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Research report

When apperceptive agnosia is explained by a deficit of primary visual processing

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ABSTRACT

Visual agnosia is a deficit in shape perception, affecting figure, object, face and letter recognition. Agnosia is usually attributed to lesions to high-order modules of the visual system, which combine visual cues to represent the shape of objects. However, most of previously reported agnosia cases presented visual field (VF) defects and poor primary visual processing. The present case-study aims to verify whether form agnosia could be explained by a deficit in basic visual functions, rather that by a deficit in high-order shape recognition. Patient SDV suffered a bilateral lesion of the occipital cortex due to anoxia. When tested, he could navigate, interact with others, and was autonomous in daily life activities. However, he could not recognize objects from drawings and figures, read or recognize familiar faces. He was able to recognize objects by touch and people from their voice. Assessments of visual functions showed blindness at the centre of the VF, up to almost 5°, bilaterally, with better stimulus detection in the periphery. Colour and motion perception was preserved. Psychophysical experiments showed that SDV's visual recognition deficits were not explained by poor spatial acuity or by the crowding effect. Rather a severe deficit in line orientation processing might be a key mechanism explaining SDV's agnosia. Line orientation processing is a basic function of primary visual cortex neurons, necessary for detecting "edges" of visual stimuli to build up a "primal sketch" for object recognition. We propose, therefore, that some forms of visual agnosia may be explained by deficits in basic visual functions due to widespread lesions of the primary visual areas, affecting primary levels of visual processing.

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1. Introduction

Visual agnosia is a modality-specific perceptual disorder, characterized by impaired shape recognition, which affects the recognition of figures, objects, faces and letters. Lissauer (Lissauer, 1890) distinguished between *apperceptive agnosia*, an impairment of perceptual processing, and *associative agnosia*, where visual perception is roughly spared, but a disorder exists in accessing stored mental representations of concepts from vision. Patients with *associative agnosia* can distinguish whether two visual stimuli are similar and can produce an accurate copy of an unrecognized figure through passive reproduction. In contrast, patients with *apperceptive agnosia* usually fail in copying and in visual discrimination tasks. However, in less severe cases, they can discriminate and compare the size of two shapes, distinguish a figure from a background and distinguish two overlapping figures. Apperceptive agnosia has further been divided into form agnosia and integrative agnosia. Form agnosia causes a deficit in shape and form discrimination, attributed to an inability to build shape representations from visual cues; integrative agnosia allows access to components or parts of shapes, but compromises integration of these component parts into a coherent whole. In general, perceptual deficits associated with integrative agnosia appear less severe than form agnosia (Farah, 2004). In either case, one dominant view in the field arguments that selective lesions to high-order modules of the visual system underlie both form and integrative agnosia (Humphreys & Riddoch, 1987a).

By definition, agnosia should not be explainable by primary sensory deficits such as poor visual acuity, visual field (VF) defects, or problems in colour, movement or depth perception (Farah, 2004). However, from a systematic review of previously reported agnosia cases (see Table 1), many patients also exhibited *visual field defects* (e.g., H.J.A. Humphreys & Riddoch, 1987b; see Bay, 1953; Ettlinger, 1956) either in the periphery, at the centre of the VF (e.g., micro-scotomas, see Campion & Latto, 1985; Campion, 1987) or both. Also, *visual acuity* was rarely formally tested, as agnosia patients are not able to perform common acuity measures (e.g., Snellen, 1862) which require letter or object recognition. Furthermore, given the absence of standard measures of visual acuity, comparing results across studies is problematic. Thus, sensory deficits may, at least partially, explain observed perceptual deficits in shape recognition.

Given the nature of lesions causing apperceptive agnosia, comorbid primary visual deficits are likely to be present. Most lesions are caused by anoxia, due to carbon monoxide intoxication or cardiac events (Caine & Watson, 2000). Such lesions often induce widespread neural loss, involving primary visual areas. Typically, patients initially suffer cortical blindness, and after some recovery, they present signs of agnosia. Thus, considering the aetiology and location of the cerebral damage, patients with apperceptive agnosia might present elementary visual deficits which can be identified with a proper psychophysical assessment.

Thus, prior to diagnosing agnosia, a deeper analysis of visual functioning is necessary to identify the mechanism underlying impaired shape recognition. Such analyses must precede attributing deficits to impairments in high-level modules of the visual system involved in shape representation from visual cues (form agnosia), or from integrating visual details into a synthetic whole (integrative agnosia). More specifically, assessments should target primary visual defects, such as spatial acuity, orientation discrimination and crowding. Diffuse neural damage to primary visual areas might reduce *visual acuity*. Low spatial acuity precludes distinguishing individual features of visual stimuli for recognition, and therefore, a deficit at this level might results in problems in form perception resembling apperceptive agnosia. Line orientation may be another factor. According to most popular models of visual perception (e.g., Marr & Hildreth, 1980), detecting the edges of a visual stimulus is the primary level of analysis necessary for shape perception; orientation-selective neurons in the primary visual areas support this function (Hubel & Weisel, 1959; also see: Ferster & Miller, 2000, for a review). Line orientation thresholds (Makela, Whitaker, & Rovamo, 1993) increase at higher eccentricities from the fovea. Thus, where defects involve the centre of the VF, perceptual deficits in apperceptive agnosia patients might actually be poor orientation perception. Finally, object recognition in healthy subjects dramatically decreases in the periphery because of the so-called "*crowding effect*" (Pelli & Tillman, 2008). The visual system recognizes objects by detecting and then combining their features. Crowding occurs when objects are too close together and details from several objects are combined in a fused, unrecognizable, percept. This effect does not occur in central vision, and increases proportionally for stimuli presented at increasing distance from the fovea. Thus, in cases of defects at the centre of the VF, apparent apperceptive agnosia might be a function of a crowding effect.

In the present study, we tested these visual functions in a patient (SDV) exhibiting both the typical lesion profile and the typical symptomatology consistent with apperceptive agnosia, in order to investigate whether his apparent agnosia could be a function of acuity, orientation sensitivity or crowding difficulties. SDV suffered a heart-attack, and consequent brain anoxia, three years before our examination. A bilateral lesion of the occipital cortex initially caused cortical blindness, which progressively recovered. At screening, SDV was able to navigate, interact with others, and he was autonomous in daily life activities. However, he was unable to recognize objects from drawings and figures. He could recognize real objects by touch, but not by vision. He could not read and did not recognize familiar faces. He was able to recognize people from their voice and movements. Motion and colour perception appeared preserved. Thus, SDV presented typical signs of visual agnosia. At formal testing, he also showed VF defects: he was blind at the centre of the VF, up to almost 5°, bilaterally; visual detection improved in the periphery.

2. Case history

SDV is a 44-year-old, right-handed man, with 8 years of schooling. He suffered an electrocution-induced heart-attack 3 years before the present examination. Following the event,