A Model of Collaboration

University Hospitals Case Medical Center’s Neurological Institute’s Skull Base Surgery Program is truly a collaborative effort. Our neurosurgeons work with a multidisciplinary team of otolaryngologists, radiation oncologists, neuro-oncologists, medical oncologists, interventional radiologists, ophthalmologists and other specialists to treat complex tumors and vascular lesions in the skull base region.

This “all for one” approach personifies our vision of “providing innovative, integrated, and individualized care that improves outcomes and the quality of life for all patients with neurological conditions.” Quite simply, patients and their overall well-being are the focal point of our efforts.

Our patient-centric, team-oriented methods are further illustrated by our commitment to minimally invasive techniques. Parasellar skull base tumors such as pituitary adenomas or meningiomas can be treated with an endoscopic approach in which the anterior segment of the skull is accessed through the nose. This method is commonly used for treating nasal and sinus conditions, but very few programs use this approach routinely for complex tumors of the skull, and, as you know, minimally invasive surgeries generally provide a decreased risk of complications, pain and scarring, and shorter hospital stays and faster recovery times.

The UH Case Medical Center’s Neurological Institute’s multidisciplinary team approach is further enhanced by a Magnet-certified, caring nursing staff. Our Reinberger Neuroscience Intensive Care Unit has earned the prestigious Beacon Award for Critical Care Excellence. UH Case Medical Center is the only hospital in the state of Ohio and one of only three nationally with five or more Beacon Award-winning intensive care units.

UH Case Medical Center is the primary affiliate of Case Western Reserve University School of Medicine, a nationally recognized leader in medical research and education, and ranks among the nation’s leading academic medical centers.

For the sixth consecutive year, UH Case Medical Center is recognized by Thomson Reuters as one of the top 15 major teaching hospitals in the country – the only hospital in Northeast Ohio to achieve this designation.

We want you to know that our interdisciplinary approach and full complement of resources are available to serve you and the needs of your patients, with convenient access throughout Northeast Ohio.

Sincerely,

Warren R. Selman, MD
Director, Neurological Institute
University Hospitals Case Medical Center
Harvey Huntington Brown, Jr., Professor and Chair, Department of Neurological Surgery
Case Western Reserve University School of Medicine

Anthony J. Furlan, MD
Co-Director, Neurological Institute
University Hospitals Case Medical Center
Gilbert W. Humphrey Professor and Chair, Department of Neurology
Case Western Reserve University School of Medicine
A 17-year-old male presented with severe headaches, hearing loss and dizziness. During summer football practice, the patient’s brother had also noticed that the patient had balance difficulties while running. A hearing test revealed complete loss of hearing in the right ear; a follow-up MRI ordered by the patient’s ear, nose and throat specialist showed a large (4 to 4.5 cm) acoustic neuroma (also known as vestibular schwannoma).

**Evaluation and Treatment**

The patient was referred to Cliff A. Megerian, MD, holder of the Richard and Patricia Pogue Chair in Auditory Surgery and Hearing Sciences; Vice Chairman, Otolaryngology–Head and Neck Surgery; Director, Otolaryngology–Head and Neck Surgery and Neurotology, University Hospitals Case Medical Center; and Professor, Otolaryngology–Head and Neck Surgery and Neurological Surgery, Case Western Reserve University School of Medicine.

“Acoustic neuromas are tumors that, although not malignant, can be fatal because they cause the brain to be compressed; the patient can then develop hydrocephalus and brain herniation,” says Dr. Megerian. “For older patients, we can sometimes remove part of the tumor to relieve pressure on the brain. But in young patients these tumors can grow rapidly. Based on the age of this patient, we chose to remove the entire tumor.”

Dr. Megerian, an otoneurological surgeon, teamed with neurosurgeon Nicholas C. Bambakidis, MD, to plan and perform the surgery. Dr. Bambakidis is Director, Cerebrovascular and Skull Base Surgery, University Hospitals Neurological Institute, UH Case Medical Center, and Associate Professor, Neurological Surgery, Case Western Reserve University School of Medicine. “Because the tumor was so large, we decided to use a modification of a standard technique, the translabyrinthine approach,” says Dr. Megerian. “We felt that adding a retrosigmoid subtemporal craniotomy would give us the best exposure and allow us to avoid injury to healthy brain tissue and to the nerve that controls facial motion.”

The 7½-hour procedure began with the retrosigmoid subtemporal craniotomy, as Dr. Bambakidis removed bone from the skull and put it aside, to be replaced later from the temporal and occipital regions. Dr. Mejerian then drilled through the mastoid and labyrinth (inner ear) structures to gain access to the posterior fossa. “This approach frees the sigmoid sinus, minimizing brain retraction,” says Dr. Mejerian. “I then began removing the lateral part of the tumor, exposing and freeing the facial nerve, and Dr. Bambakidis removed the bulk of the tumor against the brain stem.”

**Outcome**

Upon awakening from the surgery, the patient immediately asked for his family. He went home three days after surgery, his symptoms resolved (except for the hearing loss, which is permanent). One week after the procedure, he walked onto the football field at the head of his team. “We did a six-month MRI scan that looked fine, and he’s now running track,” says Dr. Mejerian. “He doesn’t appear to be at risk for recurrence, but if his annual MRI this fall is negative, we’ll continue to do the scan annually for five years.”

**Discussion**

Although acoustic neuroma in children is unusual, Dr. Mejerian says he sometimes sees four or five new adult cases per week, mostly patients in their 30s and older. “We have one of the busiest skull base acoustic neuroma programs in the nation in terms of how many patients come to us – 75 to 80 new patients in the last calendar year,” he says. “Of course, not every patient is treated surgically. Some are put under observation, and for smaller tumors, stereotactic radiosurgery can prevent the tumor from growing further. But this patient’s tumor was too large for a Gamma Knife.”

“Lesions like the one this patient had are extremely challenging to deal with,” says Dr. Bambakidis. “They require specialized care and even in the most experienced hands are high-risk procedures. It’s clear that overall outcomes are better when tumors and lesions occurring in the skull base are tackled by a team of experts in specialized centers where neurosurgeons, otologists and radiation oncologists work closely to design individualized treatment approaches. This type of approach is only available in a few places across the country, or even worldwide, for that matter.”
Targeting Skull Base Disorders
Multidisciplinary program tackles challenging conditions in a complex region

A program at University Hospitals Case Medical Center’s Neurological Institute that teams neurosurgeons with otolaryngologists is bringing a powerful multidisciplinary focus to disorders of the cranial skull base region, and providing patients with leading-edge, minimally invasive options for treatment. The UH Case Medical Center Skull Base Surgery Program also draws upon the expertise of radiation oncologists, neuro-oncologists, medical oncologists, interventional radiologists, ophthalmologists and other specialists to treat tumors and vascular lesions in this complex anatomical region.

Interdisciplinary Conferences
“We treat a broad range of disorders, including pituitary and acoustic tumors, meningiomas, brain aneurysms, and malignant tumors of the bone and sinuses, as well as genetic disorders such as neurofibromatosis and von Hippel-Lindau syndrome,” says Nicholas C. Bambakidis, MD, Director, Cerebrovascular and Skull Base Surgery, UH neurological institute, University Hospitals Case Medical Center. “In order to focus our efforts and the expertise of our specialists, we’ve launched new interdisciplinary conferences devoted to reviewing patients with certain types of disorders.” Dr. Bambakidis also is associate Professor, neurological surgery, Case Western Reserve University school of Medicine.

Minimally Invasive Procedures
In addition to providing conventional surgical expertise, the Skull Base Surgery team utilizes relatively new, minimally invasive techniques in place of open surgery in select groups of patients. “We can do procedures that previously required either a skin incision or a larger opening than we need to do now,” says Warren R. Selman, MD. “We access parts of the anterior skull base using tiny endoscopes, inserted through the nose, that give us a fantastic view and allow us to do everything that we could do before with minimal morbidity and better visualization. The endoscope allows access for instruments to work in very narrow corridors where previously we had to put in retractors and get a wider area to visualize the region. These procedures are always performed by a team, with the ear, nose and throat specialists guiding with the endoscopes and the neurosurgeons working around the anatomy and removing the tumor.”

The team uses endoscopically assisted surgery routinely for treatment of pituitary tumors and benign or malignant tumors of the bone or dura in the anterior skull base, says Chad A. Zender, MD, FACS. Although the procedure is commonly used to remove nasal polyps and treat sinus disorders, “it’s rare for a center to use it to treat complex tumors of the anterior skull, as we do.” The team’s endoscopic capabilities will be enhanced by the planned addition of a three-dimensional system of visualization that will mirror the three-dimensional view that is seen in an open surgical procedure, adding more depth and clarity, Dr. Zender says. “The 3-D monitor will help us...”
define the anatomy. So we’ll have a working port in one nostril, where the neurosurgeon will control the working instruments. The other nostril will be used for the endoscope held by the ENT surgeon, who will navigate."

The Skull Base Surgery team uses another minimally invasive procedure, endovascular treatment, as an alternative to open surgery for brain aneurysms. "Endovascular treatment allows the surgeon to close off the aneurysm by packing it full of coils or even glue material from the inside," says Dr. Selman. "It works well for a subset of brain aneurysms and patients. The factors we take into consideration are location and shape of the aneurysm, patient age and condition, and patient choice in some instances."

Endovascular treatment, he adds, is appropriate for deep-seated aneurysms that are difficult to treat surgically and for older patients who are at higher risk for complications from surgery. Dr. Zender maintains that because it’s still uncertain how the long-term results of endovascular surgery compare with those of open surgery, the latter “is still an important adjunct, particularly in younger patients or with the aneurysms that we can get to safely. So it’s critical to have expertise in both technologies available, and I think we’re one of the few institutions that have that.”

Collaboration in Cancer Treatment
When caring for patients with malignant tumors of the cranial skull base, Skull Base Surgery Program physicians work closely with oncologists at University Hospitals Seidman Cancer Center (formerly University Hospitals Ireland Cancer Center), a 120-bed facility, opened in June 2011, that’s equipped with an intraoperative MRI suite. UH Seidman Cancer Center is one of only 12 free-standing cancer hospitals that are part of a National Cancer Institute-designated Comprehensive Cancer Center.

“Treating these patients is very complicated; often they still need very large surgical exposures and multiple treatment teams,” says Dr. Bambakidis. “Following surgery they often need radiation treatment, so it requires a radiation oncologist and possibly a medical oncologist – all the resources that a comprehensive cancer center brings to the table.” The collaboration with radiation oncologists extends to providing comprehensive radiation treatment with the minimally invasive Gamma Knife and CyberKnife. In addition, patients and their families have access to genetic counseling and testing.

**NF2 Group at UH**
A diagnosis of neurofibromatosis type 2 (NF2) poses lifelong challenges not only for the patient, who may grapple with a plethora of debilitating symptoms, but also for the physicians charged with managing the disease. Most affected individuals develop bilateral vestibular schwannomas, nonmalignant tumors on the eighth cranial nerve, resulting in hearing loss, tinnitus and balance difficulties. Other possible manifestations include optic nerve gliomas that lead to blindness; spinal cord tumors that cause myelopathies or compressive sensory and motor neuropathies; and large tumor burden that leads to brainstem compression and hydrocephalus.

The course of the disease is unpredictable, with considerable variations in tumor growth rate and location. “We established a focused NF2 group that meets monthly to discuss the care of these patients because they, most of all, need interdisciplinary care, including genetic counseling and medical neurology,” says Nicholas C. Bambakidis, MD, Director, UH Cerebrovascular and Skull Base Surgery. “They require care over many years, often with repeated surgical procedures. They are exactly the type of patient who benefits from the expertise a comprehensive skull base program offers.”

**Access Our Expertise**
To discuss the transfer of your patient to the Skull Base Surgery Program, please call 216-844-1111.

Gadolinium-enhanced MRI showing bilateral large vestibular schwannomas, a posterior fossa meningioma and bilateral trigeminal nerve schwannomas.
An estimated 300,000 people in North America are afflicted with dystonia, a disorder characterized by a progressive loss of motor control. Patients with generalized dystonia grapple with involuntary muscle spasms that lead to uncontrolled twisting and turning in awkward, sometimes painful postures. Although cognition, intelligence and life span are often normal, the disorder can have a devastating impact on quality of life, as its victims frequently struggle to perform simple activities of daily living.

At University Hospitals Case Medical Center’s Neurological Institute, a research team is using advanced imaging technology to explore the complex network of brain activity relating to movement in healthy subjects and in patients with dystonia. “Normally, MRI is used to provide an image of the structure of the brain,” says Benjamin L. Walter, MD, Medical Director, Deep Brain Stimulation Program, UH Case Medical Center, and Assistant Professor of Neurology, Case Western Reserve University School of Medicine. “Functional MRI (fMRI) takes advantage of the artifact that’s created by blood flow and the oxygenation of blood. The level of oxygenation is highly correlated with neural activity in the same regions, so we can see which parts of the brain are being used.”

**Imaging Proprioception**

Dr. Walter’s current research explores two key areas: the nature of brain activity in patients with dystonia, and how that differs from activity in normal subjects; and understanding how deep brain stimulation (DBS), a leading-edge treatment for selected dystonia patients, works to quiet the involuntary spasms. Treating dystonia with DBS involves the placement of electrodes in the internal segment of the globus pallidus, a subcortical structure also targeted in the DBS treatment of Parkinson’s disease, essential tremor and obsessive compulsive disorder. “In disorders such as Parkinson’s and essential tremor, when you turn the stimulator on there’s a pretty quick benefit,” Dr. Walter explains. “That’s not the case with dystonia – it slowly improves over a long period of time, six months or longer. So there’s more of a neuroplastic effect that’s probably involved in the mechanism of DBS.”

The initial stage of Dr. Walter’s research involves using fMRI to observe brain activity in healthy subjects and in patients with dystonia who have not received DBS implants. “We’re looking to examine how sensory and motor information is handled in the brain in patients with dystonia. Dystonia is obviously a movement disorder, but there’s a lot of evidence that the integration of sensorimotor information is dysfunctional.” The research team chose to study proprioception in their subjects “because that’s very close to movement, and you get direct feedback about joint position when you move a limb.”

Using a small device that vibrates over a wrist tendon, the researchers induce a kinesthetic illusion (the false perception that the subject’s wrist is flexing) and examine the resulting fMRI images. “In our normal patients, we’re seeing that the motor cortex and the motor portion of the basal ganglia and the posterior striatum are involved,” Dr. Walter notes. “In our dystonic patients, we’ll look for changes in how the proprioceptive input is being handled. We’re hoping to discover where the signal is becoming abnormal in these patients, whether there are different anatomical structures involved, and whether there’s a different place we could put the DBS wire and get a more robust effect.”

**Defining Differences, Looking for Changes**

The next stage of the research will include fMRI imaging of patients who have received DBS treatment. “DBS is not really well understood,” Dr. Walter says. “In part you need to know where to look, and this type of neuroimaging can tell us where there are abnormal hot nodes that are involved in our proprioception paradigm and may be worth investigating using other methods. Essentially, we’re defining the differences between dystonia and normal patients, and in the dystonia patients who get DBS, we’ll be looking for changes in their brain activity over time, as the dystonia melts away.”
Preserving a Lost Art
UH neurosurgeons champion brain bypass in select patients

A microsurgical procedure that has lost some ground to advances in endovascular therapy still plays a critical role in the management of selected neurovascular disorders, according to a University Hospitals Case Medical Center neurosurgeon who performs the procedure.

“Though its indications are rare, the ability to perform brain bypass correctly can make all the difference for certain patients who have complex brain aneurysms or other cerebrovascular disorders,” says Nicholas C. Bambakidis, MD, Director, Cerebrovascular and Skull Base Surgery, University Hospitals Neurological Institute, UH Case Medical Center, and Associate Professor, Neurological Surgery, Case Western Reserve University School of Medicine.

Fewer Procedures
Brain bypass, also known as extracranial to intracranial (EC-IC) bypass, was first performed successfully in 1967 to treat an occluded internal carotid artery. The procedure involves connecting the external carotid artery to the internal carotid artery, either directly or by grafting a vein or artery. The choice of graft depends on the size of the recipient and donor vessels, graft availability and the extent of required blood flow augmentation.

In a Journal of NeuroInterventional Surgery article (2010;2:229-236) discussing the current role of cerebral revascularization for ischemic disease, Dr. Bambakidis and co-author Shakeel Chowdhry, MD, Neurological Institute, University Hospitals Case Medical Center, Resident, Department of Neurological Surgery, Case Western Reserve University School of Medicine, note that the procedure was originally envisioned as a treatment option for cerebrovascular occlusive and ischemic disease, but is now rarely used for these indications. “As newer, minimally invasive technology has come online in vascular treatment, the pendulum swings toward avoidance of surgical treatment at all costs,” says Dr. Bambakidis. “As a result, many centers are doing endovascular treatment of aneurysms, but fewer are doing brain bypass procedures. In a sense, it’s almost becoming a lost art. The expertise is becoming more and more concentrated in larger centers.”

Indications for EC-IC
While an endovascular procedure is the optimal treatment for many brain aneurysms, in select cases endovascular treatment poses a higher risk of stroke than brain bypass, Dr. Bambakidis says. “There are certain brain aneurysms that simply can’t close without the sacrifice of some normal blood vessels,” he explains. “The EC-IC bypass is designed to bypass the aneurysm and provide blood to those vessels which you would otherwise have to close off, and having that blood supply reduces the risk of stroke.”

Complex giant aneurysms (greater than 2.5 cm) are the main indication for brain bypass, although Dr. Bambakidis recently performed the procedure on a young patient with a smaller aneurysm. “This was a 14-year-old boy with a 2-cm aneurysm; we did the bypass using a radial artery from his wrist,” he notes. “Aneurysms in younger people are rare, but when we do see them they tend to be these larger ones that often require some kind of bypass or complicated open surgery.” He adds that although brain bypass is not typically indicated for ischemic brain stroke, it is a critical treatment modality for certain young patients with vascular insufficiency and resultant ischemic or hemorrhagic strokes. “These patients, who often have a disease called moyamoya syndrome, can have their symptoms improve dramatically after surgery,” Dr. Bambakidis says.

Multidisciplinary Approach
New technology such as fluorescent videoangiography is making brain bypass easier and safer, Dr. Bambakidis says, but it is still a delicate procedure that’s difficult to perform well. “Multidisciplinary expertise and coordination of care between neuroradiologists and neurosurgeons is critical to achieving an optimal result for the patient,” he says. “Because we’re known as a center that offers this expertise, our patients come from areas far beyond greater Cleveland to undergo the brain bypass procedure.”

Access Our Expertise
To have your patient with a brain aneurysm or abnormal cerebral blood vessels evaluated, please call 216-844-2724 or 1-866-UH4-CARE (1-866-844-2273).
Advanced Learning & Free CME Credits

The University Hospitals Neurological Institute Journal provides physicians with learning opportunities surrounding innovations in surgeries, treatment options, research protocols and more gained from the bench-to-bedside methodology utilized at UH.

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University Hospitals Neurological Institute

At convenient locations throughout Northern Ohio, University Hospitals Neurological Institute delivers innovative, integrated and individualized care to patients with diseases affecting the nervous system.

We are the only Neuroscience Center of Excellence in the state of Ohio with a Stage 4-Institute Designation. Our 16 Centers of Excellence offer you premier care and access to some of the country’s foremost experts in neurology, neurosurgery, neuroradiology, neuro-oncology, neuro-ophthalmology, neurootology, neuropathology, neuropsychology and related specialists. These teams work in collaboration with medical specialists at UH Rainbow Babies & Children’s Hospital and UH Seidman Cancer Center (formerly UH Ireland Cancer Center), with access to the UH Neuroscience Intensive Care Unit and Neuroscience Nursing Practice.

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