

Authors:

Reed C. Williams, MD
Riddhi Patira, MD
Eric L. Altschuler, MD, PhD

Stroke

Affiliations:

From the Departments of Physical Medicine and Rehabilitation (RCW, ELA) and Neurology (RP), Temple University School of Medicine, Philadelphia, Pennsylvania.

Correspondence:

All correspondence and requests for reprints should be addressed to: Eric L. Altschuler, MD, PhD, Department of Physical Medicine and Rehabilitation, Temple University School of Medicine, 3401 N Broad St, Philadelphia, PA 19140.

Disclosures:

The authors have no financial or other conflicts. Financial disclosure statements have been obtained, and no conflicts of interest have been reported by the authors or by any individuals in control of the content of this article.

Editor's Note:

Supplemental digital content is available for this article. Direct URL citations appear in the printed text and are provided in the HTML and PDF versions of this article on the journal's Web site (www.ajpmr.com).

0894-9115/16/9503-0230
American Journal of Physical Medicine & Rehabilitation
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DOI: 10.1097/PHM.0000000000000380

CASE REPORT

Double Disassociation of Anosognosia for Alexia and Simultanagnosia but Quantitative Awareness of Optic Ataxia

ABSTRACT

Williams RC, Patira R, Altschuler EL: Double disassociation of anosognosia for alexia and simultanagnosia but quantitative awareness of optic ataxia. *Am J Phys Med Rehabil* 2016;95:230–234.

A 66-yr-old man with a history of atrial fibrillation and a pacemaker developed sudden onset confusion, disorientation, and visual disturbance without motor weakness. Clinically, significant deficits were found in reading (alexia) and simultaneous multiobject perception (simultanagnosia), both of which the patient denied limitation in, and in vision—right hemianopsia—which he readily acknowledged. Visual acuity in the left visual field was normal. The patient also demonstrated a symptom of optic ataxia—a lack of coordination between visual inputs and hand movements—a deficit he also acknowledged. Work-up with computed topography revealed a left posterior cerebral artery infarct affecting the occipital lobe and extending to involve the parietal lobe and the splenium of the corpus callosum. The authors describe and discuss this fascinating case—the first case to their knowledge of a double disassociation of anosognosia for alexia and simultanagnosia but full, indeed quantitative, awareness of hemianopsia and optic ataxia. This case may be informative on the mechanism of anosognosia in general and supports intentional feed-forward and exemplar refference models. With regard to the rehabilitation process, appreciation that a patient has anosognosia for various deficits is crucial in recovery and health maintenance.

Key Words: Anosognosia, Alexia, Agraphia, Simultanagnosia, Balint Syndrome, Optic Ataxia, Oculomotor Apraxia

Over two millennia ago, the Roman philosopher Seneca,¹ in a letter to Lucilius, wrote, “This foolish woman suddenly lost her sight ... Incredible as it may appear ... She does not know she is blind.” Later, in 1885, von Monakow^{2,3} also alluded to the unawareness of illness. However, it was in 1914 that French neurologist Joseph Babinski ultimately described denial of insight into deficit, coining the term *anosognosia* (*a* = “without,” *noso* = “disease,” and *gnosia* = “knowledge”). Although identified in regard to many specific deficits, the most

common manifestations of anosognosia are for those basic abilities such as movement (denial of hemiparesis, as Babinski reported⁴) or vision (denial of cortical blindness, otherwise known as Anton syndrome⁵). Anosognosia is usually attributed to physiologic parenchymal damage to the parietal lobe of the brain; however, the exact mechanism of what underlies the neurocognitive pathology is unknown. As such, anosognosia remains both a vexing fundamental and clinical problem.

In 1905, Heilbronner⁶ reported a patient who failed picture interpretation but could list details and misread words but could name constituent letters, implying the inability to apprehend the details that make a whole. After seeing a similar case where a patient could see individual details of a complex scene but could not grasp the overall contextual meaning of the image, Wolpert⁷ coined the term *simultanagnosia* (*simultan* = “simultaneous” and *agnosia* = “without knowledge or ignorance to”) in 1924 to describe the inability to simultaneously synthesize elements of a scene.

Here, the authors present a most interesting case involving gross deficits of higher functions using the basic abilities of vision, represented by alexia (*a* = “without” and *lexia* = “reading”) and simultanagnosia, and movement, represented by optic ataxia—lack of coordination of visual inputs and hand movements. Remarkably, and fascinatingly, the authors report the first description of such to their knowledge, a double disassociation of anosognosia for alexia and simultanagnosia, with preserved, indeed quantitative, insight for optic ataxia. This double disassociation thus makes the case most informative as to the mechanism that may underlie anosognosia.

CASE REPORT

A 66-yr-old man with a history of atrial fibrillation and a pacemaker developed sudden onset confusion, disorientation, and visual disturbance without motor weakness and was emergently brought to the hospital for evaluation. The patient was awake and alert but disoriented to the exact nature of his hospitalization. A detailed neuromuscular examination revealed the patient to have normal strength in all extremities, normal touch sensation, and increased but symmetric muscle stretch reflexes in all limbs. Babinski reflexes were normal and he had no signs of myelopathy in his upper or lower limbs. Cranial nerve testing was normal except for a right-sided visual field cut (right homonymous hemianopsia). The patient’s visual acuity in the left field

was normal as were extraocular muscles, pupillary light reflexes, convergence, and accommodation bilaterally. With reading material presented in the left visual field, the patient was able to identify individual letters but could not read (alexia), confabulating responses that had no relation to the printed text. He could write spontaneously and to dictation but could then not read his own writing afterward. Unable to identify the color of objects (color anomia), he could correctly match them to like-colored objects. He was able to follow verbal commands and his speech was fluent. He had mild anomia when naming seen objects, but displayed none when objects were placed in either of his hands. When shown the “cookie theft/fall/kitchen spill” photograph (<http://www.ling.ohio-state.edu/~hana/x201m/other/11-cookieTheft.jpg>) and asked to describe the picture, he responded, “a boy,” “a woman,” and “a ladder (referring to a stool),” but could not, even with prompting, describe the context of the scene to any degree (simultanagnosia). Asked to reach out and touch an object in space, the patient demonstrated optic ataxia—making slow, discrete movements toward the target, while repeatedly ending up missing the point by an inch or two. Lastly, he had saccadic initiation failure (oculomotor apraxia) assessed with an optokinetic strip. The patient was aware of his visual field cut deficit, calling it an “obstruction,” but he had denial of the deficits (anosognosia) of his alexia and simultanagnosia. When told that he had not read correctly or that there was contextually more in the cookie picture, the patient blamed his errors on the visual “obstruction”—although both text and figure had been placed in the intact left visual field. Interestingly, however, the patient was precisely aware of his ataxic reaching difficulties reporting (correctly) he was “slightly off.”

Noncontrast computed topography revealed a left posterior cerebral artery infarct affecting the occipital lobe with hemorrhagic transformation extending into the parietal lobe and involving the splenium of the corpus callosum (Fig. 1). Magnetic resonance imaging, which would have been helpful in further assessing and localizing the lesion, was not performed secondary to the patient’s pacemaker. The etiology of the cerebrovascular injury was determined to be most likely embolic as the patient had been noncompliant with blood-thinning medications necessary for his cardiac arrhythmia. On day 2 of his acute hospitalization admission, physical, occupational, and speech and language pathology therapies were initiated. Seven days after acute neurologic hospitalization, the patient was admitted



FIGURE 1 Coronal noncontrast computerized tomography depicting a left posterior cerebral infarct involving the parietal and occipital lobes and the splenium (arrow) of the corpus callosum.

to the authors' institution's acute rehabilitation unit for continued therapies and medical care. In addition to the visual field cut, alexia, simultanagnosia, and optic ataxia, the patient was impulsive and had impaired balance while standing and walking. Physical therapy focused on balance and gait training. Physical, occupation, and speech therapists used cueing and scanning techniques to help the patient learn to compensate for the right visual field cut. After 2 wks of acute inpatient rehabilitation, the patient's balance had improved considerably; he was able to walk independently and he had improved compensation for the visual field cut. However, because of a need for supervision for impulsivity and problems that might result from alexia and anosognosia for alexia, the patient was discharged to a nursing facility.

DISCUSSION

Dejerine⁸ described alexia without agraphia (*a* = "without" and *graphia* = "write") in 1892, arising from a left occipital lesion rendering the left side of the brain blind and a lesion of the splenium of the corpus callosum disconnecting the right visual cortex from the language centers in the left side of the brain—presciently describing the first disconnection syndrome. Geschwind and Fusillo⁹ ascribed the inability to name seen colors while maintaining the ability to match colors to disconnection, as

per Dejerine's hypothesis. The authors' case embodies the same hemispheric disconnection secondary to the disruption of the corpus callosum at the splenium, while the remaining callosum fibers anterior to the splenium remain unaffected. Writing is preserved because the left-sided speech and motor centers are intact and remain connected. Geschwind further proposed that anomia for seen objects in alexia without agraphia was not more profound owing to pathways going first anteriorly from the right visual cortex and then crossing the corpus callosum, effectively bypassing the disrupted splenium fibers. From this, one could predict the excellent naming of objects by use of tactile cueing that the authors note in their case, as the more anterior tactile-to-language pathways remain intact. Further, with interruption of the visual-to-language pathway, the authors can explain the confabulation, per Geschwind, as the left hemisphere fabricates responses to questions regarding the disconnected right-hemispheric input. Additionally, the patient displays the symptomatic triad—simultanagnosia, optic ataxia, and oculomotor apraxia—of Bálint syndrome named for the neurologist who first identified it in 1909.¹⁰ Of note, Bálint syndrome is seen when there are two or more similarly located brain lesions in each hemisphere and it is most commonly seen in perfusion-deficient lesions affecting the parietal lobes.¹¹ It can be considered that the patient's inability to read is not classic alexia but is attributed

to simultanagnosia. However, the fact that the patient is unable to correctly name the color of a (singly) solidly colored object—a finding found in classic alexia⁹—is consistent with the patient having both alexia and simultanagnosia.

The most fascinating and important aspect of the authors' case is the previously unreported double disassociation of anosognosia for alexia and simultanagnosia, with quantitatively correct awareness for optic ataxia. The anosognosia for some deficits but awareness of others may be helpful in evaluating different models and explanations for anosognosia in general. The authors' findings support an intentional feed-forward model¹ that has been proposed for anosognosia for hemiparesis, and the authors also find it consistent with a reafference-comparator model for anosognosia: The patient has instantaneous actionable feedback of incorrect task failure in the reaching task; thus, there is no anosognosia for this, but the absence of such feedback is permissive for anosognosia of alexia and simultanagnosia. Further supporting such models is a replication¹² of the finding of the response when asked to read a selection of Chinese text: immediately responding, "that's Asian" and that he therefore could not read it.

With regard to rehabilitation of anosognosia, recovery can be difficult and problematic as compensating for a deficit is quite difficult when one is not aware of it in the first place. There are as yet no effective treatments for anosognosia. As such, rehabilitation in the absence of recognition may prove unsuccessful. Additionally, if further complicated by simultanagnosia, as is the case in the authors' patient, the inability to contextually analyze one's surroundings along with the unawareness of such inability not only could hinder them from initiating, participating in, or noting genuine therapeutic gain but also could prove dangerous. As it stands, the current therapeutic model of long-term neurocognitive rehabilitation focuses on using preserved abilities to perform functional tasks. These include teaching the patient to adapt to proceed through activities step by step, pausing after each to simplify a complex action. Patients are taught to recognize objects by tactile cues (shape, feel, and weight), rather than by color, appearance, or any writing on the object. For visual field cuts, patients are educated on the importance of head movement to elucidate as much peripheral visual field as possible as well as to adapt to low-vision rehabilitative techniques. The authors' patient's awareness of his visual field cut and optic ataxia may allow for good long-term results for these aspects of his rehabilitation. Unfortunately,

even with intense neurocognitive training, many patients require assistance. Family (and/or caregiver) training is therefore of utmost importance to successfully adapt to a patient's anosognosia for various deficits explained as an unconscious lack of insight. In the authors' case, the functional implications of anosognosia were discussed with the patient's family. For example, it is important to make sure that the family or caregivers understand that the patient might look at a sign and think he or she can read it and then act "accordingly"—whereas someone who is aware that they cannot read would ask for help for understanding what the sign says to do. This is particularly important as regards patients taking the correct medications in the correct doses.

CONCLUSION

The authors have presented the first case to their knowledge of a patient with a double disassociation of anosognosia for alexia and simultanagnosia but full awareness of optic ataxia. The authors' findings in the case are consistent with intentional feed-forward and reafference-comparator models for anosognosia. In the future, other cases of patients with a double disassociation of anosognosia for some deficits but awareness of others should be studied to gain further understanding of the mechanism of anosognosia. Possible treatments and rehabilitation strategies for anosognosia should be sought and studied.

Supplementary Checklist

CARE checklist: <http://links.lww.com/PHM/A160>

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