

Deep Brain Stimulation (DBS) for Dystonia

What is dystonia?

Dystonia is a neurological movement disorder characterized by sustained muscle contractions causing twisting and repetitive movements or abnormal postures. Dystonia can affect a specific area of the body or be more widespread throughout several muscle groups. These muscle contractions can be painful and interfere with day-to-day activities. Dystonia affects more than 500,000 people across Europe, including men, women, and children of all ages and backgrounds.¹

What are the different types of dystonia?²

There are two major categories of dystonia:

- Primary dystonia
- Secondary dystonia

A person diagnosed with primary dystonia does not have any neurologic signs or brain abnormalities. In addition, the origin of the illness is unknown. Secondary dystonia is associated with a lesion to the central nervous system often caused by an outside factor such as trauma, cerebral palsy, or stroke.

Dystonia can be even further characterized into subcategories:

- Generalized dystonia affects the entire body
- Focal dystonia affects one area of the body
- Segmental dystonia affects two or more contiguous body areas
- Hemidystonia affects one side of the body

What causes dystonia?³

The exact cause of dystonia is not fully understood. However, regardless of the category or type of dystonia, it is believed that the portion of the brain called the basal ganglia, which controls movement, is not functioning properly or has been damaged. Although there is not currently a cure for dystonia, there are a variety of treatments available that may reduce some of the symptoms associated with dystonia and improve quality of life.

What are the treatments for dystonia?²

There is currently no cure for dystonia, but there are a variety of treatments available that may provide relief of symptoms. Selection of treatment varies from patient to patient and is often dependent on the type and severity of the dystonia. The typical treatment options include drug therapy, injections of botulinum toxin, and surgery.

What is Deep Brain Stimulation Therapy?

Deep Brain Stimulation (DBS) is a surgical treatment that may help control some symptoms caused by dystonia. DBS is typically used to treat people with severe dystonia who fail treatment with pharmacologic agents and botulinum neurotoxin injections. In 2006, the UK's National Institute for Clinical Excellence (NICE) declared DBS to be a clinically safe and effective intervention for the treatment of dystonia.⁴

During a DBS procedure one or two insulated wires called "leads" are implanted in the brain. The leads are then connected to the stimulator (similar to a cardiac pacemaker), which is typically placed under the skin in the chest. The device produces mild electrical impulses that stimulate a specific region of the brain. This may help regulate incorrect signaling within the brain, resulting in improvement of symptoms. Although DBS is not a cure, it may help improve day-to-day experiences.

What is the clinical evidence behind DBS therapy?

There are several globally recognized tools used to monitor the severity of dystonia. The Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS) is a 120-point rating scale used to test the severity of dystonia in 9 body regions. This scale takes into account the severity and frequency of the dystonic movements. A higher score means greater impairment. The Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS) is another commonly used rating scale for cervical dystonia. This exam consists of three sections: Severity (range 0-30), Disability (range 0-30), and Pain (range 0-40). Similar to the BFMDRS, a higher score indicates greater impairment from the disease.

Multiple studies have demonstrated a reduction in BFMDRS and TWSTERS scores following deep brain stimulation therapy.

Primary Author	Study Design	Sample Size	Follow Up	BFMDRS or TWSTERS	Change in Mean Scores BFMDRS or TWSTERS	% Improvement in BFMDRS or TWSTERS
Cersosimo et al, 2009 ⁵	Prospective Case Series	9	6 mos.	BFMDRS (Motor Scale)	Baseline: 46.9 ± 24.3 Post DBS: 24.1 ± 16.9	Not Reported
Houeto et al, 2007 ⁶	Prospective, Controlled, Multicenter	22	1 mo.	BFMDRS (Motor Scale)	Baseline: 46.3 ± 21.1 Post DBS*: 26.7 ± 14.9	Not Reported
Vidailhet et al, 2005 ⁷			12 mos		Baseline: 46.3 ± 21.3 Post DBS: 21 ± 14.1	51
Vidailhet et al, 2007 ⁸			36 mos.		Baseline: 46.3 ± 21.3 Post DBS: 19.8 ± 17.4	58
Kiss et al, 2007 ⁹	Prospective, Single-Blind, Multicenter	10	12 mos.	TWSTERS (Severity Score)	Baseline: 14.7 ± 4.2 Post DBS: 8.4 ± 4.4	43
Kupsch et al, 2006 ¹⁰	Prospective, Controlled, Multicenter	40	6 mos.	BFMDRS (Motor Scale)	Baseline: 36.4 ± 24.6 Post DBS: 20.2 ± 18	46
Volkman et al, 2012 ¹¹			5 yrs.		Baseline: 43.4 ± 28.6 Post DBS: 15.4 ± 16.3	60
Ostrem et al, 2011 ¹²	Prospective Pilot	9	12 mos.	TWSTERS (Total Score)	Baseline: 53.1 ± 2.57 Post DBS: 19.6 ± 5.48	62.9
Vidailhet et al, 2009 ¹³	Prospective	13	12 mos.	BFMDRS (Motor Scale)	Baseline: 44.23 ± 21.12 Post DBS: 34.69 ± 21.87	24.4

*Post-surgery ventral stimulation score

What is the Vercise™ Deep Brain Stimulation System?

The Vercise Deep Brain Stimulation System is the first system engineered for precise neural targeting to customize therapy for the treatment of patients with levodopa-responsive Parkinson's disease which is not adequately controlled with medication and also now offering another therapy option for the treatment of patients with intractable primary and secondary dystonia, for persons 7 years of age and older. The Vercise System consists of a stimulator (also referred to as an "implantable pulse generator" or IPG) similar in size and shape to a pacemaker. The stimulator produces small electrical signals that travel along thin wires called "leads", which are connected to the stimulator. The stimulator is placed under the skin just below the collar bone, while the leads are positioned in a specific part of the brain.



From left to right: the Vercise Deep Brain Stimulation System consists of a remote control, stimulator with attached leads, and charger.

The Vercise DBS System features:

- **Accurate & Precise Targeting:** Multiple Independent Current Control (MICC) was designed for fine control of stimulation position and shape.
- **Innovative Design:** The Vercise lead has a robust multi-lumen construction with 8-contacts and a broad span.
- **Patient Comfort:** The Vercise IPG has a gentle contoured oval shape with the smallest footprint on the market at 20 cm².
- **Longevity:** The Vercise DBS System with unique rechargeable battery technology has a 25 year battery life to help reduce surgical interventions.
- **Patient Convenience:** The Vercise charging system and remote control are completely cordless and designed to make charging simple. The wireless charging system allows patients to be active while charging.
- **Reliability:** Only the Vercise DBS System offers Zero-Volt™ Battery Technology. The battery can be completely discharged without causing battery failure or damage – even when the patient forgets to recharge.
- **Quality:** At Boston Scientific we believe that best-in-class quality is essential to long-term viability of DBS therapy. Through substantial investments in R&D and quality, our engineers invented unique features designed to deliver unmatched reliability and convenience in a simple, rechargeable device.

About Boston Scientific

Boston Scientific transforms lives through innovative medical solutions that improve the health of patients around the world. As a global medical technology leader for more than 30 years, we advance science for life by providing a broad range of high performance solutions that address unmet patient needs and reduce the cost of healthcare. For more information, visit www.bostonscientific-international.com and connect on [Twitter](#) and [Facebook](#).

Contact

Name

Title

Phone number

Email address

References

¹ Dystonia Europe (2013). About Dystonia. Retrieved September 17, 2013, from <http://dystonia-europe.org/msa-board>

² Albanese A, Asmus F, Bhatia KP, et al. EFNS guidelines on diagnosis and treatment of primary dystonias. *Eur J Neurol* 2011; 18: 5-18

³ Janavs JL, Aminoff MJ. Dystonia and chorea in acquired systemic disorders. *Journal of Neurology Neurosurgery and Psychiatry*. 1998;65(4):436-445.

⁴ National Institute for Clinical Excellence. Deep brain stimulation for tremor and dystonia (excluding Parkinson's disease), National Institute for Clinical Excellence, August 2006, (IPG188).

⁵ Cersosimo MG, Raina GB, Benarroch EE, Piedimonte F, Aleman GG, & Micheli FE. "Micro lesion effect of the globus pallidus internus and outcome with deep brain stimulation in patients with Parkinson disease and dystonia." *Mov Disord* 2009. 24(10), 1488-1493.

⁶ Houeto JL, Yelnik J, Bardinet E, Vercueil L, Krystkowiak P, Mesnage V, Vidailhet M. "Acute deep-brain stimulation of the internal and external globus pallidus in primary dystonia: functional mapping of the pallidum." *Arch Neurol*. 2007. Sep;64(9):1281-6.

⁷ Vidailhet M, Vercueil L, Houeto JL, Krystkowiak P, Benabid AL, Cornu P, Pollak P. "Bilateral deep-brain stimulation of the globus pallidus in primary generalized dystonia." *N Engl J Med* 2005, 352(5), 459-467.

⁸ Vidailhet M, Vercueil L, Houeto JL, Krystkowiak P, Lagrange C, Yelnik J, Pollak P. "Bilateral, pallidal, deep-brain stimulation in primary generalized dystonia: a prospective 3 year follow-up study." *Lancet Neurol* 2007. 6(3), 223-229.

⁹ Kiss ZH, Doig-Beyaert K, Eliasziw M, Tsui J, Haffenden A, Suchowersky O. "The Canadian multicenter study of deep brain stimulation for cervical dystonia." *Brain* 2007, 130(Pt 11), 2879-2886.

¹⁰ Kupsch A, Benecke R, Müller J, Trottenber, T, Schneider GH, Poewe W, Volkmann J. "Pallidal Deep-Brain Stimulation in Primary Generalized or Segmental Dystonia." *New Engl J Med* 2006, 355(19), 1978-1990.

¹¹ Volkmann J, Wolters A, Kupsch A, Muller J, Kuhn AA, Schneider GH, Benecke R. "Pallidal deep brain stimulation in patients with primary generalised or segmental dystonia: 5-year follow-up of a randomized trial." *Lancet Neurol* 2012. 11(12), 1029-1038.

¹² Ostrem JL, Racine, CA, Glass GA, Grace JK, Volz MM, Heath SL, Starr PA. "Subthalamic nucleus deep brain stimulation in primary cervical dystonia." *Neurol* 2011. 76(10), 870-878.

¹³ Vidailhet M, Yelnik J, Lagrange C, Fraix V, Grabli D, Thobois S, Pollak P. "Bilateral pallidal deep brain stimulation for the treatment of patients with dystonia-choreoathetosis cerebral palsy: a prospective pilot study." *Lancet Neurol* 2009. 8(8), 709-717.

All cited trademarks are the property of their respective owners. CAUTION: The law restricts these devices to sale by or on the order of a physician. Indications, contraindications, warnings and instructions for use can be found in the product labeling supplied with each device. Information for the use only in countries with applicable health authority product registrations.

The Vercise™ Deep Brain Stimulation System is indicated for use in unilateral or bilateral stimulation of internal globus pallidus (GPI) or the subthalamic nucleus (STN) for treatment of intractable primary and secondary dystonia, for persons 7 years of age and older.