# Prevalence and etiology of sensorineural hearing loss in children with down syndrome : a cross-sectional study

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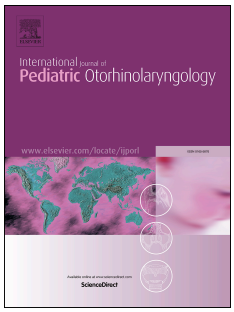


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# Prevalence and etiology of sensorineural hearing loss in children with Down syndrome: A cross-sectional study.

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**ABSTRACT**

*Background:* The prevalence and causes of sensorineural hearing loss (SNHL) in children with Down syndrome (DS) are poorly delineated.

*Objective:* To describe the prevalence, severity, laterality and underlying etiology of SNHL in a cohort of children with DS.

*Methods:* A cross-sectional study was performed among all children with DS followed at the multidisciplinary Downteam of the Antwerp University Hospital. Patients' characteristics, risk factors for hearing loss, audiometric data and results of an etiological work-up were collected.

*Results:* Among 291 patients in follow-up, 138 patients (47.4%) presented with hearing loss. In the majority this was caused by middle ear effusion and only 13 patients (4.5%) had sensorineural hearing loss, 7 boys and 6 girls with a mean age of 14.4 ± 7.4 years. Hearing loss was bilateral in 8 cases. Hearing loss severity was graded as mild in 38.5%, moderate in 30.8% and profound in 30.8% of the patients. An etiological work-up was completed in 9 children. Four patients presented with single sided deafness due to cochlear nerve deficiency. One patient had a genetic cause and in 2 patients the hearing loss was attributed to excessive noise exposure. The etiology of hearing loss was unknown in 6 patients.

*Conclusion:* Sensorineural hearing loss is uncommon in children with DS with a prevalence of 4.5%. Etiological work-up may allow identifying a specific underlying cause. Cochlear nerve deficiency was found in 4 children with DS and single sided deafness.

*Keywords:* Down syndrome, trisomy 21, sensorineural hearing loss, single sided deafness

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## 1. INTRODUCTION

The chromosomal anomaly of trisomy 21, commonly known as Down syndrome (DS), has been associated with many otorhinolaryngologic manifestations mostly due to the anatomical malformations in the head and neck region [1–4]. Regular visits to the Ear-Nose-Throat (ENT) specialist are therefore recommended [5–7]. Hearing loss is the most common ENT manifestation in conjunction with this syndrome [8–11]. Hearing loss may predispose to delayed acquisition of speech and language, thus preventing patients to reach their full potential [9,12]. The implementation of universal neonatal hearing screening has been successful in detecting hearing loss present at birth [13]. Thereafter, audiological monitoring of young children with DS is done by behavioral audiometry [14,15]. An uncooperative child should be tested by means of an auditory brain stem response (ABR) either in natural sleep or under general anesthesia to obtain an objective hearing threshold estimate [16–19]. Current guidelines from the American Academy of Pediatrics advocate that children with stenotic ear canals be seen, with ears examined under office microscope if needed, every 3 months until the ear canals grow. Audiograms are suggested every 6 months until the child is able to do "ear specific" testing and then annually if normal hearing is present [15].

The most common type of hearing loss in DS is conductive hearing loss caused by middle ear effusion (OME) with a prevalence ranging from 38% up to 78% [5,10,20,21]. In these patients, hearing acuity may improve by ventilation tube (VT) placement [22–24]. Other possible causes of conductive hearing loss are sequelae of OME such as tympanic membrane perforation, chronic otitis media/cholesteatoma, or ossicular chain abnormalities [23,25]. The prevalence of sensorineural hearing loss (SNHL) in children with DS is not clearly defined with figures ranging from 4% to 55% [4]. In a systematic review, Shott S.R. et al. [7] reported a prevalence of 4% to 20% for sensorineural and mixed hearing loss. Similarly as in non-DS children, SNHL in children with DS may be caused by a genetic defect, a congenital infection, anatomical abnormalities or may be related to perinatal risk factors or yet unidentified causes [26–29].

The hearing of patients with Down syndrome should be optimized to achieve an appropriate language development [29]. This will increase their quality of life, stimulate social interaction and promote autonomy [5]. Establishing a correct diagnosis is essential to provide the appropriate treatment and achieve these goals.

The aim of this study is to describe the prevalence and etiology of SNHL in children with Down syndrome.

## 2. METHODS

We performed a retrospective chart review of all children with DS that are followed at the multidisciplinary Down team of the Antwerp University Hospital (Belgium). Regular ENT visits are integrated in the medical care of these patients as described previously [21]. The medical record of each patient enrolled in the Downteam at September 1st, 2017 was retrospectively reviewed by one author (DSL). Patients were included if they consulted the ENT department at least once. During each ENT visit, the ears were cleared of impacted cerumen and microscopically examined by a pediatric ENT surgeon. The hearing thresholds were determined by qualified pediatric audiologists as reported in a previous study [21]. Hearing loss was classified as conductive, mixed or sensorineural. Patients with unreliable audiometric data and those with conductive or mixed hearing loss were excluded. A patient was considered lost to follow-up and thereby excluded from the database if the most recent audiological information dated from two years ago or before.

For each eligible patient, data were anonymously entered in a database. Demographic information included gender, date of birth, ethnicity, age at diagnosis of the hearing loss, presence of risk factors for hearing loss as defined by the American Academy of Pediatrics and the results of the neonatal hearing screening [30]. Consanguinity of the parents was added to the list of risk factors for hearing loss. Age appropriate audiometric tests were performed taking into account the cognitive ability of the individual child. Results of ABR are reported in dB nHL (normal hearing level). Data from pure tone audiometry are presented as pure tone average over 500, 1000 and 2000 Hz (PTA3) and pure tone average over 500, 1000, 2000 and 4000 Hz (PTA4). The last visit audiogram was compared with the first visit audiogram. The type, severity, symmetry and progression of the hearing loss was described according to the GENDEAF recommendations [31]. Hearing loss severity is graded by the hearing level of the worst ear in unilateral hearing loss and by the hearing level of the best ear in bilateral hearing loss (table 1).

After confirmation of SNHL, an etiological work-up was proposed including a search for congenital infections, genetic testing and MRI as described earlier [13]. When no cause could be identified after a complete etiological work-up, the case was classified as “hearing loss of unknown cause”. Treatment options offered were also included in the database. Descriptive statistics are reported as mean and standard deviation (SD).

## 3. RESULTS

### 3.1 Patient inclusion

On the first of September 2017, 319 patients were in follow-up at the multidisciplinary Down team. There were 291 patients with at least one visit at the ENT department. An overview of the patient flow is presented in figure 1. After reviewing each single patient, 278 patients were excluded. Ninety-six (33.0%) patients presented with conductive hearing loss and 3 patients (1.0%) presented with mixed hearing loss. Among the excluded patients were 3 children who were repeatedly uncooperative during audiometry. Their parents declined ABR under general anesthesia. In addition, the type of hearing loss could not be determined in 4 children due to the absence of bone conduction measurements. Thirteen patients (4.5%) were eligible for inclusion and these comprise the present report.

### 3.2 Patient demographics

Patient demographics are presented in table 2. There were 7 boys and 6 girls with a mean age of 14.4 ± 7.3 years. The average age at the time of hearing loss diagnosis was 9.9 ± 7.3 years. Two separate groups can be distinguished. The first group comprises 8 older patients (cases 1 to 8 as presented in table 3). These patients were examined at the ENT department for the first time after age 7. Patient 1 to 6 were not screened for hearing loss at birth, however their parents did not report any hearing loss. In these patients, onset of hearing loss is unknown and no conclusions can be drawn whether this hearing loss is congenital or postnatal. Case 7 and 8 were born after the introduction of universal neonatal hearing screening in 1998 but no information was available for case 7. Case 8 failed the neonatal hearing screening and can be considered to have congenital hearing loss. The second group includes 5 younger patients that were in follow-up before age 2. Four of them failed the neonatal

hearing screening. Patient 11 passed screening by automatic auditory brainstem responses despite having a cochlear nerve deficiency. The majority was of Caucasian ethnicity. Risk factors for hearing loss were reported in only 1 patient being born premature with a low birth weight. Information on the outcome of neonatal hearing screening was available for 6 patients.

### 3.3 Audiometric data

An ABR was conducted in 5 patients (38.5%) at an average age of 1.5 ± 1.7 years. Four patients presented with single sided deafness (SSD) and absence of ABR responses in one ear (1 left, 3 right). In 8 patients (61.5%) the most recent audiometry was measured with ear inserts or a headset. For 5 patients (38.5%), audiometric data were obtained in free field test conditions. For each patient at least one additional audiogram was available allowing evaluation of hearing loss progression. The average age at the time of the earliest audiogram was 9.3 ± 6.9 years and at the time of the most recent audiogram was 13.7 ± 7.2 years. The average time interval between both audiometries was 4.4 ± 1.7 years. Hearing loss progression was observed in 1 patient with an average annual loss of 5.3 dB over a time span of 4.7 years.

### 3.4 Hearing loss laterality and severity

There were 8 patients (61.5%) with bilateral and 5 patients with unilateral (38.5%) SNHL (table 3). Hearing loss severity is described according to the results of the most recent audiometry. The hearing loss was mild, moderate or profound in respectively 38.5%, 30.8% and 30.8% of the patients. No patient presented with severe hearing loss.

### 3.5 Hearing loss etiology

Data on hearing loss etiology are presented in figure 2. In the older patient group, the majority did not have an etiological work-up because of lack of parental interest. In case 1 and 4, hearing loss was attributed to noise exposure because parents reported repetitive listening to very loud music through head phones. Early presbyacusis could also have played a role in these patients but as with noise trauma, there is no formal proof of this. Genetic testing was performed in 3 (15.8%) patients with bilateral SNHL. One demonstrated a compound heterozygous pathogenic variant in the *GJB2* gene encoding connexin 26. Imaging studies were performed in 7 (53.8%) patients. One patient had a CT scan of the petrous temporal bone and 5 patients had a posterior fossa MRI. One patient was assessed with both imaging techniques. Magnetic resonance imaging showed unilateral cochlear nerve deficiency in all 4 patients with SSD (figure 3). One of these patients with cochlear nerve deficiency also had a history of a congenital cytomegalovirus (CMV) infection. A specific cause explaining the hearing loss could be found in 5 out of 7 patients for whom a standardized work-up was performed [13].

### 3.6 Treatment for hearing loss

A hearing aid was recommended to 4 patients (30.8%) (3 bilateral and 1 unilateral) and was successfully tolerated by 3 of them. Two patients with unilateral SNHL underwent a trial with a bone anchored hearing aid (BAHA) on a softband. Sign language was used by 8 children (61.5%) to support their non-verbal expression.

## DISCUSSION

The prevalence of SNHL was 4.5% in our population of children with DS. In 5/13 (38.5%) cases a diagnosis of congenital SNHL could be confirmed. Our data are in line with those reported by Park et al. [10] who found a 1.8% prevalence of SNHL. In a systematic review, Shott S.R. et al. [7] reported a prevalence of 4% to 20% for sensorineural and mixed hearing loss.

Profound unilateral hearing loss (SSD) was observed in 4 patients. Recent data emphasized the potential negative impact of unilateral hearing loss in speech-language development, speech perception in noise, cognition and behavior [32]. These potentially negative effects may be even more pronounced in children with a cognitive disability such as those with DS and warrant special attention to minimize these unfavorable effects.

Whereas previous studies reported on the prevalence of SNHL in DS patients, they did not provide data on the underlying cause [7,10]. Pathogenic variants in the *GJB2* gene are the most common cause of congenital SNHL and were present in one of our patients with bilateral, moderately severe hearing loss [33]. One patient with SSD had a congenital CMV infection, which is the most common non-genetic cause for hearing loss present at birth. Cochlear nerve deficiency was the most common underlying cause for SSD in our DS patients. Three of them were diagnosed with a profound unilateral hearing loss after a referral from the neonatal hearing screening. One patient who passed the neonatal hearing screening was diagnosed later at the age of 19.2 months. Cochlear nerve deficiency has been reported as the most common cause for congenital SSD in children and may mimic unilateral auditory neuropathy/dyssynchrony [32,34,35].

The inventory of etiological factors of SNHL included excessive noise exposure that is associated with a perceptive dip in the (extended) high frequencies on audiometry. The impact of such a perceptive dip is not reflected in the value of the PTA3. It has a minor influence on the value of the PTA4. These measurements therefore do not accurately demonstrate the severity of the SNHL in those patients.

Our study has several limitations because of its retrospective nature. A complete data set was not available for all subjects. Not all patients were screened for hearing loss at birth and an etiological work-up was performed in only a subset of 7 patients. For example, data on the result of neonatal hearing screening could not be retrieved from the medical records of 6 patients with SNHL and an etiological work-up was not performed in 4 patients with SNHL. In those patients where hearing screening was not performed at birth, it remains uncertain whether the hearing loss is congenital or postnatal. Parental history declining any notion of hearing loss, is unreliable especially in children with cognitive delay.

Secondly, we cannot exclude a referral bias. From 2005 to 2016 there were 500 live births of children with DS in Flanders [36]. The Downteam of the Antwerp University Hospital has a registry of 319 DS patients since 2007. Furthermore, 17 from the 31 live births with DS in 2016 are in follow-up at the Antwerp University Hospital. We therefore believe that our study population fairly represents the population of children with DS in Flanders.

A major strength of this report is that we looked for an etiology underlying the SNHL in patients with DS. Identifying an etiology for (congenital) hearing loss may direct rehabilitation strategies, may allow monitoring for hearing loss progression and can provide parents information regarding the recurrence risk [28]. The DS child with SNHL caused by pathogenetic variants in the *GJB2* gene illustrates this point. The risk for another child with SNHL is 25% even if this child does not present with DS.

According to the type of HL and underlying cause, treatment may be proposed with conventional hearing aids, a bone-anchored hearing aid (BAHA) or cochlear implant [37–41]. Assistive listening devices, for example audio induction loops in classrooms, improve the learning environment of children with Down syndrome and might be a valuable alternative to wearing a traditional hearing aid [42,43]. Speech therapy or sign language can promote language fluency and global word analysis and recognition [44]. In addition, parental education is recommended to improve the child's conditions for hearing [45,46].

## CONCLUSION

Sensorineural hearing loss was present in 4.5% of the children with DS and about 40% was found to be congenital in origin. A definite underlying cause could be identified in 5 out of 7 cases in whom an etiological work-up was performed. Cochlear nerve deficiency was a major cause of single sided deafness. This study's data illustrates the value of an etiological work-up for SNHL in children with DS since this information may be helpful for parental counseling and decision-making regarding treatment.

## CONFLICT OF INTEREST

The authors declare no conflict of interest.

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**LEGEND TO FIGURES AND TABLES**

table 1: hearing loss severity.
table 2: demographic information.
table 3: individual patient details.

(tables are placed on separate pages at the end of this article)

appendix A: fig1.TIFF
appendix B: fig2.TIFF
appendix C: fig3.TIFF
appendix D: figure captions

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Table 1: hearing loss severity.

| | |
|---|---|
| Mild | 20 - 40 dBHL |
| Moderate | 41 - 70 dBHL |
| Severe | 71 - 90 dBHL |
| Profound | > 90 dBHL |

Table 2: demographic information.

| | | | |
|---|---|---|---|
| Number of patients with SNHL (n) | | 13 | 100% |
| Gender | female | 6 | 46.2% |
| | male | 7 | 53.8% |
| Average age (yr.) | at time of study | 14.4 ± 7.4 | |
| | at 1st visit | 8.6 ± 7.1 | |
| | at diagnosis of hearing loss | 9.9 ± 7.3 | |
| Ethnicity | Caucasian | 7 | 53.8% |
| | North African | 2 | 15.4% |
| | Asian | 2 | 15.4% |
| | Hebrew | 1 | 7.7% |
| | Persian | 1 | 7.7% |
| Risk factors | weight < 1500 g or premature < 32 w | 1 | 7.7% |
| | none | 12 | 92.3% |
| Neonatal hearing screening | bilateral pass | 1 | 7.7% |
| | bilateral refer | 4 | 30.8% |
| | unilateral refer | 1 | 7.7% |
| | no data available | 7 | 53.8% |

Table 3: individual patient details.

| No. | Age (yr.) | | | Hearing loss (HL) | | | Etiology | Hearing screening | BERA (dBnHL) | | Pure tone audiometry (dB) | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | at study | 1st visit | diagnosis | symmetry | severity | progression | | | Right | Left | Right | | Left | |
| | | | | | | | | | | | PTA3 | PTA4 | PTA3 | PTA4 |
| 1 | 24.7 | 16.9 | 17.1 | bilateral | mild | no | excessive noise exposure | - | - | - | 30 | 38 | 32 | 38 |
| 2 | 23.2 | 18.2 | 18.2 | bilateral | mild | no | no etiological work-up | - | - | - | 17 | 25 | 17 | 25 |
| 3 | 21.2 | 17.4 | 17.4 | bilateral | mild | no | no etiological work-up | - | - | - | 35 | 35 | 37 | 40 |
| 4 | 20.3 | 13.5 | 13.5 | bilateral | mild | no | excessive noise exposure | - | - | - | 23 | 23 | 13 | 18 |
| 5 | 19.9 | 15.0 | 15.0 | unilateral | moderate | yes | unknown | - | - | - | 62 | 69 | - | - |
| 6 | 19.8 | 13.1 | 19.4 | bilateral | mild | no | no etiological work-up | - | - | - | 40 | 36 | 28 | 26 |
| 7 | 14.3 | 9.0 | 13.8 | bilateral | moderate | no | no etiological work-up | - | - | - | 42 | 44 | 42 | 44 |
| 8 | 12.8 | 7.2 | 7.2 | bilateral | moderate | no | Genetic (*GJB2*) | bilateral refer | - | - | 50 | 53 | 45 | 51 |
| 9 | 9.6 | 0.3 | 0.3 | unilateral | profound | - | cochlear nerve deficiency | bilateral refer | 100 | 80 | 95 | 95 | - | - |
| 10 | 8.8 | 0.1 | 4.4 | bilateral | moderate | no | unknown | bilateral refer | 60 | 60 | 53 | 53 | 48 | 48 |
| 11 | 6.9 | 1.3 | 1.6 | unilateral | profound | - | cochlear nerve deficiency | bilateral pass | 30 | 95 | - | - | 82 | 83 |
| 12 | 2.7 | 0.3 | 1.0 | unilateral | profound | - | cochlear nerve deficiency | bilateral refer | 90 | 40 | 90 | 90 | - | - |
| 13 | 2.6 | 0.2 | 0.3 | unilateral | profound | - | cochlear nerve deficiency + CMV infection | unilateral refer | 95 | 20 | 95 | 95 | - | - |

figure 1: flowchart of patient selection process.

figure 2: hearing loss etiology.

figure 3: axial T2 CISS WI (0,4mm thin slices) demonstrating aplasia of the right cochlear branch of the vestibulocochlear nerve.