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Disconnection in prosopagnosia and face processing

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ABSTRACT

Face perception is a function with significant complexity, reflected in cognitive models that propose a hierarchy of parallel and serial processing stages. Current neuroimaging data also show that face perception involves a core processing network of cortical modules, which are likely specialized for different functions involved in face processing. The core face processing network is further linked to an extended face processing network which is not solely involved in the perception of faces, but rather contains modules mediating the processing of semantic, biographic and emotional information about people. The segregation of these processes within discrete anatomic regions creates the potential for disconnection between regions to generate neuropsychological deficits involving faces. In this review we consider the types of disconnection possible both within the core face processing system and between the core and extended systems, the pattern of deficits that would be considered as evidence of such disconnections, the potential anatomy of lesions that would create them, and whether any cases exist that meet these criteria.

Face recognition is an important perceptual skill: it enables the rapid recognition of hundreds, if not thousands, of unique faces, and enables appropriate social interactions. The loss of this skill is the key deficit in prosopagnosia, the inability to recognize familiar faces, which can be acquired in later life from a variety of cerebral lesions, or be present from birth as a congenital and sometimes inherited disorder. Although there were early proposals that this symptom might arise from the convergence of non-specific deficits of vision, memory and general mental processing (Bay, 1953; Cohn et al., 1977), it is apparent now that prosopagnosia is a selective visual agnosia resulting from damage to a cortical network devoted to the accurate perception of faces (Barton, 2003).

It is also apparent that prosopagnosia is not a single functional entity but a family of disorders. Face recognition is a complex process that involves both perceptual and memory operations: dysfunction at a variety of levels may lead to the same end result of failure to recognize a familiar face. As with other visual agnosias (Barton, 2003; Lissauer, 1890), prosopagnosia has traditionally been divided into two broad subtypes (de Renzi et al., 1991). In apperceptive prosopagnosia, the patient is unable to form an accurate perceptual representation of the structure of a viewed face, a skill which is usually assessed by matching of unfamiliar faces (Benton and Van Allen, 1968). In associative prosopagnosia, formation of the facial percept is intact, but this information cannot be matched to facial memories in order to recognize that this is a face the subject has encountered before.

Just as there is more than one functional cause of prosopagnosia, there are also a variety of anatomic lesions that have been described in prosopagnosia. The two seminal small series of cases several decades ago noted an association with

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bilateral lesions of the lingual and fusiform gyri (Damasio, 1985; Meadows, 1974). Since then, there have been reports of anterior temporal lesions (Evans et al., 1995; Barton et al., 2002), unilateral right occipitotemporal lesions (de Renzi, 1986; Landis et al., 1986), and even rare left occipitotemporal lesions (Tzavares et al., 1973; Mattson et al., 2000; Barton, 2008). Likewise, recent functional imaging studies in healthy subjects have delineated a network of face processing regions rather than a single cortical region (Haxby et al., 2000; Palermo and Rhodes, 2007; Rossion et al., 2003a; Gobbini and Haxby, 2007; Ishai et al., 2005).

The fact that lesions in widely disparate locations can lead to forms of prosopagnosia lends support to network models of face processing (Haxby et al., 2000). These models consist of a core system of anatomic regions that are primarily responsible for face perception, with different perceptual roles assigned to different core modules (Haxby et al., 2000). Beyond the core system of face processing is a large extended system responsible for other functions relevant to the perception of a face, such as accessing biographical, semantic or emotional information related to the face (Gobbini and Haxby, 2007; Haxby et al., 2000). An important distinction between the core and extended systems is that the former is specialized for the perception of faces, whereas the latter is multimodal, and can be accessed by a wide range of stimuli other than faces (Gobbini and Haxby, 2007; Haxby et al., 2000).

The involvement of multiple cognitive processes and multiple anatomic regions in face recognition raises the distinct possibility that some cases of prosopagnosia may be not due to damage to specific processes or regions, but due to disconnections between processes and regions (i.e., topological *vs* hodological damage; see Catani and Ffytche, 2005). In this review we first consider historical proposals of disconnection in prosopagnosia. We then summarize current cognitive and anatomic frameworks of face processing and discuss where disconnections could arise within these frameworks, what behavioral findings would suggest such disconnections, and whether there are existing cases in the literature that fulfill these predictions. We will focus on three potential disconnections. First, we will examine the core disconnection of associative prosopagnosia, and discuss whether associative prosopagnosia can be shown to be due to disconnection between perceptual mechanisms and facial memory stores, and not simply due to damage to the memory stores. Second, we will examine the potential for a disconnection between the core face processing system and other regions involved in processing stored semantic and biographic information related to people, namely the anterior temporal lobes and the precuneus. Finally, we will discuss affective 'visual-limbic' disconnections, as reported in a hypoemotionality syndrome associated with prosopagnosia (Bauer, 1982; Gomori and Hawryluk, 1984; Habib, 1986) and in Capgras syndrome (Barton, 2003).

1. Historical concepts of disconnection as the basis for prosopagnosia

The revival of the concept of disconnection as the basis for dysfunction in alexia, aphasia and other behavioral syndromes (Geschwind, 1965) led naturally to many considerations of whether disconnection could also account for at least some cases of prosopagnosia (Aptman et al., 1977; Kay and Levin, 1982; McNeil and Warrington, 1991; Ross, 1980; Takahashi et al., 1995; Bauer, 1982; Gomori and Hawryluk, 1984; Habib, 1986). Two main proposals emerged, one focusing on *intra-hemispheric* disconnection, the other on *inter-hemispheric* disconnection.

First, regarding *intra-hemispheric disconnection*, it was noted that the bilateral lesions of the fusiform and lingual gyri described in early studies would also likely compromise the inferior longitudinal fasciculus (Catani and Thiebaut de Schotten, 2008, this issue; Catani et al., 2003), which connects regions of the occipital and temporal cortices (Benson et al., 1974). Damage to such a tract might disconnect visual processing in the occipital lobe from memory processing in the temporal lobe (Habib, 1986; Kawahata and Nagata, 1989; Kay and Levin, 1982; Meadows, 1974; Takahashi et al., 1995). If so, this intra-hemispheric disconnection might create a form of associative prosopagnosia, in which the products of intact perceptual processing of facial structure could not access residual facial memories (see also Ross, 2008, this issue). Such patients should show relatively preserved perceptual abilities and, ideally, relatively preserved facial memories as well.

Second, proponents of *inter-hemispheric disconnection* raised analogies with the syndrome of alexia without agraphia (Aptman et al., 1977). Inter-hemispheric disconnection accounts of alexia without agraphia postulate that left hemispheric regions specialized in decoding visual word forms lose access to visual input when a lesion both causes a right hemianopia and also interrupts callosal fibers from the right occipital lobe (Epelbaum et al., 2008, this issue), which process data in the remaining left hemifield (Dejerine, 1892; Geschwind, 1965). Could prosopagnosia represent the mirror-image deficit of alexia without agraphia, from deafferentation of a face processing region or network predominantly located in the right hemisphere? The initial reports of bilateral occipitotemporal damage as a prerequisite for prosopagnosia (Meadows, 1974; Damasio, 1985) would appear to cast doubt on this mechanism. However, subsequent reports of prosopagnosia with lesions limited to the right hemisphere (Landis et al., 1986; de Renzi, 1986) and functional magnetic resonance imaging studies showing more face-related activity in the right hemisphere (Haxby et al., 2000; Kanwisher et al., 1997) indicate a right-sided dominance in face processing. Thus the notion of prosopagnosia as the mirror image of alexia without agraphia cannot be so easily dismissed. Furthermore, the more variable nature of the left-sided lesions in cases with bilateral damage has suggested to some that the role of the left-sided lesion is to disconnect the left hemisphere's visual input (i.e., right visual hemifield) from face processing regions in the right hemisphere, much as splenial lesions do (Habib, 1986; Kay and Levin, 1982; Meadows, 1974). This mechanism would be particularly plausible in prosopagnosic patients with a left hemianopia. Theoretically, this inter-hemispheric disconnection could result in either an apperceptive or associative prosopagnosia, depending upon one's view of what aspects of face processing are lateralized to the right, whether it is perceptual encoding of face structure or the storage of facial memories.