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DEVELOPMENTAL COLOUR AGNOSIA

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ABSTRACT

Colour agnosia concerns the inability to recognise colours despite intact colour perception, semantic memory for colour information, and colour naming. Patients with selective colour agnosia have been described and the deficit is associated with left hemisphere damage. Here we report a case study of a 43-year-old man who was referred to us with a stroke in his right cerebellar hemisphere. During the standard assessment it transpired that he was unable to name coloured patches. Detailed assessment of his colour processing showed that he suffers from a selective colour agnosia. As he claimed to have had this problem all his life, and the fact that the infratentorial infarct that he had incurred was in an area far away from the brain structures that are known to be involved in colour processing, we suggest that he is the first reported case of developmental colour agnosia.

Key words: colour, agnosia, perception, developmenta

INTRODUCTION

Colour agnosia is one of the classical neuropsychological syndromes. A striking clinical description of what is meant by colour agnosia was given by Kleist (1934), who stated that "… *a patient who has lost the general concept of colours behaves to colour stimuli as a normal subject behaves to certain odours. We can discriminate a large number of odours but we do not have general concepts for related types of smells and no names"*. The term refers to a selective impairment in the recognition of colour that cannot be explained by perceptual, memory, or language deficits due to impairment in the internal colour space (Klein and Stack, 1953; Bruyer, 1977; Kinsbourne and Warrington, 1964; Beauvois and Saillant, 1985; Grüsser and Landis, 1991; Davidoff, 1996). It must thus be separated from cerebral achromatopsia, i.e. a selective colour perception disorder, and it can also be dissociated from colour anomia, which refers to a specific problem in naming colours as a result of a lexical impairment and not due to an impairment in the internal colour space (Roberson et al., 1999). In addition, general confusion or disorientation should be excluded as possible causes. Colour agnosia is selective in that it only affects colour, while the recognition of other primary visual cues, such as motion and shape, and higher order visual categories, such as objects and faces, remain unaffected. It is associated with lesions in the left temporo-parietal region and is therefore often but not necessarily associated with aphasia (Stengel et al., 1948; Varney and Digre, 1983).

The first reports on colour agnosia date from the end of the nineteenth century. For instance, Lissauer (1890) described a patient with an intact ability to discriminate and perceive colours, but who made errors when asked to name colour patches or to name an object associated with a visual presented colour patch. This patient was also "unable to point to the colour of a canary in the absence of colour vision impairments". This observation was against the then dominant view that the inability to retrieve the colour of an object was a language disorder (Davidoff and Fodor, 1989). Although several cases of colour agnosia were subsequently reported in the literature, e.g. Kinsbourne and Warrington (1964), Hécaen et al. (1974), and Beauvois and Saillant (1985), the consensus on what constitutes colour agnosia appears to be based more on clinical definition than on empirical testing or theoretical accounts (Beauvois and Saillant, 1985).

To dissociate colour agnosia from cerebral achromatopsia (Zeki and Marini, 1998; Bartels and Zeki, 2000), colour anomia (Beauvois and Saillant, 1985), or colour amnesia (Grüsser and Landis, 1991) performance and a variety of perceptual and neuropsychological tasks need to be taken into account. That is, visual sensory assessment of colour, but also of visual features such as shape and brightness should be normal to rule out explanation in terms of (isolated) perceptual impairments (Miceli et al., 2001). The general knowledge of colour, i.e. the definition of colour, and object-colour knowledge, i.e. the ability to name the prototypical colour of objects, such as a fire engine or a banana, should be intact to exclude colour

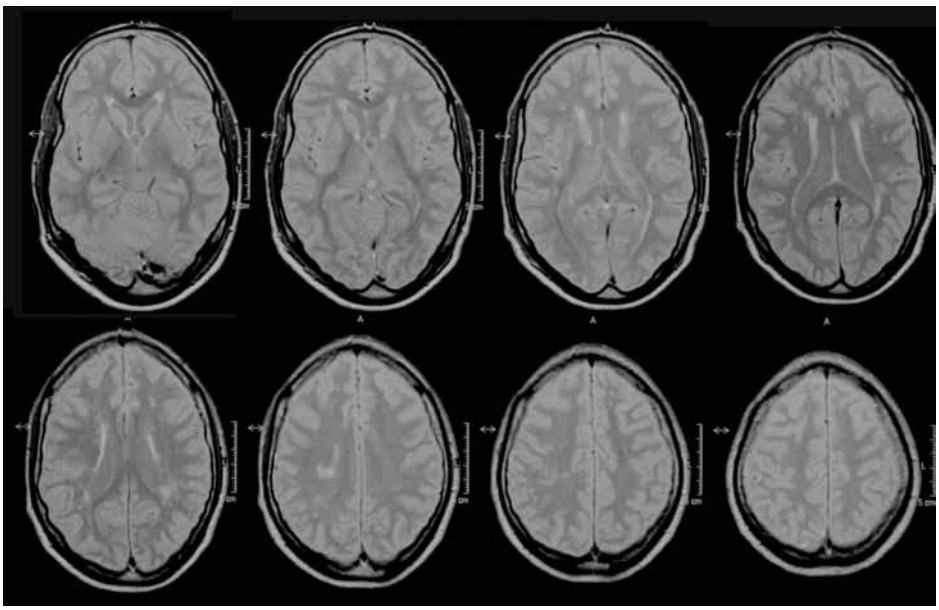

Fig. 1 – *No stroke lesion or brain abnormalities in the relevant cortical areas can be found on the MRI-slices (TSE) of the occipitotemporal and (occipito-) parietal cortices of M.A.H. at 14 days post-stroke.*

amnesia (Goldenberg, 1992; Davidoff, 1991). Furthermore, the matching of isoluminant coloured patches should be intact, in contrast to impairment in both naming and pointing to coloured patches on verbal command (Beauvois and Saillant, 1985). In addition, the ability to decide whether an object colour is veridical or not is impaired in cases of colour agnosia (Davidoff, 1991). Due to a loss of colour concepts and representation the categorization of colours is expected to be disturbed in colour agnosia, despite the ability to sort colour on the basis of individual matching (Roberson et al., 2000; Pilling et al., 2003). It is clear that a substantial analysis of the impaired and spared abilities is required before a convincing diagnosis of colour agnosia can be reached.

Until now, colour agnosia has only been described as a result of acquired damage. Other forms of higher-order recognition deficit, such as prosopagnosia (e.g., de Haan, 1999; Barton, et al., 2003) and alexia (Ley, 1938), have also been observed in healthy normal people without neurological or birth complications. These developmental cases shed light on the way in which function specialisation evolves in the brain. For instance, parallels between acquired adult-onset and developmental or childhood-onset prosopagnosia demonstrated comparable subtypes and revealed contributing perceptual encoding deficits in prosopagnosia suggestive for common functional mechanisms (Barton, et al., 2003). These parallels can only be discerned by comparison of acquired adult-onset and developmental cases, especially in the paucity of imaging evidence in developmental cases. Acquisition of colour terms and colour-object knowledge in normal subjects have been studied extensively (e.g., Bornstein, 1985; Pitchford and Mullen, 2005; Roberson et al., 2004), as well as the loss of colour names and knowledge in (semantic) dementia (e.g., Ukita et al., 1999). Moreover, the way colour knowledge evolves and is represented in the brain appears to be basically understood. However, impairments in higher order colour perception and its underlying mechanisms are not yet straightforward. Here, we report the first case of developmental colour agnosia. The diagnosis of colour agnosia is based on an extensive series of tests of colour processing.

Case History

In 1998, M.A.H. was 39 years old when he suffered from a stroke in the right cerebellar hemisphere. He is a right-handed, academically educated man, who worked at a high professional level. He came to our attention during a neuropsychological screening study in which patients were included within the first 14 days after their stroke and re-examined after 24 months (van Zandvoort et al., 2004). Apart from residual dizziness and mild memory deficits, he had no complaints about his cognitive functioning, reported no changes with respect to his premorbid level of functioning and returned completely to his premorbid professional level. On MRI-imaging (TSE) an infarction in the right cerebellar hemisphere, predominantly the lower half and the vermis, consistent with the supply area of the posterior inferior cerebellar artery (PICA) was found. No additional brain abnormalities in the occipitotemporal cortices appeared to be present on MRI (see Figure 1).