Cochlear Implants — Science, Serendipity, and Success

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The Lasker–DeBakey Clinical Medical Research Award, announced September 9, recognizes the contributions of three pioneers of cochlear implantation: Graeme Clark, Ingeborg Hochmair, and Blake Wilson. Their collective efforts have transformed the lives of hundreds of thousands of people who would otherwise be deaf.

Deafness impairs quality of life by relentlessly dismantling the machinery of human communication. Ludwig van Beethoven, plagued by deafness, wrote in 1802, "For me there can be no relaxation in human society; no refined conversations, no mutual confidences. I must live quite alone and may creep into society only as often as sheer necessity demands. . . . Such experiences almost made me despair and I was on the point of putting an end to my life."

The feelings of hopelessness, despair, and even shame that attended profound hearing loss lingered well into the late 20th century. The few who sought medical help were told there was nothing to be done for them. Today, the World Health Organization estimates that 360 million people worldwide are living with disabling hearing loss; as the population ages, the global burden of disease attributable to deafness will increase, and means of alleviating the disability will assume ever-increasing importance.

Profound hearing loss affects people of all ages. For children, hearing is central to neurocognitive development, since sound deprivation early in life degrades the multiplicity of neural circuits that are responsible for information processing, especially those involved in the acquisition of speech and language.[1] In addition, deafness impairs other key cognitive functions, such as scanning, retrieving, and manipulating verbal information — impairment that contributes to the low language levels typically achieved in people who are deaf from childhood. Since the ability to write a language depends largely on hearing its phonologic content, literacy rates among deaf children have remained intransigently low, despite the best efforts of educators. Low literacy leads to poor educational outcomes, limited employment opportunities, and restricted participation in society. For many, sign language becomes the only means of communication. Not surprisingly, deaf adolescents and young adults feel marginalized and need more psychological support than their hearing peers.

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Adults who develop profound deafness are often embarrassed by their disability and feel forced to withdraw from social exchanges with family and friends. For many of these adults, deafness may result in unemployment, imposing an additional psychosocial burden. Among the elderly, profound deafness compromises independent living, as many deaf seniors become too apprehensive to live alone. Moreover, deafness impairs cortical processing in the aging brain, especially under cognitive load, and is associated with an increased risk of dementia.

The challenge of restoring hearing to people who are too deaf to benefit from hearing aids was formidable and required an extraordinary, decades-long research endeavor. In the healthy ear, sound is collected by the external ear and amplified by the middle ear. The hair cells of the inner ear act as mechanoelectric transducers, converting acoustic energy into electrical activity that is carried to the brain through the auditory nerves. This transduction process is complex, requiring selective, time-critical

contributions from thousands of hair cells and auditory nerve fibers. In profound deafness, the hair cells are lost, and acoustic signals therefore cannot generate electrical activity in the auditory system. Could the auditory nerves be stimulated directly so as to bypass the inner ear and deliver a meaningful representation of the speech signal?

The earliest clinical attempt at such stimulation took place in Paris in 1957, when a surgeon directly stimulated the auditory nerve, causing a patient to temporarily experience crude auditory percepts. A patient brought this experiment to the attention of Dr. William House of Los Angeles, who immediately saw its potential. In the early 1960s, House successfully implanted single-channel devices to stimulate the auditory nerve through the cochlea.[2] House was roundly criticized for his work: neurophysiologists condemned it as naive and misguided — how could a handful of wires delivering crude electrical currents replace the function of thousands of hair cells and tens of thousands of auditory neurons? Clinical colleagues questioned his motives, feared the risk of meningitis, and distanced themselves so as not to sully their reputations. And the deaf community launched angry protests at what was seen as a peremptory attack on deaf culture.

An interactive graphic showing ear anatomy and the function of cochlear implants is available at NEJM.org

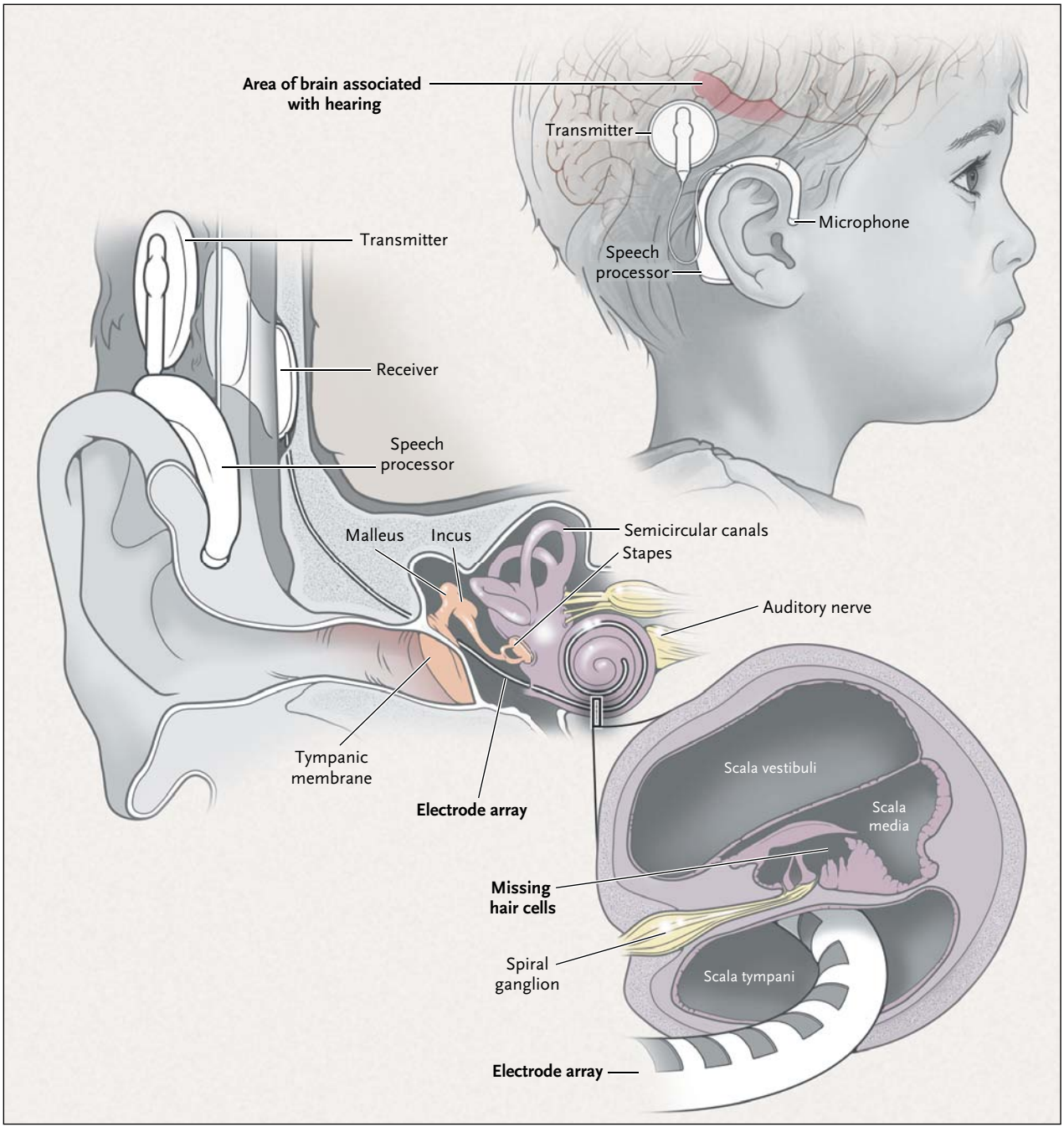
More laboratory experiments were clearly needed to bridge the huge intellectual and technological chasms facing early investigators, to pave the way for the transformational change that profoundly deaf patients needed. Biologic safety was of paramount importance, and exhaustive histopathological studies were needed to assess the safety of long-term stimulation and to inform the design of future electrode arrays. The optimization of electrical stimulation required detailed neurophysiological and psychophysical studies to guide clinical application. And many scientists doubted the sustainability of long-term stimulation, since some animal models suggested that the auditory nerve underwent retrograde degeneration due to deafness.

Early patients spent countless, laborious hours in laboratories connected to stacks of speech processors; making these processors wearable without losing computational power was an early imperative and represented an enormous engineering challenge. It became clear in the 1980s that multichannel systems allowing stimulation at multiple sites within the cochlea were needed for speech recognition. The 1990s heralded major advances in speech-encoding strategies for cochlear implants, offering speech recognition without lipreading to the majority of recipients. The realization that children who had been born deaf could also derive substantial benefit, with some developing speech and language trajectories similar to those of their hearing peers, was transformational for childhood deafness, making mainstream schooling a viable option for many deaf children.

Throughout the development of cochlear implants, the manufacturing challenges were monumental — including ensuring that the implanted electronics packages were permanently hermetically sealed, fabricating complex electrode arrays for deep insertion into the tortuous cochlea, and meeting the stringent regulatory requirements for implanted biomedical devices.

Current cochlear-implant systems are worn at ear level and contain many features of earlier prototypes. They include an implanted portion with receiver electronics attached to an electrode array placed within the cochlea, plus external components comprising a microphone, a speech processor, and a transmitter coil (see diagram). Bilateral cochlear implantation, now routine treatment in many countries, permits recipients to better understand speech in the midst of noise and to localize sound. Contemporary systems and surgical techniques allow any islands of residual hearing to be preserved, enabling electrical and acoustical hearing to be effectively combined; this combination permits better speech understanding in multitalker settings, identification of the speaker's sex, and better reception of tonal languages.

Cochlear implants have their limitations: they do not restore normal hearing, outcomes vary among patients, performance is considerably degraded by ambient noise, and music perception is limited. It's hoped that continuing research will uncover better ways of delivering the fine-structure content of the speech signal, creating more effective channels of stimulation with less electrical overlap, reducing trauma to cochlear structures through pharmacologic means, and enhancing brain responsiveness by removing molecular inhibition of plasticity.[1]



Function of a Cochlear Implant within the Ear.

These current efforts are founded on research carried out by Lasker awardees Graeme Clark, Ingeborg Hochmair, and Blake Wilson. Clark, an otolaryngologist, contributed an entire portfolio of rigorously conducted biologic and psychophysical experiments that underpinned clinical practice and informed the design of a clinical device[3]; Hochmair, an electrical engineer, contributed engineering brilliance and innovation, establishing her own company to hasten the perilous journey from bench to bedside[4]; and Wilson, a speech scientist, oversaw a giant leap forward in speech encoding

for implants that ingeniously manipulated the timing and place of stimulation so as to minimize distortion and channel interaction.[5] These three scientists had the grit to pick "impossible" projects and the courage to remain steadfast in the face of failure and criticism. Above all, they remained incurably passionate about achieving victory over one of humanity's most prevalent disabilities. They have brought sound where there was silence and hope where despair prevailed. Though they fully deserve the Lasker Award, their greatest accolade is the gratitude of 300,000 implant recipients around the world to whom they've given the gift of hearing.

Disclosure forms provided by the author are available with the full text of this article at NEJM.org.

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