Deep Brain Stimulation for Blepharospasm: What Do We Know?

Presented by Francisco Ponce, MD Reported by Hilliard Kelly Pruitt

Since the late 1980s, over 200,000 people have been treated with Deep Brain Stimulation (DBS), also known as neuromodulation, worldwide. This procedure is not new or futuristic. DBS has been around since 1986 and was approved by the FDA in 1997. The track record is that it is a safe, non-destructible, reversible, and adjustable therapy.

The first pacemaker for the heart was introduced in 1958. The deep brain stimulator is essentially a pacemaker for the brain: the device is implanted in the chest, just as the heart pacemaker, but the wires go under the skin, ascending through holes behind the forehead, through which electrodes enter the brain and basically tap into the circuit that goes awry with conditions such as dystonias.



Like many therapies, Deep Brain Stimulation is a product of an accidental discovery in medicine. During the early days of diagnosing Parkinson's disease, James Parkinson noticed that a person who suffered from a stroke that affected his entire hemisphere had stopped shaking on that side of the body. This is where the idea of the role for surgery to treat movement disorders originated. That approach was refined during the 1900s from major operations to a wire that would essentially "cook" the brain. DBS is considered more of an electrical therapy for conditions like Parkinson's and other types of dystonia rather than surgical treatment.

All movement disorders, including Parkinson's, arise in the brain. Originally, these conditions were treated through lesions by creating a small "stroke" to disrupt the circuits causing them. DBS therapies pre-dated high quality MRI imaging, before precise detail and understanding of the cellular activity of the brain, electrophysiology, and circuitry was available. Consequently, throughout the 1990s, MRIs were not used for DBS. Procedures prior to 2012 were safe and effective, although it took a long time

to pinpoint the right spot for the electrode. This required the patient to be fully conscious to provide feedback as to avoid causing irreparable damage due to a stroke. Many surgeons would only treat one side at a time due to the time commitment and patient's level of discomfort.

Thanks to new advancements such as direct targeting using MRI, the increasing ubiquity of portable CT Scanners for image guidance, and the Entropy CT Scanner GPS system during surgery to allow on-screen viewing, beautiful imaging is available with exquisite detail of the brain's anatomy. Coupled with decades of experience performing the operation, the procedure now just has to be implemented accurately. In just the last 8 years, DBS has graduated from being an intervention of last resort to a treatment that can be implemented closer to the onset of the condition.

These improvements have taken a lot of the guesswork out of the procedure. When MRI targeting is combined with a portable CT scanner used in the operating room, a high quality photo from the MRI can be combined with the CT scanner as soon as the electrode is placed in real time. This can be compared to the surrounding brain obtained beforehand to ensure accuracy. When MRI targeting is combined with a portable CT scanner used in the operating room, the most desirable result is achieved: the patient can now be asleep during the operation.

Treatment of dystonia by DBS, including blepharospasm and Meige, was approved in 2003 through a Humanitarian Device Exemption (HDE). HDE is allowed when a condition is rare enough that a multi-centered, randomized blind clinical trial cannot be launched in the same way they do for pharmaceuticals and medications. HDE allows doctors to perform DBS for dystonia, which suggests adequate data to provide a benefit for patients with the condition.

The FDA describes their approval for dystonia under the following labeling: "primary dystonia, including generalized and/or segmental dystonia, Hemidystonia, and cervical dystonia (torticollis) in patients seven years of age or above." It is under this description that conditions such as blepharospasm and Meige are treatable with DBS. Medtronic is the only company that has a Deep Brain Stimulator that has been FDA approved in the treatment of dystonia. There are about 88 patients, average age of 57, that have been described in various publications as having blepharospasm, and who have undergone DBS treatment. Most participants had more than one symptom, which fits into the category of segmental dystonia. Best results for DBS for dystonia have been achieved in children and adolescents with the DYT1 mutation. The key to best outcomes come down to good patient selection, safe and accurate lead placement, and good programming.

Class 1 evidence supports that DBS has an effect on dystonia. Insuring best outcomes with Deep Brain Stimulation requires being under the care of the right neurologist, who determines patient selection, while the movement disorder specialist is trained to tease out alternative diagnoses that might not be as amenable to these types of therapies to insure a patient's qualification as a candidate for DBS according to their experience. While the neurosurgeon's role in treating conditions with these "brain pacemakers" on the team and journey is discreet, their purpose is to make sure this therapy is implanted safely and accurately. The task of the neurosurgeon is to find how to deliver the proven electrical therapy with the minimum possible risk. This is a surgery for quality of life, so the stakes are very high. Different conditions have different latencies to the therapy. A patient with an action tremor may see the disorder stop immediately upon the pacemaker activation. With other conditions, it could be days or sometimes weeks before any results are achieved, and will see a buildup of benefits over the course of months. The goal is to restore function, and the only way to gauge an operation's success is if a patient can say their life has improved after six months, and they would do it again if they had the opportunity.

After decades of learning and research, a solid product is now being delivered. A new era of DBS is being ushered in. By speaking the language of the brain, using information about where the electrode is, and recording the activity within the brain, DBS will ultimately result in an intelligent pacemaker that knows where to go to maintain control. Advances in technology continue to open the door to smarter and more efficient programming through advanced control and understanding of the patient's symptoms. While DBS is an option for blepharospasm, recommendation by a movement disorder specialist remains critical.