Diameter of the Cochlear Nerve Canal predicts Cochlear Nerve Deficiency in Children with Sensorineural Hearing Loss

Der Durchmesser des Canalis nervi cochlearis als Marker einer Cochlearisaplasie bei Kindern mit sensorineuraler Schwerhörigkeit

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Key words
internal auditory canal, cochlear implantation, cochlear nerve dysplasia, cochlear nerve canal

ABSTRACT

Purpose Detection of cochlear nerve deficiency (CND) is usually straightforward using magnetic resonance imaging (MRI). In patients in whom MRI cannot be performed or imaging provides equivocal findings, computed tomography (CT) of the temporal bone might offer indirect evidence of CND. Our study aimed to derive a cut-off value for the diameter of the cochlear nerve canal (CNC) and internal auditory canal (IAC) in temporal bone CT to predict CND.

Materials and Methods This retrospective study included 70 children with sensorineural hearing loss (32 with CND and 38 control patients). The height, width, and cross-sectional area of the IAC and diameter of the CNCs were determined using temporal bone CT. Receiver operating characteristic (ROC) and Student's t-tests were performed for each parameter.

Results The mean diameter of the CNCs was significantly smaller in children with CND than in the control group (1.2 mm versus 2.4 mm, p < .001). The optimal threshold for CNC for separation of the two groups was 1.9 mm, resulting in a sensitivity of 98.7 % and specificity of 89.2 %. The IAC dimensions could not distinguish between children with CND and controls.

Conclusion A CNC diameter of less than 1.9 mm is a reliable predictor of CND in children with sensorineural hearing loss.

Key Points:
▪ A small cochlear nerve canal predicts cochlear nerve deficiency (CND)
▪ The size of the internal auditory canal cannot predict CND
▪ Whenever MRI is impossible or ambiguous, CT can rule out CND

ZUSAMMENFASSUNG


Material und Methoden An der retrospektiven Studie nahmen 70 Kinder mit Innenohrschwerhörigkeit teil (32 mit CND und 38 Kontrollpatienten). Die Höhe, Breite und Querschnittsfläche der IAC und der Durchmesser der CNC wurden mittels Schläfenbein-CT bestimmt. Für jeden Parameter wurde eine Receiver Operating Characteristic (ROC) und ein Student’s t-Tests durchgeführt.
Introduction
The prevalence of hearing loss in children has been reported to be between 1–6 of 1000 [1]. In congenital severe sensorineural hearing loss (SNHL) or deafness, treatment with a cochlear implant is the therapy of choice. Preoperative radiological imaging is essential for visualizing the anatomy of the inner ear. Recommendations regarding the imaging modality are inconsistent [2–5]. Most centers perform both computed tomography (CT) and magnetic resonance imaging (MRI).

In up to 79% of patients with unilateral deafness, MR-morphological deficiency of the cochlear nerve (CND) is observed [6]. However, a prerequisite for successful cochlear implantation is an intact cochlear nerve, as cochlear implantation surgery is contraindicated in the setting of cochlear nerve aplasia, whereas in cochlear nerve hypoplasia, this treatment may be considered. Therefore, precise radiological evaluation of the vestibulocochlear nerve by high-resolution MRI of the temporal bone is required.

Although imaging of the cochlear nerve with MRI is usually straightforward, in certain situations, there are obstacles such as a narrow internal auditory canal (IAC) of MRI artifacts due to fixed braces or motion of the child.

As a substitute for direct visualization of the cochlear nerve in MRI, temporal bone CT may provide indirect clues for CND, as there is an association between the diameter of the cochlear nerve canal (CNC) and the presence of hypo- and aplasia of the cochlear nerve [7, 8]. In addition, an association between CNC and narrowed IAC has been described [9]. However, the variation in the recommended cut-off values is large.

Our study aimed to derive cut-off values for the dimensions of the CNC and IAC in temporal bone CT for the prediction of CND in children with SNHL. In addition, we sought to define malformations if the inner ear was associated with CND.

Materials and Methods
Study design
For this retrospective study, the clinical database was screened for children with deafness or severe SNHL who underwent imaging diagnostics to evaluate the anatomical preconditions for implantation of the hearing system who underwent both MRI and CT examination of the temporal bone between January 2009 and June 2021. Based on the MRI findings, the patients were divided into a study group with evidence of hypoplasia or aplasia of the cochlear nerve and a control group with a regular cochlear nerve. The study was approved by the local ethics board (404/19-ek).

Imaging
CT scans were performed using a 128-slice scanner (Ingenuity, Philips Amsterdam, Netherlands). The scan parameters were 120 kV and 200 mA, with collimation of 0.625 mm. Subsequently, a multiplanar reconstruction with a slice thickness of 0.3 mm orthogonal to the IAC was made (Fig. 3). The diagnosis of CND was based on concomitant MRI if the cochlear nerve was absent (aplasia) or smaller than the other nerves within the IAC (hypoplasia) [10].

Image analysis
All measurements on the CT scans were performed with Intellispace Portal 9.0 (Philips Healthcare, Best, The Netherlands) by two experienced pediatric radiologists (25 and 30 years of experience in pediatric MRI). The height and width of the IAC were assessed in the parasagittal reconstructions at the center of the IAC as described in the literature [11, 12], except those measurements were performed in a plane perpendicular to the IAC to obtain the largest diameters of this oval shape. In addition, the area of the IAC was calculated from its height and width (Fig. 1a). Due to its round shape, only the largest width of the CNC was measured in the transverse plane (Fig. 1b).

Statistics
The mean values of the two readers were employed. The intra-class correlation coefficient was computed to assess the agreement between the two readers. Differences regarding the sex of both groups were calculated using the chi-squared test. Dimensions for CNC and IAC were tested for normal distribution for each group using the Shapiro-Wilk test. For normally distributed variables, the mean and standard deviation are given. Otherwise, the median and interquartile range (IQR) are given. Differences in central tendency were determined for normally distributed variables using a t-test for independent samples and Wilcoxon-Mann-Whitney test for non-normally distributed variables. A
p-value below 0.05 was considered significant. The diagnostic value of each variable was determined by the area under the curve (AUC) of the receiver operating characteristic (ROC) curve. The threshold to discriminate between both groups of patients was determined using the Youden index. Statistical analysis was performed using RStudio (Integrated Development for R. RStudio 1.2.5033, PBC, Boston, MA, USA).

Results

Study population
A total of 70 children with SNHL were included (median age 1.7 years, IQR 0.8–4.8, 40 males), of whom 32 were children with CND that was verified on MRI, and 38 were children with a normal cochlear nerve (Table 1). In the CND group, only the right side was affected in nine patients, the left side in 18 patients, and both sides in five patients. This resulted in a total of 37 inner ears with CND for further evaluation. Of the 37 inner ears, 30 ears showed aplasia, and seven ears showed hypoplasia of the cochlear nerve. The 38 patients in the control group provided 76 inner ears for further assessment.

Canalis nervi cochlearis
The mean CNC diameter was significantly narrower in the CND group (1.19 mm ± 0.62 mm) than in the control group (2.45 mm ± 0.33, p < .001) (Table 2 and Fig. 2a). In 89% (33/37 ears), CNCs in the CND group were smaller than 1.9 mm, whereas in the control group, the CNC diameter was at least 1.9 mm. As an outlier, the widest CNC in the CND group was 2.6 mm in a patient with a complex malformation of the inner ear, incomplete partition type 1, and high-grade hypoplasia of the IAC. In CND patients with unilateral cochlear aplasia, the CNC width of the unaffected contralateral ears did not differ from that of the control group (p = .78). The agreement between the two readers regarding the diameters of the CNCs and IACs was excellent (ICC = 0.93).

ROC analysis for CNC as a predictive parameter of CND yielded an area under the curve (AUC) of 0.96. The optimal threshold for separation of the two groups was 1.9 mm, resulting in a sensitivity of 98.7% (98.7% for each reader) and specificity of 89.2% (89.2% and 83.8% for each reader) (Fig. 3). The results for both readers are shown in Supplementary table 1–5.

Internal auditory canal
The IAC mean width (3.95 ± 1.18), mean IAC height (4.70 ± 1.40), and mean IAC area (15.6 ± 7.9) in the CND group were significantly smaller than those in the control group, with a mean IAC width of 4.82 ± 0.80, p < .001, mean IAC height of 5.22 ± 0.93, p = .04, and mean IAC area of 20.2 ± 6.5, p = .003 (Table 2 and Fig. 2b–d). The ROC analysis resulted in an AUC of 0.71 for IAC width, 0.62 for IAC height, and 0.67 for IAC area (Fig. 3 and supplementary table 1–5). The agreement between the two readers regarding the IAC dimensions was excellent (ICC = 0.97).

In children with unilateral CND, the IAC width and IAC area on the unaffected opposite side showed no significant difference.
from the values of the control group ($p = .14$, $p = .06$), whereas the difference in IAC height was weakly significant ($p = 0.046$).

**Associated malformations**

In 31% (10/32) of patients with CND, dysplasia was associated with at least one of the following structures: vestibular organ, cochlea, middle ear ossicles, and vestibular aqueduct. All five patients with bilateral CND showed associated dysplasia (Table 3). Six of ten patients with associated dysplasia exhibited malformation of the cochlea (e.g., Mondini malformation, incomplete partition).

**Discussion**

Our study was able to show for a large sample of patients with SNHL that temporal bone CT can reliably separate patients with CND from patients without CND. A CNC diameter of 1.9 mm was found to be the most efficacious parameter. In contrast, IAC dimensions do not allow conclusions to be drawn about the...
development of the cochlear nerve in individual cases. Finally, we demonstrated that CND is associated with other inner ear malformations in many patients. As the patients in the control group also suffered from SNHL our thresholds cannot be readily applied to incidental findings on CT. However, the most common clinical concern is whether cochlear implantation is appropriate in the presence of SNHL. CND, as an incidental finding on CT, seems very unlikely, especially when CT is avoided in children whenever possible.

**CNC as predictor for CND**

In 2002, Stjernholm and Muren performed measurements on rubber casts and CT scans of patients with various diseases of the head, nose, and throat [13]. They proposed a lower limit of 1.4 mm for the CNC diameter.

Komatsubara et al. (2007), based on 15 patients with SNHL including nine patients (ten ears) with CND, found that that a CNC width below 1.5 mm differentiated CND patients from controls with 89% sensitivity and specificity [14]. Later, this result was confirmed by Miyasaka et al. (2010) in 21 children (42 ears) with SNHL [7] with and without CND. The cohort of this study matches ours most closely only with considerably fewer patients (in the CND group 4 instead of 32 patients).

**Table 1** Demographic characteristics of the study population.

<table>
<thead>
<tr>
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<th>CND group (n = 32)</th>
<th>Control group (n = 38)</th>
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<tbody>
<tr>
<td>Ears for evaluation</td>
<td>37</td>
<td>76</td>
</tr>
<tr>
<td>Median age (y)</td>
<td>4.4 (IQR 1.0–6.2)</td>
<td>1.3 (IQR 0.7–3.4)</td>
</tr>
<tr>
<td>No. of males</td>
<td>22 (59 %)</td>
<td>18 (47 %)</td>
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</table>

CND: Cochlear nerve dysplasia; IQR: interquartile range.
Whether the CNC width of 1.5 mm is truly the optimal threshold for the diagnosis of CND has been challenged by the study of Yan et al. [8]. They reported that in one-third of ears with CND (35 patients, 70 ears), the CNC width exceeded 1.5 mm. Nevertheless, the authors maintained a value of 1.5 mm for the initiation of further diagnostics.

Some studies defined thresholds for CNC stenosis in CT without relating to cochlear nerve morphology, while Lim et al. reviewed temporal bone CT examinations of 42 patients with SNHL [15]. They suggested that it is reasonable to set a diameter of 1.2 mm as a cut-off for CNC for the prediction of the affected side in SNHL. Teissier et al. observed in 71 children with SNHL a correlation between CNC diameters below 1.7 mm and SNHL [16]. Of interest, a correlation was also found for CNC diameters above 2.5 mm, in agreement with our results, presumably in patients without CND. Teissier did not differentiate between normal or abnormal cochlear nerve in patients with SNHL. Kono et al. found that among 118 patients with SNHL, a CNC diameter below 1.7 mm should be considered stenotic compared to the healthy contralateral side [17]. This cohort contained only 2 patients with cochlear hypoplasia or aplasia. Regarding the contralateral healthy ears in patients with unilateral CNC stenosis, Vilchez-Madrigal et al. included CT scans from 36 children with CNC stenosis (defined by their own definition smaller than 1.0 mm) [18]. This group found a significantly smaller CNC and cochlea on the contralateral side than in a control group without inner ear anomalies. In contrast, in our CND group, there were no differences in the diameter of the CNC and IAC on the unaffected contralateral side compared to the control group.

Some studies also investigated a combination of the CNC diameter and IAC width. Tahir et al. observed that concomitant

<table>
<thead>
<tr>
<th>Table 2</th>
<th>Dimensions of the CNC and IAC in patients with CND and controls. The mean value and standard deviation are given.</th>
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<tbody>
<tr>
<td></td>
<td>CND group (n = 37 ears)</td>
</tr>
<tr>
<td>CNC (mm)</td>
<td>1.2 (± 0.6)</td>
</tr>
<tr>
<td>IAC width (mm)</td>
<td>4.0 (± 1.2)</td>
</tr>
<tr>
<td>IAC height (mm)</td>
<td>4.7 (± 1.4)</td>
</tr>
<tr>
<td>IAC area (mm²)</td>
<td>15.6 (± 7.9)</td>
</tr>
</tbody>
</table>

CNC: cochlear nerve canal; IAC: internal auditory canal; CND: cochlear nerve dysplasia; ± standard deviation.

<table>
<thead>
<tr>
<th>Table 3</th>
<th>Malformations of the middle and inner ear are associated with CND and were found in ten of the 32 patients with CND. If no laterality for an associated dysplasia is indicated, the side of the CND is affected.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Laterality of CND</td>
<td>Patient-ID</td>
</tr>
<tr>
<td>Right</td>
<td>1y male</td>
</tr>
<tr>
<td>Left</td>
<td>1y male</td>
</tr>
<tr>
<td>11y male</td>
<td>Dysplasia of the ossicles, dysplasia of the vestibulum, and almost complete absence of all semicircular canals on both sides. Normal cochlea.</td>
</tr>
<tr>
<td>4m male</td>
<td>Baller-Gerold syndrome. Incomplete partition II (Mondini malformation), aplasia of the anterior semicircular canal.</td>
</tr>
<tr>
<td>11 m male</td>
<td>Plump ossicles. Dysplasia of the cochlea.</td>
</tr>
<tr>
<td>9m male</td>
<td>Bilateral dysplasia of the vestibulum and bilateral aplasia of the semicircular canals. Normal cochlea on both sides.</td>
</tr>
<tr>
<td>1y female</td>
<td>Charge syndrome, hypoplasia of the tympanic cavity and dysplasia of the ossicles, Mondini malformation on both sides. Bilateral atresia of the semicircular canals.</td>
</tr>
<tr>
<td>4y female</td>
<td>Bilateral dysplasia of semicircular canals. Normal cochlea on both sides.</td>
</tr>
<tr>
<td>2y female</td>
<td>Incomplete partition I on both sides.</td>
</tr>
</tbody>
</table>

CND: cochlear nerve dysplasia, m: months old; y: years old.
stenosis of CNC (below 1.5 mm) and IAC (below 2 mm) was always related to CND [19].

If the threshold of less than 1.5 mm cited above from the literature was used as the criteria for CND diagnosis, about one-third (12/37 ears) of the CNC findings in the current study would have been missed. On the other hand, with our suggested threshold of less than 1.9 mm for the CNC, only one of the 76 ears with a 1.85 mm without CND would have been rated as a false positive, and three ears with CND would have been classified as a false negative. Two of these three ears came from a patient with a complex malformation of the inner ear (incomplete partition type I) and high-grade stenosis of the IAC.

IAC as a predictor of CND

Erkoç et al. presented normative values for IAC by evaluating the MRI results of 3786 healthy adult patients [20]. They reported a mean IAC diameter of 5.93 mm without differences between males and females. The lower limit for considering an IAC stenotic was determined to be 4.7 mm.

Glastonbury et al. considered a narrow IAC indicative of CND in 12 patients [9]. However, whether a narrow IAC can differentiate between patients with CND and those with other causes of SNHL remains unclear. One-half of the ears were below the lower limit (4.7 mm) in our control group despite an intact cochlear nerve. In addition, in one-third of our CND group, the IAC was larger than 4.7 mm.

According to our results, the IAC is poorly suited for the prediction of CND as the cause of SNHL. The overlap of the two groups is too large, meaning that the IAC width does not provide additional certainty in the diagnosis of CND on an individual basis.

Associated malformations

The CND is correlated with additional malformations of the auditory system. Our CND group found an associated malformation of the vestibular organ, cochlea, ossicles, or aquaeductus vestibuli in 40 % of ears. In half of these cases, malformation of the cochlea was involved.

This is consistent with the observations of Lipschitz et al., who investigated radiological abnormalities in children with unilateral deafness [21]. In this group, CND was detected in 42 of 170 patients (24.7 %). Of these, more than half (22/42) presented with another malformation of the inner ear. In 11 and 11 of the 42 patients, combined dysplasia of the cochlea and a large vestibular aqueduct were detected, respectively. Tahir et al. reported hypoplasia of the cochlea in 44/59 ears (75 %) with CNC stenosis [19]. Masuda et al. found associated malformations in the temporal bone CT in 19 of 32 (59 %) patients with CNC stenosis [22]. Together these results show that associated malformations of the bony inner ear should always be sought in the presence of CNC stenosis.

Limitations

The current study has some limitations. In addition to its retrospective nature, the precision in measuring structures in the order of a few millimeters is associated with an inherent inaccuracy, since CT can only provide a spatial resolution of about 0.3–0.5 mm.

Another limitation was the age difference between the CND and control groups. It is arguable that if the age of both groups was balanced, the differences in the IAC dimensions would be larger, rendering the IAC a better parameter. Furthermore, Kim et al. demonstrated that neither width nor height of the IAC, nor width of the CNC correlate with age [11]. Nevertheless, our results demonstrate that a CNC diameter of 1.9 mm discriminates the groups very well, regardless of the child’s age.

Conclusion

This is the first study with a sufficiently large number of patients to provide cut-off values between patients with SNHL due to CND and patients with SNHL due to other underlying pathologies. Previously published cut-off values from patients with SNHL and controls without hearing loss are valid for screening purpose. By contrast we were able to show that in a collective already diagnosed with SNHL and where MRI is contraindicated or when it yields equivocal results due to artifacts or a narrow IAC, a CNC diameter greater than 1.9 mm in CT renders cochlear nerve dysplasia extremely unlikely.

Conflict of Interest

The authors declare that they have no conflict of interest.

References


