Longitudinal linguistic outcomes of toddlers with congenital single-sided deafness-Six with and twelve without cochlear implant and nineteen normal hearing peers

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Longitudinal linguistic outcomes of toddlers with congenital single sided deafness – six with and twelve without cochlear implant and nineteen normal hearing peers

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Introduction

Each year in Flanders about 20-25 newborns are diagnosed with profound (> 90 dB HL) sensorineural hearing loss on one side and normal hearing contralaterally[1] also termed congenital single sided deafness (cSSD). In Flanders, as in many other parts of the world, there is no standard care for these children. It is, however, widely acknowledged that their ability to localize sound sources and to understand speech in noisy situations is hampered[2,3] due to absent binaural hearing. Moreover, at group level SSD has been shown to negatively affect language and cognitive development and to increase listening effort[4–9]. These results indicate that intervention should be considered.

Untreated cSSD leads to cortical reorganization that continues with increasing duration of SSD[10]. As duration of SSD is negatively associated with outcomes after intervention[11], it is advised that treatment is provided within this early critical period. This is to prevent overrepresentation of the hearing ear in the auditory system and biased input to higher order cortical areas, and to possibly restore cortical organization.

A cochlear implant (CI) is the only rehabilitative option that offers the potential to facilitate binaural hearing, as it enables sound transmission via electrical stimulation of the auditory nerve on the impaired side. In Flanders, the number of newborns with cSSD that qualify for a CI, depending on etiology, is estimated to be 5 to 10 each year[12]. Currently, it is unknown if early CI in cSSD will yield similar results as CI in a maturated auditory system[13]. First results of early implanted children with cSSD are promising[14–17].

To date, the benefit of a CI for children with SSD has only been reported for auditory skills and subjective experience. It is equally important, however, to document the benefit of CI for these children with regard to the development of language and complex cognition, given the reported significant differences to NH peers in this regard. The aim of our multicenter longitudinal study is to investigate the potential benefit of a CI in 16 children with cSSD, implanted between 8-26 months of age, with regard to language, cognition and auditory performance. It is hypothesized that provision of the CI at a very young age will partially restore binaural processing in the following years and hence yield the best conditions for near-normal auditory, linguistic and cognitive development. Although the CI is provided at a very young age, potential improvements are expected to be much more subtle than for bilateral deaf children.

Here, we present data of the first 6 implanted children who currently are 2 years of age or older (group SSD_CI). Performance is compared to that of two age-matched control groups: toddlers with SSD who did not receive a CI (group SSD_noCI, n=12) and normal hearing peers (group NH, n=19).

Methods

Ethical Considerations

The study was approved by the medical ethical committee of every participating center (B322201523727).

Participants

Characteristics of the toddlers with SSD are presented in table 1. Their auditory brain stem thresholds (air conduction) are ≥ 80 dBnHL on the affected side and ≤ 35 dBnHL on the contralateral side. With the exception of one child, none of the children suffer from comorbidities. Parents were thoroughly informed and given the current scientific knowledge about outcomes and possible risks and benefits. About a third of the initially counseled parents declined implantation.

Outcome Variables

CI use is monitored through the data logging software of the device of the SSD_CI children at their mappings sessions.

Language and cognitive performance are assessed twice a year with respectively the Schlichting Receptive test [18] and Expressive Language[19] sub tests expressive vocabulary and morphosyntactic knowledge, and the Bayley-III-NL Scales of Infant and Toddler Development[20] sub scale cognition (up to age 42 months). Testing is done at the children's home, divided over multiple sessions. All tests provide Flemish norm-referenced test scores. For interpretation and comparison purposes, the norm-referenced scores are converted into z-scores (M=0, SD=1).

Parents are asked to complete the Parents' Evaluation of Aural/Oral Performance of Children (PEACH [21] Dutch version).This questionnaire assesses communicative behavior and listening effort in daily life, using a five-point scale. Percentage scores are calculated separately for quiet and noisy situations.

Analysis

Outcomes of all children with SSD are visually and descriptively compared to average performance of the NH children ± 1 SD. Per test or questionnaire scale, the proportion of the group performing lower than the NH control group is presented separately for the SSD_CI and the SSD_noCI children. In addition, per test it is investigated how many children show a z-score ≤ -1, indicating that performance is clinically lower than average with respect to the Flemish norm data of the test itself. Both calculations are based on the child's performance at last measurement moment.

Results

Data logging shows that the SSD_CI children wore their CI for on average 6,1 ± 1,9 hours per day (across data logs), with individual CI use varying from 3,0± 1,3 to 8,1±1,1 hours per day, see table 1.

With regard to language development, the toddlers of the SSD_CI group seem to perform largely in line with the NH control group, whereas results of the SSD_noCI group appear to be more diverse, see figure 1 and table 2. For instance, while only 1 out of 6 SSD_CI children performs lower than the NH control group on language comprehension (z < NH group mean - 1SD), 6 out of 12 SSD_noCI children deviate from this mean. For one SSD_noCI child, yet none of the SSD_CI children, the score is also clinically deviant (i.e. outside the clinically considered normal performance range of z > 1). Expressive vocabulary performance deviates from the NH control group for 2 out of 6 SSD_CI children compared to 7 out of 12 SSD_noCI children, and is clinically deviant for 3 out of 12 SSD_noCI children as opposed to 0 out of 6 SSD_CI children. All of the SSD_CI children score in line with the NH group and within clinically normal performance concerning morphosyntactic knowledge. In the SSD_noCI group, however, performance of 5 out of 11 children deviates from the NH control group and out of this group for 2 out of 11 children it is below clinically normal performance.

Cognitive performance deviates from the NH control group for 1 out of 6 SSD_CI children, yet 6 out of 12 SSD_noCI children. For 3 of the 12 SSD_noCI children, compared to 1 of 6 SSD_CI children, the score is also below the clinically considered normal performance range.

Proportion of children showing lower PEACH questionnaire scores than the NH group was quite similar for the SSD_CI and SSD_noCI children, see figure 2.

Discussion

The current research presents the first data of our longitudinal study on the potential benefit of a CI in children with congenital SSD. To our knowledge, we are the first to assess linguistic and cognitive skills in children implanted at a very young age.

Preliminary data are encouraging, as five out of six SSD_CI children appear to perform largely in line with NH children on tests of expressive and receptive language. In the SSD_noCI group scores of a larger part of the children are lower than those of the NH controls. For some of the SSD_noCI children, performance is also clinically lower than average with respect to the Flemish norm data of the respective tests, especially with regard to morphosyntactic skills and expressive vocabulary. Difficulties in these branches of linguistics have recently been reported for school-aged children with unaided SSD as well[22].

Also test scores concerning cognition show lower performance compared to the NH control group for relatively more SSD_noCI children than SSD_CI children. Time will show whether differences in cognitive abilities persist and if so, whether these are caused by deprived auditory input or by other factors, such as etiology.

Equally important to the test data, the toddlers wear their device and do not seem to be hindered by acoustic input on one side and electrical input on the other. The average number of hours of CI use per day, as well as the range of individual hours of CI use, are quite similar to those reported by Polonenko et al[23]. Both their and our study have a relatively short follow up time as of yet, so it remains to be seen how CI use will be in the years to come. Nonuse (or

limited use) is reported for some children with cSSD[15,17], but these particular children were not implanted in the first years of life.

Our parent questionnaire data indicate that listening and communicating in noisy situations is still challenging for the SSD_CI children and requires high listening effort, as is the case for the SSD_noCI group. Hearing handicap presumably persists at this stage despite a CI.

An important strength of the current study is its longitudinal between-subject design which allows for careful comparison of performance of cSSD children with a CI to those without a CI and to NH children from the same population.

Our protocol is extended when the children are older to include evaluation of auditory performance, phonological processing, executive functioning and subjective CI benefit. Speech in noise understanding results of a first tested child, SSD_CI_1, show encouraging audiologically relevant (>1.5 dB[17]) differences with CI switched on compared to CI switched off in different spatial conditions.

Conclusions

Preliminary results for children with cSSD implanted at a very young age, show that the children wear their device and appear to perform largely in line with the NH children with regard to linguistic skills and cognitive milestones, whereas results of the SSD_noCI group are more diverse.

The current sample of data of the longitudinal project does not allow yet for drawing solid conclusions on the benefit of a CI in children with cSSD but is important to make evidence-based decisions regarding intervention. Long term observation of the linguistic and neurocognitive development of the children as well as their hearing abilities are of key importance to draw conclusions on CI benefit in this population**.**

Key Points:

1. This longitudinal study is the first to assess linguistic and cognitive outcomes of children with cSSD implanted at a very young age.
2. Despite the young age, these developing skills could be assessed by means of standardized test material and comparison to control groups.
3. The toddlers of the SSD_CI group wear their device and perform largely in line with the NH control group.
4. Linguistic and cognitive results of the SSD_noCI control group appear more diverse.
5. Longitudinal observation is of key importance to draw conclusions regarding CI benefit.

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Figure and Table Legends

Table 1. Participant Characteristics

Abbreviations: NHS: newborn hearing screening; cCMV: congenital cytomegalovirus; CND: cochlear nerve deficiency, IEM: congenital inner ear malformation.
†Age at implantation was generally 1 to 2 months after first test moment at inclusion.
‡ The > sign indicates no response at the highest level tested. SSD_noCI_4 was not tested beyond 70 dBnHL but additional pure tone audiometry showed no responses at 90 dB HL (250-500-1000-2000 Hz).
§ Hours of use relatively low because of family related issues
¶ SSD_noCI_7 was diagnosed with OAV syndrome.
In both SSD groups, some of the children receive(d) auditory or linguistic rehabilitation or early home based guidance. None of the SSD_noCI children wears a hearing assistive device.

Table 2. Performance of the SSD groups in comparison to the NH control group and test norm data.

Left: number of children per SSD group with, at last measurement, a score lower than performance of the NH control group (as indicated by the NH group's average score ± 1 SD); right: number of children with at last measurement a z-score below clinically considered average performance with respect to Flemish norm data (z-score ≤ -1) (only for test outcomes).

†data of SSD_noCI_6 not taken into account because cooperation was insufficient due to severe shyness and it is therefore unclear if the scores represent true abilities.

‡data of SSD_noCI_7 and SSD_noCI_12 missing.

Figure 1. Individual test outcomes.

1a: language comprehension; 1b: expressive vocabulary; 1c: morphosyntactic knowledge †; 1d: mastery of mile stone skills in cognitive development. Left picture, in blue dots: group SSD_CI, n=6; right picture, in green triangles, group SSD_noCI, n=12. Y-axis: z-score. X-axis: age at testing, in months. Each data point represents the score of one child, data points of the same child are connected. In red: average score of the NH control group ± 1 standard deviation, based on n=19, 16, 12, 11, 9, 8 and 6 resp. for measurements around 25, 31, 37, 43, 49, 55 and 61 months of age. The grey box represents the normative mean of 0 plus/minus 1 SD, scores below this box are considered clinically to be below average.

†In fig. 2C (relatively very low) scores of 1 child of the SSD_noCI group were not included in interpretation/analysis because cooperation was insufficient due to severe shyness and it is therefore unclear if the scores represent true abilities.

Figure 2. Individual outcomes on the PEACH+.

2a: auditory functioning quiet; 2b: auditory functioning noise; 2c: ease of listening quiet; 2d: ease of listening noise. Left: SSD_CI group, n=6; right: SSD_noCI group; n=9. Y-axis: percentage scores. X-axis: age at testing, in months. Each data point represents the score of one child, data points of the same child are connected. In red: average score of the NH control group ± 1 standard deviation, based on n=4,11,10,9,7 and 5 resp. for measurements around 30, 37, 43, 49, 56 and 61 months of age.

Table 1. Participant Characteristics

Participant	Time of diagnosis	Age at implantation † (SSD_CI) / Age at inclusion (SSD_noCI) (yr.mo(;d))	Side of HL	Etiology	ABR threshold, (dB nHL) affected ear ‡	CI experience (months)	CI use (average hours per day±SD)
SSD_CI_1	10 months	02.02;21	Left	Fracture of left petrous bone due to fall	>90	42,1	3,0±1,3 §
SSD_CI_2	NHS	00.08;21	Left	cCMV	>80	31,8	7,0±3,7
SSD_CI_4	NHS	01.00;26	Left	cCMV	>80	18,8	4,7±1,2
SSD_CI_5	NHS	01.02;24	Right	IEM (incomplete partition type II)	>80	17,2	8,1±1,1
SSD_CI_6	NHS	01.02;15	Right	cCMV	>80	14,4	7,0±1,6
SSD_CI_8	NHS	01.02;22	Left	cCMV	100	11,5	6,8±1,3
SSD_noCI_1	NHS	01;03	Left	CND	>85		
SSD_noCI_2	NHS	01;02	Right	cCMV	>100		
SSD_noCI_3	NHS	03;00	Right	unclear	>80		
SSD_noCI_4	NHS	01;06	Right	cCMV	>70		
SSD_noCI_5	NHS	02;11	Left	CND	>85		
SSD_noCI_6	NHS	03;01	Left	CND	>90		
SSD_noCI_7 ¶	Perinatal	01;11	Left	CND	>95		
SSD_noCI_8	NHS	02;02	Right	CND	>90		
SSD_noCI_9	NHS	02;06	Left	CND	>90		
SSD_noCI_11	NHS	02;00	Right	cCMV	>90		
SSD_noCI_12	NHS	01;06	Left	CND	>85		
SSD_noCI_13	NHS	02;00	Left	unclear	>90		

Abbreviations: NHS: newborn hearing screening; cCMV: congenital cytomegalovirus; CND: cochlear nerve deficiency, IEM: congenital inner ear malformation.

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¶ SSD_noCI_7 was diagnosed with OAV syndrome.

In both SSD groups, some of the children receive(d) auditory or linguistic rehabilitation or early home based guidance. None of the SSD_noCI children wears a hearing assistive device.

Table 2. Performance of the SSD groups in comparison to the NH control group and test norm data.

		Lower than performance NH control group		Lower than clinically considered average performance	
Test/Questionnaire		SSD_CI group	SSD_noCI group	SSD_CI group	SSD_noCI group
Language comprehension		1/6	6/12	0/6	1/12
Expressive vocabulary		2/6	7/12	0/6	3/12
Morphosyntactic knowledge †		0/6	5/11	0/6	2/11
Cognitive skills		1/6	6/12	1/6	3/12
PEACH+ ‡	Auditory functioning in quiet	2/6	3/9	-	-
	Auditory functioning in noise	3/6	7/9	-	-
	Ease of listening in quiet	2/6	4/9	-	-
	Ease of listening in noise	5/6	7/9	-	-

Left: number of children per SSD group with, at last measurement, a score lower than performance of the NH control group (as indicated by the NH group's average score ± 1 SD); right: number of children with at last measurement a z-score below clinically considered average performance with respect to Flemish norm data (z-score ≤ -1) (only for test outcomes).

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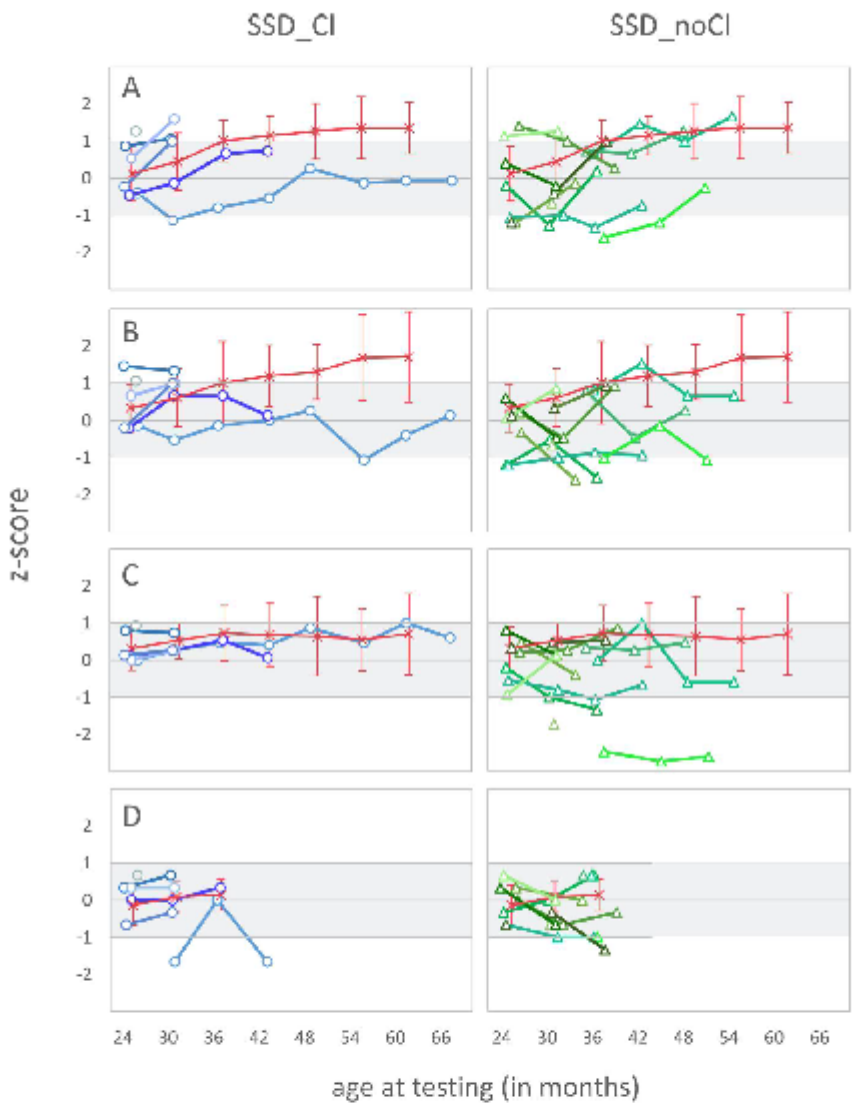
‡data of SSD_noCI_7 and SSD_noCI_12 missing.



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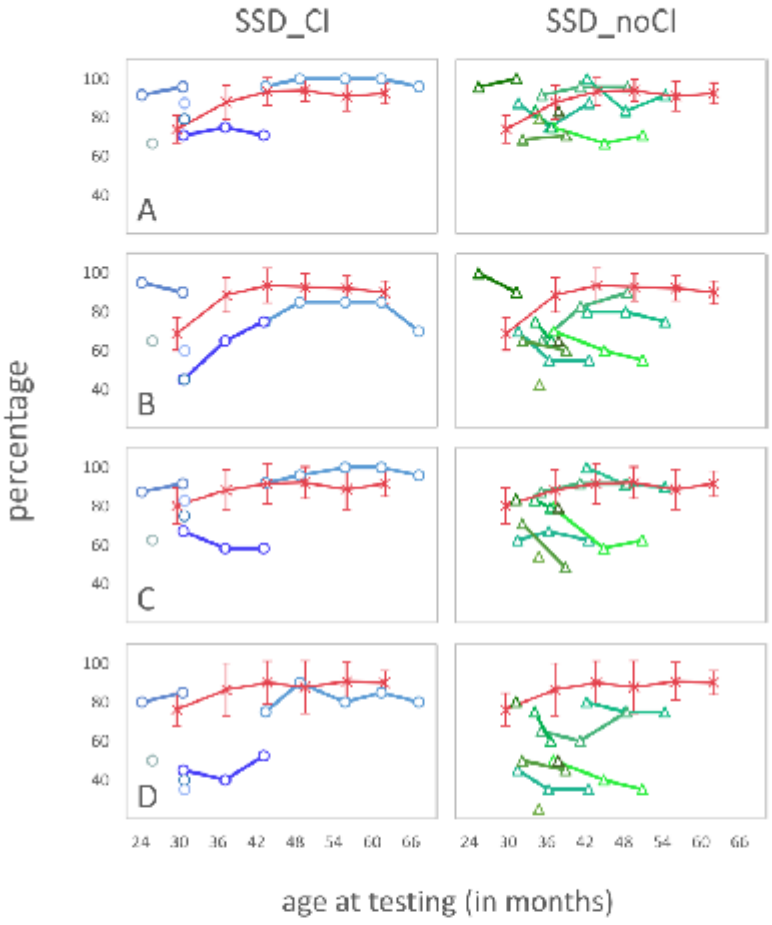

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