Prosopagnosia

KA Dalrymple, Dartmouth College, Hanover, NH, USA

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Prosopagnosia is a visual perceptual disorder defined by the inability to recognize faces in the absence of impairments to lower-level vision or intellect. Prosopagnosia can be so severe that even close friends and family members will not be recognized. Indeed, the failure of recognition often extends to the patient's own face. One such example was related by a prosopagnosic man who said, "I was at a conference and I saw this other person walking. I thought it was probably someone I knew and so I smiled at (him) and started to greet (him), only to realize then that it was my own face and I was looking at a mirrored wall" (LH).

Historical Background

The first formal report of prosopagnosia came from Wigan (1844), who described a male patient who could not recognize faces, but had otherwise normal vision. In 1867, Italian ophthalmologist Antonio Quaglino provided a description of a 54-year-old prosopagnosic man with a right hemisphere stroke who displayed an inability to recognize the faces of persons previously known to him. Although this patient displayed a left hemianopia and achromatopsia, he retained good central vision and was able to read small print. On this basis, Quaglino argued that the patient suffered from a specific disorder of face recognition that could not be ascribed to a more basic perceptual disorder. In 1947, Bodamer derived a name for the disorder from "*prosopon*" (the Greek word for face) and agnosia (the inability to recognize).

Why Faces?

As social beings, humans rely heavily on information from faces. Faces contain a wealth of information about age, gender, emotion, and identity, which are all extremely important factors in determining how we interact with each other. This abundance of information is contained in a relatively restricted stimulus set: Faces are made up of a set of two eyes over a nose and a mouth. The spatial configuration of these elements is homogeneous across individuals. As a result, a dedicated system has developed in the brain that is responsible for processing this extremely special and meaningful stimulus. Selective damage to this system can result in prosopagnosia.

Etiology and Neuroanatomy

Prosopagnosia in its acquired form often results from damage to the ventral occipitotemporal area of the brain. Damage is usually bilateral, but evidence from some patients indicates that right hemisphere damage alone may be sufficient to cause the disorder. Damage to one or more of the brain's core face processing areas can lead to severe face recognition difficulties. These core face processing areas include the inferior occipital gyrus (occipital face area (OFA)), the middle fusiform gyrus (fusiform face area (FFA)), and the superior temporal sulcus (STS). These areas are responsible for different aspects of face perception and recognition, and have reciprocal connections to each other. Recently, it has become evident that damage to the anterior temporal areas of the brain may also lead to prosopagnosia.

Prosopagnosia also exists in a developmental form. Developmental prosopagnosia ((DP), also called congenital prosopagnosia) is characterized by face recognition difficulties from an early age in the absence of known brain injury or anomalies. Some researchers have found abnormal functional activation of the core face processing areas in patients with DP, whereas others have found that reduced structural connectivity in the ventral occipitotemporal cortex may explain impaired face processing in DP. Although there appears to be considerable neural heterogeneity, it is clear that exposure to faces at an early age is critical to the development of normal face processing and that a lack of exposure to faces can lead to face recognition difficulties. For example, individuals with bilateral congenital cataracts that preclude normal visual processing from birth have been shown to develop face perception difficulties – even when the cataracts are removed at a very young age (e.g., <1 year of age). There is also evidence for a genetic basis to DP. One study found that facial recognition abilities were more highly correlated between identical twins than between fraternal twins, and other studies have shown the existence of DP in several members of the same family.

How Selective is Prosopagnosia?

Many prosopagnosics have some degree of difficulty recognizing objects other than faces, but their difficulty with faces is usually the most consequential aspect of their agnosia. The presence of comorbid object agnosia may be due to the close proximity of object- and face-specific areas in the brain. However, much research on prosopagnosia has addressed the question of whether the face recognition impairment in prosopagnosia is most salient simply because faces are an especially difficult type of object to recognize, because faces are of special importance to humans as a social species, or because prosopagnosia is truly dissociable from object recognition difficulties. The answer to this question about prosopagnosia has important implications for our understanding of the normal human visual system. If all types of visual stimuli are

recognized using a single general-purpose system, then it should not be possible for brain damage to impair face recognition disproportionately. However, if face recognition is disproportionately impaired, this suggests that the human brain has a specialized face recognition system, which when damaged leads to prosopagnosia.

To determine whether prosopagnosia is truly selective for faces, and hence whether the human brain has specialized mechanisms for recognizing faces, it is important to assess object recognition as well as face recognition in patients with prosopagnosia to determine whether recognition difficulties are unique to faces. One of the first groups of researchers to address this issue directly was McNeil and Warrington. They studied case WJ, a middle-aged professional man who became prosopagnosic following a series of strokes. After becoming prosopagnosic, WJ made a career change and became a sheep farmer. He eventually came to recognize many of his sheep, although he remained unable to recognize most humans. The authors noted the potential implications of this dissociation for the question of whether human face recognition is 'special,' and they designed an ingenious experiment to exploit WJ's newfound career. In a series of tests they assessed WJ's ability to identify famous human faces, familiar sheep, and unfamiliar sheep. They compared his performance on these tasks to age-matched controls who were also sheep farmers. Healthy subjects performed at intermediate levels in all conditions. Healthy subjects, even those who, like WJ, worked with sheep, performed better with the human faces than with sheep faces. In contrast, WJ performed poorly with the human faces and performed normally with sheep faces. These data suggest that WJ's recognition impairment does not affect the recognition of all groups of visually similar patterns but is selective for human faces.

Another notable case that provides support for the existence of a specialized face processing system in the brain is patient CK. CK had normal face recognition abilities, and even above-average face detection abilities, yet was profoundly impaired at recognizing objects. Although not prosopagnosic, CK's case provides the other side of an important dissociation between face processing and the processing of nonface objects.

The behavioral dissociation between face and nonface processing is supported by experimental inhibition of brain areas that are specialized for processing faces and nonface stimuli. Pitcher *et al.* demonstrated a particularly impressive triple dissociation among face, object, and body processing by using transcranial magnetic stimulation to systematically inhibit areas of the brain thought to be specialized for processing these categories of stimuli. The authors found that inhibition of the right OFA selectively impaired face processing, whereas inhibition of the extrastriate body area (specialized for processing body information) selectively impaired body processing, and inhibition of the right lateral occipital area (specialized for processing object information) selectively impaired object processing. This provides strong support for the idea that the handling of face information is a unique process in the brain, separate from other high-level visual systems specialized for the handling of other types of visual information.

Famous Prosopagnosics

Jane Goodall, British primatologist, ethologist, and anthropologist, best known for her study of social and family interactions of wild chimpanzees in Tanzania, described her prosopagnosia in her autobiography, *Reason for Hope*. She wrote, "I suffer from an embarrassing, curiously humbling neurological condition called *prosopagnosia*, which, translated, means I have problems in face recognition." Goodall also reported that her sister has face recognition difficulties, providing anecdotal evidence to support the empirical finding that prosopagnosia has a genetic basis.

Oliver Sacks, behavioral neurologist, best-selling author, and professor of neurology and psychiatry at Columbia University, has written of his prosopagnosia in several publications. In the August 2010 issue of The *New Yorker*, he writes of a situation similar to the one described by LH (above): "Sitting at a sidewalk table, I turned toward the restaurant window and began grooming my beard, as I often do. I then realized that what I had taken to be my reflection was not grooming himself but looking at me oddly." Like Dr. Goodall, Dr. Sacks also reports a familial link for his prosopagnosia: His elder brother also struggles with face recognition.

Conclusions

Prosopagnosia is a profound inability to recognize faces. It can result from damage to face-selective areas of the brain (acquired prosopagnosia) or exist from birth or a very young age in the absence of brain damage (developmental prosopagnosia). On the basis of lesion studies in prosopagnosic patients and supporting evidence from functional imaging investigations in healthy subjects, ventral occipitotemporal regions, especially in the right hemisphere, are implicated in the pathophysiology of prosopagnosia and in face recognition in general.

See also: Agnosia

Further Reading

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Relevant Website

www.faceblind.org

Prosopagnosia research centers at Dartmouth College, Harvard University, and University College London.