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Worldwide Trends in Bilateral Cochlear Implantation: Supplement to *The Laryngoscope*, Volume 120 Issue 5

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TRIOLOGICAL SOCIETY
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Worldwide Trends in Bilateral Cochlear Implantation

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Objectives/Hypothesis: The goal of this study is to ascertain worldwide experience with bilateral cochlear implantation (BCI) with regard to patient demographics, trends in provision of BCI to adult and child patient populations, differences and similarities in BCI candidacy criteria, diagnostic requirements, and treatment approaches among clinicians in high-volume cochlear implant centers.

Study Design: Retrospective/prospective.

Methods: An electronic survey consisting of 59 mainly multiple-choice questions was developed for online completion. It examined the implant experience and clinical opinion of expert cochlear implant (CI) centers worldwide on the indications, motivations, and contraindications for adult and pediatric, simultaneous and sequential BCI candidacy. Centers were chosen to complete the survey based on their known reputation as a center of excellence. Patient demographics were queried for two time periods to elucidate trends: 2006 and prior, and for the year 2007.

Results: Seventy-one percent (25/35) of the CI clinics approached completed the survey. Collectively, these 25 clinics represent experience with approximately 23,200 CI users globally, representing 15% of the total estimated CI population worldwide. The total number of BCI surgeries reflected in their experience (2,880) represents 36% of the estimated number worldwide as of December 2007. Cumulatively to the end of 2007, 70% of all BCI surgeries have occurred in children, with the 3- to 10-year-old age group having the highest representation (33% of all BCIs), followed in order by adults (30%), children under 3 years of age (26%), and children between 11 and 18 years of age (11%). Seventy-two percent of all BCI surgeries were performed sequentially (70% of children, 76% of adults). Children <3 years of age represent the only age group of all patients in which simultaneous surgeries predominate (58% simultaneous). For all other age groups, sequential surgeries far outnumber simultaneous (3–10 years, 84% sequential; 11–18 years, 94% sequential; adults, 76% sequential). Prior to January 2007, 68% of BCIs were performed in children. This increased to 79% for the year 2007 ($P < .001$). With regard to children only, a change is apparent over time in terms of the age group making up the majority of pediatric BCI surgeries performed. Prior to 2007, children 3 to 10 years of age made up 50% of the children undergoing BCI, whereas those <3 years made up only 33%. In 2007 this shifted more toward the younger age group (47% for those <3 years and 40% for 3–10-year-olds; $P < .001$). United States clinics had a higher proportion of adult BCI patients (59% children, 41% adults) than the non-United States clinics (78% children, 22% adults; $P < .001$). The majority of responders do not hold to a minimum or maximum age by which they limit BCI.

Conclusions: Worldwide experience with BCI is now quite extensive and provides a useful base for evaluating clinical outcomes across patient categories and for providing further support during the patient/parent counseling process.

Key Words: Cochlear, implant, bilateral.

Level of Evidence: 2b.

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INTRODUCTION

The advent of cochlear implants (CI) over 25 years ago was the beginning of a revolution in the treatment of bilateral severe to profound sensorineural hearing loss. In the early years with first generation implants, only individuals with profound bilateral deafness were considered candidates and for only monaural implantation. Subsequent years saw improvements in hearing performance occur primarily through increasing

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sophistication of implant devices and processing strategies. Such improvements allowed for the expansion of candidacy criteria to include individuals with measurable amounts of residual hearing and speech discrimination, yet who still suffered from significant functional impairment even with modern hearing aids. Today, the hearing-impaired population considered to be candidates for cochlear implantation includes individuals having widely diverse characteristics with regard to age, etiology, hearing history, quantity of residual hearing, and comorbid conditions. Extensive clinical experience over many years with large numbers of CI recipients (approximately 172,000 worldwide as of December 2008) has been required to better define candidate categories that are useful in helping to predict outcome (data from Advanced Bionics Corp., Cochlear Corp., and MED-EL Corp. databases). The defining process continues to this present day, as ongoing developments make such individual patient diversity ever more apparent. Although decision algorithms have been outlined for unilateral CI in both pediatric and adult candidates, experienced clinicians know that these categories are in no way rigid and should remain flexible to avoid excluding potential candidates who may prove to be exceptions to the trends shown from our current experience and understanding.

In the past several years another distinct phase of the CI era has developed. Significant improvements in hearing for CI users have recently been achieved, not via new implant technology alone, but through the provision and/or restoration of binaural mechanisms through bilateral stimulation. It has become apparent that a significant portion of the hearing limitations experienced by CI users in the past are related to the fact that implants were only provided unilaterally. Binaural mechanisms, such as summation, squelch, and sound localization are not possible for individuals hearing monaurally.[1,2] In addition, maximal benefit from the head shadow effect requires binaural hearing. Bilateral CI (BCI) users have demonstrated significantly improved speech understanding in quiet and in noisy environments, improved sound localization abilities, subjective reports of significantly decreased social restriction, reduced perception of hearing disability, and a trend toward reduced emotional distress compared to the unilateral implant condition.[3–14] The benefits reported in the literature to date have resulted in professional society position statements recommending BCI as accepted medical practice.[15–18] Such acceptance of a treatment approach that permanently commits both ears to CI technology is testimony to the degree to which confidence in CI treatment has grown over the past two decades.

BCI has added another level of complexity to our attempts at defining CI candidacy criteria and categories. It requires judgments to be made about two ears with varying amounts of residual hearing, varying degrees of benefit from the use of hearing aids, and possibly quite different hearing histories in the same patient. The topic of bimodal hearing (use of a hearing aid opposite a unilateral CI) has assumed greater prominence in light of BCI. Sequential BCI in children has, in particular, brought to light the pronounced effect that the different age at which each ear is implanted can have on disparate hearing outcomes between ears.[19] As has been seen with unilateral CI, a high degree of variability in outcome across patient categories can be demonstrated with some frequency.

TABLE I.
CI and BCI Population Statistics as of January 2008 From the Databases of Advanced Bionics Corp., Cochlear Corp., and MED-EL Corp.

January 2008–3 Manufacturers*	Total Worldwide (%)	United States (%)	Non-United States(%)
Total CI	153,000	59,670	93,330
Adults	81,090 (54)	36,398 (61)	48,516 (52)
Children	71,910 (46)	23,272 (39)	44,814 (48)
Total BCI	8,042	4,182	3,860
Adults	3,056 (38)	1,882 (45)	1,174 (30)
Children	4,986 (62)	2,300 (55)	2,686 (70)

Percentages are for proportion of adults vs. children for each region.

Figures for MED-EL Corp were obtained from the manufacturer up to October 2005. The company subsequently declined to provide updated figures to January 2008. Therefore an extrapolation was made to estimate final numbers by keeping the percentage of MED-EL in the total CIs and BCIs the same for the two time periods.

CI = cochlear implants; BCI = bilateral cochlear implants.

Worldwide experience with BCI includes approximately 8,000 adult and pediatric patients as of December 2008, which is approximately 5% of the CI population (Table I). A large percentage of these BCI surgeries have been performed at higher volume, more experienced CI centers around the world, many under research protocols in just the past 6 years. The goal of this current study is to ascertain overall BCI patient demographics, trends in provision of BCI to adult and child patient populations, differences and similarities in BCI candidacy criteria, diagnostic requirements, and treatment approaches among clinicians in high-volume cochlear implant centers worldwide with regard to simultaneous and sequential surgery in children and adults. T1

MATERIALS AND METHODS

An electronic survey was developed for online completion to specifically examine the implant experience and expert opinion on the indications, motivations, and contraindications for bilateral CI candidacy for four distinct groups of potential bilateral candidates: unilaterally implanted children for sequential implant, unilaterally implanted adults for sequential implant, newly diagnosed children for simultaneous implant, and newly diagnosed adults for simultaneous implant.

The survey comprised 59, mainly multiple-choice, questions divided into five sections. The first section covered general queries about overall CI experience, while investigating demographics and experience with bilateral CI users in detail. The following four sections asked questions related to clinical practices and opinion on bilateral CI candidacy issues for each of the potential candidate groups addressed above.

The survey was sent electronically to 35 CI clinics located in Europe, Australia, the United States, and Canada that were identified as expert CI clinics having experience with >250 cochlear implant users. They were invited to provide their responses to the survey voluntarily. Centers were chosen worldwide based on their known reputation as a center of excellence. There was no inquiry made or prerequisite with regard to device manufacturer preference or utilization.

In view of the variety of funding situations across and within the various countries represented by the responders, past experience and numbers of bilateral implants will inherently reflect the respective economic environments. Nonetheless for the purpose of providing clinical opinion on bilateral CI candidacy issues and the related clinical aspects, responders were asked to provide their clinical opinion while putting funding issues in the background.

Statistical Analysis

Descriptive analysis including the frequency of responses per question and per option was performed for multiple-choice questions permitting selection of one or more items and presented as a percentage of the total number of responders to the survey. For data arising from questions requesting open-ended numerical responses, statistical analysis included calculation of mean, standard deviation, and median data values for group responses. Report of the analysis of significance of differences in clinical practices examining the influence of variables, such as age at bilateral implant, sequence of bilateral implant, time interval examined, and geographical region, was performed via calculation of a 2-tailed P value via the Fisher exact test and is reported.

RESULTS

Seventy-one percent (25/35) of the CI clinics approached completed the survey. Twenty (80%) of the responders were ear, nose, and throat surgeons, and five (20%) were audiologists. Clinics were located in Europe (13), Australia (one), Canada (one), and United States (10). The European clinics include Germany (three), Switzerland (two), Spain (two), and one each in Austria, Belgium, France, Italy, Norway, and the United Kingdom.

Clinic Experience and Patient Demographics

Collectively, these 25 clinics represent experience with approximately 23,200 CI users globally. To the end of December 2007, the total number of CI surgeries performed at the responding clinics represents 15% of the total estimated CI population worldwide. All clinics had a long history of implant experience, and a variety of implant devices have been used among them. The total number of CI users to the end of December 2007 at each clinic ranged from 265 to 4,000 with a mean of 930 and a median of 700.

The number of BCI surgeries reflected in their experience (2,880) represents 36% of the estimated total number worldwide as of December 2007. BCI users per clinic ranged between one and 300, with an average of 111 and a median of 83, representing on average 13% of the total implant population at each clinic (median, 14%; range, 0.1%–21%). All but two clinics (one in France and one in the United Kingdom) had experience with 40 or more bilateral CI users to the end of December 2007. The United Kingdom clinic had 15 BCI patients, and the French clinic had one. Despite their limited BCI experience, the response data for the implant clinic in France is used in the combined analysis to reflect their clinical opinion as representative of their region. This clinic has extensive cochlear implant experience in over 550 cases but was limited at the time of the survey in offering BCI by national regulations.

Approximately half (52%, 13/25) of the clinics reported their first BCI procedure was performed before the year 2000, indicating several years of experience for their respective implant clinic. The remaining clinics reported commencing between 2 and 7 years ago. Cumulatively, just over 25% of the total BCIs reported by the responder clinics were performed in the year 2007 alone.

The proportion of BCIs performed simultaneously and sequentially across the responding clinics was investigated according to age at second implant for the following subcategories: under 3 years, 3 to 10 years, 11 to 18 years, and 18 years and older (adults). With the exception of three responder clinics, all clinics reported experience with bilateral implants in both child and adult populations. One clinic reported experience in children only, being part of a children's hospital facility, and did not answer survey questions related to BCI in adults. The remaining two clinics care for both pediatric and adult CI patients, but one clinic has BCI experience with children only and the other with adults only. These last two clinics responded to questions for all subject groups. Table II and Figure 1 demonstrate the numbers of BCIs for all clinics broken down by age, sequence of surgeries, and time period. Cumulatively to the end of 2007, 70% of all BCI surgeries had occurred in children, with the 3- to 10-year-old age group having the highest representation (33% of all BCIs), followed in order by adults (30%), children under 3 years of age (26%), and children between 11 and 18 years of age (11%). Seventy-two percent of all BCI surgeries were performed sequentially (70% of children, 76% of adults). Children <3 years of age represent the only age group of all patients in which simultaneous surgeries predominate (58% simultaneous). For all other age groups sequential surgeries far outnumber simultaneous (3–10 years, 84% sequential; 11–18 years, 94% sequential; adults 76% sequential). T2 F1

Eighty-four percent (21/25) of responders reported experience with simultaneous BCI in children, with 68% (17/ 25) reporting experience in children below 12 months of age. Experience with sequentially implanted children was reported by 84% (21/25) of responders, with the youngest cases ranging between 9 months and 4 years of age, with an average of 23 months. In these sequentially implanted children, interimplant intervals ranging between 5 and 18 years, with an average of 11.8 years, were reported. As the potential consequences of a postmeningitic infection may specifically give rise to consideration of early intervention in both ears, responders were asked to report on simultaneous bilateral experience separately in both postmeningitic and nonmeningitic children under 12 months of age. Thirty-two percent of responders (8/25) had experience with postmeningitic cases, 60% (15/25) reported experience in

TABLE II.
Number of Bilateral Implants by Age and Sequence for Three Time Intervals (Prior to 2007, During 2007, and Cumulatively to the End of 2007) Broken Down for United States, non-United States, and All Clinics Combined.

	Prior to January 1, 2007 (n = 2,147)			January–December 2007 (n = 733)			Prior to January 2008 (n = 2,880)		
Time Interval Subgroup	Total No.	Non-United States Clinics	United States Clinics	Total No.	Non-United States Clinics	United States Clinics	Total No.	Non-United States Clinics	United States Clinics
Sim child <3 yrs	272	177	95	160	114	46	432	291	141
Seq child <3 yrs	202	94	108	110	62	48	312	156	156
Sim child 3–10 yrs	131	108	23	20	14	6	151	122	29
Seq child 3–10 yrs	596	435	161	212	143	69	808	578	230
Sim child 11–18 yrs	1	1	0	18	11	7	19	12	7
Seq child 11–18 yrs	248	128	120	56	32	24	304	160	144
Total children	1,450	943	507	576	376	200	2,026	1,319	707
Sim adults	181	46	135	23	4	19	204	50	154
Seq adults	516	243	273	134	58	76	650	319	331
Total adults	697	289	408	157	80	77	854	369	485
Total patients	2,147	1,232	915	733	456	277	2,880	1,688	1,192

Sim = simultaneous; Seq = sequential.

nonmeningitic cases, and 24% (6/25) reported experience in both postmeningitic and nonmeningitic cases below the age of 12 months. Seventy-two percent (18/25) reported experience implanting postmeningitic children sequentially, with the youngest age at second implant ranging between 8 months and 10 years and an average of 3.4 years. In adults, 64% (16/25) of responders had experience with simultaneous BCI, and 84% (21/25) were experienced with sequential BCI.

The data represented in Figure 1 and Table II also show an evolving trend over time for clinical practices with respect to age at and sequence of BCI for the responding clinics. Prior to January 2007, 68% of BCIs were performed in children. This increased to 79% for the year 2007 ($P < .001$). With regard to children only, a change is apparent over time in terms of the age group making up the majority of pediatric BCI surgeries performed. Prior to 2007, children 3 to 10 years of age made up 50% of the children undergoing BCI, whereas those <3 years old made up only 33%. In 2007 this shifted more toward the younger age group (47% for those <3 years and 40% for 3–10 year olds; $P < .001$). With regard to any trend in the percent of simultaneous surgeries over time, the <3-year-old group remained unchanged (57% simultaneous prior to 2007, 59% during 2007), the 3- to 10-year-old and adult groups decreased (18% and 26% simultaneous, respectively, prior to 2007, 9% and 15% during 2007; $P < .001$ and .0025, respectively), and the 11- to 18-year-old group increased (0.5% simultaneous prior to 2007, 24% during 2007; $P < .001$).

When the clinics are broken down into United States (n = 10) and non-United States (n = 15) groups, some significant differences are noted. For the combined time period to the end of 2007, United States clinics had a higher proportion of adult BCI patients (59% children, 41% adults) than the non-United States clinics (78% children, 22% adults) ($P < .001$). With regard to children <3 years old, the non-United States clinics had a higher rate of simultaneous surgery than United States clinics (47% simultaneous for United States, 65% for non-

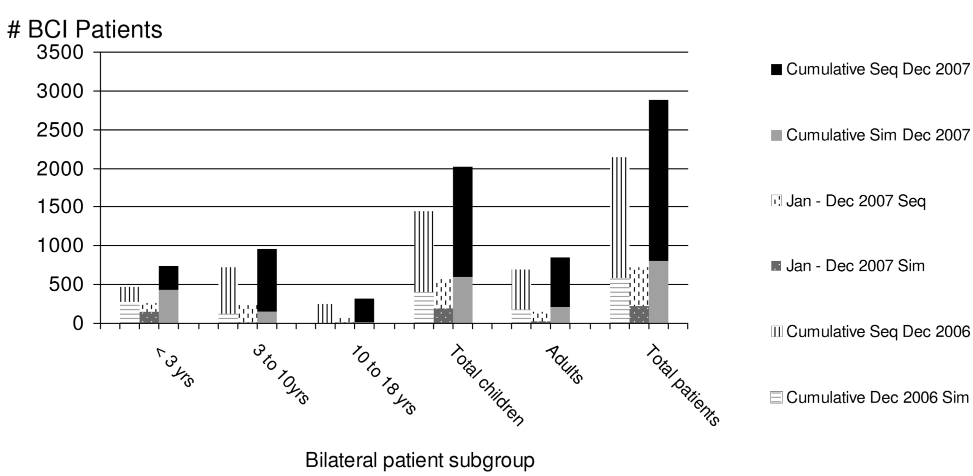

Fig. 1. Number of simultaneous (Sim) and sequential (Seq) bilateral cochlear implant (BCI) patients for each age subgroup and for each time period (prior to 2007, during 2007, and cumulative to the end of 2007).

TABLE III.
Summary of Candidacy Criteria in Relation to Subject Group.

	Subject Groups			
Candidacy Criteria	Children for Simultaneous BCI	Unilaterally Implanted Children	Adults for Simultaneous BCI	Unilaterally Implanted Adults
Minimum age				
Do not know	8%	4%	NA	NA
Not Predefined	22%	70%	NA	NA
Range	6–12 mo	6–12 mo	NA	NA
Mean	8 mo	12 mo	NA	NA
Maximum age				
Do not know	15%	11%	*	4%
No Upper limit	44%	70%	*	84%
Range	3–12 yrs	5–12 yrs	*	80 yrs
Mean	7 yrs	8.6 yrs	*	80 yrs
Maximum interimplant interval				
Do not know	NA	11%	NA	*
No limit	NA	48%	NA	*
Range	NA	5–12 yrs	NA	*
Mean	NA	8.6 yrs	NA	*
Monaural best aided speech (% word scores)				
Do not know	22%	30%	11%	7%
No limit	11%	22%	11%	11%
Range	20–60%	20–50%	30–80 %	40–60%
Mean	40%	40%	45%	50%
Binaural aided speech (% word scores)				
Do not know	26%	29%	11%	15%
No limit	11%	44%	19%	33%
Range	20–70%	40–60%	40–60 %	40–60%
Mean	45%	50%	50%	50%

*Omitted in error from the survey options.

Each criteria demonstrates the % responders who specified no limit/do not know and the range and mean calculated from the responders who specified a value. Speech scores are the range and mean of survey responses for age appropriate speech materials.

BCI = bilateral cochlear implant; NA = not applicable.

United States; $P < .001$). United States clinics have a higher rate of simultaneous surgery in adults (32% simultaneous for United States, 14% for non-United States; $P < .001$). However, it remains consistent between both United States and non-United States clinics that over the two time periods surveyed there was a significant trend of increasing representation of children in the BCI population. Prior to January 2007, 55% of United States and 77% of non-United States BCI surgeries were performed in children. During 2007, 72% of United States and 82% of non-United States BCI surgeries were in children (United States, $P < .0367$; non-United States, $P < .0096$). For both United States and non-United States clinics, children <3 years old remains the only group for whom the rate of simultaneous surgery is greater than or equal to that of sequential.

Routine Counseling on Bilateral CI Options

The majority of responders (96%) reported they routinely provide counseling to parents of newly diagnosed deaf children about the possibility of obtaining bilateral implants in either sequential or simultaneous procedures, whereas the remaining responders elected to do so only at the parents request for more information on bilateral implants. Seventy-two percent (18/25) of all responders reported counseling parents of existing unilateral CI-user children routinely during their regular annual hearing reviews in the clinic, 12% (3/25) did so only at the parents' request, and 16% (4/25) did not provide counseling on sequential bilateral implant.

In contrast, only 44% (11/25) of all responders reported counseling newly diagnosed adult CI candidates or existing unilateral CI-user adults (52%, 13/25) on bilateral CI options on a routine basis. Fifty-two percent provide such counseling only on request, whereas approximately 20% do not provide it at all for either group of adults.

Clinical Considerations for Specific Subgroups of Bilateral CI Candidates

Table III demonstrates responses to questions concerning candidacy criteria (age limits, interimplant interval, and speech discrimination scores) used for BCI. T3

TABLE IV.
Percentage of Responders Rating Each Possible Reason as a High or Very High Motivation for Considering Bilateral Implants by Parents of Deaf Children or Adult Candidates.

	Subject Groups			
Possible Motivation/Reasons Ratings	Newly Diagnosed Children	Unilaterally Implanted Children	Newly Diagnosed Adults	Unilaterally Implanted Adults
Entering mainstream school	74	85	NA	NA
Adult education	NA	NA	44	40
Employment skill needs	NA	NA	60	84
Poor speech understanding and language skills development to date/overall for adults	56	74	64	76
Difficulties hearing in noisy situations	78	78	73	80
Difficulties listening in large rooms or spaces	70	62	54	72
Difficulties localizing sounds in the daily environment	82	78	73	76
Believe their child will be/they will feel safer with two ears	77	59	46	50
Are concerned they are not doing well enough with only one ear	NA	NA	12	60
Believe if one CI does not work the other is available to fall back on	74	74	50	40
Poor school grades for school-aged children	46	50	NA	NA
One surgery, one anesthetic, one healing process instead of two	56	NA	46	NA
Parents want the best for their child and believe the best is two CIs	89	82	NA	NA
Two deaf ear diagnosed therefore two implants are requested by	37	44	19	28
To make life easier	62	62	56	67
To bring them closer to their normal-hearing peers	73	65	NA	NA
To capture the better ear	44	39	50	44
Improved quality of life	82	74	75	72
To enable learning language to occur as far as possible all the time, known as incidental learning	74	67	NA	NA

NA = not applicable; CI = cochlear implant.

It is evident that a large percentage of responders do not hold firmly to a minimum or maximum age by which they limit BCI. When specified, minimum age limits for children range from 6 to 12 months, with a mean of 8 months for simultaneous and a mean of 12 months for sequential BCI. Maximum age limits for children were specified by only 32% of responders in regard to simultaneous BCI, and only 12% for sequential BCI. The majority, 88%, do not hold to an upper age limit for adults. Speech perception scores that are used as criteria for BCI do not differ significantly across subject groups.

Hearing Aid Trials

Hearing aid (HA) trials were considered essential prior to either simultaneous or sequential BCI by 68% (17/25) of responders for children and by 52% (13/25) for adult patients. A trial period with hearing aids or bimodal stimulation (CI + HA) was not considered necessary under the following conditions: signs of early ossification/postmeningitis (mean of responders for all subject groups 61%), if wearing a hearing aid is medically contraindicated (mean 42%), when there is inoperable canal atresia (mean 29%), if auditory brainstem response (ABR) is $>$100 dB HL (mean 26%), for ABR $>$90 dB HL (mean 19%), when pure-tone average (PTA) is $\geq$100 dB HL (mean 32%), and PTA is $>$90 dB HL (mean 26%).

Motivations, Contraindications, and Rehabilitation Time

Table IV demonstrates the percentage of responders who rated as high or very high various parent/patient motivations for pediatric and adult, simultaneous and sequential BCI. For all groups together, the highest rated motivations were to obtain improved hearing in noise, sound localization, and quality of life. For children, additional highly rated motivations include mainstream school placement, having a second CI as a backup, and parents wanting the best for their child. Additional adult motivations include employment skill needs and poor performance with a first implant. T4

Table IV demonstrates the percentage of responders who consider a listed condition as a contraindication for BCI. For all groups together, the lack of an oral/aural environment and/or the use of sign language as the main mode of communication are the conditions considered to be a contraindication by the highest percentage of responses. For children, the lack of parental motivation/support and body weight $<$6 kg were additional concerns. For adults, long-term profound deafness and vestibular problems were highly rated concerns.

The estimated clinical time typically devoted to rehabilitation of BCI users relative to that required for unilaterally implanted peers is shown collectively for the responder group in Figure 2. Responders were evenly divided between suggesting rehabilitation time was either F2

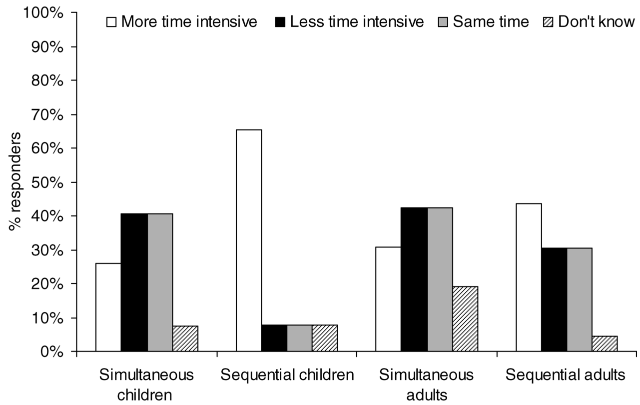

Fig. 2. Responder estimate of the relative rehabilitation time estimates for each bilateral patient group versus unilaterally implanted peers. N = 27 responders from 25 clinics.

less time-intensive or required the same time when considering simultaneously implanted children and adults. However, with regard to sequential BCI, a higher percentage of responders felt that these patients were more time-intensive, particularly in reference to sequential BCI in children ($P = .0016$).

DISCUSSION

This study involves both a retrospective analysis of clinical experience and a survey of current clinical opinion. The query of each clinic's past experience with regard to numbers and demographics of implanted patients over specified time intervals (section 1 of the questionnaire) is a retrospective analysis, and as such has sources of potential error. Responses may, as with any questionnaire based on retrospective experience, carry inherent bias depending on the responder's interpretation of the question, personal view, clinical experience, memory/recall, and the accuracy and ease of access to their clinical database. This is unavoidable to some extent in a large survey involving complex medical interventions. The electronic format of the survey was designed to allow for an open ended amount of time for completion in hopes of improving the accuracy of responses.

The query of current opinion with regard to BCI candidacy (sections 2–5) also has sources of potential error. Internal validity (accuracy of each site's responses concerning their patient data) for the answers to these questions requires that the individual completing the survey accurately represent the philosophy of BCI candidacy in operation at their clinic. Eighty percent (20/25) of surveys were completed by the lead surgeon of each CI program, and 20% (5/25) were completed by lead audiologists or researchers, all of whom are either decision makers for BCI candidacy in their program or are well versed on their clinic's criteria and involved in the patient/parent counseling process. Nonetheless, because some variance of opinion can exist among care givers, even within a single program, it is possible that the answers given may not reflect the exact criteria used in all instances. In addition, the multiple-choice format used for some questions, designed for greater ease of

TABLE V.
Percentage of Responders Considering Listed Condition as a Contraindication or Not to Performing a Bilateral Implant.

	Subject Groups			
Possible Contraindications to Bilateral Implant	Newly Diagnosed Children	Unilaterally Implanted Children	Newly Diagnosed Adults	Unilaterally Implanted Adults
Body weight <6 kg	77	73	NA	NA
Additional cognitive or other disabilities	56	59	52	68
Lack of parental motivation or support/lack of social or family network	93	93	52	52
Hygiene issues	48	56	46	52
School-aged child not in oral/aural environment/sign language main mode of	85	84	89	88
Family environment not oral/aural	67	67	NA	NA
Unemployed	NA	NA	8	4
Vestibular problems in either ear/first ear	56	67	70	80
Sudden deafness (>6 mo recovery)/in second	15	8	19	4
No local rehabilitation therapist	41	33	NA	NA
Fear of surgery/surgical risks/second ear	62	70	59	67
Cochlear anomalies in one ear/nonimplanted ear	56	56	67	75
Cochlear anomalies in both ears	54	NA	63	NA
No hearing aid trial in contralateral ear	NA	67	NA	NA
Long term profound deafness without hearing aid trial	NA	NA	96	84
Significant binaural benefit from bimodal	NA	78	NA	Not queried*

*Omitted in error from the survey options.
NA = not applicable.

completion, limits response options. Therefore responses may not fully reflect opinion on certain topics. Space was provided in the survey for narrative response in the hope of minimizing this effect.

External validity (generalizing these results to other worldwide CI clinics) is affected by the fact that the experience of these expert clinics may not be typical of other CI clinics. Higher volume, more experienced CI clinics are more likely to be involved in research and may attract a higher percentage of the pediatric population and more complex cases. Nonetheless, because it is these clinics that likely have the most BCI experience, they represent the best source of information for purposes of this survey. It is noteworthy that the demographic breakdown of BCI patients in clinics responding to this survey mirror the worldwide BCI numbers reported by the major manufacturers (Table I).

Overall, the results of the survey suggest a high level of confidence among these experienced CI clinics in the benefits of BCI. A large majority of those responding routinely provide counseling and information on BCI as a treatment option, at least with regard to children (96% do so for children, 50% for adults). The only clinic that does not provide such counseling for children is restricted from providing BCI treatment by national funding regulations. In regard to children, confidence in BCI is shown by the recent trend toward earlier application, providing BCI at a younger age, and doing so more often in younger children simultaneously (58% of all the BCI procedures performed on children under age 3 years) as opposed to sequentially. Although all responders were asked to put funding issues aside, it is unavoidable that the reimbursement policies under which each clinic operates will affect the BCI decisions that must be made. It is therefore possible that the strong focus and increasing trend to provide BCI for children as opposed to adults (68% of all BCIs prior to 2007, 79% during 2007 worldwide) is related at least in part to funding priorities. This is highlighted by the fact that United States clinics have a higher proportion of BCI procedures being performed in adults than do non-United States clinics (41% United States, 22% non-United States). However, the trends noted here may also be due to greater awareness of a growing body of evidence in scientific publications indicating the importance of age of implantation for each ear in children on hearing outcome from BCI.

Children

Children who underwent BCI in sequential procedures make up 70% of all BCI children and one half (49%) of the entire BCI user group (pediatric and adult) worldwide from the clinics answering the survey. This high representation is likely due at least in part to the relatively large number of unilaterally implanted children in existence once BCI treatment began to be utilized during the past several years. This group of sequentially implanted children has been the subject of the majority of studies and published articles on BCI treatment in children to date. With the addition more recently of a large cohort of simultaneously implanted children, a great deal has been learned from the influence of various ages and interimplant intervals present at the time of BCI. First and foremost, it has been shown that the risks and complications of BCI in children are very low in experienced hands.[20,21] Muller et al. were the first to report on early experience with BCI in a large group of 200 children, having no significant perioperative complications or protracted vestibular affects.[20] The current survey encompassing 2,880 bilaterally implanted children by experienced surgeons appears to suggest, by the confidence demonstrated, the continuing perception of BCI as a safe treatment approach.

Second, sequential BCI in children has shown the importance of the age at which the second ear is implanted on benefit received from that ear. Peters et al. demonstrated diminished second-ear speech perception performance with increasing age of second ear implantation for a group of 30 sequentially implanted children, all of whom had early first-ear implantation.[19] Other studies have also indicated an inverse relationship between either increasing interimplant interval or age at second CI on speech perception performance, spatial hearing, or binaural advantage achieved.[5,6,11,22–25] Neurophysiologic correlation has added weight to the theory that there is a neurobiologic basis for this age effect in children. Using the P1 wave latency of the cortical auditory evoked potentials as a biomarker for central auditory maturation, repeated studies have shown an age-related neurobiologic limitation for benefit received from both unilateral and bilateral implantation.[26–30] This limitation has been characterized as an age-related critical period, with the implication being that the younger a child receives an implant in each ear, the better the hearing outcome is likely to be.

In addition, recent studies have also shown an advantage to binaural brainstem pathways if the surgery is performed simultaneously or with short interval sequentially in children <2 years of age. Postulating that any limitation in development of the second ear might compromise the development of central binaural processing and the subsequent ability to integrate the information received from each ear, researchers have examined the impact of various interimplant intervals for a cohort of prelingually deafened children (i.e., receiving bilateral implant simultaneously, sequentially with short interimplant delays, and sequentially with long interimplant delays).[31] Comparison was made of the electrically evoked compound action potentials of the auditory nerve and electrically evoked brainstem responses. Preliminary results show a dependency on length of interimplant interval and age at first implant upon the rate of change of the eV latencies reflecting binaural interaction following long-term binaural stimulation. Longer interimplant intervals and older age at first implant have a negative impact, appearing more restrictive to the development of pathways in the auditory brainstem. The authors suggest this reflects a change in developmental plasticity in children with long-term unilateral implant use at the level of the auditory brainstem essentially resulting in unbalanced activity for its duration and potentially longer term, restricted or altered course of maturation evoked by the stimulation of the second side, and increased sensitivity to competition from other nonauditory modalities resulting in cross-modal plasticity on the delayed side. In due course, their findings imply that subsequent binaural listening benefits, such as speech understanding in noise and spatial hearing, may develop more rapidly for the young simultaneously implanted child under 2 years of age compared to young sequentially implanted children undergoing only implant in the first ear before the age of 2 years. Despite this demonstrated pattern, many of the studies cited herein include individual exceptions, children receiving their second implant at an advanced age with a large interimplant interval, but who perform significantly above the mean for their age group on the measures studied.[19] Factors that may help distinguish such children preoperatively have yet to be clarified.

It is not surprising then to see the trends shown in this survey with regard to BCI in children. The percentage of children undergoing BCI who are <3 years old increased from 33% prior to 2007 to 47% during 2007. In addition, simultaneous BCI is being applied much more frequently to children <3 years old than to any other age group. Although the data showing improved outcomes with BCI in younger children discussed above is likely the main motivating factor for the trend toward simultaneous surgery, reduced demands on clinic rehabilitation time may be another. Figure 2 demonstrates that 68% of clinics feel that sequential BCI in children requires significantly more clinic rehabilitation time overall than unilateral CI, but not so for simultaneous BCI. Respondents may feel that limited personnel and/or financial resources in the clinic setting may make simultaneous BCI a more efficient treatment approach.

Despite the research findings of reduced performance with increasing age of BCI, only 12% of responders consider there to be an upper age limit for second ear, sequential implantation ranging from 5 to 12 years. For simultaneous BCI, 32% reported having an upper age limit ranging from 3 to 12 years (mean, 7 years). It is possible that the 36% of responders who believe there is no upper age limit for simultaneous BCI have in mind older children with progressive hearing loss who have acquired proficient spoken language capabilities with their acoustically aided residual hearing up to the time of implantation, or alternatively, older children incurring sudden loss bilaterally (e.g., trauma, drug related, disease related). This is unlikely to be true for the 88% of responders who reported having no upper age limit for sequential BCI, because to be candidates for their first CI, these children would typically have had bilateral profound hearing loss at an early age. Regardless, it is apparent that these responders feel that the decision to provide or limit BCI in older children should not be based on age alone. With regard to lower age limit, 92% of responders appear to be comfortable with implanting both ears before the child is 12 months of age (mean, 8 months), a fact also reflected by the trend seen in this survey toward earlier BCI for children in general.

Adults

As has been mentioned previously, adults make up only 30% of the BCI user group from clinics responding to this survey, 76% receiving their implants sequentially. The data bases of the three major CI manufacturers confirm the predominance of children over adults in the BCI patient population, as well as the difference noted between United States and non-United States clinics (Table I). In light of the fact that approximately 54% of the existing CI population worldwide are adults, this may indicate significant under-representation of adults in the BCI patient population. In addition, the fact that only 50% of clinics worldwide routinely counsel adults about the option of BCI (compared to 94% for parents of children) seems to suggest decreased availability of information and opportunity for adults. From an outcomes perspective this is surprising, because postlingually deafened adults appear to be the candidates who can achieve the greatest quantitative binaural performance with BCI of all candidate groups.[8,15] In fact, it is these adults who have provided the majority of the information with regard to the subjective benefits of BCI compared to unilateral CI, such as a decrease in attention effort, decreased social restriction, reduced perception of hearing disability, and a trend toward reduced emotional distress.[4–15] It appears then that greater focus is being placed on maximizing the developmental outcomes of children through the provision of bilateral stimulation than on providing binaural benefits to adults. The degree to which this is due to the philosophical leanings of CI professionals, healthcare funding priorities, or both is unclear.

United States and non-United States Clinics

For certain demographic aspects of this survey, clinics were divided into United States and non-United States groups. This was done to analyze how differences in the healthcare system among countries might impact BCI patterns at the surveyed clinics. The United States has the most privatized healthcare system in the world and is the only developed country without a universal health care component.[32,33] Only 45% of annual healthcare expenditure in the United States is provided through government funding, which is the least of all developed countries in the world.[34] Reimbursement for medical intervention in the United States system requires primarily that a treatment be established as safe, effective, and accepted as a standard of care. Once accepted and established, prioritization of healthcare resources to one candidate group over another does not typically occur. Without a cap on annual healthcare expenditures, treatment such as CI and BCI are provided to any patient who stands to benefit as long as the patient has a source of reimbursement. The establishment of long-term cost effectiveness, which for some medical treatments takes many years, especially for children, can be determined in due course while being supported under the United States system. The sample of United States clinics in this survey represent 29% of the total estimated BCI population in the United States

(31% of children, 26% of adults). The proportion of children to adults in the BCI United States survey group is 59% children, 41% adults compared to 55% children, 45% adults in the United States as a whole ($P < .0082$) (Table I). This suggests that these expert clinics attract a higher percentage of the pediatric population than the average United States clinic involved in BCI.

The non-United States countries represented in this survey have healthcare systems that, although differing from one another in significant and tangible ways, are similar in that government determination of healthcare expenditure is dominant.[34] The individual non-United States clinics are not able to represent the average CI clinic in their country, because funding for CIs and BCIs can vary greatly in different geographic locals even within the same country. The most notable example is the Canadian clinic, which receives research funding for BCI in children, and is therefore not limited by the usual annual CI allotment determined by each individual province.[35,36] Such provincial funding for BCI was very limited in Canada during the time frame covered by this survey. In Spain, clinical funding is also regionally based. Both Spanish clinics represented in the survey are from regions where funding for unilateral CI and BCI is provided by the regional budget. This is not true in many other parts of Spain.

During the time frame covered by this survey, the French Health Ministry had CI treatment placed on their innovative technology list, which significantly limited funding for CI treatment throughout the country, especially BCI. As of March 2009, CIs have been placed on the List of Product and Presentation for Reimbursement.[37] This list covers the recommended and funded treatments to be provided nationally in France. It also lists those centers permitted to perform the CI procedure. CI is now considered an acceptable funded treatment unilaterally for children and adults, but BCI is approved primarily for adults. BCI for children is limited in France to special cases only (e.g., meningitis, Usher syndrome, trauma). The health ministry has requested more evidence on the safety of BCI before considering further application in children. France, therefore, stands in stark contrast to the overall focus of BCI in children seen in the other non-United States countries in this survey.

The United Kingdom center is another example of note. Significant geographic variations within the country existed at the time this survey was conducted, being mentioned as part of the Cochlear Implant Services Commissioning Guidelines report which stated, "At the moment, there is no national agreement on clinical criteria for cochlear implants and no national guidance. As a result, there are large geographical variations in terms of the numbers and types of cases commissioned each year. This has led to inequitable access, with long waiting times, closed lists, and a refusal to accept referrals in some areas (particularly for adults)."[38]

This is meant only to illustrate that with such diverse geographic variations in funding for CI within a single country, it is unlikely that the individual clinics responding to this survey are representative of their countries as a whole. Placing all non-United States clinics into one group is meant to distinguish the overall effect of nationalized healthcare systems on BCI trends.

The determination of long-term cost effectiveness and health-related quality of life (QoL) plays a more significant front-end role in prioritizing expenditure in health care systems represented by some clinics in the non-United States group. With regard to BCI, conflicting QoL data has been published to date for adult patients. The United States study by Bichey and Miyamoto was the first to show significant improvements in QoL and a favorable cost utility for BCI in adults.[12] This had been preceded by the study of Summerfeld et al. in the United Kingdom, which suggested that the gain in quality of life from BCI was too small to achieve an acceptable cost-effectiveness ratio.[39] However, concern has been raised within the United Kingdom about this front-end weighting of QoL measures, particularly in regard to children, in light of the need in research for more sophisticated quantitative measures that fully reflect the reported subjective benefits of binaural hearing.[40] The National Deaf Children's Society in the United Kingdom has provided a statement that says, "NDCS (National Deaf Children's Society)...would like to see measures outside of health-related QoL being considered as alternatives when assessing benefit. Health related QoL are inadequate when measuring benefit of cochlear implantation in children and comparisons with adult data should be viewed with extreme caution. We are concerned that the recommendation of bilateral implantation in children and adults done solely in the context of research will prove to be a barrier to obtaining the information required. Within funding contexts of the NHS (National Health Service) it will prove extremely difficult to obtain this data."[40]

The assertion here is that the confinement within some countries (such as the United Kingdom) of BCI treatment to funded, approved research studies, particularly those focused on evaluating health-related QoL as the prime outcome measure, presents significant logistical challenges to obtaining worthwhile data from a diverse CI patient population. Since the completion of this survey at the end of 2007, the United Kingdom's National Institute for Health and Clinical Excellence (NICE), which guides healthcare purchasing policy within their country, has made further recommendations for provision of BCI.[41] The recommendations approve funding for simultaneous BCI in children. Sequential BCI in children is approved only for those who were unilaterally implanted prior to the new recommendation in February 2009, thus establishing a precedent moving forward of providing BCI to children simultaneously. In contrast to the recent policies in France, NICE recommends against BCI in adults, viewing it as insufficiently cost-effective at this stage, except in cases such as combined deafness and blindness, or multiple handicaps.

As can be seen from Table I, the United States has 39% of the total worldwide CI population based on the three major manufacturers, but has 52% of the BCI population. The United States has a higher proportion of adults in both its unilateral and bilateral CI population than the rest of the world taken cumulatively. The reasons for this are multifactorial, but greater freedom to provide BCI to the general CI population once safety and efficacy are established may be a major one.

Hearing Aids

From the very beginning of the cochlear implant era, candidacy has been determined based on audiometric parameters obtained while wearing optimally fitted hearing aids. As CI technology has improved, the HA performance criteria of those considered to be a CI candidate has broadened. A trial of daily HA utilization for one to several months has been considered an invariable part of the candidacy process. It is interesting to see that close to one third (30% in children, 40% in adults) of responders to this survey consider a HA trial unnecessary if certain patient characteristics are met (e.g., ABR or audiometric thresholds >100 dB, early ossification). To what degree this represents a level of confidence in predicting how unaided audiometric or evoked potential testing or aided performance with stock hearing aids may correlate to aided benefit after a hearing aid trial, based on published evidence or their own personal clinical experience, is difficult to say from this survey. However, it is apparent that some responders do not consider an extended HA trial to be a rigid, inflexible requirement for all patients.

Speech Discrimination Criteria

There is no clear consensus in this survey on the level of individual ear HA speech perception performance or bimodal performance that should be used as criteria for BCI. This is partly due to the fact that several languages are represented by the responder group, in addition to a variety of test materials in use, even for the same language. One third of responders (8/25) apply no maximum bimodal performance limit. The responders who specify word test scores (9/25) specify a range from 20% to 80%, mean 45% to 50%, and appear to be applying a range of currently accepted unilateral CI criteria to each ear, instead of having separate, distinct criteria for the second ear. Such an approach is reasonable because those criteria are predictive for significant CI benefit in the majority of patients. Although questions about criteria for a level of bimodal speech perception performance were asked with regard to sequential implantation, we did not distinguish criteria for a level of bimodal gain (speech performance for CI + HA vs. CI alone conditions). In the future it may be determined that a certain level of bimodal gain is useful in predicting such a patient's perceived benefit from using an HA in the ear opposite a CI and in distinguishing candidacy for sequential BCI.

Motivations for BCI

Responders to the survey acknowledge numerous, diverse motivations for considering BCI in children and adults. Some of these motivations simply reflect the desire to provide the known auditory benefits of binaural hearing, that is, sound localization ability and better hearing in noise.[1,2] Some motivations, however, have more intangible goals in mind, such as Improved quality of life, enable incidental learning of language, parents wanting the best for their child, and making life easier. These are all goals for which 56% to 89% of responders stated they had high or very high motivation to provide through BCI. Adults and parents of children who have received BCI commonly report such intangible, subjective benefits that are difficult to quantify based on our current performance measures. As can be seen in Table IV, whenever a potential motivation was applicable to both children and adults, the number of responders rating a motivation as high or very high was at least equal to, if not greater for, children than for adults, again suggesting a predisposition toward providing BCI for children.

Contraindications

Except for the concern about the risk of surgery in low body weight infants, the dominant concerns for responders have to do with a lack of parental motivation/commitment and absence of a predominantly oral mode of communication used by the patient, family, or education environment. These are, of course, major concerns when considering a patient for unilateral CI. In addition, long-term profound deafness without a history of HA experience in adults was considered a possible contraindication by 96% of responders. This type of hearing history can apply to one or both ears of a potential candidate. BCI in adults with a history of profound hearing loss since early childhood in one or both ears is a topic that is an extension of the same discussion in older, prelingually deafened children. Concern about the effect of auditory deprivation and diminished central auditory development on CI performance in such cases is appropriate. Yet, as is true of older children with prelingually deafened ears, reports exist documenting adults who have achieved significant CI benefit in such circumstances.[42,43]

CONCLUSION

BCI in children and adults is being offered and performed frequently at the higher-volume, experienced CI clinics responding to this survey, in the majority of cases as a routine clinical procedure. Although all age groups are represented in this BCI patient population, children are receiving BCI more frequently than adults. In particular, there appears to be a trend of increasing focus on providing BCI for children <3 years of age. Sequential implantation predominates in all age groups except in children <3 years old, where simultaneous BCI is more common. The United States clinics in this survey have a higher proportion of adults in their BCI population than do the non-United States clinics. The reported motivations for BCI indicate an expectation and/or experience among clinics that BCI can provide significant quantitative binaural benefits in addition to many subjective ones. The BCI candidacy criteria for specific patient categories used by the responding clinics mirror the audiometric criteria commonly used for considering unilateral CI. Differences in healthcare funding may explain geographic differences among clinics in the use of BCI and in the provision of BCI for children over adults. As represented through this survey, worldwide experience with BCI is now quite extensive and provides a useful base for evaluating clinical outcomes across patient categories and for providing further support during the patient/parent counseling process.

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APPENDIX I: LITERATURE REVIEW SUPPORTING BILATERAL COCHLEAR IMPLANT TRENDS

INTRODUCTION

As can be seen from our main article, there are trends of increasing application of bilateral cochlear implantation (BCI) among experienced cochlear implantation (CI) centers suggested by the survey. In addition, there is a trend of increasing focus to provide BCI in children at a younger age and more often both ears simultaneously. To what extent published research on BCI supports these particular trends seen in our worldwide survey will now be discussed.

HISTORICAL TRENDS IN THE LITERATURE RELATING TO BCI

There is an extensive base of psychoacoustic literature documenting the mechanisms and benefits of binaural hearing input in normal hearing individuals, dating back to the 1970s.[A1–A4] In addition, the benefits realized by bilaterally hearing impaired individuals from the use of bilateral amplification have been well documented, such that bilateral hearing aid fitting has become standard practice for such patients.[A5–A7]

The earliest published report of a patient undergoing BCI was in 1988.[A8] As was true of such early case reports concerning BCI, the prime focus at that time was on the comparison of CI technologies, that is comparing an older, single channel device in one ear with a newer, multichannel device in the second ear.[A9] The hope was to improve on individual ear performance with more advanced CI technology in the opposite ear. In the late 1990s and early 2000s, BCI began being performed with a view toward improved individual ear performance with similar or identical CI technology in each ear. As late as 2004, studies were published in which BCI was performed with a view toward capturing better individual ear performance with similar CI technology. Ramsden et al. commented on their inability to reliably predict which ear in any given patient would perform better with a CI, and that with BCI they assured capture of the better performing ear.[A10] In such cases, the variable was not the level of sophistication of CI technology, but the differences in CI performance potential between ears of an individual patient. They also estimated that many of the patients undergoing BCI prior to 2005 had done so because of medical or technical complications with their first implant.

In the late 1990s, interest in BCI for the primary purpose of developing or restoring binaural hearing mechanisms began to emerge.[A11,A12] Most of the early reports referenced above commented incidentally on the ability of patients to integrate the sound from both implanted ears, with very different technology and signal processing from each side. It has been primarily in the last 8 years that BCI research has focused in great detail on the true benefits of two cochlear implants working together over just one alone.

BCI IN CHILDREN

Neurodevelopmental Evidence

Support for a bilateral approach to the treatment of hearing loss in children starts with that which is contained in the hearing aid literature. Strong support for the use of bilateral hearing aids was established by documentation of the negative effects that auditory deprivation has over time on the ability of individuals with hearing loss to understand speech. Silman et al. in 1984 and Gatehouse in 1992 demonstrated in adults that if only one ear receives a hearing aid when both ears have hearing loss, the untreated ear will show a significant deterioration in speech discrimination over time.[A13,A14] Children have shown this same effect with regard to hearing aid use for moderate to severe hearing loss.[A15,A16] In 1996, Willott extrapolated from animal research in postulating that central auditory system (CAS) plasticity is the underlying process that explains the loss in auditory capabilities from sensory deprivation, as well as the improvement that can occur from acclimatization and conditioning with hearing aid use.[A17]

Plasticity within the central nervous system can be viewed as having two general phases. The primary phase is that which occurs in infancy and early childhood, known as the "critical period." This expression was first used in reference to the developing mammalian visual system by the groundbreaking work of Wiesel and Hubel (1963).[A18] Theoretically, it is during this critical period of primary neuroplasticity that a neural pathway awaits specific instructional information to continue to develop normally. If such appropriate or normal experience is not gained during this time, the pathway never attains the ability to process information in a normal fashion, and as a result, the sensory perception or behavior is permanently impaired.[A19] An additional concept in this primary phase is that of a "sensitive period," which varies from a critical period in being less absolute and/or rigid. During a sensitive period, a pathway maintains a state of vulnerability to new stimuli. Whatever experience is maintained during this period leads to the pathway sustaining those adaptive changes permanently.[A20] The acquisition and integration of such neural information occurs only for the limited time frame of the sensitive period for that stimulus.

The second general phase of neuroplasticity can be called the secondary or residual phase. During this phase there is the sustained ability of the nervous system to adapt and reorganize to changes in stimulus.[A21] This phase appears to persist throughout an individual's life. Based upon data that will be discussed in succeeding sections, the extent to which secondary plasticity can reorganize neural pathways appears to be significantly limited by the initial organization that occurs during the sensitive/critical period, but can be substantial nonetheless.[A22]

Measures of Cortical Maturation

It is now well established that the CAS is maximally plastic during the primary critical or organizational phase that lasts from birth to approximately 3.5 years of age.[A23–A25] The neurodevelopmental effects of auditory deprivation on the CAS in children with congenital hearing loss have been indirectly examined by computing P1 wave cortical auditory-evoked potential (CAEP) latencies. The P1 wave is thought to originate from auditory projections in the thalamocortical pathways, and its morphology and latency are felt to be reliable indicators or biomarkers of CAS cortical maturation.[A26] Evidence from intracranial recordings in humans, as well from animal models, suggest that the neural generators of the P1 CAEP originate from the thalamocortical projections to the auditory cortex and represents current activity in the auditory cortex, including input from feedback and recurrent loops between primary auditory and association areas.[A24,A26–A28] A highly sensitive period from birth to approximately 3.5 years has been consistently demonstrated, during which time, if cochlear implantation occurs in congenitally deaf children, a normal trajectory of P1 latency development is made possible.[A23,A24]

The clinical utility of CAEP is supported by research investigating CI performance outcomes for children implanted before the age of 4 years using a series of functional tests, including open set word recognition and expressive and receptive language acquisition. These clinical outcome measures also confirm that the CAS is maximally plastic during the first 2 to 3.5 years of life, reaching its peak for receptive language development prior to the age of 2 years.[A25] Furthermore, this same data suggests that the window of opportunity for language acquisition is already starting to close by age 4 years.

Studies have more recently set out to analyze the neurodevelopmental impact of BCI on children with bilateral congenital deafness. Electrophysiologic measures have been used to assess the central auditory development in BCI children of various ages and with variable interimplant delays versus unilateral CI.[A23,A24,A29] Examination of P1 CAEP latencies for BCI children for whom both ears were implanted before 2 years of age showed that for simultaneously implanted subjects, P1 latencies were within normal limits for each ear as early as 1 month poststimulation. For children who were sequentially implanted before 2 years of age, normalization of P1 latencies took 3 to 6 months after the second CI activation.[A23] In an extended longitudinal comparative study examining the impact of either sequential or simultaneous BCI before the age of 3.5 years upon P1 latencies, researchers showed no significant differences between the simultaneous or sequentially implanted groups.[A24] By 3.5 months postimplant, both P1 latency and morphology were considered within normal limits for both groups.

This is in contrast to studies of older children receiving a sequential bilateral implant after the age of 7 to 9 years, who generally show limited plasticity in the second ear as indicated by persistent P1 latency delays, disrupted morphology of the CAEP responses, and poorer behavioral outcomes on functional assessments.[A30,A31] In essence, this implies that the CAS linked to the second CI ear has limited neurodevelopmental organization potential if implantation of that ear is delayed for several years or more.[A31] Generally, for older children approaching their teenage years who underwent early first ear CI, functional benefits derived from late sequential implantation of the second ear, including speech discrimination in quiet and in noise and measures of spatial hearing, are significantly reduced and develop at a much slower rate relative to those displayed by their first implanted ear and relative to those demonstrated by children implanted sequentially at a younger age.[A31–A34] Taken altogether, the research evidence is overwhelmingly in favor of a focus on BCI in children at as young an age as possible for both ears.

Measures of Brainstem Binaural Integration

The finding by Sharma et al. (2007) that P1 latencies develop normally for both ears in young BCI

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children regardless of whether they are sequentially or simultaneously implanted does not rule out the possibility that there may still be a functional advantage derived from simultaneous BCI. Binaural integration and interaction are additional aspects of BCI performance, over and above individual ear speech discrimination performance, which must be considered. The majority of binaural cues, such as the detection of interaural differences in timing, intensity, and frequency spectrum are encoded at the brainstem level.[A35] These cues are important for sound localization and many other aspects of listening in adverse environments.

Recent research in animals has established that monaural compared to binaural auditory stimulation results in different neural pathways being traveled.[A36] Although bilaterally deafened animals show asymmetry of the ascending projection from the inferior colliculus similar to that observed in normally hearing animals, in contrast, unilaterally deafened animals show an increase in the afferent projection from the normal cochlear nucleus to the ipsilateral inferior colliculus. Data from both animal and human models also show robust contralateral and weak ipsilateral stimulation (crossed and uncrossed fibers of central auditory pathways) after unilateral implantation.[A37] Reorganization is occurring centrally in the presence of only unilateral stimulation, which pending the duration of unilateral stimulation, may or may not be reversible. Initially, such unilateral stimulation may act as a primer, but over the long term it appears to act as a restrictor to development of the auditory pathways connected to the second ear and subsequently to binaural processing.[A23,A29,A30,A37]

As was discussed in the body of our article, any delay in the receipt of bilateral auditory input may compromise the organization of brainstem binaural processing and the subsequent ability to integrate the information received from each ear.[A29] Monaural stimulation in children appears to organize the brainstem in very different ways than binaural input.[A36] When measuring electrically evoked compound action potentials of the auditory nerve and electrically evoked brainstem responses (EABR), preliminary results show a dependency of length of interimplant interval and age at first implant upon the rate of change of the eV latencies, reflecting binaural interaction following long-term binaural stimulation.[A29] Longer interimplant intervals and older age at first implant have a negative impact, appearing more restrictive to the development of CAS pathways in the brainstem. This suggests a change in developmental plasticity in children with long-term unilateral implant use at the level of the auditory brainstem, which may not be reflected in abnormal P1 latency or morphology.

What is not clear from the literature is whether the acquisition of binaural brainstem mechanisms has a more limited or fragile critical period than does the development of cortical speech perception abilities, or if it is simply the restrictive effects incurred by prolonged unilateral stimulation that inhibit optimal integration of the later arriving second signal. The EABR studies mentioned above have yet to be correlated with behavioral outcomes of binaural processing in children. However, from a neurodevelopmental perspective they seem to suggest that even if a sequentially implanted child receives a second CI at a young enough age to acquire open set speech discrimination abilities, brainstem mechanisms may have acquired permanent limitations if the time between implants is significantly delayed. From preliminary research involving observation of a small group of 4- to 15-year-old sequential BCI children for 6 to 12 months, findings consistently demonstrated the ability to use the auditory speech information received at the second implant ear. However, the demonstration of benefit from binaural processing within this time frame was far more variable.[A34] Taken altogether, the preliminary data favors simultaneous BCI in young children over a sequential approach.

A sequential approach to BCI in children may at times be unavoidable or even advisable, depending on individual patient or parent factors. Some recent data suggests that in some cases there may be certain advantages to delaying a second implant by 1 to 2 years, as young children may benefit more in early childhood from the improvements in prosody and inflection provided by an implant and a hearing aid (in the opposite ear), compared to two early simultaneous implants.[A38] However, in general performance on speech perception, measures in the second implant ear and binaurally are viewed as not only superior but tend to develop faster for children implanted sequentially at as young an age and with as short of an interimplant interval as possible.[A31–A33] Wolfe et al. (2007) demonstrated in examining 12 sequentially implanted children (first implant <3 years of age, second <9.5 years of age) that all showed statistically significant benefits from bilateral stimulation at 1 year following BCI, but the greatest benefit was for children implanted sequentially before 4 years of age, whereas those implanted after 4 years demonstrated lower binaural benefits and greater interaural performance differences.[A33] Another 3-year follow-up study on sequential BCI children also demonstrated greater interaural performance differences on speech measures at 1 and 2 years after the second CI for children with 5 years plus interimplant intervals. In some children, interaural differences continued to improve up to 3 years after the second CI, suggesting more time may be required for children implanted sequentially at older ages than in younger children despite early intervention in the first ear.[A32] Therefore, it is possible that CAS plasticity may be preserved in some children, allowing them to perform well even after late second-ear BCI.

BCI in the Very Young

The advent in some countries of universal newborn hearing screening and screening of high-risk infants has lead to early referral and presentation of children with hearing loss. The opportunity to implant these children prior to 12 months of age more frequently presents itself. With regard to unilateral CI, surgery down to 6 months of age is being reported as safe and anatomically feasible.[A39–A42] In experienced hands, the surgical complication rate does not appear to be any higher than in other age groups. Dettman et al. (2007) report on 19 infants who received unilateral CI under 12 months of age (mean age, 0.88 years) and found that the mean rate of expressive and receptive language growth was greater than for a group of 87 toddlers who received their CI between 12 to 24 months of age (mean, 1.6 years).[A39] The same improved outcomes for children implanted before 12 months of age have been shown in other studies.[A41]

There are no published series reporting on BCI in children <12 months of age. Extrapolation from the evidence presented in this review would suggest that such early BCI intervention will likely optimize outcomes even further, as long as surgical complications are not increased. Options include simultaneous BCI or very short sequential BCI, with both devices being implanted prior to 12 months of age. Consistent with the reports of safety for CI in the very young, 92% of responders to our survey are comfortable implanting both ears of a child prior to 12 months of age.

Older Sequentially Implanted Children

A correlate to the importance of early CI and BCI in children exists in the ophthalmologic literature regarding various critical periods for components of visual integration: binocular fusion, ocular dominance, cortical retinotopic maps, direction sensitivity, and complex feature recognition.[A21] Plasticity and critical periods in the visual system are some of the most thoroughly studied cortical functions because of the ease of manipulating visual input independently in each eye. Visual acuity and complex feature recognition (such as facial recognition) in one sense can be considered analogous to auditory speech discrimination. Stereopsis is akin to binaural stereo hearing. Stereopsis is the aspect of visual perception that results from two different views of the world obtained by virtue of each eyes' different positions in the head. These two slightly dissimilar retinal images result in a binocular or horizontal disparity, that when optimally fused in the visual cortex, creates a three-dimensional perception of objects.

Childhood amblyopia can take several forms: refractive amblyopia or anisometropia (unequal refractive error between the two eyes), form deprivation (as in congenital cataracts), and strabismus (misalignment of the eyes). Failure to correct during infancy any of these abnormalities can result in permanent loss of visual acuity in the effected eye.[A43,A44] In the case of childhood strabismus, the two retinal images are too disparate for the visual cortex to create a fused image. If this were to be acquired in adulthood it would result primarily in diplopia. However because of the high, primary neuroplasticity of infants, it instead results in suppression of cortical development of the image from one eye, whichever image is of poorest quality. If correction does not begin before the closure of various visual critical periods (18 months to approximately 5 years of age), the child will have permanent loss of various visual capacities, depending upon the duration of deprivation (e.g., loss of stereopsis, strabismic amblyopia with reduction in visual acuity), even though the eye itself may be normal.[A21,A45,A46]

Although, as is true with CI in children, the best outcome is achieved if amblyopia treatment is started before age 5 years, and recent research has shown that children older than 10 years and even some adults can show improvement in visual acuity in a previously untreated eye.[A22] Children ages 7 to 12 years who wore an eye patch and underwent vision therapy were four times as likely to show a two-line improvement on a standard 11-line eye chart than amblyopic children who did not receive treatment. Adolescents aged 13 to 17 showed improvements as well, albeit in smaller amounts than younger children. The results of the study by Scheiman et al. (2005) suggest that the closure of the central visual system critical period does not mean a complete absence of plasticity.[A22] Some researchers argue that secondary or adult plasticity, that which occurs after closure of the critical period, represents plasticity that is independent of earlier critical period plasticity.[A47] It is not known whether or not the neural, synaptic mechanisms of older brain plasticity are the same as those active during the critical period in children. Many studies suggest they differ in significant ways.[A47,A48] Controlling or enhancing such adult plasticity mechanisms has numerous relevant potential applications.

Findings similar to those by Scheiman et al. (2005) have been seen in late sequential BCI children who show some ability to acquire modest open set speech capabilities in the late implanted ear. In the study by Peters et al., the mean second-ear speech perception score for children who received their second CI from 8 to 13 years of age was only 32%, but with a range of from 12% to 56%.[A31] These scores were significantly below those for younger children (the 3- to 5-year group had a mean of 83.9% with a range of 71%–96%) and below those for their first, early implanted ear. Yet some of these children perform significantly above the mean for their group, despite near lifelong auditory deprivation in the second ear. Litovsky et al. also noted in her study on sound localization abilities in a group of 13 BCI children, that one subject had a 12-year history of having no binaural hearing, yet was able to localize sounds after 23 months of using bilateral cochlear implants.[A49] Late, secondary plasticity in older children and prelingually deafened adults appears unable to achieve what is lost from deprivation in the critical period, yet modest benefits are still possible, and until more is known about predicting long-term gains in individual patients it seems appropriate that most of the professionals answering our survey are keeping an open mind with regard to an upper age limit for candidacy.

Of interest are the approaches highlighted in ophthalmologic studies attempting to maximize secondary plasticity for late acquisition of visual capabilities. It is now standard practice to penalize the better seeing, dominant eye in children with amblyopia, either by patching or pharmacologically, to promote visual integration of the weaker eye.[A45] Studies treating older

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amblyopic children and even adults also use patching of the dominant eye to achieve some visual acuity gains. Maurer and Lewis have shown that even short-term early monocular deprivation in infancy can lead to permanent ocular dominance of the unaffected eye similar to the first ear dominance seen in children with long-interval sequential BCI.[A50] They have shown that the adverse effects of the uneven ocular competition may not show up until later in the child's life when accumulated competitive interactions have begun to limit the recovery from deprivation. Of note is that in their study the ultimate visual acuity of the deprived eye was directly related to the number of hours each day of patching the dominant eye.

No consensus currently exists concerning the optimal rehabilitation techniques to use for the second ear of late, sequential BCI children. Complete auditory deprivation of the dominant, first-implanted ear for an extended period presents significant practical challenges for older children who are very dependent on the use of their best hearing in day-to-day activities. Such an approach is likely to have a low compliance rate. Yet based on findings in the treatment of amblyopia, it is possible that a rehabilitation technique of this type will in the future be shown capable of improving outcomes. Further research is needed in this area. In addition, pharmacologic manipulation to reopen or release the neural mechanisms at work during the critical period may dramatically enhance the hearing potential of patients of all ages with congenital hearing loss who present at a late age for either first or second CI.[A51]

ADULTS

There is higher representation of children among BCI patients worldwide than adults, despite the fact that more adults have CIs overall. This may indicate significant under-representation of adults in the BCI patient population. In addition, our survey seems to suggest decreased availability of information and opportunity for adults with regard to counseling about BCI. Postlingually deafened adults appear to be the candidates who can achieve the greatest quantitative binaural performance with BCI of all candidate groups.[A52–A61] For these adult candidates, the issue of auditory deprivation has lesser influence upon outcomes than it does in children, their deafness having occurred after the CAS had fully developed. However, concerns have been raised even in adults about the negative impact of long periods (20–30 years or more) of auditory deprivation in either ear upon outcomes with a CI, even when the onset of hearing loss was after CAS maturation. Zeitler et al. (2008) addressed this factor in their study and found no association in 22 sequential BCI adults between performance and the time between implants, age at second implant, or length of deafness in either ear.[A61]

Objective Measures in Adults

In the presence of a fully developed (auditorially mature) CAS, BCI has been shown capable of providing an individual with the majority of the binaural hearing mechanisms that are currently known, despite the psychoacoustic limitations of current processor arrangements and strategies. Schleich et al. (2004) studied 21 adult BCI patients (10 females, 11 males, ages 17.5–66.5 years, mean, 44 years).[A57] Duration of deafness ranged from 0.6 to 47.8 years (mean, 12.9 years). Speech reception thresholds at which a 50% correct score was achieved were measured using the Oldenburg sentence test. Speech was presented from the front, with noise either from the front, the right side, or the left side (at $\pm 90^\circ$). Performance was measured using an adaptive signal-to-noise procedure unilaterally and bilaterally. Results showed a significant head shadow effect (using Wilcoxin signed ranks tests at $P < .05$; calculated as the unilateral ear speech reception threshold [SRT] with noise to the ipsilateral side minus the score with noise to the contralateral side). There was also a significant binaural summation effect (calculated as the unilateral implant SRT, minus the binaural SRT with noise and speech originating from the front). There was a mean 6.8 dB improvement binaurally due to the head shadow effect and 2.1 dB for the binaural summation effect. A mean binaural squelch effect (calculated as the SRT when listening unilaterally with contralateral noise minus the score when listening binaurally) of 0.9 dB was also significant. There was no correlation between benefit and duration of deafness.

Litovsky et al. (2006) analyzed the data from a multisite prospective study of 37 postlingually deafened simultaneous BCI adults.[A53] Speech recognition performance using consonant-nucleus-consonant (CNC) and Hearing in Noise Test (HINT) sentences in quiet, and Bamford-Kowal-Bench Speech-in-Noise Tests (BKB-SIN) in noise were measured in unilateral and bilateral listening conditions. Testing was done with both speech and noise from a frontal (0° azimuth) speaker to examine binaural redundancy, and with speech from 0° but noise from $+90^\circ$ or -90° azimuths to examine head shadow and binaural squelch effects. By 6-months post-activation, a significant bilateral advantage was found over either unilateral condition for listening to speech in quiet. For speech in noise, a significant bilateral benefit was demonstrated when subjects were able to take advantage of the head shadow effect (i.e., when the ear opposite the noise was added for bilateral listening). Some subjects also showed evidence of binaural squelch effects (when the ear on the noise side was added for bilateral listening) and binaural redundancy (when the speech and noise were both from the front).

A few studies of BCI adults have shown either minimal or no statistically significant differences between the BCI, bimodal (CI + contralateral hearing aid), or unilateral CI conditions on various speech perception tasks.[A62,A63] Wackym et al. (2007) addressed this variation in benefit seen across BCI studies.[A64] in their study, five listening tasks of increasing difficulty were used to evaluate performance advantages of BCI in seven adult recipients. The greatest binaural benefit was shown when the most difficult listening task was used for testing (HINT sentences at 60 dB SPL in noise of 52 dB SPL). Bilateral benefit could not be shown in the easiest condition (HINT sentences in quiet) due to a ceiling effect for unilateral scores. The authors suggest that the ease or difficulty of the listening tasks used may account for the varying degree of benefit shown in various BCI studies, and that more difficult tasks may be necessary to demonstrate BCI advantages.

Overall, there is strong evidence that sound localization ability in adults is significantly enhanced with two implants compared to one CI alone.[A58–A60] Nopp et al. compared the sound localization accuracy of 20 adult BCI patients and found that accuracy was enhanced by 30% in the bilateral condition compared to the better ear alone. In addition, variability of response was greatly reduced.[A60] It is a consistent finding in the studies mentioned here that adult BCI patients have much better sensitivity to differences in level or amplitude between the ears interaural level difference (ILD) than to differences in timing interaural timing difference (ITD). This is consistent with speech testing in noise on BCI patients, which shows a greater benefit from the head shadow effect (which is more dependent on ILDs) than from binaural squelch (which is more affected by ITDs).[A53] There are also differences across CI stimulation rates. Sensitivity to ITDs is very poor in cochlear implant patients who use rates beyond a few hundred hertz. Binaural summation and central masking effects have been shown, demonstrating that fusion and use of information across bilateral implants does occur.[A61]

Subjective Measures in Adults

Subjective benefit from BCI has been evaluated in adult patients. In the previously described multicenter study of 37 simultaneous BCI adults by Litovsky et al. (2006), Abbreviated Profile of Hearing Aid Benefit (APHAB) questionnaire data was collected on 30 of the patients who had completed a 3-week period during which the patient was allowed to use only one implant on their better-performing ear, followed by a period of BCI use again.[A53] The results indicated that the bilateral users perceived their performance with the bilateral implants as significantly better than with a unilateral implant on the Case of Communication, Background Noise, and Reverberation subscales of the APHAB. There was no significant difference on the Aversiveness of Sounds (AV) subscale. It was also anecdotally noted that many of the subjects were very reluctant to be without their second implant during the deprivation period of 3 weeks, sensing much better performance with two CIs.

In 2006, the first published study intended to directly investigate perceived benefit via formal quality-of-life measures from BCI was done by Summerfield et al.[A65] They recruited 28 postlingually deafened adults from seven British hospitals who had all been successful unilateral implant users for 1 to 6 years and were seeking a contralateral ear implant. Half were randomly assigned to receive a second implant, whereas the other half were required to wait 12 months. Questionnaires used included three condition-specific measures from the Speech, Spatial and Qualities of Hearing Scale Questionnaire, and also the Glasgow Health Status Inventory, the Health Utilities Index Mark III, the Overall Quality of Life (visual-analogue scale), and the EuroQol EQ-5D. Results indicated self-reported improved abilities for bilateral compared to unilateral implants in spatial hearing, quality of hearing, and hearing for speech. Multivariate analyses showed positive changes in quality of life from improved hearing. However, in two out of the 28 patients worsening tinnitus was seen over time. Even though this negative effect was seen in only 7% of the study group, it offset the positive outcomes enough to render the overall benefit of BCI nonsignificant for quality of life measures. The use of such measures gives some indication that prioritization of healthcare funding plays a role in rates of adult BCI, particularly in the United Kingdom.

Different results have since been obtained by Bichey and Miyamoto in 2008.[A66] They identified 23 postlingually deafened adult patients with bilateral cochlear implants. Median age at first implantation was 20 years (range, 5–76 years), and median length of time to the second implantation was 6.3 years (range, 0.9–13.6 years). Patients completed a health utility index survey, which measures change in eight domains of quality of life. A score of 1.0 represents perfect health-related quality of life. Patients rated each domain for the time period just before receiving the first implant, just before receiving the second implant, and at the current time. Each patient had a clinically significant improvement in quality of life after each procedure. The health utility scores averaged 0.33, 0.69, and 0.81 at each succeeding time period ($P = .0001$). Eleven patients specifically noted greater ability to hear in noisy environments after the second procedure. Improvements were also noted in the domains of speech production, emotional well-being, cognition, and pain. The cost-utility of BCI in this cohort of patients was in the range of $23,000 to $24,000 per quality-adjusted life year. The authors conclude that the data from their study lend support to earlier studies supporting cochlear implantation as a compelling candidate for the allocation of limited healthcare resources.

One of the biggest challenges faced in assessing the benefits of BCI, both on performance and quality-of-life measures, is the great diversity of the CI candidate population. Although the patient factors that are currently thought to be predictive for CI performance are useful for patient counseling, they are not totally accurate for predicting the outcome of each individual patient. In addition, developing quality-of-life instruments that accurately measure a patient's perceived benefit in all areas that are important to the patient has proved to be very challenging. Criticisms have been raised in the past that many of these instruments can be inherently inaccurate because they do not always have a patient centered definition of quality of life.

"Many widely used measures are not patient centered because of the ways in which (questionnaire) items were generated, because a questionnaire may restrict a patient's choice, and because of the weighting system used. These limitations compromise their accuracy and usefulness because they do not measure what constitutes quality of life for all patients."[A67] Certainly then, the underrepresentation of adults in the BCI population is not due to a paucity of proven or perceived benefit. As mentioned in the main article, the philosophical leanings of CI professionals and healthcare funding priorities are likely factors. In addition, a greater awareness among professionals and parents of a critical period for binaural integration in children may be mobilizing efforts and a sense of urgency for BCI in children, with the unintended consequence being that the same urgency does not exist or is not necessary in adults.

In the healthcare economic environments that exist worldwide, it has become increasingly important to avoid applying expensive healthcare technologies to individuals who stand to derive little benefit from them. However, denying a CI or BCI to all patients within a certain candidate group based on our current understanding of predictors of benefit or value will undoubtedly deny such to some individuals whose benefit could far exceed expectations. Hopefully, in the future better preoperative predictive measures, and a better understanding of how the combination of patient historical factors predict outcome, will assist us in accurately predicting an individual patient's final benefit from CI and BCI intervention.

CONCLUSION

The worldwide trends in BCI demonstrated by our survey have a strong base of published scientific evidence supporting them, except for the under-representation of adults in the BCI population. The later point highlights the challenges faced by all healthcare systems in prioritizing care based on predicting relative amounts of benefit that may be obtained across diverse patient groups.

APPENDIX II: WORLDWIDE SURVEY—EVIDENCE BASED GUIDELINES FOR BCI

INTRODUCTION: HISTORICAL PERSPECTIVES

As can be seen from our main manuscript, there are trends of increasing application of bilateral cochlear implantation among experienced CI centers suggested by the survey. Also suggested are geographic variations in how bilateral cochlear implantation has been implemented across study centers. Before proposing current clinical guidelines for bilateral cochlear implantation based on best practices among experienced centers and the published research reviewed in Appendix I, it may be instructional to understand some of the historical developments that have affected the expansion of hearing technology, such as cochlear implants, to new patient groups and to bilateral application.

HISTORY OF STAPEDECTOMY

In 1956, Dr. John Shea and his contemporaries successfully resurrected the stapedectomy operation for the treatment of conductive hearing loss caused by otosclerosis, a procedure that had fallen into disfavor in the late 1800s prior to the development of microsurgical techniques. Before 1956, versions of this surgery were felt to have too high a risk of permanent hearing loss. By using a tissue graft to seal the oval window and a custom-made stapes prosthesis, Shea developed this into a safer and highly effective technique. The modern stapedectomy operation quickly replaced its predecessor, the endaural Lempert fenestration procedure, which prior to 1956 was the only reasonably successful surgical treatment in use for this condition.[A68,A69] The superiority of the stapedectomy operation (in terms of achieving greater hearing gain and without alteration of ear canal anatomy) was quickly evident even with anecdotal observation, ushering in the golden age of stapedectomy.[A70] Because rudimentary hearing aid technology during this time left otosclerotic patients who had moderate to severe hearing loss with few effective options, the operation was nothing short of life changing. In terms of the sheer number of patients to whom it has since been applied and the mean gain in hearing typically realized, it has become arguably the most successful hearing restoration surgery developed to date.[A71]

Within only 2 to 3 years of its reintroduction in 1956, the stapedectomy operation was being performed on the second ear of otosclerotic patients as long as they had already achieved a successful outcome on their first ear (personal communication with Dr. John Shea). This was testimony to the procedure's efficacy, becausee prior to this time no other elective hearing restoration surgery of this magnitude was considered for bilateral application. For instance, there are no available reports of the Lempert fenestration operation having ever been performed bilaterally. With its modest hearing gains and the creation of a cavity in need of permanent periodic maintenance, the benefits of a fenestration surgery in the second ear did not seem to outweigh the negatives (personal communication with Drs. John Shea and Marvin Culbertson).

The presumption that significant additional benefit could be obtained from stapedectomy on the second ear was mostly intuitive, being based on patient reports of the persistent impairment from unilateral hearing loss that remained after first ear surgery. Because of a small but real risk of permanent, total sensorineural hearing loss, sequential surgery with an intervening period of 1 year between operations to assure a stable hearing result in the first ear, was considered wisest and became common practice (personal communication with Drs. John Shea and Marvin Culbertson). Initially the additional efficacy realized after restoring hearing to the second ear was assessed primarily from subjective patient feedback, because tests of binaural hearing benefit were undeveloped. These patients, whose lives had been so positively impacted by restoration of hearing in their first ear, reported substantial additional improvements in day-to-day auditory functioning after successful second-ear surgery. Dr. Shea applied a 60/40 anecdote, estimating 60% restoration of auditory functioning from doing the first ear, and another 40% from doing the second ear (personal communication with Dr. John Shea).

DEVELOPMENTS IN HEALTHCARE FUNDING

Extrapolation of a unilateral procedure such as stapedectomy to bilateral application, and doing so based primarily on its success in unilateral application and positive patient reports of bilateral benefit, was made easy at least in the United States by the healthcare environment in the late 1950s and early 1960s. Although employer-based private health insurance in the United States was playing an increasing role in healthcare funding (covering only 9.1% of the population in 1940, increasing to 67.8% by 1960), it did not exercise any significant oversight concerning medical practice decisions.[A72] Insurers existed to fund healthcare and left decisions about complex medical interventions to individual physicians. Medical treatment decisions were the sole purview of each individual physician, often being based primarily on the sum of their own anecdotal clinical experiences and that of their colleagues. The working philosophy was, "If it might help and won't hurt, do it."

The healthcare environment in the United States and worldwide, into which bilateral cochlear implantation has been introduced, has changed dramatically since the advent of stapedectomy. These changes have mostly been related to the admirable but somewhat contradictory goals of improving access, improving quality, supporting the development of new technology, and yet controlling costs.[A72] Within the United States, the increasing focus among insurers, public officials, and employers on healthcare cost containment, began in the late 1960s.[A72] In 1965, Medicare and Medicaid were created by amendments to the Social Security Act, dramatically increasing access to healthcare and the federal government's role in funding.[A73] Between 1965 and 1975, federal healthcare spending rose from less than \$10 billion to more than \$40 billion.[A73] A major cost containment development in the future of United States healthcare occurred in 1973, when the federal government passed the Health Maintenance Organization Act, encouraging rapid growth of private managed care insurance plans with the intent of bringing healthcare inflation under control. Such plans have since become ubiquitous in the United States, now enrolling 90% of privately insured Americans.[A74]

MEDICAL NECESSITY

In the late 1960s, insurers introduced the concept of medical necessity, intended to limit the coverage they provide to include only essential or necessary services.[A75,A76] Other countries have developed similar medical and legal rules concerning coverage for medical services. Services deemed experimental or unnecessary were excluded from coverage. Initially the determination of which services were medically necessary was still left to individual physicians, as had always been the case up to that point in time.[A77] Eventually most insurers operationalized the term, reserving the right to make their own determinations of necessity or by developing guidelines that dictated to physicians which services were necessary and which were not.

The rationale behind the development of rules of medical necessity is understandable, being rooted in the soaring cost of healthcare and the unjustifiable variations in medical treatment for the same conditions seen among physicians. However, almost since its introduction, the term has been notoriously difficult to define and fraught with controversy.[A76] The definition often varied from one payer to another and exclusions frequently seemed arbitrary.

> "Using the concept of medical necessity as the contractual cornerstone of health care benefits creates problems that go far deeper than the familiar complaints about outsiders' meddling in medicine. Among those problems are medical necessity's legal vagueness, clinical artificiality, and its unreliability and restrictiveness for consumers…. Most medical decisions do not post clear choices of life versus death, nor juxtapose complete cures against pure quackery. Rather, the daily stuff of medicine is a continuum requiring a constant weighing of uncertainties and values…. Choices in this realm require a level of clinical complexity that is not reflected in simplistic notions like "necessity," and that should not be hidden under blanket categories connoting a façade of precision. To presume that a medical intervention is objectively either necessary or unnecessary belies the legitimacy of such variation in human goals and values."[A77]

The rule of medical necessity greatly complicated and often delayed the introduction of and access to new, potentially beneficial healthcare technology, particularly those with regard to non–life threatening impairments. Such limits have been particularly challenging at a time when technology cycles are becoming shorter.

> "Criticism of current definitions of medical necessity is vast and has led to more detailed definitions that are much easier for a layperson to understand. The problem with comprehensive and precise definitions is that they often exclude persons and ailments that normally would be covered under a less precise definition. Part of the problem with precise definitions is that they do not account for technological advances. A procedure can go from experimental to widely accepted very rapidly and a rigid definition will not adapt for this change leading to inappropriate exclusions of payment."[A78]

EVIDENCE-BASED MEDICINE

Another significant, parallel development affecting the adoption of new healthcare technology involves the evolution of evidence-based medicine (EBM), more specifically its use in funding decisions. Beginning with the publication of *Effectiveness and Efficiency: Random Reflections on Health Services* (1972) by the Scottish epidemiologist Professor Archie Cochrane, there has been a steady movement toward the application of scientific principles in assessing the quality of evidence concerning the risks versus benefits of various treatments used in medical practice. The Centre for Evidence-Based Medicine defines EBM as "the conscientious, explicit, and judicious use of current best evidence in making decisions about the care of individual patients."[A79]

EBM categorizes and ranks clinical evidence based on its strength and freedom from bias.[A80] For instance, medical treatments based on randomized, double-blind, placebo-controlled trials involving a homogeneous patient population and medical condition are considered to be based on the strongest evidence. A medical expert's opinion based on clinical experience is now considered to be one of the weakest forms of evidence. Systems have been developed that stratify the quality of evidence between these two extremes.

The primary intent of EBM is the improvement of quality in patient care. Interest in EBM has grown exponentially since the term was coined in the early 1990s.There was only one MEDLINE citation using the term in 1992, but 2,957 citations by February 2000. The more structured and critical approach to medical research and its clinical application outlined by EBM principles appears to improve the efficacy of healthcare, at least at the organizational level.[A81] However, many criticisms and purported limitations have also been voiced.[A82–A84] Some of these criticisms are misperceptions or misapplications of EBM principles, or are instead limitations universal to the practice of medicine itself. For instance, one misconception is that EBM denigrates clinical experience (such as that used at the onset of the stapedectomy era) in favor of research data.[A85,A86]

"Good doctors use both individual clinical expertise and the best available external evidence, and neither alone is enough. Without clinical expertise, practice risks becoming tyrannized by evidence, for even excellent external evidence may be inapplicable to or inappropriate for an individual patient. Without current best evidence, practice risks becoming rapidly out of date, to the detriment of patients."[A87]

EBM does not necessarily invalidate the motto, "If it might help and won't hurt that particular patient, do it." Neither does EBM impose upon clinicians a demand that until a treatment is supported by a randomized clinical trial (RCT), then do not do it.[A88] Instead it recognizes that,

"Some questions about therapy do not require randomized trials (successful interventions for otherwise fatal conditions) or cannot wait for the trials to be conducted. And if no randomized trial has been carried out for our patient's predicament, we must follow the trail to the next best external evidence and work from there."[A87]

"The final misperception is that only randomized trials or systematic reviews constitute the evidence in evidence-based medicine. Even the most vehement protagonist of evidence-based medicine would acknowledge that several sources of evidence may inform clinical decision making. However, the practice of evidence-based medicine stresses finding the best available evidence to answer a question, and hierarchies of evidence have been developed to help describe the quality of evidence that may be found to answer various questions."[A82]

This is an important point, because randomization of subjects is rarely done in surgical research where there is typically a strong preference for one arm of the study by either the patient or the investigator, or where randomization may be viewed as unethical.[A89] In low-incidence conditions where there is a high degree of individual variation among subjects (such as with hearing loss) it can be difficult to demonstrate a treatment effect in an RCT. Although RCTs are commonly used in pharmacologic research, they are not considered always feasible, ethical, or even the best design when studying cochlear implantation.

With regard to BCI studies in children, maintenance of proper control groups has been particularly problematic in even well-designed longitudinal studies. Once positive outcomes were seen within pediatric BCI studies and an age-related, neurodevelopmental effect became more apparent, parents tended to select out of the unilateral control group to have a second implant for their child.[A90] In addition to these study design challenges, it must be kept in mind that many aspects of medical care depend on individual factors such as quality and value-of-life judgments, which are only partially subject to scientific methods.

A relatively recent consequence of EBM that was originally unforeseen and unintended is its use and/or misuse in healthcare funding decisions as an extension of medical necessity. EBM is at its core cost indifferent, seeking primarily to apply the most efficacious treatments to maximize the quantity and quality of life of the individual patient. Yet healthcare expenditures in the United States, per capita, have more than doubled over the past two decades, and more than one third of this increase is due to an increase in the intensity of care.[A91,A92] Thus, it is not surprising that payers are increasingly focused on limiting the integration of new, high-tech treatments, especially those whose primary benefit is improved quality of life, rather than quantity.[A93] Such use has led to the criticism of EBM that it is the cause for the curtailing of healthcare services and freedom of clinical decision making.[A94]

"Some fear that evidence based medicine will be hijacked by purchasers and managers to cut the costs of health care. This would not only be a misuse of evidence based medicine but suggests a fundamental misunderstanding of its financial consequences. Doctors practicing evidence based medicine will identify and apply the most efficacious interventions to maximize the quality and quantity of life for individual patients; this may raise rather than lower the cost of their care."[A87]

CURRENT EBM APPROACH TO BCI CANDIDACY

As discussed above, clinical guidelines based on EBM should not tyrannize healthcare decisions at the level of the individual patient, nor should they be used

Table A1: Bilateral Cochlear Implant Candidacy: Children.[1]

Care of the child who is a cochlear implant (CI) candidate should include a strategy for the treatment of hearing loss in both ears. Guidelines for simultaneous and sequential implantation are listed below.[2]

SIMULTANEOUS IMPLANTATION

Unimplanted children are best considered for a simultaneous approach when they meet the following criteria:

- Bilateral hearing loss in the "profound" audiometric range of CI candidacy for both ears.[3]
- ∼6 to ∼36 months of age.[4]
- Normal labyrinthine (cochlear and vestibular), IAC, mastoid, middle ear, and ear canal anatomy.[5]
- Recent history of meningitis with otologic involvement.[5]
- No active inflammatory middle ear or mastoid disease.[6]
- No medical conditions that significantly increase surgical risk or coexisting conditions that may influence CI benefit (auditory neuropathy, cognitive or neurologic deficit, sensory processing abnormalities, etc).[4]
- Parent perception of little to no useful hearing in either ear with a hearing aid.[7]
- Parent comfort with a comprehensive CI treatment approach and the potential loss of all residual hearing.[7]

SEQUENTIAL IMPLANTATION[8]

A. Criteria for unimplanted children

Unimplanted children may be considered for a sequential approach when they do not meet criteria for simultaneous implantation because any of the following conditions exist:

- Question about the usefulness of residual hearing in either ear, (implant worst hearing ear first.)
- Older age at presentation (>∼36 months) with less predictable CI outcome, (implant ear with better hearing history/predictors first).
- Progressive hearing loss, (implant worst hearing ear first if similar hearing history/predictors in each ear).[9]
- History of vestibular disturbance that raises concern about the effect of CI on vestibular function and/or symptoms.[10]
- Presence of abnormal labyrinthine or altered mastoid/middle ear/ear canal anatomy in either ear requiring special surgical techniques for implantation, (implant best anatomically developed ear first if equal hearing in each ear).
- Concern about the influence of coexisting medical/developmental conditions (including auditory neuropathy, neurologic conditions, and sensory processing disorders) on CI benefit or that increase the risk of surgery.
- Parent perception that useful hearing exists in either ear with a hearing aid.
- Parent reluctance to sacrifice hearing in both ears simultaneously.

B. Criteria for children already unilaterally implanted

Implantation of the second ear is appropriate when the following criteria are met:

- In addition to the second ear meeting CI candidacy criteria, there should be little or no measurable binaural advantage on age appropriate speech perception measures in the bimodal condition (CI + contralateral HA) compared to the CI alone. Tests should be selected to avoid ceiling and floor effects.[11]
- In the event of continued hearing aid use in the contralateral ear, Cortical auditory evoked potential testing (CAEP) fails to show normalization of latencies for that ear (investigational).[12]
- Although good function of the first CI is preferred, implantation of the second ear can be considered in the event of less than expected first CI performance if there is hope of "capturing" a better-performing ear.[13]
- Parent/patient perception of little or no added benefit from using a HA in the unimplanted ear in conjunction with CI use in the opposite ear.[14]
- Parent/patient comfort with loss of residual hearing in the second ear.
- Parent/patient acceptance of a potentially difficult age-related adjustment period and limit to benefit, and the need to reinstitute AV therapy services.[15]

(endnotes appear on opposite side)

Endnotes

1. These guidelines are based on the bilateral cochlear implant experience of the lead author (150 BCI patients as of December 2008), worldwide CI survey results, and data published to date. Because of the variability that exists in the hearing-impaired population, these criteria should be used only as guidelines until each center can develop a base of experience of its own. The intent here is to highlight points for discussion and consideration.

2. The provision of binaural hearing as a standard of care can now be argued to include cochlear implant treatment, just as it exists for hearing aids and other ear and hearing related interventions. However, as is true for all medical interventions, the most conservative treatment that can provide binaural benefit is preferable, which in some cases may be partially provided with bimodal hearing (CI + contralateral HA). Therefore it should not be assumed that just because a patient is a candidate for unilateral implantation, bilateral implantation should automatically follow. At a minimum this stated philosophy encourages planning for the most effective use of both ears instead of limiting the focus to one cochlear implant. For the majority of patients, this approach will lead to bilateral implantation.

3. Audiometric criteria for cochlear implantation in children include bilateral hearing loss in the severe to profound range. This broad category of hearing loss is by itself inadequate to describe the numerous components of auditory functioning that must be considered in a CI evaluation—particularly now that bilateral implantation is an option. In some cases, residual hearing may be present in one ear that, although inadequate for auditory functioning as the dominant ear, may provide auditory cues that supplement a CI in the contralateral ear and may provide some degree of binaural benefit. This is particularly true if the residual hearing provides useful fundamental low-frequency information. This distinction does not inhibit proceeding with unilateral implantation but should be kept in mind when selecting candidates for simultaneous bilateral implantation or choosing which ear to use for unilateral implantation.

4. The ideal age for implantation in profoundly deaf children is as young as safely possible—currently with published reports down to 6 months of age. The upper age limit beyond which permanent delays may result from late implantation has not been definitively established and most likely varies from one child to another. However data suggests that the longer past 3 years of age the first CI takes place the less predictable the response to and outcome from implantation. Therefore, when such concerns exist in late-presenting children, it seems appropriate to use a sequential approach. This is also true in the case of coexisting neuro/developmental abnormalities, including auditory neuropathy, where outcomes are also less predictable.

5. Congenital ear malformations can require special surgical approaches for implantation and add a degree of unpredictability to outcome from a CI. A sequential approach allows an assessment of healing and benefit from one device under these atypical circumstances before proceeding with second-ear implantation. A history of meningitis with otologic involvement, on the other hand, is a strong indicator for simultaneous implantation due to the fear of impending intracochlear fibrosis and/or ossification. This is of course considering that the child has no other significant neurologic sequelae as a result of the meningitis and that the meningitis was not related to a major cochlear malformation in one or both ears. If either of these circumstances exists, treatment must be individualized.

6. Simultaneous implantation places two devices at risk during the healing phase compared to one device in unilateral implantation. This is an even greater reason that neither ear suffer from uncontrolled inflammatory disease if simultaneous implantation is considered.

7. The parents' perception of useful residual hearing in their child's ear(s) should be respected and considered, regardless of audiometric results. Current measures of auditory performance are "laboratory based" and do not tell the full story of real world functioning and benefit. Appropriate decision-making must consider a parent's comfort or reluctance with regard to the potential loss of all residual natural hearing and the simultaneous commitment of both ears to lifelong cochlear implant technology. Sequential implantation allows for a stepwise commitment and may be more appropriate for parents who do not have an "aggressive" mindset.

8. Sequential bilateral implantation may be either a planned approach decided from the beginning prior to first-ear implantation or a consideration undertaken in unilaterally implanted children who have had their first device for greatly varying lengths of time. Sequential implantation can bring into play circumstances not encountered in first ear implantation, such as second ear "critical period" effects and varying degrees of first CI performance.

9. Progressive or fluctuating hearing loss in children can present unique circumstances compared to congenital profound hearing loss. Depending on age at which the hearing deteriorates to CI criteria, decisions may be more similar to those for postlingual onset deafness. One or both ears may have significant auditory experience. There may be different rates of hearing progression in each ear, and therefore, children whose hearing loss has been monitored may have some residual and possibly fluctuating hearing in one ear or the other.

10. Vestibular disorders in children can exist as part of a complex of sensory processing disorders. The potential effects of implantation should not be overlooked when considering bilateral implantation, especially in such unusual cases as bilateral large vestibular aqueduct syndrome (LVAS) and children who have sustained a "dead ear" from prior surgical intervention.

11. Unilaterally implanted children who are being considered for second ear implantation may provide the opportunity for bimodal testing, especially in older children. Often a child's very profound hearing loss makes detailed testing unnecessary to determine candidacy. In cases of children with some residual hearing in the unimplanted ear, the optimal age-specific testing protocol has yet to be determined. Speech perception in quiet and in noise in both the CI only and bimodal conditions is a minimum.

12. The prime goal of providing binaural hearing to young children at as young an age as possible is the maximally effective "capture" of the theorized "critical period" for central auditory development of both ears. Ideally, we would hope to have measures that can quantify how successful hearing aids and residual hearing are in a particular child at providing enough quality auditory stimuli to promote central development. This is something we currently extrapolate from tests of peripheral auditory sensitivity. CAEP holds promise in this regard.

13. A child who is not performing as well as expected with their first CI presents a challenge. A multidisciplinary team approach is needed to analyze for coexisting factors that may be negatively affecting first CI performance. Depending on these factors a second CI may or may not improve such a child's performance. Decisions to attempt to "capture" a better-performing ear must be individualized.

14. Older children who present for discussion of second ear implantation are able themselves to provide some feedback in regard to the usefulness they perceive of residual hearing in that ear. The child's feedback in addition to the parent's impression of hearing aid usage patterns (which reflect a child's perceived benefit) is an important part of the evaluation process.

15. The older a child is at the time second-ear implantation is being considered, the greater the personal motivation required on the part of the child and parents to make it a successful intervention. In these older children, the second-ear CI performance will typically remain significantly below that of the first ear, and "non-use" is a risk that must be anticipated. Even for a high-performing first CI user, auditory-verbal therapy for integration of the second ear signal is likely to be essential for optimal outcomes. Therapy techniques for overcoming first ear dominance (such as deprivation) and maximizing second ear speech perception performance have yet to be clarified.

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Table A2: Bilateral Cochlear Implant Candidacy: Adults.[1]

Care of the patient who is a cochlear implant (CI) candidate should include a strategy for the treatment of hearing loss in both ears.[2] Guidelines for simultaneous and sequential implantation are listed below.

SIMULTANEOUS IMPLANTATION

Unimplanted adult patients may be considered good candidates for a simultaneous approach when they meet the following criteria:

- Bilateral hearing loss in the "profound" audiometric range of CI candidacy for both ears.[3]
- Postlingual onset of hearing loss in both ears.[4]
- Duration of profound hearing loss $<\sim$30 years in both ears.[5]
- Normal labyrinthine (cochlear and vestibular) anatomy.[6]
- No active inflammatory middle ear or mastoid disease or history of canal wall down mastoid surgery in either ear.
- History of recent meningitis with otologic involvement.
- No history of significant vestibular disorders.[7]
- No medical conditions that significantly increase surgical risk or coexisting conditions that may influence CI benefit (such as neurologic disorders).
- Patient perception of little to no useful hearing in either ear with a hearing aid.[8]
- Patient comfort with a comprehensive CI treatment approach and the potential loss of residual hearing in both ears.[8]

SEQUENTIAL IMPLANTATION[9]

A. Criteria for unimplanted patients

Unimplanted adult patients may be best considered for a sequential approach when they do not meet the guidelines for simultaneous implantation because any of the following conditions exist:

- Hearing loss in the "severe" (as opposed to profound) audiometric range of CI candidacy for either ear. (Implant worst hearing ear first.)[2]
- Prelingual or perilingual onset of hearing loss or long term ($>\sim$30 years) profound deafness in either ear. (Implant ear with best hearing history first.)[10]
- History of vestibular disturbance that raises concern about the effect of CI on vestibular function and/or symptoms.
- Presence of abnormal labyrinthine or altered mastoid/middle ear anatomy in either ear requiring special surgical techniques for implantation.
- Concern about the effect of coexisting medical conditions on CI benefit or that increase the risk of surgery.
- Patient perception that useful hearing exists in either ear with a hearing aid.
- Patient reluctance to sacrifice hearing in both ears simultaneously.

B. Criteria for patients already unilaterally implanted

Implantation of the second ear is appropriate when the following criteria are met:

- In addition to the second ear meeting CI candidacy criteria, there should be little or no measurable binaural advantage demonstrated in the bimodal condition (CI + contralateral HA) compared to the CI alone condition. Tests should be selected to avoid ceiling and floor effects.[11]
- Although good function of the first CI is preferred, implantation of the second ear can be considered in the event of less than expected first CI performance if there is hope of "capturing" a better performing ear.[12]
- Patient perception of little or no added benefit from using a HA in the opposite ear with CI.
- Patient comfort with loss of residual hearing in the second ear.

(endnotes appear on opposite side)

Endnotes

1. These guidelines are based on the bilateral cochlear implant experience of the lead author (150 BCI patients as of December 2008), worldwide CI survey results, and research published to date. Because of the variability that exists in the hearing-impaired population, these criteria should be used only as guidelines until each center can develop a base of experience of its own. The intent here is to highlight points for discussion and consideration.

2. The provision of binaural hearing as a standard of care can now be argued to include cochlear implant treatment, just as it exists for hearing aids and other ear and hearing related interventions. However, as is true for all medical interventions, the most conservative treatment that can provide binaural benefit is preferable, which in some cases may be partially provided with bimodal hearing (CI + contralateral HA). Therefore it should not be assumed that just because a patient is a candidate for unilateral implantation, bilateral implantation should automatically follow. At a minimum this stated philosophy encourages planning for the most effective use of both ears instead of limiting the focus to one cochlear implant. For the majority of patients, this approach will lead to bilateral implantation.

3. Audiometric criteria for cochlear implantation in adults include bilateral hearing loss in the severe to profound range. This broad category of hearing loss is by itself inadequate to describe the numerous components of auditory functioning that must be considered in a CI evaluation—particularly now that bilateral implantation is an option. In some cases, residual hearing may be present in one ear that, although inadequate for auditory functioning as the dominant ear, may provide significant auditory cues that supplement a CI in the contralateral ear and may provide some degree of binaural benefit. This is particularly true if the residual hearing provides useful fundamental low-frequency information. This distinction does not inhibit proceeding with unilateral implantation but should be kept in mind when selecting candidates for simultaneous bilateral implantation or choosing which ear to use for unilateral implantation.

4. One of, if not the most significant predictor of cochlear implant performance in adults is the age of onset of hearing loss. It is widely recognized that patients with adult onset (postlingual, postdevelopmental, and post-educational) hearing loss have the most predictable benefit from cochlear implantation. The prelingual onset of hearing loss as a general rule tends to significantly reduce a patient's CI performance. This predictor does not just apply to the patient as a whole, but also to individual ears in the same patient. A patient may have reached adulthood with hearing function in only one ear—the second ear being "prelingually deafened". Such disparity in the hearing history of a patient's two ears adds to the complexity of bilateral implantation decision making and may make sequential consideration more appropriate.

5. The effect of prolonged deafness and non-use of residual hearing on cochlear implant benefit in postlingually deafened adults is difficult to predict and has not been clarified in the literature. Theoretically, it may be associated with a reduction of spiral ganglion cell populations. Duration of profound deafness in one or both ears longer than 30 years does not preclude bilateral implantation but may be more appropriate for a sequential approach.

6. Abnormal or altered labyrinthine, middle ear, and/or mastoid anatomy often require special surgical techniques for cochlear implantation with a potentially higher incidence of complications and less predictable outcome. A sequential approach allows an assessment of healing and benefit with one device under these atypical circumstances before proceeding with second-ear implantation.

7. The vast majority of adult patients tolerate bilateral implantation with no significant vestibular side effects. However the potential for permanent and severe problems does exist. Theoretically, it seems likely that those with preexisting vestibular conditions would be at higher risk. Meniere's disease, prior vestibular neurectomy, labyrinthectomy, and "dead ear" after stapedectomy are examples of situations for which the potential vestibular impact must be carefully considered.

8. A patient's perception of useful residual hearing in either ear should be respected. Current measures of auditory performance are "laboratory based" and do not tell the full story of real world functioning and benefit in all cases. Appropriate decision-making must consider a patient's comfort or reluctance with regard to loss of residual natural hearing and the simultaneous commitment of both ears to lifelong cochlear implant technology. Sequential implantation allows for a stepwise commitment and may be more appropriate for patients who do not have a "comprehensive" CI treatment mindset.

9. Sequential bilateral implantation may be either a planned approach decided from the beginning prior to first-ear implantation or a consideration undertaken in unilaterally implanted adults who have had their first device for greatly varying lengths of time.

10. Prelingually deafened adults are a unique and challenging group of potential CI candidates. Most that are considered for implantation are oral deaf adults. With reasonable expectations in mind, these individuals can benefit from one CI with improved environmental sound awareness and improved lip-reading. Because of their widely variable outcomes, this is a group of candidates who in most cases may be best served with a sequential approach. This allows assessment of the benefit and satisfaction derived from one implant before deciding that a second implant is a good idea. This principle applies also to patients who have prelingual onset of hearing loss in one ear. There are individual reports of adults who are successful unilateral CI users in a postlingually deafened ear having their second, prelingually deafened ear implanted with some success after a long, difficult adjustment process. Nonetheless, this is a challenging undertaking that should be limited to carefully selected, highly motivated candidates.

11. Evaluation of a unilaterally implanted adult for second-ear implantation involves determining whether the aided hearing in the unimplanted ear is able to make significant binaural contributions to the hearing in the implanted ear. Often a patient's profound hearing loss makes the decision process straightforward. However, when residual hearing is present, the binaural contribution of the unimplanted ear with a hearing aid should be determined. However, definitive test measures and the results on which decisions should be based have not been definitively established.

12. Most postlingually deafened adults receive equal benefit from a cochlear implant in either ear. In some, however, there can be significant differences in performance potential that cannot always be predicted when choosing the first ear for implantation. Some postlingually deafened bilaterally implanted adults have significantly better performance in one ear than in the other. Therefore, if a unilaterally implanted individual's CI performance is below expectations, implanting the second ear with the goal of "capturing" a better performing ear is reasonable in carefully selected patients.

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to limit or restrict the application of a given treatment or its funding when clinical expertise dictates otherwise. This is particularly true with BCI, for which it is not feasible for such guidelines to be comprehensive enough to cover every possible clinical presentation. Of the professional society position statements recommending BCI as accepted medical practice, none propose specific candidacy guidelines and to date there has been no publication detailing such.[A95–A97] The decision to proceed with BCI in an individual patient is at times straightforward, but at other times is a complex, multifactorial process. Any guidelines must account for large variations in performance among patients unique to BCI and must include numerous qualifications. Even with detailed guidelines, the decision process demands strong clinical judgment from treating clinicians.

The assimilated, proposed guidelines for BCI in
TA1TA2 children and adults contained in Tables A1 and A2 are derived from an EBM best practices approach to current research in combination with the practice trends in the field. References include the results of our Worldwide Survey of BCI practices and the literature review in Appendix I. They serve to educate on the factors to consider in decision making, such as:

- Age at presentation
- Residual hearing and anticipated benefit of bimodal hearing (CI + contralateral hearing aid)
- Patient/parent's perception of residual hearing
- Progressive and/or fluctuating hearing loss
- Duration of profound hearing loss
- Central auditory development measures–critical period
- Vestibular history
- Anatomic abnormalities
- Comorbid or complicating conditions
- Patient/parent treatment philosophy–motivation

Center experience and future research will clarify and allow greater specificity as long-term outcomes in various patient groups becomes more available.

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