Auditory Brainstem Implant Offers Hope for Hearing

On July 13, 2007, Dr. Craig Buchman from the Department of Otolaryngology-Head and Neck Surgery and Dr. Matthew Ewend from the Division of Neurosurgery at UNC were the first surgeons in the United States to place an auditory brainstem implant (ABI) in to a patient without brain tumors resulting from Neurofibromatosis type 2 (NF2). This event marks the first patient implanted in the UNC-initiated clinical trial to study the safety and efficacy of the ABI in patients without NF2. The patient, Watson Hale, has recently had his device activated and is able to hear for the first time since suffering meningitis in 2002.

The ABI is an implantable neural stimulator, developed in 1979 by William F. House and colleagues in Los Angeles, to treat hearing loss in patients without cochlear nerves because of NF2. Patients with NF2 have bilateral acoustic tumors and they often times lose hearing because the cochlear nerve(s) are destroyed by either the tumors or the surgery needed to treat these tumors. The ABI is placed over the cochlear nucleus of the brainstem to stimulate the auditory system deep to the cochlear nerve.

Similar to the cochlear implant, the ABI stimulates the auditory system by using electrical impulses. A microphone connected to an external speech processor collects and decodes the acoustic signal (i.e. sound) into the various component parts. This information, as a digital signal, is then delivered by radiofrequency to an internal receiver/stimulator implanted under the skin behind the ear. The stimulating electrodes exit the internal device and are placed in to either the cochlea (i.e. cochlear implant) or directly onto the brainstem cochlear nucleus (i.e. ABI). Thus, the cochlear implant stimulates the cochlear nerve and the ABI stimulates the cochlear nucleus. Remarkably, specific electrical stimulation of the auditory system is recognized by the brain as sound. While cochlear implants have demonstrated a profound success in both adults and children with a variety of hearing losses, only preliminary evidence from Italy and elsewhere in Europe suggests that the ABI can be used safely in patients without NF2 and may provide patients with significant hearing abilities.

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Mr. Hale is a 62-year-old gentleman who suffered from meningitis in 2002 and associated bilateral, total profound hearing loss. He underwent a cochlear implant surgery in 2002 but was unable to gain any benefit from the device because of scarring and bone growth (i.e. ossification) inside the cochlea. Stimulation of his cochlear implant caused significant facial twitching without any hearing. Because he was a previously normal-hearing individual, we felt that an ABI could potentially help him by delivering some auditory information directly to his cochlear nucleus, thereby bypassing his diseased cochlea. Since this was not an FDA-approved use for this device in the United States, a clinical trial was initiated with the device manufacturer (Cochlear Corporation, Englewood, Colorado) and UNC.

The surgery was carried out on July 13 through an opening in the back of the patient’s skull. Undertaking an ABI surgery requires collaboration among a host of professionals from otolaryngology, neurosurgery, audiology, and industry. In the operating room, correctly placing the device requires both a detailed understanding of the anatomy by the surgeons and an ability to monitor the auditory system’s responses to electrical stimulation (EABR). Following placement of the electrodes, stimulation is carried out to see if appropriate responses are elicited. If needed, minor adjustments in the position of the electrodes can then be made in an attempt to maximize success. At UNC, operative monitoring was performed by Drs. John Grose and Holly Teagle from the Department of Otolaryngology-Head and Neck Surgery.

Following recovery from surgery, Mr. Hale underwent activation of his ABI in November by a team of audiologists from the Department including Marcia Clark Adunka, AuD, and Holly Teagle, AuD. Device activation required carefully observing the patient’s responses to varying levels of electrical stimulation on the different electrodes. Indeed, at activation, he was immediately able to discern sound from other effects and could hear different pitch levels.

On the day of activation, Mr. Hale could hear soft sounds on a hearing test and showed that the device could provide significant improvements in lip-reading and sound awareness. While these initial results were very encouraging to both the patient and the implant team members, hearing rehabilitation is expected to be an ongoing, long-term learning process. This is partially because his brain has been deprived of sound for more than 5 years and the ABI delivers a signal that his brain is unaccustomed to. Only time will tell how much progress Mr. Hale will make with his new implant, but the early results seem very promising.
I was fortunate to be a part of a recent Cleft Care mission trip led by Dr. John van Aalst of UNC to the West Bank. I am grateful for the funding that I received, including a humanitarian travel grant through our academy, the AAO-HNS, as well as the PCRF (Palestinian Children’s Relief Foundation), and Smile Train.

We arrived in Tel Aviv Sunday, December 2nd, and set off immediately for Tulkarem, a small town on the northwest edge of the West Bank roughly 40 miles northwest of Jerusalem. Our small team comprised of myself, Dr. John van Aalst, craniofacial surgeon; Dr. Ron Herring, anesthesiologist; Bilal Saib, dentist; and Pat Davison, photojournalist. We traveled past several checkpoints and arrived at the hospital in Tulkarem where patients were expecting our arrival. We spent the remainder of Sunday evening screening over 100 patients who had come from all over the region hoping to have surgery.

We divided into two triage rooms, and as only two members of our group spoke Arabic, Dr. van Aalst and Bilal Saib were invaluable as translators. Groups of 20 people congregated around us throughout the day, their arms outstretched grasping pieces of paper with only their name in Arabic listed on top. This atmosphere seemed to symbolize the anxious plea of those gathered to be seen and selected for surgery. We managed to see all those who came and out of that day set up approximately 30 surgical cases over the course of the following 6 days. Many of remainder were put on the list for the next trip, and a number also had problems such as minor scars or burns which would not be best optimized with surgery or had problems so severe such as synostosis cases whose care would require neurosurgical assistance and intensive care monitoring, neither of which were available at present time.

The ensuing days were busy. We arrived at the hospital where mandatory breakfast was served, which was fortunate as lunch was overlooked on most days because of the busy schedule of cases. We did a number of primary cleft lip and palate repairs as well as many revisions for fistulae, or improved cosmesis. We also did a number of alveolar bone grafts. One of the highlights of the trip for me was being able to do an otoplasty on a 10 year old boy with prominent ears. As an otolaryngologist, I also worked with two local ENT physicians who had been trained in the Ukraine and helped teach them some basic principles of ear surgery. We had secured an operating microscope, which was purchased in the States, in efforts to allow us to better place tympanostomy tubes at the time of cleft palate surgery. Unfortunately, the microscope was held up in customs and I was relegated to putting tubes in using a hand-held otoscope. I can only say, “big props to Dr. P” because that is no easy task to do, especially in small children, but we did manage to place around 5 sets of tubes, and did myringotomy with aspiration on another handful. We arranged careful follow-up on all of our patients with the local ENT physicians and oral surgeons, and hope that in the future these surgeons will be able to assume greater responsibilities with cleft care in the region.

On our last day, we had the first international Palestinian Cleft Society conference in Ramallah. Over 100 people attended, including several other teams of cleft surgeons from Europe, the U.S, and the surrounding region. I was able to give a presentation to the audience on the “Otologic Sequelae of Cleft Palate” and to discuss the need for improved hearing screening, as well as audiologic and otologic follow-up for these patients.

I was deeply moved by my experiences in the West Bank and feel grateful to have been a small part in this effort. I am grateful to the UNC Department of Otolaryngology, Dr. Pillsbury and the faculty, who allow and encourage such opportunities and for the great training I have obtained over the years such that I felt comfortable throughout my time in the West Bank. I look forward to continuing in similar efforts throughout my career, both in Palestine and elsewhere.
UNC audiologist, Dr. Pat Roush, accepted an invitation to present at the First Latin American Pediatric Audiology Conference in Sao Paulo, Brazil in August, 2007, where she gave two lectures; “Audiological Management of Infants and Young Children with Auditory Neuropathy/Dys-synchrony” and a second entitled: “Hearing Instrument Fitting in Infants: Practical Considerations and Challenges.” The meeting was attended by over 200 audiologists from several South American countries who came to learn more about the implementation of audiological services for young children with hearing loss identified through newborn hearing screening.

The trip included a fortuitous discovery when Pat learned, just prior to leaving for Brazil, that one of the other invited speakers, audiologist Dr. Beatriz Novares from the University of Sao Paulo, had worked with a young child, Zoe Gray, who is now being followed by Pat at UNC.

Zoe was born in Sao Paulo after the family moved there for her father’s work. She underwent a hearing screening shortly after birth, was referred for diagnostic audiology evaluation, and found to have severe bilateral sensorineural hearing loss. Hearing aids were fitted by Dr. Novares at two months of age, a remarkable achievement in South America where infant hearing screening and follow-up are not widely available. When Zoe’s family returned to the U.S. they moved to Charlotte and her parents arranged for her to be seen by Pat for continued audiological management.

Zoe was only a toddler when she left Brazil, so Pat brought a videotape of her reading a book aloud to her mother, to share with Dr. Novares and staff members who had worked with Zoe in Sao Paulo. Zoe is now in the first grade, has excellent speech and language skills, and is on par with her hearing peers in academic performance. None of this would have been possible only a few years ago. The advent of universal newborn hearing screening combined with the physiologic measures used by pediatric audiologists to define the type and degree of hearing loss at an early age and state of the art pediatric hearing aid fitting methods, have made it possible to begin auditory stimulation within weeks of birth.

The program in Sao Paulo also benefited from the generosity of Zoe’s parents, Sian and David, who contributed funds to help build a new clinic there. Pat enjoyed the opportunity to share her expertise with a dedicated team of colleagues committed to providing state-of-the-art services for infants and young children with congenital hearing loss.
5 Questions for Rupali Shah, MD

Rupali N. Shah, MD, agreed to be the first of a series of residents asked five questions in this new column in Heads Up. After earning her MD from Emory University School of Medicine, Dr. Shah joined our program and started her training in Otolaryngology/Head and Neck Surgery in June of 2007.

1. Why did you choose the field of Otolaryngology/Head and Neck Surgery?

In choosing a specialty, I was drawn to the versatility of Otolaryngology. From facial plastics to acute life-threatening airway obstruction, otolaryngology necessitates rapid adjustment to a wide spectrum of individual problems. It also allows working with a variety of patient populations from pediatrics to the elderly. Lastly, I was attracted to the ability of the otolaryngologist to be a complete doctor: making diagnoses, providing medical and surgical treatment, and maintaining patient relationships with continued care and follow-up.

2. What is your most rewarding experience so far as a first year resident?

While on the trauma service, an 18-year-old female came in to the Emergency Room after a motor vehicle accident with a transected pancreas, splenic laceration, and a pneumothorax. Her situation was life threatening. She went to the Operating Room emergently for removal of her pancreas and spleen. I was able to be involved in her care from the moment she arrived in the emergency room until her discharge. She was originally admitted to the ICU where her clinical status worsened before it got better. I followed her daily, and personally interacted with her and her family. I helped address their concerns while assisting in the care provided by the trauma team. She slowly began to improve and was eventually moved out of the intensive care unit. After a prolonged hospital stay, she had proven she was strong enough to go home. On Christmas Eve, I gave her the news, and she burst into tears with happiness and appreciation for the care the team had provided. The true reward was in watching a patient in a life threatening situation recover in time to spend the holidays with her friends and family.

3. What is the most stressful or difficult thing that has happened so far?

There are always patients who do not do as well as you hope. It is always hard to watch people not get better. During my first week of internship in the Burn Center, a 25-year-old gentleman had been struck by lightening. Despite multiple efforts at resuscitation, he passed within a few days. As the intern on call the evening he passed, I delivered the news to the family. Without extensive experience in this type of situation, it was pretty anxiety-provoking. I put into practice previous teachings on delivering bad news and learned by doing. Good communication and bedside manner with patients and family are an extremely important part of the job. With each new experience, I get a little more comfortable, but it is never easy to give bad news.

4. What has been the best thing about the UNC program so far?

I am very proud to be a part of the UNC Department of Otolaryngology. The residents and faculty have provided me with great support during my intern year. They have been approachable when I need advice or guidance with respect to everything from day to day administrative tips to help in formulating my research project for next year and grant writing.

5. What are you most looking forward to as you take on more responsibilities?

As I finish this year, I look forward to the rest of my training in becoming an otolaryngologist. Mostly, I hope to develop into a confident and capable surgeon that will provide good patient care with humility. In looking at my senior residents, I am reassured that I am in the right place with the right faculty to make that happen.
Would you trust your co-workers to physically and emotionally support you as you stood atop a 30-foot telephone poll? Members of the CCCDP and CASTLE team did just that, and much more, at a day-long retreat at the Carolina Challenge course on November 2nd, 2007.

The focus of the day was teamwork and collaboration for the purpose of furthering group and personal goals. The staff enjoyed a beautiful fall day of outside activities including chase games, balancing on a giant teeter-totter, and walking a rope bridge, but the highlight of the day was each person’s attempt to climb a 30-foot pole and jump off with the support of a harness held by co-workers.

At the end of the day, everyone had the opportunity to express their appreciation and respect for the roles fellow team members play in our working lives. A day of fresh air, along with sharing of personal joys and fears, was a refreshing and healing experience, which will help us to better address the needs of the children and families we strive to serve.

Megan Evans, Speech Language Pathologist at the Wilmington CASTLE, stands atop a 30-foot pole, ready to jump for “Tigger”, with the rest of the team holding the ropes to catch her. When she reached the top, she had trouble standing up on the pole, and was about to just sit, but she yelled, “I want to stand on this pole!” And so, she did! Not everyone made it.

This was an activity designed to simulate the process of getting a patient through the Implant Evaluation Process, using the team approach. We had to get a golf ball and then a marble (the patients) from point A to point B (the evaluation process) and into a cup (the implant surgery), using small troughs. Each member of the team had to run ahead to the end of the line after the ball passed by them to keep the process going.

Left: Francisca Hernandez, Teacher of the Deaf at the Wilmington CASTLE, climbs the pole while the rest of the CCCDP/CASTLE team braces themselves to catch her with the safety ropes and waits their turn to climb the pole and jump.
**Announcements**

*Just a few of the many exciting things happening in our Department*

**Anna Bradshaw Receives PlusPeople Award**

Anna Bradshaw was a recipient of a UNC Hospitals PlusPeople award in December of 2007. Anna works in the ENT Clinic as a Surgery Scheduler, coordinating Operating Room space and surgery times with our Otolaryngology/Head and Neck surgeons and their patients. She was nominated for the award because of the high quality of her work, devotion to her job and to the Department, her pleasant demeanor and willingness to work with faculty and staff, and the high level of care she shows towards patients. Anna’s job often involves problem solving, and she always considers the patient’s needs first. Our surgeons know they can count on Anna to utilize our OR space to the fullest. We all agree that she more than deserves this recognition. Congratulations, Anna!

**UNC Well Represented at International Pediatric Audiology Conference**

The Fourth International Pediatric Audiology Conference, held in Chicago in December, 2007, included keynote presentations by two Otolaryngology faculty. Dr. Craig Buchman’s presentation was entitled: “Hearing Loss: An Otologist’s Perspective.” Dr. Pat Roush, who directs the Pediatric Audiology Program at UNC Hospitals, gave a presentation entitled “Audiologic Management in Infancy: A Continuum of Care.” The international conference, known as “A Sound Foundation Through Early Amplification” and sponsored by Phonak, AG, in Switzerland, attracted over five hundred pediatric audiologists and related professionals from 30 nations. The conference was also attended by audiologists Corrine Macpherson, Sarah Martinho, and Paula Johnson, who specialize in pediatric audiology at UNC Hospitals.

Children with cochlear implants cannot wear their external speech processors when they are subject to static electricity. That means that plastic playground equipment is off-limits for deaf children wearing these devices. Our playground at CASTLE is in need of some updating and the staff at CCCDP and CASTLE are taking up the cause! We will be running in “The Great Human Race” in Durham on March 29th. Please visit their website to register and make a donation if you’d like to participate: [www.thevolunteercenter.org/ghr](http://www.thevolunteercenter.org/ghr)

Carlton J. Zdanski, MD, FACS, was promoted to Associate Professor on September 1, 2007. Dr. Zdanski is a pediatric otolaryngologist who joined the faculty in 2001. He is also the Surgical Director of the North Carolina Children’s Airway Center.

“A Multicenter U.S. Bilateral MED-EL Cochlear Implantation Study: Speech Perception over the First Year of Use” was published in *Ear and Hearing* in January 2008. With 22 co-authors and many more contributors, this project spanned 6 years from planning to publication. Co-investigators included members of the UNC Adult Cochlear Implant team, as well as colleagues from Vanderbilt University, University of Texas Southwestern, Carle Foundation Hospital, Dallas Otolaryngology, Medical College of Wisconsin, and Research Triangle Institute. The team from UNC included Harold Pillsbury, MD; Craig Buchman, MD; Emily Buss, PhD; Carol Pillsbury, MS; and Marcia Clark Adunka, AuD. Credit for the success of this project is also due to the study participants, who contributed many hours of their time to make this project a success, and MED-EL corporation, for providing the second device.

Comments, suggestions, or questions about *Heads Up?* Contact Elizabeth Perry, 919-966-8926, or eaperry@med.unc.edu.
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