, famous faces tasks; see Case Descriptions), RB's scanning patterns across Experiment 1 and 2 did not show an obvious resemblance to research on prosopagnosia. Instead, RB's scanning patterns indicate the effects of both ventral and dorsal simultanagnosia. RB's scanning patterns indicated that she was not using configural or featural information. Instead, compared to aged controls, RB's scanning was significantly increased in areas of the stimuli that produce the highest contrast (e.g., the border of the face against a black background), where RB sometimes shows an inability to disengage (e.g., Forehead). Although RB showed differences in scanning patterns between Experiment 1 and Experiment 2, she shows common focus on high contrast, with little configural

or featural processing.

Open-view task. In the OV task of Experiment 1, RB's scanning patterns were expected to show a resemblance to prosopagnosia, however, RB's scanning patterns appeared to fit more easily with simultanagnosia. That is, RB showed a decreased viewing in the Mouth, and increased viewing in the Forehead, which was focused along the border of the background at the top of the Forehead. These patterns were unlike prosopagnosia because of RB's reduced fixations to the Mouth, which is often associated with prosopagnosia, and a featural processing strategy (e.g., individuals with prosopagnosia focus on the Mouth). Further, individuals with simultanagnosia show scanning patterns that emphasize areas of high-contrast, and an inability to disengage from these areas once fixations are made. RB showed significantly higher fixations along the high contrast border, and a high focus in this single area (e.g., RB spent more time in the Forehead compared to other ROIs, and often made initial fixations in this area).

Further evidence for the effects of simultanagnosia, as opposed to the featural approach seen in prosopagnosia comes from the fact that RB's scanning patterns did not represent either configural or featural processing. That is, RB's highest viewed ROI, the Forehead could be considered a non-featural ROI; the Forehead does not contain a particular feature. However, RB did show evidence of configural processing, with greater fixations at the top of the Nose, when the eye region was parsed across the x- and y- axes, which is difficult to interpret. Fixations to the eye region, made by healthy individuals, tend to fall in the top of the Nose, or the area between the eyes and the nose. This pattern is associated with configural processing. In contrast, individuals with prosopagnosia who make fixations to the eye region, have been found to make very few fixations at the top of the nose, and are thought to engage in a feature-by-feature approach; individuals make many fixations to each eye, with and very few fixations to the top of

the nose. RB's puzzling fixation pattern within the parsed eye ROIs may be due to ventral simultanagnosia, which is thought to be characterized by normal fixation patterns (e.g., Navon task) without global awareness (Farah, 2004). In all, dorsal simultanagnosia likely contributes to RB's focus in a single area, and attraction to high contrast, while ventral simultanagnosia may prevent RB's awareness of global forms when fixation patterns are not disturbed,

Restricted-view task. In the RV task, despite an overall different scanning pattern, RB's scanning patterns again suggest the effects of both dorsal and ventral simultanagnosia; RB spent significantly more time viewing non-featural ROIs that contain high-contrast borders (e.g., Chin, Right Cheek, and Forehead), and lack configural information. Further, RB spent less time viewing areas that healthy individuals tend to view most (e.g., Left Eye). However, healthy individuals also viewed the Mouth more often in the RV task, which is thought to reflect a featural processing strategy. Previous research suggests that healthy individuals can accurately match novel faces using just featural information. RB may have also used more of a feature-based strategy in the RV task, however, ventral simultanagnosia and RB's extremely restricted focus (e.g., RB often attempted to identify objects based on a tiny detail) may prevent visual recognition or awareness of a single feature.

When the eye ROIs were parsed in the RV task, RB's scanning was nearly opposite to that of the OV task; significantly more viewing of the eyebrows and border of the leftmost eye ROI and reduced viewing at eye level and the area above the nose. However, these results follow a common theme of increased viewing in high-contrast areas, and decreased viewing of configural information. Also, the parsed eye ROIs in the RV task show a lack of feature-by-feature processing associated with prosopagnosia, in that, RB spends significantly less time viewing each eye. Instead, RB spent more time viewing the borders near each eye, as well as the

eyebrows.

A lack of configural processing, or awareness of global forms, is thought to account for RB's slowed VT in the OV task, and normal VT in the RV task. RB shows motor slowing, and slowed processing speed (e.g., FTT, simple reaction time task; see Table 2 and Table 3), yet, there are no significant differences in VT between RB and aged controls in the RV task. The lack of differences are likely due to the fact that RB's VT did not increase substantially (e.g., $Mdn=12,490$ and $Mdn=13,527$), as did the aged controls (e.g., $Mdn=5,603$ and $Mdn=21,630$). If RB already experiences a restricted window of visual attention due to simultanagnosia, while healthy individuals' strategies might be reduced to featural or parts-based processing, RB's strategy is likely less affected.

Open-view vs. Restricted-view task. When RB's performance was compared between the OV and RV tasks, RB's overall pattern of differences between the OV and RV tasks suggest increased viewing of the areas of the face that fall below the eye region, particularly among ROIs that border on the background (e.g., Left Cheek, Chin, Mouth, Nose, and Right Cheek). One reason for the change in scanning patterns could be that high-contrast areas are less visible in the RV task; these areas of high contrast may have been 'found' haphazardly using the viewing window in the RV task. The restriction of the viewing window may also increase feature-by-feature processing once distracting border areas are less visible. RB may not experience a much difficulty with disengaging from highly salient areas.

The results of Experiment 1 and 2 are thought to be affected by dorsal and ventral simultanagnosia; RB's perceptual impairments appear to surround a focus on single details. Across experiments, RB showed a focus on areas of high contrast, was unable to disengage from salient areas (e.g., Forehead), yet these areas often did not contain configural or featural

information. That is, during perceptual testing, RB would often focus on a single detail within an object or face, and seemed to ignore the overall image (e.g., object identification, and figure copying; see Table 2 and Table 3, see Case Descriptions), which lead to erroneous responses. This restricted focus seemed to hinder RB's ability to make sense of the images she was viewing, which fits well with both dorsal and ventral simultanagnosia.

RB's performance in Experiment 1 and Experiment 2 are not likely to be due to problems such as language and executive function; RB's performance on phonemic and verbal fluency were within a normal range. Also, RB's neurologist indicated that RB's difficulty with baseline cognitive tests occurred only when using tests that relied on vision for accurate responses. RB's lack of significant problems in language, and executive function (see Table 2 and Table 3), indicate that RB's errors were more likely due to problems in visual perception, than problems understanding the task.

Importantly, mild memory problems may have affected RB's performance across experiments, although it is thought these effects were limited to error rates. Cognitive tests such as a forward-reverse digit task, showed that RB performed below controls. Also, RB showed fewer errors on shape-matching task where items were repeated across experiments, compared to the OV and RV tasks where each trial included a new cue-target pair. RB's differences in errors could be partly due to the demands placed on memory for the face-matching task.

SS. SS, who is thought to experience symptoms of both a dorsal and ventral form of PCA, produced scanning patterns that are interpreted as the result of simultanagnosia, neglect, and optic ataxia. Despite high error rates in the OV and RV tasks, SS did not show an obvious resemblance to research on prosopagnosia. SS's scanning patterns indicated that SS was not using configural or featural information.

Open-view task. In the OV task, SS performance suggests the effects of dorsal and ventral simultanagnosia because, while initial testing suggested that SS was less impaired in face processing (see Table 2 and Table 3), SS showed difficulty disengaging from the Nose ROI across experiments. Further, SS spent more time viewing the Right Cheek, a non-featural ROI that contains a high contrast border. These scanning patterns fit with research on simultanagnosia that suggest that simultanagnosia is a restricted window of visual attention (e.g., Dalrymple et al. 2011), and leads to problems disengaging from areas of fixation, and leads to a focus on areas of high contrast.

Although healthy individuals tend to make many fixations to the Nose, SS's emphasis on this region is not taken as a sign of configural processing because SS's fixations were limited to the Nose ROI. That is, unlike previous research that suggests that healthy individuals make fixations at the top of the Nose, between the Nose and Eyes, SS's fixations were often contained within the Nose ROI. The fact that SS was lower on the Right Eye, and Mouth is taken as more evidence for simultanagnosia, as well as neglect.

It is difficult to determine whether or not SS was engaged in featural processing, while SS showed reduced fixations to the Mouth, SS also suffers from visual neglect, which affects the both lower visual quadrants (e.g., Bells Cancellation Task; see Table 3). Neglect may drive SS's pattern of emphasis at the centre of the face, due to a lack of attention to areas below the centre. The Nose and Right Cheek might be the lower limit of SS's visual attention.

SS showed a scanning pattern that is typically assumed to represent configural processing; higher viewing at the top of the nose, and significantly less viewing to the far right. However, SS's scanning patterns within the eye region are thought to be due to ventral simultanagnosia. Previous research suggests that individuals with ventral simultanagnosia are

thought to perceive global forms, but are unaware of these forms. Further, these individuals will often incorrectly identify images where fixation patterns indicate that configural information was obtained (e.g., Dalrymple et al. 2007). SS's high level of central ROI viewing could also be an artifact of SS's overall central focus, and interpreted, again, as a reflection of simultanagnosia.

Restricted-view task. In the RV task, SS's performance indicates the affects of a combination of components of both dorsal and ventral simultanagnosia, and visual neglect. SS showed greater emphasis on the high contrast border of the Right Cheek ROI, and showed a decrease in fixations to the eyes, which is also associated with simultanagnosia (i.e., Dalrymple et al. 2011). Dorsal and ventral simultanagnosia may have interfered with SS's because he might be prevented from perceiving more than a single feature, in addition to difficulty perceiving multiple details within a single feature.

Similar to the OV task, evidence of visual neglect is seen in SS's reduced scanning below the Nose. SS scanning pattern is somewhat centre-focused, and suggests a further restriction of scanning in the RV task because of the overall reduced scanning in the eye region. Simultanagnosia and Neglect in the lower visual quadrants, which may remove attention to areas such as the Mouth, may interfere with SS's ability to engage in a feature-by-feature processing strategy in that less information is available to SS.

Again, SS's performance does not reflect configural or featural processing; SS's emphasis on the Nose ROI is not interpreted as configural processing in Experiment 2 because SS spent significantly less time viewing nearby features. For example, SS spent less time viewing both eyes. Healthy individuals who make initial fixations to the Nose tended to make fixations back and forth, between the internal features. However, SS spent significantly less time looking at both eye ROIs. SS's emphasis in the single Nose ROI shows that SS is neither

viewing areas that display spatial relationships between features (i.e., configural information), nor viewing each feature in a serial fashion (i.e., feature-by-feature).

When the eye ROIs were parsed, SS showed a pattern that does not easily fit accounts of parts-based processing, but is still in line with simultanagnosia. SS spent more time viewing the ROI that was furthest to the left, and indicates greater viewing in the LVF. SS's scanning pattern does not show an emphasis on areas associated with configural processing (e.g., top of the Nose), however, SS's pattern does suggest a greater role of the right hemisphere, which is associated with configural processing. Importantly, SS's viewing of the far left x ROI also indicates a scanning pattern that is focused on high contrast, due to the border that exists here.

Open-view vs. Restricted-view task. When the OV and RV tasks were compared, SS's scanning patterns indicate a restriction in SS's scanning pattern, with even greater focus on the central areas of the face in the RV task (e.g., increases in the Nose and Left Cheek, and decreases in the Left Eye, Mouth, Forehead, and Right Cheek), and more typical form of local processing. SS's pattern of differences between tasks, similar to aged controls, may be the result of an increased reliance on local processing. For example, SS showed increases within the Left Cheek because this area falls within the RVF (e.g., vision is associated with the left hemisphere and local processing). However, SS may have been affected by the restriction of the RV task less, as seen in a less pronounced within the Nose ROI, compared to the aged controls.

These results would account of simultanagnosia as a restricted window of visual attention, where previous research has found that healthy individuals produce scanning patterns that are similar to individuals with simultanagnosia when they view images with a restricted viewing window (e.g., Dalrymple et al. 2011). If SS is already viewing the world in a way that is similar to the RV task, it is not surprising that SS would experience lowered effects of such a

task. SS's scanning pattern may have also become more focused on the Nose in the RV task, due to the lessened visibility of the high-contrast border areas. Thus, without immediate visibility of these areas, SS may have been able to use a more 'typical' form of local processing, more similar to that of prosopagnosia.

The absence of significant differences in VT for the RV task are, similar to SS's scanning patterns, assumed to be the result of a lessened effect of the viewing window on processing strategy. The lack of differences was observed despite the fact that SS was slower on tasks such as the FTT, simple reaction time task, number cancellation (see Table 2 and Table 3). The RV task is meant to restrict participants to a parts-based strategy, which appears to slow VT in healthy controls. However, if SS is already using a strategy that results from restricted visual attention, VT may not be as slowed considering that SS is not losing access to configural information to the same degree as controls.

It is important to mention that SS's performance in both Experiment 1 and Experiment 2 might be affected by SS's mild memory problems (see Table 3, Case Descriptions). For example, SS performed below the level of aged controls on a forward-reverse digit task (see Table 3). Also, despite less problems in face processing (see Table 2 and Table 3), SS made more errors than aged controls in Experiment 1 and 2. Importantly, SS made comparatively fewer errors in a shape-matching task that repeated items 12 times across trials. In contrast, the OV and RV tasks included only new cue-target pairs for each trial. SS's errors may have been affected by what SS's neurologist suggested was a retrieval deficit. That is, SS may not have been able to adequately compare the Cue and Target faces. The differences in presentation between the Cue (blurred) and Target (clear) faces in the RV may have made comparison even more difficult for SS. Additionally, during testing with the RV task, SS reported that he was trying to pay attention

to the Left Eye to make matching easier. However, SS's scanning results indicate that SS spent significantly less time viewing both eyes, which suggests that SS may not have been aware of the feature that he was viewing.

AP. AP, who was least impaired on tests of face processing, and object identification (see Table 2 and Table 3, Case Descriptions), shows evidence in the OV and RV tasks of dorsal simultanagnosia (e.g. normal performance for object naming, while errors are higher than controls for object counting), however AP's performance was also affected by motor slowing, optic ataxia, and visual neglect. AP is thought to experience more symptoms of a dorsal form of PCA, and shows more problems associated with vision for action, compared to perception. AP's scanning patterns are believed to be most affected by dorsal simultanagnosia, visual neglect, and, particularly in the RV task, optic ataxia.

Open-view task. In the OV task, the differences in scanning patterns between AP and aged controls appears to result from dorsal simultanagnosia, ventral simultanagnosia, and visual neglect. In terms of dorsal simultanagnosia, AP showed a significant increase in viewing of the high-contrast border of the Right Cheek, which does not hold configural information. AP is believed to experience dorsal simultanagnosia based on improvements in reading with use of a viewing window, problems counting objects, and difficulty navigating her environment (e.g., AP often bumps into furniture).

However, despite relatively normal face processing (see Table 2 and Table 3), and a scanning pattern that showed little difference from that of the aged controls, AP made significantly higher errors. These results are interpreted as the effects of ventral simultanagnosia. Previous research suggests that individuals with simultanagnosia often make fixations that are similar to controls (e.g., Navon task), yet produce incorrect responses in tasks where

identification is based on global forms. Ventral simultanagnosia is characterized by an unawareness of global forms. Therefore, AP may have been unaware of the global image of the faces, and able only to engage in matching of restricted areas of attention (e.g., the Right Cheek).

For the parsed ROIs, AP's scanning also fits with an account of dorsal and ventral simultanagnosia. Dorsal simultanagnosia is seen in AP's reduced viewing in the LVF (leftmost x ROI) may indicate less configural processing, because the LVF and right hemisphere is associated with configural, and global, processing. Importantly, AP is left-handed, and her neurologist suggested that, considering atrophy in the left hemisphere, AP's language may be partly located in the right hemisphere. Thus, it is unclear how to interpret AP's reduced viewing of the RVF. Ventral simultanagnosia may play a role in AP's greater fixations to the top of the Nose. This pattern, while associated with normal face processing, AP might not be aware of the configural information that she is accessing. Visual neglect likely prevented AP from attending to areas of faces that would be helpful for accurate matching.

Restricted-view task. In terms of the RV task, AP's performance is thought to reflect dorsal simultanagnosia, as well as visual neglect and optic ataxia. In line with dorsal simultanagnosia, AP's scanning pattern was higher along the border of the Forehead and in the RVF, and lower at internal areas (e.g., Nose). Increased viewing in the RVF may indicate greater emphasis on a parts-based processing strategy; however, the fact that AP's is left-handed makes these results more difficult to interpret. Similar to the OV task, ventral simultanagnosia might be the reason that AP shows high errors (20% higher than controls, although not significant), despite normal face processing.

In the OV task, AP showed a significant difference in a single ROI, but in the RV task, AP showed multiple differences. The higher number of significant differences in the RV task

could be due to problems in manual movement of the stylus. During assessment by a neurologist, AP showed evidence optic ataxia (e.g., AP often misreached), which would interfere with AP's placement of the stylus on the computer screen. AP also chose to use her non-dominant right hand in the RV task because of an injury to her left arm. These difficulties were likely compounded by visual neglect (see Case Descriptions), which may have prevented AP's attention from falling on important areas of a face.

AP's scanning pattern when the eye ROIs were parsed are more similar to patterns associated with dorsal and ventral simultanagnosia. AP showed significantly increased viewing nearest the high-contrast eyebrows, and significantly decreased viewing at eye-level. Decreased viewing of the eye region has been reported in recent research on simultanagnosia. The fact that AP showed high errors indicates that AP was not able to use a feature-matching strategy.

Open-view vs. Restricted-view task. Changes in AP's scanning pattern between the open- and restricted-view tasks indicate an increased focus on peripheral areas (e.g., decrease on Nose, increase on Forehead), and is likely the result of problems such as optic ataxia, in addition to both dorsal and ventral simultanagnosia. AP's neurologist reported that AP showed misreaching, which would have interfered with scanning in a task such as the RV, where visually guided action is required to navigate the stylus. Also, AP's injury and subsequent choice to use her non-dominant hand would have affected scanning. The aforementioned impairments are taken to be the main reasons for the appearance that AP was affected to a larger degree by the RV task, compared to aged controls.

AP's increased VT in Experiment 1 interpreted as the result of slowed motor processing and general processing speed (e.g., AP was significantly slower on FTT and simple reaction time task). AP did not show face-processing impairments, and was not significantly different in terms

of errors, suggesting that AP's VT may not have been the result of difficulty making decisions. In contrast, in the RV task of Experiment 2, despite AP's slower speed at baseline testing, and known problems such as an injured left arm, and optic ataxia (see Case Descriptions), are AP's lack of difference in VT may be due to a lack of switch in processing strategy due to both dorsal and ventral simultanagnosia. That is, AP may already engage in a largely restricted parts-based strategy, and she may not have an awareness of global forms in non-restricted viewing.

AP's errors, although only significantly higher in the RV task, are thought to result from dorsal and ventral simultanagnosia because of AP's apparent normal face processing (see Table 2 and Table 3). However, individuals with dorsal simultanagnosia are thought to show increased rates of accuracy in global perception when stimuli are smaller. AP may have showed preserved face perception at initial testing because stimuli used in baseline tests were generally smaller than those used in the OV and RV tasks. AP's increased in errors in the RV task, are taken to be the results of inadequate scanning due to optic ataxia and injury. That is, AP may have experienced difficulty viewing the areas necessary for accurate face-matching, if she had difficulty maneuvering the viewing window to areas that AP wanted to look at.

MTB. MTB, who is thought to experience the effects of both dorsal and ventral subtypes of PCA, was unable to participate in the OV task of Experiment 1 due to an inability to maintain fixation during the calibration phase. MTB's neurologist reported that MTB experienced problems in scanning. In the RV task, MTB's pattern of results is thought to be most affected by problems in motor coordination, neglect, as well as dorsal simultanagnosia (e.g., MTB often reported that single objects move out of vision, or 'jump around').

Restricted-view task. MTB's scanning patterns could be interpreted as the result of dorsal simultanagnosia in that MTB spent more time viewing high-contrast border areas, and less time

viewing areas that hold configural information. Further, MTB is known to show neglect, and a lack of awareness of global forms (see Table 2 and Table 3, Case Descriptions). Although MTB showed signs of ventral simultanagnosia, the potential effects were not seen in the RV task; MTB's scanning largely bypassed areas of configural or global information (e.g., Left Eye and Nose). MTB's performance does not show an obvious resemblance to scanning patterns associated with prosopagnosia, and appears to reflect a lack of both configural and featural processing (e.g., Chin, Right Cheek). MTB spent less time viewing areas that are associated with configural processing, and often viewed most by healthy individuals (e.g., Left Eye and Nose).

In addition to the effects of simultanagnosia, MTB's scanning patterns are most likely limited by visual neglect (see Case Descriptions), and problems such as optic ataxia. Thus, even in areas where MTB's visual neglect did not prevent attention, MTB's scanning pattern was probably affected by difficulty manipulating the stylus, and inaccurate reaching.

The parsed ROIs showed that MTB spent more time in areas that are associated with simultanagnosia and parts-based processing. That is, MTB spent significantly more time viewing the area along the eyebrows. This pattern is similar to that of simultanagnosia, considering the emphasis on the high contrast of the eyebrows, and lack of scanning on the eyes. MTB's increased scanning in the leftmost x ROI could also be interpreted as the result of simultanagnosia in that there is a high-contrast border in ROI. However, the leftmost, LVF, x ROI is also associated with right-hemisphere configural and/or global processing. The fact that MTB spent more time viewing an area in the LVF could also indicate that she is not using a parts-based strategy, viewing this area due to the high level of contrast.

Compared to aged controls, MTB produced more errors, longer VT to Cue faces. Previous research suggests that novel faces are matched successfully when either featural or

configural information is present. If MTB was engaged in featural processing, MTB's error rates would be expected to be much lower than the results of Experiment 2 indicate. Importantly, if MTB is able to engage in featural processing, other problems such as neglect, motor coordination, and optic ataxia likely interfere with the scanning that is useful in a feature-matching strategy. The different viewing conditions that exist between the Cue (restricted) and Target (open) faces may interfere with any feature- or detail-matching strategies that MTB may use. Based on a neurologist's assessment, MTB was determined to experience mild memory problems (see Case Descriptions), which may have increased errors, alongside MTB's obvious difficulty perceiving faces.

PLH. PLH's scanning pattern is interpreted as the effects of problems such as dorsal simultanagnosia, mild apraxia, optic ataxia, and visual neglect. Overall, PLH's pattern appeared to be focused on peripheral areas (i.e., border areas), with significantly less scanning in internal areas (i.e., Nose). These results are thought to be due to a focus on the contrast of the border areas, which may have reduced PLH's ability to attend to internal features, in addition to the added difficulty of using the viewing window to look at stimuli. PLH who was initially thought to experience a mixed form of dorsal and ventral subtypes of PCA, was recently diagnosed with AD, and has showed signs of diffuse atrophy. PLH was unable to participate in the OV task of Experiment 1 due to an inability to maintain fixation during the calibration phase. Thus, conclusions drawn about PLH's scanning patterns in the RV task are limited due to the manual nature of the task and PLH's problems such as mild apraxia, and optic ataxia.

Restricted-view task. PLH's performance in the RV task shows evidence for the effects of dorsal simultanagnosia (e.g., PLH is known to bump into furniture, and has difficulty finding objects on her desk), however, these results are likely also affected by significant problems such

as visual neglect, visual extinction (LVF) apraxia, optic ataxia, and memory. PLH's perceptual problems, including severely impaired face perception, were interpreted by her neurologist as being largely the result of simultanagnosia; PLH often described only a single component of an image, instead of describing an object as a whole (e.g., PLH reported that individuals presented in a picnic scene were 'flying' because she did not appear to notice the blanket on which that they were seated).

PLH's Neglect, which affects all four quadrants, most likely interfered with normal scanning because of limitations on areas of visual attention. PLH also experiences extinction in the left visual field, which may contribute to PLH's increased viewing of the Left Eye (RVF). PLH might be better able to perceive details or features on the Cue face, due to the restriction of the Viewing window, however, extinction within the left visual field may hinder matching when the Target face, which is presented with both sides of the image visible. The viewing window may reduce the effects of extinction while viewing the Cue face, similar to the use of a viewing window for reading, however, this improvement may not help in face-matching when the Target face is presented without the viewing window.

PLH's manual scanning pattern would have been affected by mild apraxia, and optic ataxia. PLH's manipulation of the stylus was affected, and her ability to move the stylus to areas she chose to view would have been impaired, in addition to problems attending to visual stimuli. PLH's neurologist reported that PLH often misreached (e.g., PLH often over-shot her reach), and showed mild apraxia (see Case Descriptions); PLH may not have been able to properly reach out and accurately place the stylus on each face.

PLH showed scanning patterns when the eye ROIs were parsed along the x- and y-axes fit with accounts of abnormal face processing, and, particularly, simultanagnosia. PLH showed a

pattern that seems similar to that of prosopagnosia, where PLH spent more time looking at the peripheral areas of the eye region (e.g., leftmost and rightmost x ROIs that contain each eye), and less time looking at the top of the nose, and eye level y ROI. However, due to PLH's significant problems with simultanagnosia, it is assumed that PLH's patterns are more likely the result of PLH being drawn to the high-contrast border area at the outermost x ROIs. Further, PLH is severely impaired at face viewing tasks (see Table 2 and Table 3), and, possibly related to problems associated with AD, PLH sometimes appeared unsure of whether or not she was looking at a face. Thus, PLH's impairments decrease the likelihood that PLH was able to determine whether or not she was looking at eyes.

PLH's errors and increased VT fit well with the results of baseline perceptual tasks (e.g., famous faces test, BTFR-SF; see Table 2 and Table 3), and tasks such as the FTT. PLH showed marked perceptual impairments, including difficulty with face perception, object perception, and simultanagnosia. Similar to most of the other individuals in the PCA group, PLH's lack of difference in VT compared to aged controls, despite signs of slowed processing (e.g., slower than aged controls on both a FTT and a simple reaction time task, which suggest slowed motor responses and processing speed), is interpreted as a reduced effect of the RV task due to PLH's already significant perceptual and motor problems.

Importantly, PLH was recently diagnosed with AD, and scored far below healthy individuals on the DRS-II. Further, when completing tasks such as the Famous Faces test (see Table 2), PLH often appeared confused about the procedure of the task at hand. The results of the RV task are also very likely to have been affected by memory problems and difficulty understanding the procedure of the RV task. That is, in addition to PLH's marked perceptual problems, and difficulty with use of the stylus, PLH may not have been able to adequately

encode or retrieve the face images she viewed.

Overall, the results of the OV task of Experiment 1 provided information about the aged controls scanning patterns, which fit well with previous research on scanning patterns in healthy samples (e.g., highest viewing in eyes, nose, mouth). In contrast, the OV task suggests that the individuals with PCA process faces differently than aged controls, and do not show an obvious similarity to scanning patterns known to occur in typical prosopagnosia. Despite differences between the individuals from the PCA group that participated in Experiment 1, all of the individuals in the PCA group produced scanning patterns that placed less emphasis on areas of configural information, often including increased viewing of high contrast areas, which is often associated with simultanagnosia.

The results of Experiment 1 appear to be related to simultanagnosia, ventral and/or dorsal, other perceptual problems (e.g., neglect), problems such as optic ataxia, and, sometimes, gaze apraxia. The differences in scanning pattern included decreases in the eye ROIs, and increased in non-featural border ROIs (e.g., Right Cheek), and SS and RB showed signs of an inability to disengage from single features (e.g., Nose and Forehead, respectively). In all, the OV task of Experiment 1 indicates that the individuals with PCA did not show evidence of typical prosopagnosia in scanning patterns. Instead, the individuals in the PCA group showed signs of simultanagnosia, in addition to other significant deficits in vision for perception.

The results of the RV task of Experiment 2 showed scanning patterns among aged controls that appeared to focus more on a parts-based processing strategy; increased viewing of RVF structures, and features such as the Mouth. Further, the aged controls appeared to use a more parts-based approach (e.g., increased viewing in the lower regions of faces, and in the RVF), still, with evidence for the inclusion of some configural processing.

The individuals in the PCA group each showed significant differences compared to the aged controls, and each displayed a unique pattern of differences. The results of Experiment 2 also appear to be related to simultanagnosia, and other perceptual problems, such as neglect. However, problems such as optic ataxia and gaze apraxia may be a larger problems for individuals with PCA in the RV task because scanning is completed with the use of a stylus. Importantly, the results from individuals such as MTB and PLH, who could not participate in the OV task, must be interpreted with caution because interference in scanning patterns from known impairments such as optic ataxia cannot be understood without a comparison to performance in a task where visual guided manual action (e.g., movement of the stylus) is needed.

For the individuals in the PCA group, the differences in scanning pattern between the OV and RV tasks did not appear similar. However, aside from MTB, each individual in the PCA group did not show significantly increased VT in the RV, despite evidence of slowed motor speed, and processing speed. These results fit with previous research that suggests that simultanagnosia is the result of a restricted window of visual attention, and a problem in global processing; the individuals in the PCA group are thought to already rely on a more parts-based approach, which could reduce the effects of the RV task. In contrast, the aged controls likely have to adjust to the reduction in the availability of configural information.

Although none of the individuals in the PCA group were found to produce scanning patterns that resemble those of prosopagnosia, an interesting similarity may exist; previous research on prosopagnosia suggests that individuals who already engage in local processing, with less or no configural processing, are not affected by a task such as the Viewing Window (e.g., Busigny, Thomas, & Roisson, 2010). For example Busigny et al. (2010) found that an individual with prosopagnosia was unaffected by face stimuli that required a switch to a parts-based

strategy, while healthy control participants showed lowered accuracy, and increased reaction times. The higher number of significant differences found in the RV task suggest that scan paths in all participants are affected when the task demands a parts-based approach. Similar to prosopagnosia, a consistent finding from all of the individuals with PCA in the present study is that they all appear to be less focused on face regions that hold configural information, and may be using a parts-based approach, which becomes even more extreme in the RV task.

The RV task is meant to limit individuals to a serial parts-based processing strategy, a strategy that is associated with deficits in recognition of familiar faces. Research suggests that healthy individuals are highly accurate at matching novel faces, even when they are limited to either configural (e.g., blurred faces; Lobmaier et al. 2008) or featural (e.g., scrambled faces) information. In the RV task, aged controls, again, produced few errors. However, the aged controls did spend more time viewing both Cue and Target faces, as well as a notable amount of time viewing the Left Eye, Left Cheek, and the Nose ROI. These increased VTs are thought to be the result of the loss of configural processing to a great extent.

All of the participants in the PCA group experience simultanagnosia (see Case Descriptions; see Table 2), despite other differences in their perceptual problems. Thus, a second important question for the present study centered on the contribution of simultanagnosia to face perception problems experienced by the individuals with PCA. A component of Bálint syndrome (Rizzo, 1993), simultanagnosia, is associated with bilateral damage to the parieto-occipital junction (Rizzo, 1993); all of the individuals in the PCA group, including RB, showed atrophy in posterior areas of the brain, including the occipito-parietal cortex. Simultanagnosia has been looked at as a problem in the perception of spatial location and spatial relationships of objects or their elements (Dalrymple et al. 2009; Dalrymple et al. 2010; Karnath et al. 2000; Huberle et al.

2010; Montoro et al. 2010), and more recently, as a restricted window of visual attention (e.g., Dalrymple et al. 2010; Dalrymple et al. 2011).

The results do not entirely fit with the idea from Dalrymple and colleagues (2011), that simultanagnosia is the result of a restricted window of visual attention. Dalrymple et al. found healthy controls' scanning patterns using a gaze-contingent window resembled scanning patterns of an individual with simultanagnosia in an OV eye-tracking task. These similarities led researchers to conclude that the effects of simultanagnosia are accounted for by restricting visual attention via the gaze-contingent window. If simultanagnosia is primarily the result of a restricted window of visual attention, we would expect to see a significantly different pattern of scanning changes for the OV vs. RV tasks, between the aged controls and each individual in the PCA group. The results of the OV vs. RV comparison does indicate that these tasks had different effects on scanning between the aged controls and the individuals with PCA. Further, the results suggest that further analysis of errors and viewing time should be included in the between task comparison, in that the individuals with PCA appear to show a smaller changes between tasks than controls in these aspects.

Facial features on real life-size faces are not close together, and faces are not generally small relative to some objects. Further, the face stimuli in the present study, while smaller relative to life-size faces, were sometimes larger than stimuli used in object identification tasks. Thus, another contribution to the PCA patients' difficulties in each task of the present study could be problems in the ability to process large stimuli, and stimuli whose elements are widely spaced. Importantly, the faces presented in the BTFR-SF and the famous faces task that was completed in our lab contained stimuli that were much smaller than life-size faces, and similar to the size of stimuli used in object identification tasks. Stimulus size may account for AP's high

scores on the BTFR-SF and famous faces tasks, yet lower accuracy in the OV and RV tasks. Researchers have found that inter-element distance, and stimulus size can affect global processing, and suggest that larger distance and size place a higher demand on visual working memory (e.g., Dalrymple et al. 2009; Dalrymple et al. 2010; Karnath et al. 2000; Huberle et al. 2010; Montoro et al. 2010)The problems experienced by RB, and the other individuals within the PCA group could be affected by the size of the stimuli, along with the spacing of facial features.

Aside from questions of similarity to prosopagnosia and simultanagnosia, all of the individuals with PCA in the present study experience multiple. The fact that only RB, PLH, and MTB experience significant face perception problems, as well as other deficits in visual perception (see Table 2 and Table 3), leaves open the possibility that their performance was affected by variables that extend beyond prosopagnosia and simultanagnosia, suggests that there may be behaviour differences between individuals in the PCA group based on differences in areas of atrophy. Also, SS and AP do not yet show significant large areas of atrophy in temporal areas, and may be experiencing a more pure form of simultanagnosia. In contrast, RB, MTB, and PLH may be experiencing simultanagnosia in combination with more perceptual deficits, or a more ventral form of simultanagnosia, which is more likely to affect perception of even the smallest details. Importantly, SS and AP showed much less impairment on tasks such as object perception, suggesting less visual impairment. Previous research on simultanagnosia may better explain the deficits of SS and AP, in that simultanagnosia is generally reported with preserved object perception. In contrast, RB was often only able to identify a single detail within an object's component part.

The fact that the PCA patients did occasionally make fixations to internal features that are associated with configural processing, yet produced poor accuracy, may suggest an overall lack

of visual awareness. Dalrymple et al. (2007) found that the fixations of an individual with simultanagnosia closely traced the shape of a global form (e.g., Navon letter), but response errors were very high. Some researchers suggest that simultanagnosia is a reduction in visual awareness, and that visual stimuli can influence eye-movements without conscious awareness of the viewer (Rizzo & Hurtig, 1987). In the present study, RB and PLH would often misidentify features during the RV task. The present study found that the PCA group produced a high number of fixations to the faces, often looking at all 8 ROIs. The results of the present study are more consistent with the idea that individual with simultanagnosia may lack awareness in global processing, compared to suggestions such as the restricted window of visual attention.

The Viewing Window Task

By using the Viewing Window paradigm, we were hoping to determine if limiting vision to a single object at a time would make a face-matching task easier for individuals with PCA. Perhaps knowing that they were only seeing a small part of the face at any one time would encourage the individuals with PCA to more fully explore parts of the face. The results did not show any improvement in accuracy for the PCA group in the RV condition, and response latencies were longer. This suggests that in at least a few cases (RB, PLH and MTB), the perceptual deficits present in these patients prevented them from even knowing what ‘part’ of the face they were looking at through the focus-window.

Two participants from the PCA group, PLH and MTB, only participated in the Viewing Window task because they were unable to maintain fixation long enough to complete calibration for the eye-tracker. In addition, the older adults in the present study were more difficult to calibrate due to prescription eyeglasses and, for some, rapid fatigue. In contrast, the Viewing

Window task does not require calibration, and completion of this task was much easier with a clinical population, compared to eye tracking.

The Viewing Window is a tool that can record scan paths during a face-matching task, without calibration, and has the added advantage of being highly portable. Previous research that used a manually operated focus-window have simultaneously recorded eye movement, and found that gaze patterns closely matched the paths of the focus-window (e.g., Baugh & Marotta, 2008; Dalrymple et al. 2007; James, Huh, & Kim, 2010). The sample size of the present study was relatively small, however, with further development (e.g., collection of normative data from the general population), the Viewing Window task could be used to provide a more detailed information regarding scan paths, which would allow for more helpful comparisons between healthy individuals and those with neurodegenerative disease.

Conclusion and Future Directions

The present study is the only known study of face perception in PCA to use an eye-tracker and restricted-window paradigm, and one of very few studies to examine face processing in this population, in general. PCA is a relatively rare neurodegenerative disorder that is likely greatly underreported due to most cases going undiagnosed, or misdiagnosed as AD. Part of the problem in detecting PCA is the fact that the earliest symptoms are similar to natural changes in vision that occur in natural aging (e.g., losing one's place while reading, blurred vision leading to a new eye glass prescription). Continued research on PCA is necessary to further delineate the early signs of this disorder, which will expose patients to health care professionals and early treatment. The more we understand how this disorder affects the elderly, the fewer barriers there will be to treatment.

It is unclear to what extent prosopagnosia or simultanagnosia contributes to impaired face perception in the PCA group, however, the present study brings up important questions that relate to theories of prosopagnosia and simultanagnosia. Our results suggested differences between the PCA group and controls, yet the scanning patterns seen in the PCA group do not clearly implicate either type of visual deficit. In fact, they suggest that it may be a case of multiple deficits influencing how faces are perceived. There are some similarities to patterns seen in prosopagnosia (e.g., more time viewing external features), and we know that the individuals with PCA experience simultanagnosia. However, no other studies have examined gaze patterns of simultanagnosia in a face-matching task, which leaves the current results without the comparisons that are available as with prosopagnosia. Also, all of the individuals in the PCA group experience multiple visual deficits, which make it difficult to isolate the effects of any specific deficit.

Both prosopagnosia and simultanagnosia involve a problem in configural, or global, processing (e.g., Farah, 1995; Le et al., 2008; Dalrymple et al. 2007), while featural or parts-based processing is more or less preserved. Simultanagnosia may be thought of, therefore, as an extreme version of parts based processing. The PCA group, particularly RB, may be using an extreme form of parts-based processing that is limited to single details. This may be made all the harder because of underlying perceptual deficits in those individuals in the PCA group with temporal lobe atrophy.

Some researchers have suggested that faces are special, and that associated brain regions are specific to face processing, while others have emphasized the role of human expertise in face processing. The results of the present study push for exploration of the idea that face processing is no more special, or expert, than the need for global processing of such stimuli.

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APPENDIX A

**INFORMED CONSENT FORM FOR BEHAVIORAL STUDY:
UNDERGRADUATE HEALTHY PARTICIPANTS**

PRINCIPLE INVESTIGATOR: Dr. Jonathan Marotta
University of Manitoba
(204) 480-7057

INVESTIGATORS: Keri Locheed,
Loni Desanghere,
Dr. Jane Lawrence,
Department of Psychology
University of Manitoba
(204) 480-1248

SOURCE OF SUPPORT: NSERC Discovery Grant

This consent form, a copy of which will be left with you for your records and reference, is only part of the process of informed consent. It should give you the basic idea of what the research is about and what your participation will involve. If you would like more detail about something mentioned here, or information not included here, you should feel free to ask. Please take the time to read this carefully and to understand any accompanying information.

PURPOSE: We are interested in how different visual representations we construct interact with one another and form our experience of vision.

DESCRIPTION: This study will take place in the Perception and Action Lab in the Duff Roblin Building on the Fort Garry Campus. During the study, you will be asked to make a series of eye- and arm-movements to target objects. For example, on some trials, you will be asked to reach out and pick up a target object. On others, you will be asked to look at target objects or visual images and identify them. An eye tracker may be used to record your eye movements when performing these tasks and an OPTOTRAK 3-D or ShapeClaw motion recording system will be used to record your finger and hand movements. Prior to this task, you will be asked to fill out a brief demographics questionnaire that inquires about your age, gender, handedness, whether you wear glasses, and your stereo acuity. The whole procedure will take less than an hour and a half to complete. You will earn 2 experimental credits for your participation in this study.

RISKS AND BENEFITS: There are no evident risks inherent in the tasks you will perform but some of the tests may be difficult. While this may be frustrating to you, there will always be an investigator with you to assist you and support you

COSTS AND PAYMENTS: There are no fees or charges to participate in this study. You will not receive payment.

APPENDIX B

**INFORMED CONSENT FORM FOR BEHAVIORAL STUDY:
STROKE PATIENTS AND AGE-MATCHED HEALTHY PARTICIPANTS**

PRINCIPLE INVESTIGATOR: Dr. Jonathan Marotta
University of Manitoba
(204) 480-7057

INVESTIGATORS: Keri Locheed,
Lee Baugh,
Loni Desanghere,
Dr. Jane Lawrence
Department of Psychology
University of Manitoba
(204) 480-1248

SOURCE OF SUPPORT: NSERC Discovery Grant

This consent form, a copy of which will be left with you for your records and reference, is only part of the process of informed consent. It should give you the basic idea of what the research is about and what your participation will involve. If you would like more detail about something mentioned here, or information not included here, you should feel free to ask. Please take the time to read this carefully and to understand any accompanying information.

PURPOSE: We are interested in how different visual representations we construct interact with one another and form our experience of vision.

DESCRIPTION: During the study, you will be asked to make a series of eye- and arm-movements to target objects. For example, on some trials, you will be asked to reach out and pick up a target object. On others, you will be asked to look at target objects or visual images and identify them. An eye tracker may be used to record your eye movements when performing these tasks and an OPTOTRAK 3-D or ShapeClaw motion recording system will be used to record your finger and hand movements. Prior to this task, you will be asked to fill out a brief demographics questionnaire that inquires about your age, gender, handedness, whether you wear glasses, and your stereo acuity. The whole procedure will take less than an hour and a half to complete.

RISKS AND BENEFITS: There are no evident risks inherent in the tasks you will perform but some of the tests may be difficult. While this may be frustrating to you, there will always be an investigator with you to assist you and support you

COSTS AND PAYMENTS: There are no fees or charges to participate in this study. You will not receive payment. If you require transportation to the Perception and Action Laboratory at the University of Manitoba we will provide a taxi to and from the study. Alternatively, we may be able to bring our equipment to your location to perform the study if mobility is difficult for you.

CONFIDENTIALITY: Your information will be kept confidential. You will be referred to by a code number. All files containing identifying information will be stored in a locked cabinet separate from data with your code number. Your files will only be accessible by the investigators and will be destroyed 5 years after the completion of the study (approximately December, 2013). All papers containing personal information will be shredded. All electronic files will be deleted. Any cds or dvds containing data will be physically destroyed.

VOLUNTARY CONSENT: If you do not wish to participate in the study, you are free to leave without consequence and we thank you for your consideration. Your signature on this form indicates that you have understood to your satisfaction the information regarding participation in the research project and agree to participate as a subject. In no way does this waive your legal rights nor release the researchers, sponsors, or involved institutions from their legal and professional responsibilities. You are free to withdraw from the study at any time, and/or refrain from answering any questions you prefer to omit, without prejudice or consequence. Your continued participation should be as informed as your initial consent, so you should feel free to ask for clarification or new information throughout your participation.

This research has been approved by the National Research Council of Canada Ethics Board and the Psychology/Sociology Research Ethics Board of the University of Manitoba. If you have any concerns or complaints about this project you may contact any of the above-named persons or the Human Ethics Secretariat at 474-7122, or e-mail Margaret_bowman@umanitoba.ca. A copy of this consent form has been given to you to keep for your records and reference.

Signature of the Participant	Date	Signature of Investigator	Date
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If you would like to receive general summary of the results from this study when it is completed, please complete your mailing address below:

Mailing Address: _____

