



Anosognosia for cerebral achromatopsia—A longitudinal case study

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ABSTRACT

Cerebral achromatopsia is a rare disorder of colour vision caused by bilateral damage to the occipito-temporal cortex. Patients with cerebral achromatopsia are commonly said to suffer due to their disturbed colour sense. Here, we report the case of a patient with cerebral achromatopsia who was initially unaware of his deficit, although three experiments with eye movement recordings demonstrated his severe inability to use colour information in everyday tasks. During two months, the evolution of his colour vision deficit was followed with repeated standardized colour vision tests and eye movement recordings. While his performance continuously improved, he became more and more aware of the deficit. Only after colour vision had almost normalized, his subjective colour sensation was inconspicuous again. The simultaneous occurrence of achromatopsia and the corresponding anosognosia and their parallel recovery suggest that both deficits were due to dysfunction of the same brain region. Consequently, the subjective experience of colour loss in achromatopsia may depend on the residual function of the damaged colour centre.

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1. Introduction

Cerebral achromatopsia denotes the complete or partial loss of colour vision after cortical damage (Zeki, 1990). Although the existence of a colour centre in the human brain has been debated for over a century (Zeki, 1990), it is now widely accepted that bilateral damage to the ventral occipito-temporal cortex causes this disorder (Brazis, Masdeu, & Biller, 2007). Achromatopsia is frequently associated with other deficits, most prominently visual field defects and prosopagnosia (Beck, Aschayeri, & Keller, 1978; Meadows, 1974; Zeki, 1990). Achromatopsia is a rare condition and most reported patients with achromatopsia either did not recover or were not followed up (for an overview see Bartels & Zeki, 2000, but see Bornstein & Kidron, 1959; Rondot, Tzavaras, & Garcin, 1967). Therefore, the natural history of the disorder is not well known.

It has been stated that patients with achromatopsia usually notice their deficit (Bauer & Demery, 2003; Hodges, 2007). Moreover, the confrontation with a visual world drained of colour has often been vividly described as a frightening and distressing experience (e.g. Pallis, 1955). Interestingly, however, there are a few published reports of patients who either did not notice their colour perception deficit (Green & Lessel, 1977; Grüsser & Landis, 1991) or did so only some time after brain damage, suggesting unawareness for a loss of colour vision (Sacks, 1996; Steffan, 1881). The unaware-

ness of a handicap after brain damage is known as “anosognosia” and has been described for the motor, sensory and visual system (Prigatano & Schacter, 1991). Anosognosia for achromatopsia and its evolution over time, however, has not been studied in depth. The study of anosognosia is important since it can inform concepts of higher brain functions, in particular models of consciousness (Bisiach & Geminiani, 1991). Along these lines, cases of anosognosia for achromatopsia may add to our understanding of visual consciousness.

We had the rare opportunity to repeatedly analyse consciousness of colour perception in a patient who suffered from cerebral achromatopsia due to bilateral ischemic damage to the occipito-temporal cortex. Using standardized colour vision tests and eye movement recordings we could show that four days after the stroke the deficit rendered him completely unable to make use of colours in order to discriminate and identify objects. Yet, despite the severity of achromatopsia, he was completely unaware of his handicap. Over a period of eight weeks, an almost complete recovery of achromatopsia could be demonstrated. During this recovery phase, objective improvement of his colour sense was accompanied by increased subjective awareness of his deficit. This observation points towards a possible role of the cerebral colour centre not only in processing but also in consciously perceiving colour.

2. Case report

A 78-year-old right-handed man presented with visual disturbances of sudden onset, which had rendered him unable to read the newspaper.

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On initial neurological examination, the patient was awake and fully oriented. He stated that his vision was currently fair and that he had noticed no specific problems apart from his reading difficulties. Binocular near vision was 0.5, and confrontation visual field testing was indicative of bilateral superior quadrantanopsia. He was unable to read sentences or words, but could read single letters of the same letter type (newspaper headlines), findings suggestive of pure alexia. Furthermore, he was not able to identify familiar faces. He did not recognise his attending physician or his wife and daughter unless they addressed him verbally.

When objects of various colours were presented, he maintained that all were greyish. However, semantic knowledge of the colours of various objects was preserved. Confronted with the colour vision deficit, he was astonished and attributed it to poor lighting. As no acknowledgement of the deficit could be obtained from the patient, his degree of anosognosia can be rated as severe corresponding to the highest possible score (3 on a 4-point scale ranging from 0 to 3) according to Bisiach, Vallar, Perani, Papagno, and Berti (1986). Magnet resonance imaging including T2-weighted images revealed bilateral posterior cerebral artery strokes with lesions involving the

fusiform gyri, lingual gyri, and parahippocampal gyri on both sides (Fig. 1).

Goldmann perimetry confirmed bilateral superior quadrantanopsia, which was more pronounced on the right side (Fig. 2).

2.1. Evolution

Four days after the stroke (T1), the patient still complained about reading difficulties and that he failed to see parts of objects, but did not report impaired colour perception. An assessment of colour vision using the Panel 16 Colour Vision Test (Precision Vision, Villa Park, USA) revealed a diffuse colour discrimination deficit (Total Error Score: 31.7; calculated according to Vingrys & King-Smith, 1988). On selective testing of colour perception in both inferior quadrants, no difference was reported. Confronted with these results, the patient admitted that he saw everything in shades of grey but had not been aware of it. Thus the unawareness score of Bisiach et al. (1986) was 2 out of 3 points. When asked whether he had not found it difficult to eat colourless food, he replied: “No, not at all! You just know what colour your food is. Spinach, for example, is just green.”

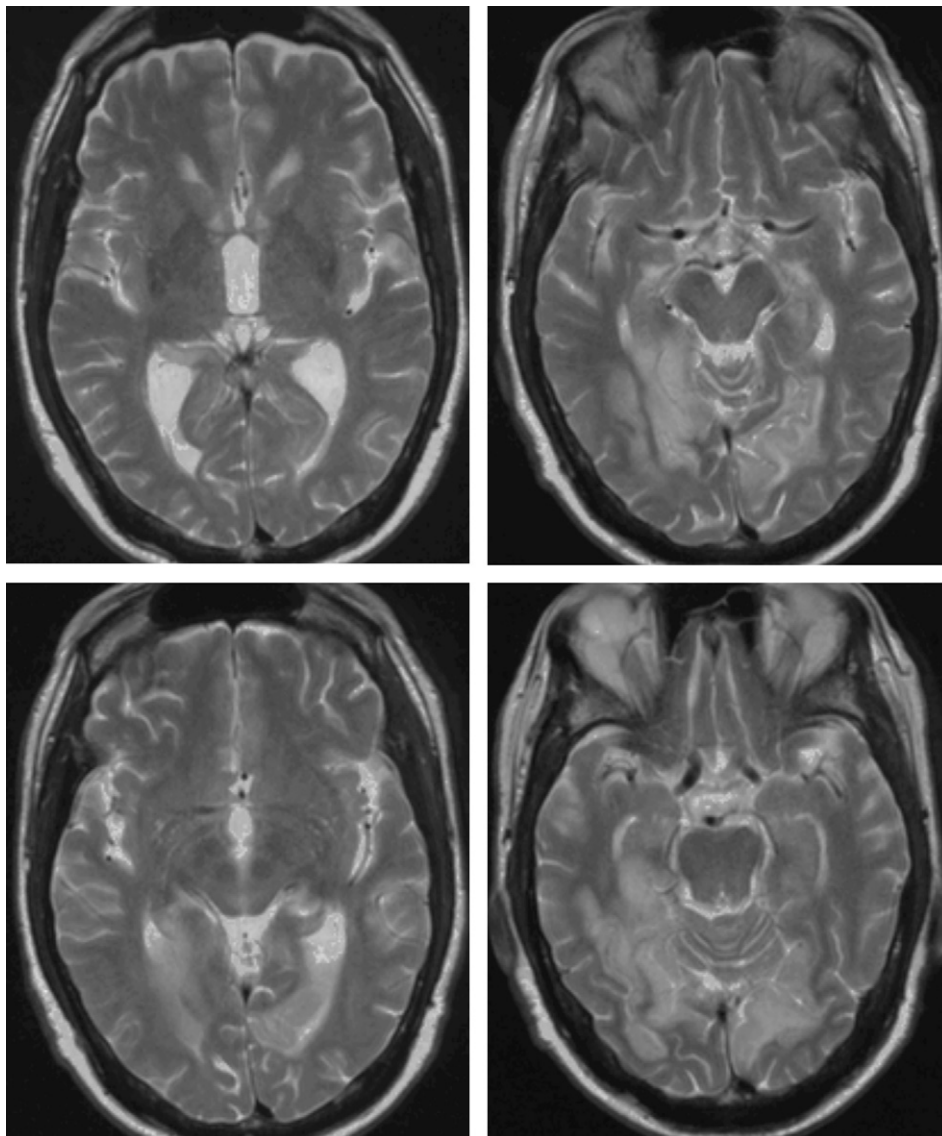


Fig. 1. Axial T2-weighted MRI scan slices of the patient two days after the stroke. Hyperintense lesions are noticeable in the distribution of both posterior cerebral arteries involving the fusiform gyri, lingual gyri, and parahippocampal gyri.

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