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**Vestibular (dys)function in children with
sensorineural hearing loss: a systematic
review.**

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Abbreviations and acronyms

Cosmin = Consensus-based Standards for the selection of health Measurement Instruments,
cVEMP/oVEMP = Collic/Ocular Vestibular Evoked Myogenic Potentials, DVA = Dynamic
Visual Acuity test, HSSC = Horizontal Semicircular Canal, HST = Head Shaking Test, RCT =

Verbecque – Vestibular (dys)function in children with SNHL

Rotational Chair Test, SNHL = Sensory Neural Hearing Loss, v(HIT) = (video) Head Impulse

Test, VOR = Vestibular-Ocular Reflex, VT = Vestibular Test

For Peer Review Only

Abstract

Objective: To provide an overview of the prevalence of vestibular dysfunction in children with SNHL classified according to the applied test and its corresponding sensitivity and specificity.

Design: Data were gathered using a systematic search query including reference screening.

Study sample: Pubmed, Web of Science and Embase were searched. Strategy and reporting of this review was based on the MOOSE (Meta-analysis of Observational Studies in Epidemiology) guidelines. Methodological quality was assessed with the COnsensusbased Standards for the selection of health Measurement INstruments (COSMIN) checklist.

Results: All studies, regardless the applied vestibular test, showed that vestibular function differs significantly between children with hearing loss and normal hearing ($p<0.05$). Compared to caloric testing, sensitivity of the Rotational Chair Test (RCT) varies between 61-80% and specificity between 21-80%, whereas this was respectively 71-100% and 30-100% for collic Vestibular Evoked Myogenic Potentials (cVEMP). Compared to RCT, sensitivity was 88-100% and specificity 69-100% for the Dynamic Visual Acuity test, respectively 67-100% and 71-100% for the (video) Head Impulse Test and 83% and 86% for the ocular VEMP.

Conclusions: Currently, due to methodological shortcoming, evidence on sensitivity and specificity of vestibular tests is unknown to moderate. Future research should focus on adequate sample sizes (subgroups >30).

Introduction

In the past ten years, interest into the prevalence of vestibular dysfunction in children with hearing loss and its assessment has increased drastically.(Christy et al, 2014; Cushing et al, 2009; Maes et al, 2014, Mäki-Torkko & Magnusson, 2005; Rine & Braswell, 2003; Shinjo et al, 2007; Singh et al, 2012; Valente et al, 2011; Wiener-Vacher et al, 2012) Because of the close anatomical relation between cochlea and vestibulum, it is not surprising that children with hearing loss may have some degree of vestibular dysfunction. The motor output of the vestibular system goes to the ocular muscles and spinal cord to serve three reflexes.(Herdman, 2007; Nandi & Luxon, 2008) The vestibular-ocular reflex (VOR) is responsible for gaze stability. The vestibulo-collic reflex (VCR) is responsible for neck stability. Finally, the the vestibulo-spinal reflex (VSR) is responsible for the orientation of the body in gravitational space and contributes to the postural tone. Acquiring sufficient control over the upright posture is necessary to attain different developmental motor milestones.

Children with vestibular dysfunction show delayed development of gross motor milestones such as standing and walking compared to children without vestibular dysfunction. This underlines the great importance of assessment of vestibular function in children.(Angeli, 2003; Nandi & Luxon, 2008) Many authors report that the prevalence of vestibular dysfunction in children with hearing loss ranges from 20 to 85%.(Cushing et al, 2009; De Kegel et al, 2012; Inoue et al, 2013; Singh et al, 2012; Maes et al, 2014) This variability in prevalence of vestibular dysfunction might be related to the different underlying pathologies or the degree of the hearing loss under investigation. Previous reports have shown that vestibular dysfunction most likely occurs in children with sensorineural hearing loss (SNHL), given the embryological and anatomical connection between the cochlea and vestibular end-organs and their shared sensory microstructure and genetics.(Cushing et al, 2008; Rine, 2009) However, conflicting results on the relationship between vestibular loss and both etiology and degree of the hearing loss have been published previously and therefore remain unclear.(Maes et al, 2014; De Kegel et al, 2012; Diepeveen & Jensen, 1968; Brookhouser et al, 1982)

Verbecque – Vestibular (dys)function in children with SNHL

To improve hearing, children with SNHL can receive a cochlear implant (CI). According to Handzel et al (2006) cochlear implantation may potentially cause vestibular injury through direct trauma to the vestibular sensory structures, alteration of the fluid balance within the inner ear, inflammatory reactions, or direct electrical stimulation. However, conflicting results regarding the influence of cochlear implantation on vestibular function in children have been stated.(Jin et al, 2006; Jacot et al, 2009; Thierry et al, 2015) Apart from these individual-related confounding factors for the prevalence of vestibular dysfunction, the choice of the vestibular test to determine the prevalence of vestibular dysfunction might also play a crucial role.(Cushing et al, 2009) In this case, the prevalence of vestibular dysfunction is determined by the diagnostic accuracy of the test under consideration in which sensitivity and specificity are of great importance. Sensitivity can be defined as the ability of a test to correctly classify an individual as 'diseased', whereas specificity refers to the ability of a test to correctly classify an individual as 'disease-free'.(Parikh et al, 2008) To determine sensitivity and specificity, a gold standard or at least a reference test is necessary. In adults, electronystagmography (ENG) or videonystagmography (VNG) during oculomotor tasks, positional testing, rotational chair testing (RCT) and caloric testing is considered to be the gold standard for vestibular testing. In general, vestibular function tests initially developed for adults, are transferred to a pediatric population, which is also the case for the gold standard. However, it remains unclear whether these tests are also suitable for use in this specific population. Due to the nature of vestibular function tests such as testing in a dark room, provoking nystagmus and nausea, they are often not well-tolerated by children and therefore require a child-friendly test protocol and corresponding pediatric normative data.(Maes et al, 2014) Thus, the protocol of the applied vestibular function test may also influence its sensitivity and specificity.

Given the many research available on vestibular dysfunction in children with SNHL and the ongoing development of new assessment tools, it can be stated that this topic has been and still is of great interest to many authors. Therefore it seems necessary to provide an overview on the currently available vestibular tests in literature for children with SNHL (with or without CI), the prevalence of vestibular dysfunction in this population classified according to the applied test

Verbecque – Vestibular (dys)function in children with SNHL

and its corresponding sensitivity and specificity. An answer will be sought to the following questions:

- What vestibular function tests and test protocols (O) are applied in children with SNHL (P)?
- What is the prevalence of vestibular dysfunction (O) in children with SNHL (P) determined by vestibular function tests (I)?
- What is the sensitivity and specificity (O) of different vestibular function tests (I) in children with SNHL (P)?

Methods

This systematic review is reported following the MOOSE (Meta-analysis Of Observational Studies in Epidemiology) guidelines.(Stroup et al, 2000)

Data sources and search strategy

Two search queries were defined: 1) (children OR adolescent) AND ("hearing loss"[mesh] OR "sensorineural hearing impairment" OR "cochlear implantation"[mesh] OR "cochlear implantations" OR "cochlear implant" OR "cochlear implants"[mesh]) AND ("vestibular function tests"[mesh] OR "vestibular testing" OR "vestibular function test") and 2) (children OR adolescent) AND ("hearing loss"[mesh] OR "sensorineural hearing impairment" OR "cochlear implantation"[mesh] OR "cochlear implantations" OR "cochlear implant" OR "cochlear implants"[mesh]) AND ((vestibulum* OR vestibule* OR vestibular*) AND (assessment OR "assessment tool" OR evaluation OR "Outcome Assessment (Health Care)"[mesh])).

Subsequently, the search queries were ran in PubMed, Web Of Sciences and Embase on October 2nd 2014 and updated twice, on September 9th 2015 and May 2nd 2016, covering Medline, Cochrane Database of Systematic Review, Cochrane Central Register of Controlled Trials, ISI Web of Knowledge, Web of Science and the biomedical and pharmacological database of Elsevier. Mesh-terminology was only used in Pubmed. Additionally, all references of the included literature were screened, to ensure that no relevant literature was overlooked.

Screening procedure

Eligibility of the search results was assessed by applying the following inclusion criteria:

1. Participants were children with SNHL (with/without CI). A child was considered to be within the age range of 0-21 years old. Articles combining adults and children needed to present results for children separately from those of adults. Studies combining children with SNHL and children with normal hearing (controls) were included, but studies exclusively consisting of children with normal hearing were not accepted as this was not supported by the search queries.
2. At least one vestibular function test was used to evaluate vestibular function. A vestibular function test was defined as a test that evaluates the VOR or the VCR, such as caloric testing, RCT, vestibular evoked myogenic potentials (VEMP), dynamic visual acuity test (DVA), or video head impulse test (vHIT). Although Computerized Dynamic Posturography is often reported as an assessment tool for the VSR, this assesses far more than solely the VSR. Therefore, articles using this technique were excluded.
3. Studies reported either of the following:
 - prevalence of vestibular dysfunction in the sample under investigation
 - differences in vestibular function between children with normal hearing and children with SNHL (with/without CI) or between different groups of children with SNHL (with/ without CI)
 - comparison of different vestibular function tests in children with SNHL (with/without CI)
 - sensitivity and/or specificity for the vestibular test under investigation
4. Original articles written in English, Dutch, French or German.

The selection of relevant studies was performed by applying the selection criteria on title and abstract (phase 1) and on the full text (phase 2). In case of uncertainties, doubts or when there was no abstract available, these references were screened on full text in phase two. Both phases of screening were performed independently by two researchers (NDB, TM), who both have a Bachelor of Science degree in Rehabilitation Sciences and Physical Therapy. After each

Verbecque – Vestibular (dys)function in children with SNHL

phase, a consensus meeting was held to compare each other's results and to discuss differences. In case of disagreement, the researchers screened the abstract or manuscript a second time and differences were discussed. If consensus could not be reached between the two researchers, a decisive opinion was provided by a third researcher (EV). All steps of the screening procedure are displayed in Figure 1.

Risk of Bias in individual studies

To evaluate the methodological quality of the articles, the Consensus-based Standards for the selection of health Measurement Instruments (COSMIN) checklist was applied.(Mokkink, 2009)

The COSMIN checklist was constructed to assess methodological quality of studies investigating the psychometric properties of assessment tools. The COSMIN contains one box for each of the following psychometric properties: internal consistency, reliability, measurement error, content validity, structural validity, hypothesis testing, cross-cultural validity, criterion validity and responsiveness. Each box comprises questions which can be answered by "excellent", "good", "fair" or "poor". For example, "*Was the sample size included in the analysis adequate?*", potential answers can be "adequate sample size (≥ 100) - excellent", "good sample size (50-99) - good", "moderate sample size (30-49) - fair" or "small sample size (< 30) - poor". The final score of a box is determined by its lowest score. The COSMIN provides an objective assessment of sample size calculations, handling missing data and the applied statistical analyses, as opposed to mere judgements.

For this review, the box regarding criterion validity (Box H) was relevant, since criterion validity refers to the extent to which the vestibular function test under consideration can be compared to another vestibular function test, either the gold standard, e.g. caloric testing, or another vestibular function test that is not considered the gold standard. Risk of bias in individual studies could only be assessed in those articles that investigated sensitivity and specificity of the applied vestibular test or in those that mapped vestibular function with two or more vestibular function tests, so that sensitivity and specificity could be calculated. In general, the caloric test is considered to be the primary reference test for vestibular function testing.

Therefore, sensitivity

Verbecque – Vestibular (dys)function in children with SNHL

and specificity of this test was not calculated and therefore not reported but only used as a reference test to calculate sensitivity and specificity for the other vestibular tests.

The articles were scored independently by two researchers (NDB or TM and EV) who were blind for each other's evaluation until the consensus meeting. To determine the inter-rater

reliability a Cohen's Kappa was calculated ($K = \frac{1-Pre(e)}{Pr(a)-Pr(e)}$).

Data extraction

For each article included in this review the following data were extracted: study population characteristics (children with SNHL: age range, number of children, etiology of the hearing loss, degree of hearing loss, presence of cochlear implant; children with normal hearing: age range and number of children), the applied vestibular function test and its methodology, the applied reference and cut-off values used to determine the presence of vestibular dysfunction, the prevalence of vestibular dysfunction, the drop-outs for each test, and the sensitivity and specificity of the vestibular function test. When sensitivity and specificity were not reported in the included literature and the required raw data were available, the authors calculated sensitivity and specificity percentages by applying the following formulas:

$$sensitivity = \frac{\text{number of true positives}}{\text{number of true positives} + \text{number of false negatives}} \times 100$$

$$specificity = \frac{\text{number of true negatives}}{\text{number of true negatives} + \text{number of false positives}} \times 100$$

Level of evidence

The criteria of the Cochrane Collaboration Back Review Group(van Tulder, 2003) were used to determine the level of evidence, but implemented as by Saether and colleagues (2014).(Saether et al, 2014) Based on the number of studies, the methodological quality and the consistency of findings, the level of evidence (strong, moderate, limited, unknown, conflicting) was assigned (Appendix 3).

Results

Study selection

The database searches revealed a total of 1704 studies. After deduplication 1086 articles were screened on title and abstract. After this screening procedure 74 articles were selected for a more in-depth second screening (including reference screening, $n=3$), leading to 38 relevant studies, of which data were extracted. The literature collection procedure is presented in Figure 1.

Risk of Bias in individual studies

Methodological quality assessment was relevant for 21 of the 38 included studies. Their quality for criterion validity (Box H) varied from poor to fair (Table 1). The degree of agreement between the two independent raters was very good ($k=0.70$).

Data extraction

Participants

The populations of all included studies consisted of children with SNHL (with/without CI) within the age range of 3 months to 21 years old and 17 studies investigated children with normal hearing between 3 months and 18 years old. Table 2 contains the population characteristics of the included studies. Five studies investigated vestibular function in a sample of children with congenital hearing loss (Jafari & Malayeri, 2011; Kaga et al, 1981; Kaga et al, 2008; Zagólski, 2007; Inoue, 2013) and two studies in children with acquired hearing loss due to meningitis (Cushing et al, 2009; Cyr et al, 1985). The other 31 studies used either a sample with unknown etiology (1 study), unreported etiology (9 studies) or a heterogeneous sample. The etiologies of the hearing loss in the samples under investigation in the individual studies are presented in Table 2. In Figure 2 the number of recruited children for each vestibular function test are graphically presented, classified according to their etiology.

Vestibular function tests

In Table 2 the applied vestibular function tests are shown per individual study and the corresponding population in which they were administered. The following vestibular function tests were applied: the caloric test (17 studies), the RCT (20 studies), the DVA (4 studies), the (v)HIT (7 studies), the head shaking test (HST, 1 study), the collic VEMP (cVEMP, 21 studies)

Verbecque – Vestibular (dys)function in children with SNHL
and the ocular VEMP (oVEMP, 3 studies). In the next paragraphs, for each test, the applied protocol, the prevalence of vestibular dysfunction and sensitivity and specificity are presented.

1 Caloric Test
Protocol

As shown in Table 2, a total of 1184 children, of which 1107 children with SNHL and 77 children with normal hearing, were tested in 17 studies with the caloric test.(Brookhouser et al, 1982; Cushing et al, 2009; Cushing et al, 2008; Diepeveen & Jensen, 1968; Hallmo et al, 1986; Holderbaum et al, 1979; Inoue et al, 2013; Jacot et al, 2009; Jin et al, 2006; Kaga et al, 2008; Lisboa et al, 2005; Sandberg & Terkildsen, 1965; Shinjo et al, 2007; Suonpaa, 1988; Thierry et al, 2015; Tribukait et al, 2004; Zagólski, 2007) The caloric test serves as a low frequency canal test that maps the sensitivity of each horizontal semicircular canal (HSCC). During the caloric test, the subject is positioned supine with the neck flexed 30°. A thermal gradient across the HSCC of one ear is created by the irrigation of the external auditory meatus using warm (44°C) and cool (30°C) water or warm (42°C) and cool (20°C) air to induce endolymphatic flow.(Brookhouser et al, 1982; Cushing et al, 2008; Cushing et al, 2009; Diepeveen & Jensen, 1968; Sandberg & Terkildsen, 1965; Hallmo et al, 1986; Suonpaa, 1988; Thierry et al, 2015; Tribukait et al, 2004; Lisboa et al, 2005) A normal reaction is that cool temperature induces a nystagmus towards the contralateral side. After warm temperature irrigation the opposite response occurs.(Herdman, 2007) When no reaction can be elicited by warm and cool water, ice water is used to identify areflexia.(Cushing et al, 2008; Cushing et al, 2009) However, four authors used ice water (4 to 5°C) exclusively and three reported application of water at 15-20°C.(Kaga et al, 2008; Shinjo et al, 2007; Holderbaum et al, 1979; Inoue et al, 2013; Diepeveen & Jensen, 1968; Sandberg & Terkildsen, 1965; Zagólski, 2007) With the original protocol by Fitzgerald-Hallpike, alternate bithermal water calorics are applied. However, this technique has shown to be uncomfortable for children, leading to excessive subject movement and a virtually unreadable recording.(Cyr et al, 1985) Therefore, some authors replaced the alternate bithermal calorics with simultaneous binaural bithermal stimulation.(Brookhouser et al, 1982; Cyr et al, 1985; Cushing et al, 2008; Cushing et al, 2009; Hallmo et al, 1986) This closed-

Verbecque – Vestibular (dys)function in children with SNHL

loop irrigation system improved the child's tolerance during the procedure in contrast to the original procedure. In adults, the eye movements as a response to irrigation are measured using ENG or VNG. When ENG is applied, the corneo-retinal potentials are measured using electrodes, whereas VNG directly measures movements of the pupils using infrared video goggles. These techniques are applied in children as well, although several authors, especially when assessing young children, report the duration of the nystagmus or the number of beats of the nystagmus as an outcome measure (directional preponderance – Formula 1) rather than the presence of canal paresis (Jonkees' formula – Formula 2) or the slow phase velocity of the eye movements. (British Society of Audiology, 2010) Directional preponderance indicates that the nystagmus response is greater in one direction as compared to the other. (British Society of Audiology, 2010) Canal paresis expresses a weakness in the induced nystagmus following caloric stimulation from one ear as compared to those obtained from the opposite ear. (British Society of Audiology, 2010)

$$\text{Directional preponderance} = \frac{(\text{warm right ear cold left ear}) - (\text{warm left ear cold right ear})}{(\text{warm right ear cold right ear warm left ear cold left ear})}$$

(Formula 1*)

$$\text{Jonkees' formula} = \frac{(\text{warm right ear cold right ear}) - (\text{warm left ear cold left ear})}{(\text{warm right ear cold right ear warm left ear cold left ear})}$$

(Formula 2*)

* formulas were derived from the recommended procedures published by the British Society of Audiology. Specifics on methodology applied in the individual studies for the caloric test are presented in Table 3.

Prevalence

Raw values for prevalence of vestibular (dys)function are shown in Appendix 1. One study compared the prevalence of vestibular loss between children who are deaf and controls. The prevalence of unilateral or bilateral vestibular loss in children who are deaf (54%) and controls (7%) was significantly different. (Suonpaa, 1988) However, no significant difference was found between the mean maximal eye velocity of the slow component of the nystagmus in children with SNHL and with normal hearing. (Suonpaa, 1988)

Verbecque – Vestibular (dys)function in children with SNHL

When integrating the results of different studies, 51.3% of the children with SNHL had normal results on the caloric test, whereas 10.8% showed unilateral loss, 8.3% showed bilateral loss, 27.8% loss without specifications on the affected side(s) and 1.1% dropped out. These percentages tend to shift towards higher prevalence of vestibular loss when the degree of the hearing loss is taken into account, as shown in Figure 3.

Quite large differences in prevalence have been reported in homogeneous groups of children with SNHL regarding the etiology of the hearing loss. *Cushing et al (2009)* reported that 62.5% of children with acquired SNHL had bilateral vestibular loss whereas 37.5% had unilateral loss, none of the children had normal caloric results. Regarding congenital SNHL, *Inoue et al (2013)* showed that 19% had bilateral loss and 17,3% had unilateral loss, whereas *Kaga et al (2008)* reported 35% unilateral loss and 50% bilateral loss and *Zagolski (2007)* 66.7% with vestibular loss without specifications on the affected side(s). According to *Tribukait et al (2004)*, 46% of children with acquired or congenital hearing loss had normal caloric responses, whereas 24% had asymmetric responses and 15% had bilateral hypofunction and 15% areflexia. Regarding vestibular function in children with CI's, *Jin et al (2006)* showed that before cochlear implantation 40% of the children had normal caloric responses whereas 20% had no responses while *Shinjo et al (2007)* reported that 15% of CI candidates had normal caloric responses and 40% had no responses at all (Appendix 1). *Cushing et al (2008)* found no significant differences between the implanted and the non-implanted side.

Sensitivity and specificity

In all 11 studies involving the caloric test, it was used as the gold standard. Therefore, no sensitivity and specificity for the caloric test is reported.

2 Rotational Chair Test (RCT)

Protocol

The RCT was administered in 1144 children of which 872 children with hearing loss and 272 children with normal hearing in 20 studies (Table 2). (Backous & Quigley, 2000; Bergholtz et al, 1977; Christy et al, 2014; Cushing et al, 2009; Cushing et al, 2008; Cyr et al, 1985; De Kegel et al, 2012; Inoue et al, 2013; Jacot et al, 2009; Janky & Givens, 2015; Kaga et al, 2008; Kaga et

Verbecque – Vestibular (dys)function in children with SNHL

al, 1981; Maes et al, 2014; Mäki-Torkko & Magnusson, 2005; Masuda & Kaga, 2014; Potter & Silverman, 1984; Rine & Braswell, 2003, Selz et al, 1996; Shinjo et al, 2007; Tribukait et al, 2004) The RCT serves as a high frequency canal test that maps the sensitivity of each HSSC. For RCT, the subject is positioned in a motorized chair with a head restraint. The rotational axis goes through the center of the head. Usually, the test is performed in the dark and the subject closes the eyes while performing mental tasks. Different rotational tests are performed. Both a sinusoidal harmonic acceleration (SHA) and step velocity protocols have been reported.(Herdman, 2007; Maes et al, 2014; Rine & Braswell, 2003)

During SHA the chair is accelerated to a peak velocity of 50°/s to 100°/s across frequencies varying from 0.01 Hz to 5 Hz both clockwise and counter-clockwise. When step velocity protocols are applied, a rapid acceleration of the chair to a constant rotational velocity is followed by a deceleration of the chair to rest. The RCT assesses the VOR using gain, phase and asymmetry outcome variables in case of SHA and gain, time constant and symmetry in case of step velocity. The gain is a measure of VOR performance calculated by the velocity of the correcting eye movement divided by the velocity of the head. The phase comprises the angle of the response which is a representation of the timing difference between the eye and the chair velocity. Asymmetry stands for the differences between VOR gain left and right. The time constant is the time that is required for the nystagmus to decrease to a predefined percentage of its original strength.(Herdman, 2007) However, some authors also reported the duration of the nystagmus and the number of beats of the post- and/or per-rotatory nystagmus. *Jacot et al (2009)* also used the Off Vertical axis rotation (OVAR), a subtest of the RCT, which evaluates otolith responses and more specifically the utricle. *Christy et al (2014)* used the modified Emory Clinical Vestibular Chair Test (m-ECVCT), which is a clinical version of the RCT.(Christy et al, 2014) During the m-ECVCT, there is no sinusoidal acceleration but a rotation of 0.5 Hz for 30 seconds to either side with a pause in between to evaluate nystagmus via infrared camera goggles.(Christy et al, 2014) Specifics on the applied RCT methodology and references to applied pediatric normative values are shown in Table 4. Numeric reference

Verbecque – Vestibular (dys)function in children with SNHL
values for gain, phase and asymmetry using the SHA are presented in Table 5.(Cyr et al, 1985; Maes et al, 2014; Selz et al, 1996)

Prevalence

Lower gain values were reported for the children with hearing loss compared to controls by *Maes et al (2014)*. This difference was only significant ($p<0.002$) for the lowest test frequency 0.01 Hz. *Cyr et al (1985)* found significant differences for gain ($p<0.001$), phase ($p<0.01$) and symmetry ($p<0.001$) between children with post-meningitic hearing loss and with normal hearing.

Raw values for prevalence of vestibular (dys)function are shown in Appendix 1. In general, 55% of the children with SNHL had normal RCT results, whereas 9.7% showed unilateral loss, 6.8% showed bilateral loss, 25.7% loss without specifications on the affected side and 2.9% dropped out.

Similar to the caloric test, these percentages tend to shift towards higher prevalence of vestibular loss when the degree of the hearing loss is taken into account (Figure 3). At 0.05 Hz rotation, gain values were significantly different for the profound hearing losses compared to moderate hearing loss ($p=0.035$).

Different percentages for prevalence of vestibular loss are reported in different etiologies. *Cushing et al (2009)* stated that only 12.5% of children with post-meningitic SNHL had normal RCT responses and 87.5% had vestibular loss, when the RCT was performed at $< 2\text{Hz}$ in the dark or at $> 2\text{Hz}$ in a light room. Regarding congenital hearing loss, normal responses varied between 32.8-79.8%, unilateral loss between 1.5-5%, bilateral loss between 19-25% and 67.2% had loss without specifics on the affected side(s).(Inoue et al, 2013; Kaga et al., 2008; Kaga et al, 1981; Shinjo et al, 2007) The latter authors used step velocity protocols.

Furthermore, significantly decreased gain values ($p=0.020$) were registered for children with bilateral CI's at 0.05 Hz rotation contrary to children with a unilateral or no CI.(Maes et al, 2014)

Comparing children with hearing loss before and after CI, *Jacot et al (2009)* described a significant decrease of the canal VOR in 17% of the cases (no values available).

Sensitivity and specificity

Verbecque – Vestibular (dys)function in children with SNHL

Sensitivity and specificity values are presented in Table 6. The m-ECVCT had a sensitivity of 63% and a specificity of 100%. (Christy et al, 2014) Using SHA protocols sensitivity and specificity were 80% compared to caloric testing. (Cushing et al, 2008) When step velocity protocols were applied, sensitivity and specificity in comparison to caloric testing varied respectively between 61-66% and 21-73%. (Inoue et al, 2013; Kaga et al, 2008; Shinjo et al, 2007) In 7 other studies the RCT was used as a reference test.

3 (Horizontal) Dynamic Visual Acuity Test (hDVA) *Protocol*

The horizontal DVA (hDVA) was performed by 217 children of which 74 children with hearing loss and 143 with normal hearing in four studies. (Christy et al, 2014; Janky & Givens, 2015; Martin et al, 2012; Rine & Braswell, 2003) Two types of hDVA need to be distinguished: a traditional or bedside hDVA and a computerized hDVA. To perform the bedside hDVA, the subject sits on a chair 3 meters from the Lea symbols chart placed at eye level. Lea symbols are a series of pediatric symbols that are designed specifically for children who do not know how to read the letters of the alphabet. During the test, the neck is flexed 30° and the head is passively moved at 2 Hz in the yaw plane. (Rine & Braswell, 2003) During the computerized hDVA the subject either moves the head actively or the subjects' head is moved passively by the investigator while wearing a rate sensor (InterSense Inertia Cube) on the head that registers the head velocities. If these head velocities exceed 120°/sec, an optotype flashes, but for no longer than 75 msec. (Janky & Givens, 2015) The test ends when the subject misses 3 of 5 optotypes on a line. Results of visual acuity measured with the head stationary are compared to results of visual acuity with the head in movement. (Christy et al, 2014; Herdman, 2007; Rine & Braswell, 2003) More than two lines difference between static and dynamic visual acuity indicates the presence of vestibular loss. (Martin et al, 2012; Rine & Braswell, 2003; Christy et al, 2014)

Prevalence

Raw values for prevalence of vestibular (dys)function are shown in Appendix 1. Normal hDVA results were found in 57.9-84.4% of the cases and Christy reported 26.3% with unilateral loss

Verbecque – Vestibular (dys)function in children with SNHL

and 15.8 % bilateral loss, whereas Martin found 15.6% of his sample to show loss without specifications of the side. Regarding the degree of the hearing loss, 15.6% of the participants with SNHL had poor hDVA, of which all with profound SNHL.(Martin et al, 2012) Children with SNHL and bilateral vestibular loss performed significantly ($p<0.008$) poorer on the hDVA than children with SNHL without vestibular loss, identified with the RCT.(Rine & Braswell, 2003) Janky & Givens (2015) reported a significant moderate correlation between vestibular loss as measured by the active DVA scores ($R^2=0.381$, $p=0.008$) and the passive DVA scores ($R^2=0.484$, $p=0.003$) and specificity

Table 6 provides an overview of the sensitivity and specificity values for the DVA. The sensitivity of the DVA test ranges from 88% to 100% and its specificity ranges from 69% to 100% compared to the RCT (and the cVEMP).(Christy et al, 2014; Rine & Braswell, 2003) For the RCT as a reference test similar protocols were applied.

4 (Video)Head Test Impulse test ((v)HIT)
Protocol

During the clinical HIT, the subject tries to keep the eyes on a visual target while its head is unpredictably and quickly turned by the investigator from the center to the left (3 times) and the right (3 times) at an amplitude of 5° to 10°. The neck of the subject is flexed 30°, assessing the HSCC. The HIT is considered positive if at least 2 corrective saccades out of 6 turns have been seen.(Christy et al, 2014) The clinical HIT was administered in 137 children of which 114 children with hearing loss and 23 with normal hearing in 6 studies.(Backous & Quigley, 2000; Christy et al, 2014; Jacot et al, 2009; Lisboa et al, 2005; Mäki-Torkko & Magnusson, 2005; Thierry, 2015) During the vHIT the subject sits 1m from the visual target wearing goggles with a high speed infrared camera, which records the eye movements and a built-in accelerometer to record the head movements. The examiner stands behind the subject and delivers randomized head impulses in a specific plane while the subject keeps the eyes on the visual target. Thus, the HSCC, posterior and anterior SCC (PSCC, ASCC) can be assessed separately.

Verbecque – Vestibular (dys)function in children with SNHL

Approximately 20 acceptable head impulses of each participant are to be recorded. Eleven children with hearing loss and twelve children with normal hearing performed the vHIT.

Prevalence

Prevalence values for vestibular (dys)function are presented in Appendix 1. Normal HIT results were found in 64-100% of the children with severe to profound SNHL, including CI candidates. (Backous & Quigley, 2000; Lisboa et al, 2005; Mäki-Torkko & Magnusson, 2005) Janky and Givens (2015) reported that respectively 55% (HSCC), 32% (PSCC) and 50% (ASCC) of the children with a CI had normal vHIT gains. These vHIT gains are significantly and strongly positively correlated ($p < 0.001$) with RCT gains for all frequencies: 0.02Hz ($r = 0.765$), 0.08Hz ($r = 0.848$), 0.16Hz ($r = 0.830$), 0.32Hz ($r = 0.774$).

Sensitivity and specificity

Values for sensitivity and specificity are presented in Table 6. For the clinical HIT sensitivity of 75% and specificity of 91% compared to combined RCT and cVEMP results are reported. (Christy et al, 2014; Mäki-Torkko & Magnusson, 2005) The sensitivity and specificity of the vHIT varies depending on the canal under investigation. For the HSCC sensitivity was 100% and specificity 100%, for PSCC respectively 67% and 71% and for the ASCC 91% and 92% compared to the RCT. (Janky & Givens, 2015)

5 Head Shaking Test

Protocol

During HST, the subject is asked to close the eyes while wearing Frenzel goggles. Subsequently, the head is pitched down 30 degrees by the investigator and then oscillated 20 times horizontally. These Frenzel goggles consist of the combination of magnifying glasses and a lighting system that is used when the subject is tested in the dark enhancing detection of nystagmus. Subsequently, the subject opens the eyes, while the therapist observes the presence of nystagmus. In case of vestibular dysfunction, nystagmus is present immediately after this procedure. (Herdman, 2007) The HST was performed in 11 children with SNHL. (Backous & Quigley, 2000)

Prevalence

Verbecque – Vestibular (dys)function in children with SNHL

Backous (2000) reported that 80% of their CI candidates have normal HST responses.

Sensitivity and specificity

No values could be calculated.

6 Collic Vestibular Evoked Myogenic Potentials test (cVEMP)
Protocol

A total of 1358 children, of which 828 children with hearing loss and 530 with normal hearing, were assessed with the cVEMP in 21 individual studies.(Christy et al, 2014; Cushing et al, 2009; Cushing et al, 2008; De Kegel et al, 2012; Inoue et al, 2013; Jacot et al, 2009; Jafari & Malayeri, 2011; Janky & Givens, 2015; Jin et al, 2006; Jin et al, 2008; Kaga et al, 2008; Maes et al, 2014; Shall, 2009; Shinjo et al, 2007; Singh et al, 2012; Thierry et al, 2015; Tribukait et al, 2004; Xu et al, 2015; Xu et al, 2014; Zagólski, 2007; Zhou et al, 2009) The cVEMP assesses function of the saccule and the inferior vestibular nerve. Air or bone conducted stimuli are presented monaurally thereby stimulating the saccule and traversing via the inferior to the vestibular nucleus in the brain stem. Via the medial vestibulospinal tract evoked impulses are sent to the neck and recorded from the ipsilateral sternocleidomastoid muscle (SCM), evaluating the recorded EMG signals (amplitude, P1/N1 latency).(Herdman, 2007; Maes et al, 2014; Uchino, 1997) Specifics on the different applied protocols and references for applied pediatric normative data for cVEMP are shown in Table 7. Pediatric normative values are shown in Table 5, which are necessary for diagnostic purposes as shorter P1 and N1 latencies and larger interpeak amplitudes have been found in children compared to adults.(Maes et al, 2014; Janky and Givens, 2015)

Prevalence

Raw values for prevalence of vestibular (dys)function are shown in Appendix 2. In general, 52.1% of the children with SNHL had normal results on the caloric, whereas 4.4% showed unilateral loss, 8.7% showed bilateral loss, 29.9% loss without specifications on the affected side(s) and 4.9% dropped out. These percentages tend to shift towards higher prevalence of vestibular loss when the degree of the hearing loss is taken into account as shown in Figure 3. Within the group of children with profound SNHL, whose cVEMP response could still be

Verbecque – Vestibular (dys)function in children with SNHL

induced, a deviant cVEMP response was present when a significantly increased threshold ($p < 0.01$) and a significantly decreased amplitude ($p < 0.05$) were identified. (Xu et al, 2015; Zhou et al, 2009) *Maes et al (2014)* reported that 33% of the children with profound hearing loss show absent responses, which is significantly higher than in moderate hearing loss ($p = 0.019$). (Maes et al, 2014)

In children who were CI candidates, 12-50% of the children had normal cVEMP responses as shown in Appendix 2 (Jin et al, 2006; Shinjo et al, 2007). Children with SNHL showed normal cVEMP responses in 46.7%-100% of the cases, whereas 15.6% to 83% of the children wearing a CI showed normal cVEMP responses. *Maes et al (2014)* found a significantly higher absence rate of 67% for children with bilateral CI compared to children without a CI using the cVEMP. (Maes et al, 2014) After implantation, 50-91.7% of the children had vestibular loss with the device on and 69.2-100% of children showed vestibular loss with the device off. (Jin et al, 2006; Xu et al, 2014) *Jacot et al (2009)* added that a significant decrease (no values available) of the otolith VOR was observed in 12.7% of the cases after cochlear implantation with prior vestibular function, whereas this was 16.7% in the study of *Thierry et al (2015)*.

Tribukait et al (2004) reported a significant but weak correlation between the caloric sum (sum of nystagmus responses to 44°C and 30°C), and the cVEMP amplitude ($r = 0.35-0.40$, $p < 0.05$) and a significant and moderate correlation between the caloric ratio (the difference of nystagmus responses to 44° C and 30° C between the left and the right ear) and the cVEMP ratio ($r = 0.60$, $p < 0.01$).

Sensitivity and specificity

All children with normal hearing showed clear cVEMP responses. (Christy et al, 2014; De Kegel et al, 2012; Jafari & Malayeri, 2011; Janky & Givens, 2015; Jin et al, 2006; Maes et al, 2014) Compared to the caloric test (8 studies), sensitivity for the cVEMP varied between 71-100% and specificity between 30-100% in children with SNHL. Compared to the RCT (4 studies) values between 48-100 for sensitivity were calculated and between 78-100 for specificity. All sensitivity and specificity results are presented in Table 6.

Verbecque – Vestibular (dys)function in children with SNHL

7 Ocular Vestibular Evoked Myogenic Potentials (oVEMP)
Protocol

Ocular VEMP was administered in 3 studies consisting of 137 children of which 85 with hearing loss and 52 with normal hearing.(Janky & Givens, 2015; Xu et al, 2015; Xu et al, 2014) The oVEMP is thought to assess the utricle and the superior vestibular nerve. Similar to the cVEMP, tone bursts are presented in the ears, except the utricle is stimulated and the motor output in the form of EMG signals of the contralateral inferior oblique muscle underneath the eye are recorded instead of the SCM.

Prevalence

Fifty-eight percent of the children with SNHL had normal oVEMP results, whereas all children with normal hearing show normal oVEMP results, which differ significantly.(Xu et al, 2015; Xu et al, 2014) Raw values for prevalence of vestibular (dys)function are shown in Appendix 2. Overall, normal oVEMP results were found in 63.5% of the children with SNHL and 45.5% of the children wearing a CI. When the implant was turned on, Xu reported normal oVEMP results in only 12.9% and 19.2% with the implant turned off.

Sensitivity and specificity

Sensitivity and specificity for oVEMP are shown in Table 6. Compared to cVEMP, sensitivity of oVEMP ranges between 83-91% and specificity between 86-95%. When the RCT is used as a reference test, sensitivity was 83% and specificity 86%.

Level of evidence

The levels of evidence regarding the sensitivity and specificity for each test are presented in Appendix 3. Moderate evidence was found for the RCT, cVEMP and DVA. Limited evidence was found for the (v)HIT and the degree of evidence remains unknown for the HST and oVEMP.

Discussion

The aim of this study was to provide an overview on the currently available vestibular function tests in literature for children with SNHL (with or without CI), to report the prevalence of

Verbecque – Vestibular (dys)function in children with SNHL

vestibular dysfunction in this population classified according to the applied test and to provide information on the corresponding sensitivity and specificity of each test. Seven vestibular function tests were identified in literature. The prevalence of vestibular dysfunction clearly depends on both population-related factors and on the applied test. Most frequently reported vestibular function tests were the caloric test, RCT and the cVEMP. However, very variable results were found for their sensitivity and specificity, suggesting these tests are rather complementary than potential substitutes for each other. The level of evidence regarding the sensitivity and specificity of the identified vestibular tests was found to be unknown to moderate, due to methodological shortcomings. Two main reasons for poor methodological quality were identified: the small sample size ($N < 30$) and the lack of adequate statistical analyses. When no correlations were calculated in case of continuous variables or sensitivity and specificity in case of dichotomized variables the study had to be scored poor, if they did, the score excellent was assigned. Fair quality was mainly related to the uncertainty about how missing data were handled.

The main goal of vestibular testing in children with SNHL is to differentiate between normal vestibular function and vestibular dysfunction, thereby providing a prevalent estimation of 0-100%. To do so, the diagnostic accuracy of the test under consideration is of great importance, which is related to the sensitivity and specificity of that test. Unfortunately, these statistical measures were only reported by two authors. Therefore, when not available, they were calculated based on the raw data presented in the original studies. Ideally, both sensitivity and specificity amount to 100%, which is, in most cases, impossible to achieve. The diagnostic accuracy can be influenced by population-related aspects such as the availability of normative data, the degree or the etiology of the hearing loss, but also by more test-related aspects such as the applied protocols and the functions or structures that are being assessed.

Population-related confounding factors

To determine the accuracy of an assessment tool, the first step comprises the evaluation of this tool in a non-symptomatic control group, in this case normal hearing children. In the control groups no vestibular dysfunction was reported, which indirectly provides information on the

Verbecque – Vestibular (dys)function in children with SNHL

specificity of the applied tests. Also, vestibular function in children with SNHL differs significantly from the vestibular function in children with normal hearing, which also provides preliminary results on diagnostic accuracy.

Another important question when it comes to pediatric normative values, concerns the developmental process of the function under investigation.(Maes et al, 2014; Janky & Givens, 2015) The vestibular system is morphologically complete at birth but its functionality continues to mature until approximately 15 years of age, depending on the investigated vestibular reflex.(Morlet 2013; Nandi & Luxon, 2008). Thus, possible age effects that may confound the test results should always be taken into account.(Maes et al, 2014) Most studies included in this review investigated horizontal VOR in children of different ages but considered them as one group, suggesting the horizontal VOR does not undergo developmental changes. The influence of age with regard to horizontal VOR function was assessed by *Rine & Braswell (2003)* through dynamic visual acuity, by *Janky & Givens (2015)* through vHIT gain values and by *Maes et al (2014)* through SHA gain, phase and asymmetry values in their children with normal hearing. All three studies revealed no age-related changes regarding horizontal VOR function, thereby confirming this hypothesis.(Janky & Givens, 2015; Hamilton et al, 2015; Rine & Braswell, 2003) This is an interesting finding as Mortlet (2013) stated that outcome variables are prone to maturational effects: saccades are inaccurate until age 2, speed of the slow component and the frequency of beats stabilize after 6 to 12 months of age, otolith VOR tends to change when children learn to walk, SHA gain decreases between two months and 11 years of age, but SHA phase remains stable.(Morlet 2013) These records suggest that especially in the first years of life, prominent changes do occur in VOR function. Also, *Casselbrandt et al (2010)* stated that “changes in gain may be related to maturation of attentional mechanisms as part of the overall process of maturation of the vestibulo-ocular system because mental alertness plays a key role in VOR function”.(Casselbrandt et al, 2010) Thus, the child’s performance or potential differences in outcome variables will depend upon the amount of attention that is needed to perform the requested task. It can therefore be hypothesized that the lacking age effects as reported by the authors included in this review, might be due to the ages of the included children

Verbecque – Vestibular (dys)function in children with SNHL

and the requested tasks: DVA in 3- to 15-year-old children, vHIT in 6- to 15-year-old children and SHA in 4- to 12-year-old children. Although *Janky & Givens (2015)* mapped vertical VOR (ASCC and PSCC) with the vHIT, no records on maturation of vertical VOR are currently available.

Regarding VCR, assessed with cVEMP, age effects have been stated from preschool years through adolescence and were also reported by the included studies in this review. These findings suggest that depending on the vestibular reflex that is being assessed and the outcome variables, age-specific reference values are in order. Only ten of the included studies (26%) reported actual pediatric normative data. Although databanks of normative data for children are (still) being collected, especially as a function of age, further expansion of these age-specific pediatric normative data seems imperative.

The degree of the hearing loss seems to be related to vestibular (dys)function. As shown in Figure 3, children with severe to profound hearing loss present with higher percentages of vestibular loss. (Brookhouser et al, 1982; Maes et al, 2014) This can be explained by the close anatomical relation between cochlea and vestibulum. Similar reasoning can lead to the assumption that congenital malformations of the inner ear and meningitis that damaged both vestibulum and cochlea, would show larger prevalence for vestibular dysfunction. (Maes et al, 2014) Some authors indeed have reported that children with congenital syndromal and acquired hearing loss more often show vestibular dysfunction. (Cushing et al, 2009; Inoue et al, 2013; Kaga et al, 1981; Kaga et al, 2008; Shinjo et al, 2007) However, based on the very heterogeneous samples in the included studies, evidence regarding the relationship between etiology of the hearing loss and the presence of vestibular loss remains inconclusive. Future research should focus on the influence of etiology or a meta-analysis performed on the existing literature on this topic might provide new insights as well.

Diagnostic accuracy of vestibular function tests

A prevalence of vestibular dysfunction varying between 0-100%, leads to lower diagnostic accuracy, which is reflected by sensitivity and specificity of the applied test. When caloric stimulation is used as a reference test, sensitivity varies between 61-80% and specificity

Verbecque – Vestibular (dys)function in children with SNHL

between 21-80% for the RCT and respectively 71-100% and 30-100% for cVEMP. When the RCT is used as a reference test, sensitivity varies between 88-100% and specificity between 69-100% for the DVA, respectively 67-100% and 71-100% for the (v)HIT and 83% and 86% for the oVEMP. This variability in sensitivity and specificity can be explained by the applied protocols and the anatomical structures that are being assessed with the applied vestibular function tests.(Maes et al, 2014)

Protocols of vestibular tests

Regarding the **caloric test**, older studies tend not to report specifics on the applied protocol whereas in more recent studies the standard protocol has been adjusted into a closed loop binaural bithermal caloric stimulation to make this test less invasive for (young) children. Water stimulation is being reported more frequently than air stimulation. It seems that agreement exists on the usefulness of the caloric test as it is often addressed as the gold standard for vestibular function testing.

For the **RCT**, a distinction can be made between SHA and step rotation. But even within these two specific test modalities, considerable differences tend to exist as shown in Table 4. It seems that each research center has its own protocol and pediatric normative data. Due to these large methodological differences, comparison of results obtained by different research groups therefore becomes quite difficult. This raises the question whether, at the moment, this test can be used as a gold standard for vestibular assessment. Nevertheless, the RCT is very-well tolerated by children, even by infants, even though testing is performed in a dark room.

Regarding **cVEMP**, the protocols, shown in Table 7, have been described clearly and only small differences were identified. Usually, air conducted cVEMP is applied with either tone bursts or clicks, for both of which pediatric reference values are available.

The protocols for **hDVA**, **(v)HIT**, **oVEMP** and **HST** are clearly described in literature. However, some specific adjustments were made to make these tests more child-friendly, e.g. Lea Vision Chart for hDVA. However, these tests were only investigated in a limited number of studies, thereby limiting generalizability of prevalence values and diagnostic accuracy.

Assessment of vestibular structures

The variability in diagnostic accuracy can be explained by the vestibular structures that are being assessed with particular tests. For example, the HSCC and the superior branches of the vestibular nerve can be tested by the Caloric test, RCT, DVA, (v)HIT and HST, the utricle with OVAR and oVEMP whereas the saccule, innervated by the inferior branches of the vestibular nerve, can only be evaluated by the cVEMP. The otoliths are sensitive for linear accelerations, whereas the HSCC is sensitive to horizontal rotational acceleration. This makes that these anatomical structures have different functions. None of the tests assess the entire vestibular apparatus. (Herdman, 2007) As could be expected, *Triboukait et al (2004)* and *Cushing et al (2008)* found low correlations between for example RCT and cVEMP and sensitivity and specificity rates for cVEMP with caloric test or RCT as a reference test tend to vary strongly.

“New” vestibular function tests

Apart from the caloric test and RCT, that are considered the gold standard, and the often applied cVEMP, several “new” assessment tools have been reported in more recently published studies. These tests have been established because of the critique on the caloric test that it is anxiety provoking in children and in case of the RCT that very specialized and expensive materials are required. However, these alternatives need to have the same diagnostic accuracy. Even though diagnostic accuracy of the DVA and (v)HIT seem promising, these results should be interpreted with caution as the level of evidence is only moderate for the DVA and limited for the (v)HIT (Appendix 3). Generalizability is limited due to the small sample sizes. The DVA shows very good sensitivity but the results for specificity vary, suggesting the DVA might serve as a good screening tool. Also, the DVA is found to be a useful test to evaluate functional VOR in uni- and bilateral vestibular deficits. However the DVA does not always identify a deficit in subjects with small lesions on one side. (Herdman, 2007)

This review revealed that the specificity of the (v)HIT is high but sensitivity is lower, which suggests the (v)HIT might serve better as a diagnostic tool. The clinical HIT, in contrast to the vHIT, can only identify overt saccades, catch-up saccades after the head movement. Covert saccades can provide extra information on vestibular function, which can be identified using the

Verbecque – Vestibular (dys)function in children with SNHL

vHIT.(Hülse et al, 2015; Hamilton et al, 2015) Moreover, the vHIT provides gain values and seems to be well-tolerated in children in contrast to the traditional caloric test and the RCT.(Hamilton et al, 2015; Hülse et al, 2015) *Janky & Givens (2015)* stated that gain values for the vHIT below 0.85 (1.5 SD below the mean value of the control group) represent vestibular dysfunction, and thereby providing 100% agreement with RCT identification. However, *Hamilton et al (2015)*, who investigated vHIT in 33 children with vestibular dysfunction of which 2 were hearing impaired, proposed a gain of less than 0.7 to represent vestibular dysfunction. The differences in reported cut-off values underline the need for further investigations regarding the diagnostic value of the vHIT, although very preliminary reports on the vHIT suggest it seems to have large potential to serve as a diagnostic tool.

Clinical practice

The findings of this review support the fact that it is necessary to develop a standard protocol when it comes to vestibular testing in children.(Cushing et al, 2009; Janky & Givens, 2015; Tribukait et al, 2004) Some suggestions for this protocol can be made, but due to the low level of evidence, these should be handled with caution. When vestibular function is assessed in children with SNHL, a battery of tests seems in order. The protocol should contain tests that assess different vestibular end-organs, such as for example a combination of caloric testing, RCT and cVEMP. Before one of the newer tests (DVA, (v)HIT) can be implemented, more research regarding their sensitivity and specificity are necessary. Furthermore, it should be taken into account that the caloric test serves as a low frequency assessment tool for the HSCC. The RCT on the other hand, can be used for high frequency assessment. However, similar prevalence values have been reported and subsequently higher sensitivity and specificity rates when an SHA high frequency protocol was applied. Thus, further clarification of which modality (SHA or step rotation) is more appropriate in children, seems necessary.

Limitations of the study

Even though the search query was ran in three databases, it is still possible that relevant studies might have been missed, which is one of the disadvantages of a systematic search. Further, the Cosmin checklist that is used for methodological assessment of the studies is a

Verbecque – Vestibular (dys)function in children with SNHL

complex and difficult assessment tool. Also, the four-point rating scale that was used is not yet validated. However, at present, in the opinion of the authors, the Cosmin checklist is the most complete checklist to assess methodological quality of studies examining psychometrics of measurement instruments.

Conclusion

Based upon the data presented in this systematic review, caloric stimulation, RCT and cVEMP have been investigated most extensively. Although the RCT is considered part of the gold standard test battery, a lot of variation exists in the protocols, which could explain the variability in prevalence and the varying diagnostic accuracy. Several attempts have been made to provide new vestibular function tests, such as the DVA and (v)HIT, that could represent a valuable alternative to the more expensive, invasive or unpleasant vestibular test that are currently applied in children such as the RCT or caloric test. However, before the use of these tests can be translated into clinical practice, far more research on the psychometric properties of these tests is required. Further research should be performed to reach consensus on a standard protocol for vestibular testing. Preferably this is done in a study with an adequate sample size ($N > 30$ per subgroup) and reported according to the STROBE statement (STrengthening the Reporting of OBservational studies in Epidemiology), a checklist of items that should be included in reports of observational studies.

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Verbecque – Vestibular (dys)function in children with SNHL

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Verbecque – Vestibular (dys)function in children with SNHL

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Figure 1: Screening procedure

Table 1: Methodological quality assessment				
Author	Cosmin box	Cosmin score by rater A	Cosmin score By rater B	Final Cosmin score
Backous, 2000	Box H	Poor	Poor	Poor
Christy, 2014	Box H	Poor	Fair	Fair
Cushing, 2009	Box H	Poor	Poor	Poor
Cushing, 2008	Box H	Fair	Fair	Fair
De Kegel, 2012	Box H	Fair	Fair	Fair
Inoue, 2013	Box H	Fair	Poor	Poor
Jacot, 2009	Box H	Fair	Fair	Fair
Janky, 2015	Box H	Poor	Poor	Poor
Jin, 2006	Box H	Poor	Poor	Poor
Kaga, 2008	Box H	Poor	Poor	Poor
Lisboa, 2005	Box H	Poor	Poor	Poor
Maes, 2014	Box H	Fair	Fair	Fair
Mäki-Torkko, 2005	Box H	Poor	Poor	Poor
Martin, 2012	Box F	Fair	Fair	Fair
Rine & Braswell, 2003	Box H	Poor	Poor	Poor
Shinjo, 2007	Box H	Poor	Poor	Poor
Thierry, 2015	Box H	Poor	Poor	Poor
Triboukait, 2004	Box H	Fair	Fair	Fair
Xu, 2015	Box H	Poor	Fair	Poor
Xu, 2014	Box H	Poor	Poor	Poor
Zagolski, 2007	Box H	Poor	Poor	Poor
<p>Legend: This table gives an overview of the methodological quality scores of the studies given by the researchers using the Cosmin 4 point rating scale for the different boxes that were used to score the articles.</p> <p>Abbreviations: Box H: criterion validity.</p>				

Author(s)	Age range	Hearing impaired children				Degree of hearing loss	CI	n	Age range	Applied vestibular tests						
		Etiology of hearing loss (n)								Semi-circular canals					Otoliths	
		C	A	U	N/R					caloric	RCT	(v)HIT	HST	DVA	cVEMP	oVEMP
Backous & Quigley, 2000	23 m/o - 10 y/o				11		x			x	x	x				
Bergholtz, 1977	1-16 y/o	3	14	13						x						
Brookhouser, 1982	3-19 y/o				170	Profound				x						
Christy, 2014	6-12 y/o	13		7		Severe-profound		23	6-12 y/o	x	x		x	x		
Cushing, 2009	4-17 y/o		9			Profound	x			x	x			x		
Cushing, 2008	3-19 y/o	17	7	16		Severe-profound	x			x	x			x		
Cyr, 1985	3 m/o - 6 y/o		21			Severe-profound		83	3 m/o - 6 y/o	x						
De Kegel, 2012	3-12 y/o	25	11	12		Moderate-profound	x	41	3-12 y/o	x				x		
Diepeveen & Jensen, 1968	6-15 y/o	9	6	40						x						
Hallmo, 1986	4-15 y/o	7	15	34		Mild-profound				x						
Holderbaum, 1979	11-14 y/o	15		11	5	Moderate-profound				x						
Inoue, 2013	20-97 m/o	94				Profound	x			x	x			x		
Jacot, 2009	7 m/o - 16 y/o	27	10	52	135	Profound	x			x	x	x		x		
Jafari, 2011	6-9 y/o	30				Profound		30	6-9 y/o					x		
Janky & Givens, 2015	6-17 y/o			11			x	12	6-15 y/o	x	x		x	x	x	
Jin, 2006	2-7 y/o	5		7		Profound	x	9	8 m/o - 10 y/o	x				x		
Jin, 2008	2-14 y/o			24			x							x		
Kaga, 2008	31-97 m/o	20				Severe-profound				x	x			x		
Kaga, 1981	<1-6 y/o	88				Profound		60	<1-6 y/o	x						
Lisboa, 2005	10-14 y/o	8	9	9		Severe-profound				x		x				
Maes, 2014	3-12 y/o	21	8	10		Mild-profound	x	48	4-12 y/o	x				x		
Mäki-Torkko & Magnusson, 2005	12-90 m/o				14	Severe-profound				x	x					
Martin, 2012	4-13 y/o				32	Mild-profound	x	32	4-13 y/o				x			
Masuda & Kaga, 2014	3-37 m/o	94	3			Profound				x						
Potter, 1984	5-8 y/o				34	Moderate-profound				x						
Rine & Braswell, 2003				11		Severe-profound		76	3-15 y/o	x			x			
Sandberg, 1965		72		14	28	Severe-profound				x						
Selz, 1996	8-17 y/o	5	5					5	8-17 y/o	x						
Shall, 2009	4-7 y/o	24	9			Severe-profound	x							x		
Shinjo, 2007	31-97 m/o	16	1	3		Severe-profound				x	x			x		
Singh, 2012	4-12 y/o		2	13		Severe-profound		10	4-12 y/o					x		
Suonpää, 1988	6-18 y/o	69	19	38	18	Severe-profound		28	7-18 y/o	x						
Thierry, 2015	1-17 y/o	26	2	15						x		x		x		
Tribukait, 2004	15-17 y/o	12	1	23		Profound				x	x			x		
Xu, 2015	3-14 y/o				43	Profound		20	4-10 y/o					x	x	
Xu, 2014	3-12 y/o				31	Severe	x	20	4-10 y/o					x	x	
Zagolski, 2007	3 m/o	18				Moderate-profound		40	3 m/o	x				x		
Zhou, 2009	2-16 y/o	12	1	10		Moderate-profound	x	12	4-18 y/o					x		
Total number of children		811	242	378	620			549		1184	1144	160	11	217	1358	137
Total number of studies		26	20	19	15		14	17		17	20	7	1	4	21	3

Legend: This table provides an overview of the characteristics of the populations under investigation and the vestibular function tests that were applied in the individual studies.

Abbreviations: C: congenital; A: acquired; U: unknown; N/R: not reported or unclear; n: number of children; CI: cochlear implant; RCT: Rotational Chair Test; (v)HIT: (video) Head Impulse Test; HST: Head Shaking Test; DVA: Dynamic Visual Acuity test; c-/oVEMP: collic/ocular vestibular evoked myogenic potentials; m/o: months old; y/o: years old

Figure 2: overview of the applied vestibular tests and the number of recruited children, classified according to the etiology of the hearing loss.

Legend: this figure shows the total number of hearing impaired children and their subdivisions according to the etiology of the hearing loss. The etiology of the hearing loss was defined as acquired (meningitis,...), congenital (Waardenburg Syndrome, Cytomegalovirus,...), unknown or not reported. Children with normal hearing are presented in white boxes with their corresponding number.

Abbreviations: RCT: Rotational Chair Test; DVA: Dynamic Visual Acuity test; (v)HIT: (video)Head Impulse Test; HST: Head Shaking Test; c- or oVEMP: collic – or ocular Vestibular Evoked Myogenic Potentials.

Verbecque – Vestibular (dys)function in children with SNHL

Table 3: overview of the applied protocols for the caloric test and corresponding cut-off values and prevalence of vestibular dysfunction

Authors	Characteristics of the applied protocol stimulus Temp. (duration)	Air/ water	specifics	Eye movements ENG/VNG	Outcome variables	Interpretation: cut-off to determine loss
Brookhouser, 1982	30°C, 44°C	water	closed loop caloric irrigation	ENG	Asymmetry Slow phase velocity Asymmetry*	unilateral loss: asymmetry > 20% Bilateral loss: nystagmic response < 7.5°/sec per ear
Cushing, 2009 Cushing, 2008	30°C, 44°C, ice water	water	binaural alternating caloric stimulation	ENG	Slow phase velocity	Unilateral loss normal: <15%, mild: 15-29% moderate: 30-50% severe: 51-100% Areflexia: no response on ice water unilateral Bilateral loss Moderate: sum of thermal caloric responses < 20°/s AND responses >3°/s with ICE water, severe: negative ice water caloric responses on both sides
Diepeveen & Jensen, 1968	30°C, 44°C + 20°C (40s (classic temp) + 2 min 20°C)	water			duration nystagmus	if present: ≥ 80 sec. Normal reaction = reaction without Bartels' glasses as well at 30°C as at 44°C; reduced reaction = reaction at 30°C. At 44°C only reaction with Bartels' glasses or no reaction at all at this temperature. Markedly reduced = no reaction at 30°C or at 44°C but reaction at 20°C. No reaction = no reaction at 20°C.
Sandberg, 1965	30°C, 44°C (40 s) + 1 min 15-20°C	water				Normal >80 sec; Slightly reduced reaction: 80 sec, small amplitude; Reduced reaction: <80, weak amplitude; Markedly reduced reaction: no reaction for 40 sec, but positive 1 min 15-20°C; No reaction to 15-20°C
Hallmo, 1986	30°C, 44°C (30 s)	water	binaural bithermal caloric test when monothermal test (44°C) > 15% between two ears.		duration nystagmus	30% limit to determine canal paresis and directional preponderance; a unilateral absent response from the HSCC in the bithermal test = canal paralysis. (Uemura (1977))
Jacot, 2009	33°C, 44°C	water			Ab-/presence normal latencies* Asymmetry*	relative valence and directional preponderance ≤ 15% > 20 % side difference
Suonpää, 1988	30°C, 40°C (30 s)			ENG	Slow phase velocity	< 14.5°/s for both sides
Thierry, 2015	30°C and 44°C (90 s)	water		VNG	summation number nystagmus*	
Tribukait, 2004	30°C, 44°C (30 s)	water- 160 ml	open-loop	ENG, VNG	asymmetry*	≥ 20 % side difference. Sum of the caloric responses (30° and 44°) < 20°/s --> bilat hypo. No response = areflexia.
Zagolski, 2007	20°C (20 s)	Water- 20 ml			frequency and duration nystagmus	> 30% difference with normal values
Holderbaum, 1979 Inoue, 2013	5°C 4°C	water water		ENG	duration nystagmus	<2SD below reference values 13-24 months: < 53,3s 25-36 months: < 54,3s 37-48 months: < 52.4 s 49-60 months: < 57.4s 61-72 months: < 48.1 s >72 months: <35.1
Kaga, 2008 Shinjo, 2007	4°C	Water- 2 ml		ENG	duration nystagmus	compared with those of age-matched controls but no further specifications.
Lisboa, 2005	42°C, 20°C, 10°C (80 s)	Air		VNG	direction nystagmus gain	
Jin, 2006	No methodological characteristics reported.					

Legend: *: Jongkees' formula

Figure 3: overview of the overall prevalence of vestibular (dys)function assessed by means of the caloric test, the RCT and the cVEMP, categorized by the degree of hearing loss.

Legend: The results of the individual studies were gathered and grouped per degree of the hearing loss. All results concerning unilateral hypofunction or areflexia were clustered into “unilateral loss”. A similar classification was made for “bilateral loss”. All absent responses, hypofunction, abnormal results in which no specifics were reported on the affected side(s) were clustered as “loss”.

Verbecque – Vestibular (dys)function in children with SNHL

Table 4: overview of the applied protocols for the rotational chair test and corresponding cut-off values and prevalence of vestibular dysfunction

AUTHORS	TYPE OF ROTATIONAL CHAIR TEST	DARK-LIGHT- VISUAL CALIBRATION	POSITIONING	(V)(E)N G	OUTCOME VARIABLES	INTERPRETATION – REFERENCE VALUES
Backous & Quigley, 2000	constant velocity of 60°/s, then 240°/s. About the vertical axis in each direction for 60 s.	DARK - / - 1.7°, 1800 mCD point of light moved at 10° at 8, 16 and 24°/s	young children on parent's lap; older children tested as adults	ENG		
Bergholtz, 1977	linear acceleration and deceleration left and right: 7.5°/s ² - 100°/s ² . Oscillating rotation, amplitude 70°/s and 7.5°/s ²				Presence/absence nystagmus	abnormal: absence of nystagmus
Costy, 2014	A series of complete chair rotations to both the right and left (100°/s for 60 seconds). (step rotation) + 5 frequencies were examined: 0.01, 0.04, 0.08, 0.16, and 0.64 Hz) at 60° per second over 3 to 9 cycles (SHA).	DARK - / - Calibration of eye movement was performed before testing.	Small children sat in the lap of an adult. The subject's head was pitched forward 30° and gently immobilized.	VNG	SHA: VOR phase, gain, asymmetry. Step rotation: PER- AND POSTROTATORY time constants (time for nystagmus to decrease to 37% of its original strength)	normal: within 1.5 SD of the comparative sample mean – Casselbrant 2010 bilateral loss: gain values < 1.5 SDs outside the comparative range unilateral loss: abnormal phase and asymmetry values in the presence of normal gain values
Quising, 2009/2008	SHA: maximum 100°/s over a frequency of 0.25 - 5 Hz. Light and Dark; low frequency (≤ 2 Hz) versus high frequency (>2 Hz)	DARK - visual target at 3.1m - 10 degree gaze deviation right & left	seated alone, head secured in padded clamps	ENG		abnormal: results outside normative data (95% CI) in at least 2 consecutive frequencies. – adult reference values
Quiring, 1985	SHA at 0.01, 0.02, 0.04, 0.08, 0.16 Hz. 0.08 Hz was used as a screening device. 4.5 cycles of rotation.	DARK - / - /		ENG	VOR gain, phase, asymmetry	Table 6
Maes, 2014	SHA: three rotations were performed at 0.01, 0.05, 0.1Hz. Peak velocity of 50°/s. To maintain alertness during rotation, children were distracted by performing mental tasks.	DARK - / - fixating a visual target 10 degrees right and left with recalibration before each rotation.	seated with safety belt and head stabilized in a head rest.	ENG	VOR gain, phase, asymmetry	Abnormal: < mean - 1.96 SD – table 6 Maes 2010 , Maes 2008 , Maes 2012
De Kegel, 2012	Max velocity 200°/s at 300°/s ² acceleration and then decayed to 0°/s by a decel. Of -4°/s ² . Twice in both clockwise and counterclockwise direction.	/ - / - /	held upright on their mother's knees with heads bended down 30°		# beats PERROTATORY NYSTAGMUS	Abnormal: <2SD beats; children > 6 years of age = < 23 beats. Kaga 1981
Doyle, 2013	chair was accelerated to a maximum rotational velocity of 200°/s, maximum acceleration of 300°/s ² ; velocity decayed to 0° over a period of 20 s. 2x clock- 2x counter clockwise.		young children on parent's lap; older children tested as adults	ENG	duration and number of beats of PERROTATORY NYSTAGMUS	Hypoactive responses: nystagmus duration < mean - 1SD. Areflexia: absent responses. No numeric values reported.
Kaga, 1981					VOR slow phase velocity	Reference values: Wiener-Vacher 1996
Jacot, 2009	40°/s ²				bias of the modulation amplitude	Reference values: Wiener-Vacher 1996
Jacot, 2009	60°/s constant rotation velocity and 13° axis rotation tilt.*				VOR	
Janky & Owens, 2015	SHA: 0.02, 0.08, 0.16, 0.32 Hz with variable maximal velocities, 70°/s, 50°/s, 40°/s, 30°/s respectively. Step rotation: in the clockwise and counterclockwise direction: acceleration at a rate of 100°/s ² up to a velocity of 100°/s. The chair maintained this velocity for 45 sec and then decelerated at a rate of 100°/s before coming to a complete stop.	DARK - / - /		ENG, VNG	VOR gain, phase, symmetry.	No numeric values reported.
Kaga, 2008; Matsuda & Kaga, 2014; Shimjo, 2007	Accelerated to a maximum rotational velocity of 160°/s ² then decayed by 4°/s ² . Test was performed once in counterclockwise and clockwise direction. 5 minute interval.	DARK - / - /	child on lap parent	ENG	# beats of nystagmus + duration PERROTATORY NYSTAGMUS	Reduced vestibular function: <50% (for both outcomes) Kaga 1981
Mäki-Torkko & Magnusson, 2005	100-120° (amplitude) with speed and periodicity of a full cycle at 2-3 s (comparable to 0.3-0.5 Hz).		child in the lap of one parent	VNG	presence of nystagmus	Abnormal: absence of nystagmus
Preyer, 1984	10 rotations to right in 20 s. After one minute rest -> left.		cross-legged. Head flexed 30° from vertical.		duration of the induced nystagmus in seconds. POSTROTATORY.	Abnormal: < 18 s postrotatory nystagmus.
Rine & Barzwell, 2003	frequencies: 0.01, 0.02, 0.04, 0.08, 0.16, 0.32 Hz at 50°/s over 3-9 cycles. Trapezoidal testing was done using a series of complete chair rotations, both right and left (100°/s) for 60 s.	DARK - / - subject had to look to center, left and right at a red illuminated dot (three repetitions)	children in lap of adult, head 30° flexed.	ENG	VOR gain, phase, asymmetry	/
Selz, 1996	frequencies: 0.04, 0.08, 0.16, 0.32 Hz with peak angular velocity of 50°/s, min 3 to max 10 oscillations.	DARK - / - /	head restrained	ENG	VOR gain, phase, asymmetry	/
Tibukait, 2004	40°/s ² , 150°/s	/ - / - /		VNG	VOR gain	Hypofunction: gain < 0.2; areflexia: no gain

Legend: *OVAR – measuring otolith function

Verbecque – Vestibular (dys)function in children with SNHL

Table 5: reported reference values for RCT and cVEMP

Authors/ research group	Vestibular Test RCT	Outcome variables (mean - SD)					
		Gain (%)		Phase (°)		Asymmetry (%)	
		Mean	SD	Mean	SD	Mean	SD
Cyr, 1985	SHA	69.70	15.4	6.86	7.42	2.17	6.90
Maes, 2014	SHA – 0.01 Hz	60.98	14.56	40.89	8.68	–1.38	16.10
	SHA – 0.05 Hz	78.67	20.05	11.93	4.10	–2.67	15.52
	SHA – 0.1 Hz	78.17	18.39	7.81	5.28	–0.38	11.15
Selz, 1996	SHA – 0.04 Hz	96.40	6.00	–1.20	2.86		
	SHA – 0.08 Hz	92.00	6.00	3.60	3.36		
	SHA – 0.16 Hz	86.60	6.00	10.20	5.15		
	SHA – 0.32 Hz	78.60	4.00	1.60	7.06		
	cVEMP	P1 (ms)		N1 (ms)		Interpeak amplitude (µV)	
		Mean	SD	Mean	SD	Mean	SD
Jafari, 2011	Tone burst	13.55	0.73	18.92	1.12	97.21	22.34
Janky & Givens, 2015		14.53	1.07	22.88	2.50	278.64	125.56
Jin, 2006	Click	10.59	0.50	16.19	1.30	181.09	90.90
Maes, 2014	Tone burst	13.19	0.82	20.78	1.47	208.38	61.53
Singh, 2012	Tone burst	15.39	1.51	23.68	2.49	160.51	57.41
Xu, 2014; Xu, 2015	Tone burst	15.00	0.98	21.68	1.36	360.88	166.57
Zagolski, 2007	Tone burst	8.3	1.70	13.5	2.5		
Zhou, 2009	Tone burst	14.66	0.72	21.54	1.01	77.40	53.36
	Click	10.79	1.36	16.82	1.79	30.11	21.64
Legend: empty cells show that this specific information was not provided by the author(s) of the individual studies.							
Abbreviations: RCT: Rotational Chair Test; cVEMP: collic Vestibular Evoked Myogenic Potentials.							

*Verbeque – Vestibular (dys)function in children with SNHL***Table 6: Overview of sensitivity and specificity of vestibular tests in children with SNHL (with/without cochlear implants)**

Author	N	Age	SNHL / CI	Test	Reference test: caloric test		Reference test: RCT		Reference test: cVEMP		Reference test: RCT + cVEMP	
					Sensitivity	Specificity	Sensitivity	Specificity	Sensitivity	Specificity	Sensitivity	Specificity
Christy, 2014	20	6-12 y/o	SNHL	m-ECVCT							63%	100%
Cushing, 2008	40	3-19 y/o	SNHL	RCT	80%	80%						
Inoue, 2013	89	20-97 m/o	SNHL	RCT	66%	73%						
Kaga, 2008	20	31-97 m/o	SNHL	RCT	61%	21%						
Shinjo, 2007	20	31m-8 y/o	SNHL	RCT	61%	21%						
Christy, 2014	20	6-12 y/o	SNHL	DVA							88%	69%
Rine & Braswell, 2003	11	Not reported	SNHL	DVA			100%	100%				
Christy, 2014	20	6-12 y/o	SNHL	HIT							75%	91%
Maki-Torkko & Magnuson, 2005	14	12-90 m/o	SNHL	HIT			80%	91%				
Janky & Givens, 2015	11	6-17 y/o	CI	vHIT-HSCC			100%	100%				
				vHIT-PSCC			67%	71%				
				vHIT-ASCC			91%	92%				
Cushing, 2008	40	3-19 y/o	SNHL	cVEMP	84%	84%	92%	95%				
Inoue, 2013	89	20-97 m/o	SNHL	cVEMP	100%	100%	48%	79%				
Jacot, 2009	22	7m-12 y/o	SNHL	cVEMP	90%	89%						
	89		CI	cVEMP	97%	98%						
Janky, 2015	11	6-17 y/o	CI	cVEMP			100%	100%				
Jin, 2006	12			cVEMP	88%	83%						
Kaga, 2008	20	31-97 m/o	SNHL	cVEMP	71%	30%	60%	78%				
Shinjo, 2007	20	31m-8 y/o	SNHL	cVEMP	71%	30%	60%	78%				
Tribukait, 2004	36	15-17 y/o	SNHL	cVEMP	80%	76%						
Zagolski, 2007	18	3 m/o	SNHL	cVEMP	100%	100%						
Janky, 2015	11	6-17 y/o	CI	oVEMP			83%	86%	83%	86%		
Xu, 2014	31	3-12 y/o	CI	oVEMP					91%	95%		
Xu, 2015	43	3-14 y/o	SNHL	oVEMP					89%	93%		

Legend: Bold percentages are reported in literature, *Italic percentages are calculations based on raw data in appendices 1 and 2.* Boxes in the same color show values on sensitivity and specificity of the same vestibular test.

Abbreviations: CI: Cochlear Implant; c- or oVEMP: collic or ocular Vestibular Evoked Myogenic Potentials test; DVA: Dynamic Visual Acuity test; m/o: months old; N: Number; RCT: Rotational Chair Test; SNHL: Sensorineural Hearing Loss; (v)HIT: (video) Head Impulse Test; y/o: years old.

1
2
3 Verbecque – Vestibular (dys)function in children with SNHL

Table 7: overview of the applied protocols for the collic Vestibular Evoked Myogenic Potentials test and corresponding cut-off values and prevalence of vestibular dysfunction								
CVEMP	BODY POSITION	SCM CONTRACTION		STIMULUS	AMPLIFICATION	BAND-PASS FILTER	CUT-OFF VALUES	REFERENCE VALUES
		UL	BL					
Christy, 2014	Semirecumbent position	x		air-conducted: 500 Hz tone bursts with 2-0-2 rise-plateau-fall; 97 dB nHL/ 105 dB nHL ; rate: 5 Hz	5000x	2 to 250 Hz. Time window of 30 ms	2 SDs from the normative mean	Valente 2007, Zhou 2009
Zhou, 2009	< 4 y/o seated on parents' lap, older children independently	x		air-conducted: 500 Hz tone bursts, 2 cycles rise/fall and no plateau. Presented monaurally, rate: 13/s, 90 dB nHL.	2000x	30-1500 Hz. Averaged 100-200 sweeps.		Table 6
Cushing, 2009	not specified	x		air-conducted: a 105 dB click		at least 100 sweeps		Kelsch 2006 P1: 8.5-14 ms; N1: 15-23 ms
Cushing, 2008								
Inoue, 2013	supine		x	air-conducted: 500 Hz tone bursts, 95 dB nHL, rate: 5 Hz		20-2000 Hz		Abnormal: <33.3
Jafari, 2011	supine	x		air conducted 500 Hz tone burst, monaurally. Rate: 5.1/s. 200 responses. Rise and fall time of 1 ms and plateau time of 2 ms.	5000x	20-2000 Hz		Table 6
Janky & Givens, 2015	Semirecumbent	x		air-conducted monaurally 5.1/s, Blackman gating window, 1 ms rise-fall time, 2 ms plateau. Intensity: 125 dB Sound Pressure Level (SPL). 100 sweeps were averaged for each cVEMP, 2 trials/cVEMP.	1000x	10-1000 Hz	absent (abnormal)/ present (normal)	Table 6
Jin, 2006; Jin, 2008; Kaga, 2008; Shinjo, 2007	supine		x	air conducted: rate: 5 Hz; rarefaction clicks: 0.1 ms, 95 dB nHL. Average of the two runs for the amplitudes. Analysis time: 50 ms. VEMPs in response to 100 stimuli were averaged twice.		20 - 2000 Hz	absolute value of the amplitude < 0.5, < 50µV (decreased VEMP)	Jin et al., 2006. Table 6
Maes, 2014, De Kegel 2012	Semirecumbent		x	air-conducted linear 500 Hz tone bursts with a rise and fall time of 1 ms and a 2 ms plateau. Intensity: 95 dB nHL (equivalent: 130 dB SPL); stimulus repetition: 5 Hz. Stimuli delivered monaurally and recorded unilaterally from 10.5 ms before 45 ms after stimulus onset. Each run: 128 sweeps were averaged.	5000x	10-1500 Hz		Maes 2011; Maes 2014, Table 6
Singh, 2012	sitting	x		air conducted: 500 Hz tone bursts, 4 ms rise/fall + plateau. Repetition: 5.1/sec	5000 x	10-1500 Hz. Time window 70 ms, 150-200 sweeps.		Table 6
Tribukait, 2004	supine		x	air conducted: 0.1 ms rarefaction clicks bilaterally presented 100 dB nHL. Stimulus rate: 4/s. 3 stimulation sequences of 64 clicks.			VEMP ratio > 0.5 -> asymmetry. Amplitude < 20 µV both sides = bilat hypo.	
Xu, 2014 Xu, 2015	supine		x	air conducted: 500 Hz tone bursts, rise/fall time : 1 ms, plateau = 2 ms, 131 dB SPL, decrease/increase dB.		1-1000 Hz, 50 stimuli averaged twice.		Table 6
Jacot, 2009				air and bone conducted: 750 Hz, 4.1/s and 6 ms duration.			present = normal; absent = abnormal.	Welgampola, 2005
Shall, 2009	supine		x	bone conducted: 10 Ω. 100 µs clicks at 95 dB nHL, 16 Hz. 200 tracings were averaged.		8-1600 Hz	amplitude ≥ 20 mV. latency: 8.3-14.4 ms.	
Thierry, 2015		x		bone conducted 750 Hz - 6 ms tone bursts. Rate: 4.1 Hz through radio ear vibration. 110 dB.			normal (type 1); partial dysfunction	Cushing 2009, Kelsch 2006

Verbecque – Vestibular (dys)function in children with SNHL

				(type 2); areflexia (type 3)	
Zagolski, 2007	supine	x	Air conducted: 500 Hz tone bursts, 90 dB, Rep. rate: 5 Hz, rise/fall time: 1 ms, plateau: 2ms	Absent/present	<u>Table 6</u>
Abbreviations: SCM: sternocleidomastoid muscle; UL: unilateral contraction of the sternocleidomastoid muscle, subjects were instructed to turn their head away from the ear being stimulated and lift their head in response to the acoustic stimulation; BL: bilateral contraction of the sternocleidomastoid muscle, subject asked to look upwards.					

Verbecque – Vestibular (dys)function in children with SNHL

List of figures :

Figure 1: provides an overview of the selection and screening process

Figure 2: shows the total number of children and their subdivisions according to the etiology of the hearing loss. The etiology of the hearing loss was defined as acquired (meningitis,...), congenital (Waardenburg Syndrome, Cytomegalovirus,...), unknown or not reported. Children with normal hearing are presented in white boxes with their corresponding number.

Figure 3: The results of the individual studies were gathered and grouped per degree of the hearing loss. All results concerning unilateral hypofunction or areflexia were clustered into "unilateral loss". A similar classification was made for "bilateral loss". All absent responses, hypofunction, abnormal results in which no specifics were reported on the affected side(s) were clustered as "loss".

Appendix 1: Overview of the prevalence of vestibular dysfunction of the semi-circular canals: raw data

Author	N	Age Range	Type of hearing loss	Type of Vestibular Hypofunction	Vestibular function tests for semi-circular canals							
					Caloric test	RCT	HIT	vHIT HSCC	vHIT PSCC	vHIT ASCC	HST	DVA
Backous 2000	11	23 months-10 y/o	SNHL (CCI)	Drop-outs		1/8					1/11	
				Normal		3/7	7/11				8/10	
				Unilateral hypofunction		1/7	3/11				2/10	
				Bilateral hypofunction		3/7	1/11				0/10	
Bergholtz 1977	30	1-16 y/o	SNHL	Normal		5/10						
				Areflexia		5/10						
Brookhouser 1982	170	3-19 y/o	Profound SNHL	Drop-outs	4/170							
				Normal	129/166							
				Unilateral hypofunction	17/166							
				Bilateral hypofuncton	20/166							
Christy 2014	20	6-12 y/o	Severe to profound SNHL	Drop-outs		1/20						1/20
				Normal		0/19						11/19
				Unilateral hypofunction		5/19						5/19
				Bilateral hypofunction		3/19						3/19
				Areflexia		11/19						
Cushing 2009	9	4-17 y/o	Profound SNHL (8/9 CCI)	Drop-outs	1/9	1/9						
				Normal	0/8	1/8						
				Bilateral areflexia	3/8							
				Unilateral hypofunction/areflexia	5/8							
				Abnormal VOR gain		7/8						
Cushing 2008	40	3-19 y/o	Severe to Profound SNHL (CCI)	Drop-outs	8/40	3/40						
				Normal	16/32	23/37						
				Mild unilateral loss	5/32							
				Moderate unilateral loss	1/32							
				Unilateral areflexia	1/32							
				bilateral areflexia	9/32							
				Abnormal VOR gain		14/37						
Cyr, 1985	21	3 months - 6 y/o	Severe to profound SNHL	Normal		0/21						
				Bilateral hypofunction		21/21						
Diepeveen & Jensen, 1968	55	6-15 y/o	SNHL	Normal	17/110							
				Hypofunction	63/110							
				Areflexia	30/110							
Hallmo, 1986	56	4-15 y/o	Mild to profound SNHL	Normal	50/56							
				Unilateral hypofunction	3/56							
				Unilateral areflexia	3/56							
Holderbaum, 1979	31	11-14 y/o	Moderate to profound SNHL	Normal	15/31							
				Abnormal	16/31							
Inoue 2013	89	20-97 m/o	Profound SNHL (CCI)	Drop-outs	14/89	5/89						
				Normal	44/75	67/84						
				Unilateral hypofunction	13/75	1/84						
				Bilateral hypofunction	18/75	16/84						
Jacot 2009	224	7m/o-16y/o	Profound SNHL	Normal	110/224							
				Unilateral hypofunction	18/224							
				Bilateral hypofunction (symm)	13/224							
				Bilateral hypofunction (asymm)	9/224							
				Unilateral areflexia	18/224							
	89	7m/o-12y/o	Profound SNHL (CCI)	Bilateral areflexia	56/224							
				Normal	54/89							
				Unilateral hypofunction	8/89							
				Bilateral hypofunction (symm)	4/89							
				Bilateral hypofunction (asymm)	3/89							
				Unilateral areflexia	5/89							
				Bilateral areflexia	15/89							

Janky & Givens 2015	11	6/17y/o	SNHL (CCI)	Normal		12/22		12/22	7/22	11/22	
Jin 2006	12	2-7 y/o	Profound SNHL	Drop-outs	2/12						
				Normal	4/10						
				Hypofunction	4/10						
Kaga, 2008	20	31-97 months	Severe-profound SNHL	Areflexia	2/10						
				Normal	3/20	14/20					
				Unilateral hypofunction	7/20	1/20					
Kaga, 1981	88	<1-6 y/o	Profound SNHL	Bilateral hypofunction	2/20	2/20					
				Bilateral areflexia	8/20	3/20					
				Drop-outs		21/88					
Lisboa, 2005	26	10-14 y/o	Severe to profound SNHL	Normal		22/67					
				Hypofunction		38/67					
				Areflexia		7/67					
Mäki-Torkko & Magnusson, 2005	14	12-90 m/o	Severe to Profound SNHL	Normal	1/26		26/26				
				Unilateral hypofunction	5/26						
				Bilateral hypofunction	20/26						
Martin, 2012	32	4-13 y/o	Mild to profound SNHL (CCI)	Drop-outs		2/14					
				Normal		9/14	11/14				
				Bilateral hypofunction		3/14	3/14				
Masuda & Kaga, 2014	97	3-37 months	Profound SNHL	Normal							27/32
				Abnormal							5/32
Potter, 1984	34	5-8 y/o	Moderate to profound SNHL	Normal		81/97					
				Hypofunction		16/97					
				Normal		14/34					
Sandberg, 1965	114		Severe to Profound SNHL	Hypofunction		5/34					
				Areflexia		15/34					
				Normal	66/114						
Shall 2009	33	4-7 y/o	Severe to profound SNHL (CCI)	Hypofunction	40/114						
				Areflexia	8/114						
				Normal	4/33						
Shinjo 2007	20	31-97 m/o	Severe SNHL	Unilateral hypofunction	7/33						
				Areflexia	22/33						
				Normal	3/20	14/20					
Suonpää, 1988	144	6-18 y/o	Severe to profound SNHL	Asymmetrical hypofunction	7/20	1/20					
				Symmetrical hypofunction	2/20	2/20					
				Areflexia	8/20	3/20					
Tribukait 2004	36	15-17 y/o	Profound SNHL	Normal	42/144						
				Unilateral hypofunction	24/144						
				Areflexia	78/144						
Zagolski, 2007	18	3 months	Moderate to profound SNHL	Drop-outs	3/36						
				Normal	15/33						
				Unilateral hypofunction	8/33						
				Bilateral hypofunction	5/33						
				Areflexia	5/33	5/5					
				Normal	12/36						
				Hypofunction	12/36						
				Areflexia	12/36						

Abbreviations: ASCC: Anterior Semicircular Canal; CCI: Children with Cochlear Implantation, c- or oVEMP: collic or ocular Vestibular Evoked Myogenic Potentials test, DVA: Dynamic visual acuity, HST: Head Shaking Test; HSCC: Horizontal Semicircular Canal; N: Number, PSCC: Posterior Semicircular Canal; RCT: Rotational Chair test, SNHL: Sensory Neural Hearing Loss, (v)HIT: (video) Head Impulse Test.

Appendix 2: Overview of the prevalence of vestibular dysfunction of the otoliths: raw data						
Author	N	Age Range	Type of hearing loss	Type of Vestibular Hypofunction	Vestibular function tests	
					cVEMP	oVEMP
Christy 2014	20	6-12 y/o	Severe to profound SNHL	Drop-outs	1/20	
				Normal	19/19	
				Unilateral hypofunction	0/19	
				Bilateral hypofunction	0/19	
Cushing, 2009	9	4-17 y/o	Profound SNHL	Areflexia	0/19	
				Drop-outs	2/9	
				Normal	5/7	
				Areflexia	2/7	
Cushing, 2008	40	3-19 y/o	Severe to Profound SNHL (CCI)	Drop-outs	15/40	
				Normal	15/25	
				Unilateral areflexia	5/25	
				Bilateral areflexia	5/25	
De Kegel, 2012	48	3-12 y/o	Moderate to profound SNHL (CCI)	Drop-outs	6/48	
				Normal	35/42	
				Areflexia	7/42	
				Drop-outs	27/89	
Inoue 2013	89	20-97 m/o	Profound SNHL (CCI)	Normal	36/62	
				Unilateral hypofunction	5/62	
				Bilateral hypofunction	21/62	
				Normal	123/224	
Jacot 2009	224	7m/o-16y/o	Profound SNHL	Unilateral hypofunction	22/224	
				Bilateral hypofunction (symm)	23/224	
				Bilateral hypofunction (asymm)	11/224	
				Unilateral areflexia	9/224	
				Bilateral areflexia	36/224	
	89	7m/o-12y/o	Profound SNHL (CCI)	Normal	55/89	
				Unilateral hypofunction	8/89	
				Bilateral hypofunction (symm)	9/89	
				Bilateral hypofunction (asymm)	4/89	
				Unilateral areflexia	1/89	
Jafari, 2011	30	6-9 y/o	Profound SNHL	Bilateral areflexia	12/89	
				Normal	14/30	
				Unilateral areflexia	4/30	
				Bilateral areflexia	12/30	
Janky & Givens 2015	11	6/17y/o	SNHL (CCI)	Normal	12/22	10/22
Jin 2006	12	2-7 y/o	Profound SNHL	Normal	6/12	
				Hypofunction	1/12	
			Profound SNHL (CCI device off)	Areflexia	5/12	
				Normal	1/12	
			Profound SNHL (CCI device on)	Hypofunction	11/12	
				Areflexia	0/12	
				Normal	1/12	
				Hypofunction	3/12	
Jin, 2008	24	2-14 y/o	SNHL (device off)	Areflexia	8/12	
				Normal	0/24	
				Hypofunction	4/24	
			SNHL (device on)	Areflexia	20/24	
				Normal	12/24	
				Hypofunction	2/24	
Kaga, 2008	20	31-97 months	Severe to profound SNHL	Areflexia	10/24	
				Normal	10/20	
				Unilateral hypofunction/ areflexia	6/20	
				Bilateral areflexia	4/20	
Shall, 2009	33	4-7 y/o	Severe to profound SNHL (CCI)	Drop-outs	1/33	
				Normal	5/32	
				Unilateral hypofunction/ areflexia	7/32	
				Bilateral areflexia	20/32	

Shinjo 2007	20	31-97 m/o	Severe to profound SNHL	Normal	10/20	
				Asymmetrical hypofunction	6/20	
				Symmetrical hypofunction	0/20	
				Areflexia	4/20	
Singh 2012	15	4-12 y/o	Severe to profound SNHL	Hypofunction	13/15	
				Areflexia	2/15	
Tribukait 2004	36	15-17 y/o	Profound SNHL	Normal	21/36	
				Unilateral hypofunction	7/36	
				Bilateral hypofunction	7/36	
				Areflexia	1/36	
Xu, 2015	43	3-14 y/o	Profound SNHL	Normal	27/43	25/43
			Severe SNHL (pre CI)	Drop-outs	5/31	
				Normal	18/26	22/31
Xu, 2014	31	3-12 y/o	Severe SNHL (CCI – CI on)	Drop-outs	5/31	
				Normal	8/26	4/31
			Severe SNHL (CCI – CI off)	Drop-outs	8/31	5/31
				Normal	8/23	5/26
Zagolski, 2007	18	3 months	Moderate to profound SNHL	Areflexia	24/36	
Zhou 2009	23	2-16 y/o	Moderate to profound SNHL	Abnormal	21/23	
Abbreviations: ASCC: Anterior Semicircular Canal; CCI: Children with Cochlear Implantation, c- or oVEMP: collic or ocular Vestibular Evoked Myogenic Potentials test, DVA: Dynamic visual acuity, HST: Head Shaking Test; HSCC: Horizontal Semicircular Canal; N: Number, PSCC: Posterior Semicircular Canal; RCT: Rotational Chair test, SNHL: Sensory Neural Hearing Loss, (v)HIT: (video) Head Impulse Test.						

Appendix 3: This flowchart shows a description of how the level of evidence was assigned. The criteria of the Cochrane Collaboration Back Review Group (*van Tulder et al 2003*) were used to determine the level of evidence, but implemented as by Saether et al 2013.

Legend: the caloric test was used as the gold standard.

Abbreviations: c- or oVEMP: colic or ocular Vestibular Evoked Myogenic Potentials test; DVA: Dynamic Visual Acuity test; HST: Head Shaking Test; RCT: Rotational Chair Test; (v)HIT: (video) Head Impulse Test.