



## **FAQ for Patients and Families: Deep Brain Stimulation for Dystonia**

### **History of surgery for dystonia**

Brain operations to treat various forms of dystonia were first performed 40-50 years ago. The operations involved a precisely controlled lesioning (destruction) of deep structures in the brain involved in movement control. Two different deep structures, called the thalamus and the globus pallidus, were operated upon. Lesioning was performed by freezing or coagulation. Surgical techniques were much less advanced in that era than they are today, so the results were not consistent and complications were frequent.

In the last 15 years there has been a resurgence of interest in the surgical treatment of dystonia. There are several reasons for this: Techniques for operating on deep brain structures have become much more precise and safe. In addition to the older technique of brain lesioning, there is a new technique, deep brain stimulation (DBS), which involves placing a stimulation electrode rather than performing a brain lesion. DBS can alter the function of abnormal brain tissue in a way that improves movement, but it is reversible if there is an unwanted effect, and it can be adjusted during a routine office visit to optimize the degree of benefit. Thus it is somewhat safer than lesioning.

Patients with other, more common movement disorders are increasingly undergoing surgical treatment. These other movement disorders include tremor and Parkinson's disease. Success with these movement disorders has encouraged greater interest in surgery for more rare movement disorders such as dystonia. Since 1995, a small number of publications in neurology and neurosurgery journals have shown that deep brain stimulation for several type of dystonia can be effective.

### **What type of brain stimulation is best for dystonia?**

The most accepted brain target for deep brain stimulation in generalized dystonia is the globus pallidus. This is a peanut-sized structure deep in the brain whose electrical activity is abnormal in dystonia. For dystonias affecting mainly the neck and face, however, the optimal target is less clear. We prefer DBS over lesioning because it is reversible and adjustable, and does not permanently destroy any part of the brain. Thus, pallidal DBS is our preferred surgery for dystonia at this time.

### **Is DBS for dystonia a standard, widely accepted therapy?**

As of 2011, only about 300 cases of DBS for dystonia have been described in the world medical literature, and the longest time any patient has had this therapy is about 15 years. Thus it cannot

be considered a standard therapy until more procedures are done and the results of them published.

### **Have other children had this surgery for dystonia?**

The most severe form of generalized dystonia often begins in childhood and may become severe by the early teen years. These patients may be excellent candidates for surgery. We have implanted DBS devices in over thirty children (age less than 18). For children under 16, the surgery is usually performed under general anesthesia, with no awake testing.

### **How does DBS improve symptoms of dystonia?**

The way that DBS works for dystonia is incompletely understood. There is a circuit in the brain, involving the basal ganglia (deep structures) and cortex (surface of the brain) that controls movement. Electrical activity in this circuit is known to be abnormal in dystonia. DBS acts as a “brain pacemaker” to over-ride the abnormal electrical activity. It turns out that the globus pallidus, a structure in the basal ganglia, is an efficient place to alter the circuit abnormalities.

### **Who should consider having surgery for dystonia?**

Patients should consider deep brain stimulation for dystonia if they meet the following criteria:

- The patient has been evaluated and treated by a qualified movement disorders neurologist who has clearly diagnosed the type of dystonia, and excluded other neurological problems.
- The dystonia adversely affects quality of life by interfering significantly with normal activities or causing social isolation.
- The patient's neurologist has attempted treatment with a variety of medications, which should at least include sinemet and anticholinergic medications such as trihexyphenidil (Artane). Baclofen and muscle relaxants such as clonazepam are also often tried before considering surgery.
- The dystonia affects too large a body area to be treated effectively with injections of botulinum toxin (botox); or attempts at injection with botox have been tried and failed.
- The patient and family clearly understand the nature and complexity of DBS therapy as well as the fact that the therapy is too new to guarantee successful treatment in any individual case.

In addition to the above considerations, DBS for dystonia appears to be more effective for certain forms of dystonia than others. Primary dystonias (patients without brain abnormality on MRI) are more likely to benefit than those with secondary dystonia (patients with brain abnormalities seen in MRI that are causing the dystonia). Common causes of secondary dystonia are stroke, anoxia, or cerebral palsy.

## **Will insurance companies pay for DBS for dystonia?**

In 2003, DBS for dystonia received a special category of approval by the Food and Drug Administration (FDA), called a “Humanitarian Device Exemption”. As a result many insurers do cover the procedure. While we have had several insurers initially deny coverage, we have found that most eventually agreed to cover after appealing the decision and supplying medical literature that evaluated the procedure. Insurance approval or denial will be determined prior to the procedure. In our experience, eventual approval is likely, but not guaranteed.

## **How is the surgery performed?**

There are several available surgical methods for DBS implantation. For adult patients, brain electrodes are most commonly implanted with the patient awake, using only local anesthetic and occasional sedation. For this procedure, a rigid frame is attached to the patient's head and a brain imaging study (MRI or CT) is obtained with the frame in place. The images of the brain and frame are used to calculate the position of the desired brain target and guide instruments to that target with minimal trauma to the brain. To maximize the precision of this method, "brain mapping" is performed in which fine microelectrodes are used to record brain cell activity in the region of the intended target to confirm that it is correct, or to make very fine adjustments of 1 or 2 millimeters in the intended brain target if the initial target is not exactly correct. Accurately “mapping” the brain using this technique requires that the patient be awake, calm, and cooperative.

At Cincinnati Children’s Hospital, we use an alternative method for DBS electrode placement, in which the surgery is performed entirely within a high resolution MRI scanner. The advantage is that patients may be under general anesthesia for the entire implantation procedure, since no “brain mapping” is performed. Rather, precise implantation is achieved using special, FDA-approved software and guidance devices and by monitoring the entrance of the electrode into the brain with “real-time” MRI. The procedure is performed in an operating room that was specially designed so that MRI can be performed during surgery.

First, the patient is put fully to sleep under general anesthesia then moved to the MRI table. The head is kept still on the MRI table with a special frame that also helps in imaging the brain. The hair is shaved and local anesthetic is injected into the skin to numb the area of the incision. Images are taken of the brain to visualize the target and plan the path for electrode placement. After the skin is opened, a small plastic frame is secured to the skull. The surgeon, with the help of special computer software, adjusts the frame so that it will guide the electrode directly to the target. All along this process, pictures are taken of the brain to ensure the electrode follows the intended path. Once the electrode is in place, it is anchored to the skull with a plastic cap, the frame is removed and the scalp is closed with sutures and staples. A final MRI is performed to confirm the final electrode placement and assess for any bleeding around the electrode.

To ensure adequate healing of each of the incisions, the incisions are covered with sterile dressings and protected with gauze wrapping around the head. The patient is then awakened from general anesthesia and brought to the post-operative care unit. The patient will then be

monitored overnight in the intensive-care unit and is usually transferred to a regular hospital room the next day.

When the patient is eating well and his/her pain is controlled with oral pain medications, he/she may go home with the family. You will be given instructions for how to care for the wounds while at home. During this time, the patient is encouraged to participate in as many normal daily activities as tolerated, though heavy lifting (>5 lbs), aggressive play, and strenuous activity are not permitted.

After the head wounds have healed sufficiently (1-4 weeks, depending on the patient), the patient is brought back to the hospital for placement of the pulse generator. Again, the patient is asleep for the entire procedure. First, a small incision is made in the scalp to identify the end of the brain electrode. This is then connected to an extension wire which is tunneled beneath the skin to another incision made just beneath the collar bone. A pocket is made in the soft tissue below the collarbone for the pulse generator to be placed. These incisions are then sutured closed so that all hardware is underneath the skin.

For older children, this may be an outpatient procedure (home the same day as the surgery). Younger children may need to spend the night in the hospital. This procedure takes about 40-60 minutes. There is usually some soreness after this procedure which lasts a few days. Again, normal, but not strenuous, activity is encouraged for 2 weeks after the procedure.

Patients with dystonia on both sides of the body, or affecting the neck or face, will usually require electrodes to be placed on both sides of the brain.

### **What are the risks of surgery?**

The most serious potential risk of the surgical procedure is bleeding in the brain, producing a stroke. This risk varies from patient to patient, depending on other medical factors, but generally ranges from 1-3%. If stroke occurs, it usually occurs during or within a few hours of surgery. Another risk is infection, especially of the deep brain stimulating hardware, which occurs in about 4-5% of patients. If an infection occurs, it is usually not life threatening, but may require immediate removal of the entire DBS system.

### **What are the benefits of surgery?**

DBS surgery does not cure dystonia in any case. When the stimulator is turned off or if it malfunctions, the symptoms return. DBS can decrease the abnormal movements and postures of dystonia but usually does not totally eliminate them. The degree of benefit appears to vary with both the type of dystonia and the duration of the symptoms. Adolescents and young adults with inherited forms of dystonia appear to get very significant benefit. For patients with dystonia due to stroke, cerebral palsy, or head trauma, the benefit may be only mild. Adults who have had dystonia for many years probably have less improvement than those with more recent onset of symptoms. Patients with fixed skeletal deformities (where a joint is frozen in place) also may get

less benefit.

### **What are the cosmetic considerations with DBS surgery?**

Complete shaving of the head is not necessary for surgery. However, a large patch of hair from on top of the head is shaved immediately before surgery when the patient is sedated. Many patients prefer to have the entire head shaved at the time of surgery so that all the hair grows back evenly (the patient should not do this themselves before surgery). When hair grows back, incisions are not visible.

There are a few incisions made for DBS surgery: two 1.5 inch incisions on top of the head, a 1 inch incision behind the ear, and a 2.5 inch incision in the chest just under the collarbone. Younger, or particularly thin, children may have an incision beneath each collarbone if two smaller pulse generators are implanted. The cap used to anchor the DBS electrode (under the scalp) forms a slight bump, which again may be somewhat visible in the case of a receding hairline.

All parts of the device are internal (under the skin), so there are no wires sticking out. In a thin person, the connecting wire running down the neck may be visible as a slight bulge when the head is turned all the way to the opposite side. The incision for the pulse generator in the chest is closed with particular attention to minimize scar formation; this incision would be visible with the shirt off, or in a swimsuit, or in a low-cut evening gown. In thin persons, the pulse generator itself forms a bulge under the skin in the area of the upper chest that may be apparent if the area is uncovered, but is not visible through clothing.

### **What tests are needed prior to surgery?**

All patients must have had a brain MRI at some time since the onset of dystonia. All patients must have a detailed videotaped neurological evaluation by one of our movement disorder neurologists: Dr. Donald Gilbert or Dr. Steven Wu. This evaluation may last several hours and documents the severity of dystonia.

### **How should the patient prepare for surgery?**

For 10 days prior to surgery, patients must not take aspirin, any aspirin containing drugs, related drugs such as ibuprofen (Advil, Motrin) or naproxen (Naprosyn), or Vitamin E. These drugs can increase the risk of bleeding. The evening before surgery, patients should wash their head, neck, and chest with hibiclens (or other soap containing chlorhexidine) in the shower. The morning of surgery, the patient should take their anti-dystonia medications. Patients should inform the surgeon if they develop a cold, cough, or any type of infection in the days prior to the surgery.

## **What type of follow-up is needed after surgery? Who will program the DBS unit?**

Patients normally leave the hospital two to three days after surgery. We ask patients to return to our clinic 1 week later for suture removal and check of the incisions by our DBS nursing specialist, and approximately 4 weeks later to see the surgeon and neurologist in the Movement Disorders Surgery Clinic. The programming is done at the first postoperative visit. Some patients have temporary disorientation for a few days after surgery due to temporary brain swelling, and if this occurs programming is deferred until the mental state completely returns to baseline. For subsequent programming needs after the initial stimulator activation, the patient is encouraged to continue in our Movement Disorders Clinic.

In the first month following DBS implantation, some patients may develop an infection of the device or of the skin over the device. This would present as drainage, increasing redness, increasing swelling, or increasing pain starting a few days to a few weeks post-surgery. It is very important to let our office know IMMEDIATELY if such signs are noted, since early wound care may be effective at salvaging the device. If such symptoms are ignored for even a few days, however, the patient will usually have to have all of the hardware removed.

Patients will typically require replacement of the pulse generator after 2-3 years, depending on the exact settings of the device. This is an outpatient procedure that takes about 30 minutes. In 2009, a rechargeable pulse generator became available, which lasts longer than the non-rechargeable pulse generators (more information below).

## **What types of pulse generators (implanted DBS control unit) are available?**

As of 2011, we are implanting 3 types of pulse generators in dystonia patients, all made by Medtronic, Inc.: Activa SC, Activa PC, and Activa RC. The choice of pulse generator for an individual patient is based on considerations of size of the device, desire for patient control over the stimulation level (see below), and need for recharging.

The Activa SC is a small pulse generator that runs a single brain electrode (one side of the brain). This may be implanted in patients who only need one brain electrode for dystonia that primarily affects the opposite side of the body. Young or small children may have two SC devices placed, one on each side, because the smaller size is better tolerated by these patients. The Activa RC

The Medtronic Activa RC is a re-chargeable pulse generator. It has the advantage of a very long interval (estimated at 10 years) prior to the need for surgical replacement. It has the disadvantage that the patient must charge it with a device strapped over the pulse generator, for a few minutes every day or for a few hours each week. At this time, we are recommending the Activa RC only for patients with existing DBS systems who have used up their non-rechargeable system less than 2 years from implantation. Patients must have a teaching session preoperatively about the recharging requirements, to make sure they are willing to do this.

### **Can patients control the DBS device themselves?**

Following surgery, the patient is given the Medtronic Access Review unit, a hand-held battery-operated unit that can be used to determine if the device is on or off, to turn it on or off, and to check battery life. All devices can be turned on or off by the patient.

Other aspects of stimulation, such as voltage level or frequency of stimulation, can be controlled by the patient depending on what type of pulse generator unit is implanted. Patients with the Medtronic Activa PC or Activa RC can control these aspects with their hand held access review device. Patients with the Medtronic Soletra cannot control any aspect of stimulation other than the on/off switch. However, the Soletra may have other advantages with respect so size (smaller than the Kinetra and Activa PC) and battery longevity (longer than the Activa PC).

### **How long does it take before the full benefit of DBS is apparent?**

For reasons that are not fully understood, it often takes several months for the full benefit of any particular programming setting to reach its maximum level.

### **Are there any restrictions on a person's activity after a DBS system is implanted?**

For at least 4 days after surgery, the patient should refrain from flying in commercial aircraft. For one week after surgery, the incisions should be kept dry, so for showering in the first week incisions must be covered with an occlusive dressing. Tight clothing or tightly fitting hats should be avoided in the two weeks after surgery. After the incisions are completely healed (2-3 weeks), the patient may return to all normal activities, including exercise. Normal physical activities will not harm the device. Security devices (such as those in airports or stores) will not harm the device or the patient, although in rare cases they may activate the on-off switch, thus turning off a DBS system that had been on. The loss of benefit to the patient may take hours or days to be apparent. When traveling extensively away from home, patients should carry their Medtronic Access Review unit so that they can easily re-activate the DBS system if it is de-activated by a security device.

### **Can I have an MRI scan after DBS?**

After DBS implantation, patients should avoid most types of MRI exams, as the exam may produce heating of the brain electrode. At UCSF, we have developed a specialized, low energy protocol for brain MRI, allowing us to perform postoperative brain MRI safely. However, we do not recommend any other forms of MRI (such as spine or chest MRI), because safe conditions for performing these have not been worked out.

## **Summary**

There are more surgical treatment options for patients with dystonia than ever before. For patients with primary dystonia, without fixed (immobile) deformities of the skeleton or tendons, DBS offers significant relief of many symptoms. The major risk is a 2% risk of stroke, due to bleeding in the brain. DBS is a relatively complex therapy requiring regular neurological follow-up and battery changes every 2-3 years. The time to consider DBS surgery is when quality of life is no longer acceptable on optimal medical therapy as administered by a movement disorders neurologist. Secondary dystonias, such as those due to stroke, trauma or cerebral palsy, may have modest benefit from DBS, but not as much benefit as primary dystonia.