FAQ for Patients: Deep Brain Stimulation for Dystonia

Contents

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. Although DBS of the globus pallidus often improves the dystonia, there can also be subtle impairments (slowing down) of previously normal limbs with this technique. Therefore, for patients with dystonia affecting mainly the neck and face, we are offering enrollment in a clinical trial of subthalamic nucleus DBS. This target may prove to be better tolerated for neck dystonia, but the clinical study is not complete.

We prefer DBS over lesioning because it is reversible and adjustable, and does not permanently destroy any part of the brain. Thus, pallidal or subthalamic DBS is our preferred surgery for dystonia at this time.

Is DBS for dystonia a standard, widely accepted therapy?

As of 2010, 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.

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 two structures in the basal ganglia, the globus pallidus and subthalamic nucleus, are efficient places 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 anticholingergic medications such as trihexyphenydil (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?

DBS for dystonia recently 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. In the most common method, implantation of the brain electrode is performed with the patient awake, using only local anesthetic and occasional sedation. The basic surgical method is called stereotaxis, a method useful for approaching deep brain targets though a small skull opening. For stereotactic surgery, a rigid frame is attached to the patient's head just before surgery, after the skin is anesthetized with local anesthetic. 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. After frame placement, MRI/CT, and calculation of the target coordinates on a computer, the patient is taken to the operating room. At that point an intravenous sedative is given, a Foley catheter is placed in the bladder, the stereotactic frame is rigidly fixed to the operating table, a patch of hair on top of the head is shaved, and the scalp is washed. After giving local anesthetic to the scalp to make it completely numb, an incision is made on top of the head behind the hairline and a small opening (1.5 centimeters, about the size of a nickel) is made in the skull. At this point, all intravenous sedatives are turned off so that the patient becomes fully awake.

To maximize the precision of the surgery, we employ a "brain mapping" procedure 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. The brain mapping produces no sensation for the patients, but the patient must be calm, cooperative, and silent during the mapping or else the procedure must be stopped. The brain's electrical signals are played on an audio monitor so that the surgical team can hear the signals and assess their pattern. The electronic equipment is fairly noisy, and the members of the surgical team often discuss the signals being obtained so as to be sure to interpret them correctly. Since each person's brain is different, the time it takes for the mapping varies from about 30 minutes to up to 2 hours for each side of the brain. The neurological status of the patient (such as strength, vision, and improvement of motor function) is monitored frequently during the operation, by the surgeon or by the neurologist.

When the correct target site is confirmed with the microelectrode, the permanent DBS electrode is inserted and tested for about 20 minutes. The testing does not focus on relief of dystonia but rather on unwanted stimulation-induced side effects. This is because the beneficial effects of stimulation may take hours or days to develop, whereas any unwanted effects will be present immediately. For the testing, we deliberately turn the device up to a higher intensity than is normally used, in order to deliberately produce unwanted stimulation-induced side effects (such as tingling in the arm or leg, difficulty speaking, a pulling sensation in the tongue or face, or flashing lights). The sensations produced at high intensities of stimulation during this testing are experienced as strange but not painful. We thus confirm that the stimulation intensity needed to produce such effects is higher than the intensity normally used during long-term function of the device.

Once the permanent DBS electrode is inserted and tested, intravenous sedation is resumed to make the patient sleepy, the electrode is anchored to the skull with a plastic cap, and the scalp is closed with sutures. The stereotactic headframe is removed. The patient then receives a general anesthetic to be completely asleep for the placement of the pulse generator in the chest and the tunneling of the connector wire between the brain electrode and the pulse generator unit. This part of the procedure takes about 40 minutes.

At UCSF, we have also been investigating an alternative method for DBS electrode placement, in which the surgery is performed entirely within a high resolution MRI

scanner. This method is being used under an investigational protocol. One advantage is that patients may be under general anesthesia for the entire implantation procedure, since no physiological testing is required.

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.

Can children have 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.

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 patch of hair from on top of the head to behind an ear is shaved immediately before surgery when the patient is sedated. Many patients elect to get a short haircut after surgery (must be at least 2

weeks afterwards) so that the hair grows in evenly. When hair grows back, incisions are not visible.

There is often puffiness around the eyes for a few days after surgery, but this goes away rapidly.

There are generally 3 incisions made for DBS surgery: a 5 cm (2 inch) incision on top of the head, a 2.5 cm (1 inch) incision behind the ear, and a 6 cm (2.5 inches) incision in the chest just under the clavicle. For adult patients with receding hairlines, a slight scar from an incision will be visible on top of the head, but is not especially prominent. 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 pectoral area that may be apparent if the area is uncovered, but is not visible through clothing.

How experienced is UCSF with deep brain stimulation?

At UCSF we have performed over 1000 DBS surgeries for a variety of movement disorders since 1998. As of 2010, we have performed 170 DBS electrode implants specifically for dystonia. We have several research studies focusing on surgery for dystonia. One is to study the outcome of an alternative surgical target, the subthalamic nucleus, in in patients with dystonias that primarily affect the face and neck. A second study focuses on abnormal electrical activity in the brains of persons with dystonia.

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 Dr. Jill Ostrem, lasting several hours, to document the severity of dystonia. Most adult patients require a formal neuropsychological evaluation, which is usually done by Dr. Caroline Racine, our neuropsychologist. Patients with dystonia that affects the neck usually require a cervical spine MRI prior to surgery.

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 not take their anti-dystonia medications. However, the patient should take any medications they normally take for other problems, such as high blood pressure. 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 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 initial programming is done at UCSF at the one week 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. For patients who live at a distance from the Bay Area and have a neurologist with a DBS programmer, we are happy to advise other neurologists regarding optimal programming strategies for dystonia.

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 2010, we are implanting 3 types of pulse generators in dystonia patients, all made by Medtronic, Inc.: Soletra, 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 Medtronic Activa RC is a re-chargeable pulse generator. It has the advantage of a very long interval (estimated at 9 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.