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Cochlear Implantation and Single Sided Deafness

Joshua Tokita¹, Camille Dunn¹, and Marlan R. Hansen^{1,2}

¹Departments of Otolaryngology-Head and Neck Surgery, University of Iowa, Iowa City, Iowa, U.S.A.

²Department of Neurosurgery, University of Iowa, Iowa City, Iowa, U.S.A.

Abstract

Purpose of review—Recently, more patients with single-sided deafness (SSD) have been undergoing cochlear implantation. We review recent studies and case reports to provide an overview of the efficacy of cochlear implants (CIs) to rehabilitate patients with SSD with regards to sound localization, speech discrimination, and tinnitus suppression.

Recent findings—There are a growing number of studies evaluating the effect of cochlear implantation for rehabilitation of the deficits associated with SSD over the past several years as more centers offer this treatment modality to patients with SSD. While individual studies have few subjects and are underpowered, the vast majority report improvement in sound localization, speech understanding in quiet and noise, and tinnitus. In some cases the outcomes with CI appear superior to those achieved with other devices including contralateral routing of sound (CROS) devices and osseointegrated implants (OIs).

Summary—Although CI is not an FDA-approved treatment for SSD, several recent studies show improvements in speech understanding, sound localization, and tinnitus. Due to the low number of cases, it is difficult to conclusively compare outcomes achieved with CIs and those provided by other devices. However, based on encouraging early results and the unique ability to restore binaural sound processing, a growing number of centers offer CIs as treatment for SSD. Forthcoming studies will help define outcome expectations in different populations.

Keywords

Bone anchored hearing aid; osseointegrated implant; contralateral routing of sound; cochlear implant; single-sided deafness; sound localization; speech discrimination; unilateral hearing loss

Introduction

Single sided deafness (SSD) causes a myriad of problems affecting an individual's ability to communicate. The most obvious impairment is difficulty hearing sounds on the affected side due to the head shadow effect. This frequently results in an individual constantly adjusting

Corresponding Author: Marlan R. Hansen, MD, 21161 PFP, University of Iowa Hospitals and Clinics, 200 Hawkins Dr, Iowa City, IA, 52242, (319) 353-7151, marlan-hansen@uiowa.edu.

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their head position in an attempt to compensate for the handicap and some cases rendering them oblivious to the presence of sound directed at the affected side¹. SSD also significantly impairs word discrimination, which varies as a function of residual hearing retained on the affected side. Those with profound SSD frequently report difficulty understanding speech, particularly in noisy environments, even in the presence of normal hearing on the non-affected side². Reducing binaural hearing to monaural hearing reduces one's ability to localize sound, and those with no functional hearing on the affected side find it difficult to determine the direction, distance or movement of sound³. The constant straining, postural adjustments and apparent lack of awareness of sound on the affected side frequently result in socially awkward mannerisms, a sense of confusion and, in some cases, may lead to social isolation.

Treatment options

Rehabilitative options for SSD include contralateral routing of sound (CROS), osseointegrated implants (OI)/bone anchored hearing aids (BAHA) including those attached as dental appliances^{4,5} and cochlear implants (CIs)⁶. Of these, CIs have been a relatively late comer to the field due, at least in part, to concerns about the ability of the brain to sort out acoustic and electric stimuli and concern that the hearing from the CI would interfere with acoustic signaling processing from the good ear. All three modalities help overcome some of the deficits associated with SSD including the head shadow effect and perception of sound on the non-hearing side. They also improve hearing in noisy environments although when speech is at the better hearing ear, the addition of any noise received by the auxiliary microphone on the CROS or OI will degrade intelligibility (Dillon, 2001).. Compared to CROS and OI devices, only CIs offer the potential to restore binaural hearing, allowing the opportunity of sound localization⁷. Although there are some data that compare outcomes in CI recipients with those provided by CROS or CI devices as detailed below, there are no large-scale studies to definitively demonstrate the superiority of CIs over CROS or OIs.

Sound localization:

Binaural hearing is essential for sound localization. Sound localization requires correct calculation of three spatial coordinates: azimuth (the angle left or right of a neutrally positioned head), elevation (angle above or below the horizontal plane) and distance. In humans with two functioning cochleae, the auditory pathway uses interaural timing and intensity differences to calculate these coordinates³, which can be further refined by monaural cues from structures of the pinna. For frequencies below 800Hz, the auditory system relies mainly on phase delays caused by interaural time differences⁸. For frequencies greater than 1600Hz, the auditory system primarily relies on interaural level differences. Both phenomena are used in the transition zone from 800Hz to 1600Hz⁹. In SSD, localization using interaural timing and intensity differences is not possible unless there is some way to replace the impaired acoustic sensor (deafened cochlea).

While the CI is the only rehabilitative option available to provide direct stimulation to the compromised ear, it is not clear that use of a CI would necessarily restore interaural time

detection differences in patients with SSD auditory system since sound processing with a CI occurs on a different time scale than acoustic processing. .

In addition to differences in distance traveled by sound to reach the right and left ear, there will also be a difference in intensity between sound waves reaching the ears. The difference in sound intensity is a function of the distance traveled from the source to the ear (intensity and distance form an inverse square relationship). The head also interferes with the incoming sound wave forming an acoustic shadow or head shadow¹⁰. The head shadow effect is more pronounced at higher frequencies than lower frequencies. Head shadow is a result of sound diffraction, which occurs when an object in the path of a sound wave has a dimension greater than 2/3 of the sound wavelength¹¹. For a human head with a diameter of 140mm, this affects sounds with a frequency higher than ~800Hz to a moderate extent and significantly affects sounds with a frequency higher than ~1600Hz. Encoding of sound loudness by a CI differs from acoustic processing and, as with interaural timing differences, it is not clear whether CIs restore a patients' ability to detect interaural intensity differences.

Monaural cues are limited to the outer ear. The structures of the pinna and the external auditory canal act as directional filters. As sound comes in contact with the pinna it is reflected or transmitted depending on frequency and the topography of the structure it encounters. The result is that different frequencies comprising the incoming sound overlap and cancel to varying extents thereby modifying the spectral composition of the sound as it enters the external auditory canal. If one has a baseline familiarity with an incoming sound, it may be possible to localize sound based on spectral changes occurring at the level of the pinna^{12,13} These mechanisms persist following SSD.

Speech perception in quiet and noise

Binaural hearing is critical for speech processing. In addition to sound localization, redundant information received by two independent acoustic sensors allows for summation and squelch. Binaural summation occurs when the same acoustic stimulus is presented to both ears. The higher order auditory processing of the redundant information provides a 2-6dB in signal threshold and is particularly beneficial in noisy environments. The squelch effect represents another form of higher order auditory processing where noise from ear perceiving a poorer signal to noise ratio is combined with noise from the ear with better signal to noise ratio^{7,11,14-16}. This helps to separate out meaningful sound from background noise and imparts the ability to discriminate speech in noise at a 2-3dB worse signal to noise ratio compared to purely monaural perception³. As with sound localization, the head shadow effect can play a significant role in speech perception in that there may be a 5-6dB decrease in threshold for sounds directly approaching the deaf ear³.

Of the available rehabilitative options for SSD, CROS, OI devices and CIs are able to overcome deficits caused by the head shadow effect. All three devices effectively place an acoustic sensor on the side of the deaf ear. However, signals detected by CROS and OI devices are ultimately routed to the better hearing cochlea and so improvements in hearing would theoretically be limited to recovering the 5-6 dB loss caused solely as a result of the head shadow effect. Since neither CROS nor OI devices use binaural signal processing there

is no expected improvement in speech perception from summation or squelch. CIs, however, allow for both an acoustic sensor as well as electrical input to the deaf side. To the degree that the auditory system can effectively combine this electrical signal with acoustic hearing in the opposite ear, CI recipients will theoretically also benefit from summation and squelch.

Effectiveness of CIs to improve sound localization and speech perception

There are several recent studies examining the effectiveness of CIs and other treatments in rehabilitating sound localization and speech discrimination in SSD^{7,11,16-27}.

Arndt et al. compared sound localization using CROS, OI devices, or CI 6 months after implantation in a cohort of 11 patients. To determine effectiveness seven loudspeakers were placed in a semicircle in front of the patients. Patients were then asked to identify which speaker was delivering the sound. Localization error, measured as the difference in azimuth angle between true and perceived sound source, was used to evaluate patients. Using this calculation, a lower score is interpreted as having better localization ability. Patients who received CIs exhibited a significantly decreased localization error (15 degrees) compared to controls (33.9 degrees, $P = 0.003$) and patients with OI (30.4 degrees, $P = 0.002$), and CROS hearing aid devices (39.9 degrees, $P = 0.001$). Speech perception was also evaluated and compared to recipients of CROS and OI devices. Three conditions were used: 1) sound and noise directed at the front of the patient's head, 2) sound directed at the normal hearing and noise directed at the deaf side and 3) sound directed toward the deaf side and noise directed toward the normal hearing ear. When noise was directed head-on, there was no significant difference in improvement seen in those with CIs versus CROS or OI devices implying all devices offer similar outcomes with this task. Patients who received CIs demonstrated significant improvement in speech discrimination over those with CROS or OI devices in the second and third conditions implying a significant improvement due to utilization of binaural summation and squelch^{7,19}.

Hansen et al. reported speech perception and sound localization outcomes in a cohort of 29 patients who underwent cochlear implantation with or without simultaneous labyrinthectomy²². Post-operative data were available for 19 patients but 12-month post-operative data were available for only 6 patients. Of 19 patients with at least 3-month post op data, 9 demonstrated improvement in sound localization based on a multiple loudspeaker test. Of the 6 with 12-month post op data, 4 patients showed improvement in sound localization. Sound localization appeared to improve with experience, suggesting plasticity and adaptability in the auditory system with regards to processing of acoustic and electric stimuli for sound localization. There was also an improvement in speech discrimination. Of the 19, 13 showed improvement in word score. The overall improvement on CNC word scores was 28% $\pm$ 5.1 (mean $\pm$ SE) with individual improvement ranging from 64% to -26% ($p < 0.05$). Fourteen of 19 showed improvement in sentence scores as measured using AzBio sentences. Overall improvement on AzBio was 40% $\pm$ 6.6 with individuals ranging from 92% improvement to -57% ($p < 0.05$). While the small sample size limits the power of the study there is an overall trend toward improvement in sound localization and speech discrimination at least at the 12-month post operative mark. It should be noted, however,

that many of the subjects had a short duration of implant experience at the time this paper was written²².

In a study examining 10 adults with unilateral hearing loss who underwent cochlear implantation, Firszt et al. reported that 7 demonstrated an improvement in sound localization. Interestingly, these same 7 had post-lingual deafness in contrast to 3 who did not exhibit any improvement in sound localization and who had either pre or peri-lingual deafness. This study suggests that perhaps there are some limitations in improving localization in patients with pre or peri-lingual deafness²¹. A more recent study by Firszt reported 3 of 5 adolescents with unilateral hearing loss had improved speech recognition and sound localization²⁸.

A recent study by Tavora-Vieira et al. examined the benefits of cochlear implantation in nine post linguually deafened patients with unilateral hearing loss and found that all nine patients exhibited an improvement in sound localization ($p = 0.001$) although a limited setup using a left, right and central speaker were used. All nine subjects also exhibited objective and subjective improvement in speech discrimination ($p = 0.008$)²⁹.

Hassepass et al. evaluated 3 children with non congenital unilateral hearing loss and reported that cochlear implantation imparts improved speech discrimination in noise as well as improved localization as determined by a multi-speaker sound field test²³.

Jacob et al. in 2011 follow a cohort of 13 patients for 8 months and compared sound localization and speech discrimination in patients who received CI versus a control group with unaided SSD. Patients in the cohort exhibited improvement in sound localization as well as a subjective sense of greater ease in listening in noisy environments. He further noted that placing a CI did not have any impact on the normal hearing side³⁰.

A recent review by Vlastarakos et al. identified 27 studies evaluating the impact of CIs single sided deafness. After controlling for duplicate subjects (a handful of patients were included in more than one study) 6 studies evaluated 63 patients for the ability to localize sound. All 6 studies reported a trend in improved sound localization, however only 2 of these studies, involving 25 patients, found the improvement to be statistically significant. Seven studies representing 85 patients evaluated speech perception in noise. Of these, 6 demonstrated improved speech discrimination with 4 of them (representing 50 patients) being statistically significant^{31,32}.

Tinnitus

Tinnitus is a frequent sequela of hearing loss and ranges in severity from mild to severe with regard to impact on daily activities. Most individuals with SSD suffer from some degree of tinnitus. Several studies have suggested that CIs reduce the severity of tinnitus³³ and indeed in early studies, potential tinnitus suppression was a motivation to offer CIs to patients with SSD^{16,18,20,22}.

Several studies have sought to evaluate the effect of CIs on tinnitus suppression. Tavora-Vieira had seven patients who reported severe tinnitus prior to having a CI placed. All seven

reported a reduction in tinnitus when the processor was activated^{25,29}. Ramos et al. in 2011 reported that in a cohort of 10 patients, 2 experienced suppression of tinnitus, 7 experienced reduction in tinnitus intensity and 1 experienced no change after placement of a CI³⁴. In a study by Arndt et al, 9 patients in the cohort had tinnitus of which 6 experienced improvement in tinnitus severity and 3 reported no change^{7,19}. Van de Heyning et al. in 2008 studied a cohort of 22 patients with tinnitus and found that it was completely suppressed in 3 patients, improved in 18 and unchanged in one^{16,18}. Hansen et al. in 2013 reported that while 12-month post-operative tinnitus questionnaires were not yet available, most patients in the 29 patient cohort reported improvement in tinnitus after activation of the CI. Biasco and Redleaf, in a recent meta-analysis drawing data from studies by Arndt, Ramos and Van de Heyning found that CIs had a statistically significant improvement on the severity of tinnitus ($p < 0.00001$)²⁶. Although the majority of the studies to date are promising, all of them have few subjects and consequently, lack statistical power.

Other considerations

While there is increasing evidence that patients with SSD benefit from CIs, there are several factors to consider in deciding whether a patient represents a suitable candidate. First, there are few cases of patients with congenital SSD who have received CIs. Whether the results achieved in patients with post-lingual SSD translate to this population, especially with regard to the processes that depend on binaural auditory processing, requires further study. Duration of deafness has been shown to be a significant determinant of CI performance in patients with bilateral deafness³⁵ and this likely holds true for patients with SSD. Thus, patients with long-standing SSD may not be suitable candidates for CI and would likely derive more benefit from other rehabilitative options (e.g. CROS or OI devices).

Patients with severe, refractory Ménière's disease represent another population that warrants consideration for CIs. Our group has offered simultaneous CIs for those patients that are considered candidates for a surgical labyrinthectomy based refractory vertigo or drop attacks and poor hearing²². Surgical ablation of the labyrinth, while addressing vertigo and drop spells, resulted in profound SSD. CIs effectively restored hearing to the deafened ear²². Thus, patients were provided relief from the vertigo attacks and remediation in hearing in the affected ear. In many patients, the CI also diminished the tinnitus that occurred in Ménière's disease, which can be debilitating.

Patients with a CI in one ear and good acoustic hearing in the opposite ear provide a unique opportunity to explore sound encoding and adaption with a CI. For example, assessing pitch mapping performance in the CI ear over time that can be compared to a normal hearing ear should help us understand the mechanisms and extent to which patients can utilize and adapt to different CI mapping strategies. Also, comparison to the normal hearing contralateral ear will allow identification and refinement of CI processing strategies that best mimic acoustic hearing.

Conclusions

Patients with SSD frequently suffer from communication problems that arise from significantly diminished sound localization and speech perception in the presence of background noise. As a result, many patients seek rehabilitative options. Of the currently available treatment options for SSD, CROS and OI devices are the most frequently used. While these options provide a significant benefit by overcoming the head shadow effect to detect sound on the deafened side, they fail with regard to providing the benefits of binaural sound processing. Thus they are unable to restore sound localization or improve speech discrimination through summation and squelch. CIs also offer the potential to diminish tinnitus. Careful attention to patient selection criteria will assist in identifying those patients most likely to be benefited by a CI or a different device. As the number of individuals with CIs for SSD increases, there will be more opportunities to compare the outcomes of CIs with those provided by CROS or OI devices.

Abbreviations

BAHA	bone-anchored hearing aid
CI	cochlear implant
CROS	contralateral routing of sound
OI	osseointegrated implant
SSD	single sided deafness

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Key points

1. SSD compromises sound localization and speech perception in quiet and noise.
2. CIs offer the potential to restore binaural sound processing, sound localization and suppress tinnitus in patients with SSD.
3. Careful selection criteria will help identify patients most likely to benefit from CIs for SSD.
4. Current studies, while limited in the number of subjects, are encouraging with some studies demonstrating superiority of CIs to CROS and OI devices.