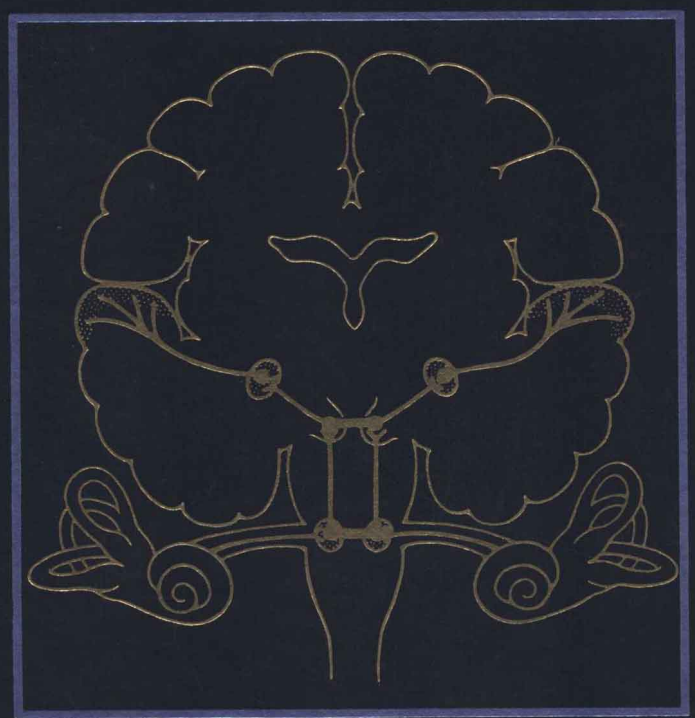
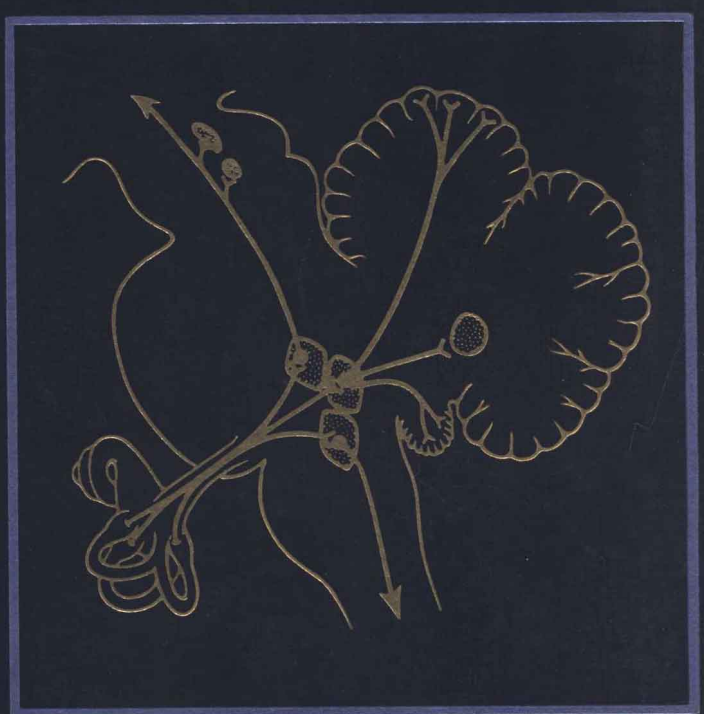
NEUROTOLOGY





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Neurotology

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P R E F A C E

Neurotology is a relatively new clinical field that is undergoing rapid evolution. In a simple definition, neurotology comprises the neurology and neurosurgery of the ear. Much as neuro-ophthalmology has become well-recognized as a subspecialty of ophthalmology, in recent years neurotology has become established as an area of special expertise within otolaryngology. The field of neurotology has achieved a considerable measure of maturity in recent years, in large part through the efforts of an ever-increasing number of intelligent, energetic physicians and scientists who have made it their life's work. As the body of knowledge has expanded geometrically, both the scope of clinical practice and the issues that deserve attention through research have become increasingly well-defined. The creation of postresidency fellowships in neurotology has done much to disseminate neurotologic expertise and foster excellence in the field.

Our goal in organizing this book was to provide the field with its first comprehensive text. Until now, clinicians who wished to attain expertise in neurotology have had no alternative but to digest large files of journal reprint material. Many important clinical and basic science subjects have seldom, if ever, been summarized in review articles. Because interest in neurotology cuts across many specialties, articles on any given neurotologic topic are often far flung throughout the medical literature. The text is intended to serve as a comprehensive resource for neurotologists and other interested clinicians (e.g., neurologists, neurosurgeons, ophthalmologists, audiologists, etc.) on the state-of-the-art in medical and surgical neurotology. Although the book is comprehensive, efforts have been made to avoid exhaustive treatments of the subject in order to improve readability and make the material stimulating for those who wish to read it "cover to cover." The 84 chapters were authored by foremost experts in the field who were selected not only for their expertise, but also for their literary ability. Efforts were made to ensure the proper s
of coverage as well as to achieve as much
style as can be obtained in a multiauth
editors with to thank the one-hundred t
tributors for their arduous efforts and gooc
the editorial process. Special thanks ar
Christine Gralapp, medical artist, for her
tions, which depict complex neurotolo
with remarkable clarity and realism. Her
will be featured in a full-color atlas of
niques in neurotology and cranial base su
in preparation as a companion to this text

Robert
Derald E. B

It is with great pleasure that we dedicate this book to William F. House, M.D. Very few are fortunate enough to know someone who has changed the course of his particular field as Bill has. We have had the good fortune not only to be under his professional tutelage, but also to know him well personally.

Upon completion of his residency, Bill joined his brother Howard in his busy otologic practice. He first concentrated on middle ear disease and developed the facial recess approach for the management of the mastoid in chronic otitis media. Soon thereafter, he decided that the future of otology lay in the treatment of diseases of the inner ear.

Bill introduced the operating microscope and otologic techniques to the field of neurosurgery in first developing the middle fossa approach and then the translabyrinthine approach for the treatment of acoustic tumors. The advantage of the microscope and microsurgical techniques was apparent, and the mortality rate for acoustic tumor removal fell dramatically regardless of the approach employed. Preservation of the facial nerve became the rule rather than the exception.

These two basic approaches were expanded to the extended middle fossa approach and the transcochlear approach for tumors involving the skull base. At about the same time, as if his other activities were not enough to keep him occupied, Bill developed an interest in the cochlear implant. A patient brought a report of Djourno and Eyries to him and, with Jack Urban, he worked tirelessly for many years to perfect a clinically applicable cochlear implant. Bill currently continues his work on the cochlear implant.

His latest achievement is the development of the auditory brainstem implant for patients who do not have an auditory nerve that can be stimulated. Bill Hitselberger has been Bill's coworker on this project, along with many others.

Bill's creative genius is equaled by his personal qualities. He has unselfishly trained a whole generation of colleagues in the field of neurotology. Whenever a new technique or device was developed, Bill would immediately disseminate the information to others. His goal was always to improve patient care.

The patient's welfare was always primary in Bill's mind. We have seen him anguish when a patient was not doing well and celebrate with the patient when all was going well.

It is with great respect and admiration that we dedicate this book to Dr. William House, the father of neurotology.

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F O R E W O R D

THE NEUROTOLOGY SAGA: A PERSONAL PERSPECTIVE

During the past few years, I have heard myself introduced on occasion as the "Father of Neurotology." If this is true, then it is also true in my case that being this kind of a father is not something that was planned; it just happened.

In looking back, I realize now that it all started during my third and last year of ear, nose, and throat (ENT) residency at Los Angeles County Hospital. The year was 1955. I was 31 years old. By then June and I had been married for 10 years, and Karen was 8 and David 7. I had completed dental school, served 2 years in the Navy as a dentist, finished medical school, and taken the ENT residency so that I could become a plastic surgeon. June was helping to hold the household together by working part time in my brother Howard's office as an RN.

ENT was not a sought-after residency at that time because it was widely believed that penicillin was eliminating the sinus, mastoid and tonsil, and adenoid problems that occupied the eye, ear, and nose specialists. In fact, we had so few residents that I was obliged to be on-call at the hospital every other night. Fortunately, the library was well-stocked so I had time to read the latest ENT literature.

There was no full-time ENT faculty, but there was usually an attending physician to help in the clinics and in surgery several times a week. As I look back, I can now call the residency a *learning residency* instead of a *teaching residency.*

During the last year of my residency much of my learning came through Howard, who (being 15 years older than I) had finished the 2-year Los Angeles County residency in 1939 and had established a large practice that was 95% otology. He was doing seven or eight fenestration operations a week; this included Saturday morning surgery. In addition to this, he usually had two or three ENT doctors taking a 1-month course from him in otology. The students would observe surgery during the day and come to the County Hospital morgue at night to do fenestrations on cadavers. I would often help in the morgue, and Howard encouraged me to do cadaver head and neck dissection and surgical procedures. On Saturdays, I often observed his surgery.

Howard also introduced me to a very remarkable, older ENT practitioner, Gilbert Roy Owen. He had become very interested in temporal bone and sinus x-ray. As time permitted I would go to his office, and he would take me through his remarkable collection of x-ray pathology. One of the things he repeatedly showed me was the enlarged internal auditory canal of acoustic tumors. I remember going to the library and reading as much as I could find on acoustic tumors. The recurring theme was that these were serious, although benign, lesions that should be operated on as early as possible.

During the last few months of my residency, a remarkable event occurred that was to change my life. Howard had heard of some interesting work going on in Germany called "tympanoplasty." He visited Dr. Wullstein in Wurtsburg and for the first time saw the Zeiss microscope. He immediately ordered a microscope (I believe for the large sum of $2,000) and invited Dr. Wullstein to come to Los Angeles to demonstrate his techniques. It was my job to chauffeur the doctor to his demonstrations. The films that he showed of temporal bone surgery through the microscope were astounding to me in terms of what you could see of the temporal bone structures over what we had been seeing with the headlight and loops. He took these films by working to a point in the tympanoplasty and then swinging in a microscope with a camera mounted on one eyepiece. While he worked through the other eyepiece, he filmed the procedure.

When Howard's microscope arrived he began using it in the new stapes mobilization procedures, and I would take the microscope to the morgue at night to explore the wonders of the temporal bone.

On completion of my residency in July 1956, I joined Howard in his office. By then I had become fascinated with otology, and—because of Howard's practice—I was able to spend all my time in otology. The first few hours of each day were spent making the rounds of several Los Angeles hospitals to change the dressings of Howard's numerous fenestration patients. The remainder of the day

was spent in the office cleaning fenestration cavities and seeing a never-ending stream of otology patients.

STARTING PRACTICE AND DEVELOPMENT OF THE FACIAL RECESS APPROACH

It was an exciting time to be starting an otology practice. Wulstein and Zolner had introduced "tympanoplasty" surgery using a Zeiss microscope a few years earlier, and Howard was doing a few skin grafts to the middle ear at the time of mastoid surgery. John Shea (whom I had gotten to know during my residency because he had spent some time with Howard learning fenestration surgery) had boldly introduced stapedectomy because so many of the stapes mobilization procedures that had been introduced by Rosen a couple of years before were refixing. Howard, who for years had been doing 10 or 12 fenestrations a week, asked John to come to Los Angeles and demonstrate his revolutionary stapes removal operation. As the word spread of the remarkable results these stapedectomy patients were getting, the patients came flocking to Howard's office. I was immediately busy working up these patients and getting them on Howard's surgery schedule.

It soon became apparent that Howard had no time for anything else except otosclerosis surgery. No one wanted the kid brother to do their stapes surgery. Many of the patients, hoping to get their hearing restored by this new miracle surgery, turned out to be chronic ear patients. After all, antibiotics except for sulfa drugs had only been in widespread use for 10 years. Howard was more than willing to turn these patients over to me and encouraged them to have me do their mastoid surgery.

During my residency, the goal of radical and modified radical mastoid surgery was to open the mastoid to allow it to drain rather than back up into a brain abscess. Antibiotics and tympanoplasty procedures using the microscope were challenging these concepts that had been the standard of practice for the past 75 years. The microscope made it possible to see the facial nerve more clearly and, therefore, allowed much more complete removal of cholesteatoma and granulation from the middle ear rather than leaving it wherever the facial nerve might be. Indeed, I remember my instructors during my residency telling me that if you so much as touch the facial nerve, it will become paralyzed.

Tympanoplasty procedures were now advocating grafting over the middle ear with skin and, thus, violating the leave open for drainage principal. It was soon found that grafting over infected granulation even with vigorous postoperative antibiotics was unsuccessful. It became obvious to me that if all the middle ear granulation and cholesteatoma was to be removed it was necessary to know where the facial nerve was and treat it as a friendly landmark. The microscope made this possible through identifying the horizontal canal and lifting the granulation to locate the facial nerve in the tympanic segment. Visualization was also enhanced by developing continuous irrigation suction where the amount of water flow and suction were controlled by rotating the thumb over the suction hole. Until then, mastoid surgery had been done by drilling using two hands on the drill as taught by Lempert; then irrigating and suctioning. This was a slow, tedious process, and I remember taking 3 or 4 hours to do a mastoid in the laborious way. However, using one hand on the drill and one hand on the suction was sometimes viewed as reckless surgery. However, Howard let me do it and often came to my defense in discussions with colleagues.

The principal of chronic ear surgery thus changed to remove the disease and cure the infection. The tympanoplasty grafts were now much more successful, but there were still problems. Mastoid cavities are subject to accumulation of dibris and recurrent infection. It was discouraging to see a nice tympanoplasty result, with some improvement in hearing, be wiped out by a recurrent infection that destroyed the graft. To overcome this problem, many different mastoid obliteration procedures such as swinging in muscle from the temporalis, and various plastic and tissue inserts were advocated. I tried all of these procedures and found them all to be wanting.

It became obvious to me that the best answer was to avoid creating a cavity. After all, we were no longer simply opening things for drainage; we were now after a cure for the infection. But if we left the posterior boney ear canal intact, could we see well enough to remove all the infected tissue in the middle ear? The answer was "no," so it was back to the dissection lab again. I had seen Wullstein on his type I tympanoplasties, that is, those with an intact ossicular chain drill a control hole from the mastoid just lateral to the facial nerve and ending in the middle ear. He called this a "control hole" and was able to visualize through this disease in the middle ear. This was done quite blindly, and it frightened me because I could visualize a good chance of hitting the mastoid part of the facial nerve. In the dissection lab, I learned how to skeletonize the mastoid part of the facial just inferior to the horizontal canal and open the area widely for good visualization of the stapes incus and posterior part of the middle ear. The chorda tympani nerve and the annulus of the ear drum were used as landmarks. I named this the "facial recess approach" because I wanted to emphasize the facial nerve as the basic landmark.

Fortunately, this approach is now widely used and has become the standard in cochlear implant surgery. However, leaving the canal wall intact led to new prob-

lems. Wullstein had advocated four types of tympanoplasty. Type 1 would now be called a myringplasty because the ossicular chain is intact. Type 2 was a graft to the head of the stapes. Type 3 was a graft to the promintory to leave the oval window open if the stapes superstructure was gone. Type 4 was a fenestration of the horizontal canal if the oval window was obliterated. It is obvious that these procedures were designed to avoid reconstructing the ossicular chain. Leaving the canal wall intact made it necessary to reconstruct the ossicular chain since the graft was now in the location of the previous ear drum. As intact canal wall procedures became more widely used, a number of otologists, including myself, devised a number of prosthetic reconstruction procedures.

Such reconstruction of the ossicular chain led to another problem. If the middle ear did not become aerated, the hearing result was poor and the graft would adhere to the promintory. There would also be a retraction of the graft into the attic and a new cholesteatoma formation. I tried a number of procedures to avoid this retraction, including placing wire mesh in the attic to prevent the retraction. I called the procedure the "iron curtain procedure." Months later, to my horror, if the middle ear did not aerate I saw the skin retract through the mesh and, thus, become a worse problem than the original cholesteatoma. I learned that aeration, not obliteration, is essential for successful chronic ear surgery.

I told the story of my experiences with the development of chronic ear surgery in a book dedicated to neurotology to illustrate how temporal bone surgery had developed after the microscope was introduced. The use of amplification, continuous suction irrigation, and use of the facial nerve as a landmark allowed us to develop temporal bone procedures to move through the temporal bone with dispatch and, thus, develop the next generation of temporal bone surgery. The retrolabyrinthine, translabyrinthine, transcochlear, and middle fossa approaches would not have been possible before today's chronic ear surgery was developed.

DEVELOPMENT OF THE MIDDLE FOSSA APPROACH

Obviously, otosclerosis has been a big part of my life. To find out more about it, I read a two-volume series of articles on otosclerosis that had been collected by the American Otologic Society. One of the articles that caught my eye was a study that detailed how otosclerosis lesions commonly occurred around the cochlea above the internal auditory canal and compressed the cochlear nerve. This was theorized to be a possible cause for the sensorineural loss that I was frequently seeing in otosclerotic patients. It seemed logical to me that if you could drill away the otosclerosis and relieve the pressure on the eighth nerve, you might reverse some of the hearing loss.

I have always been an early riser, and I remember sitting one morning and looking at a dissected skull that my father had given me in dental school. There was a yellow line for the greater superficial petrosal nerve, and it occurred to me that this might be the key to follow back to the geniculate ganglion and then on along the labyrinthine part of the facial to the internal auditory canal.

By this time we could afford a babysitter, so June and I started going to the morgue to see if I could get to the internal auditory canal without damaging the hearing or the facial nerve. Because of my experience with diamond burrs and irrigation in dentistry, I had already adapted these procedures to my work in chronic ear surgery. June would set up the microscope and instruments and act as the scrub nurse to facilitate the dissection. Since the approach called for elevation of the temporal lobe, I recruited the help of a young neurosurgeon, Ted Kurze, and after a number of dissections I felt we were ready for our first case.

For this case, I selected an attorney who had changed careers to become and accountant after he went completely deaf. His medical records clearly showed that he had otosclerosis. On August 1, 1958, June's birthday, we did the first middle fossa decompression at St. Vincent Hospital in Los Angeles. I was very honored and not a little scared that Dr. Carl Rand, the dean of Los Angeles neurosurgery, Cushing's last resident, and Dr. Kurze's associate, came to watch the surgery.

As far as I know, this was the first intracranial procedure in which the operating microscope was used. During the procedure, using the old Jordan Day drill with belt-driven engine arm and hand-piece, enough static electricity developed that it caused frequent stimulation of the facial nerve. I could feel facial contractions through the drapes. I had experienced this a number of times before during chronic ear surgery, so it was not an unknown phenomena to me, but I was already so nervous that I came very close to aborting the procedure.

Fortunately, the patient recovered well, with no facial weakness. I remember his wife telling me how sexy she thought he looked with his completely shaved head. Unfortunately, he did not recover any hearing. Some years later he was to become one of my first cochlear implant patients.

DEVELOPMENT OF ACOUSTIC NEUROMA SURGERY

My discouragement because the operation did not recover any hearing was offset by my realization that the middle fossa approach had a number of other possibili-

ties, such as vestibular nerve section and identification of the facial nerve during acoustic tumor surgery.

Early in practice I had seen a very handsome young fireman with a unilateral hearing loss. I sent him to Dr. Owen for x-rays. The report came back that he had an enlarged internal auditory canal. I referred him to Dr. Kurze who concurred with my diagnosis of an acoustic neuroma. He told me that he did not want to operate at that time because the patient would trade a little hearing loss and tinnitus for certain facial paralysis and possibly ataxia. Within 2 years he had developed facial numbness and early papilledema. I attended the surgery, which was performed with the patient in the sitting position and took some hours. Unfortunately, the patient stopped spontaneous respiration and died several days later. In a later discussion with Dr. Kurze, we both agreed that he had done all he could. I will never forget what he said, "You have to realize we were dealing with a large tumor."

I realized in the aftermath of the loss of this patient that the key to early acoustic tumor surgery was preservation of the facial nerve. I remember that I dreaded seeing and evaluating patients with unilateral hearing loss because I felt that if a diagnosis of acoustic neuroma was established the patient's doom was sealed.

June and I continued our sessions in the morgue. I was trying to work out an approach to the cerebellopontine angle through the middle fossa. The concept was to identify the facial nerve at the beginning of the procedure and then to dissect the acoustic tumor away from it. I had never operated on a acoustic tumor, but I teamed up with a young neurosurgeon, Jack Doyle, who had just finished his residency at the Mayo Clinic. We did our first acoustic in January 1960, using the microscope, with the patient in the sitting position. Drilling out the labyrinth and internal auditory canal down to the jugular bulb using a slow Jordan Day drill is a long and very tedious procedure. Jack Urban, a fantastic engineer who died some years ago and whom I still miss very much, helped me develop a special retractor and a seat with arm rests. It was a partial removal. The patient had some facial weakness but recovered well. During the next 3 years, we were to do another 20 tumors this way. The histories and the results are chronicled in our first monograph.

DEVELOPMENT OF THE TRANSLABYRINTHINE APPROACH

Operating in the sitting position and removing the labyrinth through the middle fossa was very onerous to me. My experience with mastoid surgery, with the patient prone on the table, seemed to present some interesting possibilities. So it was back to the dissection lab and to investigations on how to remove the labyrinth and open the internal auditory canal using mastoid procedures. I soon found that the facial nerve could be skeletonized, the labyrinth removed, and the internal auditory canal opened, without having to retract any brain or drill away any bone with the dura open.

Fortunately at this time, Bill Hitselberger came into my life and, for the first time, I could work with a neurosurgeon who really wanted to learn temporal bone surgery and be able to apply the expertise of neurosurgery to the problems of acoustic neuroma surgery. We soon recognized that it was safer and much easier to operate with the patient in a supine position and to approach the angle through the mastoid and the labyrinth. This eliminated the constant worry of air embolism and considerably shortened the dissection down to the angle.

By now, I too, had begun teaching in Howard's courses, and many students wanted to learn the temporal bone approaches. One of these students was Frank Ellis from Sydney, Australia. During each night of dissection, I would emphasize that the key to establishing the exact location of the facial was to identify it at the beginning of the tumor dissection at the point where it entered the fallopian canal at the lateral end of the internal auditory canal. I would say, "Frank you've got to see that bar of bone (the vertical crest) at the end of the canal." It was Frank who dubbed it Bill's bar, a name that has stuck. It is the key to saving the facial nerve and making early acoustic neuroma removal possible.

Bill Hitselberger and I developed a very close working relationship and, when faced with a complication of death, we would carefully explore what we should have done differently or return to the morgue for a new look at a particular part of the surgery. We were backed up by Jack Urban who often observed surgery and developed instrumentation, microscope viewing tubes, and camera and television equipment. It seemed like he could do it all, if it involved engineering.

Over the next few years, we did a number of acoustic neuroma surgeries using the translabyrinthine approach. After 50 cases, we decided it was time to publish our results. I was very impressed with Cushing's volume, published in 1917. Each patient that he had operated on up to that time was documented in detail and in sequence. I tried to emulate this example by publishing each of my cases in the same way. Fortunately, Dr. George Shambaugh, who was at that time the editor of the Archives of Otolaryngology, suggested that we devote an entire issue of the journal to these cases. This issue was the first significant recognition of this work, and I shall always be grateful to him. It set forth clearly the value of microsurgery in acoustic tumor surgery. This heralded a very significant change in intracranial surgery and was not met with enthusiasm by the neurosurgical community.