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Treatment Options for Vestibular Schwannoma



Colin L. W. Driscoll, MD, and
Michael J. Link, MD

Acoustic neuromas, more appropriately called vestibular schwannomas, are benign tumors that arise from overproduction of perineural Schwann cells. They account for 6% to 8% of all primary intracranial tumors. Within the cerebellopontine angle they represent 80% of tumors. The incidence of vestibular schwannomas is hard to estimate, but it is probably somewhere between 10 and 15 per 1 million population.

The tumors typically present in patients between 40 and 50 years old. But they can occur in children and occasionally in patients in their 80s. Irrespective of the age of the patient, common symptoms include unilateral hearing loss, tinnitus, and impairment or loss of balance. Hearing loss that is bilateral and symmetric is unlikely to be caused by a vestibular schwannoma.

Says otorhinolaryngologist Colin L. W. Driscoll, MD: "Irrespective of the age of the patient, the most common presenting symptom is gradual or sudden-onset unilateral hearing loss and tinnitus. More rarely, patients may experience vertigo or mild imbalance." Adds his colleague, neurosurgeon Michael J. Link, MD: "If a vestibular schwannoma becomes large enough, it can produce facial numbness, severe ataxia, and even dysphagia by impinging on the nerves that mediate those functions."

Diagnosis and Treatment Options

The best modality available to confirm the diagnosis of vestibular schwannoma is MRI of the head, with and without contrast. The tumor is almost always seen as a brightly enhancing lesion in the cerebellopontine angle that enlarges and extends to the internal auditory canal.

The selection of treatment is highly individualized—and sometimes subject to controversy. The first option is simply to observe and note the patient's progress with a follow-up MRI and a hearing test in 6 months. This is particularly true for patients older than 65 years who have small tumors.

For patients younger than 65 years who have small tumors and good hearing, there are 2 main options. First is stereotactic radiosurgery, a 1-day, outpatient, focused radiation treatment. This treatment does not destroy or remove the tumor. Rather, it stops or slows its growth. At Mayo Clinic, more than 300 patients have been treated for vestibular schwannoma using Gamma Knife stereotactic radiosurgery. The overall results are very good, with an expected greater than 95% chance of long-term tumor control. The risk of facial weakness is less than 1%.

Second is an open surgical approach (Figure 1). Mayo Clinic neurologic surgeons operate on all sizes of tumors, ranging from a few millimeters to greater than 6 cm. If the tumor measures less than 2 cm and the patient has useful hearing in the affected ear, an approach that preserves hearing is attempted—either a retrosigmoid approach or a middle fossa approach. The goal of

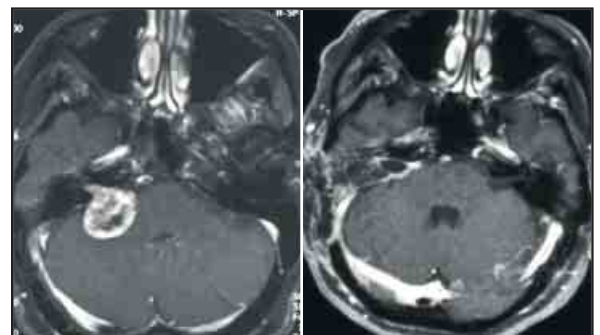


Figure 1. Left, Preoperative axial T1-weighted image with gadolinium shows a 2.5-cm right vestibular schwannoma. Right, Postoperative axial T1-weighted image with gadolinium 2 years after tumor resection shows complete tumor removal with no evidence of recurrence.

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the surgery is to remove all the tumor, thus curing the patient and preserving useful hearing without adding new neurologic deficits.

Depending on the size and configuration of the tumor, hearing preservation ranges between 10% and 50%. The risk of facial nerve weakness varies from less than 1% in patients with very small tumors to 20% to 25% in patients with very large tumors. The risk of other surgical complications is less than 1%, and the typical hospital stay is 3 to 5 days.

If the patient does not have useful hearing, a translabyrinthine approach may be recommended. This involves operating through the inner ear, and subsequently no hearing is preserved in that ear. This approach has some benefit in that it exposes the internal auditory canal well, which tends to make this portion of removal of the tumor easier.

There is a third open surgery scenario as well: subtotal resection (Figure 2). Says Dr Link: "In some cases we may elect to do subtotal resection on a very large tumor. We may elect to leave a small portion of the tumor attached to the facial nerve to avoid inducing new facial weakness after surgery. Then, once we have reduced the tumor to an acceptable size, we treat the remnant with Gamma

Knife radiosurgery if it shows growth on future imaging."

A neurosurgical and otorhinolaryngologic team performs all operations for vestibular schwannomas at Mayo Clinic. These specialists have extensive experience treating tumors of this type and a good collaborative approach to treating these patients. The alliance of advanced specialties working together—neurosurgeons, otorhinolaryngologists, radiation oncologists—is a hallmark of the Mayo Clinic approach to the treatment of vestibular schwannomas. Says Dr Link: "Patients benefit tremendously from preoperative consultation by all members of the team so they have a thorough understanding of the treatment options from the very beginning."

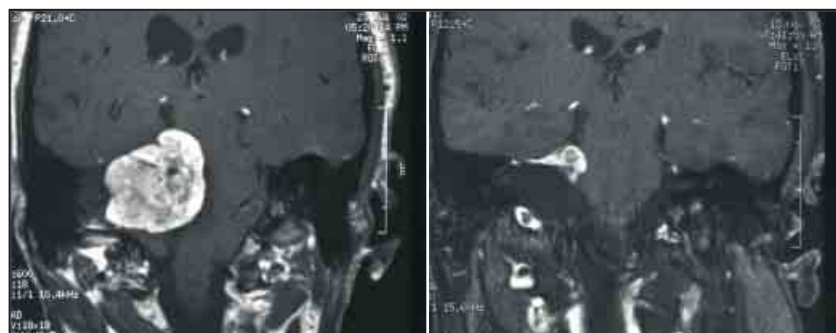


Figure 2. Left, Preoperative coronal T1-weighted image with gadolinium shows a very large, 4.5-cm right vestibular schwannoma with severe compression of the brain stem and cerebellum and moderate obstructive hydrocephalus. Right, Coronal T1-weighted image with gadolinium 3 months postoperatively shows a small tumor remnant along the course of the right facial nerve. Hydrocephalus has resolved, and the patient has normal facial nerve function. The tumor remnant is now a size that could be treated effectively with radiosurgery if it shows signs of growth.

Surgical Management of Brain Tumors That Cause Epilepsy

Brain tumors are among the possible causes of epilepsy, defined as chronic and recurring seizure activity, in both adults and children. Various types of brain tumors can be involved in seizure generation. Common epilepsy-related tumors arise from glial cells and include low-grade gliomas such as pilocytic astrocytoma, ganglioglioma, dysembryoplastic epithelial tumors, and oligodendrogliomas. These tumors are generally slow growing, tend to be benign and well defined, and are successfully removed by

surgery. An estimated 70% of tumors that cause epilepsy are slow-growing gliomas.

Favorable surgical outcomes depend on the completeness of resection of both the lesion and the epileptogenic zone involved in seizure initiation. Complete resection requires a highly skilled, experienced, and integrated surgical neuroscience team accustomed to working together throughout the continuum of care. Individualized care—from presurgical evaluation to planning and executing a specific surgical strategy unique to each

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patient through postoperative care and follow-up—is the hallmark of the Mayo Clinic Epilepsy Center neurosurgical team.

Because of advances in neuroimaging, epilepsy-associated brain tumors are more easily detected. However, these may not be visualized by CT scan. MRI is indispensable for visualizing exact anatomic details of the tumor. Says W. Richard Marsh, MD, Mayo Clinic neurosurgeon: “Every patient with a chronic seizure disorder deserves to have an MRI scan. If a brain tumor can be identified as a cause of the seizure disorder, good surgical treatment exists.”

Adds neuropsychologist Max R. Trenerry, PhD: “In addition to the anatomic detail that MRI can provide, neuropsychological evaluation helps patients understand the nature and extent of cognitive changes associated with the tumor and seizure disorder.”

Evaluating the Epileptic Patient

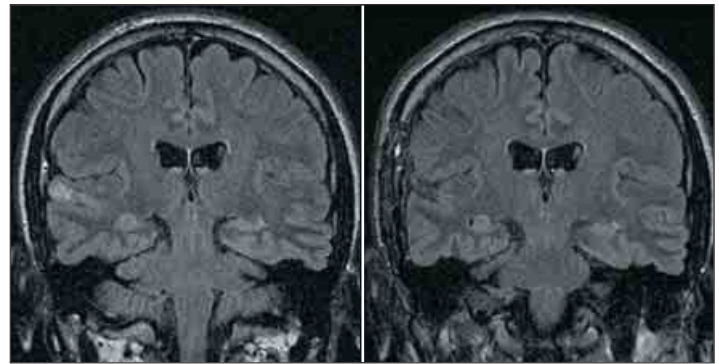
At Mayo Clinic, a patient seeking care for brain-tumor-caused epilepsy sees at least 3 specialists from the neurosciences team. Consultation with a neurologist includes careful history taking and a thorough examination. A neurosurgeon discusses details of the various surgical approaches, including recovery, risks, and benefits. Consultation with a neuropsychologist and detailed neuropsychological testing assess the potential effects of surgery on cognitive abilities, including intelligence, memory, attention, and reasoning.

Preoperative testing includes MRI as well as prolonged EEG recordings to co-localize the patient’s seizure onset with identified tumors. Occasionally, patients need intracranial monitoring to localize the extent of the epileptogenic zone around the area of the identified tumor. This monitoring can include the insertion of either deep brain or surface electrodes over the portion of normal brain in the region of tumor.

Surgical Options

If the tumor is near eloquent areas of brain—such as primary language areas or primary motor or sensory areas—it is usually performed with the patient sedated but awake to minimize the risk of neurologic deficit.

If tumors are located in tissue outside



MRI scans of left temporal lobe preoperatively (left) and postoperatively (right) after successful removal of tumor.

eloquent areas, surgery proceeds with the patient under general anesthesia.

Precision during the surgery is assured by computer-assisted monitoring. This technology provides information about the volume, shape, and configuration of the tumor and is used to identify the margins of the tumor, which may not be apparent under direct visualization. With this understanding of the physical space within the patient’s brain in which the tumor exists, surgeons can excise it successfully and thoroughly—thus increasing the likelihood that seizures will cease once the tumor and the epileptogenic zone are removed. In the future, intraoperative MRI will be used to confirm complete tumor removal.

Results

The usual hospital stay after tumor removal is 4 days. Most patients can resume full activity within a few months. Some patients are able to stop taking their antiepileptic medications within the first several years after surgery. About 70% of epileptic patients with tumors and seizures become seizure-free as a result of brain surgery.

Complications and Risks

Complications and risks are unique to each patient. They depend on the area of brain involved and the exact type of tumor and will be discussed in detail with the patient by the neuroscience team.

When to Refer

If a patient has an abnormal MRI showing a benign tumor, he or she should be referred as early as possible. This is especially true for pediatric patients. Says Dr Marsh: “With a child who is in a learning-growth-development phase, seizures adversely affect development. If there is an identifiable, curable cause, treatment is best undertaken sooner to get the child back on an ascending track.”



W. Richard Marsh MD, and
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Interdisciplinary Brachial Plexus Clinic Uses Novel Approaches to Attempt to Restore Hand Function

The brachial plexus is a complex network of interconnecting nerves that innervate the arm from the shoulder to the hand. The C5 through C8 and T1 spinal nerve roots form the basis of the brachial plexus.

Injury and lesions to the brachial plexus are fairly common and arise from a variety of causes. Traumatic lesions are typically caused by high-speed motor vehicle accidents, such as those involving motorcycles and snowmobiles. Perinatal cases occur in 1 in 2,000 births and are often related to shoulder dystocia. Other causes include tumors, irradiation, and nerve entrapment.

In traumatic lesions, critical motor function in the shoulder, elbow, wrist, and hand may be lost, and sensation in the fingers impaired. Incapacitating pain may also result from stretching or rupture of the nerves of the brachial plexus or avulsion of the nerves from the spinal cord. Lesions in the upper trunk (C5 and C6) result in the loss of shoulder and elbow flexion, whereas injuries to the lower trunk (C8 and T1) impair hand function. Injuries to the complete brachial plexus paralyze the entire upper limb.

Whatever the cause, these injuries can inflict severe disability in the shoulder, elbow, and hand. "In addition to the physical problems, patients may also have considerable psychological distress and economic hardship," says neurosurgeon Robert J. Spinner, MD. "The good news is that recent advances in the diagnosis and operative management and the depth of our multidisciplinary Brachial Plexus Clinic neurosciences team all work together to improve function and ease the patient's psychological distress."

Adds neurologist C. Michel Harper, MD: "Communication and coordination of patient care

across medical and surgical specialties clearly enhance the care, outcome, and satisfaction of our Brachial Plexus Clinic patients."

Mayo Clinic's Interdisciplinary Brachial Plexus Clinic

The interdisciplinary Brachial Plexus Clinic at Mayo Clinic in Rochester addresses the complex problems of patients who have brachial plexus lesions. Specialists from several areas—neurosurgery, orthopedic surgery, neurology, radiology, physical medicine and rehabilitation, physical and occupational therapy—work cooperatively to design a treatment and recovery plan that is unique to each patient. It is the mission of the Brachial Plexus Clinic to help patients recover as much pain-free function and quality of life as possible.

The team of surgeons that evaluates each patient also operates together. Having performed more than 100 procedures in 2003 alone (including 25 contralateral C7 transfers and 30 free-functioning muscle transfers in the past 2 years), this team has extensive experience and expertise to provide state-of-the-art care for patients with brachial plexus injuries.

Says Dr Harper: "The multidisciplinary approach is absolutely essential when evaluating and treating patients with complicated brachial plexopathies. Each patient is evaluated by a neurologist with a special interest in peripheral nerve disorders. The same team of neurologists performs all necessary electrodiagnostic studies—both preoperatively and during surgery—to help the surgical team make critical decisions regarding localization, prognosis, and treatment options for the patient."

Treatment Options

The diagnosis of brachial plexus injury can be established soon after injury. Evidence of complete root avulsion indicates early surgical intervention is necessary, and the sooner it is undertaken, the better the outcome. Waiting longer than 6 months for surgery is not advisable because the cumulative effects over time of muscle atrophy, motor end plate degeneration, and neuronal death contribute to inferior outcomes.

Surgery

Surgical options include neurolysis for neuroma in continuity, nerve repair for lacerations, and

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nerve grafting to bridge gaps that result from traumatic ruptures, tumor excision, or severe stretch lesions (which do not conduct impulses across the lesion). Nerve regeneration is slow, however, occurring at a rate of approximately 1 inch per month.

Nerve Transfers

When direct nerve grafting cannot be performed or is less likely to provide a satisfactory result, transfer of “expendable” uninjured nerves allows the rapid recovery of key muscles. Nerves can be moved from an uninjured portion of the brachial plexus or from a number of other sites to be used for recovery of both motor and sensory functions.

Surgeons may direct nerve fibers to a specific motor or sensory “target,” improving chances for recovery of function.

Potential nerve transfers include the spinal accessory nerve, intercostal motor and sensory nerves, and the phrenic nerve. In addition, the Mayo Clinic team also uses the contralateral C7 transfer from the opposite (uninjured) side (Figure 1). Transferring a part of the uninjured C7 nerve root, usually combined with a vascularized nerve graft, provides the possibility of restoring grasp function in patients with total plexus avulsion.

In a patient with an upper trunk injury, other innovative techniques that are done closer to the

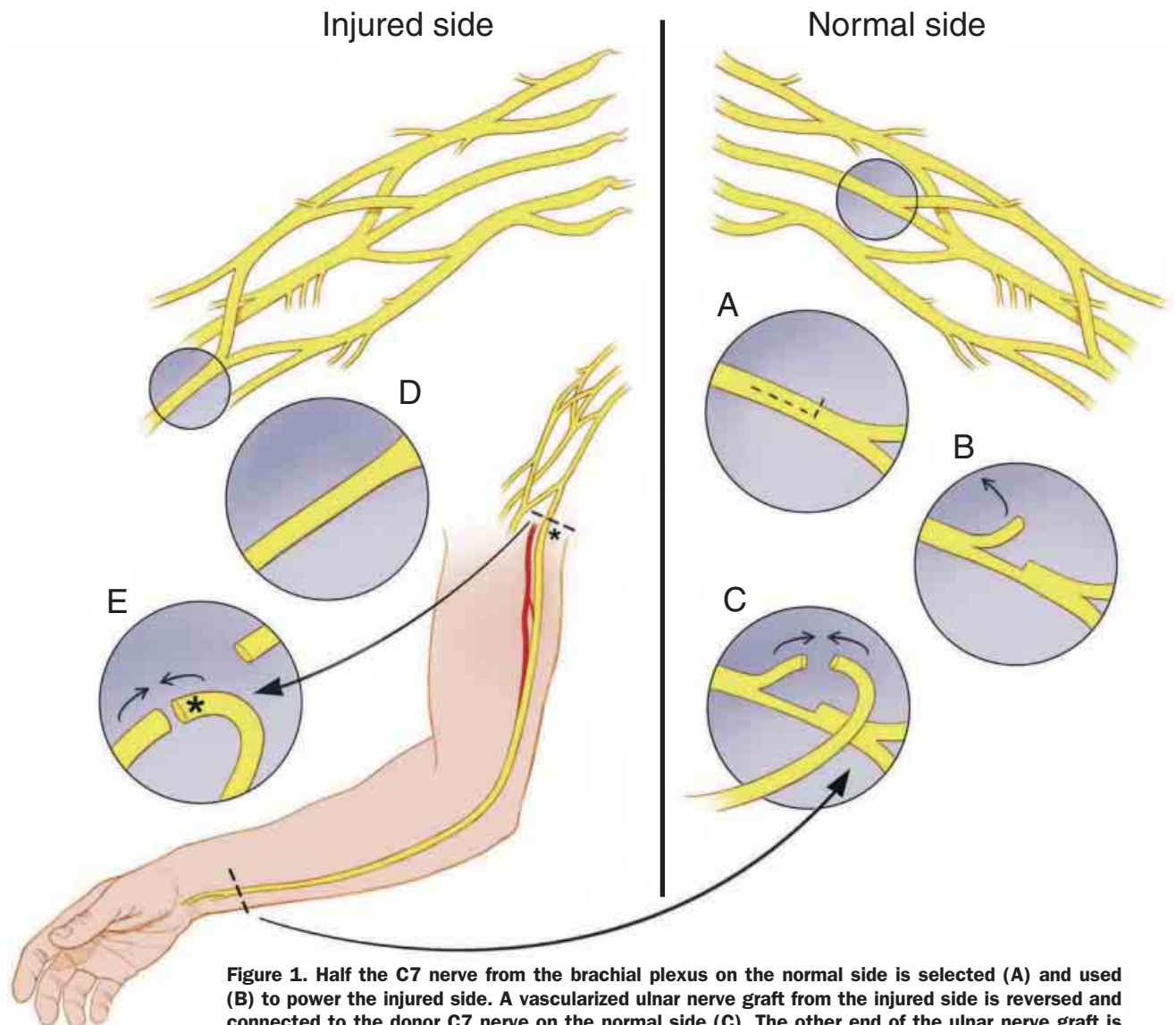


Figure 1. Half the C7 nerve from the brachial plexus on the normal side is selected (A) and used (B) to power the injured side. A vascularized ulnar nerve graft from the injured side is reversed and connected to the donor C7 nerve on the normal side (C). The other end of the ulnar nerve graft is then connected to the median nerve on the injured side (D and E). This complex technique potentially allows patients with severe brachial plexus injuries to regain sensation in the hand and movement in the fingers on the injured side.

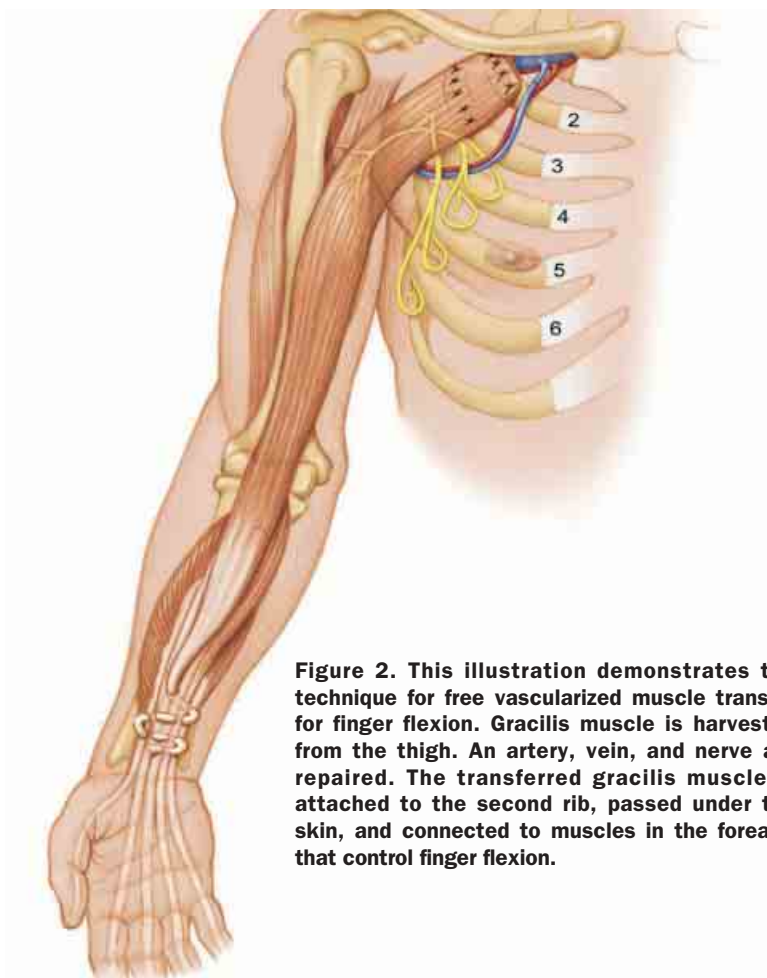


Figure 2. This illustration demonstrates the technique for free vascularized muscle transfer for finger flexion. Gracilis muscle is harvested from the thigh. An artery, vein, and nerve are repaired. The transferred gracilis muscle is attached to the second rib, passed under the skin, and connected to muscles in the forearm that control finger flexion.

target muscle may be used to improve function. For example, a fascicle of the ulnar nerve or the median nerve may be used to reinnervate the biceps muscle; in addition, one of the nerve branches supplying (a part of) the triceps may be used to reinnervate the deltoid muscle.

Muscle Transfers

Another advanced method is the microsurgical transfer of a healthy muscle. With circulation restored and nerve repairs performed in the arm, the muscle transfer can provide needed motor function when delay in or previous unsuccessful treatment has resulted in irreversible muscle atrophy in the arm or when improvement of hand function is desired.

To do this, the surgical team transfers an expendable muscle, such as the gracilis muscle from the thigh, along with its nerve and blood supply, to animate the elbow, wrist, and hand (Figure 2). Collectively, these methods often restore shoulder stability, limited but useful shoulder abduction, full elbow flexion, and, in some patients, hand function and protective sensation.

For information about the Brachial Plexus Clinic or to refer patients for evaluation, contact 507-538-1988.

Carotid Angioplasty With Stent Placement

Carotid angioplasty with stent (CAS) placement is an emerging alternative to carotid endarterectomy for the treatment of patients with carotid artery occlusive disease. Mayo Clinic neuroradiologists began using it in 1996 for patients at high risk for surgery.

“The Cerebrovascular Clinic in the Department of Neurology has a multidisciplinary CAS placement protocol in which a vascular neurologist, an interventionalist, and a surgeon—either a vascular surgeon or a neurosurgeon—meet with the patient to help clarify the best treatment approach. Cardiology colleagues may also be involved if the patient has cardiac symptoms. This is not uncommon, since so many patients with carotid occlusive disease also have coronary artery occlusive disease,”

says neurologist Robert D. Brown, Jr, MD. Adds Harry J. Cloft, MD, PhD, neuroradiologist: “The protocol has been highly successful in allowing us to select patients carefully and appropriately as we move forward with this emerging technology.”

Indications for CAS Placement

Patients who have a severe carotid artery narrowing are candidates for CAS placement, especially those who have had symptoms such as transient ischemic attack or cerebral infarction caused by that narrowing. “However, we also occasionally use CAS placement to treat severe carotid stenosis even though a patient has had no symptoms, just as we may recommend carotid endarterectomy for asymptomatic carotid disease,” Dr Brown says.

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How CAS Placement Works

Most patients arrive at the Cerebrovascular Clinic after carotid ultrasonography or MR angiography has shown narrowing of the carotid artery. After thoroughly examining the patient, the multidisciplinary neurosciences team members decide whether CAS placement is the appropriate treatment. If it is, the patient proceeds to the interventional neuroradiology suite in Saint Marys Hospital for further evaluation.

The patient is sedated but awake, and a small plastic catheter is inserted in a groin artery and tracked through the aorta to the carotid arteries. Next, contrast material is injected to delineate the anatomy. If the angiogram confirms severe narrowing that could be best treated with CAS placement, then the procedure begins.

First, a protection device may be deployed distally in the carotid artery—this device functions something like a minute net umbrella to catch material that may break free when the angioplasty is performed. Then the angioplasty balloon is brought across the plaque and inflated to push the plaque aside, thus reducing arterial narrowing. The stent—a small metallic scaffolding device—is brought up to keep the material pushed aside so the artery remains open. The procedure ends with withdrawal of the distal protection device and the catheter.

Typically, the patient is hospitalized for 1 day. Aftercare involves taking daily clopidogrel and aspirin for 1 month to prevent blood clots from forming at the CAS site and then aspirin alone indefinitely thereafter. Mayo Clinic specialists follow each patient long term, both to assess durability of the stent and to determine whether narrowing recurs. Follow-up includes annual carotid ultrasonography that begins several months after the procedure.

Results, Risks, and Complications

The concept of CAS placement is a logical extension of the balloon stenting used for coronary artery disease. Initially, in the early 1990s, CAS placement was performed on patients who were at high risk for



Left, Preoperative image of narrowed carotid artery. Right, Postoperative image showing stent in place and open artery after CAS placement.

conventional surgery. The outcomes of these early cases were excellent and the risk of stroke and death was extremely low. Because these measures of success were so similar to the standard treatment of carotid endarterectomy, the use of CAS placement was cautiously and carefully expanded.

Since then, Mayo Clinic experience with CAS placement suggests that, when performed by an experienced, multispecialty team on carefully selected patients, the procedure is approximately equal to carotid endarterectomy in terms of effectiveness, risks, and complications.

Future Directions

The National Institutes of Health has selected Mayo Clinic to participate with 25 US medical centers in the formal evaluation of CAS placement in the Carotid Revascularization Endarterectomy Versus Stent Trial (CREST). The goal of CREST is to determine how CAS placement compares with carotid endarterectomy, the standard treatment for carotid artery stenosis. A key question is whether the risk of recurrent narrowing after CAS placement is as low as the extremely low risk of recurrent narrowing after carotid endarterectomy.

CREST will also evaluate the comparative risks of stroke associated with CAS placement and carotid endarterectomy. Because CAS placement requires the interventionalist to work within the artery, the possibility exists for stroke during the procedure. Carotid endarterectomy, when performed by an experienced neurosurgeon or vascular surgeon,



Harry J. Cloft, MD, PhD, and
 Robert D. Brown, Jr, MD

carries a very low risk of stroke. The goal of the CREST protocols is to determine these issues conclusively.

Mayo Clinic's Cerebrovascular Clinic is now accepting enrollees into the CREST study. The Cerebrovascular Clinic also evaluates other

patients with carotid artery narrowing or occlusion, including those who do not wish to participate in the CREST study. To find out more about participating in CREST or referring a patient for evaluation, call the Cerebrovascular Clinic at 507-284-1588.

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