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Developmental prosopagnosia: A study of three patients

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Abstract

We studied perception in three patients with prosopagnosia of childhood onset. All had trouble with other 'within-category' judgments. All were deficient on face matching tests and severely impaired on tests of perception of the spatial relations of facial features and abstract designs, indicating a deficit in the encoding of coordinate relationships, similar to adult-onset prosopagnosia with lesions of the fusiform face area. Two had difficulty perceiving feature colour, which correlated with reduced luminance sensitivity. In contrast to adult-onset patients, saturation discrimination was spared in two and spatial resolution impaired in two. Curvature discrimination was relatively spared. Contrast sensitivity showed variable reductions at different spatial frequencies. We conclude that developmental prosopagnosia is similar to the adult-onset form in encoding deficits for the spatial arrangement of facial elements. Deficits in luminance perception and spatial resolution are more associated with defective encoding for basic object-level recognition, as shown on tests of object and spatial perception.

Keywords: Perception; Prosopagnosia; Developmental

1. Introduction

Prosopagnosia is the inability to recognize familiar faces in the absence of more generalized cognitive dysfunction. While some degree of visual (de Haan, Young, & Newcombe, 1987) or memory disturbance (Bauer & Verfaellie, 1988; Bruyer et al., 1983; de Haan et al., 1987; Malone, Morris, Kay, & Levin, 1982; Rizzo, Hurtig, & Damasio, 1987) for stimuli other than faces can be present, the recognition deficit is more severe and sometimes highly specific for faces. Most cases of this unusual problem are acquired through lesions of the right (de Renzi, 1986; Landis,

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Cummings, Christen, Bogen, & Imbof, 1986; Takahashi, Kawamura, Hirayama, Shiota, & Isono, 1995) or both(Damasio, Damasio, & van Hoessen, 1982; Meadows, 1974) occipitotemporal cortices, involving regions ranging from the anterior temporal lobe to the fusiform and lingual gyri. Just as normal face recognition is thought to involve a series of processing stages, acquired prosopagnosia is considered now to have several functional subtypes (Damasio, 1985; Takahashi et al., 1995). The broadest and most frequently used division is into two categories (Damasio, Tranel, & Damasio, 1990; de Renzi, Faglioni, Grossi, & Nichelli, 1991): apperceptive prosopagnosia, in which impaired formation of the facial percept prevents recognition, and associative prosopagnosia, in which an adequately formed facial percept cannot be compared with stores of facial memories.

Though rare, there are numerous well-studied cases of acquired adult-onset prosopagnosia in the literature. Developmental or childhood-onset prosopagnosia, however, has not been reported as often. The first case of this developmental form, AB, was described in 1976 (McConachie, 1976) and in later reports (Campbell, 1992; de Haan & Campbell, 1991). Since then, at least seven additional patients have been reported in single case studies. These cases are summarized in Table 1.

Although the numbers are few, comparisons with adult-onset prosopagnosia reveal some interesting points. The majority of the developmental cases are thought to have deficits at the level of structural encoding of the facial percept. Supportive evidence for this stems mainly from impaired performance on the matching of unfamiliar faces (the Benton Face Recognition Test, or BFRT), poor judgments of facial affect, gender or age (Ariel & Sadeh, 1996; de Haan & Campbell, 1991; Kracke, 1994; Young & Ellis, 1989), and deficient recognition of objects under more demanding conditions, as in line drawings, unusual views, and overlapping or incomplete figures (Ariel & Sadeh, 1996; de Haan & Campbell, 1991; Young & Ellis, 1989). Also, within-category judgments for non-face objects were impaired in at least three patients (de Haan & Campbell, 1991; Temple, 1992; Young & Ellis, 1989), suggesting that the defect was not as face-specific as reported for some adult-onset prosopagnosic patients (Farah, Levinson, & Klein, 1995; McNeil & Warrington, 1993).

Despite these parallels with adult-onset prosopagnosia, imaging in the few developmental cases in which this was obtained has not shown lesions of occipitotemporal cortex similar to those in the adult-onset form. Also, other deficits commonly associated with occipitotemporal lesions are unusual in childhood prosopagnosia. Only one patient had dyschromatopsia and superior altitudinal field defects (Young & Ellis, 1989), and only two had topographagnosia (de Haan & Campbell, 1991; Young & Ellis, 1989) while two others did not (Kracke, 1994; Temple, 1992). Residual covert or unconscious knowledge of familiar faces has not been found in any developmental prosopagnosic patients tested (Bentin et al., 1999; de Haan & Campbell, 1991; Young & Ellis, 1989), though it has been shown in several adult-onset cases (Bruyer, 1991; Young, 1994).

The paucity of other functional markers or imaging evidence of occipitotemporal damage raise the question of whether the type of perceptual encoding deficits in developmental prosopagnosia are similar to those in adult-onset prosopagnosia. It is not always the case that developmental disorders share the same pathophysiology as their adult counterparts. For example, childhood-onset dyslexia and the acquired pure alexia of left occipital lesions are considered different disorders. In adult-onset prosopagnosia, the type of perceptual encoding dysfunction continues to be debated. We have recently found that patients with lesions of the fusiform face area are severely impaired in their ability to discern the spatial relations of facial features (Barton, Press, Keenan, & O'Connor, 2002). These spatial relations are important in distinguishing the subtle variations in the basic configuration of features that all faces share. An orientation-specific expertise with perceiving these spatial relations