Developmental prosopagnosia with normal configural processing

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The configural processing hypothesis proposes that prosopagnosia results from a domain-general impairment in configural processing, and so predicted that all prosopagnosics would have impaired configural processing. In order to test this prediction, tests of face recognition and configural processing were presented to a developmental prosopagnosic. He was severely impaired in face recognition, but his normal performance on three tests of configural processing disconfirmed the configural processing hypothesis. Additional tests of low-level vision and object recognition found no evidence of impairments with material other than faces. The pattern of spared and impaired face recognition indicates that this case of developmental prosopagnosia is caused by a domain-specific inability to match novel views of faces with previously derived representations. *NeuroReport* 11:79–83 © 2000 Lippincott Williams & Wilkins.

Key words: Configural processing; Developmental prosopagnosia; Domain specificity; Face recognition

INTRODUCTION

Prosopagnosia, a loss in the ability to recognize faces, is easy to diagnose but difficult to explain. Some investigators argue that it is caused by an impairment of a recognition system specialized for faces [1–3] Others argue, in contrast, that it is caused by an impairment of a system or systems essential for face recognition, but not specialized for it [4,5]. The resolution of this debate is important for two reasons: it will illuminate the nature of the cognitive deficit(s) that cause prosopagnosia, and it will reveal the extent to which normal face recognition is performed by cognitive processes that are domain-specific and functionally specialized for that task.

Recently, a number of cases that strongly argue for the existence of a face-specific system have been published [1,6], but this evidence has been challenged by new empirical findings and methodological arguments that are consistent with a more domain-general causal account [4,7]. Herein, I report a case that contradicts one of the leading domain-general accounts: the configural processing hypothesis (CPH).

According to the CPH, the visual system contains no components that are specialized for processing faces. It does, however, have procedures specialized for configural processing: procedures that allow the recognition of an item as a whole in one glance. Tests of configural processing require identification of objects with deleted portions so that the perceiver must combine a number of individually meaningless parts to form a structured percept. According to the CPH, prosopagnosia results from a loss of configural processing [5]. In consequence, it predicts that all prosopagnosics should be severely impaired in tests of visual closure [5]. This prediction applies to all cases of prosopagnosia whether acquired or developmental, because the CPH is concerned with explaining the cause of prosopagnosia regardless of a case's particular genesis. In support of the CPH, Levine and Calvanio's review of the literature revealed no cases of prosopagnosia without configural processing difficulties [5], and an updated review [4] also failed to find any dissociations between configural processing and prosopagnosia.

Although no cases of prosopagnosia with detailed evidence for intact configural processing have been reported, a number of cases indicate the possible existence of this dissociation. One prosopagnosic performed normally on the Street completion test [8], two were normal with Gollin's incomplete drawings [2,9], and one was normal on both the Street test and Gollin's drawings [9]: these tests assess configural processing. Unfortunately, all of these findings were presented unaccompanied by the data from the control subjects, and these prosopagnosics were not given a range of tasks probing their configural processing. Therefore, it is premature to reject the prediction that all prosopagnosics will evidence difficulties in configural processing.

Herein, I report findings from B.C., a developmental

prosopagnosic, that thoroughly test the CPH. Unlike acquired prosopagnosia, the face recognition impairments of developmental prosopagnosics are not the result of brain damage occurring since birth. Only three cases of developmental prosopagnosia have been reported in detail, so in addition to its theoretical value, this case provides information about a rarely studied condition.

In fact, the paucity of reported cases may not properly represent the prevalence of developmental prosopagnosia. The individual reported upon in this article cofounded an internet support group for prosopagnosics with approximately 50 members, and the great majority of these individuals are developmental prosopagnosics. While many of these individuals have not been formally tested and so their diagnosis is not assured, their numbers indicate that developmental prosopagnosia merits more attention than it has received. Socially, developmental prosopagnosia can be a crippling deficit, and unfortunately it often goes undiagnosed due to its obscurity. Theoretically, it may provide us with crucial evidence regarding the developmental course, computational specificity, and genetic basis of face recognition.

CASE HISTORY

B.C. is a 52-year-old ambidextrous male with no history of head trauma who has been unable to recognize faces his entire life. When he does successfully recognize someone, it is through identification of his hairstyle, facial hair, or jeans. I say 'his' because B.C. is almost completely unable to recognize women: B.C. recognized male elementary schoolmates via their jeans, and he reports still being able to imagine each male classmate's jeans. He recently attended a street fair with numerous longhaired men and was able to recognize many of the longhaired men upon re-encountering them. He also reports that the presence of long hair and facial hair on men allows him to lipread and recognize facial expressions much better than he normally can.

Although he reports no other visual difficulties, his prosopagnosia is accompanied by central auditory processing deficit (CAPD) and motor difficulties. His CAPD makes it difficult for him to understand speech in noisy settings. His motor difficulties are most clearly manifest in a gait abnormality, and as a boy, he never enjoyed playing sports. Both conditions, as well as his prosopagnosia, appear to have a genetic rather than a prenatal origin, because other family members were reported to share these impairments. Unlike many prosopagnosics, B.C. has no navigational difficulties and in fact is an avid hiker. Previous testing of his eyesight showed him to have 20/20 vision, but he became farsighted in his forties.

His prosopagnosia has caused great difficulties throughout his life. B.C. is a very personable man, but his inability to recognize faces resulted in him continually losing friends. Upon entrance in the Navy, his reliance on facial hair, long hair, and jeans left him unable to cope with his colleagues' uniform appearances and he had a nervous breakdown after 5 days. Throughout B.C.'s life, he knew that he was different from others but was unable to determine what this difference was. With the help of friends, he identified his prosopagnosia in his late forties, and this realization has been very comforting for B.C. He is now quite interested in aiding other prosopagnosics, and he has constructed a very detailed website about his experiences with prosopagnosia (http://www.choisser.com/faceblind) as well as cofounding the internet support group mentioned earlier.

B.C. possesses a quite remarkable intelligence. On a recent IQ test, he scored 131, and on college and law school entrance examinations he scored extremely well on both verbal and mathematical tests. He received a BSc in electrical engineering and graduated from law school. Following an unsuccessful stint as a lawyer, he engaged in a very successful career as an electrical engineer.

TESTS AND RESULTS

To confirm B.C.'s prosopagnosia and determine whether deficits in configural processing were the cause of it, I tested him with a number of tasks probing his face recognition, and a battery of configural processing tasks. Following discussion of these tasks I report tests of low-level vision and object recognition used to assess the face specificity of his visual impairments. All of the tasks were presented on a computer screen except for the commercially available tasks. Control means were obtained for the commercially available tests from their respective manuals; for the tasks created in the laboratory, control means were obtained from undergraduate and graduate student subjects.

Is B.C. prosopagnosic? Consistent with his personal history, B.C. displays severe impairments on some tests of face recognition, even though he can perform normally on tasks that rely solely on mechanisms that operate early in the face recognition process (as defined by Bruce and Young's [10] model of face perception). Accordingly, he scored 50/50 on the word portion of the Warrington Recognition Test [11] and 46/50 on the face portion of the test. His normal performance on these tasks indicates that he has no trouble recognizing identical patterns of words and faces (which requires only the ability to match individual features in a spatial array).

He was mildly impaired on a test created in our laboratory in which he was presented with a frontal view of a face and then required to choose one three-quarter profile photo out of three that depicted the individual. The controls averaged 27.1/30 (s.d. 1.3); B.C.'s score of 24/30 placed him more than 2 s.d. below the control mean. Benton's Facial Recognition Test [12] requires matching target individuals with 1–3 identical photographs taken from different perspectives or under different lighting conditions. Although B.C.'s score of 43/54 on this task places him in the normal range, the test revealed his impairment. His matching was extremely slow (he usually took more than 1 min per item) and he reported using a feature matching strategy, focusing in particular on the eyebrows, rather than matching facial configurations.

In order to test B.C. with a more ecologically valid task, I created the One in Ten task. This requires the subject to recognize 15 photographs of a target individual, which differ in illumination, out of 150 photographs that are presented one at a time. In the study phase, three photographs of the target individual were cycled through three times for 3 s per photograph. Following this, he was presented with 150 test faces, one at a time, and he was

asked to respond as quickly as possible with a mouse click whether or not the photograph displayed the target individual. The 150 test photographs were broken into three groups of 50. There were an average of five photographs of the target individual in each set of 50 and none of the 150 test photographs were repeated following a target. A signal detection analysis was used to determine B.C.'s ability to discriminate between target and distracter individuals. Compared with the control subjects, B.C. was significantly less able to discriminate between the individuals, and he was also much slower. The mean d′, the measure of discrimination, was 3.61 (s.d. 0.486) for the controls and 2.15 ($z = -3.0$) for B.C., indicating far less ability to discriminate targets from distracters. Control subjects' 'yes' responses averaged 774 ms (s.d. 121) and their 'no' responses averaged 530 ms (s.d. 87); B.C. took almost twice as long, averaging 1399 ms ($z = 5.17$) for yes and 1012 ms ($z = 5.54$) for no. B.C.'s normal performance on other timed tasks, discussed below, demonstrate that his slow reaction times were not produced by a general psychomotor slowness, but rather reflect his deficit in face recognition.

As a final test of his face recognition abilities, he was asked to name famous faces, each of which was presented for 10 seconds. B.C. was severely impaired on this task, naming only six of 25 photographs (24% correct). Control subjects averaged 94.4% correct (23.6/25; s.d. 1.41), placing B.C.'s score more than 12 s.d. below the control mean. Following this, a paper and pencil matching task was presented to B.C. which asked him to match the names of the individuals in the famous faces task with their profession or another distinguishing characteristic. B.C. scored 25/25 (100%), demonstrating that he was familiar with all of the individuals on whom he had been tested. This means his poor performance on the photo task resulted from problems with face recognition, rather than from a lack of knowledge about the identity of the test targets.

Is B.C.'s prosopagnosia caused by deficits in configural processing?: To test the CPH prediction [5] that prosopagnosia is a byproduct of defective configural processing, I presented B.C. with the same three tasks from the Kit of Factor-Referenced Cognitive Tests [13] that Levine and Calvanio used with the prosopagnosic they tested. On the Gestalt Completion Test (which is very similar to the Street test), subjects have to identify a common object from a group of black blotches created by deleting parts of the object. (See Fig. 1 for examples from the three tasks.) B.C.'s score of 18 places him almost 1 s.d. above the control mean of 15.2 (s.d. 3.6). In the Concealed Words task, in which subjects must identify a word based on fragments of a printed word, B.C. scored 23 whereas controls averaged 23.6 (s.d. 6.4). Finally, on the Snowy Pictures task, subjects must identify objects from an outline drawing that is partly obliterated by snow-like splatters. B.C.'s score of 13 places him almost 2.5 s.d. above the control mean of 5.7 (s.d. 3.0). In summary, B.C. scored at or above the mean on the three tests of configural processing. He manifested no deficits in configural processing; therefore, his prosopagnosia cannot be the result of defective configural processing. This impaired face recognition with intact configural processing cannot be reconciled with the configural processing hypothesis.

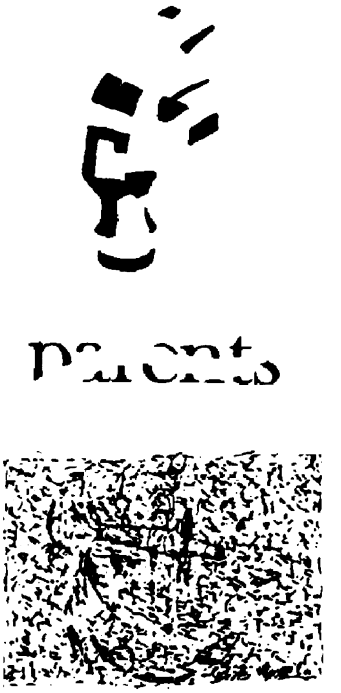

Fig. 1. Sample items from the Gestalt Completion task (hammer), the Concealed Words task (parents), and the Snowy Pictures task (anchor). Copyright 1976 Educational Testing Service, Princeton, NJ 08541 Reproduced under license.

Does B.C. manifest any visual deficits other than prosopagnosia?: The results so far eliminate one domain-general explanation for B.C.'s prosopagnosia: the CPH. They do not, however, establish that his prosopagnosia is caused by a defect in a recognition system that is specialized for faces. To establish this, other domain-general hypotheses must be eliminated. For example, if B.C. were to display deficits in tasks using materials other than faces, then one would have to seriously consider the possibility that his prosopagnosia was the result of a more general, non-face specific deficit, although not one involving configural processing. (Of course, such data would also be consistent with an impairment of multiple independent systems, one of which is specialized for faces). His normal scores on the Warrington word and face tasks, as well as his performance recognizing the objects and words in the configural processing tasks, suggest that his non-face specific processes are normal. To investigate domain-general alternative hypotheses further, I presented him with a variety of tasks testing low-level vision and object recognition.

On tests of perceptual matching drawn from the Birmingham Object Recognition Battery (BORB) [14] B.C. scored slightly above the control mean for each test. He scored 29/30 on the Length Match task, 29/30 on the Size Match task, 28/30 on the Orientation Match task, and 38/40 on the Position of Gap Match task.

On the BORB's Overlapping Figures task, difficulty with figure–ground segmentation is indicated by greatly elevated times for the naming of overlapping figures compared with times for non-overlapping figures. The figures consist of letters, geometrical shapes, and common objects. B.C.'s performance was normal on five of the six tasks. The

one anomalous score was not the result of difficulty with figure–ground segmentation. Although B.C.'s difference score for one of the two pages of common objects was high, this resulted from his difficulty with just one item on that page, and his performance on the other page of overlapping common objects was quite normal. In addition, although his difference score for this page was not normal, his time to name the overlapping figures on this page was still 6 s faster than the control average. He made no naming errors on any of the pages.

B.C. also scored normally on tasks from the Kit of Factor-Referenced Cognitive Tests [14] which require flexibility of closure. These timed tasks require the subject to hold a percept in short-term memory in order to disembed it from other more complex figures. His scores on these three tasks (Hidden Figures, Hidden Patterns, and Copying Task) placed him well above the control means.

On a variety of tests of object recognition, B.C. displayed no difficulties. He answered quickly and correctly named 256/259 objects from Snodgrass and Vanderwart's [15] corpus of line drawings. His scores on four tests of object recognition from the BORB were all within the normal range. He scored 25/25 on the Minimal Feature Match, 25/25 on the Foreshortened View Match, 26/32 on Part A (Hard) of the Object Decision task, and he drew objects from memory normally. In summary, B.C. displayed no difficulties with any of the tasks tapping low-level vision or object recognition. This indicates that B.C.'s visual deficits are restricted to the recognition of faces, and so places the locus of his deficits in face-specific mechanisms.

DISCUSSION

B.C.'s impaired performance on the face recognition tasks that require recognition of individuals in novel views, as well as his severely impaired performance on the famous faces task, demonstrate his prosopagnosia. In order to test the claim of the configural processing hypothesis [5], which states that prosopagnosia is caused by difficulties in configural processing, such that all prosopagnosics will fail tasks involving configural processing, I tested him with the Gestalt completion task, the Concealed Words task, and the Snowy Pictures task. His performance on these tasks clearly shows that B.C. does not have configural processing difficulties, and thus demonstrates that prosopagnosia can exist without configural processing decifits.

It is certainly plausible that configural processing deficits may be the cause of prosopagnosia in some individuals. However, B.C.'s results indicate that this is not invariably the case: higher-level impairments can cause prosopagnosia as well. B.C. has no evidence of deficits in low-level vision, in object recognition, or in configural processing; indeed, no visual deficits other than his difficulty recognizing faces. This strongly suggests that the cause of B.C.'s prosopagnosia is an impairment in mechanisms specialized for face recognition.

As other authors have suggested [4,16], it would be quite surprising if a process as complex as face recognition could be impaired in only one manner. The heterogeneity of face recognition impairments and lesion loci indicates that prosopagnosia is not a unitary disorder [17], and therefore any hypothesis specifying one underlying cause is unlikely to be correct.

B.C.'s performance on the different face recognition tasks indicates that he is able to structurally encode faces and recognize identical views, but that he has difficulties recognizing a face when it is presented in a novel view. B.C. scored normally on the Warrington Recognition Memory Test, which requires recognition of identical photos, whereas he displayed deficits with the famous faces task and the two tasks requiring recognition of novel views. This difficulty contrasts with his normal ability to match different views of objects in the Minimal Feature Match and the Foreshortened View Match, as well as his ability to recognize individuals through non-face routes. Although the evidence points to pure prosopagnosia, it is still premature to conclude that he has normal object recognition capabilities, because some authors [4] have argued that standard object recognition tasks (on which B.C. performed normally) test recognition at the level of basic categories, whereas face recognition requires subordinate level recognition.

The level of B.C.'s face recognition deficit is unlike that of the three other pure developmental prosopagnosics that have been thoroughly investigated. The inability of A.B. [18,19] and Y.T. [2] to recognize new faces in the Warrington task and their poor performance on the Benton task indicate an inability to structurally encode faces. Dr S [20], like B.C., performed normally on the Warrington task; but unlike B.C., Dr S also performed normally on the Benton Face Recognition task, thus demonstrating that she can recognize faces over changes of viewpoint and illumination. A famous faces task was the only test that Dr S failed. Thus, it appears that B.C..s inability to achieve face constancy with novel views places his impairment at a stage between that of A.B. and Y.T. in structural encoding and that of Dr S in accessing person-specific information.

CONCLUSION

The configural processing hypothesis proposes that prosopagnosia is not produced by face-specific deficits, but rather is produced by a more general deficit in configural processing. B.C.'s prosopagnosia with intact configural processing contradicts the configural processing hypothesis. His prosopagnosia appears to be the result of impairments in a face-specific recognition system. His normal ability to recognize faces when presented with identical photos, along with his inability to recognize faces across changes of viewpoint and illumination, implicate an impairment in matching novel views of faces with previously derived representations. This deficit is unique among the developmental prosopagnosics reported in the literature, and so reinforces the view [2] that, like acquired prosopagnosia, developmental prosopagnosia is not a unitary disorder.

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