TO THE EDITOR: I applaud Segar et al. (Segar DJ, Chodakiewitz YG, Torabi R, et al: Deep brain stimulation for the obsessive-compulsive and Tourette-like symptoms of Kleefstra syndrome. Neurosurg Focus 38(6):E12, June 2015) in reporting the beneficial effects of deep brain stimulation (DBS) in a patient with a rare genetic condition, Kleefstra syndrome. The patient saw a reduction in anxiety, compulsive symptoms, and vocal tics following the procedure. Her overall functioning, including communication and social skills, improved. This case highlights the potential for DBS to treat neuropsychiatric conditions, suggesting the potential for expanding the indications for DBS. I have cared for this same patient for more than a decade and have been impressed with her outcome following placement of the DBS system. In addition to the excellent review of her history provided by the authors, there is one additional fact in this case that is worth emphasizing. At the time of DBS surgery, the patient had not yet been diagnosed with Kleefstra syndrome. Rather, her primary diagnosis at that time of surgery was autism.

The patient had displayed core features of an autism spectrum disorder since early childhood, including a restricted pattern of behaviors and impairments of communication and socialization. At the onset of adolescence, she saw a dramatic worsening in her level of functioning. Her language and socialization declined dramatically as coprolalia and obsessive-compulsive features increased. She spent an inordinate amount of time repetitively flushing toilets and watching traffic outside. She continuously rubbed her hands together, causing calluses to form and her fingers to bleed. She was socially withdrawn and stopped interacting with peers. In short, her core features of autism became more pronounced, and she saw an overall decline in daily functioning. For these symptoms, she was trialed on multiple different medications, including atypical antipsychotics, selective serotonin reuptake inhibitors, benzodiazepines, and antiseizure medications. She saw no benefit from the medication trials and instead declined further. At the time she underwent surgery, she had an extremely poor quality of life and was desperate for any improvement in her condition.

For most patients, the core symptoms of autism improve over time. In a subset, however, behavioral regression occurs during adolescence. This phenomenon has been well reported, and symptoms are often refractory to medical management. In one report of a 13-year-old boy with autism, dangerous self-injurious behaviors refractory to medical management improved after he underwent DBS of the basolateral complex of the amygdala. Like our patient, he, too, demonstrated benefits in emotional, social, and cognitive domains over the 24-month postoperative follow-up period. Implantation of our patient’s DBS system took place more than 4 years ago, and she has continued to maintain improvements through that time. The DBS leads were placed in the ventral capsule/ventral striatum, but our patient’s core symptoms were different from those of the other reported patient, consisting primarily of obsessive-compulsive disorder and Tourette’s syndrome as opposed to self-injurious behaviors. DBS of the basolateral nucleus of the amygdala has been advocated elsewhere as a potential intervention in cases of severe autism. Although these 2 cases suggest that DBS may be effective for severe, medically refractory symptoms in patients with autism spectrum disorders, the target for lead placement may need to be personalized to the patient’s primary symptoms. Based on the experience of this patient, further research into DBS in autism is warranted. Careful review of the patients’ genetic assessments and their phenotypic expression will be required to better predict the response to surgery and determine the optimal targets for lead placement.

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**Disclosures**
The author reports no conflict of interest.

**Response**

We are grateful to Dr. Gaitanis for his thoughtful comments and his extraordinary care of this unfortunate patient. It was his dedication and expert treatment of her that eventually led to her referral for DBS. What is most extraordinary in this patient is that her severe autism and substantial baseline cognitive impairment would have been significant contraindications for surgery. The original DBS surgery was performed by another surgeon, and when her behavior declined approximately 1 year after the initial implantation, the subsequent treatment team was admittedly cynical about the initial indications for surgery. The discovery that one DBS lead had migrated off target and the other lead had fractured prompted a carefully considered decision to re-implant both leads. The subsequent restoration of sustained clinical benefit once the DBS leads were properly replaced provided additional validation to the efficacy of DBS treatment in this patient and offers hope to other patients with severe autism spectrum disorders. As Dr. Gaitanis has pointed out, however, the optimal target and stimulation parameters remain undefined and may need to be individualized for patient-specific symptoms.

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