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Recent Developments in Bionic Hearing Restoration from the Round Window to the Inferior Colliculus

Colletti V^{*}, Shannon RV^{**}, Mandalà M^{*}, Carner M^{*}, Veronese S^{*}, Colletti L^{*}

^{*} ENT Department, University of Verona, Italy

^{**} House Ear Institute

Modern auditory prostheses range from implants that impart mechanical energy to the cochlea via the ossicular chain or bypassing a damaged ossicular chain via the round window (round window implants) to implants that, bypassing severely damaged inner ear cells, electrically stimulate the auditory nerve within the cochlea (cochlear implants). Other auditory prosthesis are suitable for electrical stimulation of the central auditory pathways: the auditory brainstem implants which, bypassing a damaged cochlea and auditory nerve auditory, stimulate the brainstem nuclei, and the inferior colliculus implant or auditory midbrain implant which, bypassing damaged brainstem nuclei stimulate the inferior colliculus.

With the refinements in implant technology, patient selection criteria for the various different implant devices need to be periodically reconsidered with a view to obtaining increasingly high levels of speech recognition for the different etiologies. We review the latest outcomes, obtained with various implant devices, and propose guidelines for device selection for different etiologies of deafness.

Key words : electrical stimulation of the central auditory system, round window mechanical stimulation, cochlear and auditory brainstem implants, inferior colliculus implants

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INTRODUCTION

Auditory restoration with implant technology has been a great success story in otology and the ultimate limits of the technology are still not known. Today there are a wide variety of approaches and auditory prostheses for restoration of hearing, each with a precise indication for specific degrees and sites of hearing loss (Figure 1). Modern auditory prostheses range from implants that impart mechanical energy to the cochlea (middle ear implants, MEIs) via the ossicular chain or bypass a damaged ossicular chain and vibrate the round window directly (round window implants, RWI). Cochlear implants (CIs) bypass damaged inner ear cells and electrically stimulate the auditory nerve

within the cochlea. The auditory brainstem implant (ABI) bypasses a damaged cochlea and auditory nerve auditory and directly stimulate the brainstem nuclei. The inferior colliculus implant (ICI) or auditory midbrain implant (AMI) bypass damaged brainstem nuclei to stimulate the inferior colliculus in the midbrain.

With the refinements in implant technology, patient selection criteria for the various implant devices need to be periodically reconsidered with a view to obtaining increasingly high levels of speech recognition for the different etiologies. In general it is thought that the more peripheral the implant, the better the chance of success. Indeed the more peripheral implants (RW, MEIs, CIs and ABIs in non-NF2 patients) have been

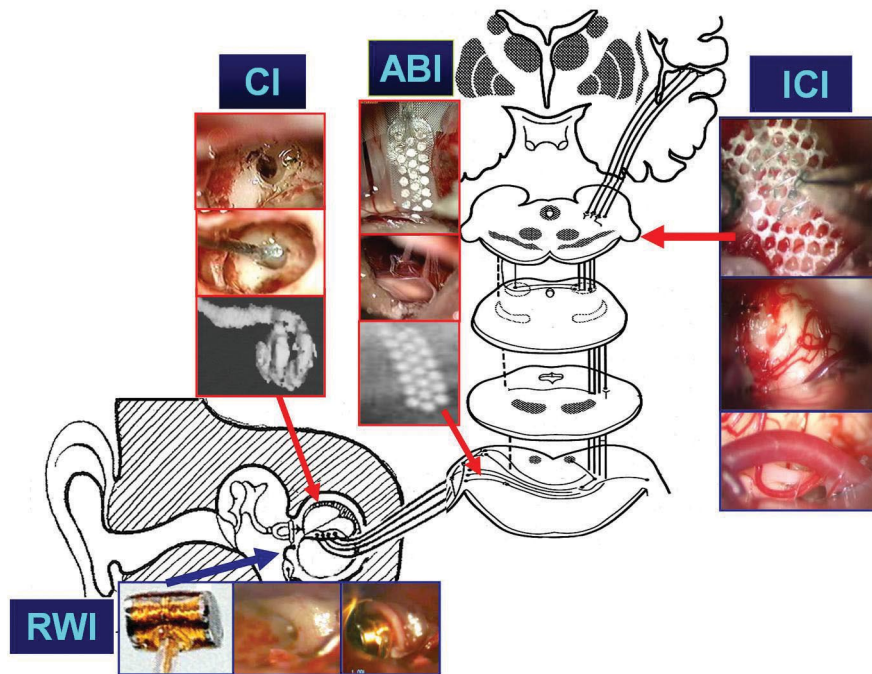


Figure 1. Schematic diagram of modern auditory prosthesis from the round window implantation, RWI to CI, to ABI and to ICI. The arrows indicate the respective sites of implantation. The sequence of the picture summarizes the surgical procedure for each implantation.

highly successful. More central implants (ICI and AMI and ABI in NF2 patients) have so far not resulted in high speech understanding performances.

In this paper we review the latest outcomes obtained with the various types of implants available and guidelines are proposed for device selection for different etiologies.

MIDDLE EAR IMPLANTS (MEIs)

MEIs have been developed in otology to circumvent some of the common problems of hearing aid users, such as annoying feedback, poor sound quality, stigma of handicap, and poor speech understanding in background noise. Successful MEI application in patients with sensorineural hearing loss has been reported by various authors¹⁻⁴. Recently, Colletti et al.⁵⁻⁷ have extended the indications of MEIs to patients with extensive damage of the ossicular chain and moderate to severe conductive and mixed hearing loss who could not benefit from conventional BCHA and ACHA and BAHA (e. g. in children less than 2 years). For many years, patients suffering from chronic suppurative

otitis media (CSOM) or with radical cavity (RC) or with severe malformation of the external and middle ear with mild-to-severe hearing impairment (congenital aural atresia, CAA) have been treated unsatisfactorily with ossiculoplasties (OPL)⁸ procedures and/or air and bone conductive hearing aids. The difficulty of achieving significant and stable improvements of hearing by means of OPL is related to the biology, pathophysiology, and progressive pathology that characterize CSOM, RC and severe ear malformations. Surgical treatment of functional deficits in severely malformed ears is also still not satisfactory either in residual conductive or mixed hearing loss. For patients with an absent or severely altered ossicular chain the location of the floating mass transducer (FMT) has been shifted from the standard position on the long process of the incus to a novel location onto the RW, thereby bypassing the diseased and malformed ossicular chain (Figure 2). Details of device specifications and surgical approaches for placing RWI are presented in previous papers⁵⁻⁷.

The outcomes of RWI^{5-7,9} demonstrate that with the

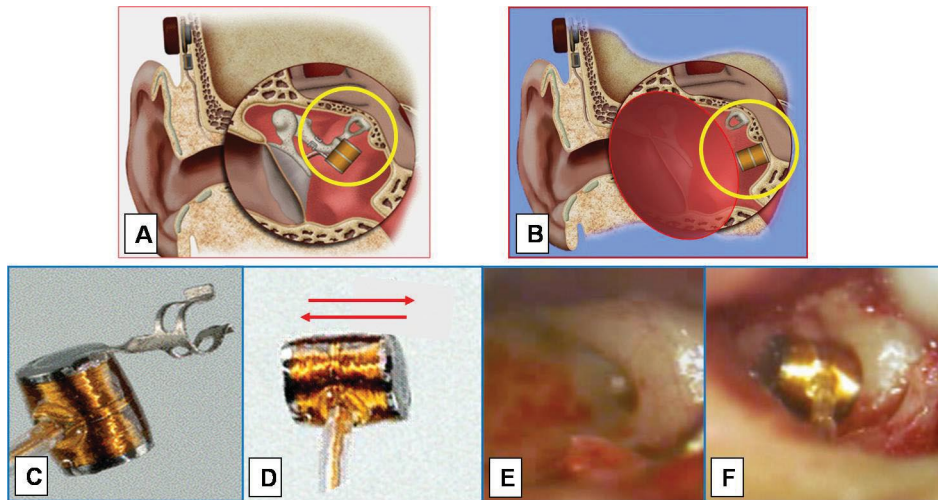


Figure 2. Technical details for fitting the Medel Vibrant Soundbridge middle ear implant. The floating mass transducer is fixed on the long process of the incus, as originally developed (A) and located onto the round window as proposed by Colletti et al. in 2005 (B) after cutting the crimper. The crimper of the FMT allows the fixation on the incus (C) and is removed (D) to make the FMT suitable for RW implantation (C-D). The RW area (E). FMT on the RW (F).

present technique congenital or acquired middle ear defects associated with permanent dysfunctions of the middle ear are not an issue because the RW area containing the FMT is completely obliterated by either muscle or fascia. Migration of the FMT from its position on the RW could still be a long term complication associated with the surgery, but it has not been reported up to 48 months of follow-up observation. Mixed hearing impairments of any etiology with sensorineural components of at least 50-60 dB can be treated successfully with RWI. The majority of patients can achieve 100% intelligibility in speech audiometry and high levels of intelligibility at normal conversational speech levels^{5-7, 9}. The post-operative results suggest that RWI with the Med-El VSB offers a viable treatment option for patients with severe mixed hearing loss regardless of the etiology (congenital or acquired) and previous surgeries. The RWI bypasses the normal conductive pathway to the cochlea by delivering vibratory energy directly to the cochlea via the RW and allows compensating for the conductive and, with amplification, for sensorineural component.

COCHLEAR IMPLANTS

CIs represent today the most widely adopted tech-

nique to restore auditory function in severe to profound hearing loss patients (Figure 3). With improving outcomes, cochlear implant candidacy has been expanded to additional patient populations with significant residual hearing¹⁰⁻¹². Over the last three decades, CIs have improved steadily to the point where the average sentence recognition with modern multi-channel devices is nearly 100% correct¹³. Many patients are able to recognize 100% of simple sentences presented in quiet and must now be tested in conditions of added noise to evaluate the limits of their performance. Despite an impoverished pattern of peripheral activation both in term of spectrum and distortion of the information, it is now well known that the central auditory processing can achieve high levels of speech pattern recognition. Recent findings show further improvements in performance when combining electric stimulation delivered via CI with residual acoustic hearing¹⁴.

Binaural hearing is important for localizing sounds in space and for understanding speech in noisy surroundings. Bilateral CIs allow some localization of sounds as well as a modest improvement of speech recognition in noise¹⁵.

Infants fitted with CIs before one year of age show normal speech and language development¹⁶ and are

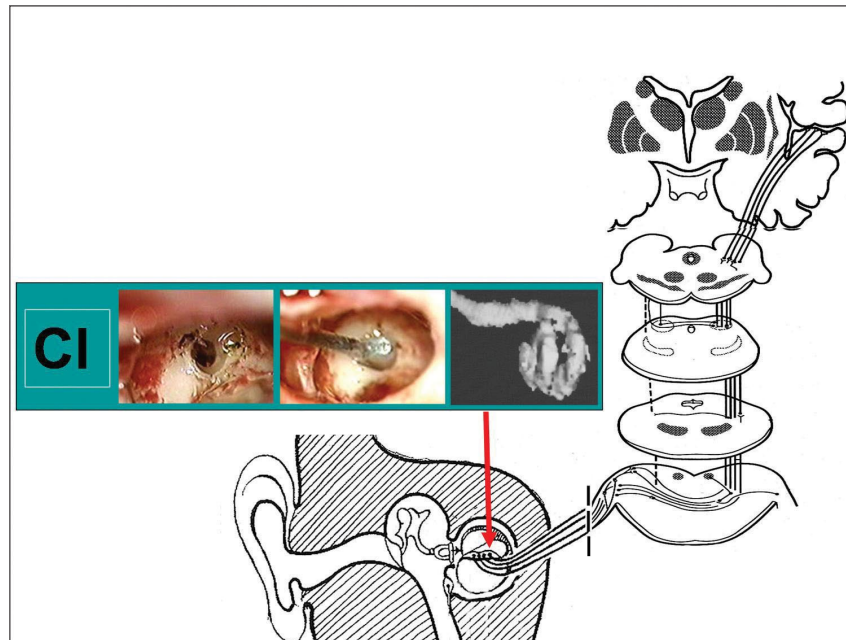


Figure 3. CI procedure and site of implantation in the auditory system. Cochleostomy is shown (A). Insertion of the electrodes array through the cochleostomy (B). Volume rendering CT reconstruction of the electrodes array after insertion into the cochlea (C).

usually able to attain a normal educational experience with normally hearing peers. It is now clear that congenitally deaf children who receive the implant as adolescents derive limited benefit from the implant even after many years of daily use. The earliest possible identification of hearing loss and implantations are important for optimal use of CIs in children.

Although overall CI speech recognition is excellent, the variability in performance across listeners is still quite large. Physiological factors may be responsible for the wide variation in CI outcomes. It is possible that low-performing CI patients have damage to the residual VIII nerve that limits their speech recognition. It is worth considering whether some patients who perform poorly with a CI might achieve a better out-

come with an ABI.

AUDITORY BRAINSTEM IMPLANTS

Restoring hearing function in neurofibromatosis type 2 (NF2) patients was the main reason for the development of ABIs¹⁷⁻²⁰. Removal of a vestibular schwannoma, that characterize this genetic disorder, usually severs the auditory nerve so that a CI is of no value (Figure 4). Following tumor removal, either via a translabyrinthine approach or via a retrosigmoid approach, a multichannel ABI electrode array can be placed on the surface of the cochlear nucleus (CN). In general, speech recognition performance of NF2 ABI patients is much poorer than the excellent level commonly achieved with CIs. Most NF2 ABI patients recognize less than 5% of the words in sentences, even

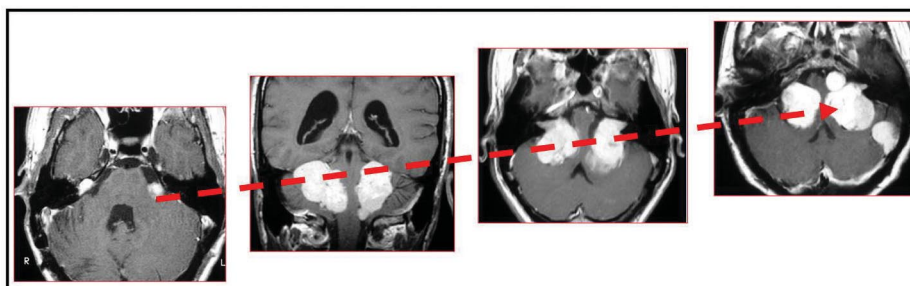


Figure 4. MRIs showing bilateral schwannomas of different size in the cerebellopontine angle of patients with NF2. As a result of the tumor growths, NF2 patients face the prospect of total bilateral deafness.

after many years of ABI experience. NF2 ABI patients receive sound awareness, some environmental sound discrimination-identification, and a significant improvement in face-to-face communication when the ABI is combined with lip reading²¹⁻²³. In an attempt to improve ABI performance a new ABI was developed with penetrating microelectrodes (PABI) to improve the selective activation of the tonotopic strata in the cochlear nucleus. Unfortunately, patients fitted with the PABI, despite achieving the goals of low stimulation thresholds, excellent tonotopic selectivity and distinct pitch across electrodes, have not demonstrated the expected high levels of open-set speech recognition²⁴.

Selection criteria for ABI candidates have been today widely expanded to include non-tumoral (NT) patients^{25, 26} (Figure 5): patients with skull base fracture, severe ossification of the cochlea or modiolus and lost function of the auditory nerve. Many NT ABI users are capable of excellent speech understanding, similar to the high levels seen in CIs²⁷⁻²⁸.

The limited performance of ABIs in NF2 patients with both with conventional and penetrating electrodes, combined with the excellent speech recognition in some NT ABI patients, suggest that the tonotopic selectivity of the surface ABI electrode was not the

factor limiting NF2 ABI performance. It is reasonable to assume that the NF2 tumor growth and/or removal may damage neural elements in the cochlear nucleus that are essential for speech recognition. The excellent results obtained by NT ABI patients demonstrate that effective prosthetic stimulation is possible at the CN, despite the missing cochlea and auditory nerve processing and the highly unnatural activation patterns in the CN.

Based on these excellent speech recognition results in adults, clinical trials are now underway to evaluate the efficacy of ABIs in children²⁹⁻³². Children with cochlear nerve aplasia or severe ossification following meningitis may have no auditory nerve available for activation by a CI. In some cases, children with these conditions have received CIs without benefit³⁰. Initial results with ABI in children show sound awareness, an increase in vocal production, and discrimination of sounds²⁹⁻³². In one child with congenital absence of cochlea and auditory nerve, the ABI provided similar progress on a battery of auditory tasks compared to children of the same age implanted with CIs³³. Some children with ABIs have been able to achieve open-set speech recognition²⁹. Furthermore, preliminary results show that auditory brainstem implants contribute to the development of auditory-verbal skills in prelingual-

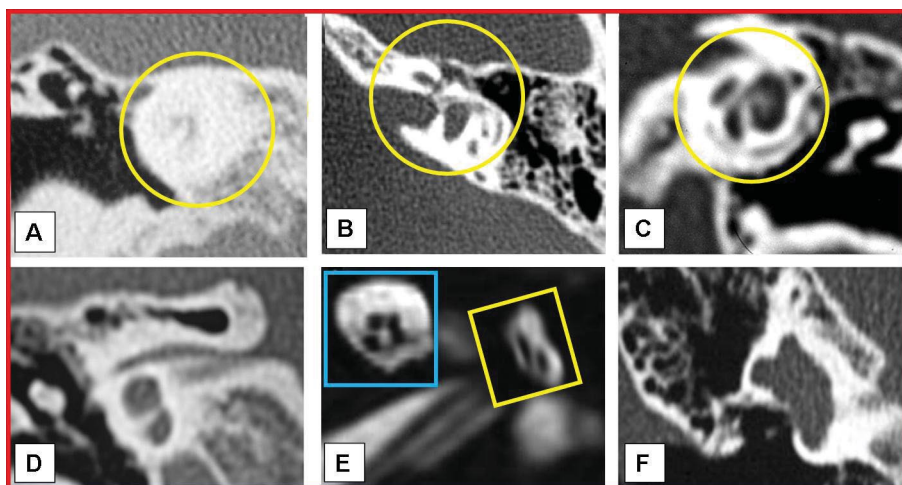


Figure 5. Extended ABI indications based on CT and MRI findings. CT scan showing severe post-meningitic cochlear ossification (A). CT scan demonstrating post-traumatic temporal bone fracture (B). CT scan of advanced otosclerosis with severe demineralisation of the otic capsule (C). CT scan showing a narrow internal auditory canal associated with cochlear malformation (D). MRI showing an internal auditory canal with cochlear nerve aplasia (yellow rectangle). A normal internal auditory canal is represented in the blue rectangle (E). CT scan demonstrating a severe cochlear malformation: common cavity (F).

ly deaf children³¹.

INFERIOR COLLICULUS IMPLANTS (ICI)

To date, ICIs represent the only option that might restore hearing in NF2 patients bypassing the CN. Damage at the level of the CN could be the reason for the limited open set speech understanding in ABI NF2 patients. If this is the case, it might still be possible to restore speech recognition by bypassing the region of

damage and electrically stimulating the auditory mid-brain at the inferior colliculus (IC). Similarly to the development of the surface and penetrating ABI, different strategies have been proposed to deliver electric stimulation to the IC. The first successful case of hearing produced by electrical stimulation of the human midbrain through ICI with surface electrodes was described by Colletti et al. in 2007³⁴ (Figure 7). A clinical trial is underway with the auditory midbrain

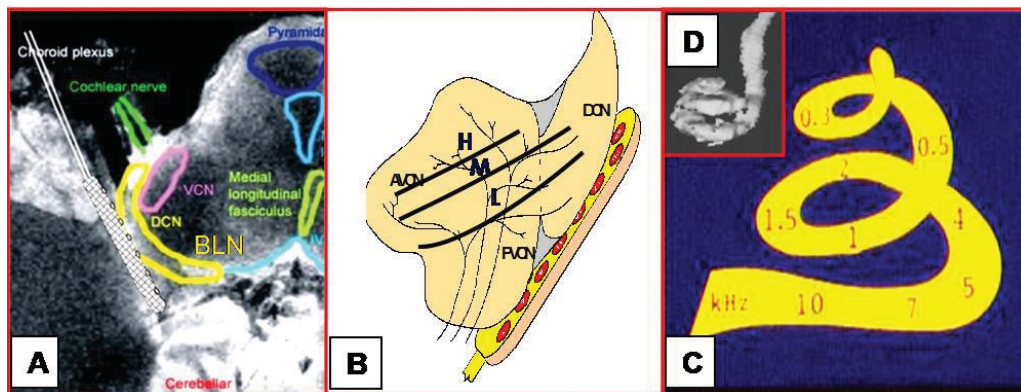


Figure 6. MRI imaging of the cerebello-pontine angle showing the anatomical location of the cochlear nucleus and the area of ABI insertion (A). Iso-frequency regions run at a shallow angle almost parallel to the CN surface and ABI location (B). Schematic diagram showing the distribution of tonotopic organization along the turns of the cochlea (C). Volume rendering CT reconstruction of the electrodes array after insertion into the cochlea (D).

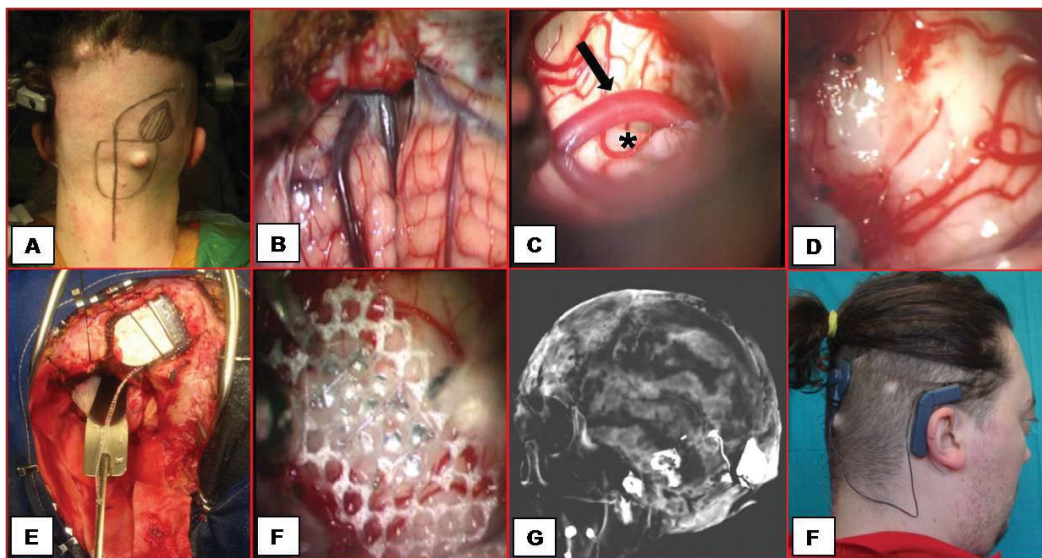


Figure 7. Sequence of pictures obtained during the infratentorial supracerebellar median surgical approach for ICI procedure. Long midline incision from the spinous process of third vertebrae body up to the occipital region (A). After suboccipital median craniotomy the cerebellar vermis is freed from the tentorium and gently depressed to establish an unobstructed corridor to the pineal region (B). The exposure of the Cranial Nerve IV (arrow) and superior cerebellar artery (star) to identify and expose the IC (C). Placement of the internal processor of the ICI (D). The electrodes array is placed on the dorsal surface of the inferior colliculus area (E). CT scan performed after the ICI procedure (F). The patient is wearing the external processor of his implant (G).

implant (AMI), which uses a penetrating electrode array³⁵. Unfortunately, so far, no patients have achieved significant open set speech recognition with electrical stimulation of the IC. Future research is necessary to evaluate the potential of electric stimulation of the auditory midbrain for restoring useful hearing.

CONCLUSIONS

Guidelines proposed for device selection for different etiologies of hearing loss are summarized in table 1. At almost every stage of prosthesis development the potential benefit of the new technology has been underestimated. For decades OPL and hearing aids have been the only procedures to restore hearing in patients suffering from conductive or mixed hearing loss due to severe acquired or congenital ossicular chain defects. For many years it was thought that the CI would never allow normal conversation by telephone, but that is now an expected outcome. For long time it was believed that ABI would never allow the same level of speech recognition as CIs, but now many NT ABI show comparable performance to CIs. Will the RWI replace OPLs procedures as well as hearing aids and BAHA as the procedure of choice in congenital or acquired mixed hearing loss? Will binaural CI implantation under 1 year of age become the standard procedure for deaf children with normal auditory

nerve function? Will children with cochlear and cochlear nerve disorders obtain speech understanding outcomes with the ABI comparable to those implanted with CI at the same age? Will NF2 patients achieve open set speech recognition with ICI or AMI? Implant technology has been a great success story in Otolaryngology and the ultimate limits of the technology are still not known. More implant devices and locations are now available and more research is needed to further refine the process of matching an individual patient to the most appropriate implant device.

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Table1 A proposed framework for selecting the appropriate implant based on the site of damage to the auditory system.

SITE OF DAMAGE	IMPLANT TYPE
Congenital or acquired moderate to severe mixed hearing loss	RWI
Hair cells	CI
Mild ossification in the cochlea	Drill-out CI
Severe ossification in coclea/modiolus/internal auditory meatus	CI or ABI
Auditory neuropathy	CI or ABI
Skull Base fractures that sever VIII bilaterally	ABI
NF2 with VIII nerve preservation	CI(?) or ABI or ICI
NF2 small tumor	ABI or ICI
NF2 tumor > 2 cm or CN Damage	ICI

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