CONGENITAL VISUAL AGNOSIA AND PROSOPAGNOSIA IN A CHILD: A CASE REPORT

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ABSTRACT

We present an eight years old child, L.G. with congenital agnosia and prosopagnosia. This is a special case of perceptual deficits in a child which are discrete and exist in the context of a very high verbal intelligence. L.G. was administered an extensive battery of tests of cognitive functioning. He has intact basic visual skills, although his visual analysis is sometimes slow. L.G. can read, write and do math at age level or above. Four normal eight years old boys were used as controls on a selection of the perceptual tests, administered to L.G., which did not have normative data. L.G.’s object recognition skills bear the hallmarks of adult apperceptive agnosia. His visual memory and imagery are normal. Tests of face processing skills reveal, unlike adult prosopagnosics, severe deficits in addition to the familiar face recognition problem. L.G.’s agnosia and prosopagnosia are compared to the relevant literature.

Visual agnosia refers to an impairment of object recognition that cannot be explained by sensory loss. Patients with visual agnosia are often classified into one of two diagnostic categories: “apperceptive” and “associative” agnosia (Lissauer, 1890). Apperceptive agnosia is defined as “Any agnosia in which perceptual impairments seem clearly at fault.” Associative agnosia is defined as “Any of a number of conditions in which patients cannot name a seen object, despite the absence of gross visual impairments” (Farah, 1990, p. 161).

Cognitive models of normal visual information processing are increasingly used as frameworks for explaining neuropsychological impairment. Marr’s (1982) computational theory of visual perception is one of the influential models in the field. This model postulates that there are at least three levels of descriptions involved in the recognition of objects. Warrington and Taylor (1973) elaborate on his model. They believe that in many cases of visual agnosia initial structuring of the percept is intact (Marr’s first two stages of object description). Failure of identification is associated with Marr’s third level of processing, the “viewpoint independent” descriptions of objects. This third stage involves the assignment of a common identity to various object representations. Davidoff and Wilson (1985) and Warrington and Taylor (1978) propose four stages of processing of visual information which lead to object identification. Impairment in one of the first three is associated with apperceptive agnosia, in the fourth with associative agnosia. Some authors object to the historic dichotomic classification of visual agnosia. Benton and Tranel (1993) point out that it “... assumes a definable point of demarcation between perception and recognition”.

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Bauer (1993) believes that the field of agnosia research has transcended the notion of a dichotomy. He suggests that visual object recognition be viewed as a complex outcome of parallel processing.

Descriptions of children with visual agnosia rarely appear in the neuropsychological literature, presumably because they are rarely encountered. Visual perception achieves adult level efficiency in infancy. It seems feasible, therefore, that visual agnosia in children (specifically the apperceptive type), due to a brain lesion following infancy will resemble visual agnosia in adults. Indeed Young and Ellis (1989) describe a child with prosopagnosia and agnosia and conclude that her deficits are very similar to those of adult agnosics. A potentially different pattern of skills may be exhibited when the lesion occurred before birth. Agnosia, in this case, is not a disruption of a previously normal perceptual system, but rather visual processing that has developed abnormally since birth. Congenital visual agnosia is thus a disorder with potentially important implications both for the study of agnosia and for the understanding of normal development of object recognition.

Prosopagnosia is an impairment of the ability to recognize familiar faces. The specificity of prosopagnosia and its dissociation from various other cognitive deficits is still debated. De Renzi, Faglioni, Grossi et al. (1991) believe that we should conceptualize prosopagnosia as a dichotomy, parallel to the one we have for visual agnosia. Apperceptive prosopagnosia, in their opinion, is a type of visual agnosia and therefore not specific, while associative prosopagnosia is a deficit of a specific category of semantic memory. They suggest that when recognition impairment is restricted to faces, the deficit should be considered a mnestic dysfunction, associated with the category of faces. Benton and Tranel (1993) present a similar view. They believe that unknown face discrimination and familiar face recognition represent discrete and possibly unrelated functions. They are willing to accept only deficient recognition of familiar faces as an index of prosopagnosia. Just as in the field of agnosia, the descriptions and explanations of prosopagnosia often rely on cognitive models of normal functioning. Bruce and Young (1986) present a comprehensive cognitive model of normal processing of faces leading to recognition. Prosopagnosia may result from lesions at different sites on this model.

Prosopagnosia in children is also rarely described, although it is probably more frequent than reported, since there are several reports of adults with developmental prosopagnosia.

The relationship between prosopagnosia and visual agnosia has been the focus of many theoretical and clinical debates. Many researchers consider prosopagnosia to be part of a more general visual recognition disorder. Humphrey and Riddoch (1987) argue that since differences between faces are minute they are simply more difficult to tell apart than objects, yet the basic recognition deficit is the same. Other researchers believe that deficient recognition of familiar faces cannot be explained by an inability to notice the minute differences of facial features, since some patients can distinguish between very similar faces, yet not recognize familiar ones. Prosopagnosic patients can also judge gender and emotional facial expressions (Berhman et al., 1992; Humphreys et al., 1993; Sergent and Villemure, 1989; Tranel et al., 1988). Sergent et al. (1992) believe
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