Pallidal Deep Brain Stimulation

Treatment for Dystonia

Dystonia

Dystonia is a neurological disorder characterized by involuntary muscle contractions resulting in abnormal postures. These movements may be painful. Although the exact cause of dystonia is unknown, it is believed that there is an alteration of the nerve impulses in the part of the brain called the basal ganglia, which is deep in the center of the brain.

Some forms of dystonia are genetic (called primary dystonia) and to date there have been 13 genes identified, some of which can be identified through genetic testing. More commonly, primary dystonia is non-genetic and the cause is unknown. This is referred to idiopathic dystonia. In secondary dystonia, dystonia is due to injury or medication use such as neurolyptics (such as Haldol) and the cause is known.

Classifications of Primary Dystonia

Primary dystonia is classified descriptively by the age of onset and the distribution of symptoms.

Early onset is when symptoms appear before age 26. If symptoms appear after this age, they are considered late onset. The two most common types of dystonia are generalized and focal dystonia.

Generalized dystonia begins in one or both legs and gradually spreads to other parts of the body, usually the back and neck. It usually begins in children or teens and occurs in approximately 3:100 000 people in North America.

In focal dystonia only one area of the body is affected. For example it can affect the neck (cervical dystonia), hands (writer's cramp), eyes (blepharospasm). Focal dystonia is the more common with an estimated 30:100 000 people have a form of focal dystonia.

Treatments

Dystonia can sometimes be treated with medications such as Trihexyphenidyl (Artane), Diazepam (Valium), Lorazepam (Ativan), or Levodopa (Sinemet). The most common and current treatment for dystonia is Botulinum Toxin injections (Botox). These wear off after 3-4 months needing repeat injections. Botox can also be very expensive if you are not covered by medication insurance.

If medication or injections have lost effect or do not work then surgical intervention is an option.

History of Pallidal Deep Brain Stimulator

Surgery on the globus pallidus began in the 1950's with what was called a pallidotomy. The surgeon would make a small lesion in the target area which was shown to help alleviate dystonia. However side effects were high and the outcomes were questionable.

Neurosurgeons now have much improved techniques for brain scanning (CT and MRI), stereotatic head frames, and electrophysiology, to assist in locating the precise target and decreasing complications.

Pallidal Deep Brain Stimulation (DBS)

DBS is a technique using a stereotactic procedure. A small lead (a very thin insulated wire electrode) is implanted into the globus pallidus (GPi) replacing the pallidotomy. The lead is connected to an impulse generator (battery) by way of a small wire. The impulse generator delivers electrical pulses which stimulate the globus pallidus resulting in an improvement in the dystonia. It is, therefore, somewhat like a brain pacemaker.

To accomplish this, precise measurements of the patient's brain structures are obtained by the placement of a stereotactic head frame and an MRI of the patient's head prior to the surgery. From these measurements a computer calculates the exact angles and depths of the trajectories of the micro-recording electrodes to place them in the globus pallidus.

Two small holes are made in the skull on either side of the midline behind the hairline. A tiny recording electrode is guided down into the target area of the globus pallidus. Recordings are obtained from the brain cells in the area to determine where the overactive cells are, and to identify the location of other important structures in the area. When the target area has been identified on both sides of the brain the DBS electrode (1.2 mm diameter insulated wire) is implanted into the globus pallidus.

The impulse generator delivers electrical impulses which stimulate the target areas. The stimulation reduces the abnormal postures, movements and pain on the opposite side of the body.

This is a 2 part surgery with the implantation of both leads followed by connection and implantation of the lead extension and battery a few days later. If the target areas cannot be identified during surgery the leads will not be implanted. If all goes well the patient will return to the operating room, on a day arranged by the neurosurgeon, to have the lead extensions and the battery implanted under a general anesthetic. The stimulator leads are tested during surgery and again prior to implanting the batteries. The amount of stimulation needed to improve movement will need to be adjusted by our nurse coordinator. These adjustments will require visits back to the Movement Disorders

Clinic. Initially these adjustments may need to be done frequently until the proper balance of medications and stimulation is reached.

This treatment is not a cure for the underlying condition; however it can significantly improve the patient's quality of life. It is also important to note that it may take up to 3 months to determine the final outcome of the surgery.

Risks of Surgery

All surgery carries some element of risk. For GPi-DBS the risks of complication are low but may include any of the following:

- 1:200 (0.5%) chance of death due to hemorrhage
- 2 to 3 % chance of a major complication such as a stroke, hemorrhage/bleeding
- 10% chance of permanent numbness, weakness, speech and/or swallowing problems, drooped face, bladder urgency or frequency, or difficulty with concentration and memory
- 40% chance of mild, transient occurrences of the above side effects
- Small chance of seizures or infection
- Transient confusion
- Headache

Initially, periodic adjustments in programming the stimulator will be needed. During this adjustment period some people may experience the following side effects:

- Transitory, but mild changes in sensation in a limb or a small area on the face
- Occasional transient dizziness or feeling of being lightheaded

Risks due to the Implanted Stimulator

The overall risk of any these hardware-related complications (all of which are treatable but may require repeat surgery) over 4 years is 20%, meaning about 5% per year.

- Infection / rejection / breakage of hardware
- Malfunction of hardware

The stimulator requires a battery which will last 2-4 years, and replacement will require a minor surgery. It is expected that within the next few years a re-chargeable battery will be developed.

Preparation for Surgery

A decision to consider surgical treatment is made with your doctor. The patient is then referred to the Surgery for Movement Disorders Program. The patient will be assessed by Dr. Kraft, and Dr. Kiss to determine whether he/she meets the criteria for surgery. If the criteria are met and the patient is agreeable to proceed, a number of baseline assessments are arranged and the patient is booked for surgery. The baseline assessments include: a CAT or MRI scan of the brain, neuropsychological testing, and a

video taping session along with some additional testing and counseling by the nurse coordinator. More detailed counseling may be arranged with a psychiatrist if needed.

In Hospital

The patient will be admitted to hospital early in the morning the day of surgery. After admission the patient will go to a day unit and from there will be taken to the MRI department in radiology where he/she will be fitted with a special frame which will be secured to the head using local anesthetic to freeze the scalp. An MRI will be done with the frame on to obtain the necessary coordinates to be used for calculations during surgery.

The patient will then be taken to the operating room where the procedure will take place. It is difficult to determine exactly how long the surgery will take but it often takes until late afternoon, early evening to operate on both sides of the brain. One of the nurses from the Movement Disorder Clinic will keep the relatives informed as to the progress of the surgery.

The patient will stay in hospital until he/she returns to the operating room within a few days to complete the second part of the procedure. It is best to provide a phone number where the patient's relatives are available and the surgeon will call them after the procedure. Relatives may visit with the patient several hours after returning from surgery.

Both the family and the patient need to be aware that it is not uncommon for patients to be confused for a few days following this surgery. This confusion, if it occurs, usually clears and the patient returns to his/her preoperative state.

The patient will return to the Movement Disorder Clinic one week after discharge to see the nurse-coordinator for suture removal and incision check. The nurse co-coordinator will continue to keep contact by phone to make an appointment for programming. Programming can take weeks to months making small adjustments in stimulator settings weekly in combination with changes/reduction in medications.

The Cost

In Canada, the surgical procedures and any support services are covered by the health plans of all provinces. If you are not an Alberta resident, the cost of the Deep Brain Stimulators may not be covered and you will need to discuss this with your Provincial Health Care Plan. If you are not a resident of Canada contact the administrators of your health insurance plan to ask if you are eligible for coverage.

Summary

Surgical procedures are now available for Dystonia as well as for Essential Tremor and Parkinson's disease. The type of surgery that would be most appropriate for the patient is decided on an individual basis, in consultation with the program neurologist, neurosurgeon and the patient.

At present, surgery is being offered only to those patients who are in good health and who have moderate to moderately severe dystonia. Risks are higher for those who are frail and older than 70 years of age. While complications can occur, the procedure is relatively safe and effective.

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