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## Research report

# Anosognosia for hemiplegia with preserved awareness of complete cortical blindness following intracranial hemorrhage

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## ABSTRACT

A 51-year-old woman presented with anosognosia for hemiplegia (AHP), neglect, and a complete loss of vision, for which she was almost immediately aware. Neuroimaging studies revealed intracranial hemorrhages in the medial temporal lobes bilaterally, extending back to the occipital cortex, but sparing the calcarine cortex. A large right frontal-parietal hemorrhage which extended to the posterior body of the corpus callosum was also observed. The patient's vision slowly improved, and by 11 months post onset, formal visual fields revealed improvement primarily in the left upper quadrants only. In contrast, resolution of her AHP occurred between the 26th and 31st day post onset. Awareness of motor impairment was correlated with her ability to initiate finger tapping in her left hemiplegic/paretic hand. During the time she was unaware of her motor deficits but aware of her visual impairments, her dreams did not reflect concerns over visual or motor limitations. The findings support a "modular" theory of anosognosia.

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## 1. Introduction

Early reports of anosognosia for left hemiplegia were made by von Monakow, Anton, Pick, and Babinski (Papagno and Vallar, 2003). Subsequent group studies on anosognosia for hemiplegia (AHP) noted that this clinical condition is frequently associated with large cerebrovascular accidents (CVAs) typically involving the distribution of the right middle cerebral artery (MCA) (Pedersen et al., 1996; Orfei et al., 2007). AHP can exist in patients who are aware of other neurological and/or neuropsychological disturbances. Bisiach et al. (1986) demonstrated that AHP could exist in patients with partial visual field loss (e.g., a homonymous hemianopia) who were

aware of their visual impairments (and vice versa). This finding led to two theoretical propositions. First, "unawareness of a failure of a particular function betrays a disorder of the highest levels of organization of that function" as initially proposed by Anton (see Bisiach et al., 1986, p. 480). Second, that "awareness" of a given function may be "decentralized and appportioned to the different functional blocks to which it refers" (Bisiach et al., 1986, p. 480). This suggests a "modular" model of anosognosia (Bisiach and Geminiani, 1991). Berti et al. (2005) presented neuroimaging findings supporting this proposition. While the debate continues as to the crucial neural networks responsible for AHP (Prigatano, 2009), lesions involving the right frontal-parietal cortex and

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insular cortex are commonly associated with AHP (Berti et al., 2005; Karnath et al., 2005; Vocat and Vuilleumier, 2010).

Anton's syndrome is another classic form of anosognosia (Prigatano, 2009). In this case, the patient is completely cortically blind but unaware of their blindness. The natural course of Anton's syndrome and its neuroimaging correlates have not been well studied (Prigatano and Wolf, 2010). Yet, Anton's syndrome is typically associated with bilateral posterior cerebral artery (PCA) occlusions/hemorrhages (e.g., Argenta and Morgan, 1998).

We examined and followed a patient who presented with left AHP and complete cortical blindness for 1 year post stroke. Unlike Anton's syndrome patients, this patient was aware of her blindness. We were interested in determining the neuroimaging correlates of this latter clinical condition and how her vision might change with time.

In addition, we monitored the resolution of AHP and its behavioral correlates. If Berti et al.'s (2005) assertion is correct, the patient should become aware of her motor limitations once she is able to carry out an intended motor (finger) movement. If AHP resolves because other associated neurological and neuropsychological disturbances improve (i.e., neglect diminishes and memory gets better), then spontaneous recovery from AHP would not be specifically associated with the ability to initiate movement.

Finally, we explored whether the patient's dreams during the time she presented with AHP, neglect, and awareness of her complete cortical blindness revealed an implicit awareness of her motor deficits, despite her explicit verbal denial of her hemiplegia. It has been suggested that implicit awareness may exist, even when explicit denial of a motor deficit is present (Ramachandran, 1994; Vocat and Vuilleumier, 2010). Dream material might reveal implicit awareness of an impaired function.

## 2. Case report

SL was a 51-year-old, right-handed female who was discovered to have a large right parietal arteriovenous malformation (AVM) in 2004. The patient underwent embolization and partial resection of the AVM at that time, with no neurological sequelae. The patient was reevaluated in 2007, and a residual right parietal AVM was identified and again resected. Following that surgery, the patient was described as having short-term memory difficulties which resolved within approximately 3 months. No motor weakness or visual disturbance was noted.

In March of 2009, the patient was found unconscious and unresponsive. An intracranial hemorrhage was documented. While she improved for a period of time, she then deteriorated and developed a left hemiplegia and "cortical blindness". Her sister, who was with her right after these events, indicated that the patient was not initially aware of her blindness. Within a few hours, however, she became aware of her blindness, via her sister's report. Her sister also noted at the time the patient could not move her left side (arm and leg), but was unaware of her inability to do so. Her clinicians also noted left hemispatial neglect.

Subsequent to these events, the patient underwent a third craniotomy in which the AVM was embolized and resected.

The hematoma was evacuated. Post-surgical imaging revealed partial evacuation of the right parietal hematoma, bifrontal pneumocephalus, evolving infarcts in the right MCA and the PCA distribution, and deep white matter hyperintensities in the parietal and temporal areas bilaterally.

Visual evoked potentials (VEP) were performed using a pattern reversal checkerboard stimulus. Light flashes were of 150  $\mu$ sec duration at 1.8/sec. The latency of the P100 wave was not reproducible from either eye stimulation during the study. No cortical responses were noted following pattern VEP flash.

Approximately 2 weeks after her last surgery, and 18 days post onset of cortical blindness and AHP, the patient was transferred to an inpatient neurorehabilitation unit. At that time she continued to report complete visual loss, but no restriction in her left arm or leg, despite her dense left hemiplegia. She also had her head turned to the right compatible with unilateral left neglect.

## 3. Assessment of anosognosia

During serial behavioral assessments, the patient was asked a variety of questions concerning her vision and motor functioning. Her responses were recorded, and then later scored according to the Bisiach's Scale of Anosognosia (Bisiach et al., 1986) as it relates to motor (upper/lower limb) impairments. The scoring system was as such:

Score: 0	The disorder is spontaneously reported or mentioned by the patient following a general question about his complaints.
1	The disorder is reported only following a specific question about the strength of the patient's left limbs.
2	The disorder is acknowledged only after its demonstration through routine techniques of neurological examination.
3	No acknowledgement of the disorder can be obtained.

Anosognosia related to her loss of vision was also scored in an analogous manner. As noted above, she would spontaneously mention that she could not see, and therefore her score was 0 by the time she was evaluated.

## 4. Assessment of neglect

Given that the patient was completely blind, many traditional tests of neglect could not be administered (Mesulam, 2000). In order to assess the degree of neglect in this woman, a motor exploration task was utilized. The patient was asked to take her right hand (for which she had complete apparent functional use) and touch her left hand (a measure of personal neglect, see Bisiach et al., 1986; Spinazzola et al., 2008). Repeated trials were performed each day. The patient's responses were recorded and later drawn by a medical

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