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William F. House, DDS, MD—The Father of Neurotology

Published: August 29, 2011

This is an opinion editorial by Douglas L. Beck, AuD, Web Content Editor, American Academy of Audiology.

Editor's Note: As Web Content Editor for the American Academy of Audiology, I've been honored to meet and interview hundreds of peers and colleagues, scientists and authors and many others over the last few years (See interviews). However, in the case of Dr. William F. House, the traditional Q and A interview format doesn't provide enough information and latitude to truly represent his contributions to dentistry, otolaryngology, otology, neurotology and skull-base surgery, as well as the impact he's had on almost 200,000 people (globally) who use cochlear implants daily. Therefore, this article is written to present an overview of the amazing story of a contemporary genius, who I've been honored to call my friend for 27 years.

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I met Dr. House in the early 1980s when I secured my first professional job, right out of graduate school (The State University of New York at Buffalo). Throughout graduate school, I had been reading amazing and controversial stories about the not-yet-FDA-approved cochlear implant (CI) and the absolutely tremendous uproar associated with it. Although many people said it was clearly among the greatest ideas of all times, others said it was the work of the devil. Really, it was that contentious.

Professionals and lay people had very strong pro and con opinions, and apparently nobody was afraid to voice their concerns…the controversy was almost beyond description and in looking back on it now (in 2011), it's very hard to believe. Sort of reminds me about the vast professional divide regarding audiologists dispensing hearing aids (40 years ago) and the explosive issues surrounding the AuD degree (20 years ago).

Here's my recollection….

I was hired as a research audiologist at the House Ear Institute (HEI, now called the House Research Institute) in Los Angeles. Working at "House" was simply an extraordinary experience. I quickly learned there were three Drs. House; Howard P. House (the older half-brother of William), William F. House (the subject of this article) and John W. House (Howard's son.) As a natural consequence of having three doctors who answered to the name "Dr. House," we (the staff and others) referred to them by their first names to minimize confusion. Therefore, William F. House became forever known to many of us, as "Dr. Bill."

The 1980s were an exciting time at HEI. The cochlear implant was FDA approved for adults (1985) and was soon to be FDA approved for children (1990), we had computers in our labs (not a common finding in the 1980s) and we often had rock stars, movie stars, actors, comedians, dignitaries and politicians from across the globe visiting "the Institute." In fact, when a very important person (VIP) was about to tour the facility, we heard the page across the public address system (almost daily), something like, "Dr. Ready, please dial the operator," which meant "VIP in the building." We snapped to attention, poured the coffee down the sink, straightened ties and quickly got to where we were *supposed* to be.

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That's how I met Dr. Bill. He was walking through the research labs with some very famous Hollywood people. As the procession walked past my desk, Dr. Bill looked at my name tag, extended his hand and said "Hi Doug. It's a pleasure to meet you. My name is Bill House. Can you tell us about the work you do?"

I was floored. First of all, he's Bill House. Second, he introduced himself to me (yeah, like I didn't know who he was!). Third, the living legends standing next to him were amazing (sorry, can't say their name, probable violation of patient confidentiality). Fourth , all I was doing was running cochlear implant patients on our speech perception test protocol in a basement level lab. It didn't seem very exciting. I mean, don't get me wrong, it was very important, and we worked long and hard on bench-testing different processing schemes (such as the "high fi" strategy)…but it seemed pretty boring compared to the work Dr. Bill was doing!

After I caught my breath (I was just a simple country audiologist, not accustomed to being in the presence of the rich and famous), I explained we had to start by designing special speech-based tests for CI patients (as did many other CI groups across the world such as the groups in San Francisco, Utah, Melbourne, and more) because traditional tests used for hearing-impaired people didn't give us enough information about what CI patients were actually hearing.

One test I was particularly fond of was the "Danhauer-Beck Sentence Test" (DBST). The DBST has long since faded into history along with the Edsel, 8-Track tapes and New Coke. The DBST counter-balanced all English-language phonemes in third-order approximations, using male and female talker presentations. We had visual-only presentations (to determine the speech reading ability of each patient) auditory-only (simply turn off the picture on the TV!) and combined AV presentations. We used a Snellen eye chart to make sure our CI research patients has normal or corrected vision while presenting visual presentations of the DBST. I explained we needed to know much more about CI patients and their auditory perceptions in the real world. The goal was to experiment with different speech processing strategies to see which ones provided the most speech information to CI patients. That is, they all had the same internal components and we were changing the processing parameters of the external (speech processor) components.

I explained that we (sorry if I get the names wrong, it was a long time ago….Jeff Danhauer, Marc Pratarelli, Edward Cudahy, me and many, many others) had speculated about what seemed to work best in hearing aids

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(30 years ago) and tried to apply that knowledge to CIs. Specifically, it made sense to try to emphasize (band pass) the most important speech information (the second formant frequency) through the cochlear implant, so as to minimize background noise while maximizing consonant perception. Of course, the early processors were designed to deliver information from about 300 to 2700 Hz, so our hands were somewhat tied with respect to the amount of high frequency information we could deliver. Nonetheless, as we varied the spectral response, some of the patients achieved nearly perfect scores on auditory-only presentations (from the reel-to-reel tape recorder, patched through the bench-model prototype and plugged directly into their wearable CIs) using the "high fi" strategy. Others didn't. All of them had single-electrode CIs. I was amazed when one of the research patients (JA) *phoned* me to cancel her appointment one day—using her single electrode CI.

Dr. Bill paid attention to every word I said. He nodded at the right times and seemed very pleased with the work we were doing. Then he thanked *me*, and I got to shake hands with some really famous people. The procession walked on. They headed to the electrophysiology lab where Manny Don, Yvonne Sininger, and many other very bright people were working.

That same afternoon, Dr. Bill came back to the lab without the entourage and without "Dr. Ready." Jeff and I were reviewing our notes (Jeff's were always on a yellow legal pad) and Dr. Bill asked if we had any preliminary results. Jeff Danhauer, PhD, was (and is) a professor at UCSB and he was (at that time) a consultant to the HEI. Jeff was one of the guys that made sure the work we did was meritorious and had a strong scientific foundation. We (mostly Jeff) spoke with Dr. Bill for a very long time and he explained the details and his preliminary analysis. Dr. Bill listened carefully to every word he said, asked insightful questions and he appeared pleased with the discussion and the quality of the work.

I remember another time Dr. Bill came into the auditory research lab. He decided that to really understand what the audiologists were doing and to maximally help his patients, he should get a hearing aid dispensing license. Of course, he didn't *need* a hearing aid dispensing license and he assured us he had no intention of personally dispensing hearing aids, but he wanted to know more about hearing aids so he could better advise his patients. Fair enough.

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Donna Eskwitt and I (mostly Donna) worked with Dr. Bill with regard to hearing aids and a year or so later, he did indeed pass the exam and got his hearing aid dispensing license. I've never heard of any other neurotologist who did the same. When I met with Dr. Bill in August 2011, I reminded him about this story and the fact that he actually had to study and learn lots of new hearing and hearing aid information while he was developing surgical techniques and tools and while he was operating (many times daily) on people for cochlear implants and acoustic neuromas, and doing endolymphatic sac, vestibular neurectomy, glomus jugulare, and many other surgeries. I said "Dr. Bill, you really didn't need to get your hearing aid license in the middle of all that!" He said "*Now*you tell me!"

Dr. Bill's accomplishments are staggering.

In 1946, as a Navy dentist, he realized that sitting alongside a patient was fine while working on their lower teeth, but while working on the upper teeth, it made a lot of sense to have the patient recline. His captain cautioned that although having the patient lying down might be okay for males, the female patients might be concerned Dr. Bill would rape them! (Yes, the captain actually told Dr Bill that!). Of course, every dental patient across the world now has their teeth operated on while reclining. Another improvement he made with regard to dentistry was a diagnostic X-ray holder, which has been modified and remains in use today.
In 1948, Dr. Bill entered medical school and in 1953 he was awarded his MD. The antibiotic era had just started and many physicians were thinking that otolaryngology was about to become a profession of the past—very few people sought ENT residencies. In fact, he was the only applicant for an ENT residency at the Los Angeles County Hospital in 1953. In 1956, he finished his ENT residency at 32 years of age.

In the mid-1950s Howard House traveled to Germany and witnessed the use of a microscope in surgery. The microscope was originally designed for gynecologic cancer and Professor Wullstein had adapted the same for use in otology. Howard ordered the microscope and it became the first surgical microscope used in the United States. Dr. Bill took the microscope to the morgue in L.A. and he learned to use it while performing temporal bone dissections on unclaimed inhabitants. That was arguably the start of microsurgery in the United States.
Dr. Bill and a local engineer named Jack Urban worked together to design the "sidearm," which was the first operating microscope accessory that allowed an observer to look into the same microscope from alongside the surgeon to correctly (right side up) see what the surgeon was seeing. Of note, the surgeon's view was binocular,

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whereas the side arm was monocular and did not allow the observer depth perception. Years later, Jack Urban was the first to adapt TV cameras to the operating microscope. I suspect every operating room in the United States (and every major hospital across the globe) now has a version of the House-Urban microscope complete with TV viewing for observers for use in neurotology, otology, neurosurgery, orthopedics, and more.

Dr. Bill pioneered the "facial recess" approach. His goal was to improve the "state of the art" with regard to radical mastoidectomy—which was previously achieved via hammer and chisels to perform a canal wall down (ear canal obliterated) procedure to allow the infected mastoid to drain, thus avoiding a brain abcess. Prior to Dr. Bill's involvement with radical mastoidectomy, Lempert used a constant speed hand-held drill (held in both hands) to better control the mastoid dissection. Dr. Bill applied his dental knowledge and skills to the issue-at-hand and soon started using the bi-directional drill with a diamond stone to better control the fine dissection with regard to bone removal, while reducing blood loss and increasing facial nerve protection. Further, Dr. Bill designed the first hand-held suction-irrigator (i.e., the "House Irrigation-Suction"), which wets the bone dust (so as to not float through the room), keeps the bone cool (so as to not heat up the structures from the drill activity) and suctions out the wetness so as to maintain an excellent visual field within the operation. As usual, Dr. Bill honed his skills and "bench-tested" these ideas and thoughts in the morgue prior to applying them. When he did apply these new concepts to patients with ear disease, he developed the tympanoplasty. The facial recess approach is used today for many otologic and neurotologic procedures, such as the traditional approach for cochlear implantation and for facial nerve decompression in Bell's Palsy.

Dr. Bill saw many patients with Meniere's Disease, and there were very few treatments that consistently worked. Nonetheless, Dr. Bill read that in 1925 a French surgeon (George Portmann) had successfully performed an endolymphatic shunt, which "cured" the patient of Meniere's Disease. There were no other reports of this event and there were no reports of a second patient: back to the morgue! After working out the surgical approach, he tried it. It worked very well with regard to alleviating dizziness and the operation preserved hearing in about 70 percent of his patients. Of note, Dr. Bill did a subarachnoid shunt in which the endolymphatic sac was opened, a shunt was placed into the arachnoid space, and the endolymph was shunted into the spinal fluid. Many surgeons over the years have performed various modifications (mastoid shunt, sac decompression, etc.) and some have given up endolympatic sac surgey altogether in favor of vestibular nerve section for patients with intractable vertigo.

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The controversy over endolymphatic sac surgery remains. Ching, Fayad, Linthicum, and colleagues (2011) recently reported 15 cases of shunt surgery examined post-mortem. In five surgeries, the endolymphatic sac (ES) was not even exposed. Nonetheless, four of the five patients had reported relief of their vertigo. In eight surgeries, the ES was exposed, yet the shunt didn't reach the ES lumen and four (of the eight) had reported vertigo relief. The shunt had been successfully placed in the ES lumen of the two remaining patients, yet neither had experienced relief from vertigo.

Of note, Alan Shepard (astronaut and command pilot of Apollo 14) was disabled by Meniere's Disease. He was treated by Dr. Bill. Alan Shepard received an endolympahtic subarachnoid shunt—he was one of the first to have the operation—after which his hearing improved by 40 dB and he was returned to duty, eventually flying to the moon and back, symptom free. Without the operation, as Alan himself noted, he would have been grounded, likely never flying again, much less piloting an Apollo capsule to the moon.

The diagnosis of an acoustic neuroma (AN) back in the first half of the 20th century was more-or-less a death sentence. That is, by the time the tumor was large enough to be diagnosed (prior to CT and MRI) the tumors were often quite large, had already compressed the brain stem and produced an elevated intracranial pressure (often causing blindness and hearing loss) and their removal generally meant a sub-occipital approach that likely involved sacrificing the facial nerve—and as if that wasn't' bad enough—carried a 40 percent mortality rate.

Dr. Bill understood that if these tumors could be diagnosed sooner (i.e., when they were smaller), surgical removal would be more efficient. The first challenge was developing a patient-positioning radiologic technique that would allow pantopaque (a dye) to travel from a lumbar spinal injection site towards the internal auditory canal. Dr. Bill pioneered the technique despite outcry from many neurosurgeons that an inflammatory response was certain to occur. It did not.

Although years earlier, Dr. Bill had pioneered the middle fossa craniotomy approach (MFC) for otosclerosis (which didn't work out very well), he reasoned that by using the MFC approach to gain access to the entire internal auditory canal (IAC), he could follow the facial nerve into the tumor, while protecting the nerve and

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dissecting the tumor—which proved to be ideal for removing small acoustic tumors. The first removal of an acoustic neuroma via the MFC was in 1961. Eight tumors were removed using this approach by early 1962, but it was apparent larger tumors could not be successfully managed this way. Back to the morgue. Dr. Bill developed the translabyrinthine craniotomy (TLC) for the removal of larger tumors. Both techniques are still used today in operating rooms across the world.

By 1964, Dr. Bill had published his first monograph detailing the diagnosis and TLC surgical treatment of acoustic neuromas, reporting on 51 patients.

Despite the fact that Dr. Bill was working with Dr. Bill Hitselberger (a Harvard Medical School graduate and Mayo Clinic trained neuro-surgeon) and that they had spectacular results, the head of neurosurgery at the University of Southern California School of Medicine (who was also on staff at St Vincent's Hospital, Los Angeles) insisted that if Dr. Bill was allowed to admit and treat patients with brain tumors, that he (the head of neurosurgery) would resign his position at St. Vincent's. Dr. Howard House told the St Vincent's medical board that if Dr. Bill *wasn't* allowed to treat acoustic neuroma patients, Howard would resign. The executive committee decided in favor of Dr. Bill. The neurosurgeon resigned.

The first cochlear implants ever attempted and reported were done in 1957 (in France) by Drs. Journo and Eyries. Dr. Bill read about the procedure and quickly became enthralled with the concept. Dr. Bill did his first cochlear implants in 1961 (50 years ago!). Both patients reported perceiving sound, and although it was very exciting, the implants were not bio-compatible and eventually had to be removed. Dr. Bill did not pursue a patent on CIs as he did not want others to be restricted from developing these important devices. By 1968, biocompatible materials were available for pacemakers, hydrocephalus shunts and more…and it was time to apply these technologies to cochlear implants.

Dr. House asked Hallowell Davis for academic guidance and input on the CI project. Dr. Davis said he did not believe in CIs and he reportedly told Dr. Bill that working with Dr. Bill would "stain" his reputation. Merle Lawrence (Head of Kresge Labs, Ann Arbor) told Dr. Bill the CI was useless as the inner ear and hearing of those same patients was deteriorating and would be totally destroyed via electrical stimulation. Hal Schuknect,

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MD (Massachusetts General Hospital), told Dr. Bill that only a vast team of neuroanatomists, neurophysiologists, electrical engineers, and an enormous bankroll could make this a viable project.
Dr. Bill again enlisted the help of Jack Urban to work together to design the CI for the benefit of mankind. Jack never charged Dr. Bill a dime for his services. Dr. Bill never charged any of the patients.

The first device was a multi-electrode device, a five-electrode array insulated within medical grade silicone. Three patients were selected and implanted in 1969-1970. Over the next year or two, many permutations of electrode stimulation were attempted, compared and reviewed.

Of vast importance—an *amazing observation* was made, which ran exactly contrary to the well-known tonotopic (place) theory from von Bekesy, and is worthy of consideration, today. The tonotopic theory states that the basal end of the cochlea is where the high frequencies receptors are located and the apical end is where the low frequencies are located. von Bekesy theorized that as the mechanical traveling wave moved along the basilar membrane, areas along the basilar membrane are stimulated, correlating with the frequency of the stimuli. Therefore, it seemed likely that one should deliver the high frequency information to the basal end of the cochlea and the lower frequency sounds to the apical end.

In keeping with this theory, Dr. Bill and Jack Urban put together a five-electrode device, and of course, each electrode was placed into the patient's cochlea at a different spot along the spiral. However, stimulating all five electrodes simultaneously while routing the ground to a location outside the cochlea worked as well or better than stimulating pairs of electrodes along the basilar membrane. Dr. Bill found the best speech perception responses were obtained by stimulating the entire cochlea at once, and doing so allows use of a single short electrode (which promised to better preserve residual hearing) and required less total electrical energy.
Of course there were, and there remain, many people opposed to single electrode use, but as Dr. Bill wrote in his new (2011) book (page 82), "I did not and still do not believe that using a single electrode to deliver information to the hearing system necessarily limits the processing sense—any more than having a single eardrum to transmit hearing in the normal ear limits the ear's ability to understand a complex speech signal."

Here is a bit of his rationale:

Clearly, vast spectral information from sounds are transmitted through the normal tympanic membrane *simultaneously* and the brain working in tandem with the inner ear figures it out and makes sense of it. The brain is, of course, the ultimate speech processing computer and secondary to neural plasticity and the benefit of time and training, remarkable things can happen. Further, despite astonishing speech perception abilities with multi-electrode cochlear implants it is reasonably well known that people with CIs do not do remarkably well with pitch perception even *with* multiple electrode systems.

Want, Zhou & Xu (2011) recently reported their results with regard to pitch perception as measured in semi-tones (which might be thought of as inclusive of every key, black and white, on a piano keyboard). They reported on 19 CI users and 10 people with normal hearing. The CI users demonstrated a mean threshold for pitch discrimination of some 5.5 semitones. Again, to use the piano analogy, to detect the difference in two notes, they had to be as far apart (on average for the CI group) as the pitch change from A, to $B^{\flat}$, to B, to C and $C^{\natural}$. They reported CI users had an enormous range of performance from less than one semi-tone to 19.6 semitones. Normal hearing participants demonstrated average thresholds of 0.4 semitones. However, people do extraordinarily well with modern speech processors and cochlear implants – despite having very limited pitch perception, as shown by Want, Zhou and Xu.

Tuning curves presented by Kiang, Sachs and Peake (1967) indicate that individual feline auditory neurons fire across a range of range of frequencies and the range varies based on the intensity of the signal. That is, although the neurons may respond to a very narrow range of frequencies centered at 450 Hz when the stimulus is presented at a threshold or near-threshold intensity, with greater intensity, for example at 40 dB, it may respond to stimuli from 300 to 600 Hz. At still greater signal intensity, for example at 60 dB, may respond from 180 to 900 Hz.

Halpin (2002) noted "off-frequency" thresholds are well known and are supported by animal research, and inner hair cell sensitivity to acoustic energy from another frequency happens. Halpin noted that Kiang, Sachs and Peake reported each auditory nerve had a "best frequency" which required minimal energy for neural firing and there was also a "broad excitation area" and that off-frequency excitation required increasingly higher intensity. Further, Halpin notes most of the tuning characteristics of auditory neurons have to do with the inner hair cells, to which they're attached. Halpin notes "…every inner hair cell in the human cochlea responds to a wide range

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of frequencies, as long as the stimulus is made sufficiently intense…"(page 57). Further, he notes audiometric thresholds are based on a very limited binary detection protocol (present or absent), and patients are *able and expected*to detect pure tone stimuli from remote cochlear cells, if the local cells have failed.

Further, and of significant importance, remember that a single electrode does not imply that a "single frequency" is being delivered to the ear. Chute, Hellman, Parisier and Selesnick's (1990) reported on 126 children using CIs. The majority of the children had multi-electrode systems. The child referred to as "TL" was in the top three children with regard to speech perception – and none of the children achieved higher scores than she did. She wore a single electrode 3M/House cochlear implant with a modified processor. Imagine what might have happened if all the children with single electrode systems had advanced processors with a band pass range from 300 Hz to 6000/8000 Hz?

One last story…

As if the facial recess approach, reclining dental patients, cochlear implants, the translabyrinthine and middle fossa craniotomies, and the suction-irrigator and being the first person to introduce microsurgery to the USA, weren't enough, Dr. Bill invented the Auditory Brainstem Implant (ABI). It's use was (and remains) rare, limited to patients with bilateral acoustic neuromas, such as is often found in patients with von Recklinghausen's Disease (also called Neuro Fibramatosis II). Unfortunately, many of these people lose all their hearing secondary to tumor growth and removal. That is, they often lose both cochleas, rendering the cochlear implant useless. In one of these cases in 1979, Drs Bill House and Bill Hitselberger first placed an electrode directly on the cochlear nuclei of the brainstem. It was astonishing.

What was equally astonishing was something I learned 25 years later (in 2005) when I was chatting with Dr. Bill Hitselberger (known to many of us as at House as "Hitsy"). In his career, Hitsy operated on more than 6,000 skull base tumors.

I asked how the two of them (Drs. Bill & Hitsy) were able to get new procedures and protocols (like the auditory brainstem implant and the cochlear implant) approved by the hospital's Institutional Review Board (IRB).

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Hitsy smiled, looked around the room, waited a little bit and said…"I thought Dr. Bill talked to them." Dr Bill smiled and said, "I thought Hitsy got it cleared."

I don't know the answers to the question about single versus multiple electrode cochlear implant systems, but the fact that Dr. Bill is still asking these questions at age 87 after studying the literature for 50 years makes me wonder about it, too.

In failing to widely develop single electrode cochlear implants, I wonder, did we throw out the baby with the bathwater? That is, perhaps there should be multiple solutions (relatively inexpensive single-electrode and tremendously sophisticated multi-electrode solutions) to address this vast problem. Multiple electrode cochlear implants are excellent, highly sophisticated and provide fantastic results - yet they are expensive. Unfortunately, the vast majority of CI candiates (across the globe) are poor. The evidence is strong that simple, inexpensive, single electrode cochlear implants could provide significant benefit to those who will never be able to afford "the standard of care." Perhaps commercial availablity of both systems would underscore Dr. Bill's long years of service to mankind--and his goal, "so all may hear."

Dr. Bill and I were chatting late in the spring of 2011 and he invited me to visit him on August 20, 2011, in Oregon. Nothing on the planet could've prevented me from being there. Thanks Dr. Bill. You are an amazing person, an inspiration, an example of "The Right Stuff," and I am (and have always been) so proud to call you my friend. Hope to see you again, soon!

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