
***SURGICAL INTERVENTIONS
FOR DYSTONIA***



**Dystonia Medical
Research Foundation**

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TABLE OF CONTENTS

I. Introduction 2

II. Peripheral Surgeries 3

Cervical Dystonia/Spasmodic Torticollis 3

 The Bertrand Procedure: Selective Peripheral Denervation
 Rhizotomy
 Microvascular Decompression

Spasmodic Dysphonia/Laryngeal Dystonia 7

 Selective Laryngeal Denervation and Reinnervation
 Thyroplasty

Blepharospasm 10

 Myectomy Surgery

Generalized Dystonia & Hemidystonia 12

 Intrathecal Baclofen

III. Brain Surgery: Lesioning Procedures & Deep Brain Stimulation 15

Lesioning Procedures: Pallidotomy & Thalamotomy

Deep Brain Stimulation

Comparing Lesioning & DBS

Children & Brain Surgery

IV. Conclusion 25

V. Appendix 26

Dystonia

The Nervous System

Anatomy

 Eyes

 Neck

 Larynx

VI. Sources 31

I. INTRODUCTION

As research about dystonia progresses, great attention is being paid to the role of surgical interventions for alleviating symptoms. Surgical treatments for dystonia may be an option for cases that do not respond to oral medications or botulinum toxin injections. Researchers are actively refining current techniques and collecting information about which patients may benefit the most from surgical treatments.

There is no single surgical procedure that can be applied to all forms of dystonia. Surgical procedures for dystonia can be divided into two broad categories: *brain surgery* and *peripheral surgery*. Peripheral surgery includes procedures that target parts of the body other than the brain.

In both brain and peripheral procedures, the goal of surgery is to interrupt the faulty communication between the brain and muscles that causes involuntary muscle movements. Surgery intends to treat symptoms and improve function but does not cure the underlying condition.

Because dystonia is a chronic disorder, the management of symptoms is an ongoing, lifelong process. Just as medications and botulinum toxin injections are often not singular solutions to an individual's dystonia, surgery is one component of the total management of dystonia. Surgery does not necessarily eliminate the need for additional forms of treatment. However, in many cases surgery improves quality of life and reduces the need for medications or botulinum toxin. Like all surgical interventions, operations to treat dystonia are associated with the risk of certain complications.

The patient selection process for determining if an individual is a candidate for surgery is deliberate and precise. Only a neurologist or neurosurgeon who specializes in movement disorders can recommend surgery for dystonia. The cost of surgery varies by procedure and medical center, and coverage is often on a case-by-case basis for Medicare and private insurance. The success of any surgical procedure lies heavily in proper diagnosis, the experience of the clinical team, and the skill and artistry of the surgeon.

II. PERIPHERAL SURGERY

The symptoms of dystonia occur when muscles of the body receive faulty information from the brain causing them to contract involuntarily. These faulty messages originate most commonly in a part of the brain called the basal ganglia. These messages are conveyed over brain pathways to the spinal cord and, from the spinal cord, extend into the muscles via nerves.

Peripheral surgeries occur outside the brain and generally target the specific nerves and muscles affected by the incorrect messages from the brain. Peripheral surgeries are generally used to treat focal dystonia. An exception is intrathecal baclofen, which targets the spinal cord and is used to treat generalized or hemidystonia. However, for the purpose of this publication, a discussion of intrathecal baclofen is included under the category of peripheral surgeries.

CERVICAL DYSTONIA/SPASMODIC TORTICOLLIS

The Bertrand Procedure: Selective Peripheral Denervation

Selective peripheral denervation surgery for cervical dystonia is commonly referred to as the *Bertrand procedure*. In the 1970s, Dr. Claude Bertrand, with the collaboration of Dr. Pedro Molina-Negro, developed this procedure as a peripheral approach to treat cervical dystonia. The term *selective* refers to the care taken to identify the muscles of the neck affected by dystonia, and the term *denervation* refers to cutting the nerves that supply those muscles. The purpose of the Bertrand procedure is to reduce abnormal contractions in the affected muscles by severing the nerves to these muscles. The goal of the procedure is to leave intact the supply of nerves to unaffected or less-affected muscles.

This procedure is tailored to address the unique needs and symptoms of each patient. The initial approach is often to denervate the muscles causing the most prominent dystonic movement, knowing that some

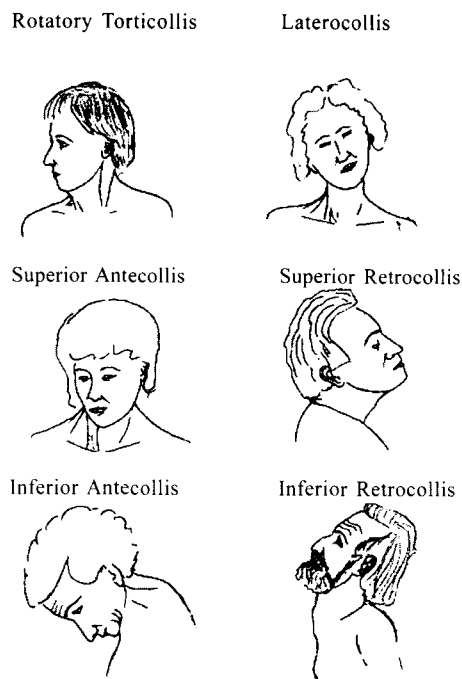
residual movements may remain from lesser-affected muscles. If the results do not sufficiently alleviate symptoms, a second procedure may be performed. In many cases, the initial surgery is enough to significantly improve the abnormal posture. More aggressive surgeries, in which all cervical muscles involved in the dystonia are denervated in a single operation, may result in temporary weakness in the neck.

An essential part of the procedure is the pre-operative evaluation to properly identify the muscles involved and to assess if the procedure will benefit the individual. Patients who may be eligible for the surgery are observed clinically by the physician and with EMG equipment to monitor muscle activity and pinpoint the muscles affected by the dystonia.

One basic element of the Bertrand procedure is to cut rootlets of the spinal accessory nerve, which

supply sternocleidomastoid muscles in the neck, and to spare the nerves to the trapezius muscle. The spinal accessory nerve is one of 12 cranial nerves that originate in the brainstem, which is the junction of the brain and the spinal cord. A second element of the Bertrand procedure is cutting the posterior rami (branch) of one or more spinal nerves along the cervical vertebrae. (This element of the procedure is called posterior *ramisectomy*.) Spinal nerves are arranged in pairs along the length of the spinal cord and supply muscles and organs. Some research

Figure 1



The six elemental forms of spasmodic torticollis.

suggests that the ramisectomy increases the improvement in persons who have become resistant to botulinum toxin therapy.

To date over 2,000 cervical dystonia patients have undergone this procedure. Some centers report significant improvement in as many as 88% of cases. Although the procedure may benefit individuals with a range of symptoms, the categories of patients who may have the best results from the Bertrand procedure are individuals in which:

- Symptoms mainly affect the neck
- Symptoms have stabilized for 3 years
- The head turns to one side (rotational torticollis)
- The head is tilted (laterocollis)
- The head turns and is pulled backwards (rotational torticollis with superior retrocollis)
- The head turns and tilts forward (rotational torticollis with superior antecollis)
- The head is pulled back (superior retrocollis)

Dystonia in which the head turns both to the side and either back or forward may have the best outcome. Individuals who respond to botulinum toxin therapy as well as non-responders may be eligible. The procedure may also be an option for a small number of patients with generalized dystonia who have very defined symptoms in the neck.

Side effects may include numbness in the back of the head, tightness at the surgery site, some remaining movements, difficulty swallowing, and lack of benefit. Patients are often able to go home after two or three nights in the hospital.

Studies have demonstrated that the Bertrand procedure can significantly improve the posture of the neck with a better range of motion. Physical therapy following the procedure is very important to preserve range of motion.

Rhizotomy

Since the late 19th century, physicians have attempted to treat cervical dystonia surgically by cutting the spinal nerves that supply the contracting muscles. While the ramisectomy element of the Bertrand procedure involves excising specific branches of the cervical spinal nerves near the muscle (away from the spinal canal and spinal cord), pioneering surgeons initially attempted to remove the nerve at the root (inside the spinal canal near the spinal cord). The procedure of cutting a nerve at the root is called a *rhizotomy*.

Each spinal nerve has two roots: a dorsal (posterior) sensory root and a ventral (anterior) motor root. The sensory root conveys sensory information from the muscles, joints and skin to the spinal cord, and the motor root conveys signals from the spinal cord to the muscles. Cutting the sensory roots does not alter dystonia but does help spasticity. Cutting the motor roots—which means cutting 85-95% of the root innervating a dystonic muscle—will effectively denervate the muscle but at the cost of inducing significant weakness.

Ventral rhizotomy for dystonia was used widely between the 1930s and the 1970s and was often combined with a denervation of the accessory nerve. By destroying the nerve at the root, the effect on the muscle is more generalized and may cause a greater degree of weakness. The results of these procedures were overwhelmingly disappointing and caused a high incidence of complications in dystonia patients. The ventral rhizotomy as a treatment for cervical dystonia was eventually replaced in the 1970s by the Bertrand procedure.

However, select medical centers continue to incorporate rhizotomies in their surgical approaches to cervical dystonia, and researchers continue to explore the effects of severing the nerves at different locations along the cervical vertebrae in order to provide additional surgical options for dystonia patients. Selective dorsal rhizotomies are commonly done to treat spastic stiffness of the limbs (diplegia).

Microvascular Decompression

Microvascular decompression surgery for cervical dystonia is based on the idea that various blood vessels compress and irritate some of the cranial nerves (particularly the spinal accessory nerve), resulting in dystonic symptoms. This surgical procedure involves relocating the blood vessels without injuring the vessels, nerves, or muscles. The relocated blood vessels are held in place with small implants. Sectioning of nerves may or may not be included in the procedure. There is very little published data about this procedure, and it has been largely abandoned for dystonia patients. Adverse effects include lack of benefit, cerebrospinal fluid leakage, and stroke. There is evidence to suggest that if a patient undergoes decompression surgery and is not satisfied with the results, the partial removal of the occipital bone and scarring that results from microvascular decompression may make it difficult for a subsequent surgeon to perform a safe and effective Bertrand procedure.

SPASMODIC DYSPHONIA/LARYNGEAL DYSTONIA

Selective Laryngeal Denervation and Reinnervation

Selective laryngeal adduction denervation and reinnervation (SLAD/R) is a surgical procedure to treat adductor spasmodic dysphonia/laryngeal dystonia by cutting (denervating) selected end branches of the recurrent laryngeal nerve, which is a branch of the vagus cranial nerve.

The first attempts to reduce the spasms of spasmodic dysphonia by severing the laryngeal nerve took place in the 1970s. Cutting the laryngeal nerve paralyzed the muscles controlling one side of the larynx so that the larynx could not contract excessively. Early results were good, but symptoms reappeared in many patients. Subsequent pioneers in the field sought to improve the procedure by varying the method by which the nerve was separated from the muscle. Recurrence of symptoms as well as breathy voice continued to be a problem in many patients.

The element that distinguishes SLAD/R from previous incarnations of the surgery is that, after the recurrent laryngeal nerve is cut away from the thyroarytenoid and lateral cricoarytenoid muscles, the nerve stumps are hooked up to another nerve (reinnervated), one that is not associated with the dystonia. Supplying the muscle with another nerve prevents the problematic branch of laryngeal nerve from growing back and reconnecting to the muscle. Preventing the laryngeal nerve from communicating to the muscle prevents the spasms from returning and helps to change the closing forces of the larynx. It is important to note that the procedure is performed bilaterally, unlike previous nerve operations performed for adductor SD. Because the disorder originates centrally in the brain, it likely exists bilaterally in the larynx. It is therefore logical to treat both sides.

The procedure is accomplished by making an incision in the neck and then creating a small window into the laryngeal cartilage to expose the underlying nerves and muscles. An operating microscope is often used to aid in identification and suturing of the tiny nerve branches. The procedure takes three or four hours to complete. Great care is taken to preserve the back part of the cartilage that protects the nerve branches to the breathing muscles.

SLAD/R is best suited for individuals with spasmodic dysphonia without a tremor. It may be an option for persons who are not satisfied with botulinum toxin treatments. More than two hundred persons with spasmodic dysphonia have undergone SLAD/R over the course of about 10 years. During the initial recovery period, all patients experience temporary voice breathiness and some experience swallowing difficulty. These issues resolve over a few months and the patient is left with a near-normal voice, free of spasm. Studies have indicated that as many as 85-90% of patients are very satisfied with the results of surgery, and the results, so far, have been life long.

Thyroplasty

Thyroplasty surgeries include a group of surgical techniques to modify the cartilage surrounding the larynx. These adjustable and reversible procedures involve manipulating the cartilage by implanting wedges or shims to hold the tissue in place. A number of variations of this procedure are currently used and are effective for restoration of the voice after paralysis or in changing the pitch of the voice.

Type I thyroplasty has been used for the abductor variety of spasmodic dysphonia. In this procedure, the vocal cords are brought closer together in hopes of decreasing the effect of the abductor spasms. Results are mixed, with some patients getting good relief and others having minimal effect.

Type II thyroplasty is a procedure for adductor spasmodic dysphonia that involves spreading the vocal cords apart by inserting a shim that prevents them from contacting each other during the spasms that occur with this disorder. Although some patients have reported good relief of vocal strain, others feel the trade off to a breathy and weak voice is excessive.

Researchers in the US and abroad continue to investigate thyroplasty procedures. The advantage of these procedures is that they are largely non-destructive and do not alter the muscles or nerve supply of the larynx. They work through adjustment of biomechanics alone and are theoretically reversible, although in practice the reversibility may be limited by scarring.

BLEPHAROSPASM

Myectomy

Surgical removal of the eyelid and brow-squeezing muscles is referred to as a *myectomy* procedure and is used to treat blepharospasm. Myectomy prevents the muscles surrounding the eyes from being stimulated by removing the muscle.

Before the availability of botulinum toxin, myectomy was essentially the only treatment option for blepharospasm. The introduction of botulinum toxin injections in 1989 benefited many persons with blepharospasm thereby changing the population of individuals eligible for myectomy. Candidates for myectomy became those for whom botulinum toxin is not sufficient.

Just as the patient selection changed, the procedure itself evolved. Initially, the procedure involved removing all eyelid-squeezing muscles in both upper and lower lids as well as the brow area at one time. At the present time, the procedure is tailored to the needs of the patients. It is most common for the surgeon to remove the muscle in the upper eyelids and brow (*full upper myectomy*) and then re-evaluate the need for a *lower myectomy* at a later date. Patients heal faster when the procedure is done in stages, and some individuals do not require the lower myectomy.

The full upper myectomy may be done entirely through an eyebrow incision. The incision lies immediately adjacent to the brow hair and allows access to the upper lid orbicularis muscle, and part of the lower lid orbicularis muscle as well as the procerus and corrugator muscles in the brow area. Most of the orbicularis muscle is removed during the eyelid surgery. A strip of dense muscle is left at the margin of the upper eyelid to help maintain some voluntary closure and to protect the eyelash roots.

A *limited upper myectomy* is a partial upper myectomy. It is available for those individuals who are benefiting from botulinum toxin but need something extra to restore function of the eyes. It may be helpful for those patients who have apraxia (difficulty opening the eyes) or for those who in addition to blepharospasm have ptosis (drooping lids). Partial removal of the orbicularis may subsequently decrease the need for botulinum toxin in these patients. A limited myectomy is done through an upper eyelid crease incision and involves removal of the orbicularis muscle within the upper lid area only. Because there is less tissue removal than the full upper myectomy, patients recover in less time. A limited myectomy also gives more predictable cosmetic improvement because less tissue is removed. It is not designed to replace a full upper myectomy. Most patients will still require botulinum toxin injections following the limited myectomy procedure.

Persons who have stopped responding to botulinum toxin as well as those rare individuals who fail to respond at all may be eligible for myectomy. Individual surgical centers have treated hundreds of blepharospasm patients with myectomy. Techniques used for cosmetic surgery, such as sculpting the fat beneath the brow and manipulating the placement of the brow, may be implemented to provide a beneficial aesthetic as well as functional result.

Myectomy surgery can be done under local or general anesthesia. The healing process following a myectomy may take up to a year. In most cases, the patients are able to keep their eyes open immediately following the operation. However, considerable swelling, hematomas (blood accumulation in lid), lymphedema (tissue fluid), and bruising may be present early in the post-operative period and prevent complete eyelid opening. Cool compresses in the first four to five days followed by warm compresses are very helpful at settling the lid swelling and bruising.

There are numerous potential side effects associated with myectomy surgery that are predictable and, to some degree, occur in most patients. Numbness of the forehead region often occurs and is usually temporary

but may last a year or more. Loss of tissue volume in the eyelid area may occur with the muscle removal, but the improved brow, lid position, and decreased eyelid wrinkling generally gives an improved cosmetic appearance. Decreased eyelid closure occurs as a result of eyelid muscle removal and may require the need for additional artificial tears and lubricating ointment. As the eyelid swelling resolves, the eyelid closure improves and the dry eye symptoms generally improve. Chronic lid swelling which may last six months or longer in some patients can be a chronic and troublesome complication. Chronic lid swelling is much less severe and persistent in the modern myectomy practices in which upper and lower lid myectomies are performed separately. Infection, hematoma, brow hair loss, and abnormal positioning of the lower lid can occasionally occur but are uncommon.

Patients continue to improve in function as well as in appearance for about six months to a year after myectomy surgery. Reports have shown that visual disability is improved in approximately 90% of patients. Some patients have more improvement than others. Touch-up procedures are required in some cases, and some individuals continue to require botulinum toxin injections.

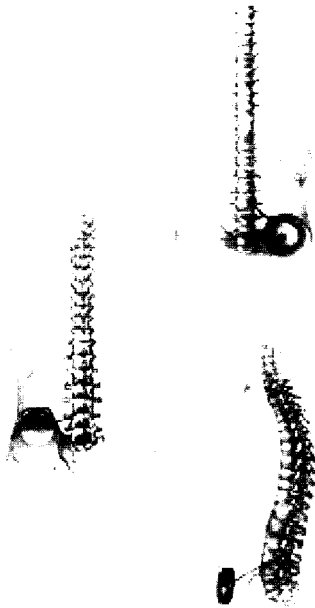
GENERALIZED DYSTONIA & HEMIDYSTONIA

Intrathecal Baclofen: The Baclofen Pump

Baclofen (Lioresal®) is a medication introduced in the late 1960s as a treatment for spasticity. The medication is also commonly used to treat select cases of dystonia. Baclofen in the spinal fluid around the brain and spinal cord supplements the body's supply of a chemical neurotransmitter called GABA, which relaxes muscle movement. The drug may be given orally, but very high doses must often be used to ensure that the drug saturates the blood stream and reaches the spinal fluid. High doses of baclofen may cause intolerable side effects such as muscle weakness and fatigue. A surgically implanted baclofen pump delivers baclofen directly to the spinal fluid, and only very small doses are needed. (The term *intrathecal* means in the spinal fluid.)

Intrathecal baclofen therapy is a non-destructive, adjustable, and reversible treatment. Several hundred dystonia patients have been treated with intrathecal baclofen over the course of about 10 years. It has been used for children and adults with generalized dystonia (both primary and secondary) and hemidystonia who respond to baclofen. Many persons treated with intrathecal baclofen have a combination of dystonia and cerebral palsy. Intrathecal baclofen may be used to treat dystonia affecting the upper and lower limbs.

In order to determine if an individual is eligible for intrathecal baclofen, he/she will undergo a screening test to observe the body's response to baclofen. A response to the oral drug may necessitate a screening test to observe the body's response to a small dose injected directly into the spinal fluid. The medication is injected using a standard lumbar puncture or spinal tap. The screening test procedure involves injection of the medication followed by several hours of observation. Relaxation of the muscles indicates that an implanted baclofen pump will likely be effective. The effects of the screening test are temporary and may last several hours after the injection. If a patient does not respond at all to the screening test, a second test using the same procedure may be tried the next day or at a later date.

Figure 2

Intrathecal baclofen hardware in body.

Some physicians use a continuous intrathecal infusion of baclofen as a screening method, since more patients respond to continuous infusion than to single injection screening doses. In the infusion technique, a small catheter is inserted into the spinal fluid and is connected to an external pump that infuses baclofen in increasing doses over two to three days.

Starting intrathecal baclofen therapy involves surgically implanting a programmable pump in the body. The device is usually placed either to the right or left of the belly button, beneath the skin and fat of the abdomen. The pump is connected to a thin tube that is tunneled around the side of the body to the back. A small needle introduces the tube to the spinal canal. Once the surgical incisions are closed, the pump is adjusted by a remote computerized device to deliver the amount of medication appropriate for the individual. The procedure takes one to two hours, and the hospital stay may range from four to seven days. Modest improvement of symptoms may be noticeable before the individual is discharged from the hospital, and it may take six months or more to achieve the full extent of benefit.

Regular maintenance is a key component of intrathecal baclofen therapy. Regular exams and physical therapy may be a component of postoperative care. Pumps must be refilled every one to four months in the physician's office as a straightforward outpatient procedure. The pump is refilled by inserting a thin needle through the skin, into the pump. The frequency of refilling the pump depends on the dose required. If necessary, the doctor may adjust the delivery rate of the pump at the time of the refill by remote control. The pump battery lasts approximately seven years, depending on how much medicine is programmed to be delivered each day. Before the battery runs out, the pump will need to be replaced with a new pump through a surgical procedure. The catheter can usually stay in place and be reconnected to the new pump.

Baclofen in the spinal fluid relaxes muscles throughout the body, and appears especially effective for targeting dystonia in both the upper and lower half of the body. ITB may be more effective for treating secondary dystonia than for primary dystonia.

Studies have shown that intrathecal baclofen can dramatically improve symptoms and quality of life. Some centers have reported significant improvement in as much as 85% of patients. However, like any surgery, the procedure is not without risks. Hardware complications may also arise including infection and catheter breakage and disconnection. In a small percentage of cases, symptoms may resume or worsen within the first year. The most common side effects are constipation, decreased muscle control, and drowsiness.

III. BRAIN SURGERY: LESIONING PROCEDURES & DEEP BRAIN STIMULATION

The goals of brain surgery for persons with dystonia are to decrease muscle spasms, increase mobility and function, and improve pain.

There are currently two categories of brain surgery for dystonia: *lesioning procedures*, which involve selective destruction of targeted, abnormal brain tissue, and *deep brain stimulation (DBS)*, which mimics the effects of lesioning by manipulating selective brain areas with non-destructive electrical pulses.

Although risks exist, case studies have shown that both lesioning procedures and DBS can result in marked improvement of dystonia with minimal complications. Some patients are able to decrease or altogether stop drug therapy following surgery.

Dystonia most often originates in a part of the brain called the *basal ganglia* which are involved in the coordination and control of muscle movements. The basal ganglia are a group of structures that include the *globus pallidus* (also called the pallidum), the *thalamus*, and the *subthalamic nucleus*. Lesioning procedures for dystonia usually target the globus pallidus or the thalamus; deep brain stimulation usually targets the globus pallidus or subthalamic nucleus. The globus pallidus is responsible for the output of messages from the basal ganglia. The recipient of this output is the thalamus. The subthalamic nucleus is a tiny structure located directly beneath the thalamus and is connected to the globus pallidus.

Different parts of the brain work together to help the body accomplish a specific task, such as tapping the foot. The parts of the brain communicate via pathways of individual brain cells that transmit chemical messages from one to the other. In an individual with dystonia, the pathways that facilitate the movement of the foot are disrupted by abnormal activity. The goal of brain surgery is to free up the pathways so that the brain and body may accomplish the intended function—in this case, moving the foot.

Brain surgery may be performed unilaterally (on one side of the brain) or bilaterally (on both sides). The effects of surgery occur on the side of the body opposite to the surgical site.

To date, most persons who have undergone brain surgery for dystonia were treated for generalized dystonia. However, individuals who may be eligible for brain surgery include persons with focal, segmental, or generalized dystonia with significant, disabling symptoms that do not respond satisfactorily to other therapies. Adults as well as children with primary and secondary dystonia may be eligible.

Based on the limited available data, different categories of patients may respond differently to brain surgery. Although cases of both secondary dystonias (including tardive dystonia) and focal dystonias may be eligible, it appears as though persons with DYT-1 generalized dystonia are the best candidates for brain surgery—either lesioning or DBS. Studies have shown as much as 60-90% improvement in DYT-1 patients treated with lesioning or DBS. Patients with secondary hemidystonia may be eligible for brain surgery, though they may not benefit as much as those with DYT-1 dystonia. Researchers are examining the possibility that persons with secondary dystonia may get greater benefit from lesioning or DBS to the thalamus rather than the globus pallidus.

There is limited data about the long-term effects of each approach. Brain surgery for dystonia is an evolving science, and investigators are continually collecting information.

Lesioning Procedures: Pallidotomy & Thalamotomy

The practice of lesioning parts of the brain in dystonia patients was very common in the 1950s and 1960s, since at that time it was essentially the only available treatment for severe cases. These procedures, as practiced 50 years ago, had mixed results. In some cases the improvement was spectacular; in other cases complications developed; and in still other cases repeated procedures were necessary. By the 1980s, brain surgery for dystonia had fallen out of favor and was not widely practiced. However, the increased understanding of the basis of movement disorders such as Parkinson's disease and the success in treating it with surgical approaches, plus the development of brain imaging technology, led to a re-evaluation of surgery as an option for patients with dystonia.

The procedure that involves creating a destructive lesion in the globus pallidus is called *pallidotomy*, and the procedure that involves creating a lesion in the thalamus is called *thalamotomy*. A permanent lesion is made in the brain tissue by heating the tip of an electrode and coagulating the intended tissue.

When lesioning surgery is chosen, pallidotomy is now preferred over thalamotomy and provides a reasonable alternative to pallidal DBS for patients who are averse to the cosmetic appearance of the implanted pulse generator or do not want to be burdened by repeated battery replacements. Bilateral pallidotomy has shown an average of 67-80% improvement in the Burke-Fahn-Marsden dystonia rating scale in patients with generalized dystonia. Primary generalized patients respond better than focal or secondary dystonias. In Parkinson's patients, bilateral pallidotomies are avoided because they cause hypophonia, a quieting of speech. This has not been observed in dystonia patients, and many dystonia patients have had bilateral pallidotomies without significant worsening of speech.

Although thalamotomy was once the most common brain surgery performed for dystonia, it is now used almost exclusively in cases of stable hemidystonia, and a very specific site in the thalamus is targeted. The

procedure is performed unilaterally. Bilateral brain surgeries increase the risk of complications, and bilateral thalamotomies in particular are known to often cause speech impairment.

The primary factor that distinguishes modern lesioning procedures from those of 50 years ago is that surgeons are able to locate the lesioning target more accurately. The following factors make it much easier for the surgical team to locate the target within the brain, which is crucial to reducing the risk of complications:

- *Stereotactics*—Surgeons are able to target the precise area of the brain with a computerized, 3-dimensional scale using MRI and CAT scans.
- *Microelectrode recording and brain mapping*—The surgical team has the ability to listen to the sounds of brain cells firing messages to one another. Cells in different parts of the brain fire at very specific rates and in characteristic patterns, and by listening to these cells the surgeon knows exactly where the electrode is within the brain. Several recording tracts may be necessary to identify the precise target.

Once a physician has recommended brain surgery, and pre-operative screening tests and preparations are complete, the basic plan of operation for pallidotomy and thalamotomy are the same. The individual is fitted with a head frame under general or local anesthesia. The brain is mapped with imaging technology to create a blueprint for planning and measuring the placement of the electrode. Under local anesthesia, the electrode is inserted through a small hole in the skull into the brain. The brain itself does not feel pain, and the patient is awake during most of the procedure. The surgical team interacts with the patient throughout the procedure, and the patient provides feedback about symptoms and how he/she feels. Microelectrode recording is used to confirm the target. The mapping procedure alone may take up to several hours. Once the target is defined, the surgeon inserts the thermal electrode and creates a lesion. The thermal electrode is removed and the procedure is complete. A bilateral

procedure may be done in a single surgery or in two separate surgeries. If a second target is to be lesioned, the mapping procedure is repeated for that specific target. Most patients are in the hospital for two or three days. Medications may be temporarily resumed, and after a short time the patient returns to the neurologist for a follow-up exam.

There is a small but real risk of complications associated with lesioning. The most serious risk is a 1-2% incidence of stroke or hemorrhage during the mapping phase of the surgery. Also, the target of the pallidotomy, the internal segment of the globus pallidus, is located right above the optic tract which may be damaged if the electrode is not targeted precisely. There also exists the risk that the pallidotomy will not improve the symptoms. However, the procedure has been shown to dramatically improve dystonia in some patients.

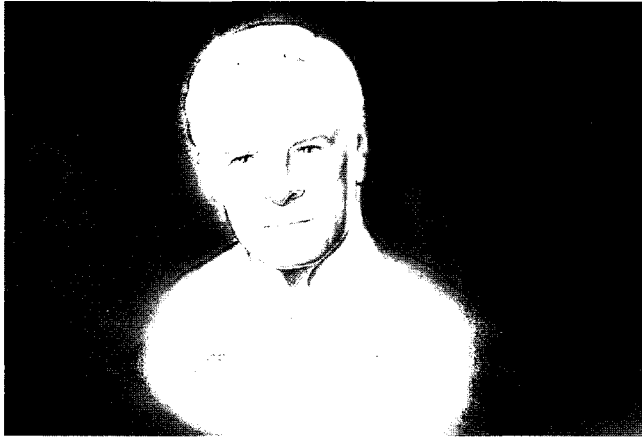
Deep Brain Stimulation

Deep brain stimulation (DBS) involves implanting stimulating electrodes into selected targets in the brain in order to mimic the effects of lesioning. Surgeons began using DBS in place of lesioning for Parkinson's disease patients in the mid-1990s. DBS also has applications to tremor and pain. Whereas DBS has been used to treat thousands of persons with Parkinson's disease, the procedure began being applied to dystonia only in the late 1990s. The results of more than 200 dystonia patients have been published as case studies in medical journals.

Bilateral pallidal DBS produces significant benefit in dystonia with average improvements of about 50-60% in the Burke-Fahn-Marsden dystonia rating scale. Some primary generalized patients have been reported to have up to 90% improvement. DBS has also been performed on persons with secondary dystonias, cervical dystonia, segmental dystonia, and myoclonic dystonia with encouraging results.

The complete DBS apparatus includes the DBS electrode, a connecting wire, and a pulse generator (a.k.a. "brain pacemaker" or stimulator) that contains a battery. The initial procedure to implant DBS is identical to that

Figure 3



Deep brain stimulation hardware in body.

of the pallidotomy and thalamotomy. Once the brain target is mapped and identified, instead of creating a lesion, the surgeon places the DBS electrode into the target. The wire and pulse generator may be implanted at the same time as the electrode or at a later date. The generator is implanted under the collarbone or in the abdomen. The wire is then tunneled up the neck, behind the ear, and to the site of the electrode (the patient is under general anesthesia for this part of the procedure). The wire is connected to the electrode, and the incisions are closed. Most DBS procedures involve the implantation of two generators and are done in two surgeries. It is possible to implant both generators in a single surgery, but this is a very demanding approach for the patient. Immediately after the operation, the patient may temporarily resume medications. The patient may be discharged the next day.

Once the generator is implanted, the patient must wait a week or two before the batteries are activated. This waiting period is necessary to allow the swelling that normally occurs with the surgery to diminish. The DBS electrode conveys electrical pulses into the brain using power produced by the battery in the generator. A series of visits to the hospital are required to adjust the voltage settings to the needs of the individual. It may take several weeks or months to achieve the correct settings. The

patient can check the status of the generator using a handheld device that resembles a TV remote control. Using this device, the patient can determine if the generator is on or off, and can turn it back on in the event that it shuts down unexpectedly. (Certain phenomenon such as magnetic fields caused by security devices may cause the battery to temporarily stop working.)

The expected life span of a battery at a typical voltage is about 3-5 years. At a very high voltage, the battery may need to be replaced after a year; at a very low voltage, perhaps up to seven years. Replacing a battery can be done under general or local anesthesia as an outpatient procedure.

Dystonia does not respond to DBS in the same manner as other movement disorders do. For example, persons treated for tremor will generally improve within seconds of turning the generator on. In patients with dystonia, improvement may be delayed for days, and weeks or months may pass before the full extent of the benefit is reached. DBS does not necessarily eliminate the possibility of subsequent drug or botulinum toxin treatments.

Side effects are minimal, but no procedure is without risks. The main risk in DBS is a fatal hemorrhage. However 99-99.5% of patients do not have significant bleeding. Despite vigorous efforts to avoid it, infection is a risk in approximately 2% of patients. Infection can be serious and warrant the removal of the hardware. If this happens, it may be possible to re-implant the hardware once the infection is treated. Hardware failure is also a concern, though this is rare and precautions are in place in the event of situations such as a battery failing. It is estimated that in 5% of DBS procedures for dystonia some complication may arise, most of which can be addressed without removing the hardware.

Although no longer considered “investigational” for dystonia by the Food & Drug Administration, DBS is in its infancy as a treatment for this disorder. The preliminary results are quite positive, and the procedure is expected to evolve over time as more patients are treated and more data is collected.

Comparing Lesioning & DBS

Studies have shown that both lesioning and DBS can dramatically improve dystonia. Both approaches are associated with a small, but real, risk of complications. There has not been a clinical study to compare the results of lesioning procedures and DBS, and the advantages and disadvantages of each remain an open issue.

Lesioning procedures and DBS have many elements in common including:

- Patient selection criteria
- Area of brain targeted
- Basic surgical procedure
- Potential for profound benefit to eligible patients
- Risk of complications including hemorrhage during surgery, hemiplegia or hemiparesis, sensory impairments, speech/language impairment

In both cases, the chance of benefit must be weighed against the risk of complications. No two cases of dystonia are alike, and determining the specific approach to treatment—in this case lesioning or DBS—must be decided after careful discussions among the patient, family members, neurologist, and neurosurgeon.

Of the dystonia patients who are eligible for brain surgery, more individuals are currently being recommended for DBS than pallidotomy. The pallidotomy, however, is by no means an obsolete procedure. Unless a patient is against having hardware installed in his/her body, the tendency is to try DBS before proceeding to the pallidotomy because DBS is adjustable and reversible.

Financial and geographical issues cannot be overlooked. Persons who have DBS must visit the doctor regularly for maintenance check-ups. People who live in remote areas or areas not in proximity to a major movement disorder center may be at a disadvantage. Travel to and from the center—and the expense of this travel—is a part of the ongoing management required of DBS patients.

Because lesioning creates a permanent change in the brain tissue, there is a slightly higher risk of permanent complications during the surgery such as swallowing difficulty, speech difficulty, and cerebral hemorrhage. Because DBS involves the implantation of hardware, complications associated with the apparatus are possible, including infection, erosion through the skin, hardware breakage, and stimulator failure. The risk of hardware complications exists for as long as the hardware is implanted.

It remains to be seen whether the pallidotomy or DBS is more effective than the other. The experience of the surgeon and medical team are the most important determinants of success and risk. The lowest incidence of complications occurs in major medical centers that perform these procedures often. Patients should choose a center with a long-standing expertise in movement disorders and a clinical team devoted to surgery for dystonia and movement disorders. A movement disorder neurologist and a surgeon should be specially trained in functional surgery, and an electrophysiologist should be on staff for brain mapping. An experienced nursing staff is also important.

Patients of both categories of brain surgery may benefit from physical therapy and supportive therapy following the procedure.

Comparison Table	
Lesioning	DBS
Permanent	Reversible
Controlled destruction of brain tissue	Non-destructive
Not adjustable	Adjustable
No hardware	Implanted hardware
Little post-op maintenance	Extensive, ongoing maintenance
Few post-op restrictions	Common-sense restrictions regarding activity; must avoid magnetic fields and diathermy
No cosmetic issues	Hardware may be slightly visible beneath skin in some people

Children & Brain Surgery

Children over the age of seven are eligible for lesioning and DBS, although the longer one waits, the less brain and skin growth will occur after the operation. However, there is little data available about long-term effects of DBS and how a child's development may affect the hardware. Steps can be taken during surgery to ensure that the apparatus can accommodate the child's growth. Children and adolescents may be at a slightly higher risk of complications from DBS because general rather than local anesthesia is used during implantation and post-operatively children are more likely to engage in rough play that may affect the hardware.

Surgery does not necessarily have to be considered only as a last resort. Certainly, if an individual is satisfied with how symptoms respond to a less invasive treatment such as botulinum toxin or medications, there is no need to consider brain surgery. However, especially in children, early intervention may significantly improve quality of life. The benefits of brain surgery include more than improved mobility—a child's ability to function comfortably at school (both academically and socially), to make friends, and to be active are important factors to consider. In both children and adults, brain surgery can drastically improve pain, which is often a major component to a person's quality of life.

IV. CONCLUSION

Having surgery is a very significant step for an individual to take in the treatment of dystonia. If you are considering surgery or if surgery has been recommended to you by a movement disorder specialist, the following questions may help you initiate discussions with your doctors:

- What is the name of the operation and what does the name mean?
- Why is this specific surgery appropriate for my case?
- What are the advantages of having surgery?
- What benefits might I expect?
- What are the risks?
- What happens if I don't have the surgery? Are there alternative treatments?
- Where can I get a second opinion?
- What is the experience of the medical center and surgeon with this procedure?
- Does the medical team publish the results of surgical case studies?
- Where will the surgery be done?
- What kind of anesthetic will be used (general or local)?
- How long is the recovery and what rehabilitation is necessary?
- How much will the surgery cost and who will pay for it?

Surgical procedures may improve function and better the lives of patients who do not receive adequate relief from medications and/or botulinum toxin injections. A patient who is considering surgery must weigh the opportunity for benefit and the risk of complications. Careful discussions with movement disorder specialists and being as knowledgeable as possible about dystonia and surgery may aid in the consideration process. The Dystonia Medical Research Foundation can provide names and contact information of dystonia specialists.

Until a cure for dystonia is achieved, researchers are working diligently toward developing treatment options to improve the lives of affected individuals. Surgery is an area of research in which vast progress is being made and in which the prospect of developing more effective treatments is tremendously promising.

V. APPENDIX

Dystonia

Dystonia is a neurological movement disorder that causes muscles in the body to contract or spasm involuntarily. The involuntary muscle contractions cause twisting, repetitive, and patterned movements as well as abnormal postures.

Dystonia is not a single disease but a syndrome—a set of symptoms that cannot be attributed to a single cause but share common elements. Some forms of dystonia may affect a specific body area, such as the neck, face, jaw, eyes, limbs, or vocal cords. When dystonia affects a single body area, it is called *focal* dystonia. Focal dystonias include cervical dystonia, blepharospasm, oromandibular dystonia, writer’s cramp, and laryngeal dystonia (spasmodic dysphonia). *Segmental* dystonia affects two or more adjacent body areas. If two or more non-adjacent body areas are affected, the dystonia is termed *multifocal*. *Generalized* dystonia refers to dystonia that may affect the limbs, trunk, and other major body areas simultaneously. When dystonia only affects muscles on one side of the body, it is called *hemidystonia*. Although the outward appearances of the various forms of dystonia may appear very different, they all share the element of repetitive, patterned, and often twisting involuntary muscle movements.

Dystonia affects men, women, and children of all ages and backgrounds. Dystonia may be genetic or caused by factors such as physical trauma, exposure to certain medications, or other neurological conditions.

Dystonia is the third most common movement disorder after Parkinson’s disease and tremor, affecting an estimated 250,000 persons in North America. Nonetheless, dystonia is often misunderstood by the public and misdiagnosed by medical doctors. Dystonia is neither a psychological disorder, nor does it affect intellect. Dystonia is not fatal, but it is a chronic disorder that causes varying degrees of disability and pain, from mild to severe.

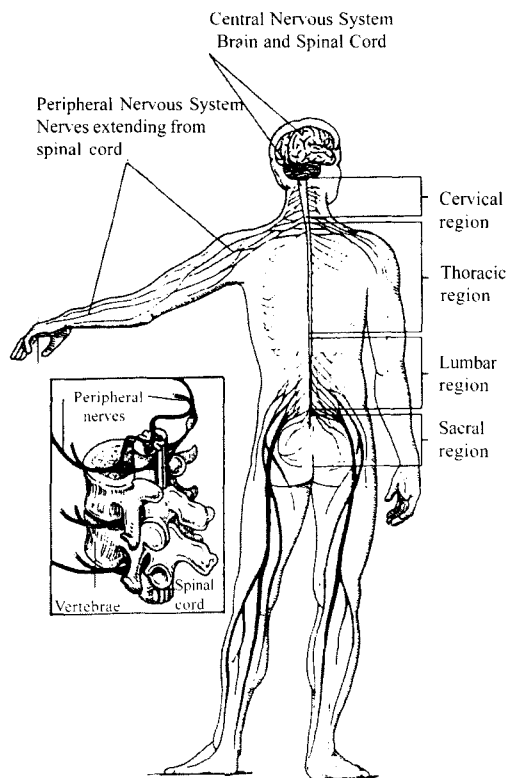
Figure 4

The nervous system is divided into two parts: the central nervous system (CNS) and the peripheral nervous system (PNS). The CNS consists of the brain and the spinal cord. The PNS consists of the nerves extending from the spinal cord.

These two systems are responsible for all bodily activities, ranging from heart rate and muscle movement to emotions and learning.

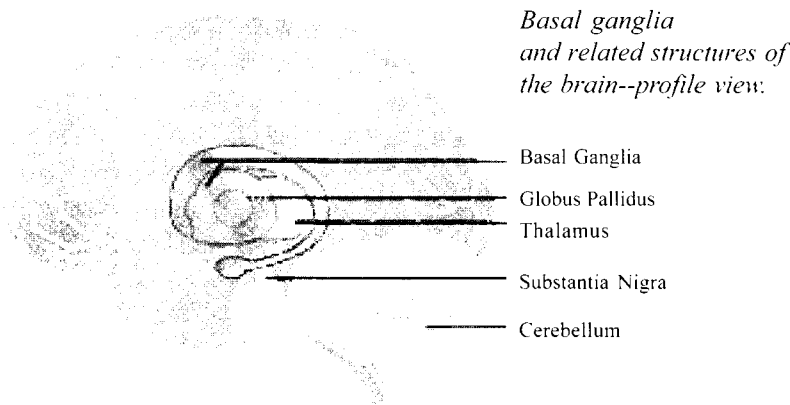
The brain is the most complex and intricate organ in the human body. The wrinkled gray mass that makes up 80% of the brain is called the *cerebral cortex*. This part is responsible for activities such as thinking, perceiving, and producing and understanding language. The cerebral cortex is divided into two sides or “hemispheres”—the right and the left. Although both hemispheres appear identical, they differ in purpose and function. Language, reasoning, and logic capabilities originate in the left side of the brain whereas appreciation of shapes and textures and artistic talents originate in the right side.

The cerebral cortex is further divided into four sections or “lobes”—frontal, parietal, temporal, and occipital. The frontal lobe is involved in movement



Anatomy of the nervous system.

Figure 5



and decision-making skills; the parietal lobe interprets touch, pain, and temperature; the temporal lobe is involved in hearing and memory; and the occipital lobe contains the vision center.

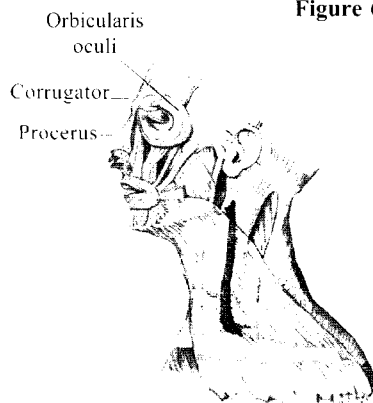
Beneath the cerebral cortex lie structures that help us move, sleep, wake, breathe, smell, hear, see, taste, and eat. Dystonia affects an area in this deep part of the brain believed to regulate movement called the basal ganglia. An imbalance of brain chemistry causes the basal ganglia to send inappropriate messages to the muscles, causing them to contract and spasm involuntarily.

Anatomy

Eyes

The orbicularis oculi muscle encircles the opening of the eye socket and acts to close the eyelids. The corrugator muscle draws the eyebrows together and wrinkles the brow. The procerus muscle is a facial muscle between the eyebrows and down the nose.

Figure 6



Eye muscles in profile.

Neck

The sternocleidomastoid and trapezius muscles are major muscles in the neck. The two sternocleidomastoid muscles are thick muscles on each side of the neck that act to bend, rotate, flex, and extend the head. The trapezius muscle moves the shoulder blades upward in a shrug.

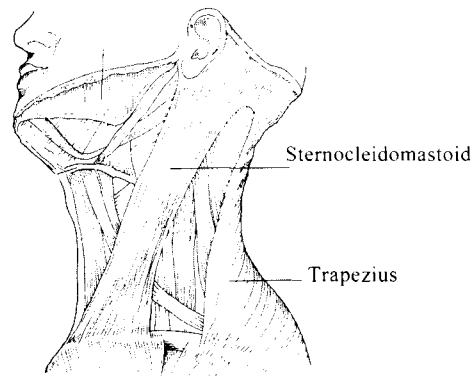


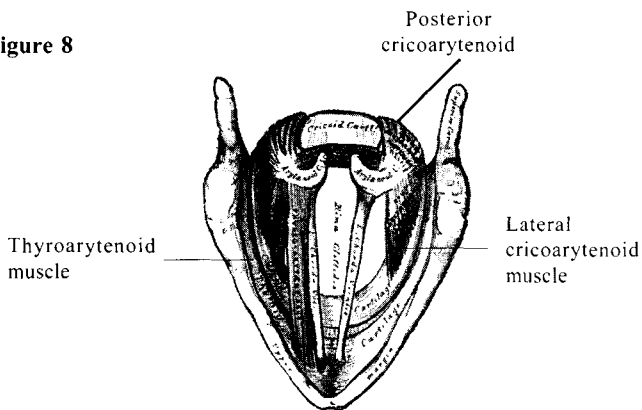
Figure 7

Anatomy of the neck.

Larynx

The larynx or “voice box” is an organ in the neck that plays a crucial role in speaking and breathing. The framework of the larynx is made up of the thyroid cartilage. The front portion of the thyroid cartilage is visible in some people as the “Adam’s apple.” The vocal cords are located in the center of the larynx. The thyroarytenoid muscle is responsible for closing the vocal cords, and the posterior cricothyroid muscle is responsible for opening the vocal cords.

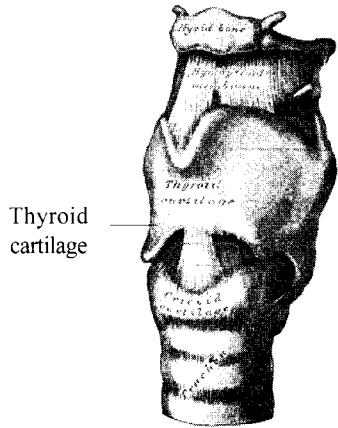
Figure 8



Inside larynx, view from above.

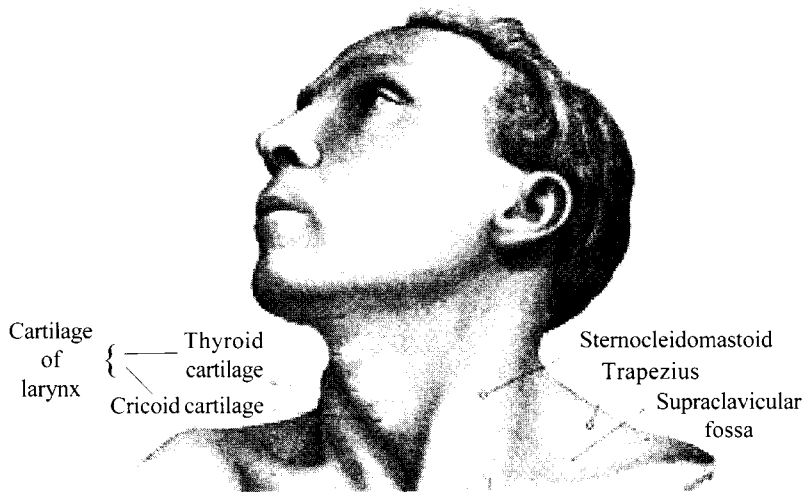
Larynx (*continued*)

Figure 9



Outer cartilage of larynx, angled view from front.

Figure 10



Surface of the neck.

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