Curr Surg Rep (2014) 2:75
DOI 10.1007/s40137-014-0075-9

Cochlear Implants in Single-Sided Deafness

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Abstract Single-sided deafness presents a unique challenge to otolaryngologists and audiologists. While the normal hearing ear may allow listeners to perform adequately on audiometric screening, individuals with only one functioning cochlea suffer when resolving speech in noisy environments and in sound localization—which both contribute to a reduced quality of life. Though there are a variety of strategies that provide contralateral routing of sound signals, the cochlear implant is the only treatment to truly restore binaural hearing. Only very recently has cochlear implantation (CI) for single-sided deafness begun in earnest, with encouraging results that demonstrate the strengths and pitfalls of implantation over traditional extracochlear methods. The purpose of this review is to update the field by emphasizing binaural benefits, discussing historical treatments of single-sided deafness, critically evaluating recent data on outcomes of CI for single-sided deafness, and recommending indications for cochlear implants in single-sided deafness in children, adults, and subjects with concurrent ipsilateral tinnitus.

Keywords Single-sided deafness · Unilateral cochlear implant · Speech in noise · Sound localization · Binaural hearing · Unilateral tinnitus

This article is part of the Topical Collection on *Ear Surgery*.

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Introduction

Single-Sided Deafness

Single-sided sensorineural deafness, hereafter referred to as SSD, is defined as a severe-to-profound hearing loss (>70 dB normal hearing level, nHL) in one ear with preserved audiometric thresholds in the contralateral ear. Though the precise incidence and prevalence of SSD is unknown, its prevalence has been estimated anywhere from 3 to 6 % of the population [1], and among children and teenagers ranges from 2 to 5/1000 [2••]. Incidence is estimated at 1 in 3,700 newborns being born with congenital SSD [3] and the incidence increases with age [4]. Sudden sensorineural hearing loss (SSNHL) is the most common cause of SSD, though a myriad of other causes are known, including intractable Ménière's disease, unilateral vestibular schwannoma, trauma leading to temporal bone fractures, unilateral noise damage, and ototoxic drug exposure.

Historically, hearing limitations posed by SSD were largely understated, as it was assumed that one normal hearing ear provided sufficient auditory input for a relatively normal hearing experience [5]. It is now widely known that individuals with SSD face significant challenges to hearing in complex or noisy environments and in localizing or lateralizing sound sources [6•]. Not surprisingly, individuals who suffer from SSD consistently rate lower in quality of life inventories such as the speech, spatial, and qualities hearing scale (SSQ) [7–10]. Deficiencies in speech comprehension are chiefly a result of impaired binaural squelch and summation, whereas deficiencies in azimuth localization are mostly due to impaired calculation of interaural phase differences (IPDs), time differences, and intensity differences [6•, 7, 10, 11••].

Binaural Benefit to Speech Perception

Speech comprehension is vastly benefited from having two separate sound inputs, specifically with respect to binaural squelch and summation. The squelch effect describes the ability of the brain to selectively filter noise from a desired sound when there is binaural hearing input, particularly in a hearing environment where noise and speech are originating from different azimuthal or vertical locations, due to interaural level, phase, and time differences. The advantage conferred with binaural hearing due to squelch is a 2–5 dB gain in signal-to-noise ratio (SNR) in speech discrimination testing [6•, 7]. Binaural summation is a psychoacoustic phenomenon wherein one acoustic stimulus reaching two ears provides the brain with redundant information that adds to the net perceived sound. This provides an additive effect to higher-order processing centers and results in a 2–6 dB improvement in speech reception thresholds compared to the monaural condition [6•, 7]. The head shadow effect refers to the reduction in intensity of sound reaching an ear opposite to the side of the sound source because the head functions as a physical attenuator. In subjects with SSD, speech and other sounds projecting directly at the deaf ear are primarily detected at the functioning contralateral cochlea. However, because of the head shadow effect, these sounds are up to 10–16 dB lower than the signal source, which makes speech more difficult to understand, especially with ambient background noise [7]. In addition to diminished sound levels at the contralateral hearing ear, limitations in comparing interaural sound intensity levels also affect a subject's ability to localize sounds.

Binaural Input Necessary for Horizontal Sound Localization

Sounds produced at an azimuth off-axis will arrive in each ear at slightly different times, at slightly different phases, and due to the head shadow effect, at slightly different intensities. Different binaural mechanisms are used to help localize sounds in the horizontal plane, depending on the acoustic signal quality (Table 1). For low stimulus frequencies, auditory phase resolution is robust enough to allow cross-correlation of interaural phase differences in order to infer the azimuthal location [6•]. The ability to differentiate between phases is maximized when the peak-to-peak distance of a single wave spans the largest physical range of place-coded neurons in the superior olivary complex. As such, IPD is best suited to localize low-frequency sources and becomes less effective at frequencies greater than 800 Hz, at an approximate wavelength that is shorter than twice the interaural distance of the human head [6•]. For acoustic stimuli greater than 16 kHz, phase resolution is nearly impossible. As such, the time delay between signal detection of each ear, called the interaural time difference (ITD), helps inter the sound source in the horizontal plane. Frequencies within the range of 800 Hz to 16 kHz use a combination of IPD and ITD. In addition to IPD and ITD, larger differences in sound intensity between ears correlate with larger azimuths in a frequency-dependent manner. A normally functioning auditory system will expect attenuation between ears because of the head shadow attenuation and correlate larger interaural intensity differences with larger azimuth angles. Without two functioning cochleae, the head shadow effect, along with other binaural mechanisms, are ineffective in aiding horizontal localization.

Table 1 Several independent mechanisms of binaural integration assist in horizontal sound localization, depending on the acoustic stimulus.

Acoustic quality	Binaural azimuth localization mechanism
Low frequency (<800 Hz)	Interaural phase difference
Mid frequency (800 Hz to 16 kHz)	Combination of interaural phase difference and interaural time difference
High frequency (>16 kHz)	Interaural time difference
High intensity (any F)	Interaural level difference

Summarized from content in [6•]

Statement of Purpose

In the context of a recently changing political and healthcare climate, cochlear implantation (CI) candidacy has expanded to include subjects with relatively preserved hearing thresholds but reduced speech perception (Hybrid CI or electric acoustic stimulation). Demonstrable benefits of hearing in noise, sound localization, and quality of life from studies on CI for SSD similarly argue for an expansion of the indications for implantation. The purpose of this review is to critically evaluate the most recent literature on CI for SSD and to form conclusions regarding CI for subjects with SSD that will inform clinical practice and policy makers. Though the focus of this report is not to investigate the efficacy of unilateral CI for tinnitus reduction, because many of the studies were conducted with subjects who suffered from SSD and concomitant ipsilateral tinnitus, some discussion of the efficacy of CI for tinnitus reduction will follow as well.

Treatments for Single-Sided Deafness

Historical Treatments

In the past, treatment modalities to restore binaural hearing relied on amplification devices that transmit sound, through air or bone, from the deafened ear to the contralateral, normal ear. Such technologies include air conduction contralateral routing of sound (CROS), transcranial CROS (t-CROS), and bone conduction technologies including the Bone-Anchored Hearing Appliance (Baha, Cochlear, Inc, Macquarie University, NSW, Australia, both percutaneous and transcutaneous solutions), the Sophono device (Sophono, Inc, Boulder, CO, USA), and the SoundBite dental conduction device (Sonitus Medical, Inc, San Mateo, CA, USA). An air conduction CROS hearing aid (HA) is decades-old technology that consists of a microphone placed in the deafened ear which transmits sound via a wire or wirelessly to a receiver that is placed in the normal hearing aid, thereby averting the negative head shadow effect in monaural listeners. Due to inexpensiveness, and ease of use and fitting, CROS hearing aids typically are the first line intervention for SSD [12]. Though users have endorsed subjective benefits from restoring the head shadow effect by allowing sound awareness from the deaf side [6•, 12, 13], this success has been tempered by its drawbacks, including the need to occlude the better ear canal and relatively poor overall improvement in hearing experience, particularly in regard to hearing in noise and sound localization [13, 14]. Transcranial CROS transmits a signal received by an air conduction hearing aid in the deaf ear to the contralateral cochlea via bone conduction. Theoretically, transcranial CROS represents an improvement over air conduction CROS because of a smaller reduction in the sound signal across bone versus electronically or wirelessly [6•, 12]. However, its use is not well established because there are only a limited number of trials with small sample sizes and conflicting data regarding improvements in speech perception, sound localization, and patient satisfaction relative to air conduction CROS [15, 16].

More recently, the Baha has gained traction for auditory rehabilitation in SSD. The Baha is a type of osseointegrated bone conduction device wherein an osseointegrated titanium implant is placed into the skull. An external sound processer transmits signals from the deafened ear through the titanium implant and eventually to the normal hearing cochlea on the contralateral side via bone conduction. In contrast to t-CROS, there is a voluminous literature on the efficacy of Baha. In addition to consistent reports of patient satisfaction and increased speech perception in noise, studies show moderate improvements in sound localization ability after Baha [17–19]. In comparative studies on the efficacy of Baha versus CROS, investigators have uniformly recommended Baha over CROS or t-CROS for the management of SSD [20–22]. While hearing aid and conduction technologies simply transfer sounds to the single functioning cochlea, CI in the deaf ear uniquely affords the opportunity to utilize two independent cochlear generators.

Cochlear Implantation in Single-Sided Deafness

Despite the improvement in hearing in noise and localization reported by Baha and CROS users, CI implantation holds promise as the definitive treatment for SSD. Although the first studies of unilateral CI implantation, dating back to 1957, were in patients with bilateral deafness [23–25], CI for SSD began fortuitously in the 2000s as an experimental treatment for incapacitating and intractable tinnitus [26] when users reported marked gains in sound localization and speech perception in complex listening environments [7]. Because hearing loss co-exists in up to 85 % of individuals with tinnitus [27], hearing improvements were likewise reported in subsequent studies of CI in tinnitus patients with ipsilateral SSD [9, 10, 28]. The small number of studies that directly compare benefits with CI versus Baha or CROS strongly endorse CI over these more rudimentary treatment strategies [10, 29]. A number of studies have since reported on substantial improvements in sound localization, hearing in noise, and quality of life measures due to restoration of binaural squelch, summation, and the head shadow effect [6•, 8, 10, 30]. Despite these advances, candidate selection and anticipated benefits are not as clear and predictable as implanting a bilaterally deaf individual.

Speech Benefits after Unilateral Cochlear Implantation

Several recent studies investigate the utility of CI for SSD in both pediatric (Table 2) and adult (Table 3) populations. In short, the main benefits for speech perception with regard to a unilateral implant for SSD are an increased speech understanding, especially in noisy environments, and a decreased effort to hear [6•, 11••].

When solely analyzing the SSD ear, Hansen et al. report a 28 % increase in CNC scores and a 40 % increase in AzBio sentences when comparing unilateral free-field audio stimulation pre-implantation to electrical CI stimulation post-implantation [31]. Despite this increase in cochlear function of the SSD ear, the binaural benefits of free-field speech comprehension gleaned after implantation are limited in quiet environments, likely because the normal hearing ear doesn't need to rely on binaural squelch and summation when noise is not present. In a case series with 3 subjects implanted for SSD, Firszt et al. reported only 1 patient who showed statistically significant improvements in CNC speech scores in the bimodal, quiet condition compared to either unilateral NH or CI-alone conditions [8]. In the same study, adding noise to the speech tests resulted in 2/3 patients showing significantly better CNC scores in the bilateral condition than solely with either the normal hearing ear alone.

Table 2 Outcomes of children and adolescents with SSD receiving unilateral CI

Authors and year	Study type	Population	Etiology	PTA in nonimplanted ear in dB nHL (range)[a]	Device brand	Speech perception results	Localization/ lateralization results	Subjective improvements	Comments
Hassepass et al. 2013 [2••]	Case series	3 children with SSD	Mixed	13 (10–18)	Cochlear	Significant binaural improvement in noise for several configurations, including speech and noise at the front and speech at the CI ear and noise at the NH ear	Significant improvement in 2/2 tested at 12 mos	Evaluations suggest that children like the CI more than HAs	One subject had difficulty concentrating on subjective questionnaires
Plontke et al. 2013 [36]	Case report	One 8 year child	Post-traumatic	n/a, “normal”	Cochlear	In quiet, multisyllabic word score was 100 % within 6 mo and monosyllabic word score was 90 % within 6 mo. In noise, significant improvement in OLSA score within 3 mo	Slight reduction in angle detection error when noise presented frontally or on CI side. No improvement on NH side	No objective measures, but describes high degree of patient satisfaction	Indication for CI was lateral skull base fracture with imminent fibrosis
Cadieux et al. 2013 [37]	Case series	9 adolescents with asymmetric HL; 3 with HA	Mixed	53 (25–75)	3 ABC 2 Cochlear	Significant gains in quiet and in noise for bimodal vs. CI or HA alone, but only for 3 subjects with long CI use and w/o congenital HL	Significant increases with bimodal vs. CI or HA alone, only for 3 subjects with long CI use and w/o congenital HL	n/a	2 subjects with congenital HL did worse, and were implanted at 12 and 15 years old. All have HL on contralateral side
Tzifa and Hanvey 2013 [38]	Case series	8 children with SSD, all HA users	Mixed	59 (38–75)	Cochlear	1. 7/7 tested with increase in CAPII scores post-op 2. 6/8 with improvement in SIR scores post-op	n/a	n/a	All had HL on contralateral side (2/8 with severe HL)

[a] For manuscripts not reporting contralateral unaided PTA, these values were calculated from audiograms appearing in the text, and are 3-tone averages (500, 1000, and 2000 Hz)

ABC Advanced Bionics Corporation, *AzBio* Arizona State University/Advanced Bionics Corportation sentence test, *BAHA* bone-anchored hearing aid, *BKB(-SIN)* Bamford-Kowal-Bench (Speech in Noise) test, *CAPII* Categories of Auditory Performance II, *CI* cochlear implant, *CPA* cerebellopontine angle, *CROS* contralateral routing of sound device, *HA* hearing aid, *HL* hearing loss, *HSM* Hochmair, Schulz, and Moser sentence test, *IAC* internal auditory canal, *NH* normal hearing, *PTA* pure tone average, *SIR* speech intelligibility rating, *SPHL* severe-to-profound hearing loss, *SSD* single-sided deafness, *SSNHL* sudden sensorineural hearing loss, *SSQ* speech, spatial, and qualities hearing scale, *TIMIT* texas instruments/massachusetts institute of technology sentence test, *VAS* visual analog scale, *VS* vestibular schwannoma

Table 3 Outcomes of adults with SSD receiving unilateral CI

Authors and year	Study type	Population	Etiology	PTA in nonimplanted ear in dB nHL (range)[a]	Device brand	Speech perception results	Localization/lateralization results	Subjective improvements	Comments
Tavora-Vieira et al. 2013 [9]	Case series	9 adults with SSD and tinnitus (7/9)	Mixed	16 (3–28)	MED-EL	9/9 significant improvement in noise, esp. with noise and speech from front	6 with “near 100 % correct choice of stimulus side”	All 3 subscales of SSQ improved significantly	7/7 with tinnitus improvement after surgery
Tavora-Vieira et al. 2013 [32•]	Case series	5	Mixed	15 (13–27)	n/a	Significant speech in noise benefits in 3 free-field configurations: speech/noise in front, speech in front and noise at the normal hearing ear, and speech at the implanted ear and noise at the hearing ear	n/a	Subjective evaluation at 3,6, and 9 months post-op revealed scores similar to long-term bilateral implants and even normal hearing individuals	Patients with long-term post-lingual unilateral deafness can still achieve robust outcomes with a CI as long as the contralateral ear functions
Hansen et al. 2013 [31]	Case series	29 adults with post-lingual SSD	14/29 ISSNHL 10/29 MD	26 (6–33)	13 ABC 16 Cochlear	Avg. 28 % increase in CNC word scores and 40 % increase in AzBio sentences for electric stimulation of CI-only vs. same ear preoperatively with audio	Most subjects improved postoperatively Performance improved from 3 to 12 months post-op	n/a	Only 5/29 subjects had full 12 month data collection
Di Lella et al. 2013 [45]	Case series	10 adults with VS or other IAC/CPA tumor with deafness on contralateral side	VS or IAC/CPA tumor resection	120 (n/a)	5 Neurelec 4 Cochlear 1 MED-EL	Speech benefits after contralateral implantation are minimal when implantation occurs before the ipsilateral, tumor-bearing ear significantly deafens	n/a	n/a	SSD subjects who became bilaterally deaf after tumor resection
Firszt et al. 2012 [8]	Case series	3 adult males with SSD and tinnitus	SSNHL	22 (15–27)	Cochlear	In quiet, bimodal hearing better in 1/3 patients In noise, bimodal hearing better in 2/3 patients No binaural benefit on BKB-SIN	Significant improvement for 3/3 with bilateral compared to NH ear alone	1/3 with improvement in 3 SSQ subscales	Tinnitus improved in all 3 after implantation

Table 3 continued

Authors and year	Study type	Population	Etiology	PTA in nonimplanted ear in dB nHL (range)[a]	Device brand	Speech perception results	Localization/ lateralization results	Subjective improvements	Comments
Firszt et al. 2012 [35]	Case series	10 adults, 3 with pre-/ peri-lingual HL	Mixed	56 (27–88)	8 Cochlear 1 MED-EL 1 ABC	1. 7/7 post-lingual HL subjects with significant improvement with CI + HA versus unaided or aided better ear alone for TIMIT sentences in quiet 2. No differences between CI + HA versus unaided or aided better ear alone for any test for 3 patients with pre-/peri-lingual HL	Significant improvement in bimodal compared to HA-alone condition for post-lingual patients only	1. Significant benefit in all 3 SSQ domains for all post-lingual patients 2. Benefit in spatial domain for all pre-lingual patients	5/7 post-lingually deafened patients with HA experience in better ear; 0/3 pre- or peri-lingually deafened patients with HA experience in better ear
Stelzig et al. 2011 [33]	Case series	4 adult males with SSD	Mixed	n/a (5–30)	MED-EL	Small increase in speech scores in the bilateral condition compared to CI-only in all 4 patients	n/a	VAS scale showed overall CI acceptance	Limited sample size negates statistical analysis
Roland et al. 2011 [30]	Case series	3 adults with SSD	SSNHL	20 (n/a)	Cochlear	No significant differences in CNC or BKB-SIN scores from pre-op to post- for 2/3 patients	n/a	n/a	Limited sample size negates statistical analysis
Arndt et al. 2011 [10]	Case series	11 adults with SSD and tinnitus with prior Baha and CROS trials	6/11 with SSNHL	13 (7 – 30)	Cochlear	Significant increase in HSM sentence scores in CI versus unaided, CROS, and Baha only in condition where speech is presented to the SSD ear and noise presented to the NH ear	Significantly improved with CI compared to unaided, CROS, and Baha alone	SSQ scores: CI significantly better than unaided, CROS, or Baha	Stringent selection criteria
Buechner et al. 2010 [28]	Case series	5 adults with SSD and tinnitus	3/5 with SSNHL	n/a	ABC	3/5 with significant improvement when noise presented to NH side and speech to front	n/a	n/a	2/5 with sustained tinnitus improvement
Vermeire and Van de Heyning 2009 [7]	Case series	20 adults with SSD and tinnitus; 9 HA users	Mixed	37 (10 – 79)	MED-EL	1. Summation effect not significant in CI + HA and CI + NH groups 2. Squelch effect of adding the CI in the HA users but not the NH group	n/a	Significant improvement in SSQ domains in both NH and HA groups	Did not measure tinnitus reduction

Table 3 continued

Authors and year	Study type	Population	Etiology	PTA in nonimplanted ear in dB nHL (range)[a]	Device brand	Speech perception results	Localization/ lateralization results	Subjective improvements	Comments
Ching et al. 2004 [34]	Case series	21 adults with bilateral SPHL but only unilateral CI; 12/21 HA users	Mixed	107 (73 – 117)	Cochlear	1. CI + HA scores were significantly better than CI alone for BKB sentences 2. CI scores were significantly better than HA-alone scores 3. CI + HA significantly better than CI alone when noise at CI and speech at HA side	CI + HA was significantly better than CI or HA. No significant differences between experienced and inexperienced CI + HA users	Functional Performance in Real Life: CIHA significantly better than CI or HA; CI significantly better than HA. No significant differences between inexperienced and experienced users	Was not designed to study benefit of CI for SSD. Minimum duration of CI use 1 year

[a] For manuscripts not reporting contralateral unaided PTA, these values were calculated from audiograms appearing in the text, and are 3-pure tone averages (500, 1000, and 2000 Hz)

ABC Advanced Bionics Corporation, *AzBio* Arizona State University/Advanced Bionics Corporation sentence test, *Baha* bone-anchored hearing aid, *BKB(-SIN)* Bamford-Kowal-Bench (Speech in Noise) test, *CAPII* Categories of Auditory Performance II, *CI* cochlear implant, *CPA* cerebellopontine angle, *CROS* contralateral routing of sound device, *HA* hearing aid, *HL* hearing loss, *HSM* Hochmair, *Schulz, and Moser sentence test*, *IAC* internal auditory canal, *NH* normal hearing, *PTA* pure tone average, *SIR* speech intelligibility rating, *SPHL* severe-to-profound hearing loss, *SSD* single-sided deafness, *SSNHL* sudden sensorineural hearing loss, *SSQ*, speech, spatial, and qualities hearing scale, *TIMIT* Texas Instruments/Massachusetts Institute of Technology sentence test, *VAS* visual analog scale, *VS* vestibular schwannoma

When speech and noise are presented in varying spatial configurations, the true benefit of CI in SSD becomes apparent. Tavora-Vieira et al. report better BKB-SIN speech comprehension with CI-on versus CI-off, especially when speech and noise are presented directly in front of the listener [9]. In the CI-on state, Tavora-Vieira et al. also report improvements in speech comprehension in three free-field conditions: speech and noise in front, speech in front and noise at the NH ear, and speech at the CI ear and noise in the NH ear [32•]. Similarly, other groups reported that implantation afforded binaural benefits when noise was presented to the NH ear and speech was presented either to the SSD ear [10] or to the front [28]. Stelzig et al. assessed German Freiburg monosyllable tests, dichotic listening tests, Hochmair-Schulz-Moser (HSM) sentence tests, and Oldenburg Sentence Tests (OLSA) in noise with 4 subjects and demonstrated overall gains in the binaural hearing condition compared to unilateral CI-on condition and CI-off conditions [33].

While most studies report significant gains with speech comprehension in noisy environments, controversy still exists around the utility of CI in SSD for all patients [32•]. Roland et al. report that only 1 of 3 of their study subjects receiving a CI for SSD showed significant improvements in the free-field BKN-SIN comprehension test when comparing preoperative scores to postoperative CI-activated scores [30]. Overall, however, the argument for the advantage of unilateral CI with respect to speech in noise is thoroughly convincing. A recent and robust meta-analysis of speech comprehension in various noisy environments by Vlastarakos et al. reports CI insertion in the SSD ear leads to better speech perception when speech is presented directly to the SSD ear or from the front, while concurrent noise is presented to the front or at the normal hearing ear (Fig. 1) [11••].

In patients with asymmetric hearing loss, the addition of a contralateral hearing aid is beneficial as long as a post-lingual onset of hearing loss is present. Ching et al. report that the CI + contralateral HA condition affords better BKB sentence scores than either the CI-only or HA-only cases when speech was presented to the CI side and noise to the HA side [34]. Vermeire and Van de Heyning demonstrate that contralateral hearing aids significantly aid speech in noise due to improvements attributed mainly to the squelch effect [7]. However, Firszt et al. demonstrate speech gains in implantation for pre-lingually deaf subjects are less robust than post-lingually implanted subjects [35].

In children the speech benefits after implantation for SSD are profound, although the number of studies is limited (Table 2). Plontke et al. present a case where a child with a lateral skull base fracture and imminent fibrosis was implanted [36]. Improvements in speech discrimination in both quiet and noise, localization, and patient satisfaction

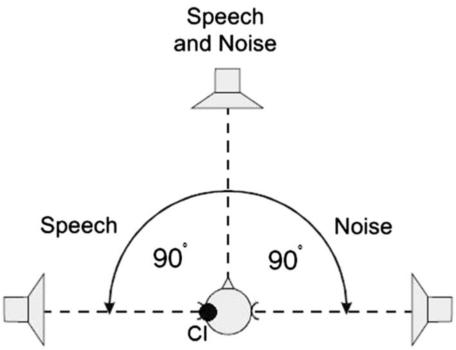

Fig. 1 Benefits of Binaural hearing after CI implantation are most beneficial when resolving speech in noise. Specifically, maximal benefit compared to the pre-implanted condition occurs when speech is presented to the implanted ear and at angles up to the front of the listener, while concurrent noise is presented at angles between the front and the normal hearing ear. *CI* implanted ear, contralateral ear is the normal hearing ear. Summarized from content in [11••]

were reported. To assess CI efficacy in various pediatric age groups, Hassepass et al. performed HSM sentence tests and (OLSA) in noise in 3 children/adolescents aged 4, 10, and 11 [2••]. Differences in speech resolution, localization, and subjective assessments demonstrate significant improvements in the post-implant CI-aided condition relative to the unaided preoperative condition. In children with asymmetric hearing loss, Cadieux et al. report significant CNC and BKB-SIN speech improvements when children listened bimodally versus CI-only or HA-only conditions [37]. Concordant with Cadieux, Tzifa and Hanvey report improvements in CAPII (7/7) and SIR (6/8) scores in children with asymmetric hearing loss utilizing an implant [38].

Sound Localization Benefits after Unilateral Cochlear Implantation

Because binaural hearing is necessary for horizontal sound localization, unilateral CI drastically improves a SSD subject's ability to identify sound sources. Outcomes of localization in adults are compared in Table 3. In patients with adequate localization ability in the HA-only condition, binaural benefit from CI was limited. However, in patients where localization was poor pre-implant, localization increased significantly. At a basic level, patients implanted for SSD are able to recognize on which side (CI ear or NH ear) sound was being presented to [9]. Firszt et al. found significant improvements in localization with CI compared to HA-only localization [35]. Additionally, patients with CIs performed significantly better with their CI when localizing than in unaided, CROS, and Baha conditions [10]. In a case series of 3 patients, Firszt et al. demonstrated improvements in localization in 10° increments when sound was presented to the CI ear, NH ear, and bilaterally [8].

A thorough meta-analysis of implantation in SSD patients found subjective improvement in localization, and improvements in localization when sound sources were presented to the CI ear at angles approaching 90° to the front of the patient, even when noise was presented to the contralateral quartile of the normal hearing ear [11••]. Hansen et al. found that the greatest improvements in localization were during the 3–6 months postoperative period, inferring an experience dependence [31]. However, the study was limited because only 5 of the 29 patients had full localization assessments through the entire 12-month trial as it was designed. In patients with asymmetric hearing loss, Ching et al. showed that adding a contralateral hearing aid improved localization compared to CI-alone or HA-alone groups [34]. It is interesting to note that localization ability is maximized when ears are matched by bilateral acoustic (HA) or bilateral electric (CI) input, compared to unilateral CI and contralateral HA [39].

Localization benefits are slightly better in adults than children [40], though the number of studies quantifying localization ability in children is limited (Table 1). In a study with 3 children aged 4, 10, and 11, localization of the older two children was assessed by presenting OLSA sentences in 7 equidistant speakers placed at 30° increments [2••]. Localization deviation from the source decreased in both of the two subjects who were tested. In noisy conditions, Plontke et al. reported improvements in angle detection error when the noise was presented frontally or to the CI side [36]. Finally, Cadieux et al. demonstrate that for 3/5 children implanted with asymmetric hearing loss, adding a contralateral hearing aid significantly increased hearing ability compared to the CI-alone or HA-alone conditions [37].

Unilateral Cochlear Implant Candidacy

Children with Single-Sided Deafness

Although adult outcome studies by far outnumber pediatric reports, the argument to implant in children appears robust. As such, children with SSD should be implanted—ideally as soon as possible because the best outcomes in SSD children have been associated with early implantation [38]. Many children with SSD demonstrate delays in speech and language comprehension [41, 42] as well as an increased likelihood of academic difficulty [2••, 43]. As unilateral hearing requires an increased effort to comprehend speech in noisy situations, it is not unsurprising that a 2010 study of prospectively followed children with SSD demonstrated they had attention fatigue, behavioral problems, and academic weakness in 25 % compared with bilaterally-hearing peers [44]. Children with post-lingually acquired SSD who were implanted with a CI performed objectively better with speech in noise and localization, and subjectively better in their perceived hearing ability or patient satisfaction [2••, 36]. Early detection of SSD in children seems a key factor in subsequent speech perception outcomes, as intervention at a young age allows for greater neural plasticity to take advantage of input from the implanted ear. Further, in cases where a second CI is indicated in the contralateral ear, as in the case of progressive hearing loss, cat models suggest that only the total duration of bilateral deafness (i.e. time to first implant) is associated with auditory outcomes [4].

In children for whom the unilaterally deafened ear does not respond to amplification, cochlear implants clearly show substantial benefits in speech, academic performance, and quality of life. Similar to CI in bilaterally profoundly hearing impaired children, the decision to undergo implantation hinges on parental investment in, and institutional availability of, postoperative auditory rehabilitation with specialized pediatric personnel so that children may maximize the auditory gain from their devices. These factors should be weighed against the speech and behavioral gains associated with implanting a child with SSD.

Adults with Single-Sided Deafness

While studies of children consistently show psychosocial and behavioral improvements after implantation, the claim that implanting every adult will conclusively bring benefits is not as definitive. For example, the addition of an implant and restoration of binaural input seems less efficacious in prelingually deafened adults [35]. The extent to which unilateral deafness affects an individual's activities of daily living, and the capacity that binaural benefits will significantly affect his or her quality of life, must be weighed on a personalized level.

Interestingly, auditory neural pathways are preserved bilaterally as long as the only hearing ear has substantial ability to stimulate unilateral auditory pathways [32•]. In a unique meta-analysis of patients with SSD and slowly growing vestibular schwannomas in the contralateral (normal hearing) ear, only duration of total bilateral deafness was associated with hearing outcomes [45]. One recent study even showed drastic speech improvements in noise in patients with unilateral deafness as long as 40 years before single-sided implantation [9, 32•]. As such, unlike bilateral deafness, duration of unilateral deafness has not been definitively associated with implant outcomes—so there is less of a time sensitivity to make the decision to implant if a patient is unsure. However, the decision to implant may be expedited by concurrent medical conditions which require surgery in the neighboring anatomical areas, such as implantation during an indicated labyrinthectomy in patients with recalcitrant Ménière's Disease in the treatment of vertigo [31].

Single-Sided Deafness with Ipsilateral Tinnitus

The first accounts of CI implantation serendipitously reducing tinnitus in SNHL patients were reported in 1976 by House et al. [46] and later characterized by Brackmann in 1981 [47], despite tinnitus not being the primary indication for implantation. Over the past decade, in patients with SSD and concurrent ipsilateral debilitating tinnitus, the main indication to implant is more of an attempt to suppress the tinnitus than to strictly replace hearing. Indications for implanting a patient with SSD for the primary goal of suppressing tinnitus were thoroughly analyzed by Punte, Meeus, and Van de Heyning, with main inclusion criteria being severe tinnitus for less than a decade and concurrent ipsilateral SNHL, and main exclusion criteria being patient's inability/unwillingness to attend regular follow-up and CI rehabilitation [48]. With recent knowledge that a CI-induced reduction of tinnitus in the SSD ear increases speech perception in the NH ear, indications for treating SSD are expanding [49]. A thorough meta-analysis of recent literature relating to CI in treating unilateral SSD patients with tinnitus was authored by Arts et al. [50•], who showed significant decreases in tinnitus after implantation assessed by Visual Analog Scale (VAS) at 1-, 3-, 6-, and 24-month post-op compared to the tinnitus VAS levels in the pre-implant condition.

While traditional attempts to preserve the integrity of the cochlear apex by using short CIs may be beneficial in the patient with preserved low-frequency thresholds, using a short electrode cannot be recommended if the patient has ipsilateral tinnitus because full-length cochlear stimulation may be necessary to suppress tinnitus [51]. Although the suspected mechanisms of tinnitus reduction after implantation include habituation and cochlear reorganization [52], research is warranted to develop CI stimulus algorithms specifically aimed at suppressing tinnitus. Studies describe modified CI stimulus paradigms with both high frequency [53] and low-frequency [54] currents optimally suppressing tinnitus in case reports, and variable outcomes of tinnitus suppression with standard stimulus patterns [28].

Improving Postoperative Outcomes in Single-Sided Deafness Patients

Major contributing factors to post-implant outcomes are the attempts to preserve hearing during implantation, therapy during the postoperative auditory neuroplastic window, and the use of additional acoustic amplifiers. Preserving hearing via surgical techniques can reduce trauma to the cochlea and ultimately aid in speech and localization benefits. Postoperatively, the auditory cortex demonstrates the greatest extent of reorganization during the first 6 months post-activation, so it is crucial to keep implanted subjects' motivation high during this critical period of speech therapy [55]. In addition to training, combined electro-acoustic stimulation with an acoustic amplifier may be beneficial in certain populations. In cases where subjects have some degree of preserved low-frequency hearing, an acoustic amplifier in the ipsilateral ear may assist in the detection of low-frequency tones post-implant [56]. When asymmetric bilateral hearing loss exists, combining an amplifier in the contralateral ear is associated with increased hearing abilities with respect to speech in noise in both children [37, 38] and adults [2••].

Conclusions

While hearing aids and similar amplifiers are reasonable strategies for initially attempting to correct hearing loss with minimal risk to the patient, CI is the definitive treatment for replacing a nonfunctioning cochlea and providing binaural hearing benefits. Squelch and summation aid in increasing the signal-to-noise ratio of incoming sounds and interaural comparisons assist in localization after the head shadow effect is eliminated. Because the treatment progression from hearing aids to CI is a large decision for both patients and providers, it is important to evaluate the most recent information regarding who will benefit most. The anticipated benefit of implantation, namely speech in noise, localization, and a decreased effort to hear, should be weighed on an individualized level against the potential risks. For children, the argument to implant is compelling. For adults, the duration of unilateral deafness may not be as strong a contributor to outcomes as duration of bilateral deafness, so the decision to implant may not require quick judgment.

Compliance with Ethics Guidelines

Conflict of Interest Christopher K. Giardina, Eric J. Formeister, and Oliver F. Adunka have declare no conflict of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by the authors.

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