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CONGENITAL PROSOPAGNOSIA: FACE BLINDNESS

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Abstract

A lifelong and severe weakening of face processing that is evident from birth is known as congenital prosopagnosia (CP), and it disregards intact visual and intellectual ability. The fusiform gyrus, which especially stimulates in reaction to faces, is the brain region typically linked to prosopagnosia. Because of the fusiform gyrus's functions, most people can distinguish faces from similarly complicated inanimate items with greater detail. The strategy used by people with prosopagnosia to recognize faces relies on their less sensitive object recognition system. Conversant facial recognition involves the right hemisphere fusiform gyrus more frequently than the left. Acquired prosopagnosia, which is most commonly encountered in adults, is caused by damage to the occipitotemporal lobe. It is unknown if this impairment is limited to the fusiform gyrus's role in face recognition or if it is also complex in highly handled visual stimuli. Prosopagnosia is divided into two categories: associative and apperceptive. The person with congenital prosopagnosia never fully acquires the ability to distinguish faces. Research on neuropsychology in great detail revealed that people with CP perform less well on face tasks such as matching faces, encoding/retrieving new faces, and counting renowned faces. Using ERP for neurophysiological analysis, it was discovered that people with CP generally had lower voltage of the N170 component, which is a sign of face processing in the temporal lobe. Nonetheless, among people with CP, there were significant differences in the N170 values.

Keywords: Face Recognition, Fusiform Gyrus, Occipitotemporal Lobe, Visual Stimuli.

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Introduction

The failure to recognize faces can have devastating significances for individuals suffering from this deficit. Prosopagnosia, as this disorder is termed, although rather rare has usually been familiar in individuals who have sustained brain damage in adulthood [2]. Acquired prosopagnosia (AP) has been recognized for a long time and has providing a unique window into the psychological and neural substrate of face processing. In recent years, attention has been paid to an analogous impairment, congenital prosopagnosia (CP), which refers to the impairment in face processing that is apparent from birth in the absence of any brain damage, and occurs in the presence of intact sensory too intellectual functions [2]. As is true in AP, CP individuals are typically talented to acknowledge that a face is present but are unable to identify the face and, hence, rely on voice or other cues such as clothing, gait or accessories for person identification [1].



Figure 1: The contribution of early visual experience to the development of complete configural face process faces are complex visual stimuli and their processing involves several different psychological processes (5).

Nature and extent of deficit in CP:

Even though an impairment in face processing is de facto required for the diagnosis of CP, face processing is not massive and can involve, for example, detecting whether a face is present among non-face stimuli, determining whether two faces are the same or different or recognizing the individual identity of a face [15]. Which of these

processes is concerned in CP is unclear and this is explored in some recent papers.

Models of Face Recognition

Face recognition is a multistage process ending with the identification of a person. These stages are imitated in cognitive models of face recognition, the most important being that of Bruce and Young [34]. Each box in the model represents a distinct cognitive process: while it is dispensable that these different stages occur in separate anatomic structures, some neuroanatomic models suggest that this may be the case. The model begins with producing a "facial percept", the encoding of the structural information about the face. This perception is matched to stores of face memories, termed "face recognition units", to determine whether the face has been seen before.

Tests of face familiarity

Previous tests of face recognition may have been less sensitive because their stimuli could allow subjects to use alternative strategies to circumvent poor face recognition, such as remembering hairstyles and clothing. Newer tests have addressed those boundaries by minimizing those extraneous cues. The most frequently used test of familiarity for recently viewed faces is the Cambridge Face Memory Test (CFMT), a test with high internal reliability. Although the original version of this test used only adult Caucasian faces, other versions have been created, such as the CFMT Chinese, CFMT-Australian, and versions, such as the CFMT-C and the CFMT-Kids.

Tests of face perception

Tests of face perception – that is, the capability to perceive differences between faces – do not establish the diagnosis of prosopagnosia. What they can do is validate if prosopagnosia is due to impaired encoding of the facial structure, also therefore is an apperceptive variant, or if such encoding is intact, which would point to an associative variant [12].

Neural investigations of CP

Only a small number of studies have led neural investigations of CP. Two studies have used induced response potentials (ERP) with specific exploration of the potential, recorded from a relatively circumscribed region at the posterior-inferior aspect of the temporal lobe. In one study, subject YT, observed the frequency. Only a small number of studies have conducted neural investigations of CP [18]. Two studies have used evoked response potentials (ERP) with specific exploration of the N170 potential, recorded from a relatively constrained region at the posterior-inferior aspect of the temporal lobe. In one study, subject YT, monitored the frequency.

Methods:

Computer tasks were run on a Macintosh G3, using any e prime 5.2 at a viewing distance of approximately 65 cm from the screen. In all experiments, stimuli persisted visible until the subject responded by pressing designated keys on the keyboard or by providing a verbal response. Instructions were to respond as speedily and accurately as

possible and practice trials were given before each experiment.

Face-related Tasks Famous face identification

Subjects remained shown two sets of famous faces. For the first set, 97 photographs were presented individually on a computer screen until subjects named each face, provided any other relevant information about the face (e.g., profession), or said they did not know anything about the face. A response was careful correct if the correct name was provided. For the second set, 56 photographs were of famous individuals and the remaining 56 were unfamiliar (e.g., famous actors or celebrities from other countries). Photographs were chosen so as not to cover any salient or diagnostic cues [17]. Roughly half the photographs were in colour and half in monochrome. Half the faces were male and the other half female. Images were collected to fit into a black oval to maintain equivalent size across pictures.

Greeble Discrimination

In each trial, two Greebles (or a Greeble and an object) look as if on the screen for a same/different judgment. Stimuli could differ at a basic level (a Greeble and a car), or could both be Greebles but differ at the gender (different directions of appendages), family (different main bodies), or separate (same direction of appendages and main body but different shape appendages) level [17]. There were 30 trials of each kind.

Configural Tasks

Low-level Visual Testing

To create log contrast thresholds across the range of spatial frequencies from 1 to 30 cycles per inch (cpi), a pair of images was presented in sequence, one of which contained the frequency-modulated image and subjects performed a forced-choice discrimination of first against second image. If the response was correct, a more difficult discrimination (decreased contrast by 0.2) was obtainable on the next trial. If the response was incorrect, the contrast was increased by 0.2. Each frequency bin (0.3, 1, 3, 10, 30) was tested in a separate block and a log contrast threshold was determined for each cpi using method of limits where threshold is defined as the value of contrast that produces 82% accuracy in response [24].

Another method Participants

All participants with CP

The three participants with CP were KM (CP1), a 60-year-old female, MT (CP2), a 42-year-old male and TM, a 28-year-old male (CP3), all of whom have joined in our previous experiments. All are right-handed with no neurological or neuropsychological devices aside from the impairment in face processing.

Participants with AP

The two patients with AP were RN (AP1) and SM (AP2). RN, defined previously, is a 49-year-old right-handed male who suffered an anoxic episode following myocardial infarction in 1998. A recent MR scan exposed several gross abnormalities (enlarged ventricles, widespread atrophy, most marked in bilateral parieto-

temporal regions) and density in homogeneities in occipitotemporal areas.

Neurologically normal control participants

Control participants were community volunteers and undergraduate students with no history of neurological or psychiatric sickness. For each experiment, two age and gender-matched controls were used for each AP and CP individual, and data from further 20 neurologically normal individuals were also added to form a large comparison sample [34]. All participants gave written consent and all had normal or corrected-to-normal visual acuity by self-report. Community volunteers were paid for their participation, and students participated in return for course credit.

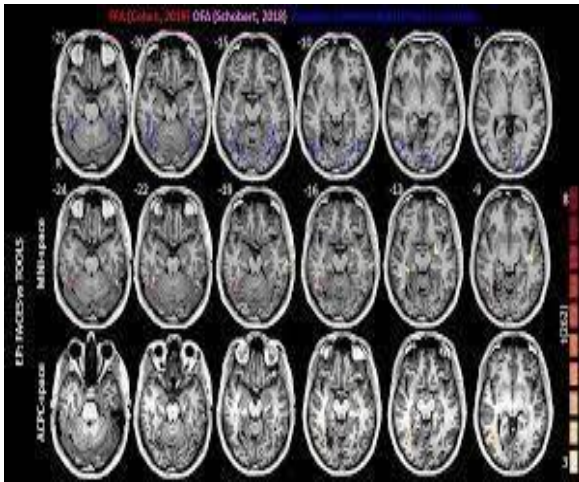


Figure 2: Structural MRI scans for AP patients AP1 (RN) b AP2 (SM). Scans were taken after behavioural testing, and several years post-onset for both individuals.

Face Identity Discrimination and Recognition

To imitate that the prosopagnosia participants were impaired at face processing, we administered a famous face recognition task and an unacquainted face discrimination task to each individual and to the control individuals. The Benton Face Recognition Test was also administered to all CP and AP participants, even though it should be noted that it is possible for prosopagnosia individuals to score in the normal range on this task. An additional face discrimination task was administered to the CP individuals and their matched controls [12].

AP1 was imprecise at between category discrimination and AP2 at both within and between category discrimination. AP2, but not AP1, also fell extra two standard deviations below the control mean in terms of d, and AP2 was also atypically slow in both conditions. CP1, CP2 and CP3 were within normal precision limits, in terms of both accuracy and precision. All images were taken from the internet and were collected with a standard black oval to remove non-facial cues. Unevenly half the faces were male and half female, and the number of Caucasian and non-Caucasian faces was equated across the familiar and unfamiliar sets.

Other possible responses were an incorrect name or 'do not know'.

Facial Expression Recognition

Having confirmed that the facial identity processing of the CP (and AP) individuals is impaired, we then went on to investigate their facial expression processing, using a very fine-grained task, cover all six basic facial expressions (anger, disgust, fear, happiness, sadness). Specifically, morphed expressions from all possible pairwise combinations of the Ekman and Friesen (1976) six straightforward facial expressions were used. These stimuli have been extensively used in previous research with other brain injured populations, and have been shown to serve as a very sensitive test of expression recognition. The stimuli were presented independently in a random order for a six-alternative forced-choice recognition response.

Results and Discussion

A group of five CP individuals participated in three series of studies, each speaking one of the issues laid out above. In all five individuals, CP was manifested without any obvious original neurological cause, and visual acuity and basic visual processing abilities were unimpaired (see Table 1 for biographical details and results of low-level visual tasks). In addition, to provide a clear indication of the extent of the impairment in the CP individuals, we also compared the discrimination of face and of nonface stimuli of the CP group with that of three individuals.

Table1: Congenital Prosopagnosia

TM	M	27	normal	6.
KM	F	60	Normal	7
NI	M	41	Normal	5.5
MT	M	40	Normal	6

Table 2: Acquired prosopagnosia

SM	M	27
CR	M	21
RN	M	41

The thresholds were firstly established for control subjects; any threshold for CP which exceeds 95% confidence interval at any of the tested CPIs would be considered different. All CP subjects performed in the normal range for all CPIs. Average (CI) for controls (07). Note that the lower the, the better the performance.

Face Discrimination and Recognition

Because standardized tests of face processing have

not proven effective in causing the disorder in prosopagnosia and measures of accuracy alone are often considered deficient, we tested the subjects on a series of customized face credit and discrimination tasks, obtaining measures of RT, accuracy, and sensitivity (A0). Also, because the summary statistics (mean, SD) yielded by a small control group is usually not reliable enough to reflect population parameters, we followed the procedure used by many researchers to assess abnormal performance in individual patients (Barton, Press, et al., 2002) and calculated the 95% confidence interval from the control data.

Discussion

The goal of this investigation was to address several outstanding questions about CP, a disorder in which individuals are disproportionately impaired at face processing in the absence of an underlying neural concomitant or cognitive deficit and in the absence of a low-level visual processing disorder. The specific questions pertain to the nature and severity of the face processing impairment, its specificity, and its relation to a more fundamental deficit in deriving configural information from visual images. The answers to these questions not only have relevance for this population, but can be used to inform theories of object recognition and the psychological and neural representation of faces versus other objects.

Nature and Severity of Face Processing Impairment

The data obtained across three experiments suggest that the CP individuals are impaired at assigning identity to faces and at discriminating between novel faces, especially as the discrimination becomes more taxing (differentiating gender vs. individual identity). Moreover, the representations derived by the CP individuals differ from those of normal individuals:

Specificity of Face Processing Impairment

Not only are the CP individuals impaired at face processing but, as is apparent from the experiments with common objects and with Greebles, they are also all affected to some extent in their recognition of nonface visual objects. Note, however, that their nonface processing is not as severely affected as their face processing and the deficit is largely evident in their slowed processing rather than in accuracy. Also, the deficit is not apparently a function of general perceptual slowing as the basic-level discriminations are performed well.

Relation of Face Processing Impairment Configural Representation:

The idea that faces are processed in a configural fashion whereas nonface objects are processed in a more piecemeal or feature-based way has received widespread support. The absence of an inversion effect for faces in our CP group is suggestive of a deficit in deriving the relational or second-order statistics from faces. Other case studies report similar results in AP and CP (18). Although there remains some disagreement on what constitutes "configural" most researchers agree that face inversion

affects configural processing and the absence of the inversion decrement suggests an abnormal reliance on a featural mode of representation.

Summary and Conclusion

Examining CP patients allows us to investigate the brain mechanisms underlying face recognition by fMRI and other imaging modalities, which are not impacted by injuries that could interfere with normal blood flow or neurovascularization (14). Ultimately, a significant number of CP patients who have been evaluated thus far, both in our research and in the literature, have a family member who also has face processing impairments (16). There is a lot of interest in this familial component, and research on the genetic process and propensity will help us begin to connect behavior to cortical development. As far as we are aware, there have been no reports of molecular DNA and linkage research, but the only genetic investigations reveal that the cumulation segregation ratios are consistent with a basic autosomal dominant mode of inheritance.

Author contributions

All authors are contributed equally.

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Declaration of Competing Interest

The authors have no conflicts of interest to declare.

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Reference

1. Behrmann M, Marotta J, Gauthier I, Tarr MJ, McKeeff TJ. Behavioral change and its neural correlates in visual agnosia after expertise training. *Journal of cognitive neuroscience*. 2005 Apr 1;17(4):554-68.
2. Duchaine BC, Nakayama K. Developmental prosopagnosia and the Benton Facial Recognition test. *Neurology*. 2004 Apr 13;62(7):1219-20.
3. Behrmann M, Avidan G, Leonard GL, Kimchi R, Luna B, Humphreys K, Minshew N. Configural processing in autism and its relationship to face processing. *Neuropsychologia*. 2006 Jan 1;44(1):110-29.
4. Bacon WF, Egeth HE. Local processes in preattentive feature detection. *Journal of Experimental Psychology: Human Perception and Performance*. 1991 Feb;17(1):77.
5. Singh Y, Meher JG, Raval K, Khan FA, Chaurasia M, Jain NK, Chourasia MK. Nanoemulsion: Concepts, development and applications in drug delivery. *Journal of controlled release*. 2017 Apr 28;252:28-49.
6. Bruce V, Valentine T, Baddeley A. The basis of the 3/4 view advantage in face recognition. *Applied cognitive psychology*. 1987 Apr;1(2):109-20.
7. Biran Y, Masters CL, Barnham KJ, Bush AI, Adlard PA. Pharmacotherapeutic targets in Alzheimer's disease. *J Cell Mol Med*. 2009 Jan;13(1):61-86. doi: 10.1111/j.1582-4934.2008.00595.x. Epub 2008 Nov 18. PMID: 19040415; PMCID: PMC3823037.

8. Barton JJ, Press DZ, Keenan JP, O'Connor M. Lesions of the fusiform face area impair perception of facial configuration in prosopagnosia. *Neurology*. 2002 Jan 8;58(1):71-8.
9. Suryasagar G, Bhargavi S, Kanakaiah B, Sowmya DK, Naresh M, Nama S, Rao CB. A review ON resealed erythrocytes. *Int. J. Biol. Pharmaceut. Res.*. 2013;4:290-6.
10. Behrmann M, Avidan G, Leonard GL, Kimchi R, Luna B, Humphreys K, Minshew N. Configural processing in autism and its relationship to face processing. *Neuropsychologia*. 2006 Jan 1;44(1):110-29.
11. Behrmann M, Kimchi R. What does visual agnosia tell us about perceptual organization and its relationship to object perception?. *Journal of Experimental Psychology: Human Perception and Performance*. 2003 Feb;29(1):19.
12. Hwisa NT, Gindi S, Rao CB, Katakam P, Rao Chandu B. Evaluation of Antiulcer Activity of Picrasma Quassioides Bennett Aqueous Extract in Rodents. *Vedic Res. Int. Phytomedicine*. 2013;1:27.
13. Behrmann M, Marotta J, Gauthier I, Tarr MJ, McKeeff TJ. Behavioral change and its neural correlates in visual agnosia after expertise training. *Journal of cognitive neuroscience*. 2005 Apr 1;17(4):554-68.
14. Bruce V, Valentine T, Baddeley A. The basis of the 3/4 view advantage in face recognition. *Applied cognitive psychology*. 1987 Apr;1(2):109-20.
15. Bruce V, Young A. Understanding face recognition. *British journal of psychology*. 1986 Aug;77(3):305-27.
16. Namballa M, Adimulapu A, Jesudasan RE. QbD Assisted Optimization of Microwave-assisted Synthesis of Polyacrylamide Grafted Tragacanth: Characterization and Instrumental Analysis. *Current Microwave Chemistry*. 2024 Apr 1;11(1):16-29.
17. BODAMER J. Die Prosop-Agnosie; die Agnosie des Physiognomieerkennens [Prosop's agnosia; the agnosia of cognition]. *Arch Psychiatr Nervenkr Z Gesamte Neurol Psychiatr*. 1947;118(1-2):6-53. German. doi: 10.1007/BF00352849. PMID: 18901747.
18. Duchaine BC, Nakayama K. Developmental prosopagnosia and the Benton Facial Recognition test. *Neurology*. 2004 Apr 13;62(7):1219-20.
19. Duchaine B, Nakayama K. Dissociations of face and object recognition in developmental prosopagnosia. *Journal of cognitive neuroscience*. 2005 Feb 1;17(2):249-61.
20. Duchaine BC. Developmental prosopagnosia with normal configural processing. *Neuroreport*. 2000 Jan 17;11(1):79-83.
21. Harris AM, Duchaine BC, Nakayama K. Normal and abnormal face selectivity of the M170 response in developmental prosopagnosics. *Neuropsychologia*. 2005 Jan 1;43(14):2125-36.
22. Hasson U, Avidan G, Deouell LY, Bentin S, Malach R. Face-selective activation in a congenital prosopagnosic subject. *Journal of cognitive neuroscience*. 2003 Apr 1;15(3):419-31.
23. Ariel R, Sadeh M. Congenital visual agnosia and prosopagnosia in a child: a case report. *Cortex*. 1996 Jun 1;32(2):221-40.
24. Kress T, Daum I. Developmental prosopagnosia: A review. *Behavioural neurology*. 2003 Jan 1;14(3-4):109-21.
25. Marotta JJ, Genovese CR, Behrmann M. A functional MRI study of face recognition in patients with prosopagnosia. *Neuroreport*. 2001 Jun 13;12(8):1581-7.