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# Cochlear Implantation in Children with Single-Sided Deafness: Preoperative Assessment and Device Use

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**Keywords**

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- Etiology
- Cochlear implant
- Vestibular function
- Data log

## Abstract

**Objectives:** To characterize the first cohort of cochlear implanted pediatric patients with single-sided deafness (SSD) at Aarhus University Hospital, Denmark. Data includes etiology, age at implantation, balance testing, and language assessment prior to implantation as well as data log of Cochlear Implant usage.

**Methods:** Clinical data from 34 children with SSD (21 females (62%) and 13 males (38%)), implanted with Cochlear Implant (CI) during 2018-2023, was analyzed.

**Results:** Acquired prenatal and postnatal sensory disorders toxoplasmosis, rubella, cytomegalovirus and herpes simplex (TORCH) risk factors were associated with sensorineural hearing loss (SNHL) in 19 cases (56%). Congenital Cytomegalovirus (cCMV) DNA was diagnosed in 17 samples (50%). The cause of the SNHL remained unknown for 11 (32.4%) children. In 6 (66.7%) of the cases with unknown etiology, CT scan showed inner ear malformations (IEM).

The mean (SD) age for implantation was 4.2 ± 3.5 years. For the children with congenital SNHL, the mean (SD) age for implantation was 2.6 ± 1.6 years. For the children with acquired SNHL, the mean (SD) age for implantation was 6.9 ± 4.3 years. Data log showed that children with SSD and CI wore their processor 6.2 ± 3.73 (1 SD) h/day.

**Conclusions:** Further work is needed in order to assess how etiology, age at implantation, and device use affect outcome. Understanding of the long-term outcomes associated with CI in children with SSD can contribute to more thorough counselling on outcome expectations and the best auditory rehabilitation.

## INTRODUCTION

Single-sided deafness (SSD) is characterized by severe to profound sensorineural hearing loss (SNHL) (>70 dB HL) in one ear and normal hearing (<25 dB HL) on the contralateral ear. Recent evidence indicates that children with SSD perform poorer linguistically and academically than their normal hearing peers [1-3]. Binaural hearing enables localization of sound and provides access to information needed to hear and understand speech in complex listening situations, such as places with noise. Children with normal hearing benefit from the head shadow effect, binaural squelch and binaural summation effect to derive spatial characteristics of a sound. Children with SSD lack these binaural cues and may experience difficulties with speech perception in noise and sound localization compared to children with normal hearing [4,5]. Previous studies of language skills in children with SSD have shown challenges particularly on expressive language and grammar, whereas their language development is age-appropriate on receptive language [6,7].

Other treatments of SSD such as contralateral routing of signal hearing aids (CROS) and bone-anchored hearing systems (BAHS) do not restore binaural hearing as the auditory input is sent to the contralateral ear. Currently, there is doubt as to whether treatment of young children with CROS or BAHS may be more disturbing than beneficial for their hearing perception, as the sound from the contralateral ear may disturb the sound in the normal hearing ear [8-10]. A Cochlear Implant (CI) enables binaural auditory stimulation of the brain in patients with SSD. Experience with CI in younger children (< 4 years) with SSD is limited; only few case series of smaller heterogeneous cohorts have been published. However, studies of children with SSD and CI have indicated improved speech recognition in noise [6,11] and improved sound localization [6,11,12].

Since 2017, CI has been offered to children with SSD. Initially CI was only implanted in children with SNHL caused by bacterial meningitis or etiologies with risk of development of SNHL on the contralateral normal ear. These etiologies included

cytomegalovirus (CMV) or inner ear malformations (IEM) such as enlarged vestibular aqueduct (EVA), and incomplete partition (IP). Since 2023, children are considered for cochlear implantation despite etiology, if they have an intact auditory nerve confirmed by magnetic resonance imaging of the posterior fossa and are younger than 4 years at time of surgery.

The aim of the current paper was to provide an overview of the first pediatric cohort of SSD patients with CI at Aarhus University Hospital, Denmark (West Danish CI Center).

## MATERIAL AND METHODS

This study is a retrospective analysis of children who underwent cochlear implantation from January 2018 - June 2023 because of SSD. All children implanted with unilateral CI because of SSD in this period were included in the study.

All children with SSD and CI received auditory verbal therapy from a Listening and Spoken Language Specialist or an auditory verbal practitioner during the first year of CI use.

Data was collected with permission from the local institutional review board and included auditory and language assessment, etiology, imaging, cochlear implant device, age at implantation, balance testing and data log.

### Pre-CI evaluation

Auditory and language assessment

All children participated in Auditory Brainstem Response (ABR, Eclipse EP25 Evoked Potential System Interacoustics, Denmark) and behavioural audiometry (Visual Reinforcement Audiometry or Conditional Play Audiometry). Language assessment was conducted as part of our standard baseline-testing before cochlear implantation. As a baseline for language level, children in the age group 2.5-5.0 years and children above 13 years were tested with Peabody Picture Vocabulary Test, 4th Edition (PPVT-4), which is a norm-referenced test of receptive vocabulary [13]. Children between 5 and 13 years were tested with Clinical Evaluation of Language Fundamentals, 4th Edition (CELF-4) [14]. CELF-4 is a standardized measure of receptive and expressive language skills.

### Etiology

CMV status was determined in all children with either Guthrie card (until 2022) or with urine and blood samples before the age of three months (from 2022). All children were offered evaluation for variants in genes associated with non-syndromic or syndromic SNHL. For children with positive cCMV testing, only 23.5% underwent genetic testing. The primary gene test targeted GJB2, GJB6 and SLC26A4. If the test was negative, a larger gene panel with 160 genes was employed. Depending on whether an underlying syndrome was suspected or not, testing with either a syndromic SNHL gene panel or a non-syndromic gene panel was performed. Of the patients with unknown etiology, only 45.5% got a large gene panel testing.

### Imaging

High resolution Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) were performed on all patients, if necessary in general anesthesia. Both modalities were carried out with specific inner ear/temporal bone protocols.

### Surgery

All patients were implanted by a team of experienced CI-surgeons. All patients had complete electrode insertion using a round window approach. Electrode placement was confirmed intraoperatively by Trans Impedance Matrix (TIM) measurements followed by Neural Response Telemetry (NRT) to assess neural integrity.

### Vestibular function

Vestibular tests were performed when the child was able to participate. Saccular function was evaluated with cervical Vestibular Evoked Myogenic Potential (cVEMP), (Eclipse EP25 Evoked Potential System (Interacoustics, Denmark). Stimulation with 65 db nHL, 500Hz, bone conduction, (B-81, Interacoustics, Denmark). cVEMP was considered absent when a biphasic waveform was absent. Function of the semicircular canals was evaluated with Video Head Impulse Test vHIT (EyeSeeCam, Interacoustics, Denmark) or head impulse test with a video recording of the eyes and manually evaluated for catch-up saccades.

### Post CI assessment

Data logs containing average daily CI use were extracted from the Custom Sound Pro fitting software (Cochlear Ltd.).

## RESULTS

### Participants

The current study includes data from 34 children with SSD (21 females (62%) and 13 males (38%)), implanted with CI during 2018-2023. Data of auditory assessment, etiology, imaging, cochlear implant device, age at implantation, balance testing, and data log is shown in Table 1.

The youngest child with SSD was implanted at the age of 6 months and the oldest at the age of 16.2 years. The mean (SD) age for implantation in children with SSD was 4.2 ± 3.5 years (median = 3.0 years, one sided distribution). Overall, the children can be divided into two groups depending on the time of the onset of hearing loss (congenital or acquired). In 22 (64.7%) children the hearing loss was found at the newborn hearing screening (NHS). Ten (29.4%) children passed the NHS. In two children the NHS wasn't administered, but time for onset of hearing loss is known for one of the children (acquired, see Table 1, CI-17).

For the 22 children with congenital SNHL, the mean (SD) age for implantation was 2.6 ± 1.6 years (median = 2.0 years, one sided distribution). For the 11 children with acquired SNHL, the

**Table 1**: Subject Demographics

| Subject ID | Sex | Etiology | NHS | Onset of Diagnosis | Age at implantation (years) | Side | Vestibulogy | Pre-Op Pure Tone average CI (dB) | Pre-Op Pure Tone Average contra (dB) | Implant | Speech processor | Daily use |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| CI-01 | F | Meningitis | Pass | acquired | 0.56 | L | Not accomplished | 74 | 4 | CI512 | N7 | 2,2 |
| CI-02 | F | cCMV | Failed unilat | congenital | 3.14 | L | cVEMP -/-, vHIT -/- | 87 | 25 | CI 512 | N7 | |
| CI-03 | M | cCMV | Failed unilat | congenital | 3.96 | L | Not accomplished | 99 | 18 | CI 532/CI 622 | N7 | |
| CI-04 | F | cCMV | Failed unilat | congenital | 2.96 | R | Not accomplished | 70 | 9 | CI 512 | N7→Kanso 2 | 2,1 |
| CI-05 | F | Waardenburg | Failed unilat | congenital | 1.3 | L | Not accomplished | 84 | 1 | CI 632/CI622 | N7 | 10,1 |
| CI-06 | F | cCMV | Failed unilat | congenital | 1.39 | R | Not accomplished | 85 | 13 | CI 632 | N7 | 6,8 |
| CI-07 | M | cCMV | Failed unilat | congenital | 1.3 | R | Not accomplished | 71 | 10 | CI 632 | N7 | 4,3 |
| CI-08 | F | cCMV | Pass | acquired | 10.57 | L | vHIT +/+ | 108 | 9 | CI 622 | N7 | 0 |
| CI-09 | F | cCMV | Failed unilat | congenital | 1.72 | R | cVEMP+/- HIT +/+ | 66 | 14 | CI 632 | N7 | 9,5 |
| CI-10 | F | cCMV | Failed unilat | congenital | 4.6 | L | cVEMP -/- HIT +/+ | 79 | 13 | CI 632 | N7 | 4,1 |
| CI-11 | M | cCMV | Failed unilat | congenital | 1.33 | L | cVEMP -/-, HIT: pat bilat | 75 | 15 | CI 622 | N7 | 7,0 |
| CI-12 | F | cCMV | Failed unilat | congenital | 4.79 | L | cVEMP+/-, HIT:+/+ | 70 | 13 | CI 622 | N7 | 7,1 |
| CI-13 | M | cCMV | Pass | acquired | 6.87 | L | cVEMP +/-, vHIT: +/- | 70 | 10 | CI 632 | N7 | 1,5 |
| CI-14 | M | cCMV | Failed unilat | congenital | 6.55 | R | cVEMP +/+, vHIT +/+ | 83 | 13 | CI 622 | N7 | 4,8 |
| CI-15 | F | Unknown | Pass | acquired | 7.05 | R | cVEMP+/+, vHIT +/+ | 93 | 5 | CI 632 | N7 | 3,9 |
| CI-16 | M | cCMV | Failed unilat | congenital | 1.76 | R | cVEMP -/-, HIT (can't participate) | 68 | 11 | CI 622 | N7 | 10,0 |
| CI-17 | F | Unknown | Not tested | acquired | 7.98 | L | cVEMP +/+, vHIT +/+ | 63 | 8 | CI 632 | N7 | 9,7 |
| CI-18 | F | Unknown | Not tested | unknown | 11.04 | R | cVEMP -/+ V-HIT+/+ | 93 | 0 | CI 632 | N7 | 6,2 |
| CI-19 | F | cCMV | Pass | acquired | 7.64 | L | cVEMP-/+, V-HIT +/+ | 109 | 3 | CI 632 | N7 | 3,2 |
| CI-20 | M | cCMV | Failed unilat | congenital | 1.03 | L | cVEMP +/+, HIT +/+ | 63 | 16 | CI 632 | N7 | 6,9 |
| CI-21 | M | COL4A, ANSD | Failed unilat | congenital | 4.95 | L | cVEMP +/+, HIT +/+ | 110 | 10 | CI 622 | N7 | 10,8 |
| CI-22 | M | cCMV | Failed unilat | congenital | 1.53 | L | cVEMP +/+, HIT: (can't participate) | - | 15 | CI 632 | N7 | 5,9 |
| CI-23 | M | Unknown | Pass | acquired | 4.01 | R | cVEMP: (can't participate) HIT +/+ | 85 | 15 | CI 632 | N7 | 11,7 |
| CI-24 | M | SLC26A4 | Failed unilat | congenital | 2.47 | R | cVEMP +/+ HIT +/+ | 83 | 13 | CI 632 | N8 | 6,0 |
| CI-25 | F | Meningitis | Pass | acquired | 1.38 | R | cVEMP +/+ HIT +/+ | 54 | 10 | CI 622 | N8 | 7,5 |
| CI-26 | F | Unknown, ANSD | Failed unilat | congenital | 2.46 | L | cVEMP +/+ HIT +/+ | 85 | 22 | CI 632 | N8 | 3,1 |
| CI-27 | M | Unknown | Pass | acquired | 5.37 | L | cVEMP +/+ HIT +/+ | 87 | 25 | CI 622 | N8 | 11,1 |
| CI-28 | F | Unknown | Pass | acquired | 16.23 | L | cVEMP+/+, vHIT +/+ | 94 | 3 | CI 632 | N8 | 0,5 |
| CI-29 | F | Unknown | Failed unilat | congenital | 2.15 | L | cVEMP +/+ HIT +/+ | 80 | 20 | CI 622 | N8 | 3,9 |
| CI-30 | F | Unknown | Pass | acquired | 7.77 | R | cVEMP -/+, vHIT:+/+ | 95 | 12 | CI 622 | N8 | 16,7 |

| | | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| CI-31 | F | Unknown | Pass | congenital | 3.78 | R | cVEMP -/-, HIT Not accomplished | 79 | 12 | CI 622 | N8 | - |
| CI-32 | M | cCMV | Failed unilat | congenital | 3.93 | L | cVEMP: -/-, HIT: +/+ | 85 | 10 | CI 632 | N8 | - |
| CI-33 | F | Waardenburg, MITF | Failed unilat | congenital | 1.34 | R | cVEMP+/+, HIT: +/+ | 70 | 18 | CI 622 | N8 | - |
| CI-34 | F | Unknown | Failed unilat | congenital | 0.89 | R | cVEMP -/-, HIT: -/- | 68 | 21 | CI 622 | N8 | - |

NHS = new born hearing screening, IEM= inner ear malformations, L=left, R=right, N=Nucleus

mean (SD) age for implantation was 6.9 ± 4.3 years. For one child the time of onset of hearing loss remains unknown (see Table 1, CI-18).

## Auditory assessment

ABR and/or behavioural audiometry were performed in all children. Pure Tone Average (4 frequency PTA at 0.5, 1, 2, and 4 kHz) values in Table 1 are from either ABR or behavioural audiometry depending on the last test performed before surgery. The mean PTA for the children with SSD and CI in this group was 12 dB (range 0-25 dB) on the normal hearing ear and 81 dB on the ear with SNHL (range 54-110 dB). The child implanted with a PTA of 54 dB had meningitis and severe SNHL in the high frequencies (2 and 4 kHz) and was implanted early in the process due to the risk of fibrosis/ossification of the cochlea.

## Language level prior to implantation

Sixteen children were tested prior to implantation; eight children with CELF-4 and eight children with PPVT-4. Twelve children were too young to participate in conventional testing. Five children were not able to participate in testing.

The mean standard score on CELF-4 for the 8 children was 98 (range 82-114), which means that all children except one were within the range of average for the children's chronological age. For the children tested with PPVT-4, the mean standard score was 106 (range 81-122). The children tested with PPVT-4 scored above average for the children's chronological age.

## Etiology

Congenital CMV infection was detected in 17 samples (50%), all of whom had asymptomatic cCMV infection. (TORCH) risk factors were associated with HL in 19 cases (56%). SNHL due to meningitis was found in two children (5.9%). Hereditary SNHL was diagnosed in four cases (11.8%); with pathogenic variants in the SLC26A4 gene in one case, Waardenburg variants in two cases, and COL4A6 in one case. The cause of SNHL remained unknown in 11 (32.4%) children (Figure 1).

## Imaging

In total, inner ear malformations (IEM) were found in 9 cases (for details see table 2). In 6 (66.7%) of the cases the underlying etiology of the SNHL remains unknown.

We found different IEM (unilateral and bilateral) such as incomplete partition-II (IP-II), enlarged vestibular aqueduct (EVA) and dilated vestibule including combinations of these [15]. In addition, CT scan showed a possible apical turn anomaly that could not be classified. Therefore, this child (CI-29) is not included in the total number of children with IEM.

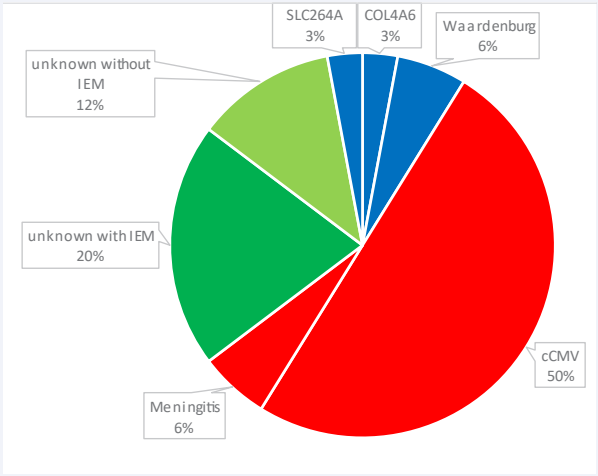


**Figure 1** Shows the etiology of all 34 participants. Red indicates environmental induced HL; blue heriditary HL associated with specific gene mutation; green unknown. IEM= Inner ear malformations

## Vestibular tests

*Cervical vestibular evoked myogenic potentials (cVEMP)*

Saccular function measured as cVEMP was assessed in 25 children. Two children failed to complete the test while the remaining patients were operated prior to routine inclusion of saccular testing in our laboratory. CVEMP response was absent in 44% (11/25) of the deaf ears. Two children (8%) with cCMV had a normal cVEMP response on the deaf ear, but no response on the contralateral normal hearing ear. Six children (24%) had absent cVEMP responses bilaterally (See Table 1).

*Video Head impulse test (vHIT) and Head impulse test (HIT)*

Eleven children were assessed and completed the vHIT. One patient (9%) had a bilateral pathologic test with low gain value and corrective saccades. The same patient had absent cVEMP responses. The remaining 10 children (91%) had normal vHITs. Ten were assessed with HIT and two of them (20 %) had corrective saccades bilaterally. The remaining nine children (90%) had a normal HIT (See Table 1).

In total, 29 children had their vestibular organ evaluated and 13 (45%) of them had an abnormal response in one of the tests.

### Cochlear Implant device

All children in the group received a Cochlear Nucleus implant (CI512, n = 3; CI532, n = 1; CI632, n = 17; CI622, n = 13) and sound processor (Nucleus 7, n =23; Nucleus 8, n=11).

Two children (CI-02+03, table 1) with congenital SSD due to cCMV experienced a progressive SNHL in the better (non-implanted) ear, both approximately 2½ years after implantation, for which one is now fitted with a hearing aid and the other has received a second CI. Two children were re-implanted; one because of device failure (one year after first implantation) and the other because of abscess/infection (three years after implantation). Four years after implantation one child exchanged her N7 to a KANSO 2. Both children who were re-implanted received a CI622 after first being implanted with a CI532 and a CI632 respectively (see Table 1).

### Daily device use

Daily device use was based on data logging in Custom Sound Pro®. Average daily hours of use were measured during a programming visit to the implant centre. Device use was only measured in 28 out of 34 children, since CI was not switched on in four children at read-out (CI-31-34) and two children experienced a progressive SNHL in the better (non-implanted) ear (CI-02+03). Recipient age and CI experience at data logging read-out is not equal across recipients, see Figure 2 and Figure 3. At data logging read-out the participants' mean age (SD) was 5.36 (3.49) years and median CI experience of 0.73 years (because of skewed dataset, not normally distributed).

On average the children with SSD and CI wore their processor 6.2 ± 3.73 (1 SD) hours per day with individual device use between 0.5 and 16.7 hours per day. Based on data logging 11 out of 28 children are "regular" uses (>7 hr/d), 15 out of 28 children are "limited" users (1-7 hr/d), and 2 out of 28 children are "nonusers" (<1 hr/d). Data from the child who used the processor 0 hours per day is not part of the averages calculated above (CI-08, Table 1). Data log (see table 1) showed that children with CI implanted <2 years of age (N=10) have a mean (SD) use at 7.02 (2.51). The largest amount of daily use was found in the group of children implanted between 4 and 6 years of age (N=5) with mean (SD) h/day at 8.96 (3.26). Children implanted at 2-4 years of age (N=4) and above 6 years of age (N=9) used CI less with 3.77 (1.66) h/day and 5.81 (h/day), respectively.

## DISCUSSION

Until 2023, CI was only implanted in children with SSD with risk of development of SNHL on the contralateral normal ear in Denmark. This impact the results of this study where cCMV is the cause of the SNHL in 50% of the children. Cushing et al found a comparable prevalence of cCMV in 43% of the children in their study [17]. A recent study by Gordon et al., including 57 children also found cCMV as the most common cause of SNHL

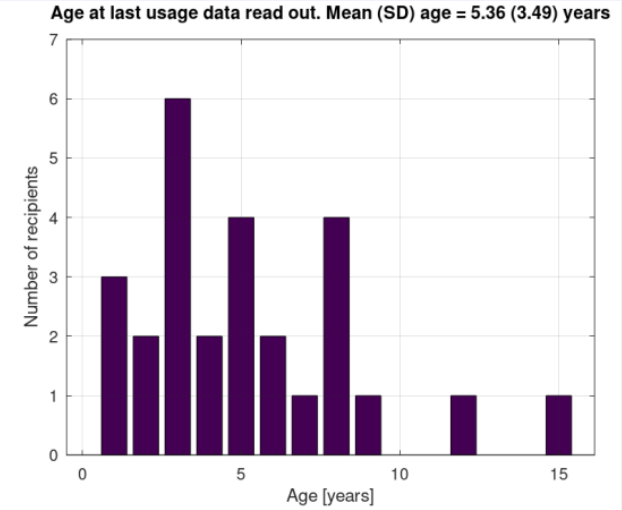


**Figure 2** Age at last usage data read-out.

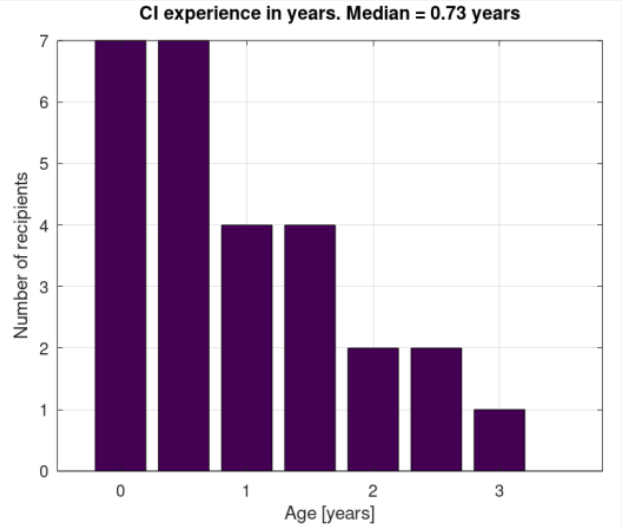


**Figure 3** CI experience at usage data read-out.

with 19 children with congenital, and two children with acquired SNHL [18]. The second most common cause in their study was genetic SNHL; the third was meningitis while the cause remained unknown in 15 (26%) cases. Park et al found only two cases of cCMV out of the 20 children in their study and had 11 (55%) cases with unknown etiology [8]. In the study by Arras et al including 12 children, 67% had SNHL due to cCMV and the cause was unknown in 17% [6]. In the present study the cause of SNHL was unknown in 11 (32.4%) children. This indicates that this study has a higher proportion of children with unknown etiology than the studies by Gordon et al and Arras et al but a lower percentage than the study of Park et al does. The high percentage (66.7%) of children with inner ear anomalies in the group with unknown etiology should be considered in further testing. EVA is part of the IEM in all six children with unknown etiology. According to a review of non-syndromic EVA by Roesch et al, identifying the causative gene in Caucasian cohorts with EVA could be challenging [19].

**Table 2:** Subject Imaging

| Subject ID | CT | MR |
|---|---|---|
| CI-01 | NAD | Hydrocephalus |
| CI-02 | NAD | WML |
| CI-03 | NAD | WML |
| CI-04 | NAD | WML |
| CI-05 | Bilateral dilated vestibule | NAD |
| CI-06 | NAD | NAD |
| CI-07 | NAD | NAD |
| CI-08 | NAD | NAD |
| CI-09 | NAD | NAD |
| CI-10 | NAD | WML |
| CI-11 | NAD | WML |
| CI-12 | NAD | WML |
| CI-13 | NAD | WML |
| CI-14 | IPII bilateral + EVA bilateral | IPII bilateralt + EVA bilateral |
| CI-15 | EVA bilateral | NAD |
| CI-16 | NAD | WML |
| CI-17 | EVA sin | NAD |
| CI-18 | EVA bilateral + dilated vestibule | EVA dxt + dilated vestibule |
| CI-19 | NAD | NAD |
| CI-20 | NAD | NAD |
| CI-21 | NAD | NAD |
| CI-22 | NAD | NAD |
| CI-23 | IPII bilateral+ EVA dxt | EVA dxt |
| CI-24 | IPII dxt + EVA dxt | EVA dxt |
| CI-25 | Fibrosis lateral semicircular canal (SCC) dxt | Inflammation in cochlea dxt |
| CI-26 | NAD | NAD |
| CI-27 | NAD | NAD |
| CI-28 | EVA sin | EVA sin |
| CI-29 | Possible anomaly in apical turns bilateral | NAD |
| CI-30 | IPII, EVA, dilated vestibule dxt and IPII, dilated vestibule sin | EVA dxt |
| CI-31 | NAD | NAD |
| CI-32 | NAD | NAD |
| CI-33 | NAD | NAD |
| CI-34 | NAD | NAD |

WML = White matter lesion, EVA= Enlarged vestibular aquaduct, IP= Incomplete Partition, NAD= No abnormality detected

Biallelic mutations in the pendrin gene were strongly correlated with Pendred syndrome, but were only found in one-fourth of patients with non-syndromic EVA. In the majority of patients with monoallelic pendrin mutations but also in some patients with no mutations in causative genes, a specific Caucasian EVA (called CEVA) haplotype seemed to be responsible for unilateral or bilateral EVA [20-22].

In this study, the mean (SD) age for implantation for all the children was 4.2 ± 3.5 years (median = 3.0 years, one sided distribution). For the children with congenital SNHL, the mean (SD) age for implantation was 2.6 ± 1.6 years (median = 2.0 years, one sided distribution); and for the children with acquired SNHL, the mean (SD) age was 6.9 ± 4.3 years. In the study by Park et al [8] the mean (SD) age was 5.0 ± 1.1 and in the study by Gordon et al [18] the mean (SD) age of implantation for early-onset (prelingual) group was 2.47 ± 1.58 and the late onset (postlingual) 11.67 ± 3.91 years. The mean (SD) age of implantation in the group of children with congenital SNHL corresponds to the mean age found in the study by Gordon et al, whereas the mean (SD) age of implantation in the group of children with acquired SNHL variates when the studies are compared.

In Denmark children with SSD are not routinely evaluated post operatively with audiological testing such as localization and speech discrimination in noise. The assessment of language in the present study showed similar results to Arras et al [2] and Halliday et al [7].who both showed with-in average receptive language skills. Therefore, the used test battery can´t capture language improvement post CI. Due to limited audiological testing and assessment of complex language skills, it is not possible to assess how age at implantation affects these outcome measures in the present study. Further studies of this cohort should look into outcome of CI and understanding of more complex language skills such as grammar and narrative skills.

Data logging of daily device use for children with SSD and CI shows on average 6.2 ± 3.73 (1 SD) hours per day with individual device use between 0.5 and 16.7 hours per day. Arras et al 2022 showed that children with SSD and CI wore their speech processor on average 8.9 ± 2.7 hours per day, with individual averages ranging between 2.9 and 12.2 hours per day [6]. The study by Park et al., showed device use ranging from 7.1 to 12.0 hours per day [8]. Gordon et al included 57 children with SSD in their study [18]. They aimed to use data logging of daily CI use to assess acceptance of the CI in children with SSD. Average daily use was 6.5 hours per day. Compared to other studies the children in the present used their CI less. This might be explained by the fact that the children in this cohort had less experience with their CI. The median time was 0.73 years, whereas the median time was 3.1 years in the study by Arras et al., [6]. Furthermore, data logging was made as a single read-out, which means that it does not show average device use over time as in the study by Arras et al., [6]. Data log gives an idea of degree of acceptance of the device, which again is influenced by different factors, such as hearing outcome but probably also psycho-social factors among others. For children with SSD and CI, we plan to implement both patient related outcome measures and objective measures that brings us closer to evaluating the possible benefits of this treatment.

Twenty-nine of the children in our cohort underwent vestibular testing, and 45% of the cohort demonstrated an abnormality of the vestibular organ, primarily on the deaf ear. This finding is consistent with Sokolov et al. [24], and the results further support the idea that the prevalence of vestibular dysfunction in unilateral deafness is high and similar to that of children with bilateral deafness.

## CONCLUSION

This study describes a cohort of children with SSD in regards to etiology, age at implantation, vestibular function, and language assessment prior to implantation as well as device use post CI.

Further work is needed to assess the benefits of CI for children with SSD. Follow-up should focus on spatial hearing and language skills to understand the long-term outcomes associated with CI in children with SSD, and provide thorough counselling on outcome expectations and the best auditory rehabilitation.

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