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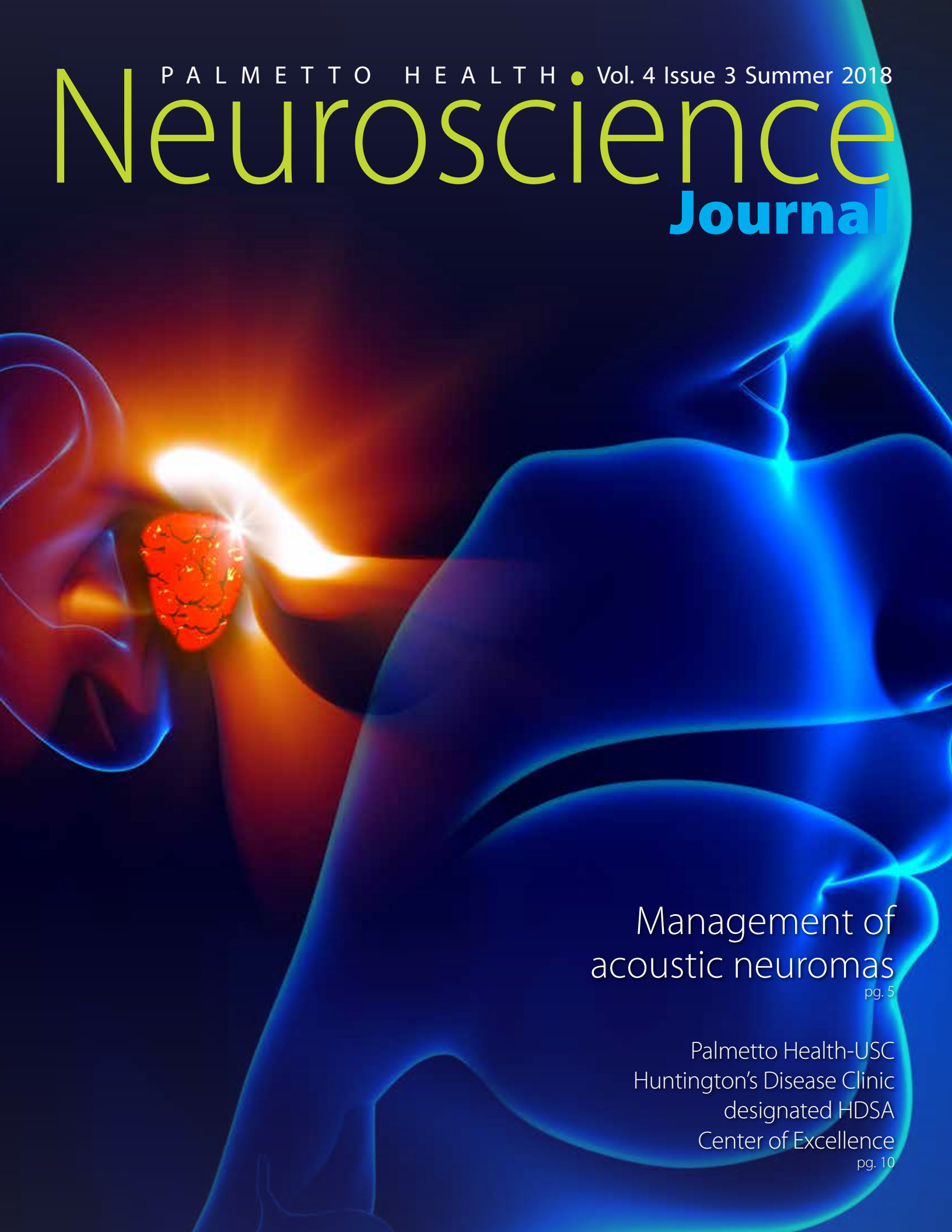
Neuroscience Journal



Management of acoustic neuromas

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Palmetto Health-USC Huntington's Disease Clinic designated HDSA Center of Excellence

pg. 10

As physician co-leaders of Palmetto Health's neuroscience service,

we share a vision to provide the most advanced neurology and neurological surgery treatments available to the residents of South Carolina. We are excited to share this edition of our neuroscience journal featuring articles about acoustic neuromas and the first Huntington's Disease Society of America Center of Excellence in South Carolina.

Souvik Sen, MD, MS, MPH
Chair of Neurology,
Palmetto Health-USC Neurology
Professor of Neurology,
University of South Carolina School of Medicine

Roham Moftakhar, MD
Chief of Neurosurgery,
Palmetto Health Richland
Medical Director,
Palmetto Health-USC Neurosurgery
Associate Professor of Clinical Surgery,
University of South Carolina School of Medicine

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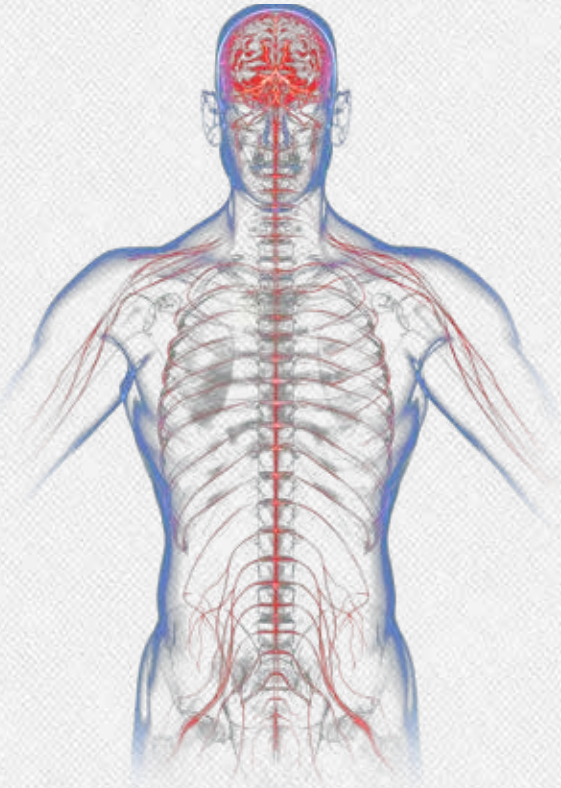

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Management of acoustic neuromas

by Roham Moftakhar, MD, chief of neurosurgery, associate professor of surgery, Palmetto Health-USC Neurosurgery

Acoustic neuromas, also known as vestibular schwannomas, originate from the vestibular nerve and are most often benign tumors. The nerve involved in the majority of cases is the superior vestibular nerve. The incidence of clinically recognized acoustic neuroma is 1–2 in 100,000 person-years, and in the U.S., acoustic neuromas comprise 80 to 90 percent of cerebellopontine angle tumors. The majority of these tumors are unilateral (more than 90 percent). Bilateral variants are most common in patients with neurofibromatosis-2 (NF-2). These tumors are slow growing with average growth rate of 1.9 millimeters per year.

The signs and symptoms of a patient with acoustic neuroma could be decreased hearing, complete hearing loss, tinnitus and imbalance. Very rarely, there is unilateral facial numbness. The main imaging method to evaluate acoustic neuroma is magnetic resonance imaging (MRI) of the brain with and without contrast, with special attention to the internal auditory canal.

Once the acoustic neuroma is diagnosed, a multidisciplinary approach should be instituted for its management. Most of the time this team consists of skullbase neurosurgeons, neuro-otologists and radiation oncologists. Patients should receive a hearing test to determine if the patient's hearing is serviceable. Most practitioners use 50 percent or greater hearing discrimination and less than 50 decibels as the definition of serviceable hearing. Whether or not the patient has serviceable hearing could alter the surgical approach and overall management.

Management of acoustic neuromas can be broken down into three main categories: observation with serial imaging, microsurgical resection and stereotactic radiosurgery.

Observation usually is reserved for patients with incidental finding, who are asymptomatic. Elderly patients or those with medical problems preventing surgical treatment might wish to have their tumor observed over time. Since acoustic neuromas are benign slow-growing tumors, imaging is done every six months to once a year to follow for tumor growth. The best

imaging for acoustic neuromas are MRI of the brain with and without contrast. If the patient cannot get an MRI, then computed tomography (CT) with contrast would be a second choice.

Microsurgical resection of acoustic neuroma is mostly for patients who are symptomatic and/or patients with brainstem compression. There also is data that suggests patients with smaller tumors in the internal auditory canal could benefit from resection, since hearing preservation down the line is higher with microsurgery (*Journal of Neurosurgery*, 1993). Microsurgical approaches can be divided into three categories: retrosigmoid craniotomy, translabyrinthine approach and middle fossa. Retrosigmoid craniotomy could be used if the patient has serviceable hearing and the bulk of the tumor is in the cerebellopontine angle. The translabyrinthine approach could be used in patients without serviceable hearing and in cases where the bulk of the tumor is in the internal auditory canal. Finally, the middle fossa approach is reserved for cases where there is serviceable hearing and the tumor is within the internal auditory canal. According to Sammii and Matthies in their series of 1,000 surgically resected patients with vestibular schwannoma, facial nerve preservation was found in 93 percent of patients and cochlear nerve preservation in 68 percent. However, hearing preservation was achieved in 47 percent (Sammii and Matthies, Neurosurgery, 1997). Recurrence rate after gross total resection is less than 1 percent.

Radiation therapy with stereotactic radiosurgery (SRS) is another option for treatment of acoustic neuroma. Studies suggest that SRS can be used to successfully control tumors up to 3 centimeters in diameter (Lederman et al., Stereotactic and Functional Neurosurgery, 1997). For tumors less than 3 centimeters, tumor control has been reported very similar to microsurgery upwards of 95 percent (Pollock, et al. 2006 neurosurgery). The exception to this is when there is compression of the brainstem

or when the tumor is adjacent to the brainstem where radiation could have deleterious effects on the normal tissue. Long-term hearing preservation has been reported to be higher with SRS (57.5 percent in SRS versus 14.4 percent in microsurgery group) (Nonaka et al., Neurosurgery, 2013). In this study long term tinnitus was higher in patients after SRS than microsurgery. However, facial nerve palsy and post-operative complications were higher in the microsurgery group (47 percent versus 4.6 percent).

In conclusion, treatment of acoustic neuroma should be performed in a multidisciplinary manner. The team usually consists of neurosurgeons, neuro-otologists and radiation oncologists. Acoustic neuromas could be monitored if they are small and without compression of the brainstem or other symptoms. Treatment options include microsurgery and radiation therapy. Radiation therapy is reserved most of the time for smaller tumors without symptoms related to brainstem and compression. Microsurgery is reserved for larger tumors with brainstem compression and for patients with symptoms related to tumor compression.

Case example

A 38 year-old man presents with pressure in his ear and tinnitus. MRI of the brain demonstrated right acoustic neuroma measuring 1.8 centimeter x 1.5 centimeter (Figure 1). The options of microsurgery, stereotactic radiosurgery and monitoring with serial imaging were discussed. Due to size of the tumor and patient's young age, we recommended microsurgical resection of the tumor. The patient underwent right retrosigmoid craniotomy for resection of the tumor. Gross total resection of the tumor was performed. Post-operative MRI confirmed gross total resection (Figure 2). Post-operatively, the patient had a symmetrical facial function and was neurologically intact. ◄

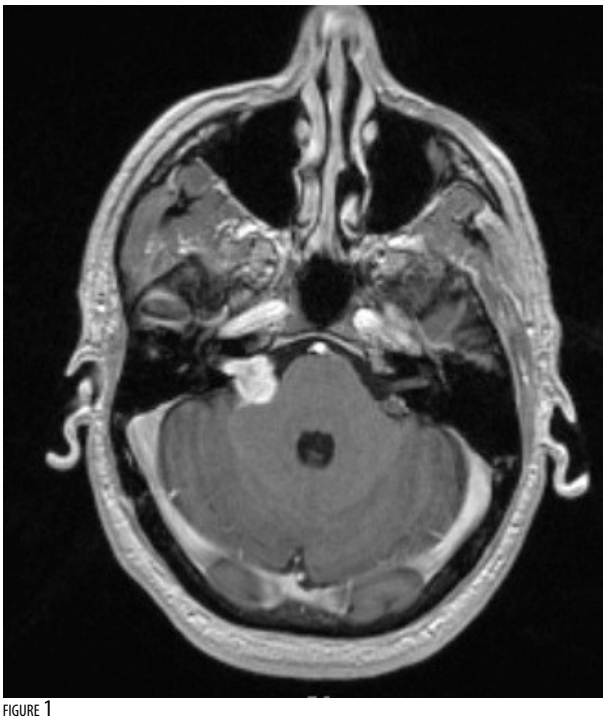

FIGURE 1

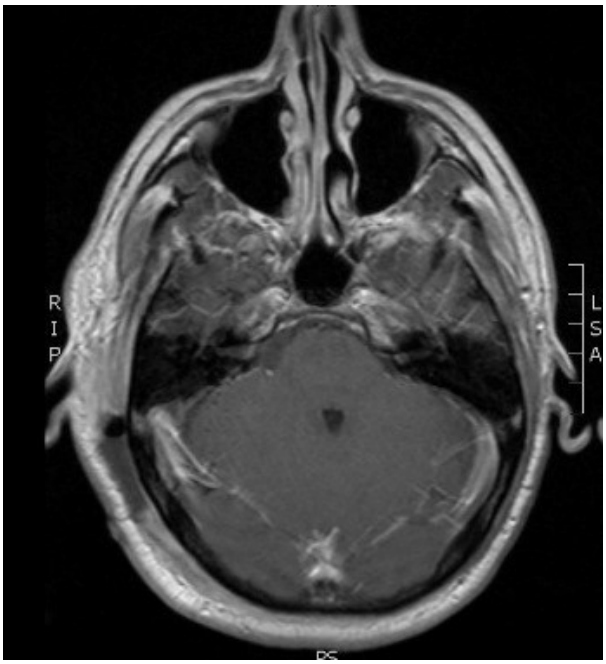

FIGURE 2

Skullbase surgery and complex brain tumor surgery

Skullbase surgery is employed when a tumor is located at the base of the skull, the underside of the brain or the upper vertebrae of the spinal column. These are complicated areas that involve the utmost care and precision when being treated.

At Palmetto Health-USC Neurosurgery, we believe in the multidisciplinary team approach when diagnosing and treating both malignant and benign tumors of the brain and base of the skull. Our fellowship-trained neurosurgeons are highly specialized and dedicated to finding the most minimally invasive techniques possible.

Additionally, we incorporate the expertise of physicians from otolaryngology, occuloplastic surgery, ophthalmology, radiation oncology and endocrinology. This collaboration leads to precise evaluations and better outcomes for our patients.

Conditions treated

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- Epidermoid (congenital cholesteatoma)
- Esthesioneuroblastoma
- Glomus jugulare tumors
- Facial nerve disorders
- Jugular foramen tumors
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- Meningioma
- Petrous apex tumors
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Physician leaders

Roham Moftakhar, MD | Palmetto Health-USC Neurosurgery
Dr. Moftakhar is fellowship trained in skullbase, cerebrovascular and endovascular neurosurgery. He completed his residency in neurological surgery at the University of Wisconsin and his fellowship training at the University of Miami. Dr. Moftakhar is the chief of neurological surgery at Palmetto Health-USC Medical Group.

Erwin Mangubat, MD, MPH | Palmetto Health-USC Neurosurgery
Dr. Mangubat is fellowship trained in epilepsy, functional neurosurgery and endovascular surgery. He completed his residency in neurosurgery at Rush University Medical Center in Chicago and his fellowship training at both the Swedish Neuroscience Institute in Seattle and Rush University Medical Center.

David Straus, MD | Palmetto Health-USC Neurosurgery
Dr. Straus is fellowship trained in skullbase surgery and neurosurgical oncology. He completed his residency in neurological surgery at Rush University Medical Center and his fellowship training at the University of Washington Harborview Medical Center. He has advanced training in stereotactic radiosurgery from the Cleveland Clinic. Dr. Straus is the surgical director of the Gamma Knife Center.

Jonathan King, MD | CENTA Medical Group, PA
Dr. King is a rhinologist and otolaryngologist with specialized experience in anterior skullbase surgery. He completed his residency in otolaryngology at the University of Florida and previously served as chief of ENT at Maxwell Air Force Base.

Scott Thompson, MD | Carolina Eye, Ear, Nose & Throat Associates, LLC
Dr. Thompson is an otolaryngologist with fellowship training in skullbase surgery. He specializes in neuro-otology and lateral skullbase surgery. He completed his residency training in otolaryngology at the University of Oklahoma and his fellowship in neurotology at the University of Pittsburgh Medical Center.

Skullbase surgery team members

- Gamma Knife Center: Brandon Stone, MD; Eric Wooten, MD; Kurt Meyer, MD, PhD; Ben Wright, MD
- Oliver Simmons, MD, plastic and reconstructive surgery
- Rakesh Patel, MD, oculoplastic and reconstructive surgery
- Mark Robinson, MD, neuro-ophthalmology
- Charlotte Thompson, MD, neuro-ophthalmology, oculoplastics and orbital surgery
- Brooke McAdams, MD, endocrinology
- C. Blease Graham III, MD, neuroradiology, Pitts Radiology

How to refer

- Contact us at 803-360-0023.
- We are committed to providing prompt evaluation and treatment planning.
- Neurosurgical consultation will be available within two business days of referral.

First Huntington's Disease Society of America Center of Excellence in South Carolina

by Miroslav Cuturic, MD, assistant professor of clinical neurology, University of South Carolina School of Medicine, director, Palmetto Health-USC Huntington's Disease Clinic

This year, our Palmetto Health-USC Huntington's Disease Clinic received the Huntington's Disease Society of America Center of Excellence designation. The Huntington's Disease Society of America (HDSA) is the national leader and premier nonprofit organization dedicated to improving the lives of those affected by Huntington's disease. HDSA's network of Centers of Excellence provides assistance, education and outreach to families and health care professionals across the United States. The HDSA Centers of Excellence program provides an elite multidisciplinary approach to Huntington's disease care and research. HDSA currently has 43 Centers of Excellence across the United States, and our clinic is the first such center in South Carolina.

Although designated as an orphan disorder (affecting fewer than 200,000 people nationwide), Huntington's disease (HD) is the most common hereditary neurodegenerative disorder, three times as common as the more well known amyotrophic lateral sclerosis (ALS). HD is a globally widespread disorder, with a worldwide prevalence of 5–10 per 100,000 people, with some regional variability. With South Carolina's estimated population of five million, within the state, we can expect to have up to 500 individuals affected by Huntington's disease, and two to three times as many pre-symptomatic gene carriers who will develop the illness within the next decade.

HD is a very complex neurodegenerative disorder exhibiting a highly penetrant autosomal-dominant inheritance pattern, with both sexes having a 50 percent chance of inheriting the genetic defect from the affected parent. The genetic defect consists of CAG trinucleotide expansion on the short arm of chromosome 4, which translates into polyglutamine chain expansion in the mutant huntingtin protein, resulting in abnormal protein aggregation and neurodegeneration. The course is manifested by progressive motor, cognitive and psychiatric deterioration, with chorea being the most prominent feature. The illness emerges around mid-life, with a mean age of 39 years, but earlier in each generation and earliest for sons acquiring the gene from their fathers. Juvenile onset, before the age of 20 years, is seen in less than 10 percent of cases. Currently there is no FDA-approved cure or disease-modifying therapies for HD. However, multidisciplinary intervention is beneficial. This includes symptomatic therapy, lifestyle interventions and supportive management.

Our HD clinic was founded in 1985 at the South Carolina Department of Mental Health in Columbia, SC, by Ruth K. Abramson, PhD. The clinic was one of the first of its kind in the southeastern United States, and has provided multidisciplinary on-site services to HD patients and their families in South Carolina for decades. Seeking a stronger academic and clinical research-oriented environment, the clinic was relocated to the University of South Carolina School of Medicine Department of Neurology in 2010. Currently, our HD clinic is located on the Palmetto Health Richland campus in the Palmetto Health-USC Neurology department. We provide multidisciplinary services that include on-site neurological evaluations and treatment, genetic counseling, neuropsychological,

psychological and social work consultations, as well as physical, speech and occupational therapy. Additionally, on the campus of our medical center, we provide psychiatric consultations. Furthermore, our genetic counselors provide genetic counseling and predictive HD testing for individuals at risk, as well as genetic counseling and family planning for families at risk. We generally see new patients and have follow-up appointments with patients every Friday; however, in case of need, arrangements can be made for an HD patient to be seen any day of the week.

Our clinic has an "open door policy" that allows selected support group leaders to schedule HD patients directly for an initial neurological consultation, without the need for referral from a primary physician. For individuals with HD who meet the criteria, we provide an indigent program that follows national guidelines as defined by the National Health Service Corps sliding fee discount program. Through our affiliations, we participate in patient care, including in-house visits, of institutionalized HD patients within the South Carolina Department of Mental Health, as well as the South Carolina Department of Corrections. We also provide bedside visits of patients in terminal stages of Huntington's disease residing in affiliated nursing homes. Finally, we provide in-house consultations for hospitalized HD patients in the Palmetto Health system, the largest integrated health care system in the South Carolina Midlands region. Our clinic currently serves patients and their families from all of South Carolina and adjacent areas of North Carolina and Georgia, and remains the principal center for specialized HD care in our state.

In our HD clinic, we provide training to medical and genetic counseling students, as well as neurology, internal medicine and psychiatry residents. Recently, we also have included students from USC's College of Social Work. The USC School of Medicine Department of Neurology currently is funding a student-based research program for the development of a nematode (C. elegans) model of Huntington's disease in collaboration with the Department of Pharmacology, Physiology and Neuroscience.

The program recruits honor-roll, pre-med, postgraduate and medical students from the University of South Carolina to study the neurobiology of Huntington's disease. The aim of the program is to attract our university's most gifted and motivated students to the field of Huntington's disease research. The Palmetto Health-USC Neurology Clinic is a certified research site of the Huntington's Study Group, a world leader in facilitating high quality clinical research trials in HD. Since July 2016, we have been participating in Enroll-HD, an international observational study and database. Our HD clinic staff has supported and participated in all annual HDSA sponsored events within the South Carolina HD community over the last several years, including the Team Hope Walk, Strike Out HD Bowling Bash, and Hope for HD Walk and Bike-a-Thon. This year HDSA recognized our long-term commitment and awarded us a Center of Excellence designation.

With our Palmetto Health and USC School of Medicine partners, we strive to further advance our services to HD patients, their families and the community. We want to ensure that our patients and their families receive the highest standard of care. Our goal also is to facilitate the access of our patients to cutting-edge research and multi-center clinical trials, which is additionally enhanced by networking within the HDSA Centers of Excellence program. This affiliation augments our outreach capabilities through HDSA programs and contributes to the long-term viability of our program. Synergy with the Centers of Excellence program further diversifies our research, teaching and educational pursuits. While the Center of Excellence designation is recognition of our long-term commitment to providing the best possible care to our patients, it also fills us with a sense of responsibility to meet the high standards set by the HDSA. As the director of the clinic, I would like to invite our medical community to support and utilize the services of our clinic and help us build the program around the needs of our HD patients and their families. ◄

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