DEI Op Ed- Rationale for Moderation in Application of CI to SSD in Children and Adults

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Note: What follows was originally written as a DEI internal discussion among the physicians and audiologists of DEI. Therefore, it is a professional level discussion. It is published here on our website for patient/public information.

It is my opinion that the application of cochlear implant (CI) technology to the problem of single sided deafness (SSD-normal or near normal hearing in one ear with severe to profound hearing loss that is poorly aidable in the opposite ear) is not near as intuitive or "natural" as it may seem on the surface nor that all the hype and recent FDA approval would imply.

In 2006 I wrote this position paper on the Rationale for Bilateral Cochlear Implantation (BCI) in Children and Adults. Up to that time, unilateral CI was the norm. In this monograph, I argued for why both children and adults who meet cochlear implant criteria should be considered for bilateral application. In it, I used the deficits caused by SSD as one of the arguments for BCI. I also concluded that the provision of binaural hearing for all patients whenever possible should be the norm, whether that be in the bimodal condition (CI with contralateral hearing aid) or BCI. Point being, I am a strong advocate for binaural hearing.

Why then do I now feel the need for moderation about CI in SSD? Why the sudden conversion from Dr. Maverick to Dr. Conservative on this topic? Why is providing binaural hearing to patients with bilateral severe to profound hearing loss intuitive and a natural evolution, but not as much so in SSD? I will try to explain the details for my opinion in what follows. The summary is this: in my opinion the application of CI to SSD demands a level of performance from and confidence in CI technology that is on the border of overconfidence. It is knocking hard on the practical ceiling of what benefits CIs are currently able to muster. In stock trading terms, I consider CI in SSD to have a very narrow "upside vs. downside" potential.

The Problem of Experience

Our CI program has now done approximately 1900 CI surgeries (of which 700 are BCI) over a 30 year period and we have participated in over 20 IRB-monitored clinical trials. Over that time period, we have pushed the envelope here and stretched the indications there in order to move our understanding forward of when, in whom, and how CIs provide benefit, and more importantly when they do not. We have changed the lives of many by being a "maverick" in the CI domain and it has taught us a great deal. We have generated data but also acquired a wealth of anecdotal experience.

When caring for patients over many years, the negatives tend to resonate in memory more than the positives. Certainly, the happy patients and positive stories are the ones that help us get up and come to work every morning. It is easy to overlook the fact that many of the positives we were blessed to participate in would not have happened if not for pushing the boundaries somewhat, which then also caused some of the negatives. But it is the poor performing, unhappy patients that are the ones who cause inordinate reflection in us as we try to figure out how to keep the disappointing experiences to a minimum. I believe this reticence is as it should be. Critical reflection of one's work, rooted in a soft

skepticism that understands the human limitations at discovering truth, is a very important character trait in any field of science. This is true especially when the data supports moving forward with a treatment strategy, the benefits of which my own experience causes me to suspect a large percentage of patients will not obtain.

> There is a large stock on hand; but somehow or other, nobody's experience ever suits us but our own. Letitia Elizabeth Landon

> "**Good judgment comes** from **experience**, and a lot of that **comes** from bad **judgment**." - Will Rogers

Evidence Based Medicine (EBM) when applied to the CI domain has added its share of confusion to this clinical experience dilemma. Don't get me wrong. EBM has its great strengths and will be with us in some form for the foreseeable future. It has and continues to propel our understanding forward on many fronts. The data generated by formal studies is invaluable. But in a field of clinical medicine with small subject numbers and relatively short time frames of monitoring like CIs, data apart from clinical experience can become a practical and even ethical "hall pass" to a generation of younger professionals.

Research to Date on CI in SSD

There is now a significant body of published clinical trials on the efficacy of CI applied to patients with SSD, in both children and adults. These studies are designed exactly the same way most all clinical trials have been carried out in the CI/ BCI era- audiometric measures (discrimination in quiet and noise with binaural and localization measures) in the pre and postop condition, usually out to 12 months of CI use. There are usually also quality of life (QoL) questionnaires out to 12 months. In most studies to date, these measures are positive, modestly so, for showing binaural benefit of CI in SSD with improved patient satisfaction at 12 months.

With the data published to date, offering a CI to **certain** patients with SSD is reasonable. But it is very important to keep in mind what most of these studies are not telling us, either because of limitations inherent in their design or due to their short-term period of observation.

We learned this related to our study on sequential BCI in children. All of these prelingually deafened children received their first CIs prior to 3 years of age and were good first CI users. The data presented was a breakdown of pre and post second CI performance by age of second ear implantation. (Sequential BCI has many, applicable analogies to CI in SSD). Although earlier age of second ear CI resulted in better second ear and binaural performance, all age groups showed improved scores in the BCI condition. By the end of this one-year study, all children were "benefiting" from the second CI based on audiometric measures and parent questionnaires.

But subsequent data told a different story over the long term for some of these children. Even though this older group showed improved second CI and binaural performance after 1 year of use, 4 out of 7 of them (57%) had become minimal or non-users of their second device 3 years after surgery. It has taken many years since the broad implementation of BCI to tease this out, but this high non-user rate has been reported here and here also.

I have been able to find only one publication that reports on the long term wearing patterns of patients who received a CI for SSD, and that only in children. As you can see from this report, even though all children showed significant binaural gains and parent satisfaction level was high, of the 5 children who had reached the 3-year point, 3 of them (60%) were minimal or non-users. I have been unable to find any reports documenting the percent of adult SSD subjects who are consistent users of their CI over 3 years after their completion of a study.

My point is that although significant improvement on audiometric measures and positive questionnaire data are helpful to know, and can garner FDA approval, they do not tell the long-term story on a patient's subjective, practical benefit. The "observation effect" (Hawthorne effect) that is well known to be present in a clinical trial of this nature, is good in some ways and yet can also be deceptive. It is good because during the typical 1-year period of a clinical trial when all subjects are being regularly tested, they are held accountable for wearing and using their CI device. Patients enrolled in a research study want to please and show themselves a worthy participant, especially when the device and surgery may be provided for free. Compliance and performance are optimized. It is a highly accountable period of experience that helps them through the difficult accommodation period and maximizes their performance through consistent device use.

But the observation effect can also be deceptive because although it may help the study show good, measured benefit from the intervention through "forced use" so to speak, it may not sustain itself practically over time as the patients are eventually after the study left to their own in deciding whether or not it is worth it to them to continue using the device based on their subjective day to day perception of benefit. I am just pointing out the frequent lack of correlation between measured benefit and practical usefulness.

So, if a CI indication has a 50% long-term non-user rate, should we not do it? What about the 50% who do continue to use and benefit from it? Won't they be left out if focus is on the non-users?

If in the early years of the CI era there had been a 50% or greater non-user rate, this would have been considered abysmal and would have threatened the broad acceptance of CI as a valuable technology for the treatment of hearing loss. But times are different now. We are in an age in which the big battle of CI acceptance has been won and now the CI era is slowly expanding its borders. But there are major holes that remain in the candidacy selection criteria of any application that may have this kind of frustrating, long-term non-user rate.

SSD in Children

Med El's FDA approval for use of CI in SSD covers down to 5 years of age. Keep that in mind. It is currently not FDA approved for infants. Doing a CI in the deaf ear of a 5,7, or 13 year old SSD child who may have been deaf in that ear since birth is a completely different scenario from the same discussion in a 1 year old. Our Sequential BCI data has clearly shown that.

Another limitation that nearly all binaural research done to date has (and that most effects pedi CI studies) is the paucity of good, longitudinal data. The challenges to speech/language/psychosocial development and educational outcomes caused by SSD are well documented by several observational studies which are referenced in our papers linked above. Despite all that is known, there are many essential questions that remain unanswered:

1. Although studies document that 35-50% of children with SSD will at some time in childhood experience developmental, psychosocial, behavioral, or educational problems, (which is 10 times higher than in children with normal hearing), it still means 50-65% of SSD children do not ever show these problems. Do we apply a surgical treatment strategy to all infants or children with SSD even though only half of them or less will ever show deficits worthy of aggressive intervention? How do we tease out those who will be the most affected and do so at an early enough age to make CI most effective for them?
2. To complicate the discussion further, some studies (good review discussion here) indicate that the early struggles and delays of children with SSD tend to correct themselves over time. Colletti et al compared a group of adults with congenital SSD to adults with normal hearing in both ears. They found no between-group differences in the areas of scholastic achievement, types of employment, social problems, or substance abuse. How does information like this affect our decision about treatment options for SSD in children?
3. Also concerning for us to contemplate is whether the child's personal experience of having an aggressive surgical intervention, their perception of the implanted artificial device in their head, and the social aspects of wearing a visible hearing device, may cause psychosocial issues of their own, particularly in the group of normally developing SSD children that would not have otherwise happened to them if not for this aggressive intervention mentality. (Premum non nocere!)
4. To date, there are still no controlled, longitudinal studies that show whether any binaural intervention (BAHA, BCI, or CI in SSD) improves the life outcomes of those affected children compared to the unilateral condition. None of us know whether children with BCI have better developmental outcomes than do those with one CI. To properly study CI in congenital SSD, a control group of children with SSD who do not receive a CI is essential. Why? Because of the extensive support services typically received by children who undergo a CI. The control group who does not undergo CI would need to receive the same attention and support services in order to distinguish the effect of the CI itself. Maybe the only thing children with SSD who are experiencing negative impact need in order to do well is the same kind of early recognition, attention, and therapy received by typical CI recipients. Both groups would have to be followed well past elementary school. It is unlikely we will have such data anytime soon. Parents need to know this when being counseled about options.

The Problem of Ear Dominance

There are reports that claim having normal hearing on one side does not detract from the acceptance of a CI signal on the opposite ear in SSD patients. I am skeptical of this weakly-founded conclusion. I can accept that when used in SSD, some patients may be willing to use and attempt to benefit from a CI on the deaf side, but the claim that normal, highly-superior hearing in the opposite ear does not make acceptance of the CI very challenging does not fit with our experience nor that of other prior reports. There does seem to be a significant relationship between one ear auditory dominance and the patient's subjective perception (not measured performance) of binaural benefit and their desire to use the poorer performing CI ear.

In our CI program we have seen the auditory dominance phenomenon in our postlingually deafened adult patients with asymmetric SNHL who receive a CI opposite an ear with better than typical residual hearing. If we choose to implant a patient's bad ear before the hearing in their better ear has deteriorated to a certain severe level, we always have concerns they will not want to use the CI. Our experience in asymmetric hearing loss has led us to adopt the approach that if we cannot reasonably

expect the prospective CI ear to become their better hearing ear, there is a chance the patient will not want to use the CI and would rather just stay with their hearing aid for the time being.

Even though the hearing a CI provides is a godsend to patients with bilateral profound hearing loss, it is a greatly inferior quality signal to normal hearing, even with moderate or moderately severe hearing loss using a hearing aid on the opposite side. Just because real binaural benefit can be measured from a CI device in many situations does not mean that over time the patient will feel the combination of their natural hearing on one side and the CI on the other is something they want to put up with. Even with moderate hearing loss serviced with a hearing aid on the better side, the CI signal has difficulty competing quality-wise for the patients listening attention and may even subjectively seem to interfere, despite measured benefits.

So consistent was this problem in sequentially implanted BCI children (1st implanted better hearing ear dominance interfering with the assimilation and performance of the poorer performing 2nd CI ear), that 2 new approaches arose in the time period after our study:

1. First ear deprivation- we would tell the parents of these children that leaving off the 1st CI may be necessary in order to "drive" the attention to and performance of the second CI ear. This approach can be very difficult for the patient but is effective. It has also been used at times in sequentially implanted post-lingual adults whose second CI performance is struggling.
2. Early simultaneous BCI- the experience of auditory dominance with sequential BCI in children was a major impetus behind further binaural neurodevelopmental research and early simultaneous BCI surgery prior to 12 months of age.

Enter now the SSD indication where the opposite ear is actually a **normal** hearing ear. There is no way their CI will become their dominant hearing ear. It is like Lloyd and Harry riding a minibike up the mountain pass to Aspen, trying to keep pace with a Porsche. And, the normal ear cannot be "turned off" for deprivation therapy. With commitment, effort, and time, the patient can eventually get some benefit, but I would expect for the majority it will be fraught with frustration.

This hits at the main concern I mentioned in the introduction, that of overconfidence in what CI technology can do. I am asked frequently by prospective patients, "what is the success rate of cochlear implants?" I tell them it is instead more useful to speak of the range of CI outcomes as a school grade. There are A, B, C, D, and F users. The "F" users are those whose hearing and perceived performance is worse than they were before surgery. Thankfully, with our current candidate selection criteria (bilateral severe to profound hearing loss with poor discrim scores), the number of "F" users in our program is less than 1%. But even among the rest of patients who are functioning somewhat better than before surgery, there is a wide range of outcomes we cannot fully predict. We have plenty of "C" users, mostly "B"s, and a good share of star "A" users that are so impressive. They are all happy because they are doing significantly better than preop.

My fear is that if an SSD patient (with normal hearing in their opposite ear) does not achieve an A or B performance level with their CI, they will be unhappy and not want to use it. And they are less likely to achieve this level of performance because their normal ear dominance will be sucking assimilation attention away from it. They can still show improved binaural performance on audiometric measures and rate satisfaction as positive in the first year or two after surgery (sunken effort phenomenon) but once they have sailed off into the sunset, the processor may spend most its time in their drawer.

This does not mean we shouldn't ever do it or that no patients will find it beneficial, but we sure better counsel patients accordingly and help set their expectations in this regard, lest dissatisfaction abound. Even with that kind of counseling, my prediction, which may be wrong, is we may see that which I have heard reported at CI meetings about other series of CI in SSD, a long term non or minimal-user rate around 50%.

Risk Benefit Analysis

Cochlear implants in patients with bilateral profound SNHL have great potential for being a paradigm shifter. This intervention takes an individual confined to one realm and enables them to function in another. The potential benefit here is enormous. Patients' lives can be forever changed. Touching news stories are done on them so the world can see the tears of one who goes from isolation and silence to the world of sound. Sometimes their auditory performance does not even have to be all that impressive (B or C user) and they are still very thankful because it is much better than what they had before. That is how bad they were. It has changed their stars.

Yet even in this widely agreed upon indication there are disappointments which vex us. A small number of these patients, despite our best intentions, do not do well with their CIs. An even smaller number may feel they have other symptoms that are worse than their state prior. This is the professional challenge of every CI audiologist and any CI surgeon who cares enough to actually get ongoing outcomes feedback. But at least in their case the upside potential was huge. Their unfortunate, poor outcome is a known downside statistic we have yet to solve, cannot fully foresee, and must accept if we are going to help the 98% who do well.

Of all the options for SSD that have been promoted, marketed, and implemented over the decades, CI is the first option that has had much of any risk at all. CROS hearing aids have zero risk. BAHA's (see Footnote 1 at the end of this paper) are fairly innocuous also. A CI is the first option for SSD that has a small but real risk of causing some vexing, regretful side effects (worsened tinnitus, vestibular symptoms, chronic pain) in the patient, and this for a deficit that in no way compares in life impact to that caused by profound bilateral SNHL. Even if the ear is already deaf, I would sure want the patient to know of these potential risks relative to the incremental benefit they may receive.

The binaural deficits caused by SSD are well known and have been categorized thoroughly. I do not want to diminish them. Patients have real, negative impact on their lives from this. But I have also had the privilege of watching hundreds of children born with SSD grow up, many of whom go on to graduate first in their class from High School and go to the best colleges in the country. I have guided innumerable adults through the first, somewhat devastating year of sudden SNHL, and monitored them for decades, watching them learn to adjust and move on with life.

The challenge for us as clinicians (as the patient's investment advisor) is helping them balance the upside and downside risk that exists, and which has much smaller margins in SSD than in any other prior CI indication. I don't think there will be any tearful patient news stories done on this, no patient visits with doctors or audiologists where the parent or patient thanks you for having categorically changed their life. Rather, the best we can hope for is some "pretty goods," "a little better than before," and "I wear it most of the time." The lesser downside is if the poorer quality signal of the CI seems to interfere at times with their normal hearing in the opposite ear and so they become a non-user of a $30k device. The nightmarish downside, which is a potential in a fraction of patients, is if the surgery causes

frustrating postop symptoms they did not have before and makes them wonder why the they ever did this and why we would have ever recommended it just to get some modest binaural benefits. “My life was fine before all of this!!!” Those patients always forget having said preop, “well the ear is already deaf so it can’t get any worse.” There is always such a thing as worse.

If our patient selection and counseling are not done well and from a solid body of both EBM and clinical experience, we will have that which occurs any time professionals overreach the horizons of an expensive technology like CI- more unhappy patients than we wish and many others with a very expensive unused device in their head. Our professionalism and advocacy in service to our patients must be experienced and mature enough to carefully parse the pertinent issues on their behalf.

Footnote 1: All the attention now being garnered upon CI in SSD also happened exactly this way regarding BAHA for SSD in recent years. This is déjà vu all over again. Go back and look at the numerous studies showing real, audiometric and some binaural benefits, as well as positive patient satisfaction questionnaires, for use of BAHA in SSD. Our adult non-user rate with this indication is over 50% and many have asked for their abutments to be removed. Then, BAHAs for pedi patients with SSD started to enter the conversation. Imagine how I felt about that after our adult experience! It has certainly not been a game changer for them either.

Granted, we do have a smattering of BAHA SSD patients who use their devices long term and are probably happy they did it. I still tell patients about BAHA as an option for SSD, with a dearth of rosiness. If interested, I put them through our SSD counseling process. If after such full disclosure they are still interested in a BAHA as their first choice for SSD, I will do it. Which is my whole point of this discussion- full disclosure and careful patient selection. Professionals must resist getting caught up in any hype nor kid themselves about what we may likely think of all this in a few years.

Footnote 2: The most intuitive application of CI in SSD patients in my opinion is in those who also have chronic, intractable tinnitus in the SSD ear. This seems the most reasonable application and is probably the only treatment approach with hopes of providing benefit for the tinnitus and shifts the risk/benefit ratio in the patient’s favor. On the opposite end of the benefit continuum is the practice I have heard recently of the routine placement of a CI at the time of acoustic neuroma removal. Placing a CI in an SSD ear with good prognostic indicators is challenging enough for the performance outcome of that ear. Doing so in an ear with a poor prognostic factor such as an acoustic neuroma, and then expecting the resultant CI hearing to be well received opposite a normal hearing ear, reveals either a lack of CI experience or is unwisely glib. The only acoustic neuroma patients in whom I have placed a CI at the time of tumor removal are those with bilateral severe to profound hearing loss, usually whose tumors were discovered on routine MRI as part of their CI candidacy evaluation. In other words, they were headed toward getting a CI anyway. With good cochlear nerve preservation, reasonable auditory thresholds can be achieved with the CI after tumor removal, but the average discrimination scores are significantly worse than in ears without an AN. If the hearing is near normal in the opposite ear, I would predict the non-user rate to be extremely high, and afterwards you have a magnet in the head that even when it is “MRI compatible,” creates a large image void that makes imaging surveillance challenging at best.