



Review Paper

Prosopagnosia

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ABSTRACT

This article provides an overview of the existing literature on developmental prosopagnosia, a condition characterized by the lifelong condition that can be diagnosed solely on the basis of facial information. Given the rarity of this syndrome there is a limited number of case reports available. The review examines the current data and proposes assessment strategies for individuals experiencing prosopagnosia. It is known to have 2 types of prosopagnosia they include acquired and developmental types. Acquired prosopagnosia is a condition that can occur due, to brain injury leading to difficulties, in recognizing faces, whereas Neurodevelopmental impairment leads to developmental prosopagnosia, characterized by a lack of facial processing techniques. Rather than being a uniform condition, developmental prosopagnosia manifests in different sub forms with identifiable functional impairments. This article reviews the available evidence in detail, examining hypotheses regarding the developmental origins of prosopagnosia and the specific deficits associated with face recognition.

Prosopagnosia is characterized by difficulties in recognizing faces, and currently, no specific treatment is available. However, individuals facing this situation can adopt certain strategies and practices to enhance their ability to recognize people. It is also called face blindness.

Exploring and integrating these strategies into daily life could be useful for those affected by prosopagnosia, leading to improved communication and improved navigational skills in face-to-face encounters

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1. INTRODUCTION

Specialized face recognition

The amygdala, the fusiform frontal lobe, the posterior pituitary frontal cortex, a section of the ventromedial temporal cortex, and the superior temporal sulcus are among the brain regions that are particularly skilled in

processing facial images in humans. The above mentioned regions enable us to recognize and memorize unique facial patterns for numerous individuals². The perception of human faces relies on the recognition of distinct facial characteristics like the eyes, nose, and mouth, as well as the interpretation of their particular spatial configuration.

The historical background & emerging recognition

Despite the fact that challenges in recognizing faces were observed more than a century ago, our comprehension of prosopagnosia continues to evolve.

Historical milestones in understanding prosopagnosia, with thanks to Oliver Sacks:

1790: Franz Joseph Gall, physiologist, proposes the idea that mental functions originate not in the heart or liver but in the brain³.

1865: Paul Broca established a link between specific areas of the brain and specific neurological and cognitive functions.

1872: Hughlings Jackson described a typical clinical case involving faces and areas.

1947: German neurologist Joachim Bodamer coined the term 'prosopagnosia' and described three patients who had difficulty recognizing faces due to head injuries (acquired prosopagnosia), with no other difficulties with visual processing use of the character

1955: Christopher Pallis, an English neurologist, records a patient's experience (recorded in a diary) of the onset of prosopagnosia after a stroke.

1963: American actor and filmmaker Brad Pitt is reported to have prosopagnosia.

1980s: Emergence of CT and MRI scans, corroborated by earlier autopsy findings, revealed damage in the "fusiform face area" of the brain in individuals with prosopagnosia

1990s: Developmental (or congenital) prosopagnosia is recognized as a distinct neurological disorder present since childhood, common in many family members. In addition, in October 1996, Bill Choicer begins to accept the term "face blindness"; for this condition, the earliest known use is from an 1899 medical paper

In the chapter "Face-Blind" of his book "The Mind's Eye," Oliver Sacks, a neurologist who personally experiences face blindness, presents a comprehensive review of advancements in prosopagnosia research. He offers an accessible explanation of the neuroscience discoveries and discussions that have contributed to our current understanding of prosopagnosia⁴⁻⁵.

Prosopagnosia can manifest with additional challenges, encompassing difficulties such as:

Recognizing emotions: Individuals with prosopagnosia may encounter challenges in identifying and interpreting emotions displayed on people's faces.

Discerning age and gender: Difficulty may arise in accurately perceiving and distinguishing the age and gender of individuals.

Following TV programs or films: Recognition of characters and comprehension of plotlines in TV programs or films may be impaired, contributing to challenges in understanding and enjoying visual narratives. Oversight of familiar individuals⁶⁻⁷.

Challenge in recognizing or recalling faces, both in person and in photographs.

Types of prosopagnosia

They are divided mainly into

1. Acquired type of prosopagnosia
2. Developmental type of prosopagnosia

1. Acquired prosopagnosia

In the pre-21st century era, the documented instances of prosopagnosia were predominantly associated with brain injuries resulting from events like head trauma, stroke, or degenerative diseases. Such cases, known as acquired prosopagnosia, involved individuals who previously possessed initially normal face recognition abilities that later became impaired. People with acquired prosopagnosia often recognize their deficit because they had prior experience with normal face recognition⁸⁻¹⁰. If you have noticed a significant decline in your ability to recognize faces, it is advisable to consult a neurologist. Any sudden decline in this capacity could potentially indicate an underlying condition requiring immediate attention.

The acquisition of prosopagnosia, a condition marked by the absence of the ability to recognize faces, is a direct consequence of brain injury.

The first recorded case of acquired prosopagnosia was reported 150 years ago^{11, 12} but modern understanding of this condition received a major boost with the Bodamer¹³ report in 1947.

Since then it has been recognized that prosopagnosia can be acquired and can be caused by a variety of diseases such as depression, epilepsy, encephalopathy, epilepsy, Degenerative atrophy, or temporal lobe amputation¹⁴.

There are 3 subtypes of acquired prosopagnosia

A perceptual prosopagnosia

The problem is that a person has difficulty creating a clear picture of someone's face from what they see. This makes it hard for their memory to recognize the face properly. The memory of the face is not strong enough, so it doesn't reach the level needed to signal that the face is familiar or to access more information about the person.

Associative prosopagnosia

The patient can see and understand faces clearly, but the problem is connecting this detailed facial information with their memories of faces. Sometimes, experts think this happens because there's a disconnection between what the person sees and what they remember^{15,16}.

Amnesic variant

In some cases, it could be that the patient has simply lost their memories of faces. Some people might call this as an "amnesic variant" of prosopagnosia¹⁷.

2. Developmental prosopagnosia

Developmental prosopagnosia also called as congenital prosopagnosia, it is distinguished by difficulties in identifying faces that manifest early in life. These challenges come from the neuronal development impairments that mainly affect the processing of face mechanisms.

Researchers have identified common DNA mutations in individuals with congenital prosopagnosia, challenging initial doubts regarding a genetic relation to this condition which tend to run in families.

Notably some mutations related with congenital prosopagnosia are not inherited but are "de novo" mutations, occurring spontaneously in individuals without any family history condition.¹⁸

In individuals face recognition abilities widely vary with typical functioning, ranging from excellent to poor with most falling in between somewhere these extremes.

Individuals with developmental prosopagnosia generally fall at the end of the face recognition spectrum. At the opposite end of the spectrum are “super recognizers” who exhibit exceptionally high facial recognition skills.

Developmental prosopagnosia patients have never had normal facial recognition, making their damage less obvious to them. As a result, many are still unaware of their prosopagnosia even in the adulthood, creating challenges in diagnosing the condition in children.

Adults with developmental prosopagnosia frequently report traumatic life experiences because of their inability to recognize others. This leads to chronic anxiety, shame, guilt and limited social group¹⁹.

Anatomical basis of prosopagnosia²⁰

There is a strong evidence supporting the importance of bilateral lesions in the overall etiology of prosopagnosia. The clinical prevalence of bilateral disease, and the consistent bilateral lesions in autopsies to date, strongly suggest that this pattern is not coincidental and that it is not difficult to explain how the rarity of this condition is clarified by the prevalence of bilateral lesions. However the location of the left hemisphere lesion asymmetry remains puzzle. Although it is plausible to speculate that it may be important because of the approximate placement of the left hemisphere lesions to those in the right hemisphere, the cases in which left hemisphere lesions were non- occipitotemporal can be considered coincidental free and unrelated, as seen in common autopsy findings (Hecaen), 1957; 1962.²¹

Prosopagnosia causes

It refers to a condition where there is a problem caused in the specific site of brain that provide information regarding faces

Recent studies have confirmed that right hemisphere damage to the specific temporal-occipital areas mentioned above is sufficient to induce prosopagnosia. MRI scans of patients with prosopagnosia showed lesions isolated to the right hemisphere, while fMRI scans showed that the left hemisphere was functioning normally²²

The most common type of prosopagnosia occurs when individuals are unable to develop the ability to recognize faces. This form of prosopagnosia can be hereditary and run in families. It is important to note that prosopagnosia is distinct from memory dysfunction, memory loss, impaired vision, or learning disabilities.

The disorder is believed to stem from congenital factors or damage to a particular brain fold known as the right fusiform gyrus, which appears to play a role in coordinating the neural systems responsible for facial perception and memory.

If you do not develop the ability to recognize faces – this is the most common type and may run in families. Prosopagnosia is not related to memory dysfunction, memory loss, impaired vision, or learning disabilities.

The disorder is thought to be the result of congenital influence, damage, or impairment in a fold in the brain that appears to coordinate the neural systems controlling facial perception and memory (right fusiform gyrus).

- Few prosopagnosia symptoms are normally seen in children suffering from Autism and Asperger’s syndrome²³.
- Additionally, children with prosopagnosia can have a difficult time at school, as many school professionals are not well versed in prosopagnosia, if they are aware of the disorder at all.^[23] Recently, a database of children's faces and test for child face perception has been developed, which may offer professionals a way to evaluate if a child has prosopagnosia²⁴⁻²⁵.

Living with prosopagnosia

Living a successful life with this type of condition involves developing new ways and strategies in order to navigate.

The most common way to deal with this is to learn and find alternatives for identifying people around them, using unique techniques such as voice, clothing, or markings like tattoos.

In some cases, ethnicity is a factor - some people with prosopagnosia find it easier to recognize certain ethnic groups. There are some support groups on social media for people with this condition, as well as professional organizations with resources.²⁶

Diagnosis

The diagnosis of prosopagnosia is made clinically. There are a few neurocognitive diagnostic tests that can be used to aid with the evaluation.

1.Bielefelder Famous Faces Test (BFFT): This assessment involves presenting the subject with 70 standardized “Famous” faces, prompting them to either identify the individual or provide a description of the face²⁷.

2. Benton Facial Recognition Test (BFRT): In this test, the subject is exposed to a target face alongside multiple test faces, and their task is to correctly match the target face with the corresponding test face. .

3.Cambridge Face Memory Test (CFMT): Similar to the BFRT, the CFMT evaluates the patient’s capacity to match unfamiliar faces.²⁸ it is noteworthy that both the BFRT and CFMT focus on facial matching rather than recognition. Consequently these tests are more apt for assessing associative prosopagnosia, wherein patients can still perceive faces without difficulty.

In addition to these formal tests, there are free online questionnaires available for individuals to self-evaluate prosopagnosia. It is essential to note that, at present, there is no university accepted diagnostic standard, and further research is required to establish the reliability of these tests.

Neuroimaging

The introduction of functional imaging has revolutionized cognitive neuroscience, particularly in the realm of the research. It has precisely delineated the boundaries of the active networks and regions involved in facial perception. This includes a fundamental face network comprising the fusiform face area(FFA)²⁹ Occipital face area (OFA), and the posterior segment of the superior temporal sulcus ^{30,31}.

Additionally, an extended network encompasses the anterior temporal face area along with regions such as the inferior frontal gyrus and precuneus.^{32, 33} Although both hemispheres exhibit activation in response to faces,^{34, 31} the right hemisphere tends to manifest a stronger effect.

The advancement in functional and structural capabilities of Magnetic Resonance Imaging (MRI) has significantly contributed to the understanding of acquired prosopagnosia.

An important observation is that prosopagnosia can be caused by various types of lesions,³⁵ a fact that aligns with the understanding of the extensively distributed networks engaged in face processing. From the study of acquired prosopagnosia, two significant observations have emerged.

1. Lesions can occur on one side (unilateral) or both sides (bilateral), with a significantly higher likelihood of unilateral lesions being on the right side.^{32, 36, 37} While there have been instances of prosopagnosia subjects with left – sided lesions, these occurrences are rare, and most of the individuals in such cases have been left handed.³⁸⁻⁴⁰ This observation raises the potential consideration that these individuals might have had an anomalous hemispheric lateralization from the outset.

2. A crucial distinction arises between damage to the occipital- temporal region and damage to the frontal lobe. Recent functional MRI studies reveal that damage to the temporal area in the occipital cortex results in reduced activity in critical regions like the fusiform face area and occipital face area.⁴¹ Intriguingly, damage to the frontal areas does not impact activation despite the injury.⁴²

The fusiform face area, responding selectively to specific faces, is situated just above the cerebellum on the underside of the cerebral cortex. This intricate network of regions and their differential responses contribute to our evolving understanding of the neural processes underlying face perception.

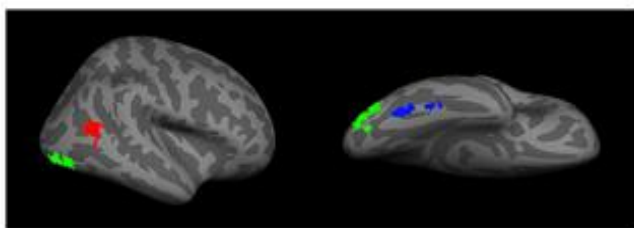
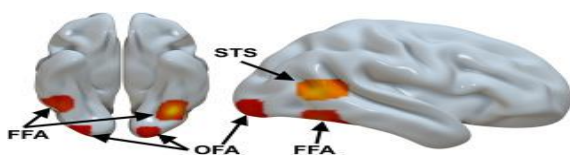


Figure no.1 above diagram represents the core face network

Include fusiform face area (blue), the occipital face Area (green) and the posterior superior temporal sulcus (red)



While looking at the right side of the brain, especially in lower and outer part (right hemisphere on the right) and examining it from the right lateral view, certain areas are more active when processing faces are compared to objects. It is observed from a functional MRI experiment.

The key face processing network includes specific regions such as Fusiform face area (FFA), occipital face area (OFA), and superior temporal sulcus (STS)

Measures to be taken:

- Inform in advance about the condition before meeting with someone.
- Seek assistance from family and friends to help in identifying people.
- Request introduction of them when you greet or meet someone.
- People with prosopagnosia must rely on voices and body language to distinguish people.
- Make a note of distinctive features and unique characteristics such as jewelery, accessories hairstyle.
- Use name tags or written notes about names, details especially at work.

These strategies can help navigate social situations when dealing with facial recognition challenges.

2. CONCLUSION

The prosopagnosia is an intriguing and intricate condition characterized by the inability to recognize faces, the examination offered valuable information regarding cognitive and neuronal mechanisms that include face perception. The literature on prosopagnosia emphasizes the diverse nature of this disorder, with both acquired and developmental forms, shedding light on the complex interplay between genetic predispositions and environmental factors.

The research on prosopagnosia not only enhances our understanding of specialized brain mechanisms for face recognition but also has practical implications for developing therapeutic interventions. As we unravel the neural pathways and cognitive processes involved in processing faces, there is potential for innovative rehabilitation strategies to emerge, providing hope for individuals grappling with the social and emotional difficulties posed by prosopagnosia.

Moreover, this review highlights the significance of interdisciplinary collaboration by incorporating insights from psychology, neuroscience, genetics and clinical practice to comprehensively address the multifaceted nature of prosopagnosia. As our knowledge continues to advance, it becomes increasingly clear that uncovering the mysteries surrounding prosopagnosia not only contributes to a broader understanding of the human brain but also holds promise for developing targeted interventions that can significantly enhance the quality of life for those affected by this condition.

Ongoing research will be crucial in the quest for these advancements, as it will help uncover new aspects of prosopagnosia and create opportunities for better diagnosis, treatment and support for individuals facing difficulties with recognizing faces.

3. CONFLICT OF INTEREST

The authors declare that there are no conflicts of interest.

4. ACKNOWLEDGEMENT

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