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Prosopagnosia

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Abstract

Prosopagnosia is a cognitive disorder that affects one's ability to recognize faces. Prosopagnosia can be caused by a congenital defect, or it can be acquired as a result of brain damage. Much research has been devoted to discovering the specific causes and effects of Prosopagnosia. Many case studies have been performed in order to determine the specific effects that each case of Prosopagnosia causes for various individuals suffering from the disease. This article discusses the various aspects of Prosopagnosia; specifically focusing on the behavioral, anatomical, and neurological implications.

Introduction

Prosopagnosia, also called face-blindness, is a cognitive disorder of face perception. A person's ability to recognize faces is impaired, while other aspects of visual processing and intellectual functioning may remain intact. There are two types of Prosopagnosia: Congenital or Developmental Prosopagnosia, and Acquired Prosopagnosia. Congenital Prosopagnosia is a face-recognition deficit that is life long, beginning in early childhood, and is not attributed to brain damage. Acquired Prosopagnosia refers to a condition that follows acute brain damage, usually brain damage specifically to the occipito-temporal lobe. The term Prosopagnosia is derived from the Greek words for "face" and "lack of knowledge." Although individuals with this disorder may be able to see normally, their ability to recognize faces is deficient. This disease can be life altering, as individuals with this disease may have limited social and behavioralinteractions. However, there are some Prosopagnosiacs who are able to lead normal, happy lives despite their inability to recognize faces. Scientists have performed many case studies and much research has been done to discover the underlying causes of Prosopagnosia. Both Congenital and Acquired Prosopagnosia are complex conditions caused by various factors. What are some of the neural, behavioral, and anatomical implications of Prosopagnosia?

Materials and Methods

In order to answer the question proposed above, many research papers and journal articles with relation to this topic have been read. Touro College's library database and the NYU library database were used to search for relevant studies and reviews. The next step taken was to look for articles that were referenced by those obtained through the Touro College Library and NYU search engines that seemed relevant. All of the articles and information that was accumulated through this research have been used in the attempt to conclusively determine the neurological, anatomical, and behavioral results of Prosopagnosia.

Results

Background on Congenital/ Developmental Prosopagnosia

Congenital Prosopagnosia (CP) refers to the impairment in face processing that is evident from the time someone is born. CP is not due to any brain damage and can occur in a person with intact sensory and intellectual functions. Individuals that suffer with CP are usually able to acknowledge that they are looking at a face; however they are unable to identify the specific face. As a result, they have come to rely on other cues such as voices, clothing, or specific accessories (hairlines, eyebrow shapes, etc.) to help recognize a person. CP can even affect the recognition of even the most familiar faces, such as close family members. In extreme cases, one may not even be able to recognize his- or herself. An important fact to understand regarding those with CP is that they have never experienced normal face perception. Therefore, research that is conducted relies largely on self-testimony and the testimony of parents (Behrmann, Avidan, 2005).

Developmental Prosopagnoisa, a more general label, refers to a disorder caused by anomalies occurring at any time during development, whereas the title of CP automatically assumes that prosopagnosia occurred at birth or early infancy (Susilo, Duchaine, 2013). To be categorized as someone suffering with Prosopagnosia, impairment in face recognition is required. However, it is important to note that face recognition is complex and there are multiple types of face recognition and face perception. For example, one needs to know whether a face is present among non-face stimuli, to determine whether two faces are the same or different, and be able to recognize the individual identity of a specific face. Studies show most patients with CP are able to detect faces from among other objects. Additionally, some individuals with CP find it easy to match faces (usually because they are able to identify key features), but when the reaction time or more specific measures of discrimination are measured and the tasks demanded are more difficult, the deficit becomes quite obvious. However, most CP patients fail in the area of identifying specific faces (Behrmann, Avidan, 2005).

Congenital Prosopagnosia: The Thatcher Effect

Case studies have been performed regarding the discussion on whether CP is specific to faces, as opposed to other objects. It is known that face processing typically involves individual identification, where one is shown a face and one responds with the individual's identity. This process is different than that for identifying others objects that are usually recognized at a basic level (for example, as a chair, an apple,or a house); face recognition requires the perception of very detailed features. People are able

to perceive and discriminate between objects based on either a single feature, involving a feature process, or based on the relationship between features, involving configural processing.
Scientists have theorized based on case studies that related configural processing to face perception. Face perception is often described as a configural process and it has been suggested that a loss of configural processing may be a clear indication of Prosopagnosia. In an effort to determine the relationship between configural processing and Prosopagnosia, a case study was performed. The 'Thatcher Effect' is a phenomenon where it becomes more difficult for a non- Prosopagnosiac to detect local changes in an upside down face, despite identical changes being obvious in an upright face. An experiment was performed to demonstrate that prosopagnosiacs, both CP and AP patients, do not suffer from the 'Thatcher Effect'. A control group of people who had no indication of prosopagnosia was formed. Another group of mixed CP and AP sufferers were collected. Then, two pictures of faces were held in the upright position with pictures of the same face flipped upside down next to them.

In order to recognize faces, each brain has developed configural processing to be able to distinguish different details of individual face features such as eyes, nose, etc. When a face is turned upside down, configural processing cannot take place and so those seemingly minor differences are very difficult to detect. Patients suffering with CP however, do not suffer from this effect. This can be because the area of their brain that controls configural processing and face perception is impaired. These results were also found among the patients suffering from AP, and those patients exhibited an even easier time detecting the similar facial features between the right-side-up faces and the upside down ones than the CP patients (Behrmann, Avidan, 2005).

Congenital Prosopagnosia: The Part-Whole Effect

Impairments in configural processing may affect other visual stimuli as well. Recognition of faces depends on the spatial relations between the components which need to be represented to distinguish between individuals. Another experiment was conducted to in order to obtain data regarding configural impairment in other areas. Five CP individuals and five controls were shown four compound letters at global and local levels. Two of the stimuli had identities that were consistent at the local and global level and two had identities that were inconsistent. This means that a large letter is composed of smaller letters, and can either be categorized as consistent or inconsistent. A letter consistent at both global and local levels would be a large letter H made up of small letter H's, while an inconsistent letter would be a large letter H shaped by small letter S's.

CP individuals showed normal speed at recognizing and identifying the local letters, however they were extremely slow at deriving the global whole from the local elements. They were able to identify the small components, but had difficulty in reading the large letter formed by the small parts. This can be another indication of a failure in representing the spatial elements of a display. This demonstrates that CP patients can usually identify or recognize individual aspects and features of a face, but cannot perceive the global picture and recognize the face (Behrmann, Avidan, 2005).

Another case study was performed to demonstrate a similar idea. Unlike most types of objects, faces are represented as a perceptual whole, meaning that there are many individual features that come together to form one image of a face. Therefore, the recognition of a face is referred to as holistic or configural face processing. This raises the possibility that face recognition deficits in CP patients may result from abnormal holistic face processing. To test this theory, a performance test was conducted called the 'Part-Whole Effect'. A control group of 38 non-Prosopagnosiacs were selected along with 38 CP patients. Participants were briefly shown a target face and then asked to choose which of two images placed side by side was the target face that they had been shown. This test was repeated with a picture of a specific facial feature instead of an entire face. Both the control group and the CP patients demonstrated the 'Part-Whole Effect' (Avidan et. al. 2011).

This refers to a greater ability to discriminate in the whole condition than in the part condition. Each group was slower to recognize target features than a target face. Although both groups were affected, the CP group did have a lower average accuracy than the control group. Interestingly, this low accuracy for discriminating in the part condition varied based on the specific feature. The patients with CP were suffered from the 'Part-Whole Effect' only when the target features were the mouth, eyes, and nose. With all other facial features, they were able to identify the target features as accurately as they could identify a target face (Susilo, Duchaine, 2013).

Congenital Prosopagnosia: Anatomical Implications

The ability to recognize faces is so important in humans that the brain appears to have an area solely devoted to that task, the fusiform gyrus. The fusiform gyrus is positioned between two lobes and is therefore a part of the temporal and occipital lobes. This area is believed to comprise the core visual representation system for faces. Specifically, this core is attributed with recognition of facial features. The fusiform gyrus is also responsible for face selectivity, meaning that it responds to images of faces more strongly than to any other objects.

In order to determine the functional relationship between the anatomy of the brain and facial perception, experiments have been performed utilizing fMRI technology. Brain imaging studies consistently find that the fusiform gyrus becomes active when people look at faces. Studies have also shown a strong correlation

between activity in this area and face recognition, leading scientists to believe that the fusiform gyrus performs essential functions for facial perception.

Face selectivity and repetition suppression, both forms of neural processing, were measured using fMRI technology. Their relationship to face recognition ability was then determined in a case study. A control group of 15 non-Prosopagnosiacs was selected along with a group of 15 CP patients. The CP group showed reduced face-selective responses than the control group. In addition, it was demonstrated by these studies and analyses that there is a correlation between high performance on tasks involving facial identity processing and fMRI face selectivity activity in the fusiform gyrus. This brain-behavior relationship is associated with behavioral factors relating to face identification. Further tests were performed that measured quantitatively the relationship between behavioral skill in facial recognition and fMRI test results. The tests and analyses that were performed successfully presented correlations between the anatomy of the brain, specifically the fusiform gyrus, and facial recognition, selection, and perception. Despite this, the scientists who performed these studies admit that they would like to improve their approach to this research. This could be done in a variety of ways, including greater number of participants, a greater number of behavioral measures performed by those participants, and a greater diversity of the behavioral measures. These improvements would help produce more accurate data regarding the brain-behavior relationship (Furl et. al., 2012).

Congenital Prosopagnosia: Behavioral and Social Implications

Face recognition plays an important role in social and behavioral cognition. Therefore, tests have been performed in an effort study face recognition abilities in CP patients. Specifically, scientists were interested in processing of facial stimuli with emotional valence and its differences from non-facial stimuli. This is important in order to understand the impact of emotional facial expression on the success of long-term memory. A group of 49 people were selected; 24 of them were CP patients and the rest were non-Prosopagnosiacs. The participants in this study were shown a series of images including neutral faces, positive and negative faces, and building facades. The pictures were shown for 3.82 seconds each, and the participants were asked to perform certain tasks. For example, for the faces, they had to determine the gender of the face. For the building facades, they had to decide whether it was a public building or a private one. The reaction times were studied, and there was a slightly lower performance by the CP patients than by the non-prosopagnosiacs. Interestingly, the slow response by the CP patients was only present with the images involving faces; for the non-face stimuli (i.e. the building facades) the CP patients had a quicker response time than the control participants.

Once the responses were analyzed, voxel-based morphometry was used to relate anatomical differences to memory success. This revealed that CP patients had a lower grey matter density. This low density spanned a variety of areas, including the right middle temporal gyrus and the left precentral gyrus, which is associated with the Brodmann area. The scientists then used fMRI data to compare the face processing of the CP patients and the control participants. There were three major areas of the brain that correlated with the decreased face recognition activities of the CP patients. These areas are the left fusiform gyrus, the right lateral occipital complex in the core face processing area, and the right DLPFC.

This data conclusively determined that CP patients have impaired long-term memory for faces, and for complex visual stimuli as well, although impaired memory is more severe with regards to faces as opposed to buildings (Dinkelacker et. al., 2010).

Background on Acquired Prosopagnosia

Acquired Prosopagnosia is a neurological syndrome which does not allow a person to recognize faces as a result of brain injury. This is in contrast to CP patients who have never had the experience of face recognition. Patients with AP can have many different types of lesions which cause their acquisition of this disease. Research has shown that the most common injury to the brain that results in AP is damage in the inferior or medial occipito-temporal cortex. This is not surprising, as the core area for face processing is found in that region. The core is comprised of the fusiform gyrus, also called the fusiform face area (FFA), as mentioned above, as well as the occipital face area (OFA) and the posterior superior temporal sulcus (pSTS).

The exact relationship between damage to each of these areas and the acquisition of AP is not very clear, but it has been proven that most cases of AP do result from injury to these areas. As is the case with many other acquired diseases, there are a multitude of possible injuries that can act as the causing factor. Therefore, it is difficult to determine what the extent of damage in Prosopagnosia patients is in these specific areas. For example, in the case of a particular patient with AP, it was determined that Prosopagnosia was caused by a right hemisphere lesion to the inferior occipital gyrus. As a result, there was no activity in the occipital face area, and the patient became unable to carry out face recognition. However, there was still activation of the right fusiform face area, containing the fusiform gyrus, and the pSTS. This phenomenon is hard to explain according to the current understanding of the hierarchical view of face perception. This means that a healthy human brain may be able to form direct pathways to bypass activation in the OFA, as opposed to a hierarchical pathway (Prieto et. al., 2011).

Acquired Prosopagnosia: The N170/M170 Effect

According to the recently discovered view of branched pathways involved in face recognition, there should be an early onset time to preferential activation to faces. This activation time should not be dependent at all on the integrity of the OFA. In order to test the accuracy of this statement, researchers endeavored to determine the time course of brain activation in these areas in both healthy people and in those with AP. Using electroencephalography and magneto-encephalography, the N170 was identified. This is a potential of negative polarity on the human scalp in the occipito-temporal area. The negative polarity appears between 130 and 170 ms after the presentation of the stimulus (i.e. the vision of a face) and continues to increase in amplitude when a face is presented along with another object. This is a marker of the first electrophysiological response to faces by the brain and is referred to as the 'N170/M170 Effect'.

Researchers are not sure which areas of the brain are the sources of this neural event, but they have identified the posterior and middle sections of the fusiform gyrus as primary contributors. A study was performed that reported the electromagnetic recordings on the scalp of patients with AP during stimulation by faces and other objects. During the course of this study, the researchers also wanted to prove definitively whether a lesion in the OFA prevents activation of the FFA or pSTS. This could be determined by testing whether early face-preferential responses, the N170/M170, were observed for those areas.

The patient that participated in this study had extensive brain damage and a severe case of AP. However, she also exhibited the N170 amplitude increase as a response to faces compared to non-face stimuli (i.e. other objects). These results and others demonstrate that the deformations in the patient's skull and scalp caused by her injuries, as well as the lesions in her cortex, did not affect her N170. When presented with a face and other object, a larger response was measured via neuroimaging studied in the fusiform gyrus and pSTS. In addition, fMRI data of this patient appears similar to that of healthy people. This information is understood when presented along with the electromagnetic findings of this study. They demonstrate that the patient's brain is able to differentiate between faces and other objects within 200 ms of seeing the image. This response time is as early as that of normal observers. Therefore, this confirms that the N170 face effect is related to the initial reaction to a stimulus, to the activation of a generic face representation. However, the patient's difficulty in recognizing faces lies in individualizing faces. This would mean that the electrophysiological response to identical faces would be the same as the response to different faces. When healthy people are presented with subsequent images of the same face, the amplitude of the N170 is reduced when compared to the amplitude when shown different faces one after another. However, in a patient with AP, the amplitude of N170 remains the same irrelevant of the faces being shown.

These results also demonstrate that the OFA is not involved, or not essential, in generating the N170. According to fMRI data alone, the FFA, the OFA, and the pSTS are all involved in preferential activation to faces. However, the electrophysiological findings of the N170/M170 help explain the relationship between the activation of these brain regions and face recognition. Because the patient in this study exhibited normal N170/M170 amplitude despite her lesion in the OFA, that area can be viewed as unessential for generating the N170/M170 increases. This supports other claims that have hypothesized that at least two of these three areas must remain functionally intact in order to generate the N170 face effect. Despite these positive findings, the researchers caution that some inconsistencies arose during this study. For example, despite the fact that the N170 face effect is possible without OFA activation, there is evidence of OFA contribution to the generation of N170/M170 in a non-Prosopagnosiac brain. Therefore, more research must be devoted to understanding the relationship between OFA activity and N170 generation (Prieto et. al., 2011).

Acquired Prosopagnosia: Residual Function

The three major areas of the brain that relate to face recognition are the FFA, the OFA, and the pSTS, as previously discussed. Within these regions, many aspects of face recognition take place. For example, perception of facial identity is linked to the fusiform gyrus, while initial perception of facial structures is linked to the OFA, and perception of facial expression is linked to the pSTS. These divisions of activity have been decided based on anatomic models but they may not be completely accurate. Therefore, it is interesting to study the surviving face-selective regions in patient with AP (Sorger et. al. 2007).

A study was performed using fMRI technology to assess the specific functions of cortical regions. The researchers set out to determine whether patients with AP exhibited any residual sensitivity to facial identity or facial expression in their surviving face-selective regions. Three patients with AP were involved in this study, two of whom had uninjured fusiform gyrii. Only one similar study had ever been performed and in that case, the fMRI data on the AP patient found that residual sensitivity to facial identity changes were present in an object-selective region of the ventral lateral occipital cortex. This is not the expected area; one would assume that the residual activity would take place in the fusiform gyrus area that was undamaged and therefore functionally intact. However, the two patients with intact fusiform gyrii involved in this study exhibited residual sensitivity to facial identity in the fusiform gyrus, consistent with its role in identity processing. This study successfully demonstrated that there is residual activity in the surviving face-selective regions of patients with AP. However,

because each patient's injuries are different, and because of the small group of patients involved in this study, it is difficult to determine the overall accuracy of the study (Fox et. al., 2013).

Conclusion

This review has gathered much research and information, concerning the anatomical, behavioral, and neurological aspects of Prosopagnosia. However, this disorder is extremely complex. Each individual with Congenital Prosopagnosia was born with their specific brain malfunction and the individuals with Acquried Prosopagnosia have different brain lesions and damage. As a result, it has become extremely difficult to understand the exact causes and results of Prosopagnoisa due to the specificity of this disorder. Research is continuing, however, to try and improve the understanding and the causes, along with an attempt to find a cure for the individuals with Prosopagnosia.

Abbreviations

CP	Congenital Prosopagnosia
AP	Acquired Prosopagnosia
DP	Developmental Prosopagnosia
OFA	Occipital Face Area
pSTS	Posterior Superior Temporal Sulcus
fMRI	(functional) Magnetic Resonance Imaging

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