

## VIEWPOINT

# ANOSOGNOSIA: THE NEUROLOGY OF BELIEFS AND UNCERTAINTIES

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### ABSTRACT

Anosognosia is a common, fascinating, and ill-understood disorder following brain damage, where patients who suffer severe deficits such as hemiplegia may remain unaware of and deny their handicap. Many studies including recent work published in this journal have attempted to determine the neurological, cognitive, and motivational bases of anosognosia. These studies have typically focused on descriptive correlations between anosognosia and various clinical factors, but did not identify a consistent pattern of brain lesion or dysfunction. Rather, the results have emphasized the complex and multifaceted nature of anosognosia. This review discusses the implications of existing results, and proposes a general “ABC” framework for anosognosia where various problems in Appreciation, Belief, and Check operations may contribute to abnormal cognitive and affective appraisal of a deficit. New experimental approaches and new therapeutic tools are needed to better understand the neurocognitive mechanisms responsible for our awareness of normal functioning and failures.

Key words: Anosognosia, denial, awareness, neglect, motivation, belief

Anosognosia is a perplexing condition. It can affect patients with various neurological impairments who appear unable to notice and acknowledge the existence of their deficits, often despite blatant evidence for their handicap. Typical examples are hemiplegic patients who may assert that their paralyzed limbs are still functioning normally. Other patients with bilateral cortical blindness may claim that their vision is intact, or amnesics may contend that their memory is excellent (for detailed review see Prigatano and Schacter, 1991; Vuilleumier, 2000). In fact, patients may be anosognosic for virtually any neurological deficit following brain injury, including aphasia (Lebrun, 1987), prosopagnosia (Young et al., 1990), or apraxia (Berti et al., 1996).

Anosognosia for hemiplegia (AHP) is thought to be relatively common, encountered in at least 20-30% of hemiplegics after an acute stroke (Cutting, 1978; Stone et al., 1993). However, we still have a very poor understanding of anosognosia. This is perhaps not too surprising given the heterogeneity of this phenomenon. But more surprisingly, only few systematic studies have been carried out in these patients with the aim of better characterizing the crucial clinical features and their possible underlying cognitive mechanisms. Most existing studies have been descriptive (e.g., examining the correlation with different lesion types, severity of neurological impairment, etc.), whereas experimental investigations have been rare or performed only in a few single cases (for review see Vuilleumier, 2000).

In this issue of *Cortex*, a paper by Marcel and colleagues provides us with a complex set of new data that revisit many important questions about anosognosia for hemiplegia. Marcel et al. (2004,

this issue) rightly considered that we have no secure grip on the characteristics of what has to be accounted for in anosognosia, neither on how to account for it, and therefore wished to remedy this poor knowledge by a systematic investigation in a group of 64 hemiplegic stroke patients. A valuable aspect of this investigation was to employ a number of original experiments that were designed to test specific hypotheses (e.g., general performance monitoring abilities, mental flexibility, first vs third person perspective). Marcel et al. (2004, this issue) point to many interesting tracks that need further explorations. However, this new study also clearly shows that much work remains to be done. The reported results underscore the complex cognitive picture of anosognosic disorders, and raise many new questions in addition to offering tentative answers to some interrogations that have recurrently been debated over the last century. I will review a few of these questions here, and discuss whether existing data can now allow us to incriminate any particular neurological, cognitive, or motivational factors in anosognosia.

### *What is the Role of Concomitant Neurological Deficits?*

A question posed ever since Babinski (1914, 1918, 1924) first described anosognosia for hemiplegia concerns the role of sensory deafferentation, especially proprioceptive loss. An early and reasonable belief based on clinical impression was that a lack of sensory inputs might deprive the patients of direct feedback about their affected limbs' state and impair the conscious representation of their half-body, resulting in “asomatognosia”. However, many subsequent

studies (Bisiach et al., 1986; Starkstein et al., 1992) as well as the new report by Marcel et al. (2004, this issue) have shown that anosognosia does not correlate with severity of primary sensory loss. A fascinating case study by Garcin et al. (1938) also showed long ago that a patient's inability to recognize his left hand as his own could resolve without any improvement in proprioceptive sensation, in sharp contrast with a deficit in intentional motor use that appeared much more determinant for the lack of self-hand recognition. Yet, informal clinical impression would still compel many neurologists to believe that prototypical anosognosic patients present with an impaired sensory experience of their affected limbs. This is why Marcel et al. (2004, this issue) and other investigators (Starkstein et al., 1992; Stone et al., 1993; Small and Ellis, 1996) have repeatedly set out to re-examine this issue. However, a question may still remain unsatisfactorily addressed in these studies, namely, what is the type of "sensory loss" that most matters in anosognosia. Obviously, anosognosia is usually not associated with complete sensory deafferentation due to peripheral lesions, although it can occur after brachial plexus damage (Laplante and Degos, 1984) or pedunclopontine stroke (Bakchine et al., 1998) when there is a concomitant confusion or dementia. Perhaps, a more specific sensory deficit or a combination of deficits might play a critical role. For example, allesthesia and tactile extinction appear more often correlated with AHP than true perceptual loss (Vuilleumier, 2000), suggesting a problem in integrating sensory inputs with spatial or bodily representations at a higher-level of processing. Global measures collapsing all sensory functions into a single number on a three-point scale, as employed by Marcel et al. (2004, this issue) and other group studies (Bisiach et al., 1986; Starkstein et al., 1992; Stone et al., 1993; Small and Ellis, 1996) are certainly too crude to provide an appropriate characterization of deficits associated with AHP.

Likewise, a majority of studies including the work by Marcel et al. (2004, this issue) have demonstrated a lack of reliable correlation between the severity of motor loss and the presence of AHP (Cutting, 1978; Starkstein et al., 1992; Small and Ellis, 1996). Here again, however, the most relevant type of motor dysfunction might concern higher-level processes, related to the subjective correlates of action planning and motor intention (Heilman, 1991; Daprati et al., 2000; Frith et al., 2000), rather than just the degree of weakness. In line with earlier observations by Garcin et al. (1938), Heilman and colleagues (Heilman, 1991; Gold et al., 1994; Adair et al., 1997) have suggested that abnormal motor intention might disrupt some "feedforward" signals conveying a subjective sensation of volitional effort, and thus prevent the patient from detecting a mismatch between the required and performed motor action.

By analogy, a deficit in initiating and monitoring active memory searches might also contribute to anosognosia for amnesia in some patients (Vuilleumier, 2000). However, EMG measures in patients with AHP suggest that covert planning of actions may still take place (e.g., during mental imagery of bimanual actions) even though patients are unable to execute such actions voluntarily (Hildebrandt and Zieger, 1995). Altogether, these findings suggest that any abnormalities in motor control associated with AHP are likely to implicate a complex representation of intended actions as made available in conscious awareness, rather than just a weaker motor command that can easily be scored on the MRC scale. Indirect measures of motor intention as used by Heilman and colleagues (Heilman, 1991; Gold et al., 1994; Adair et al., 1997) or measures of motor planning in bimanual tasks as proposed by Marcel et al. (2004) and others (Hildebrandt and Zieger, 1995; Ramachandran, 1995) might be more revealing, although tricky to design and interpret. Marcel et al. (2004, this issue) reported that patients with right brain damage and AHP tended to overestimate their bimanual abilities during a structured questionnaire, but since data for unimanual abilities were not reported, it is unclear whether there is anything special to this "bimanual" condition as compared with the basic propensity of patients with AHP to overestimate usage of their contralesional hand alone. It would be interesting to know whether anosognosia for left hand function is exacerbated by imagining or planning concurrent right hand actions.

In this respect, contralesional spatial neglect is a notable suspect in anosognosia. Neglect is a condition where perceptual experience can be suppressed despite well-preserved sensory inputs, and willed action compromised despite well-preserved motor strength. Therefore, contralesional sensory and motor extinction associated with neglect might be more important than elementary sensory or motor loss in excluding information about current states of the limb from the patient's awareness. The results from Marcel et al. confirm several previous studies indicating a consistent correlation between neglect and AHP (for review: Feinberg, 1997; Vuilleumier, 2000; see also Cocchini et al., 2002). However, dissociations have been reported. A few patients with AHP have been observed who showed no signs of personal or extrapersonal neglect (Bisiach et al., 1986; House and Hodges, 1988; Small and Ellis, 1996; Dauriac-Le Masson et al., 2002), and AHP during the Wada test is not consistently accompanied by neglect (Adair et al., 1995). It has been argued that neglect and AHP are two dissociable conditions (Bisiach et al., 1986; Feinberg, 1997; Dauriac-Le Masson et al., 2002). But here too, the measures and criteria used to diagnose neglect vary among the studies, and are probably often insufficient given the large

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