Right Brain: A descriptive account of two patients' experience with and adaptations to Bálint syndrome

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We provide a narrative account of 2 patients' experiences with Bálint syndrome, a rare and debilitating neurologic disorder characterized by optic ataxia, ocular apraxia, and simultanagnosia.

It was a quiet Thursday afternoon when A.S., a 68-year-old woman from a suburb of Chicago, awakened from a nap to the realization that something was terribly wrong. "I went to lie down, and when I got up, I couldn't find where the cabinets were, or the doors," she remembers. Over the next 2 days, A.S.'s confusion heightened as she increasingly lost her ability to name or even distinguish household objects that she'd been surrounded by for years. Unable to read the numbers on her telephone or to see where the bedroom wall ended and the bedroom door began, A.S. naturally thought that there was something wrong with her eyes and made an appointment with the ophthalmologist later in the week. She then did what so many stroke victims do; she put herself to bed early, hoping that everything would be better in the morning.

When the meeting finally came, it became clear that whatever A.S. was afflicted with, it was not a simple matter of visual impairment. "We took her to an eye doctor, because we didn't know what it was at the time," recalls her husband, Michael. "They ran tests on her eyes, and she passed them with flying colors!" A.S.'s visual acuity test tells the same tale; her vision tested out at 20/30 in each eye, and when she put on her glasses, it rose to 20/20. A.S. and her husband left the doctor's office with normal examination results and a neurologic referral in hand, wondering what sort of ailment could rob her of her ability to see the bathroom sink while leaving her with what we typically think of as perfect vision.

At 66 years old, J.D. was a robust and active man, who was described by those who knew him as a workaholic who could not sit still. After emigrating to the United States from the former Soviet Union, where he had been mayor of a large town, J.D. began his quest for the American Dream as a truck driver. Fast-forward 30 years and he is again a pillar of his com-

munity, where he was heading a large extended family while owning and operating his own trucking business.

J.D. was often on the road for long hours, and so his family thought little of it when he began swerving erratically on the long drive to his son's house for Thanksgiving dinner. "He worked double shifts for all 3 days before Thanksgiving because his workers wanted them off," his daughter recalls. "We just thought he was tired." Nor did his family know what to make of his bizarre attempts to serve himself at the table. "He was holding the spoon upside down. We all laughed," she recalls.

What happened next was no laughing matter, as J.D. began to experience left-sided weakness, facial drooping, and eventually a loss of consciousness that caused his family to rush him to the emergency room. That scene at the table is his last memory of the 2 months that followed. During that time, J.D. remained unconscious and confined to bed, until suddenly and unexpectedly awakening one afternoon and deciding that it was time to get up and stretch his legs. Since then, his road to partial recovery of powers of speech and the use of his left extremities has been a slow one.

The stories of A.S. and J.D. appear dissimilar at first glance, but they are in fact both descriptions of the onset of a rare neurologic disorder known as Bálint syndrome. Bálint syndrome is generally caused by one or more ischemic strokes to the parietal and occipital cortices. Magnetic resonance (MR) studies showed that A.S. had had a series of cardioembolic infarcts involving the parietal and occipital cortices, bilaterally (figure, A). J.D. probably had a series of ischemic infarctions due to severe atherosclerotic vertebrobasilar disease (figure, B).

The resulting syndrome is characterized by a triad of symptoms: 1) oculomotor apraxia (difficulty initiating voluntary eye movements toward an object), 2) optic ataxia (inability to reach for objects under visual guidance), and 3) visual simultanagnosia (constriction of the visual attentional field). Every neurology resident will have been acquainted with this syndrome, but to truly understand the Bálint

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Figure Axial magnetic resonance fluid-attenuated inversion recovery

Images from patient A.S. (A) and patient J.D. (B) show bilateral para-calcarine ischemia intermixed with secondary minor hemorrhagic changes stretching parietally.

patient is to witness the devastating effect that these deficits have on ability to function.

A.D. begins her day as we all do, by attending to her hygiene. However, because of her profound oculomotor apraxia, she cannot judge her location relative to the bathroom sink, mirror, and walls by sight, but only through the guidance of her other senses. Accordingly, she has learned that she must touch the sink at all times in order to remain oriented—otherwise her eyes are as good as closed. Likewise, during her daily shower, she must keep her hand on the shower bar at all times. She once failed to do this, misplaced her foot, fell, and broke 2 vertebrae. “Now I’m afraid of falling all the time,” she relates, adding that while she’s walking, the tension that accompanies this fear can be more exhausting than the act of walking itself.

Having showered and towed off with some difficulty, A.S. explains how, because she cannot trust her eyes to direct the toothpaste to her toothbrush, she must put the toothpaste directly in her mouth, and by trial and error move the toothbrush to meet it. At breakfast she uses her hands for most everything, and cannot eat soup at all. Nor is tidying up after herself any easier, as is evident from A.S.’s account of her attempt to pick a pen up off the floor: “I see it, and when I go to pick it up, it’s not there! I was aiming for the pen, and I see it, but I can’t pick it up!”

Because J.D.’s strokes are more recent and more extensive, his word-finding ability has not improved as much as A.S.’s has. During examination, his wife’s presence is immensely helpful to him; she conveys his meaning, alleviating the frustration that his expressive aphasia causes. The biggest obstacles for J.D. to surmount in his morning routine are dressing and brushing his teeth. While he has no difficulty removing articles of clothing, putting them on is an adventure: “I can’t find the front and the back—it’s all mixed up!” he exclaims.

Nor are his activities of daily living all that have been affected. Once an avid reader, J.D. can no lon-

ger direct his eyes to scan a line of text. Moreover, when asked to read a written word aloud, J.D. might only see one letter of it, or even just one line from that letter, depending upon the distance at which it is held from his face. This is because, as part of his simultanagnosia, J.D. is only able to consciously attend to objects placed within a tiny window of his visual field at any given time.

For J.D., another result of the stroke that caused his Bálint syndrome has been depression. “I was never sad like this before,” he said, and his wife confirms it. “He never once cried before, but now he cries often.” J.D. is having trouble adjusting to a life in which he cannot work to provide for his family as he once did, but rather requires significant assistance in performing simple tasks. “I think that’s the worst part for him because he’s a man with a lot of pride who worked all his life, and now he can’t do anything,” his daughter tells us.

Adaptation is key to recovered function in these patients. A.S. still finds a number of ways to enjoy herself. “The Library of Congress sends me audiobooks, and I can listen to the books anytime I want. They also sent me a radio so that I can listen to the news, because I tried to look at the paper and I can’t ... So when they told me I could get the books [on tape], I was so enthusiastic!”

Some of her adaptive strategies have been contributed by her large and devoted family. Several months ago, A.S.’s sons found a solution to her problem with distinguishing adjacent objects such as a doors and walls, or kitchen cabinets. “I can see yellow very well, so my son put yellow tape on the doors.” By taping the edges of a door, sink, or other object, A.S. is better able to distinguish object from surround, so that her house is no longer as treacherous as it once was.

At the end of our interview we asked A.S. if there was anything she would like to say to people interested in learning more about Bálint syndrome. She replied, “I’m talking to you for one reason—because I think if more people know about it, they won’t go through what I’ve gone through.” A.S. cherishes the hope that her story might motivate physicians to seek better treatments and therapies that might allow her and other Bálint patients to take up the threads of the former lives, and to “see again.”

AUTHOR CONTRIBUTIONS

Jason R. Cuomo was involved in the design of the study, in data collection and analysis, and in drafting and revising the manuscript. Dr. José Biller was involved in the conceptualization and design of the study, in data collection and analysis, and in revising the manuscript. Dr. Murray Flaster was involved in the design of the study, in data analysis, and in revising the manuscript.

DISCLOSURE

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