Anosognosia

Lorenzo Pia and Anna Berti (2015), Scholarpedia, 10(4):6586. doi:10.4249/scholarpedia.6586 revision #149167 [link to/cite this article]

- **Prof. Lorenzo Pia**, SAMBA (SpAtial, Motor & Bodily Awareness) Research Group, Psychology Department & Neuroscience Institute of Turin (NIT), University of Turin, Turin, Piedmont, Italy
- **Dr. Anna Berti**, Psychology Department & Neuroscience Institute of Turin (NIT)

Contents

Introduction

Doctor: *"Why you are in the Hospital?"*

Patient: *"I have just felt unwell"*

D: *"Can you move your left arm?"*

P: *"Yes!"*

D: *"Would you please reach my hand with your left arm.....have you done it?"*

P: *"Yes, I have"*

This sounds like an ordinary clinical conversation between a neurological patient and a doctor. However, the astonishing characteristic of such dialogue is that the patient, affected by a complete paralysis of the arm and leg of the left side of the body due a right-hemisphere stroke, firmly denied the motor disability. This is an example of a remarkable well-known phenomenon termed anosognosia, that is the unawareness of the existence of a neurologically based disability (from the Greek words "nosos" disease and "gnosis" knowledge; an- / a- is a negative prefix).

In historical terms, the first report of unawareness of a neurological deficit seems to be the one described by Seneca in Liber V, Epistula IX (Bisiach & Geminiani, 1991). In a letter to his friend Lucilius, he described a woman who obstinately denied her blindness.*"....You know that Harpestes, my wife's fatuous companion, has remained in my home as an inherited burden....This foolish woman has suddenly lost her sight. Incredible as it might appear, what I am going to tell you is true: She does not know she is blind. Therefore, again and again she asks her guardian to take her elsewhere because she claims that my home is dark.....It is difficult to recover from a disease if you do not know to be ill...."*. Interestingly, this description already underlined some of the most important features of anosognosia: the denial, the confabulation generated to justify the problem ("home is dark"), and the negative impact on the recovery ("It is difficult to recover from a disease if you do not know to be ill").

Two thousand years later, Babinski (1914) introduced the term anosognosia in order to describe the denial of the paralysis. *"I want to draw attention to a mental disorder that I had the opportunity to observe in cerebral hemiplegia, which consists in the fact that patients seem unaware of or ignore the existence of their paralysis"*.

It is worth noting that anosognosia was firstly described as a symptom of a more general disorder of body schema build up by anomalous somato-sensory experiences. Hence, the impairment was interpreted as a direct consequence of the brain damage. However, in the second half of the last century, it was proposed the possible role of motivational reactions in the development of unawareness. So-called motivational theories (Weinstein & Kahn, 1955) assumed that premorbid components of personality were causally related to the illness development. Therefore, anosognosia was considered a form of organic suppression or an avoidance reaction against the stress caused by the illness. Such theories interpreted the disorder as the activation of a psychodynamic reaction rather than directly linked to the brain lesion, and therefore, as an effect of a damage to a given cognitive system. Accordingly, during those years there was a decline of pure neurological approaches to the phenomenon in favor of more psychiatric perspectives.

At the end of the century, however, several clinical observations questioned such a pure psychodynamic interpretation. First, anosognosia can be very selective. In other words, patients with multiple neurological and neuropsychological impairments may deny one deficit, but spontaneously report another. However, it is difficult to explain the existence of a psychological reaction against a single disorder when others, often very disabling, are present. Secondly, anosognosia tends to ameliorate with time, and it is observed much more frequently in the first period after the stroke than during the chronic phase. A defense mechanism would predict the opposite temporal pattern, insofar as time is needed for establishing a psychodynamic reaction. The third observation comes from the amytal testing, a diagnostic tool that, through a pharmacologic intervention (i.e., injection in the brain circulation of a barbituric) produces a temporary suppression of the activity of one hemisphere, including motor functions. In this case, anosognosia for hemiplegia appears immediately after the amytal injection and mainly when it is the right hemisphere (left hemiplegia) that is knocked down. A psychodynamic theory would predict anosognosia for both left and right hemiplegia. Finally, a physiological manipulation, the injection of cold water into the left ear of the anosognosic patient provokes an instantaneous, although temporary, remission of the denial. This effect, triggered by a vestibular reflex, lasts a few minutes during which a left side nystagmus (left side reflexed ocular movement) is observed. In such cases, the patients astonishingly admit their paralysis but again 'forget' it when the effect of the stimulation is over while a psychodynamic reaction should not be influenced by a physiological manipulation. These and other data suggest that anosognosia might be better explained as a specific cognitive deficit directly caused by a brain damage, rather than a normal functional reaction triggered by the emotional strain (for a review see Prigatano, 2010).

In the present article, we will first mention some famous cases of anosognosia. Then, we will discuss anosognosia for hemiplegia as paradigmatic to indicate the selective and specific aspect of denial of illness. Finally, we will be briefly describe other forms of anosognosia.

Famous Cases and Media

Although no diagnosis has been accounted for the death of the novelist Charles Dickens, his varied symptoms has recently suggested a right parietal-temporal disorder including anosognosia for a whole range of bodily symptoms (McManus, 2001). On September 3, 1867, he wrote to a friend: *"I never was better in my life – doubt if any body ever was or can be better – and have not had anything the matter with me but that squeezed foot, which was an affair of a few days [sic]"*.

The Italian director, Federico Fellini, suffered a right-hemispheric stroke that left him hemiplegic on the left side. Despite he was aware of the deficit, he displayed negative feelings towards his paralysed limb, which he, in one occasion, called *"a bloated, damp bunch of asparagus"* (Loetscher, Regard et al., 2006).

In a recent New York Times article, [1] (http://opinionator.blogs.nytimes.com/2010/06/20/the-anosognosics-dilemma-1/) It has been speculated that Thomas Woodrow Wilson, the 28th U.S. president suffered from anosognosia for hemiplegia.

During the 1997 trials, Prof Amador supplied the court with mounting evidence that Theodor Kaczynski, otherwise known as the unabomber, suffered from anosognosia for his schizophrenic sympthomatology (Amador and Paul-Odouard, 2000). *"Mr. Kaczynski suffers from anosognosia…It reflects brain dysfunction rather than a calculated plan to manipulate authority for personal gain. If the patient with anosognosia does not perceive the evaluation as a challenge to his self-concept (his understanding of his innate capacities and abilities), he will usually submit without argument. Undoubtedly, that is likely the reason that Mr. Kaczynski submitted to a neuropsychological evaluation. It did not threaten his self-concept"*.

At present (2012), we have found three songs named anosognosia. The first [2] (http://itunes.apple.com/it/album/anosognosia/id255469691?i=255470033) is in the seventh album ('The useless lesson') by an electronic musician and composer, Kerry Leimer. The second [3] (http://itunes.apple.com/it/album/anosognosia/id319668059?i=31966) is in ('Alligators') by Allan Dawson, a London singer, songwriter and guitarist. The third [4] (http://itunes.apple.com/it/album/anosognosia/id365900118?i=365900302) is the second track of 'Twenty twelve', the debut album of Broadcast the Nightmare, a metalcore band placed out of Atlanta. Interestingly, we have contacted the band in order to know something more and this is the answer *"Yes you are right. It is a certain type of brain disorder. The lyrical content is metaphorical reference to people who choose to be ignorant in their ways, whether it be religious, spiritual or political aspects of life"*.

Anosognosia for Hemiplegia

Probably the most striking instance of anosognosia can be found in brain-damaged patients, affected by a complete contralesional hemiplegia (for a review Bottini et al., 2010). These patients may deny that there is anything wrong with their contralesional limbs and claim of being able to perform any kind of action. In addition, when asked to perform a purposeful movement with the paralysed limb they may appear convinced of having accomplished the requested action despite unambiguous evidence to the contrary coming from different sensory channels (see [5] (https://sites.google.com/a/lorenzopia.com/Lorenzo-pia/Anosognosia%20for%20hemiplegia.avi?attredirects=0) for a video). It is worth noticing that such "on line" confabulations, recently termed "illusory limb movements" (Feinberg, 2007), are not present in all anosognosic patients.

The symptomatology of anosognosia for hemiplegia can vary between and within patients. This disturbance is frequent after damage to the right hemisphere, with a prevalence ranging from 20% to 50% of hemiplegic patients, depending on the studies, the differences being related to the time of evaluation (acute vs. chronic phase of the illness) and selection criteria (e.g., Pia et al. 2004). When explicitly questioned about the condition of their limbs, patients may show different degrees of denial ranging from emotional indifference (anosodiaphoria), in which the motor problems may be admitted but without any concerns, to resolute and intractable unawareness of the disease. Additionally, productive symptoms, as verbal confabulations about the possibility of moving the plegic limb, and delusional beliefs may coexist. In this latter case, patients may experience their limb as not belonging to them (asomatognosia) or attribute their own body parts to other persons (somatoparaphrenia). The content of the confabulation can be very bizarre and patients may even claim that somebody else is lying on their beds or may show violent attitude against those 'alien' limbs (misoplegia).

The interpretation of anosognosia for hemiplegia is not straightforward. Theories that explain it either as a secondary consequence of sensory feedback deficits or as due to the co-occurrence of different kinds of neuropsychological disorders are not thought to be exhaustive explanations. Indeed, double dissociations have

been shown between anosognosia for hemiplegia and sensory/cognitive deficits (see Prigatano, 2010). It is worth noticing, however, that the motivational factor account has been recently re-proposed in terms of abnormal affective regulation or altered emotional and attitudinal processes implicated in self-attribution of perceptual experiences (e.g., Turnbull et al., 2005). A lesion to a right-lateralized emotion-regulation system might crucially contribute to the emergence of anosognosia for hemiplegia. Consistently, some anatomical data show an overlap between areas underpinning motor awareness (see below) and those subserving emotions. Additionally, right brain-damaged patients often develop emotional changes and difficulty tolerating aversive emotional states (Turnbull et al., 2005). Thus, although the emotional theories do not explain the full-fledged syndrome of anosognosia for hemiplegia, it is possible that the complex interactions between individual predispositions and brain dysfunction would shape the way in which denial manifest itself in different patients.

Recently, it has been proposed that anosognosia for hemiplegia might be explained as a domain specific disorder of motor control (Berti and Pia, 2006). Anatomo-clinical correlations and lesion analyses have shown that anosognosia for hemiplegia follows brain damages within lateral premotor and insular cortex (Berti et al., 2005; Karnath et al., 2005; Vocat et al., 2010). A possibility is that this damage would alter the monitoring of voluntary actions, thus preventing patients to distinguish between movement and no-movement states. Moreover, the (non-veridical) feeling of movement would arise from an intact motor intentionality subserved by a normal activity of the brain structures that implement the intention-programming system (Berti and Pia 2006). Evidence of preserved movement intentionality in anosognosic patients comes from the fact that they may show normal proximal muscle electrical activity in the affected side when they believe they are moving the plegic limb (Berti et al., 2007). Moreover, it has been shown that in bimanual action (Garbarini & Pia, 2013) the motor execution of the anosognosic patients' intact hand is affected by the intention to move the paretic hand. Interestingly, such an intentional stance dominates their subjective experience of willed actions because patients falsely detect movement of their plegic arm when they intend to move it, respect to when they do not (Fotopoulou et al., 2008).

Other forms of Anosognosia

Patients affected by hemianaesthesia (i.e., the loss of tactile/proprioceptive sensibility on one side of the body), can be completely unaware of their sensory impairment (e.g., Marcel et al., 2004; Spinazzola et al., 2008). Interestingly, unawareness for hemianaesthesia can be dissociated from anosognosia for hemiplegia, unilateral neglect, intellectual impairment and general self-monitoring on both functional and anatomical ground. This form of denial seems to be more frequently associated to lesions of those brain structures underpinning sensory-spatial processing (e.g., insular, temporal and subcortical lesions mainly affecting basal ganglia). Hence, as for the cerebral circuits of self-monitoring processes for primary sensory functions are located in areas involved both in the execution of the primary functions and in the emergence of awareness related to the monitoring of the same functions. Present theories suggest that patients affected by anosognosia for hemianaestesia have a nonveridical sensory awareness generated as an illusory experience by the failure to distinguish between an imagined sensation and a real, physical one (Pia et al., 2014). The brain structures underpinning sensory-spatial processing would be the neural basis of sensory self-monitoring allowing the distinction between 'veridical' and 'non-veridical' sensory awareness.

Anosognosia may occur in cerebral achoromatopsia, a quite rare neurological disorder characterized by a complete or partial loss of colour vision after a bilateral cortical damage to the ventral occipito-temporal cortex. Patients affected by this disorder claim to see the world in black and white and are usually aware of their visual deficit. However, there are a few descriptions of patients who either did not notice their colour perception deficit or did so only some time after the brain damage, suggesting unawareness for the loss of colour vision. Despite verbal and nonverbal/perceptual testing showing the presence of severe color blindness, the patients may still claim a normal color vision, even when faced with their errors in naming colors. It is worth noticing that color vision may improve with time and a parallel improvement in awareness can be observed. The simultaneous

occurrence of achoromatopsia and anosognosia, their parallel recovery, and their lesion site in visual areas suggest that both deficits were due to dysfunction of the same brain region, implying again that normal perception and normal monitoring share common anatomical substrates.

As mentioned above, anosognosia may occur also within more complex cognitive disorders as, for instance, schizophrenia and Alzheimer's disease. In these pathologies, it is very hard to individuate a common core explaining the different denial behaviors. The fact that in many instances there is a severe intellectual impairment is of course a confounding factor. However, several data seems to support the idea that also in these cases unawareness can be selective and neurologically based.

In schizophrenia, it has been often used the term 'lack of insight' to indicate a broad construct encompassing unawareness of the disorder, of the effects of treatment, of social consequences, of the occurrence of psychiatric symptoms and difficulty in labeling them. Recently, however, numerous data suggest that unawareness in schizophrenia might have similarities with the concept of anosognosia: persistence despite all contrary evidence, confabulations to explain the symptoms, frontal lobe impairments and domain specificity (e.g., Pia & Tamietto 2006). For instance, there can be dissociations between unawareness for negative and positive symptoms. Moreover, unawareness for pathological involuntary movement (i.e., tardive dyskenisia) can be present for some muscular group and not for others akin to what has been reported for patients with anosognosia for hemiplegia who may be aware of the motor deficit affecting the lower limb and not of the contemporary presence of the motor impairment of the upper limb and viceversa.

Similarly, anosognosia in Alzheimer's disease has not been initially considered as a domain specific impairment Indeed, data from early/acute stages of the disease and from different aspects of functioning including cognition, mood, behavior and daily activities have been often grouped in a single anosognosia index. However, it has been recently suggested that also unawareness in Alzheimer's disease might be interpreted as a selective and neurologically based condition. It seems to be strictly connected to frontal lobe impairments (e.g., patients with anosognosia and Alzheimer's disease often show a reduction of cerebral haematic flow in the frontal regions, deficits of executive functions and extrapyramidal signs) and can be observed for some deficits and not for others (see Kaszniak & Edmonds, 2010; Starkstein & Power 2010, Pia & Conway, 2008 for reviews).

To conclude *"Understanding the biological and neuropsychological mechanisms responsible for anosognosia in its various forms may reveal important insights into brain organization and how human consciousness (subjective awareness of the self and the environment) is possible. The comparative study of anosognosia in patients with identifiable brain disorders in comparison to patients with psychiatric disorders may provide rich insight into the body-mind problem"*. George Prigatano The study of anosognosia. Oxford: Oxford University Press, 2010.

References

Amador XF & Paul-Odoard R. Defending the unabomber: Anosognosia in schizophrenia. The Psychiatric Quarterly, 71: 363-371, 2000.

Babinski J. Contribution à l'étude des troubles mentaux dans l'hémiplégie organique cérébrale (anosognosie). Revue Neurologique, 27: 845-848, 1914.

Berti A, Bottini G, Gandola M, Pia L, Smania N, Stracciari N, Castiglioni I, Vallar G & Paulesu E. Shared cortical anatomy for motor awareness and motor control. Science, 309: 488-491, 2005.

Berti A and Pia L. Understanding motor awareness through normal and pathological behavior. Current Directions in Psychological Science, 15: 245-250, 2006.

Berti A, Spinazzola L, Pia L & Rabuffetti M. Motor awareness and motor intention in anosognosia for hemiplegia. In Haggard P., Rossetti Y., and Kawato M. (Ed.), Sensorimotor foundations of higher cognition: Oxford University Press, 15(5): 245-250, 2007.

Bisiach E & Geminiani G. Anosognosia Related to Hemiplegia and Hemianopia. In Prigatano GP & Schacter DL (Ed.), Awareness of Deficits After Brain Injury. Oxford University Press, 17-39, 1991.

Bottini G, Paulesu E, Gandola M, Pia L, Invernizzi G, and Berti A. Anosognosia for hemiplegia and models of motor control: insights from lesional data. In Prigatano GP (Ed.) The Study of Anosognosia. Oxford: Oxford University Press, 363-379: 2010.

Feinberg TE. The "Hard problem" Of anosognosia: Delusional confabulation and anosognosia. Cortex, 43: 1099-101, 2007.

Fotopoulou A, Tsakiris M, Haggard P, Vagopoulou A, Rudd A & Kopelman M. The role of motor intention in motor awareness: An experimental study on anosognosia for hemiplegia. Brain, 131: 3432-3442, 2008.

Green GJ & Lessel S. Acquired cerebral dyschromatopsia. Archives of Ophthalmology, 95: 121–128, 1977.

Garbarini F and Pia L. Bimanual coupling paradigm as an effective tool to investigate productive behaviors in motor and body awareness impairments. Frontiers in Human Neuroscience 7(737): 1-5, 2013.

Grüsser OJ & Landis T. Visual agnosias and other disturbances of visual perception and cognition. In J. Cronly-Dillon (Ed.), Vision and visual dysfunction. Basingstoke, UK: Macmillan, 1991.

Karnath HO, Baier B & Nagele T. Awareness of the functioning of one's own limbs mediated by the insular cortex?. Journal of Neuroscience, 25: 7134-7138, 2005.

Kaszniak AW & Edmonds EC. Anosognosia in Alzheimer's Disease: Behavioral Studies. In Prigatano GP (Ed.), The Study of Anosognosia (pp. 189-227). Oxford: Oxford University Press, 2010.

Loetscher T, Regard M & Brugger P. Misoplegia: A review of the literature and a case without hemiplegia. Journal of Neurology, Neurosurgery and Psychiatry, 77: 1099-1100, 2006.

Marcel AJ, Tegnèr R & Nimmo-Smith. Anosognosia for plegia: Specificity, extension, partiality and disunity of bodily unawareness. Cortex, 40: 19-40, 2004.

Mc Manus ID. Charles Dickens: A neglected diagnosis. Lancet, 358: 2158-2161, 2001.

Pia L & Conway PM. Anosognosia and Alzheimer's Disease. Brain Impairment, 9(1): 81-88, 2008.

Pia L & Tamietto M. Unawareness in schizophrenia: Neuropsychological and neuroanatomical findings. Psychiatry and Clinical Neurosciences, 60(5): 531-537, 2006.

Pia L, Spinazzola L, Garbarini F, Bellan G, Piedimonte A, Fossataro C, Livelli A, Burin D, Berti A. Anosognosia for hemianaesthesia: a voxel-based lesion-symptom mapping study. Cortex 61: 158-166, 2014.

Prigatano GP. The study of anosognosia. Oxford: Oxford University Press, 2010.

Spinazzola L, Pia L, Folegatti A, Marchetti C & Berti A. Modular structure of awareness for sensorimotor disorders: evidence from anosognosia for hemiplegia and anosognosia for hemianaesthesia. Neuropsychologia, 46(3): 915-926, 2008.

Starkstein S & Power BD. Anosognosia in Alzheimer's Disease: Neuroimaging Correlates. In Prigatano GP (Ed.), The Study of Anosognosia (pp. 171-187). Oxford: Oxford University Press, 2010.

Turbull OH, Evans CE & Owen V. Negative emotions and anosognosia. Cortex, 41: 67-75, 2005.

Vocat R, Staub F, Stroppini T & Vuilleumier P. Anosognosia for hemiplegia: a Clinical anatomical prospective study. Brain, 133(12): 3578-3597, 2010.

Weinstein EA & Kahn RL. Denial of Illness-Symbolic and physiological aspects. Springfield: Charles C. Thomas, 1955.

Author's web pages

- Lorenzo Pia: http://my.unito.it/unitoWAR/page/dipartimenti8/D008_en/D008_EN_research2?path=/BEA%20Repository/5630049
- Anna Berti: http://my.unito.it/unitoWAR/page/dipartimenti8/D008_en/D008_EN_research2?path=/BEA%20Repository/5630049

Other web pages

- Patrick Vuilleumier (http://labnic.unige.ch/nic/htms/vuilleumier.html)
- Hans-Otto Karnath (http://homepages.uni-tuebingen.de/karnath/Sektion.html)
- George Prigatano (http://www.thebarrow.org/Research/Hypothalamic_Hamartoma/203962)
- Katerina Fotopoulou (http://www.iop.kcl.ac.uk/staff/profile/default.aspx?go=11433)
- Salvatore Aglioti (http://w3.uniroma1.it/aglioti/SCNL/People/Post%20Doc.html)
- Paul Jenkinson (http://psydb.herts.ac.uk/staff_list/FMPro?-db=staff_list_email&-format=recorddetail.html&-lay=details&-sortfield=surname&-max=2147483647&-recid=33712&-findall=)
- Maria Donata Orfei (http://www.researchgate.net/researcher/39512110_Maria_Donata_Orfei)
- Vilayanur S. Ramachandran (http://cbc.ucsd.edu/ramabio.html)

Sponsored by: Eugene M. Izhikevich, Editor-in-Chief of Scholarpedia, the peer-reviewed open-access encyclopedia

Reviewed by (http://www.scholarpedia.org/w/index.php?title=Anosognosia&oldid=0) : Anonymous

Reviewed by (http://www.scholarpedia.org/w/index.php?title=Anosognosia&oldid=144750) : Dr. Maria D Orfei, Department of Clinical and Behavioural Neurology, IRCCS Santa Lucia Foundation, Rome, Italy

Accepted on: 2015-04-01 07:22:36 GMT (http://www.scholarpedia.org/w/index.php?title=Anosognosia&oldid=149167)

Categories: In development (Neuroscience) | Awareness of intention | Free will