SPECIAL ISSUE: ORIGINAL ARTICLE

PROSOPAGNOSIA WITHOUT APPARENT CAUSE: OVERVIEW AND DIAGNOSIS OF SIX CASES

Christian Dobel[1], Jens Bölte[1], Mona Aicher[1] and Stefan R. Schweinberger[2]

([1]Westfälische Wilhelms University, Münster, Germany; [2]Friedrich-Schiller University, Jena, Germany)

ABSTRACT

We compared six cases of congenital prosopagnosia to unimpaired participants using standardized test batteries, tailor-made experimental paradigms, and clinical questionnaires. Every prosopagnosic participant displayed deficits in recognizing famous faces and retaining novel faces over short periods of time. Other aspects of face perception such as judgment of emotional expression, speech reading and memory for faces and names were impaired to a lesser degree or only in single cases. No evidence was found for general visual deficits or social dysfunctions. Two of our six cases are first order relatives, and a further three report first-order relatives suffering from prosopagnosic symptoms. The results are in line with the idea of a genetic component to congenital prosopagnosia.

Key words: congenital, developmental, prosopagnosia, face perception

INTRODUCTION

The term prosopagnosia refers to an impairment in distinguishing and recognizing faces in the absence of generalized cognitive dysfunctions (Bodamer, 1947). For most cases in the literature, this condition results from brain lesions located bilaterally or unilaterally (right hemisphere) in the occipitotemporal cortices (Damasio et al., 1990; Farah, 1990), typically involving the fusiform and lingual gyri. Another type of prosopagnosia, which might be more common and which cannot be attributed to an acquired neurological damage, has been referred to as developmental (Duchaine and Nakayama, 2005), congenital (Hasson et al., 2003; Kress and Daum, 2003; Behrmann and Avidan, 2005; Behrmann et al., 2005), or familial prosopagnosia (Barton et al., 2003)[1]. This impairment has received relatively little attention until recently and typically single case studies have been reported with few exceptions. Even though it is assumed that it exists from earliest childhood on, only very few children were described in the literature (Ariel and Sadeh, 1996; Jones and Tranel, 2001).

In the present study, we report six participants with difficulties in recognizing faces from childhood onwards and compare their performance with that of normal, unimpaired participants. Anticipating the results, our prosopagnosic group showed difficulties in recognizing famous faces and in delayed matching to sample of faces. Other aspects of face processing such as judgement of emotional expressions, speech reading, or memory for faces were impaired to a lesser degree or only impaired in single cases. Within category discrimination of cars and pairs of glasses was found to be preserved. Before providing more detail on reported cases of congenital prosopagnosia, we briefly introduce the face recognition model originally suggested by Bruce and Young (1986), which, together with its more recently updated versions, provides the theoretical framework for this study.

Face Recognition

Taking data from healthy participants and prosopagnosics into account, Bruce and Young (1986; see also Burton et al., 1990, 1999; Schweinberger and Burton, 2003) developed a face recognition model for the perception of known and unknown faces as well as further abilities involved in face perception. In this model, a structural code (a 3D-representation of the visual stimulus face) is formed in order to access face recognition units (FRUs). These activate person identity nodes (PINs), which have to exceed a critical activation threshold for face recognition to take place. Name and semantic information retrieval follows person identification. Other independent functions, receiving input from the structural analysis, process emotional expression, facial speech, gender, age, or race.

[1]We will use the term congenital to stress that the impairment is likely to be present from birth. There are cases reported in the literature which are referred to with "developmental". The impairment is likely to result from an early brain damage.

The distinction between apperceptive and associative prosopagnosia (De Renzi et al., 1991) continues to be used frequently, and these two conditions are thought to arise from deficits at different levels of the model. In apperceptive prosopagnosia, a deficit in structural encoding hampers FRU activation (or face recognition) and also compromises other aspects of face processing. In associative prosopagnosia, familiar face recognition is also impaired, but other aspects of face processing such as matching or recognition of age, gender, or expression are largely preserved. Thus, structural encoding seems to operate at normal levels but FRU activation is either insufficient or is unable to trigger conscious person identification.

Congenital Prosopagnosia

To provide a basis for evaluating our own data, we will outline the abilities and deficits of prosopagnosic individuals reported in previous publications. We will only consider cases exhibiting prosopagnosic symptoms since childhood and for which no brain lesion, developmental or psychiatric disorder has been evident. Based on this criterion, we excluded A.B. (de Haan and Campbell, 1991; McConachie, 1976), H.D. (Kracke, 1994) and T.A. (Duchaine et al., 2003a) because they fulfilled the diagnostic criteria for Asperger syndrome, and A.L. (Joy and Brunsdon, 2002) because he displayed a range of developmental, motor, and cognitive problems, suggesting that his prosopagnosic symptoms are part of a more general deficit. However, it will become evident that even the remaining cases exhibit diverse deficits in various areas suggesting that a sound assessment of the pattern of deficits requires extensive and diversified testing.

A diagnostic cornerstone in all publications is the famous or familiar faces test. All prosopagnosic participants recognized fewer faces of famous or familiar persons than control participants.

In some cases, other aspects of face perception such as the recognition of gender, emotion, or age were impaired. Case R.P. had difficulties in judging gender and emotion from a face (Laeng and Caviness, 2001). T.A. (Jones and Tranel, 2001) could not classify gender. Duchaine et al. (2003b) tested N.M. and found only an impairment for facial identity, but not for emotions and objects. Further aspects of face perception were also tested in different studies, but performance typically seemed to be within normal limits. The "Benton Facial Recognition test" (BFRT; Benton et al., 1983), is often used with acquired prosopagnosia despite the fact that the name of this test is misleading, as it assesses the simultaneous matching of one or more exemplars of one face across different viewpoints or lighting conditions, but not face recognition. Face matching in the BFRT was abnormal in one case of congenital prosopagnosia [V.A. (de Gelder and Rouw, 2000)]. Duchaine and Nakayama (2004) observed normal performance for 7 of their 11 congenital prosopagnosic subjects.

Face memory has been assessed using the Warrington test (Warrington, 1984) and found to be impaired for four cases: Y.T. (Bentin et al., 1999), A.V. (de Gelder and Rouw, 2000), E.P. (Nunn et al., 2001), and S.O. (Kress and Daum, 2003). On the other hand, B.C. (Duchaine, 2000) and T.A. (Jones and Tranel, 2001) were unimpaired in this test.

Problems with object perception in addition to face processing were reported in two cases: Dr. S. (Temple, 1992), and T.A. (Jones and Tranel, 2001). Dr. S. displayed difficulties in making judgments about orientations of lines and in memorizing complex visual information (Temple, 1992). T.A. showed impaired or borderline performance in visuo-constructional and visuo-motor tasks. R.P., reported by Laeng and Caviness (2001), did not show any deficits during neuropsychological and neurophthalmological examinations and performed normally on a number of complex tests involving objects. His performance was impaired, however, when more fine grained testing methods were used (see below).

The question of dissociations between object and face perception was also addressed in two recent group studies on congenital prosopagnosia by Duchaine and Nakayama (2005) and Behrmann et al. (2005). In both studies small groups of congenital prosopagnosics were tested with a wide range of tasks involving face and object perception and recognition. Duchaine and Nakayama (2005) found in four of seven subjects a strong dissociation between face and object tests and took this finding as "support for the hypothesis that face and nonface recognition relies on separate mechanisms" (p. 249). In contrast, the five subjects of Behrmann et al. (2005) were impaired at object discrimination tasks and novel object tasks, however, to a lesser degree than at face discrimination. Consequently, the authors concluded that "the deficit may be attributable to a more fundamental perceptual disorder" (p. 1130). Duchaine et al. (2004) were able to corroborate their conclusion with a single case study, in which they studied a person with severe prosopagnosia who was able to perform within category learning as control subjects. Subjects had to learn within 10 hours to distinguish an artificial class of stimuli called greebles. The prosopagnosic subject was able to perform this task as well as controls. Therefore, it was concluded that all subjects including the prosopagnosic person "relied on ordinary object recognition mechanisms to recognize the greebles" (p. 472). These results provide further support for a dissociation between face and object recognition.