Surgery for Cochlear and Other Auditory Implants

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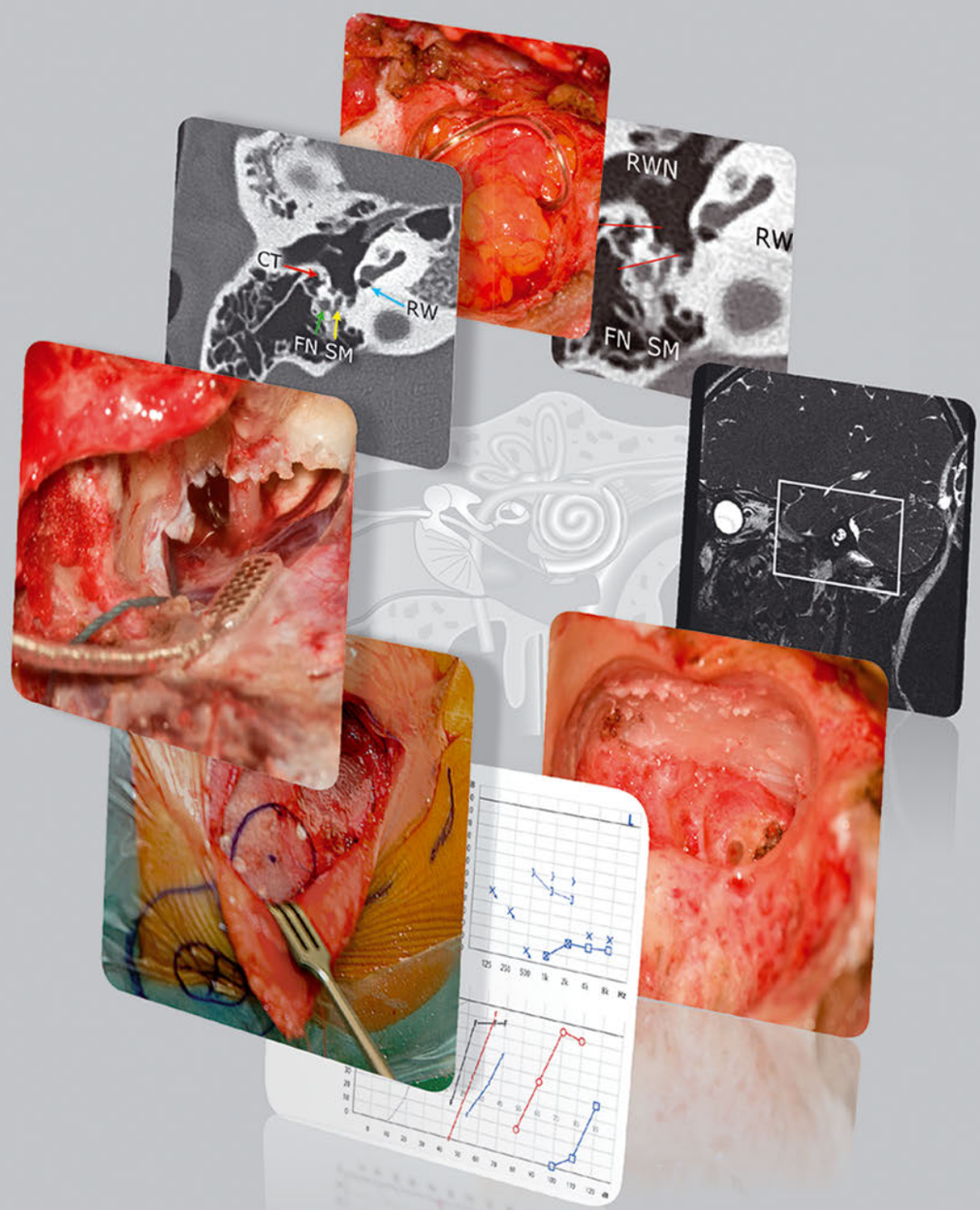

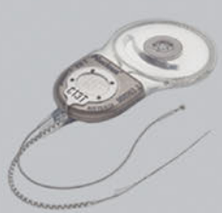

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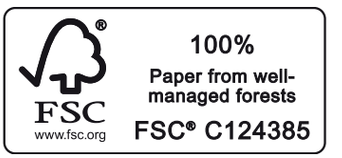

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Preface

Perhaps no other branch in medicine has made such rapid developments in such a short span of time as that of hearing implantology and rehabilitation. In the little more than half a century since the first cochlear implant was devised and placed in the round window by William House in 1961, we have come a long way into the era of auditory brainstem implants and a range of other implantable hearing devices. Giant strides have been made both in the understanding of deafness and in the technology used to treat it. Researchers have published exhaustively on this subject, more so in the last decade. It was in the light of this that we decided to compile this book, and I am extremely pleased to see *Surgery for Cochlear and Other Auditory Implants* come to fruition. It has been the collective effort of my team at the Gruppo Otologico in Piacenza and Rome that has made it possible. Above all, this book is a tribute to the legendary William House—surgeon, inventor, and teacher par excellence. It is due to his pioneering work in the science of implantology, including the cochlear implant and the auditory brainstem implant, that we are what we are. There is a tendency to forget the memory of the giants of the past on whose shoulders we stand today. This book is a reminder to all its readers of the genius and tireless efforts of remarkable physicians and surgeons like William House and many other before and after him.

This book is based on our vast experience of over 20 years with cochlear implants, auditory brainstem implants, and other implantable hearing rehabilitation devices that were developed during that time. The book is targeted at the otological surgeons keen to pursue the discipline of hearing implantology; hence the main focus of the book is on the surgical aspects of hearing implantology and not on electrophysiology and rehabilitation, for which there are other excellent textbooks already available. Having said that, we have left no stone unturned to include all the possible facets relating to surgery, making this book an exhaustive surgical compilation. This includes preoperative protocols, radiology, decision making, intraoperative techniques, difficult surgical situations, complications, and special considerations in implantology. The text describes surgery of all the implantable devices available today including cochlear implants, auditory brainstem implants, active middle ear implants, and bone anchored hearing devices. It also discusses a wide range of surgical situations from routine indications to more challenging ones such as inner ear malformations, congenital absence of the cochlear nerves, implantation in neurofibromatosis (NF) II, implantation in the only hearing ear in solitary vestibular schwannomas, and implantation in chronically discharging ear and in cases with CSF leaks. Special emphasis is given to the radiology of inner ear malformations. The role of subtotal petrosectomy in implantation is discussed in detail. A special chapter is written on single-sided deafness after surgery and its rehabilitation with bone anchored hearing aids and cross hearing. Algorithms have been developed to lay out the decision making process in various situations so as to make it easy for the reader to comprehend. Finally, all this is laid out in a pictorial format with high-quality illustrations that has been popular with the readers of our earlier textbooks and has come to be the hallmark of books from our stable.

The support of Cochlear, Med-El , Advanced Bionics, Neurelec, Oticon Medical, and Sophono has been vital in the making of this book and I thank them. I also thank Paul Merkus and Rolien Free from the Netherlands for their valuable help and coordination with the members of the Gruppo Otologico. I thank Thomas Roland from the United States for participating in this book. Last but not the least I thank my coauthors who worked tirelessly on this book for the last couple of years to produce a very satisfying product.

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About the Authors

Mario Sanna is Professor and Director of the Otolaryngology Clinic at the University of Cheiti, Italy and the Founder & Director of the Gruppo Otologico (Piacenza-Rome, Italy), an internationally acclaimed center for otology, hearing implantology, and skull base surgeries. Professor Sanna is a master in his field, known across the world for his pioneering work in otology and skull base surgery. He is a founder member of the European Skull Base Society and the Italian Skull Base Society and an honorary member of the French and Spanish otolaryngological societies.

Professor Sanna has authored 15 books and over 375 articles in peer-reviewed journals. His illustrated textbooks are extremely popular and some of them have won international awards. He has been an invited faculty in over 200 conferences and academic meetings. Over 100 fellows and 500 observers have been trained at the Gruppo Otologico. Professor Sanna has contributed tremendously to the knowledge of otology and skull base surgery and is considered by his peers to be one of the pioneers of modern skull base surgery.

Paul Merkus is a Dutch ENT surgeon (VUmc Amsterdam, the Netherlands) with longstanding experience in adult and pediatric cochlear implant surgery. He has published several papers on cochlear implantation and auditory brainstem surgery and his team is expert in decision making in difficult cases. His knowledge in imaging and ear surgery and his educational talents are well evidenced in this book.

Rolien Free is a Dutch ENT surgeon, who was trained at the University Medical Center Groningen, the Netherlands by Professor Frans Albers. She specialized in otology/neurotology and pediatric ENT surgery and has published several papers on cochlear implantation, otology, and pediatric ENT. Since 2010 she is codirector of the Cochlear Implant Team Northern Netherlands. She has an extensive experience of 10 years in both adult and pediatric cochlear implantation surgery. In 2011 and 2012 she worked as a neuro-otological fellow in Professor Mario Sanna's Gruppo Otologico, Piacenza, Italy. The neuro-otological stipendiums of the Heinsius Houbolt Foundation (Wassenaar, the Netherlands) and the A.I.N.O.T. (Associazione Italiana Neuro-Otologica, Italy) made these fellowships possible. She has tutored several years with Professor Ugo Fisch in his otology and skull base microsurgery courses in Zurich, Switzerland and is tutoring in the cochlear implant and advanced temporal bone dissection course led by Professor Louis Hofmeyr in Pretoria, South Africa. She is currently secretary of CI-ON (National Committee of Dutch Cochlear Implant Teams) and of the Dutch Flemish Society for Pediatric Otorhinolaryngology.

Maurizio Falcioni is an Italian ENT surgeon with more than 20 years' experience in middle ear and skull base surgery. He has published several papers on cochlear implantation and auditory brainstem surgery. His experience is particularly focused on cochlear implantation in difficult cases (concomitant chronic otitis, cochlear ossification, malformation).

1 History of Auditory Implantation

Studying the history of auditory implantation and following the developments that have taken place in only some 50 years is exciting and teaches us about the courage, vision, and endurance of some special individuals in and from different parts of the globe. It also makes us realize that the cochlear implant (CI) has been developed through multidisciplinary efforts and would not have existed if other developments had not paved the way; for example, without the experience with silicone-covered leads in pacemakers, biocompatibility studies, and the existence of antibiotics, this development would not have been possible. The reactions of disbelief and skepticism in the community of auditory professionals, and later the reactions of anxiety and anger in the deaf community to these new developments—which felt like a threat and an offense to their culture and way of living—also demonstrate the sociological impact that the cochlear implant has had and sometimes still has. Finally, this chain of development could not have taken place without serendipity lending a hand, and without some very courageous patients who decided in close consultation with their doctors to become objects of research for this possible new treatment.

Five Eras in the Development of Auditory Implants

- Forerunners
- Pioneers and experimentation: 1957–1960s
- Feasibility studies, safety studies, evaluation of auditory gain: 1970s
- Development of the commercial multielectrode cochlear implant: 1980s
- Development of the auditory brainstem implant: 1990s

1.1 Forerunners

1.1.1 Alessandro Volta (1745–1827) (Fig. 1.1)

Interest in stimulating hearing by use of electricity started in 1790 when Alessandro Volta, an Italian physicist and Count of the region of Lombardia in northern Italy, stimulated his own auditory system by connecting a battery with potential difference of ~50 volts to two metal rods that were inserted into his ears. Apparently he heard a "boom" within his head and then a hissing sound as if "soup was boiling"; he found the experience quite uncomfortable and moreover it lacked tonal quality!

Volta had had an interest in electricity from a very young age and at the age of 18 years was already corresponding with various academics and scientists on the subject. He invented the electrical battery (the voltaic cell; 1800), and also developed an interest in chemistry. He was also the person who discovered methane in a nearby swamp as a gas that could be used as fuel (1778). The son of a family of nine children who was expected to become a priest like five of his siblings, Alessandro Volta contributed significantly as a scientist and became professor in physics at the University of Pavia in 1779 and professor in philosophy at Padova (Padua) in 1815. In his recognition, the unit of potential difference (or "voltage")—the *volt*—has been named after him since 1881.

Fig. 1.1 Alessandro Volta with his voltaic cell.

Some other experiments involving electrical stimulation of the auditory system were described in those years, all without tonal quality of sound. In 1855 Duchenne of Boulogne (1806–1875), based in Paris, stimulated the ear with alternating current instead of direct current, leading to a sensation of "the beating of a fly's wings"; still an unsatisfactory outcome.

Several discoveries in the early 20th century (around the 1930s), the era of the development of the telephone, influenced the final development of the auditory implant.[1]

1.1.2 Homer Dudley: the Vocoder

Working as a researcher at the Bell Telephone Laboratories in New York, Dudley described and designed in 1939 a real-time voice synthesizer that produced intelligible speech. The fundamental frequency of speech, the intensity of its spectral components, and

its overall power could be extracted using a specially designed circuit. This synthesizer was named the *vocoder* (encoding the voice) and its operating principles formed the basis for the early speech processing schemes in auditory implants.[2]

1.1.3 Wever and Bray: the Cochlear Microphonic

In 1930, Ernest Glen Wever and Charles Bray recorded electrical potentials in the cochlea that reproduced the sound stimulus and described this phenomenon, which was later called the Wever–Bray effect.[3] These experiments were performed on a cat, with an electrode introduced into the auditory nerve. While they thought to record the discharges of the auditory nerve, following the "telephone theory" (signals carried along the cable of the ear, i.e., the auditory nerve), in fact it was the cochlear microphonic that they recorded, produced by the outer hair cells of the cochlea. The telephone theory was later rejected, but Wever and Bray did inspire several CI pioneers.[4]

1.1.4 S.S. Stevens: Electrophonic Hearing

In the 1930s Stanley Smith Stevens and his colleagues described "electrophonic hearing," thought to be the mechanism by which cochlear structures respond to electrical stimulation to produce hearing and therefore only present in intact cochleae. It is now known that electrophonic hearing is a result of the mechanical oscillation of the basilar membrane responding to voltage changes. Before 1957, efforts to stimulate hearing electrically were performed on patients with at least a partially functioning cochlea. The results therefore could have been based on electrophonic hearing instead of on direct stimulation of the auditory nerve. The early pioneers had to prove the effect of their auditory implant as being a result of stimulation of the auditory nerve and not electrophonic hearing.

1.2 Pioneers and Experimentation

1.2.1 In France

Andre Djourno and Charles Eyries (1957)

The first direct electrical stimulation of the auditory nerve was performed in the 1950s by the French-Algerian surgeons Andre Djourno (1904–1996) and Charles Eyries (1908–1996). They each had a different interest and background. Whereas Djourno was a scientist who had studied medicine, and had been interested in medical applications of electricity and nerve stimulation for a long time, Eyries was a trained otolaryngologist and was more of a clinician. He had more interest in the facial nerve embryology and function and ways to restore this function.

Before they worked together, Djourno had already produced a device to continuously measure pulse, had used electroencephalogram to study narcolepsy, and had originated the use of electricity for removal of metal pieces from bones. He also invented some sort of artificial respiration by direct stimulation of the phrenic nerve. With these inventions he showed his deep interest in neural stimulation by the use of prostheses. His next project of interest was the making and testing of "implantable induction coils" (which he called "microbobinages"). He made these coils himself to use for "telestimulation," and tested them in rabbits. The induction coil was placed under the skin and the stimulation was delivered transcutaneously. Subsequently, the rabbit jumped after stimulation of the sciatic nerve. He studied several aspects of telestimulation, among which were electrode biocompatibility, the effect of long-term telestimulation of a nerve, and the effect of stimulus frequency. Because with higher frequency the muscle did not contract and with lower frequency the contraction was painful, he determined that the ideal stimulus frequency was around 450 Hz, finally using his own voice as the telestimulus because it was of the appropriate frequency. This finding might have prepared him for the idea of stimulating the auditory nerve to restore hearing.

A 57-year-old male patient, who presented postoperatively with both bilateral loss of hearing and bilateral loss of facial mobility after bilateral resection of large cholesteatomas, brought Djourno and Eyries together. In the quest for a possible nerve graft, Eyries met Djourno at the laboratories of the medical school in Paris and they decided together with the patient on a surgical procedure with the purpose of restoring facial as well as acoustic nerve function.

This procedure was performed on February 25, 1957, by Eyries, during which the induction coil was placed under the temporal muscle and the active electrode was placed in the stump of the auditory nerve, while at the same time the facial nerve graft was applied. The procedure restored facial nerve function by the graft. The reports on the auditory outcome, also partly tested intraoperatively, showed encouraging responses. After extensive rehabilitation with a speech therapist, auditory sensations were present; discrimination between lower and higher frequencies was detected, but there was no speech perception. Unfortunately, after some weeks the implanted electrode broke down, and a subsequently implanted second device also broke down. Eyries held Djourno responsible for the breakdown of the electrodes and refused to perform a third implantation; this marked the end of their working partnership and communication.

Djourno, however, went on and was approached by a colleague who proposed to organize funding and engineering support for further development of the implant in collaboration with industry, in exchange for exclusivity. A true academician, Djourno refused; he was not interested in profiting from his inventions and detested industry. Out of principle he implanted one more patient together with a different, third surgeon. This implantation was not successful and because of lack of funding Djourno finally stopped working in the field of auditory prostheses.

The activities of Djourno and Eyries were followed up in Paris by Claude-Henri Chouard, a student in Eyries's laboratory. He became instrumental in the development of one of the first functional multichannel implants, the Chorimac-12 (now Neurelec/MXM-Oticon). He considered Charles Eyries as his major source of inspiration. Because of the work and implantation of Djourno and Eyries in 1957, that is seen as the year in which the development of auditory implants started.

1.2.2 Early American Years

The discoveries of Djourno and Eyries did not reach the outside world or the United States quickly: they published their findings in a French medical journal and the connections of Djourno, the scientist rather than the surgeon, with American otolaryngologists were not close. Eyries, the clinician, on the other hand did not show much enthusiasm for the project and that only briefly. William House in California serendipitously was given a translated English summary of the French manuscript via a patient around 1959. The manuscript was very positive on the findings and House became inspired.

William F. House (Fig. 1.2)

At that time, House (1923–2012) was a young dentist-turned-otologist who had completed his residency in 1956 and had just started working in Los Angeles at the Otologic Medical Group, with his half-brother Howard House. Although early in his career, he had already made some significant contributions to the field of otology/neuro-otology by developing the facial recess approach and later he also developed the middle fossa approach. Bill's bar at the fundus of the internal auditory canal, a landmark for the identification of the labyrinthine segment of the facial nerve, carries his (nick) name.

When the manuscript from Paris reached him he was working with John Doyle, a neurosurgeon, on the middle fossa approach. They were working on recording the cochlear nerve response to sound, associated with tinnitus. James Doyle, the brother of John Doyle and an electrical engineer, took charge of the electrical recordings during surgery. Having found success recording sound-induced potentials from the cochlear nerve, they planned on stimulating the nerve to restore hearing.

They first experimented with electrical stimulation to restore hearing during stapes surgery, using promontory or opened oval window stimulation. This was successful: these patients reported that they were able to hear the stimuli. This stimulated the researchers to implant a patient with a device.

The first patient who consented to being implanted was a 40-year-old man with severe deafness due to otosclerosis. Promontory stimulation of the right ear showed responses on January 5, 1961, and 4 days later a gold wire electrode was inserted under local anesthesia via retroauricular approach through the round window into the cochlea. The electrode left the ear via the retroauricular skin. The patient heard electrical stimuli but had poor tolerance of loud noise. After several weeks the implant was removed. A second patient, a woman suffering from congenital syphilis, followed during the same month: she also heard the stimulus. Eventually the wire was removed because of fear of infection. Hoping to be able to produce and study discrimination of the higher frequencies, the first patient was implanted again, this time with a five-wire electrode: the results were not very encouraging and this device also needed to be removed because of risk of infection. This problem with the biocompatibility of the materials used presented a major concern for the follow-up of their experimental procedures.

The thought behind implanting a five-wire electrode was to be able to spread the high-frequency stimuli along spatially separated electrodes; subpopulations of nerve fibers would thus be stimulated, summing to a high-frequency response along the whole nerve. An early patent application for this multielectrode implant was submitted by James Doyle and Earle Ballentyne in 1961. The patent process took time and it was only accepted in 1969. The supporting theory, however, was proven to be wrong because the same signal was applied to all electrodes and thus did not lead to pitch discrimination.

Fig. 1.2 William F. House.

In the meantime the press got hold of the story of these two patients: overenthusiastic articles were printed, leading to deaf patients calling Dr. House and Dr. Doyle for "this cure for their deafness." At the same time, investors became interested in making money from this new medical device. House was worried that these developments were far too optimistic, certainly in the light of the struggles with biocompatibility. Differences in opinion on how to proceed and handle this situation led to a separation between House and the Doyle brothers. The development of the implant became less of a priority to House, who also was running a busy otologic practice, while the Doyle brothers continued with experimenting and implanting, helped by an otologist from Los Angeles, Frederick Turnbull. Although they presented their results optimistically, no systematic analysis was performed. In 1968, James and John Doyle abandoned their research activities due to lack of funding.

F. Blair Simmons at Stanford University

F. Blair Simmons (1930–1998) was the first to stimulate the cochlear nerve in the United States in 1962.

During medical school at Harvard, Simmons worked in the laboratory of S.S. Stevens as a research associate and later also in the Walter Reed Institute before starting his residency in otolaryngology at Stanford University. He became an assistant professor at

Stanford in 1962 and within a month was presented with the chance to stimulate the cochlear nerve during a surgical procedure in an 18-year-old man with a cerebellar ependymoma. This recurrence of tumor presented with a mild hearing loss. An exploratory craniotomy was planned under local anesthesia, giving an opportunity for cochlear nerve stimulation and for receiving feedback from the patient. After the intention was explained, the patient agreed to the intraoperative stimulation and received preoperative training. During the procedure the patient described the bipolar square-wave stimulation directly on the auditory nerve as auditory sensations and was able to discriminate frequencies of stimulation up to 1 kHz. Simmons's first implantation then followed in 1964; the 60-year-old male patient had been deaf on one side and was now losing the hearing in the better-hearing ear, but also suffered from severe loss of sight due to retinitis pigmentosa. He agreed to the procedure wholeheartedly. A six-channel percutaneous device developed by Simmons was implanted via combined retroauricular and transmeatal approach under local anesthesia. A 2-mm cochleostomy and then a 0.1-mm hole in the modiolus gave access to the cochlea and auditory nerve. Testing was performed at Stanford and at Bell Laboratories. The testing was very difficult due to the patient's combination of disabilities, but Simmons proved, by stimulating each electrode separately, that stimulation of the six different parts of the cochlea led to different frequency/pitch perceptions. His implant was a percutaneous device able to stimulate all six electrodes separately, in contrast to the five-wire electrode of House with which all electrodes received the same signal at the same time.

After these experiments and their results, Simmons was pessimistic about the possibility of achieving speech perception and thought that biocompatibility studies first had to be performed before further work on a cochlear implant would be worthwhile.

1.2.3 Later American Years

William House Reengages

Although House had left the field of auditory implant development in frustration and had focused more on clinical practice in the 1960s, with the development of pacemakers and ventriculoperitoneal shunts he sensed that return to the field of cochlear implantation might be possible as the issue of biocompatibility seemed to have been solved. In the 1970s he developed a single-channel device together with engineer Jack Urban. This led in 1972 to the House/3M single-channel device, approved by the U.S. Food and Drug Administration (FDA) in 1984. This was the first implant worldwide to be implanted in a large group of patients.[5]

Robert Michelson

The third implanting surgeon in the field of auditory implants in the United States was Robert Michelson, working in Los Angeles. His preliminary report to the American Academy of Otolaryngology on cochlear implantation using gold wire electrodes in three patients brought a storm of protest; the old-school auditory theories could not explain how electrical stimulation of neural tissue could create cortical auditory perception. The fact that he did not first perform his experiments on animals but operated directly on humans added to the criticism.

Some of his experiments were performed in the Coleman laboratory at the University of California, where he met Francis Sooy.

Implant Team University of California

Around that time in San Francisco, Francis Sooy, the newly appointed head of the ENT department of the university hospital of the University of California–San Francisco (UCSF), was building his own cochlear implant team. His vision was to gather strong individuals of different, multidisciplinary backgrounds and he found and hired Robin Michelson, C. Robert Petit, Mel Bartz (an electrical engineer), and Michael Merzenich (a neurophysiologist) to work together.

In that era there was great skepticism in the audiologic and ENT community and a disbelief in the success of these implants. The lay press had mainly been writing of the miracle, but no analyses were presented for publication in any scientific journals. Robert Petit, a student assistant, joined after meeting Michelson in his laboratory and telling Michelson of his belief in and dream of a multichannel electrode. However, Michelson did not believe that multiple channels were necessary. Nevertheless, they teamed up and worked with a two-channel implant, which was now imbedded in silicone. Merzenich, the neurophysiologist, performed neurophysiologic tests on the cats implanted by Michelson and Petit: one side with normal hearing, the other side with a cochlear implant. Merzenich, at first a skeptic, was convinced that the auditory input from the implant reached the brain of the cat, but it remained difficult to understand what exactly humans heard. Petit searched for new tests; he hired a music professor to create simple tunes with different sound envelopes and different pitches and loudness levels. In carefully controlled and filmed laboratory conditions, one of the newly implanted patients was presented with a song through the implant: it was recorded on film that the patient hummed the melody and tapped the rhythm of the song with a pencil. This movie prompted Francis Sooy to further support the cochlear implant team and was shown at a meeting of otologists in 1972. It finally convinced the professional field that it was indeed possible to stimulate the cochlear nerve and produce auditory cortical perception, although concerns and doubts remained.

1.3 Feasibility, Safety, and Evaluation Studies

1.3.1 The Bilger Report

Francis Sooy was responsible for arranging a meeting in 1974 sponsored by the National Institutes of Health (NIH). It was decided that at this stage cochlear implantation surgery was still experimental, that inclusion criteria needed to be formulated first, and that all further implantation had to cease until a strict and scientific evaluation was performed on hearing outcome and success of the procedure in the patients already implanted.

In 1975, NIH organized this thorough evaluation of the patients that had been implanted thus far in the United States: 13 in total were implanted with single-channel devices by either Robin Michelson (2) or William House (11). The contract to perform this assessment was awarded to a team from the University of Pittsburgh, directed by Robert Bilger. The patients were sent to Pitts-

burgh for a week for extensive evaluation: psychoacoustic, audiologic, and vestibular tests were performed.

After the study of these 13 patients, three conclusions were drawn: (1) a cochlear implant does give support in lip-reading skills; (2) it improves the quality of life; and (3) (surprisingly) patients also improve in their own speech production after implantation. Overall, the Bilger report concluded that a single-channel device helped deaf people, with minimal risks. This opened finances and funding for research to develop collaboratively a multichannel device. Also, for the time being, the single-channel implant was officially sanctioned.

1.4 Development of a Multichannel Device

1.4.1 United States, Australia, Austria, and France

After the Bilger report and its conclusions, cochlear implant research became more legitimate as did the funding. Questions of the safety and feasibility of electrical stimulation of the auditory nerve in the long term needed to be answered and also the best material for electrical biostimulation needed to be identified. Several groups started working on the development of a multielectrode cochlear implant: among others, the UCSF group led by Francis Sooy, including Merzenich, Michelson, and Robert Schindler, and the Melbourne group led by Graeme Clark. Helped by technological advances in the computer and aerospace industry, they worked on minimizing the receiver device and improving the safety and durability of the electrode array. The physicist Adam Kissiah, working for the NASA/Kennedy Space Center, played an important role: he patented an electronic digital hearing aid in 1977. His design is still used.

Around the same time, work on a multichannel implant was conducted in Vienna, Austria, leading to the implantation of one of the first multichannel devices (3M/Vienna) in December 1977 by Professor Kurt Burian, head of the department of ENT at Vienna University. The implant was developed by Ingeborg and Erwin Hochmair; the later founders of MED-EL in 1989, who before 1988 worked together with the U.S. 3M company, with whom they teamed up during a stay at Stanford University.

In France, Claude-Henri Chouard together with Patrick MacLeod also worked on a multichannel implant. As early as 1976 a French paper was published by Pialoux, Chouard, and MacLeod detailing experiences with a multichannel (8- and 12-channel) device, the Chouard–Bertin device (also called the Chorimac-8 and Chorimac-12).[6] In 1977 a patent was filed for this multichannel device and its electrophysiologic technique. With the help of the French government this group developed its implant system further, which later became known as the Neurelec/MXM (now Neurelec/Oticon) implant system (there is no FDA approval at the time of writing in 2014).

William House, however, continued working on his, now approved, single-channel device and, together with Jack Urban, refined the House 3M single-channel implant, which was the first FDA-approved implant. More than 1,000 were implanted from 1972 to 1985, after which the age criteria of the FDA began to be lowered (see the timeline box in the next column).

The activities in Australia (Melbourne group) and separately in the United States (UCSF) led eventually to the introduction of the Cochlear Nucleus device from Australia and the Advanced Bionics Clarion device from the United States. They were introduced from 1984 onward and overtook the single-channel devices because of better spectral perception and speech recognition abilities as demonstrated in large clinical studies with adults. In 1985 first FDA approval was granted for adults (Cochlear Nucleus), and in 1990 for children from the age of 2 years onward (Cochlear Nucleus).

FDA Approval Timeline

- 1984: single-channel electrode array in adults (House/3M)
- 1985: multichannel electrode array in adults (Cochlear Nucleus 22)
- 1985: first implantation in children (5 and 10 years)
- 1990: multichannel implantation from 2 years onward (Cochlear Nucleus)
- 1996: approval of the Clarion device in adults (Advanced Bionics)
- 1997: approval of the Clarion device in children (Advanced Bionics)
- 1998: multichannel implantation from 18 months onward (Cochlear Nucleus)
- 2000: multichannel implantation from 12 months onward (Cochlear Nucleus)
- 2000: multichannel ABI approval (Cochlear Nucleus)
- 2001: approval of the Combi 40+ Med-el device

Given that with the FDA approvals safety was no longer a major research question, attention was directed to other questions: speech processing was one of these items; earlier implantation in congenitally deaf children was another.

Combined work in universal neonatal hearing screening programs, earlier diagnosis of deafness and a team approach, education and rehabilitation of implantees, and a growing acceptance of the deaf community followed.

1.4.2 Graeme Clark, Melbourne University Australia (1935)

On the other side of the globe, Graeme Clark, an otolaryngologist, grew up with a deaf father who was a pharmacist. From a very young age he witnessed the difficulties his father had with communication and this energized him in his future goals. As a medical student he read the work of Blair Simmons and his discovery of the tonotopy of the cochlea and was impressed. In 1969 he studied single and multichannel devices in experimental animal studies at the University of Sydney and documented in his thesis that single-channel devices had limited utility. He searched for a systemic scientific approach to develop a multichannel device, looking at speech processing strategies, electrode array design, and development of a reliable implantable receiver. His efforts led to one of the first multichannel devices, implanted in 1978, and funded by what was to become Cochlear Company, the University of Melbourne, and the Australian government.

Some of the important findings of Clark and his group were the round window insertion resulting in less trauma, and the use of platinum electrodes for safest long-term stimulation. In 1981 he showed that without use of lip reading some open set speech understanding with the multichannel implant was possible.[7]

1.4.3 Anxiety and Hostility in Deaf Communities

Whereas at the start and during the earlier development of cochlear implants the professionals—who, limited at that time by their concepts and knowledge of the auditory system, did not believe the reports that were being put forward—were the resistant group, later it was the deaf community itself that raised objections. Protests were organized at conferences and meetings concerning cochlear implantation in the late 1980s and early 1990s. The professionals working at that time on cochlear implants found very strong and active opposition. In particular the decision to implant congenitally deafen children at a very young age without their own consent, implying the concept of disability, was offensive. The term "cultural genocide" was even used to express the threatened feelings within the deaf community, which mainly consisted of prelingually deafened people using sign language. Critics from the deaf culture argued that the cochlear implantation and the rehabilitation focusing on mastery of hearing and speech production would lead to a poor self-image by the child as a "disabled" person rather than a proud deaf person using sign language. In New York and the United Kingdom these revolts turned into battles.

Nowadays cochlear implants are more mainstream and many congenitally deaf, early-implanted children attend mainstream education instead of deaf schools or schools for the hearing deprived.

1.5 Development of the Auditory Brainstem Implant

1.5.1 Auditory Brainstem Implant

Although during the pioneering years several surgeons had already stimulated the cochlear nerve directly at the modiolus or at the cerebellopontine angle, or stimulated the inferior colliculus,[8] the development of the auditory brainstem implant (ABI) started in 1979. In that year, a female patient with neurofibromatosis type 2 (NF2) asked for placement of a single-channel electrode on her brain in a quest for hearing restoration. Although the outcome was thought to be very doubtful, her perseverance led to the first auditory brainstem implantation in 1979 by William House and the neurosurgeon William Hitselberger at the House Ear Institute.[9,10] A single ball-type electrode was placed within the cochlear nucleus complex and led to hearing sensations. However, probably due to a shift of the electrode, these effects were lost and a new design of electrode, better situated to the anatomy, was developed, resulting in a Dacron mesh/platinum ribbon two-electrode array (in collaboration with the Huntington Medical Research Institute). Around 25 patients were implanted with this system and their outcomes proved to be comparable to the results of the single-channel cochlear implant (sound awareness and support of lip-reading skills). When the number of electrodes in the ABI was increased to three it became clear that the auditory outcome and especially pitch might vary according to electrode location, showing tonotopy to be present in the cochlear nucleus also. The first multichannel brainstem implant used was presented in 1991[11] and was slightly modified in 1993.[12] This led to better speech perception performance and sound quality. Clinical multicenter trials started in 1994 with a multichannel brainstem implant (Nucleus Multichannel ABI, Cochlear). The implant was approved by the FDA as an investigational device in June 1994, but this device was eventually a hearing solution for NF2 patients. Whereas the ABI used in the United States started out with 2, 3, and later 8 electrodes, the current unified ABI of Cochlear is the former European ABI with 21 electrodes developed in 1991–1993.

Nowadays, around 1,000 ABIs have been placed, with around 100 in congenitally deaf children. As well as NF2 patients, non-NF2 patients with congenital cochlear malformations, cochlear nerve aplasia, cochlear aperture stenosis, and extensive cochlear ossification have also been implanted with an ABI. Final FDA approval was granted in 2000 for adults (18 years and older) and NF2 only, but is now granted for NF2 patients of 12 years or older with "reasonable expectations." In Europe and Asia at least 144 ABIs have been implanted in children and adults for other indications besides NF2.[13]

A remaining problem with the current ABI is the fact that the surface electrodes do not reach the cochlear nucleus and its tonotopy as well as does the cochlear implant. The higher frequencies, located below the surface of the nucleus, in particular are difficult to reach. To address this, a Penetrating ABI (PABI) was designed (House Ear Institute, Cochlear, and Huntington Institute; 2008), with a 10-microelectrode array to penetrate 1–2 mm into the cochlear nucleus (which is 2 × 8 mm) in addition to the 12 surface electrodes on a separate array. Although thresholds were lower, activation of higher pitches, and more selective stimulation were attained, the overall speech understanding was not significantly better than with a normal ABI.[14]

Additionally, for patients with damage of the cochlear nucleus or brainstem (mostly due to NF2) the auditory midbrain implant (AMI; 2015) or inferior colliculus implant (ICI) was designed. Around six patients have been implanted and showed disappointing results, although sound awareness and limited support of lip-reading skills are found to be comparable to the early ABI results in NF2 patients.[15,16]

1.5.2 Summary of Discoveries and Developments

The development of auditory implants has truly been a multidisciplinary, international endeavor. Alongside the development of the auditory implant, knowledge of the function and physiology of the cochlea and the auditory pathway grew; greater knowledge of the process of speech perception went hand in hand with advances in material and electrical technology. A summary of the most significant discoveries and most important professionals is presented in the timeline in **Fig. 1.3**.

1.5.3 New Developments and Future

Developments in auditory implants have not come to a standstill. Since hearing preservation and electroacoustical stimulation are already becoming mainstream, the next horizons will be the total implantable cochlear implant (TICI), enlargement of the experience with ABI in non-NF2 patients, regulation of the vast number of patients all needing regular service, improvement of the per-

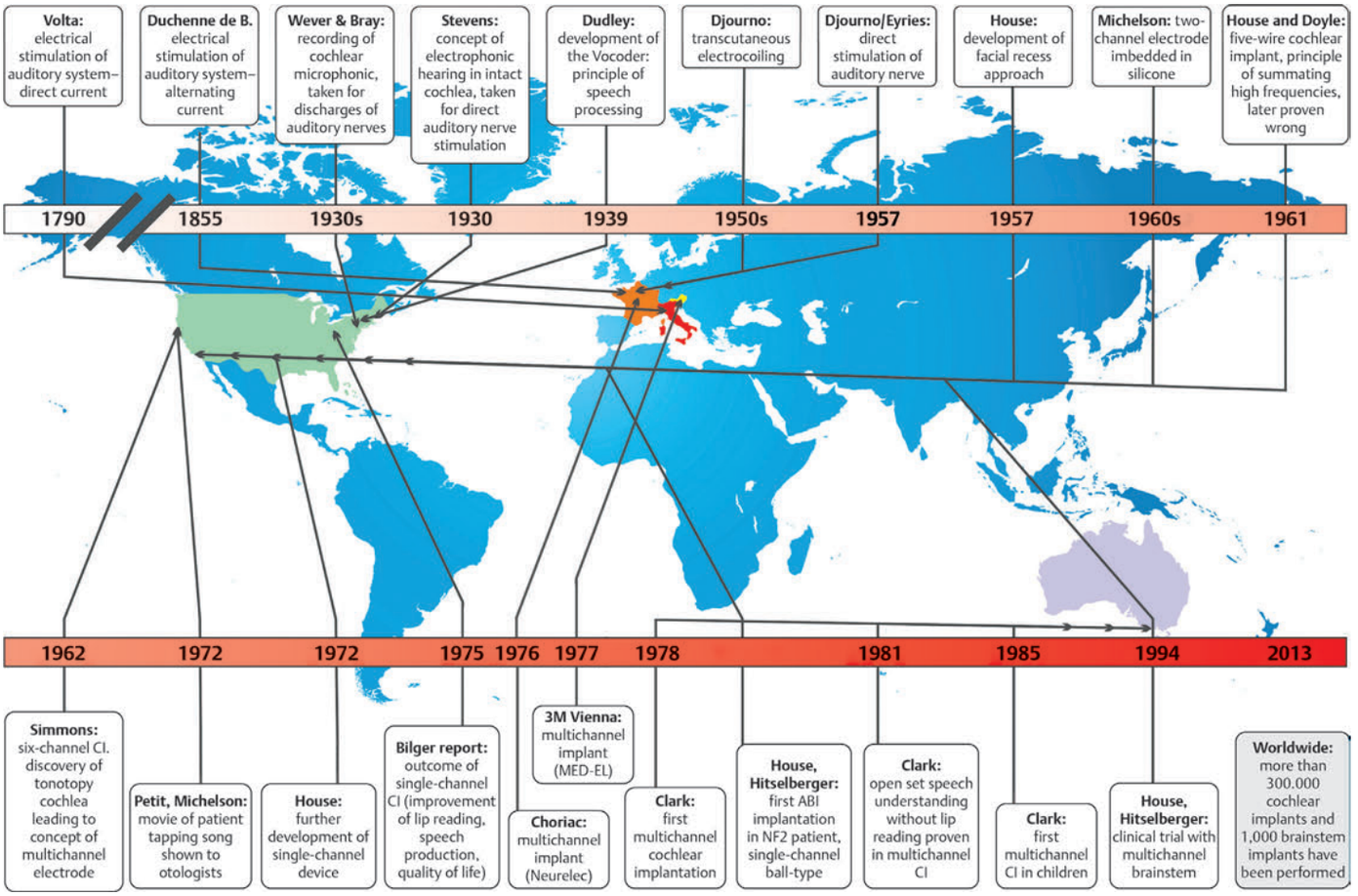

Fig. 1.3 Developments in auditory implantation on a timeline and world map.

ception of music with cochlear implants, and widening the availability of hearing rehabilitation in the developing countries as well.

Today more than 300,000 cochlear implants have been inserted, around 1,000 auditory brainstem implants have been surgically placed, and in the Western world bilateral implantation in children is becoming standard practice because better incidental learning conditions at school and better environmental hearing are proven benefits. The development in the field of auditory implantation over the past 50 years has truly been remarkable and has led to a profound change in the daily life, working conditions, and quality of life of the deaf or hearing-deprived patient.

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2 Surgical Anatomy in Auditory Implantation

Auditory implantation requires good knowledge of the anatomical structures that may be encountered during middle ear implant surgery, cochlear implant (CI) surgery, or auditory brainstem implant (ABI) surgery. Because the 3D anatomy of the middle ear, temporal bone, cochlea, cerebellopontine angle, and the brainstem is complicated, it is difficult to learn the anatomical composition only from a book. Intensive work in a temporal bone dissection laboratory and study of temporal bone instructional DVDs are mandatory to gain better insight into these anatomical relations. The dissections shown in this chapter have been performed especially to give appreciation of the anatomical relationship of the structures needed in auditory implantation.

2.1 Anatomy of the Middle Ear and Mastoid

2.1.1 The Mastoid (Fig. 2.1)

The usually well-aerated cells of mastoid and antrum are a perfect route for access to the middle ear and epitympanum. The landmarks for opening up the mastoid cavity are, posteriorly, the sigmoid sinus and, posterior fossa dura; superiorly, the middle cranial fossa dura; and, anteriorly, the posterior wall of the external ear canal. For easiest access to the facial recess, the posterior wall of the external ear canal needs to be thinned well. The digastric ridge is the most caudal landmark in the mastoid cavity, defining the facial nerve leaving the mastoid via the stylomastoid foramen, situated just anterior-medially to this ridge.

In case of a well-aerated cavity without pathology, a limited mastoidectomy with proper access will suffice for cochlear implantation. In case of pathology in the mastoid, all pathology needs to be removed meticulously.

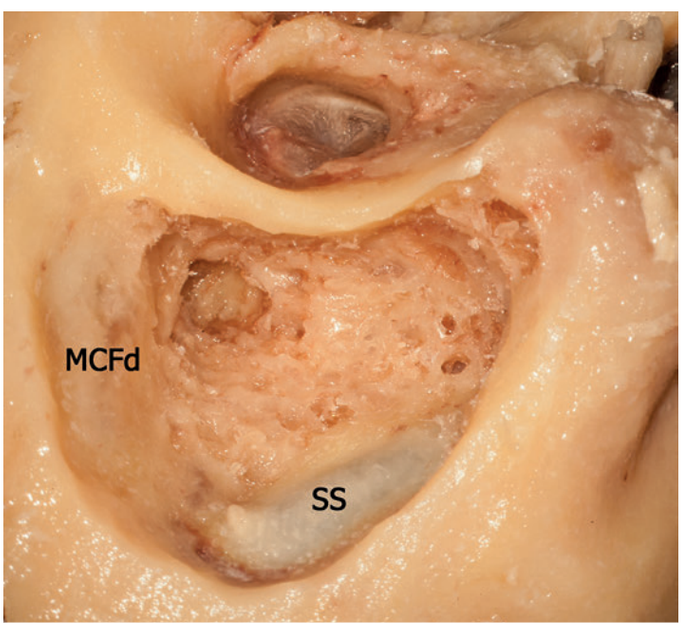

Fig. 2.1 Mastoid and antrum. SS, sigmoid sinus; MCFd; middle cranial fossa dura.

2.1.2 The Antrum (Fig. 2.1)

The antrum is the largest mastoidal cell and connects the mastoid air cells with the tympanic cavity via the aditus ad antrum ("the entrance of the antrum"). It is located just posteriorly to the epitympanum, inferiorly to the middle fossa plate, and posterior-lateral to the labyrinth. Since the antrum is very consistent and there is no important structure lateral to it, it serves as one of the most important landmarks in the initial stage of mastoidectomy. On the medial wall of the antrum is found the prominence of the lateral semicircular canal: one of the most important landmarks for identification of the facial nerve and the incus. The prominence of the lateral canal, together with the digastric ridge, defines the vertical segment of the facial nerve. Both structures can be used to identify the facial nerve before starting to drill the posterior tympanotomy.

2.1.3 The Tympanic Cavity

The mesotympanum is the middle portion of the tympanic cavity located just medial to the tympanic membrane (**Fig. 2.2**). It is bordered superiorly by the epitympanum (attic), which hosts the corpora of malleus and incus, and inferiorly by the hypotympanum, which starts inferior-laterally from the inferior border of the round window. The mesotympanum and epitympanum are separated by the tympanic segment of the facial nerve. The protympanum, located in the anterior part of the tympanic cavity, harbors the tympanic orifice of the eustachian tube, just inferior to the semicanal of the tensor tympani muscle and lateral to the genu of the carotid artery. The anterior epitympanum can be found anterior from James's cog, a bony eminence at the roof of the epitympanum. The cog is sometimes also regarded as a landmark for the facial nerve; it points at the facial nerve.