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Cochlear Implantation in Children with Single-Sided Deafness: Does Aetiology and Duration of Deafness Matter?

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Key Words

Aetiology, deafness · Cochlear implant · Congenital cochlear nerve deficiency · Single-sided deafness

Abstract

For adult patients with single-sided deafness (SSD), treatment with a cochlear implant (CI) is well established as an acceptable and beneficial hearing rehabilitation method administered routinely in clinical practice. In contrast, for children with SSD, CI has been applied less often to date, with the rationale to decide either on a case-by-case basis or under the realm of clinical research. The aim of our clinical study was to evaluate the longitudinal benefits of CI for a group of children diagnosed with SSD and to compare their outcomes with respect to patient characteristics. Evaluating a pool of paediatric SSD patients presenting for possible CI surgery revealed that the primary aetiology of deafness was congenital cochlear nerve deficiency. A subgroup of children meeting the CI candidacy criteria for the affected ear (the majority with acquired hearing loss) were enrolled in the study. Preliminary group results suggest substantial improvements in speech comprehension in noise and in the ability to localise sound, which was demonstrated through objective and subjective assessments after CI treatment for the group, with results varying from patient to patient. Our study shows a trend towards superior outcomes for children with acquired hearing loss and a shorter duration of hearing loss compared to congenitally deafened children who had a longer duration of SSD. This indicates an interactive influence of the age at onset, aetiology and duration of deafness upon the restoration of binaural integration and the overall benefits of sound stimulation to two ears after CI treatment. Continued longitudinal investigation of these children and further studies in larger groups may provide more guidance on the optimal timing of treatment for paediatric patients with acquired and congenital SSD.

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Introduction

Over the last several years, evidence has emerged showing a significant benefit of cochlear implant (CI) surgery in adult patients with single-sided deafness (SSD) and asymmetric hearing loss (AHL). Initially, CI were used to treat intractable tinnitus in patients with SSD, which is the most extreme case of AHL where the poorer ear presents with total deafness while the contralateral ear exhibits normal hearing. Apart from the suppression of tinnitus, many of the tinnitus patients treated with a CI derived additional hearing benefit from binaural hearing [Van de Heyning et al., 2008; Vermeire et al., 2009].

Only recently, CI treatment has been introduced in children with AHL [Hassepas et al., 2013]. AHL or even SSD in children can have a negative impact upon the normal development of the auditory cortex in the young child. Furthermore, the ability to develop and use binaural hearing and its subsequent hearing abilities in daily life can be affected. Especially when entering full-time education, children with SSD display behavioural problems and academic weaknesses, as well as increased needs for speech therapy in comparison to their normal-hearing peers [Lieu et al., 2010, 2012]. A recent review by Kuppler et al. [2013] examined articles from 1986 to 2012 and identified increasing evidence that SSD can be a critical dynamic factor negatively affecting the academic success of children, particularly in the early school years; important factors included specific listening difficulties, such as comprehension in noise, as well as secondary difficulties, such as low self-esteem and increased fatigue [Kuppler et al., 2013].

Results from CI surgery in young children with acquired SSD suggest results comparable to those of adults suffering from acquired SSD [Cadieux et al., 2013; Hassepas et al., 2013; Plontke et al., 2013; Tzifa and Hanvey, 2013; Távora-Vieira and Rajan, 2015]. Evidence of rehabilitation of binaural hearing skills in quiet and noise as well as improvement in the localisation of sounds has been demonstrated. Moreover, CI surgery remains the only treatment option available to individuals with SSD that may enable children to regain or improve binaural hearing [Arndt et al., 2011b; Cadieux et al., 2013; Gordon et al., 2013; Plontke et al., 2013; Tzifa and Hanvey, 2013]. Hassepas et al. [2013] reported improvements in speech comprehension in noise and localisation of sound following CI surgery in 3 children with unilateral hearing loss (UHL). Their preliminary findings prompted us to test their study design in a larger cohort of children with SSD presenting clinically for CI treatment of acquired or congenital hearing loss. A comparison of rehabilitation outcomes with respect to individual patient characteristics, including the duration and aetiology of their hearing loss, is made.

Patients and Methods

The retrospective clinical analysis presented in this paper was conducted in accordance with the guidelines of the Declaration of Helsinki (Washington, World Medical Association, 2002) and to

S. Arndt and S. Prose contributed equally to this work.

Table 1. Demographics of children evaluated for CI candidacy and children meeting CI candidacy

Groups	All SSD children evaluated for CI candidacy (n = 50)			SSD children meeting CI candidacy (n = 32)		
	congenital	perilingual	postlingual	congenital	perilingual	postlingual
Number of children	24	6	20	6	6	20
Mean age at evaluation (range), years	7.2 (1.3–15.6)	7.2 (2.2–7.4)	10.9 (5.3–18)	4.0 (1.0–13.0)	7.2 (2.2–7.4)	10.9 (5.3–18)
Mean duration of deafness (range), years	7.2 (1.3–15.6)	5.9 (0.3–7.4)	4.1 (0.3–10)	4.0 (1.0–13.0)	5.9 (0.3–7.4)	4.1 (0.3–10)
<i>Imaging</i>						
HRCT: IAC normal, MRI: normal	6	6	20	6	6	20
HRCT: IAC normal, MRI: CND (no.)	9	–	–	–	–	–
HRCT: IAC narrow MRI: CND (no.)	5	–	–	–	–	–
<i>Aetiologies</i>						
Unknown	5	1	3	2	1	3
CMV infection	4	–	–	4	–	–
Labyrinthitis	–	1	3	–	1	3
Hypoplasia/aplasia cochlear nerve (CND)	14	–	–	–	–	–
Premature birth	1	–	2	–	–	2
EVA	–	1	5	–	1	5
Cranio-cerebral injury	–	2	–	–	2	–
Meningitis	–	1	1	–	1	1
Varicella zoster infection	–	–	1	–	–	1
Auditory neuropathy	–	–	1	–	–	1
Sudden hearing loss	–	–	2	–	–	2
Mumps	–	–	3	–	–	3
<i>CI candidacy</i>						
CI feasible, n	6	6	20			

IAC = Internal auditory canal.

ISO 14155 concerning the procedures of clinical studies [Clinical testing of medical products on humans – good clinical practice: part 1 and 2].

All children suffering from AHL (including SSD) receiving pre-examination concerning their suitability for CI treatment at the University Hospital Freiburg during 2008–2014 were also included in this study, including the 3 cases previously reported by Hassepass et al. [2013]. The examination included high-resolution computed tomography (HRCT) of the temporal bone and magnetic resonance imaging (MRI) of the head with contrast enhancement. Audiological testing consisting of pure-tone thresholds (250–8 kHz), evoked auditory brainstem responses and electrocochleography was performed preoperatively to confirm CI candidacy of the dysfunctional ear. In some cases, age and compliance of the child made general anaesthesia necessary for those examinations. During the same anaesthesia, where indicated and required, adenoidectomy, paracentesis and/or insertion of ear grommets were also performed. An additional neuropaediatric examination was initiated if developmental disorders were demonstrated or suspected. Prior to surgery, all families were informed thoroughly about alternative treatment methods, such as bone conduction implants or conventional hearing aids, e.g. CROS (contralateral routing of signals). Furthermore, the necessity for participation in a mandatory rehabilitation phase at the certified Implant Centre in Freiburg afterwards was emphasised during parent counselling, and formal consent of the patients was obtained on behalf of their child.

Objective Evaluation of Speech Audiometry and Localisation. The Oldenburg Sentence Test (OlSa) was used with an adaptive procedure in three test conditions with background noise set at 65

dB SPL: S_0N_0 (noise and speech from front), $S_{NH}N_{SSD}$ (speech from the normal-hearing side, noise from the deaf/implanted side) and $S_{SSD}N_{NH}$ (speech from the deaf/implanted side, noise from the normal-hearing side). Testing was performed unaided (preoperatively) and 12 months after surgery. The test set-up has been described in detail previously by Hassepass et al. [2013] and Arndt et al. [2011b]. For speech perception in noise using the OlSa in an adaptive procedure, a difference of 1 dB or greater ($p < 0.05$) in the speech reception threshold (SRT) L50 is taken to be outside of normal test-retest variation and considered a significant difference [Wagener et al., 1999; Brand et al., 2002; Wagener et al., 2005].

Localisation measurements were performed in a sound field with a frontal semi-circle (2 m in diameter) consisting of seven, equidistant loudspeakers in a horizontal plane at intervals of 30° positioned at the subject's head level, as described by Arndt et al. [2011b]. The speech stimuli, OlSa sentences, were presented at a sound level of 65 dB SPL. The ability to localise was recorded by localisation error in degrees.

Subjective Evaluation. Subjective evaluation was conducted with a child self-report and a parent report using the adapted versions of the Speech, Spatial and Qualities of Hearing Scale (SSQ) by Galvin et al. [2007] and the adult version of the SSQ originally developed by Gatehouse and Noble [2004]. Questionnaires were administered in consideration of the age-appropriate nature of the questionnaires preoperatively and 12 months after CI surgery; for children older than 9 years, only the adult SSQ was used. For analysis, the average subscale scores for the SSQ are calculated per test interval, per questionnaire version per test subject and subgroup, where group size permits. Statistically significant changes between

Table 2. Demographics of the CI-treated children (n = 20)

Onset of deafness	Gender	Duration of deafness, years	Age at implantation, years	Ear	Aetiology	Implant	Available test results (12 months after CI)
<i>Congenital</i>							
Child A	F	4.3	4.3	Right	CMV infection	Nucleus CI24 RE (CA) (Cochlear)	Yes
Child B	F	13.8	13.8	Right	CMV infection	Nucleus CI24 RE (CA) (Cochlear)	Yes
Child C	F	3.2	3.2	Left	CMV infection	HiRes90k HiFocus MS (Advanced Bionics)	Surgery too recent
Child D	F	1.8	1.8	Left	CMV infection	Nucleus CI512 (CA) (Cochlear)	Too young for testing
<i>Perilingual</i>							
Child E	F	7.4	8.6	Left	Meningitis	CONCERTO FLEX ²⁸ (MED-EL)	Yes
Child F	M	7.0	9.6	Right	Unknown, progressive	Nucleus CI422 (Cochlear)	Yes
Child G	M	0.3	2.3	Left	Craniocerebral injury	Nucleus CI24 RE (CA) (Cochlear)	Surgery too recent
<i>Postlingual</i>							
1	F	2.5	6.6	Right	EVA	Nucleus CI24 RE (CA) (Cochlear)	Children's sentence test, omitted
2	F	3.0	8.6	Right	EVA	Nucleus CI24 RE (CA) (Cochlear)	Yes
3	F	0.4	10.2	Left	Labyrinthitis	Nucleus CI512 (CA) (Cochlear)	Yes
4	M	0.8	11.4	Left	Sudden hearing loss	Nucleus CI422 (Cochlear)	Yes
5	F	5.1	14.4	Left	Sudden hearing loss	Nucleus CI422 (Cochlear)	Yes
6	M	3.0	15.7	Right	Premature birth	CONCERTO FLEX ²⁸ (MED-EL)	Yes
7	F	0.8	16.9	Left	Varicella zoster infection	CONCERTO FLEX ²⁸ (MED-EL)	Surgery too recent
8	F	1.0	18.0	Right	EVA	Nucleus CI24 RE (CA) (Cochlear)	Yes
9	F	1.5	11.7	Left	EVA	Nucleus CI512 (CA) (Cochlear)	Yes
10	M	1.2	5.7	Left	Meningitis	Nucleus CI512 (CA) (Cochlear)	Surgery too recent
11	F	0.5	7.0	Left	Unknown, progressive	CONCERTO FLEX ²⁸ (MED-EL)	Surgery too recent
12	F	10.0	11.0	Right	Unknown	Nucleus CI24 RE (CA) (Cochlear)	Yes
13	F	0.3	8.1	Right	Premature birth	Nucleus CI422 (Cochlear)	Yes

EVA = Enlarged vestibular aqueduct.

pre- and post-CI self-evaluations with the SSQ are reported only where subgroup size permits (i.e. postlingual subgroup). Precise methods were described by Hassepass et al. [2013].

Data Analysis. Data were analysed with SAS 9.2. Results are described for each test measure for three subgroups of children divided according to the onset of their hearing loss: congenital, perilingual (2–4 years) and postlingual (>4 years). The results of each individual of the various test measures were compared between baseline (i.e. prior to CI) and the 12-month follow-up (post-CI). In view of the small group size of the congenitally and perilingually deaf children, statistical analysis of their group data was not possible.

For the postlingual SSD children, normality of pre- and post-operative differences could not be assumed for many of the investigated items; therefore, outcomes with the CI were compared to preoperative performance using Wilcoxon's signed-rank test for paired observations, and p values were calculated based on the exact distribution of the test statistics.

Results

From a total of 50 children (30 females and 20 males) presenting with SSD at the University Hospital in Freiburg for CI pre-examination between 2008 and 2014, 24 cases presented with right-ear deafness and 26 with left-ear deafness. After extensive workup, 32 children qualified for CI surgery. To date, 20 children have received a CI, and the remaining 10 are scheduled for CI surgery.

The type of CI provided was Cochlear Nucleus [CI512, CI24RE (CA) and CI422; Cochlear Ltd., Sydney, N.S.W., Australia] for 15 children, a CONCERTO FLEX²⁸ from MED-EL (Innsbruck, Austria) for 4 children and a HiRes90k HiFocus MS from Advanced Bionics (Valencia, Calif., USA) for 1 child.

Aetiology and Demographics

From the pool of 50 SSD children evaluated, 24 children had congenital UHL (i.e. onset <2 years), 6 children had perilingual UHL (i.e. onset 2–4 years), and 20 children had postlingual hearing loss (i.e. onset >4 years). In table 1, demographic data and aetiological factors are detailed for all children evaluated for CI candidacy as well those meeting CI candidacy criteria. Table 2 shows the detailed demographic data of the 20 children treated with CI surgery to date subdivided by age at onset.

Congenital Deafness (n = 24). More than 50% (n = 14) of the congenitally deaf children showed aplasia/hypoplasia of the cochlear nerve in MRI; therefore, they did not qualify for CI surgery. All 4 implanted children suffered from cytomegalovirus (CMV) infection.

CI surgery for the oldest child (13.8 years) was performed at the express request of the child and the parents due to increased social and school-related distress. After intensive counselling concerning the relatively low expectations of a positive audiological outcome and in light of a positive result in promontory testing, CI candidacy was confirmed for this child.

Fig. 1. Results of OISa for the critical signal/noise ratio in each configuration and localisation error before (unaided) and 12 months after CI surgery (CI) in congenital SSD children: both children suffered from congenital CMV infection; child A was implanted at the age of 4.3 years and child B at 13.8 years. * $p < 0.05$ vs. baseline.

Seven of the 18 non-implanted children received a CROS hearing aid, 2 a Bonebridge (MED-EL), 1 a bone conduction device, 1 an iSense and 1 a Baha Softband. Six children remained unaided.

Perilingual Deafness ($n = 6$). Three children have been treated with a CI, 1 awaits CI surgery, 1 child received a CROS hearing aid and 1 a Baha. The 1st of the implanted children suffered from pneumococcus meningitis, the 2nd from a fracture of the temporal bone and the 3rd from progressive deafness of unknown aetiology (note: classification as a possibly late-discovered, congenital SSD was not entirely eliminated).

Postlingual Deafness ($n = 20$). All of the 20 children with postlingual deafness qualified for CI surgery after the pre-examination. As of the writing of this report, 13 were treated with a CI.

Speech Audiometry and Localisation

Altogether 20 children received CI surgery; however, surgery was too recent for the inclusion of test results for 5 patients. One child was too young for testing on the measures of binaural hearing available, another child could only be tested using a children's sentence test (Oldenburger Sentence Test for Children) and, therefore, their results could not be combined with the group results and were excluded from group data analysis. Repeated-measure analyses of complete data sets were available for 13 implanted children, including 2 of 4 congenitally deaf, 2 of 3 perilingually deaf and 9 of 13 postlingually deaf children.

Congenital Deafness. Complete data sets are available for 2 of 4 children to date. One child was too young to perform the available tests, and the other received surgery within the last year. Both children (A and B) are poor performers in audiological testing. Compared to the unaided situation, child A showed poorer results in S₀N₀ and significantly poorer results in the S_{NH}N_{SSD} condition 12 months after implantation and a minimal non-significant increase in speech comprehension in the S_{SSD}N_{NH} with CI. Testing of child B showed close to equal performance in the S₀N₀ and in the S_{SSD}N_{NH} coincidental speech-in-noise test condition and signifi-

cantly poorer results in the S_{NH}N_{SSD} condition 12 months after CI compared to the preoperative condition.

In terms of localisation abilities, child A showed a substantial increase in the localisation error, and child B showed a substantial decrease in the localisation error 12 months after CI surgery compared to the unaided condition (fig. 1). Child A uses the CI only at school and child B is a non-user.

Perilingual Deafness. Two children have completed evaluations; for the 3rd child the follow-up time is too short for data inclusion. Figure 2 depicts the results of child E, who acquired deafness perilingually due to meningitis; there was a significant improvement in S_{SSD}N_{NH} as well as in S_{NH}N_{SSD}, and a non-significant improvement in S₀N₀ with the CI at the 12-month follow-up compared to the unaided situation. Child F had no improvement in S_{SSD}N_{NH} but significantly better performance in S₀N₀. In S_{NH}N_{SSD}, speech comprehension decreased significantly with the CI compared to the unaided situation. Localisation ability of child E was substantially improved 12 months after CI surgery. The results of child F improved marginally with the CI.

Child E demonstrated overall good results and uses the CI daily, whereas child F uses the CI only at school and, generally, showed poorer results in test outcomes.

Postlingual Deafness. Group results for speech recognition for the postlingual subgroup ($n = 9$) are shown in figure 3. Statistical analysis of group data revealed significant improvements in S_{SSD}N_{NH} ($p = 0.007$). A trend to improved results 12 months after CI surgery ($p = 0.06$) was noted regarding the S₀N₀ condition. The ability to localise increased significantly after CI treatment ($p = 0.0076$).

Speech, Spatial and Qualities of Hearing Scale

The subjective impression of hearing ability was evaluated with age-appropriate versions of the SSQ scale before and 12 months after CI surgery for 11 children: 9 using both the child and parent versions and 2 teenagers received the adult version of the SSQ only. The results of the SSQ assessment for children with congenital and

Fig. 2. Results of OISa for the critical signal/noise ratio in each configuration and localisation error before (unaided) and 12 months after CI surgery (CI) in perilingual SSD children performed at the age of 8.6 years (child E) due to meningitis-induced SSD and at the age of 9.5 years (child F) due to progressive UHL of unknown origin. * $p < 0.05$ vs. baseline.

Fig. 3. Box-and-whisker plots of the critical signal/noise ratio and localisation deviations of the children with postlingual SSD ($n = 9$), with a tendency to significance in S₀N₀. Wilcoxon's signed-rank test.

perilingual SSD are displayed in figure 4. Child A from the congenital subgroup had almost no improvement in all three subcategories after CI surgery compared to the preoperative condition. Parent ratings for child A showed no improvements with the CI, while parent ratings were not determined for child B, a congenitally deaf teenager, who used the adult version of the SSQ. Child B rated her hearing ability essentially equal for the speech subcategory, scored lower for spatial hearing and slightly higher for hearing quality with the CI compared to the unaided situation.

In contrast to the congenitally deaf child ratings, ratings for children with perilingual deafness revealed overall subjective im-

provements after CI surgery for 1 child (child F) and no subjectively perceived improvement for child E in the speech and hearing quality subcategories. Spatial hearing showed essentially equal points after CI surgery.

Examination of parent ratings for the individual children in both congenital and perilingual groups showed overall similar tendencies for improvement and deterioration compared to those reported by their respective children. Closer examination indicates the parents of congenitally deaf child A and perilingually deaf child E rated spatial hearing abilities in the pre-CI unaided condition comparatively poorer than the self-assessment of their child would

Fig. 4. Subjective self-assessment by SSQ for SSD children before (unaided) and 12 months after surgery (CI). **a** Congenital SSD: for child A via child and parent versions of the SSQ, for child B assessment by the adult SSQ alone, due to the age of the subject. **b** Perilingual deafness (child E and F).

suggest. Furthermore, self-assessment of child E reported a decrease in spatial hearing with the CI, while their parents observed an increase. Overall ratings tended to be lower by parent proxies compared to their children's own assessments.

SSQ scores for the postlingual subgroup showed statistically superior results in all three subcategories (fig. 5). The child self-evaluation revealed significant improvements with the CI for speech hearing ($p = 0.027$), spatial hearing ($p = 0.028$), hearing quality ($p = 0.028$) and the total scores ($p = 0.028$). Parent proxy evaluations also showed significantly better scoring in all three subcategories: speech hearing ($p = 0.028$), spatial hearing ($p = 0.028$), hearing quality ($p = 0.028$) and for the total rating ($p = 0.028$). In contrast to the parent proxy ratings for patients with congenital and perilingual deafness, on average the parent proxy ratings for children with postlingual SSD tended to be higher compared to the child self-ratings with the CI.

The older teenagers with postlingual deafness completing the adult version of the SSQ demonstrated improvements with the CI in all three subcategories (speech hearing from 5.7 to 7.7, spatial hearing 2.9 to 5.9 and quality from 5.3 to 9.1).

Discussion

There is still ongoing discussion on the topic of SSD in children and the suitability of CI surgery as a treatment option. To date, scientific reports on rehabilitation results with CI in children suffering from SSD are limited to case reports and small cohorts. This report represents the first study in a relatively large cohort of 20 CI-treated SSD children, with 13 already having complete 12-month data. Analysis of aetiology demonstrates a very heterogeneous spectrum of causes for UHL in children that is also dependent on the age at onset of the hearing loss.

Fig. 5. Box-and-whisker plots for the mean values of subjective self-assessments by and for each subsection of the SSQ for postlingual SSD children via child and parent versions of the SSQ before (UA) and 12 months after surgery (CI). * $p < 0.05$ (Wilcoxon's signed rank test).

The most frequent cause of deafness in congenital SSD children was aplasia or hypoplasia of the cochlear nerve. Clemmens et al. [2013] described unilateral cochlear nerve deficiency (CND) as a common cause of SSD. Furthermore, Levi et al. [2013] stated CND as an important factor of SSD aetiology in children. They reported on a total of 18 children with cochlear nerve dysplasia; of 15 patients with unilateral dysplasia, 11 had a severe to profound hearing loss. Both MRI and HRCT are indispensable tools in the pre-examination assessing the patient's suitability for CI surgery, especially to exclude children with hypoplasia/aplasia of the hearing nerve, as an intact cochlear nerve is required for successful hearing rehabilitation after CI surgery. A small inner ear canal may be indicative of CND in HRCT, but might also be of average width [Pagarkar et al., 2011]. In order to be able to differentiate the beneficial information obtained from MRI and HRCT, Pagarkar et al. [2011] suggested a combination of the diagnostic imaging tools with audiological tests for the evaluation of the integrity of the hearing nerve. Bilaterally deaf children with uni- or bilateral cochlear nerve aplasia/hypoplasia showed overall poorer results in comparison to their peers having other malformations, such as an enlarged vestibular aqueduct (EVA) [Buchman et al., 2011; Valero et al., 2012]. Therefore, the authors do not recommend CI surgery in SSD patients with proven CND in view of the anticipated poor outcomes which in turn may negatively impact bilateral hearing compared to the unaided condition.

Another main aetiology for congenital SSD in the present study was congenital CMV infections. There are no publications to our knowledge that report the incidence of CMV infections in SSD patients. In children with bilateral congenital deafness, CMV infection is reported to be a common cause by Deltenre and Van Maldergem [2013] and Yoshida et al. [2009]. Intrauterine CMV infection is estimated to affect approximately 0.5–2.5% of all live births [Hagay et al., 1996]. At this point in time, viral and/or host

inflammatory mechanisms involved in the pathogenesis of auditory dysfunction remain unclear [Cheeran et al., 2009].

In the group of perilingually deafened children, various causes of deafness were found in the aetiology analysis. The exact time of onset of deafness remains often vague – children with UHL regularly fail to provide clear evidence of the presence and subsequent impact of their deafness, which is in contrast to bilaterally deaf children. As long as there is no certain proof of perilingual hearing loss (for example, craniocerebral injury or meningitis), it might prove more useful to classify and treat a child as having a congenital hearing loss in order to provide ideal treatment in the case of suspected UHL during perilingual development.

In the present study, the major aetiological factor associated with postlingual SSD in children was EVA ($n = 4$), whereas the most common causes for SSD in adults have been reported to be sudden hearing loss and labyrinthitis [Arndt et al., 2011b].

Not only aetiological factors, but also the onset of deafness should be taken into consideration to estimate the audiological outcome of CI treatment in SSD children. Both children suffering from congenital SSD in the present study showed poor results in speech audiometry. In almost all tested speech-in-noise situations, results were inferior compared to their preoperative situation in both. In addition, localisation ability improved only slightly for child B after CI while child A showed even deterioration.

To date, a single publication reports on CI outcomes in children with SSD: 3 were congenitally deaf with unknown aetiology and 1 had postlingual SSD due to meningitis [Távora-Vieira and Rajan, 2015]. Both congenitally deaf children with measurable results were implanted after 4.5 and 6.8 years of deafness. Not only did they demonstrate poor results in audiological testing, one of them was also a non-user [Távora-Vieira and Rajan, 2015]. The poorer audiological results and the duration of deafness of our 2 congenitally deaf children (4.3 and 13.8 years old) are comparable

to the results described by these authors. In the presented study, 1 of 2 children with congenital SSD is a non-user while the other only wears the CI at school.

In contrast to these findings, our youngest child with congenital SSD who received the CI at 1.8 years, as well as the youngest child (aged 14 months at implantation) of Távora-Vieira and Rajan [2015], exhibits clinical evidence of binaural integration through behavioural responses to sounds. Moreover, both children wear their CI all the time (both were too young for audiological testing).

Summarising the results of Távora-Vieira and Rajan [2015] and the results from our children with congenital SSD, congenital SSD children older than 4 years have shown to be poor performers using speech audiometry and sound localisation. According to current clinical evidence, albeit obtained from a small number of congenitally deaf children, CI intervention should take place before the age of 4 years. One reason for the poor outcome in older congenital SSD children might be the inability to exploit the critical sensitive period for the development of binaural auditory skills, as described by Gordon et al. [2013]. For the young bilaterally deaf child, this critical window is projected to be before the age of 4 years with the proviso that one implant is obtained before the age of 2 years. If this also applies for the congenital SSD child the recommended period for implantation to potentially optimise outcomes for binaural hearing development is likely to be substantially shorter than the age of 4 years.

In cats with UHL, Kral et al. [2013] described that if the onset of unilateral hearing is early (before or around the peak of functional synaptogenesis) than massive reorganization of aural preference in favour of the hearing ear was found. The effect was reduced if the onset of unilateral hearing was in the intermediate period, and it disappeared if the onset was late. In early-onset UHL, the better-hearing ear became functionally dominant. This might explain the inferior outcome of CI for the late-implanted congenitally deaf ear in children. They recommended that periods of monaural stimulation should be kept as short as possible, and training of the CI ear should commence after implantation [Kral et al., 2013]. Comparable results were also reported by Grothe et al. [2010] in mammals. They explained that in the case of implantation after a critical period, the children might permanently lose their opportunity to attain bilateral integration of hearing benefits. It remains to be determined if possibly the windows of sensitive periods of central auditory development might be wider or narrower for congenital SSD than for binaural profound hearing loss.

Besides the timing of implantation, as a matter of fact, the aetiology present may play a central role in the outcome of cochlear implantation. Moreover, another reason for the poor audiological results of our congenitally deaf children might be the congenital CMV infection, since other authors, e.g. Yoshida et al. [2009], reported inferior outcomes of CI surgery in bilaterally deaf children due to congenital CMV infection compared to children with non-CMV-related deafness. In agreement, Viccaro et al. [2012] described long-term results for CI surgery in 6 children with congenital CMV-related deafness; only 2 of them showed appropriate use of grammar and syntax, the other 4 used only single words. They assumed that the persisting performance gap between the CMV children and non-CMV peers was associated with effects on the central nervous system as a result of CMV infection [Viccaro et al., 2012].

In the group of children with perilingual deafness, the results were rather different among the children. The child with acquired deafness due to meningitis presented overall good results and uses the CI daily. The second child uses the CI exclusively at school – all in all, the child also showed poorer results in testing, which were comparable to the results of children with congenital disease; this child was implanted at the age of 9.5 years for undefined, progressive hearing loss. Because of the progression and nature of the hearing loss, it is not entirely clear if the hearing loss was indeed perilingual or rather a late-discovered congenital SSD.

It remains to be seen if the age at the onset of deafness and the age of implantation are also important factors for the children with perilingual deafness, and, therefore, if CI should be performed within a small time window after the onset of deafness.

In the present evaluations, most children with postlingual SSD demonstrated substantial improvements in all test configurations for speech audiometry and localisation with the CI compared to the unaided condition. The results for speech comprehension in noise and sound localisation are comparable to those of adult CI patients with SSD.

Apart from the study of 3 children by Hassepass et al. [2013] (all 3 children are included in this study), there are, to our knowledge, only case reports for acquired SSD in children, such as the study by Plontke et al. [2013]. In SSD adults, several studies demonstrated improvements in qualities supporting higher-level processing and integration of both acoustic and electrical signals leading to improved overall hearing. The improvements in performance reported for younger CI patients with acquired SSD suggest that results were equivalent to those seen for SSD adults and show evidence of rehabilitation of binaural hearing and similar improvement in the quality of speech comprehension in noise and the ability for sound localisation [Arndt et al., 2011a, b; Punte et al., 2011; Stelzig et al., 2011; Firszt et al., 2012; Plontke et al., 2013].

However, the results of our SSQ reports cannot be compared to those of other SSD children due to the absence of published SSQ data for children. For the congenitally deaf children, no or only slightly increased improvements in the SSQ were noted after CI surgery. These results are consistent with the objective audiological assessments for these children demonstrating no or only minimal improvement in any test condition. It is further corroborated by the daily CI use reported, with one child being a non-user and the other a part-time user only.

While objective outcomes did not always substantiate subjective ratings for perilingual SSD children, a tendency to a better outcome was noted in those who use their CI more consistently.

For the postlingual SSD child group, agreement between subjective and objective assessments was good, with all subjects reporting daily use of their device. In conclusion, daily use is a simple yet reasonably accurate indication for the success of hearing treatment.

In comparison, subjective assessment via the SSQ in implanted SSD adults with acquired hearing loss, as reported by Arndt et al. [2011b], demonstrated a significant improvement in speech and spatial hearing. In contrast, no changes were observed for the quality of hearing after CI surgery. In 2013, Hassepass et al. demonstrated improvements in the perceived hearing ability in all subsections of the SSQ after CI surgery compared to before. The present study confirms the improvement in subjectively perceived hearing for all three SSQ subcategories for children with postlingual SSD treated with CI surgery.

As parent proxy ratings of their postlingually deaf child's post-CI hearing ability via the SSQ were comparatively higher than those of their child, proxy ratings should be interpreted with caution due to inherent emotional bias of proxy judgement.

In view of the small number of children enrolled and evaluated to date, caution should be applied regarding the interpretation of the present study results on CI surgery in congenitally and perilingually deaf children and the long-term treatment effects. It also remains to be seen if the benefits of treatment increase with follow-up duration and intensive counselling to encourage more consistent device use despite poor initial outcomes. However, to sum up the collective findings of the 4 children with congenital SSD from our study and those from Távora-Vieira and Rajan [2015], it seems to be important that these children are diagnosed and treated with CI as early as possible. Results might be limited if the children are not implanted within a certain time window. To be able to define this critical window, a larger group of patients has to be studied.

In conclusion, the selection criteria for CI in SSD children requires further refinement regarding the aetiology, duration and onset of deafness on a case-by-case basis while taking the anticipated results into consideration.

Conclusion

Irrespective of age, MRI of the head is an essential prerequisite before CI surgery to exclude aplasia or hypoplasia of the hearing nerve. More than 50% of congenital SSD children suffer from CND. The results also indicate that the duration of deafness is an important determinant of CI treatment outcome, especially in the group of children with congenital disease: the earlier the cochlear implantation, the less cortical remodelling occurs.

The recommended period for implantation in congenital and perilingual SSD children to enable rehabilitation of binaural hearing skills has yet to be determined and is likely to be shorter than for children presenting with early-onset bilateral deafness.

For children with postlingual SSD, CI surgery presents the only opportunity to restore binaural hearing abilities.

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