REVIEW ARTICLE

MEDICAL PROGRESS

Profound Deafness in Childhood

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In childhood, profound hearing loss (a hearing level of >90 dB) has far-reaching, lifelong consequences for children and their families. The most striking effect of profound hearing loss is the lack of development of spoken language, with its impact on daily communication; this, in turn, restricts learning and literacy,[1] substantially compromising educational achievement and later employment opportunities.[2,3] There is a high prevalence of psychosocial problems among deaf children.[4] Fortunately, recent interdisciplinary developments are transforming outcomes, offering many more opportunities for deaf children. Recent advances[5] suggest that deafness may be considered a model system for understanding neurosensory restoration. For example, cochlear implants can bypass the sensory end organ, stimulate the neurobiologic and neurocognitive substrates for speech and language processing, and consequently promote cognitive development.

The children with the best results from cochlear implantation are among those who have received implants before 2 years of age. These children will enter first grade with expressive and receptive spoken language skills that are close to those of children with normal hearing[6]; their successful participation in mainstream education has become a realistic expectation.

Causes and Pathophysiology of Profound Childhood Deafness

The prevalence of permanent childhood hearing loss, which is mainly due to loss of cochlear function, is 1.2 to 1.7 cases per 1000 live births.[7] Between 20 and 30% of affected children have profound hearing loss. The prevalence increases up to 6 years of age as a result of meningitis, the delayed onset of genetic hearing loss, or late diagnosis. In developing countries, the prevalence is greater because of a lack of immunization, greater exposure to ototoxic agents, and consanguinity; about half the disabling cases of hearing loss worldwide are preventable. Approximately 30% of deaf children have an additional disability — most commonly, cognitive impairment.[8]

Hearing loss can result from interference with the transmission of the acoustic signal at any point between the outer ear and the auditory cortex. Sound energy is collected by the outer ear and is amplified by the middle ear for transmission to the cochlea (Fig. 1). This energy transfer initiates a traveling wave along the basilar membrane, resulting in shearing forces between the tectorial and basilar membranes. The shearing forces tilt the stereocilia of the hair cells, stretching tip links and opening potassium channels. Potassium inflow generates a receptor potential in the hair cells, which in turn leads to secretion of glutamate into the synaptic cleft. Action potentials are generated in the spiral ganglion cells, activating the central auditory system. The "battery" of this process is the stria vascularis, which actively secretes potassium into the endolymph.

Audiologic testing can identify the site of the lesion and permit characterization

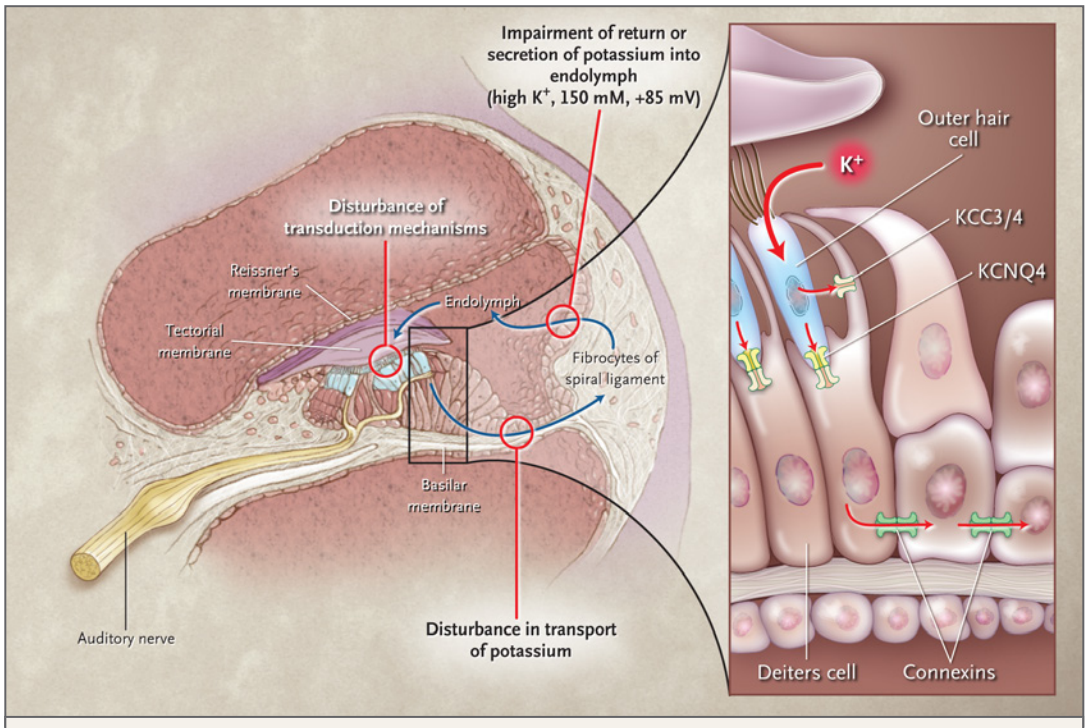


Figure 1. Impaired Molecular Processes in Deafness.

Cochlear function relies on a number of molecular processes that can be disturbed in deafness. Hearing function requires the inflow of potassium into the hair cells (inset). In the stria vascularis, potassium is extracted from blood and actively secreted into the endolymph. Electrochemical forces then drive potassium into the hair cells. The potassium is recycled: it leaves hair cells through channels (KCNQ4) to enter Deiters cells (through the KCC3 and KCC4 channels) and pass through connexins into surrounding cells and fibrocytes. Deafness may be caused by disturbance of transduction mechanisms, by disturbance in the transport of potassium from hair cells through connexins, or by impairment of its return or secretion into the endolymph.

of individual hearing losses (Table 1). It is now possible to detect "dead regions" in the cochlea (the result of a discrete loss of inner hair cells[9]) and to distinguish between dysfunction of hair cells and dysfunction of the auditory nerve (by using otoacoustic emissions and auditory brainstem responses[10]). Etiologic classification has been enhanced by developments in molecular medicine[11] that have helped characterize previously indistinguishable causes of deafness (Table 2). Inherited deafness affects a variety of molecular processes (Fig. 1), including gene mutations known to interfere with the function of transcription factors, potassium and chloride channels,[12] connexins,[13] and stereocilia.[14] In addition, there is evidence that genetic variations (e.g., mitochondrial mutations) may lead to increased sensitivity to ototoxic agents. A single gene (*GJB2*), which encodes the connexin 26 molecule, is commonly involved in deafness; mutations in this gene interrupt potassium recycling, resulting in the accumulation of potassium and, ultimately, cell death.

CENTRAL NERVOUS SYSTEM CONSEQUENCES OF CONGENITAL DEAFNESS

In contrast to the cochlea, the brain is immature at birth[15] and develops over many years.[16] Although some of the ability to differentiate auditory stimuli is inborn,[17] a range of sensorimotor, perceptual, and cognitive abilities are acquired during childhood. The acquisition of spoken language requires auditory input and interaction with the environment (e.g., an understanding of communicative intent and opportunities for role-play and imitative learning[17,18]). On the basis of the ability to differentiate sounds, children learn to abstract and categorize stimuli by learning to "recognize"

Table 1. Audiologic Assessment in Children.

Assessment	Technique	Clinical Usefulness
Objective tests		
Otoacoustic emissions	With sensitive microphone placed in ear canal, detection of mechanical energy propagated outward by metabolic activity in outer hair cells	High sensitivity and specificity make this test an essential screening tool; handheld automated versions are available; no behavioral response needed; takes about 10 min per ear; presence of emissions may indicate auditory neuropathy, a condition characterized by normal outer-hair-cell function but deficient conduction along the auditory pathway
Automated brain-stem auditory evoked response	Measurement of electrophysiological responses to acoustic stimuli generated in auditory nerve and brain stem	Used to determine hearing threshold; sedation often required, and testing takes about 15 min for both ears; ear-specific results can be obtained
Auditory steady-state response	Measurement of electrophysiological responses to rapidly modulated auditory stimulation with steady-state stimuli	Allows delivery of stimuli at high intensity (125-dB hearing level), with frequency-specific and ear-specific estimation of steady-state response; complements test of auditory brain-stem response in assessing profound hearing loss
Tympanometry	Recording of middle-ear impedance as pressure in ear canal is raised or lowered	Used to assess status of middle ear
Acoustic reflex	Measurement of increased stiffness of middle ear due to contractions of middle-ear muscles in response to loud sounds	Useful for estimating hearing threshold or identifying sites of auditory dysfunction from middle ear to brain stem
Cortical evoked response	Measurement of physiological activity in a range of sites beyond the brain stem (e.g., auditory cortex)	Primarily used in research to assess higher-level auditory functions (e.g., neurologic dysfunction); may be used to monitor maturation of auditory system
Behavioral tests		
Observational audiometry	Assessment of change in state of activity in response to sound in very young infants	Useful in combination with other tests; not ear-specific; responses may be misinterpreted
Visual-reinforcement audiometry	Use of a head turn in response to an acoustic stimulus, which is then reinforced by a visual reward	Can be performed in children as young as 6 mo of age; provides frequency-specific and ear-specific information; should always be used as soon as possible to confirm objective tests

a phoneme, their mother's voice, or a favorite musical instrument as being distinct from background noise. As a result, auditory "objects" emerge perceptually.[19]

Auditory objects are formed in the cerebral cortex,[20] which is also responsible for conscious experience and sensory learning. Cortical development continues until adulthood,[16] with extensive developmental changes both at the cellular and microcircuitry levels (Fig. 2). In addition, the auditory cortex is composed of several functionally and histologically distinct Brodmann's areas. These areas are tightly interconnected and together represent one functional unit; lower-order areas activate higher-order areas (bottom-up interactions), and higher-order areas modulate those below (top-down interactions[19]).

Congenital deafness changes the functional properties of the auditory system[24-27] and impairs cortical development[21] (Fig. 2), affecting the mutual interaction of the cortical areas.[25,28] Complex auditory functions and speech perception cannot be comprehensively established when hearing is restored late in life in congenitally deaf persons, since some aberrant developmental steps in synaptic counts, plasticity, and network properties have taken place without hearing (Fig. 2). Stimulation of the auditory system during periods of maximal receptiveness (sensitive periods) is central to its normal development.[25,29]

Sensory modalities have extensive interconnections with other brain regions. Real-world events typically generate simultaneous auditory, visual, or somatosensory responses.[30] Such multimodal

Table 2. Classification and Features of Hearing Loss.

Variable	Comments
Site of lesion	
Conductive	External or middle ear
Sensorineural	Cochlea or auditory nerve
Neural	Auditory nerve (as in auditory neuropathy); may be nongenetic (e.g., developing after hyperbilirubinemia) or genetic (e.g., due to a mutation of the otoferlin gene *OTOF*)
Central	Due to difficulties with perceptual processing of auditory information in the brain
Onset	
Congenital	Present at birth; can be detected by neonatal screening
Acquired	Develops any time after birth (e.g., after infection or head trauma)
Cause	
Genetic	Attributable to inherited disturbance of molecular mechanisms in the inner ear; genetic causes account for at least 50% of cases of permanent hearing loss in childhood; molecules encoded by involved genes include the gap-junction protein connexin 26 (a *GJB2* mutation), motor molecules (actin and myosin), and transcription factors; inheritance is usually autosomal recessive (80% of cases) but may be dominant (15%) or X-linked or mitochondrial (<1%); deafness may be present at birth or may develop in later life; about 4% of children with genetic hearing loss have an inner-ear malformation
Infectious	May be prenatal (due to cytomegalovirus infection, rubella, syphilis, toxoplasmosis, or other viral infection) or postnatal (e.g., due to measles, mumps, or meningitis); meningitis may obliterate the cochlea with new bone, with major implications for cochlear implantation
Environmental	Extracorporeal membrane oxygenation, noise; may be associated with admission to neonatal unit
Ototoxic agents	Aminoglycoside antibiotics (with the 1555A→G mutation of the 12S rRNA [*MTRNR1*] gene conferring genetic susceptibility in some children) and chemotherapeutic agents such as cisplatin
Miscellaneous	Sepsis, craniofacial anomalies, prematurity, low birth weight, anoxia, rhesus incompatibility
Clinical features	
Syndromic deafness	Associated with other recognizable clinical findings (e.g., disturbance of vision [in Usher's syndrome], disturbance of thyroid function [in Pendred's syndrome], or cardiac arrhythmia [in Jervell and Lange-Nielsen syndrome]); accounts for 30% of cases of hereditary hearing loss; about 400 syndromes have an associated hearing loss
Nonsyndromic deafness	Deafness as an isolated finding
Language	
Prelingual deafness	Occurs before development of spoken language
Postlingual deafness	Occurs after acquisition of spoken language
Severity	
Mild, moderate, or severe deafness	A hearing level of 20–40 dB indicates mild hearing loss, 41–70 dB moderate loss, and 71–90 dB severe loss; mild-to-severe loss is generally permanent, but hearing aids can compensate for the deficit; the level of loss may fluctuate, as in the large vestibular aqueduct syndrome (often associated with Pendred's syndrome), in which minor head trauma or air travel may cause precipitous loss of hearing
Profound deafness	Hearing level >90 dB; may require cochlear implants to access speech

interactions are established postnatally[31] and require multisensory input.[32] Deafness affects the functional coupling among the sensory systems: multimodal interactions do not develop, and other sensory systems completely or partially overtake some auditory cortical regions.[33] Furthermore, since information cannot be represented through sound in deaf persons, it becomes represented in reference to other modalities, a process that has adverse cognitive effects.[34,35] Thus, deaf children have difficulty scanning and retrieving phonologic and lexical information in their working memory; they also differ from their peers with normal hearing in their ability to sustain visual atten-

tion[36,37] and in visual sequence learning.[38] Consequently, the auditory system becomes uncoupled from other systems, affecting key cognitive functions.

CLINICAL EVALUATION

A comprehensive history taking that explores relevant risk factors is an essential first step in evaluation. Diagnosis of hearing loss is usually achieved through a battery of objective tests (Table 1)[39] with the use of the time-honored "cross-check principle" (in which the diagnosis is supported by more than one objective test). It has become possible to determine the degree and type of hearing loss in each ear, as well as the site of the lesion, even in the first few weeks of life. The audiologic test battery should include age-appropriate behavioral testing as soon as feasible.[40]

Universal neonatal hearing screening (UNHS) aims to identify hearing loss early in life, which facilitates early intervention. A combination of otoacoustic emissions and auditory brain-stem responses is used, often in a two-stage process.[41] The superiority of UNHS over conventional distraction testing is now undisputed, and the yield from newborn screening (1.2 cases identified per 1000 infants; 95% confidence interval, 0.8 to 1.7) is close to the expected prevalence. The proven benefits of early identification and intervention (at <6 months of age) in terms of later language outcomes,[3,42] reading ability, and communication have confirmed the effectiveness and cost-effectiveness of UNHS and its potential to transform the life opportunities of children whose deafness would not otherwise have been diagnosed until later in life.[43] It has been proposed that neonates in whom deafness is detected undergo comprehensive diagnostic evaluation and intervention at no later than 3 months of age.[44] In many health care systems, substantial investment in and redesign of children's hearing services as they now exist[45] will be needed to meet such a target.

Once deafness is established, a systematic approach to determining the cause is best undertaken within a dedicated multidisciplinary setting. Given that half the cases of congenital hearing loss have a genetic basis, genetic testing may be very useful, although some families may decline such investigations. Identification of the cause may provide substantial benefits, such as determining the prognosis, identifying associated risk factors (e.g., cardiac conduction defects) or coexisting conditions (e.g., impaired vision), preventing further hearing loss (e.g., by identifying ototoxic susceptibility of the 1555A→G mitochondrial mutation), and facilitating genetic counseling. In about 30 to 40% of cases, the cause remains unknown.

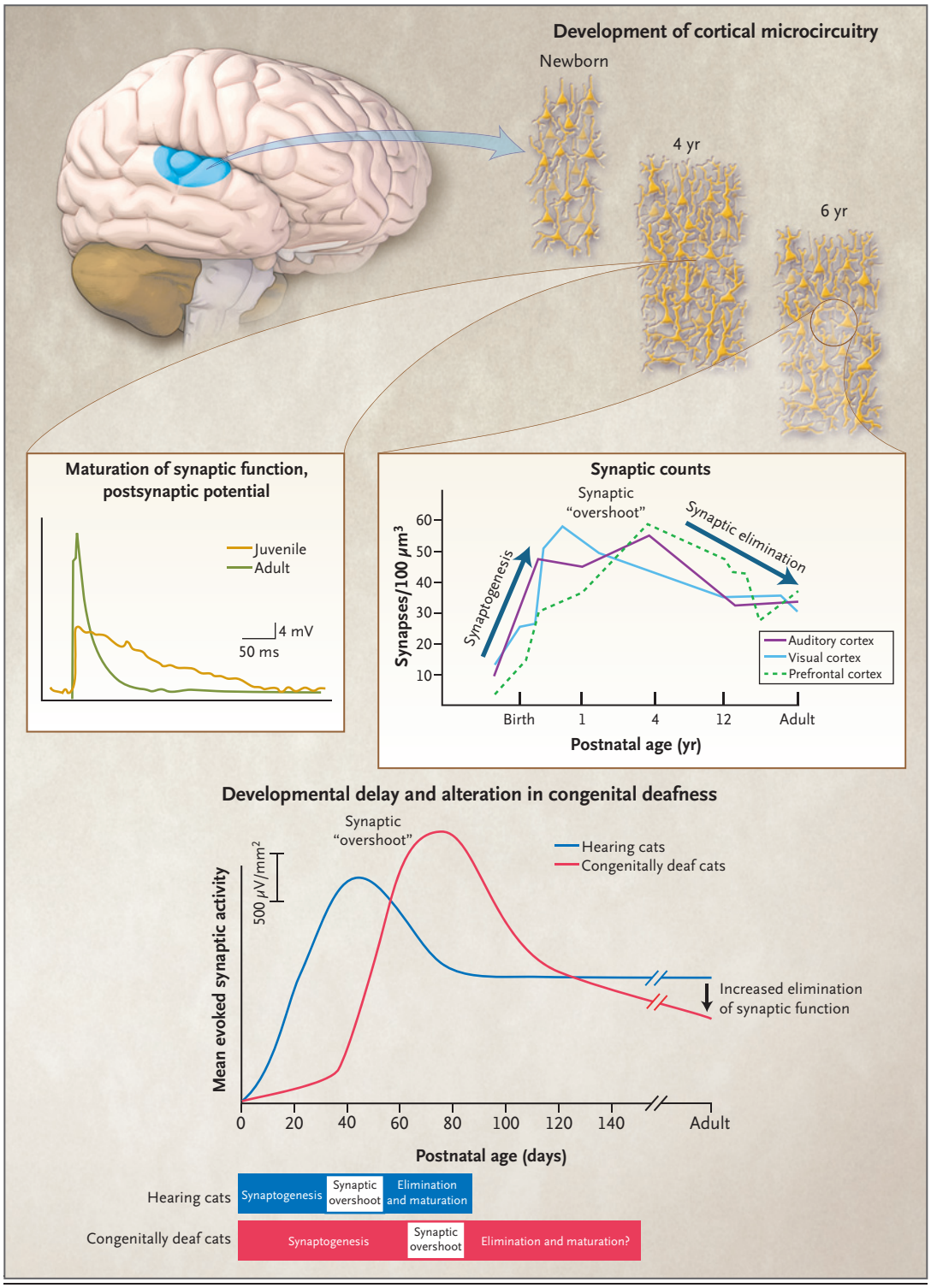
Deafness is a family matter. Listening to parents' views and valuing their roles and input remain the most helpful clinical intervention[46]; thus, time for parental engagement should always be given priority.

Figure 2 (facing page). Development of the Auditory Cortex.

The effects of postnatal development on the circuit properties of the auditory cortex are shown. The density of dendritic trees is highest at the age of 4 years in children with normal hearing. Synaptic counts reflect the circuit changes and demonstrate the changing computational power of cortical networks. Peak synaptic density has been observed at 2 to 4 years in children with normal hearing. Subsequently, synaptic counts decrease; unused synapses are eliminated, reflecting the brain's need to specialize its functions to accommodate prevailing conditions (demands). This specialization is accompanied by changes in synaptic function that have been confirmed in animal models. In juvenile animals, synaptic potentials have a longer duration, leading to higher synaptic plasticity. In adult animals, the synaptic potentials are shorter, leading (along with other molecular changes) to reduced plasticity. The auditory cortex develops differently in animals and persons with congenital deafness. In congenitally deaf cats, the overall synaptic activity (a measure sensitive to all the effects shown) shows two main effects of deafness: a developmental delay with retarded and exaggerated synaptic overshoot and a consequent increased elimination of synaptic function, starting after the development of overshoot and continuing into adulthood. To what extent the maturation of synaptic function contributes to this functional elimination is unclear at present. The horizontal bars show the temporal windows of these developmental processes in hearing and deaf cats. The eventual consequence is impoverished cortical activity in deaf adults. Data are from Huttenlocher and Dabholkar,[16] Kral et al.,[21] Conel,[22] and Aramakis et al.[23]

BASIC PRINCIPLES OF COCHLEAR IMPLANTATION

Cochlear implants partially restore hearing by bypassing the nonfunctional organ of Corti. They electrically stimulate the auditory nerve fibers that survive the loss of hair cells (Fig. 3). More than 80,000 children worldwide have cochlear implants. Contemporary systems have up to 24 electrodes, can record evoked signals from the auditory nerve, and contain several speech-encod-



ing algorithms that transform sound into electrical stimuli.[47] Some algorithms allow signals to be directed to preselected auditory nerve regions. The processor encodes the speech signal as pulse trains delivered to individual electrodes that are arranged by frequency to exploit the cochlea's normal tonotopic distribution of fibers (high frequencies at the basal end and low frequencies toward

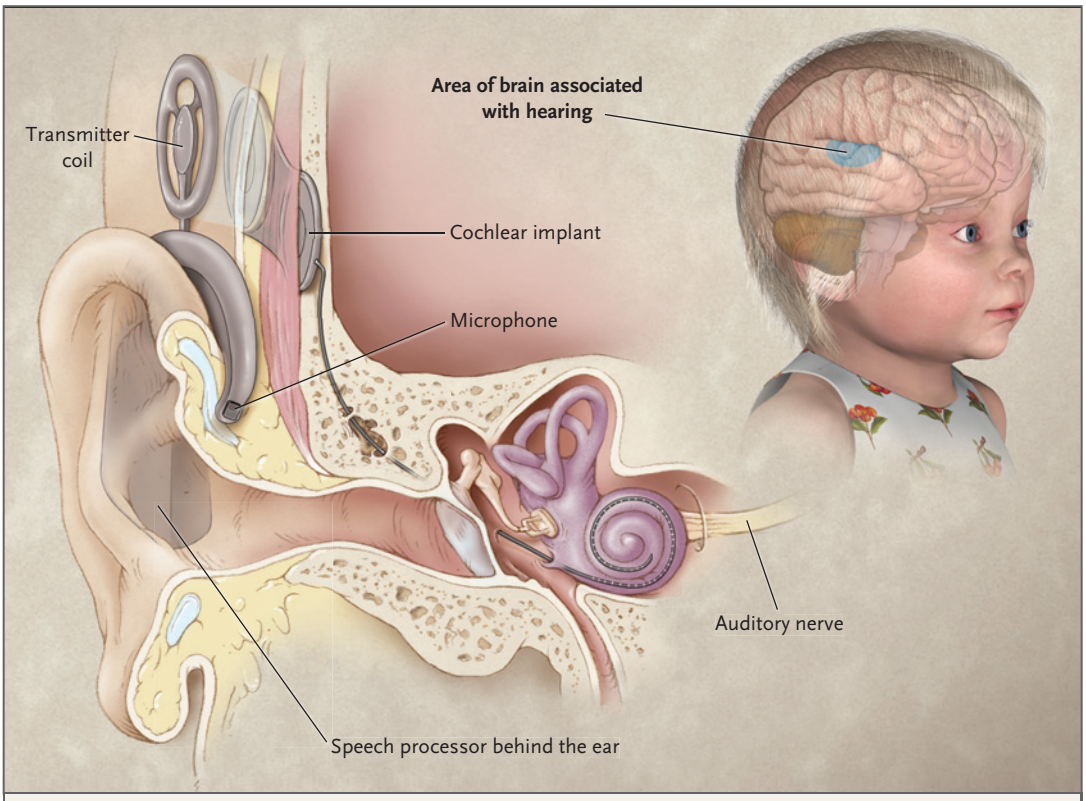


Figure 3. Position of a Cochlear Implant in the Human Ear.

The auditory cortex is shown in blue. The speech processor (with batteries) is located behind the pinna. Sound is picked up by the microphone located above the pinna, processed by the processor, and led to the transmitter coil. This coil transmits the signals to the subcutaneous receiver by magnetic induction. From there, the signals are carried by an electrode array, which is surgically positioned in the cochlea. Power is supplied by the batteries in the processor located behind the ear. Batteries require replacement every 2 to 3 days.

the apex). To avoid interactions with adjacent electrical fields, stimulation is provided by short, interleaved pulses. Cochlear implants provide physiologically useful intensity, frequency, and timing cues that are required for speech comprehension. However, the temporal fine structure, which is important for an understanding of speech against background noise and for music appreciation, is poorly represented in current devices; also, the encoding of very-low-frequency signals, such as the pitch of a voice, is limited. Electrical stimulation thus evokes an impoverished pattern of auditory nerve activity, as compared with acoustic stimulation.[48]

CANDIDACY FOR COCHLEAR IMPLANTATION

Deafness has wide-ranging implications for the child and his or her family, and evaluation should thus be multifaceted, embracing their social and emotional needs, lifestyles, communication preferences, and expectations. Cochlear implants should be considered for children younger than 24 months of age who, without hearing aids, can hear only sounds that are louder than a 90-dB hearing level at frequencies of 2 and 4 kHz[49]; however, in the era of UNHS, determinations are more often based on objective measures than on behavioral thresholds. In older children, less conservative criteria are often applied.[50] The decision to perform an implantation should be supported by evidence of the lack of development of speech (or its precursors), language, and listening skills that are appropriate to the child's age, developmental stage, and cognitive ability after appropriate hearing-aid use. Since outcomes are known to vary, physicians should help parents set achievable, evidence-based expectations.

Cochlear implantation typically results in com-

plete loss of residual hearing, so certainty about the audiologic findings before implantation is essential. Hearing losses are often complex, as in auditory neuropathy (in which auditory-nerve activity is desynchronized[10]), and some infants may simply be difficult to test by virtue of coexisting disease. In children with residual hearing, deciding when cochlear implantation would predictably outperform hearing aids can be difficult; repeated testing and observation over time may be necessary. With such complex decisions being made so early, largely based on objective testing in neonates, the onus on physicians to provide accurate determinations of hearing capacity early in life is considerable. Thus, candidacy is generally best decided by an experienced multidisciplinary team, with active collaboration with professionals in the child's educational environment. Empowering parents and placing them at the center of the decision-making process remains essential for long-term success.

AUDITORY PLASTICITY AND SENSITIVE PERIODS

Cochlear implants can alleviate the deficits in the auditory system[24] and promote cortical maturation in deaf animals[25,26]; electrophysiological studies in humans are consistent with these laboratory findings, showing maturation of evoked responses with cochlear-implant stimulation.[51] However, a period of maximal receptiveness to auditory stimulation (sensitive period) occurs both in animals and in humans[25,29]; this is controlled by genetically determined processes that prevail even in the absence of auditory experience (Fig. 2). Eventually, the consequence of these changes is a substantial reduction of synaptic activity (computational power) of the cortex in deaf animals as compared with hearing animals. Delaying implantation beyond this window markedly decreases brain adaptability and speech understanding.[25,29]

Several sensitive periods are proposed in the development of the human auditory system, relating to the auditory, phonetic and phonologic, syntactic, and semantic aspects of language.[52] These periods probably reflect the differential maturation sequence of the various cortical areas. Normal brain maturation also requires the capacity to respond through appropriate multimodal interaction, which is affected by deprivation.[32] In persons who have become deaf after the acquisition of spoken language, brain activity evoked by a cochlear implant can be observed in nonauditory regions, with visual centers contributing to comprehension of speech through lip-reading.[53,54] However, by taking over auditory neuronal resources, such cross-modal reorganization can also degrade auditory performance in persons and animals with congenital deafness.[55,56]

On the basis of these findings and linguistic outcomes, implantation is recommended in the first 1 to 2 years of life[57,58]; with the use of UNHS, implantation is feasible in the first year of life.[59] Major complications occur in about 5% of patients, and the most common complication is infection.[60] Contemporary implant systems carry a low risk of meningitis,[61] but pneumococcal vaccination is highly recommended before surgery.[62]

BILATERAL COCHLEAR IMPLANTS

The ability to localize sounds in the auditory space provides cues that humans use to segregate auditory objects, especially in conditions involving multiple sound sources or in noisy environments. The physiological mechanisms are complex, relying on minute intensity and timing differences between the two ears. The neural pathways that subserve binaural hearing are only partially degraded by sound deprivation[63] and can be salvaged, at least in part, by early sensory restoration.[64] To optimize outcomes, bilateral implantation is best undertaken early, either simultaneously or with a short interval between the two procedures.[65] Studies of bilateral cochlear implantation in children has shown that sound localization is improved by 18.5%; crucially, an average 20% improvement in the ability to hear speech against background noise has been reported under rigorous test conditions.[66] Concerns remain regarding cost-effectiveness, given that the lifetime costs approach $90,000 and commensurate health benefits remain to be demonstrated.

DEVELOPMENT WITH A COCHLEAR IMPLANT

Cochlear implants deliver new stimulation to specialized cortical areas in the brain that are responsible for auditory, phonetic, and phonologic processing, eventually enabling the recipient to encode, process, and reproduce speech signals. The development of proficiency in spoken language

requires processes that link objects, actions, and language together. The goal is to optimize the development of age-appropriate spoken language skills in the years after implantation, usually by adopting a hierarchical approach from simple detection through understanding spoken words to their typical use in conveying thought. A matter of much contention among therapists and parents is the choice of the form of communication (signed or oral) that facilitates spoken language development. Effective communication (interaction) during the preimplantation period, even if expressed through signs, facilitates the later acquisition of spoken language.[67] However, signing alone does not allow the development of phonetic and auditory functionality. The child should be immersed in an environment rich in oral communication; each child's learning style should be considered, with intervention being guided by what works best. For children who are unable to achieve proficiency in spoken language after implantation, early introduction of signed communication (either alone or to supplement oral communication) becomes necessary.[68]

Regardless of the strategy used, the outcome varies considerably, despite the successful restoration of peripheral hearing. This variation reflects the effect of auditory deprivation on the multiple information-processing subsystems and neural circuits underpinning the development of spoken language.[38] Children with cochlear implants have many cognitive processes that differ fundamentally from those of age-matched controls with normal hearing. For example, their short-term memory shows important processing delays in scanning and retrieving verbal information, and their working memory is characterized by a reduced rate of encoding phonologic and written information.[38] Even on nonauditory tasks, such as sustained attention and visual-sequence learning, deaf children underperform as compared with their peers with normal hearing.[36-38] New approaches to auditory training that help a child with a cochlear implant reallocate attentional resources to the auditory system and exploit multimodal interactions may improve language outcomes.

Auditory training programs embedded in computer games may enhance language outcomes. Such packages, which are designed to improve speech perception by presenting a series of increasingly challenging discrimination tasks,[69,70] may be made available through cell-phone-based portable technologies or through the Internet.[71] The Internet can also be used to program the device remotely, thus reducing the need for visits to the center where the child received the cochlear implant.[72] Remote technologies may also help to deliver care when there is a shortage of skilled personnel (e.g., in developing countries).

OUTCOMES OF COCHLEAR IMPLANTATION

SPEECH AND LANGUAGE

One of the most gratifying outcomes of cochlear implantation is the restoration of a child's ability to understand speech. Technological advances, an earlier age at intervention and implantation, and relaxation of audiologic criteria to permit implantation of cochlear implants in children with limited residual hearing have all improved spoken-language outcomes.

Children who have received cochlear implants at a younger age have faster and more age-appropriate development of spoken language than children who have received implants later.[6,73] The effect of cochlear implantation on spoken-language outcomes is currently being evaluated in a multisite, prospective study in the United States.[74] Given the many factors that contribute to the process of learning language, it is critical that studies of language acquisition account for covariates and adjust for confounding factors. Long-term studies (10 to 14 years after implantation) have shown mean word-recognition scores of 80% in a quiet setting and 45% in a noisy setting[75]; 60% of children with cochlear implants are able to use a telephone with a familiar speaker. However, many children continue to have difficulties with the more complex language constructs, such as syntax, semantics, and pragmatics.[76] Measures of speech production after cochlear implantation confirm continuing improvements in the years after the procedure. Ten years after implantation, about 77% of children had speech that was intelligible to a listener.[77] The presence of additional disabilities that affect generalized developmental learning greatly constrains language development[78]; however, children with such additional disabilities still have benefits in terms of enhanced environmental awareness and social engagement.

There is considerable variation in spoken-lan-

guage abilities among children with cochlear implants.[79] The best outcomes are obtained after early cochlear implantation, especially among children with normal cognition who are constantly exposed to high-quality spoken language and are supported by committed parents and caregivers.

EDUCATION

There has long been disagreement about the optimal education for deaf children; some advocate for fostering oral speech, whereas others advocate for sign language. Without previous knowledge of deafness, many people are surprised by how much hearing loss affects educational attainment. That the average 18-to-19-year-old deaf student reads at a level of an average 8-to-9-year-old student with normal hearing illustrates the devastating impact of deafness on the acquisition of such a vital life skill.[80] Deaf children have great difficulties in matching the phonologic content of spoken language to written language.[81]

Evidence suggests that hearing-impaired children with normal cognition who receive cochlear implants early have enhanced reading ability[57,82]; the literacy skills of most children with implants, however, still lag well behind children of the same age who have normal hearing.[1] For many children with implants, mainstream schooling has become a realistic option. Success depends on having sufficient proficiency in spoken language and possessing the cognitive abilities to make regular school feasible. Even if children with implants surpass deaf children of the same age who have hearing aids and similar hearing losses, implants do not guarantee the development of academic skills that are similar to those of children with normal hearing.[83]

PSYCHOSOCIAL ISSUES

Psychological disorders are two to five times as common in deaf children as in children with normal hearing, and the prevalence is especially high among deaf children with additional disabilities or disruption in the family, such as divorce.[4] When parents with normal hearing receive the news that their child is deaf, the trauma can be so great as to provoke a bereavement response, and such parental upset can be sensed by the deaf child. Deafness may also disrupt mother-child bonding, paving the way for emotional difficulties later. Adolescence presents additional challenges; many deaf teens have reduced self-esteem and uncertainty about their identity. Do cochlear implants enhance psychosocial adaptation in deaf children? Certainly, for children who develop competence in spoken language and have strong family support, the outcomes are favorable.[84] However, no well-validated, health-related quality-of-life instruments that are specific to deafness are currently available to formally explore such issues and capture the diversity of outcomes.[85] Psychosocial research is needed and appears to be a vital step in improving outcomes for hearing-impaired children.[86]

FUTURE DEVELOPMENTS

Research in the molecular biology of hearing loss could deliver low-cost tools based on DNA chips to screen populations for the most common gene mutations causing deafness. DNA sequences that render certain persons susceptible to environmental agents (e.g., noise and ototoxic drugs) causing deafness may emerge, and molecular markers identifying the children at greatest risk for the development of a hearing loss in later life may be discovered. The possibility of treating deafness by triggering hair-cell regeneration or through stem-cell therapy remains an elusive goal at present but is likely to become a reality in the decades ahead. Cochlear implants are likely to become multifunctional, combining drug-delivery (e.g., neurotrophic factors) and cell-delivery capabilities to rescue spiral ganglion cells or even generate new ones[87]; innovations in design will allow better encoding of temporal fine structure, improving speech perception against background noise and the ability to enjoy music.[88,89] Hearing conservation through cochlear implantation should allow the synergistic combination of acoustical and electrical stimulation of the same ear[90]; further technological advances[87,91,92] will probably improve the outcome of implantation. Objective markers of brain maturation[93] or response to complex sounds[94] (e.g., with the use of electroencephalographic measures) may guide future decisions about candidacy for cochlear implantation. Actively harnessing the brain's computational capacity through the development of multimodal cognitive "brain training" exercises will probably further enhance outcomes.[69,70] Emerging evidence suggests that auditory brain-stem implantation may be of value in children who do not have cochlear nerves.[95]

SUMMARY

Profound childhood deafness is not just a sensory loss but has a lifelong effect on many levels of brain function. Many developments are transforming the management of profound deafness; these include universal neonatal screening, early intervention, and advances in diagnostic neuroaudiology, molecular biology, and integrative neuroscience. Cochlear implantation has transformed developmental outcomes, providing access to spoken language for the majority of children who receive implants early in life. Laboratory investigations have elaborated the neurobiologic processes that follow auditory deprivation, particularly a physiological uncoupling of the auditory system, resulting in degradation of its functional connectivity with key centers in the brain.

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