ISSN 1857-9345

BONY COCHLEAR NERVE CANAL AS A PREDICTOR FOR COCHLEAR NERVE STATUS IN PRELINGUALLY DEAF CHILDREN

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ABSTRACT

Objectives: The presence of a functional cochlear nerve is a key issue in the preoperative evaluation of pediatric candidates for cochlear implants. Correlations between cochlear nerve deficiency (CND) and bony abnormalities of the labyrinth or bony canal of the cochlear nerve are not yet well understood. The aim of this study was to determine whether the width of the bony cochlear canal (BCNC) can serve as a reliable predictive factor for the existence of a CND.

Materials and methods: A total of 11 children with a confirmed diagnosis of prelingual, severe sensorineural hearing loss were included in this study. In all patients, indication for CI was confirmed and according to the preoperative protocol, high-resolution CT and MR were performed. Reconstructions at a distance of 0.6 mm of the axial plane and images from the HRCT of temporal bones were used for measuring the width of the BCNC. The cochlear nerves were evaluated on axial and sagittal – oblique T2 – MRI images and classified as normal, hypoplastic or aplastic. Two factors were reviewed retrospectively: the presence of inner ear anomalies and the relationship between BCNC stenosis and the existence of CND.

Results: From a total of 22 temporal bones analyzed (22 ears in 11 patients), inner ear malformations were detected in 6 ears from 3 patients (27.27%). All three children had a bilateral malformation, in one it was Michel deformity and in two it was IP2 (incomplete partition 2). The BCNC diameter ranged from 0.1mm to 2.33mm with a mean value of 1.46±0.6mm. CND was recorded in 4 of 22 ears and all were associated with stenosis of the BCNC. In a total of three ears with a stenotic canal, we obtained a normal finding for the cochlear nerve on MR.

Conclusion: Children with BCNC stenosis have a high incidence of CND. A narrowed BCNC on CT can be an indicator for the selection of children with sensorineural hearing loss who will need to be additionally referred for MRI in order to definitively assess the status of the cochlear nerve.

Keywords: bony cochlear nerve canal, profound sensorineural hearing loss, cochlear nerve, children

INTRODUCTION

Sensorineural hearing loss is one of the most common abnormalities present at birth, occurring in 1-3 out of every 1000 newborns [1]. The causes of hearing loss in children can be different. Deafness can be congenital or acquired, and it is thought that up to 20% of all cases of congenital sensorineural hearing loss are due to anomalies of the inner ear that affect the bony labyrinth [2]. With the introduction of universal neonatal hearing screening, congenital deafness can be diag-

nosed in the first months of birth, which allows early treatment and minimization of the negative implications of this anomaly.

Cochlear implants, which enable direct transmission of the sound signal to the neurons of the spiral ganglion and the cochlear nerve, is considered the state-of-the-art treatment of choice for patients with severe sensorineural hearing loss. The main outcome of cochlear implantation in prelingually deaf children is to achieve normal or near-normal speech and language development. But the expected results may differ depending on a series of individual factors as well as the post-implantation rehabilitation and education of these patients.

The presence of a functional cochlear nerve is a key issue in the preoperative evaluation of pediatric candidates for cochlear implants. Conditions for cochlear nerve deficiency, defined as completely absent (aplasia) or thinned (hypoplasia) cochlear nerve, have been described in children with congenital deafness. Children with CDN show poor results after cochlear implantation, and the absence of the cochlear nerve (aplasia) is considered an absolute contraindication for cochlear implant implantation. Previous studies have reported that the benefits of a cochlear implant in patients with CND were poorer than in children with severe sensorineural hearing loss who had normal-sized cochlear nerves, and these varied greatly between individual children [3]. Among CND patients, only some can achieve simple open-set speech perception skills, most patients show only improvements in sound awareness, and some patients may show no benefits after implantation [4].

Determining the caliber of the cochlear nerve as well as determining its status is extremely important for the diagnosis and treatment of sensorineural hearing loss in this group of patients. Currently, the diagnosis of CND is established by precise preoperative radio diagnostic evaluation and direct visualization of the vestibulocochlear nerve (VIII) with magnetic resonance (MR). There is a deficit when the cochlear nerve is absent (aplasia) and cannot be seen in the internal auditory canal (IAC) or is smaller (hypoplasia), then the ipsilateral facial nerve in the IAC [5]. In situations where MR cannot be performed because of the costs its availability and the fact that recording itself takes longer and often requires anesthesia for the pediatric patients, computed tomography (CT) of the temporal bone can provide indirect evidence of existence of CND. It is believed that there is a relationship between the diameter of the BCNC and the size of the cochlear nerve. A stenotic BCNC indicates possible hypoplasia or aplasia of the CN [1].

In the previous study , we determined the borderline values for stenosis of the BCNC, and the purpose of this current study is to make a correlation between the findings of CT and MRI whether the narrowed (stenotic) BCNC can be used as an indicator of the existence of a cochlear nerve deficiency.

MATERIALS AND METHODS

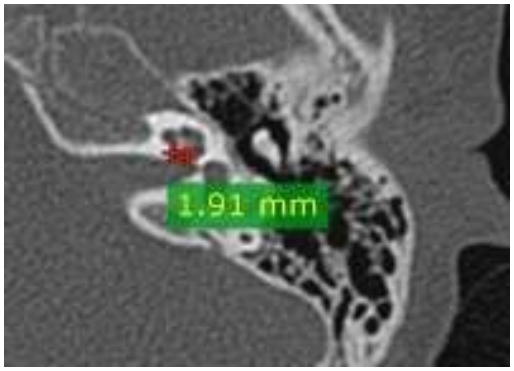
A total of 11 children aged 2 to 7 years, with a diagnosis of prelingual, severe sensorineural hearing loss, candidates for CI, were included in this study. The diagnosis of prelingual, severe sensorineural hearing loss was made using ABR method in the Audiology Center of University Clinic of Ear, Nose and Throat in Skopje, and all of them underwent appropriate preoperative preparation and evaluation.

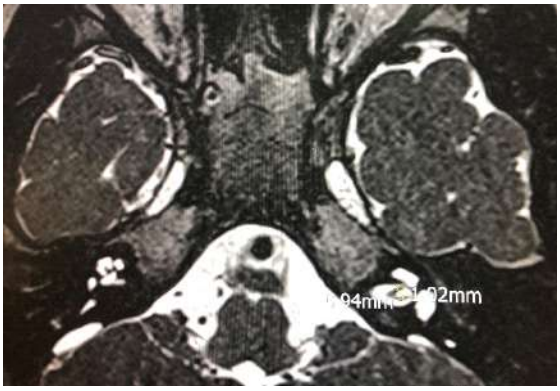
Radiological evaluation

In all patients, according with the preoperative protocol, high resolution computer tomography (HRCT) of the temporal bones and MR of the inner ear were performed in order to show the structures of the bony labyrinth, detect anomalies of the inner ear and determine the status of the cochlear nerve.

HRCT examinations were performed on a 64-slice Somatom Definition AS+ computed tomography scanner (Siemens Healthiness, USA) at the Institute of Radiology following a standard protocol for temporal bone evaluation. BCNC width is a radiological parameter defined as the distance measured between the inner edges of the bony walls of the canal in the mid-section between the base of the modulus and the bottom of the IAC (Figure 1) [6]. For the measurements, the sections in the axial plane, that is, their reconstructions realized at a distance of 0.6 mm, will be used.

In this study, the BCNC width, i.e., a distance measured between the inner margins of the bony walls in the central part of less than 1.5 mm, will be considered as the limit value for stenosis. Then we assume that there is a high probability of the existence of a deficiency of the cochlear nerve [1, 7, 8]. Additionally, each cochlea was examined for bony abnormalities of the inner ear.





In all patients, in order to directly visualize and pre-operatively assess the state of the cochlear nerve in the internal auditory canal, an MRI of the head with an emphasis on the pontocerebellar angle and the IAC (internal acoustic canal) was performed. These recordings were made on a SIMENS Avanto-fit 1.5T magnet, with a matching head coil, in the neutral position, on the back. Fast spin-echo T2-MRI images with high resolution provide an excellent view of the facial (VII) and the three segments of the vestibulocochlear nerve (VIII) – the superior and inferior vestibular as well as the cochlear nerve, in the part of the pontocerebellar angle and the internal auditory canal [9, 10]. These four nerves can be easily recognized in the distal or middle part of the internal auditory canal (MAI) in normal patients, using an oblique plane image with a sagittal view. In the lateral segment of the MAI, the facial nerve lies in the anterosuperior part, the cochlear nerve lies in the anteroinferior part, and the superior and inferior vestibular nerves lie in the posterior part of the canal. The site of separation of the vestibulocochlear nerve into its three components is variable, and the division is complete only in the lateral aspect of the IAC [9, 10]. The cochlear nerves will be evaluated on axial and sagittal-oblique T2-MRI images and the horizontal diameter of the cochlear and facial nerves will be measured at the point closest to the fundus of the MAI (Figure 2). At the same time, the cochlear nerve will be described as normal, hypo, or aplastic. According to Kim et al., the CN is larger than either the superior or inferior vestibular nerve in 90% of normal ears and is of similar size or larger than the facial nerve in 65% of them [10]. Deficiency of the cochlear nerve implies aplasia (absence) or hypoplasia (thinning) of the cochlear nerve. The cochlear nerve will be considered hypoplastic if it is smaller in diameter relative to the adjacent facial nerve at the level of the middle to lateral third of the MAI. The cochlear nerve will be considered aplastic if it is not visible in the IAC in any plane on the MR images.

RESULTS

11 children with congenital, bilateral severe sensorineural hearing reduction (BSHL) were included in the research, of which 8 (72.7%) were female and 3 (27.3%) were male, with an average age of 4 years (age between 2-7 years). From a total of 22 temporal bones analyzed (22 ears in 11 patients), inner ear malformations were detected in 6 ears from 3 patients (27.27%). All three children had a bilateral malformation, and in one it was Michel deformity and in two it was IP2 (incomplete partition 2) (Table 1).

Since the analyzed data represent matched pairs (two measurements were obtained from one patient – left and right BCNC), separate and finally summarized calculations were made for the mean value of the width of the bone canal on the CT images. BCNC diameter ranged from 0.1mm to 2.33mm with a mean value of 1.46±0.6mm. In half of the patients, this diameter was less than 1,575 mm (Table 2).

Stenotic bony cochlear nerve canal, BCNC ≤1.5 mm, was measured in 7 ears (31.82%), 3 (27.27%) on the right and 4 (36.36%) on the left ear (Graph 1). In three cases it was bilateral stenosis and in one it was unilateral. Among patients with bilateral stenosis, one had Michel deformity, one IP2, and in the third patient with bilateral stenosis, no bony labyrinth anomaly was detected. In the second patient with IP2, normal values for the width of the bone canal were measured.

Table 1. *Summarized data from Ct and MRI*

Patient N°	Age/ gender	Inner ear malformation		Width of the BCNC mm				Diameter and status of the CN on MRI			
		right	left	right		left		right		left	
1. A.K	4/f	no	no	0.8	**stenotic**	≤0.5	**stenotic**	0,7	**hypoplastic**	0,1	**aplastic**
2. T. I	4/f	**Michel deformity**	**Michel deformity**	0.2	**stenotic**	0.1	**stenotic**	-	**aplastic**	-	**aplastic**
3. N.B	4/f	no	no	1.59	normal	1.56	normal	1,1	normal	1	normal
4. A. S	4/m	**IP 2**	**IP 2**	1.1	**stenotic**	1.1	**stenotic**	1	normal	1	normal
5. E.I	7/f	no	no	1.77	normal	1.68	normal	1,1	normal	1,04	normal
6. K.K	5/m	**IP 2**	**IP 2**	2.16	normal	1.64	normal	1,06	normal	0,95	normal
7. T. S	4/f	no	no	1.69	normal	1.60	normal	1,15	normal	1,15	normal
8. Z. S	3/f	no	no	1.55	normal	1.56	normal	0,9	normal	1	normal
9. A.Gj	2/f	no	no	1.55	normal	1.44	**stenotic**	0,9	normal	0,9	normal
10. S. A	2/m	no	no	2.02	normal	1.97	normal	1,11	normal	0,95	normal
11. E. B	3/f	no	no	2.22	normal	2.33	normal	0,9	normal	0,9	normal

Table 2

BCNC width in mm	mean±SD	min – max	median (IQR)
right	1.51 ± 0.6	0.2 – 2.22	1.59 (1.1 – 2.02)
left	1.41 ± 0.6	0.1 – 2.33	1.56 (1.1 – 1.68)
total	**1.46 ± 0.6**	**0.1 – 2.33**	**1.575 (1.1 – 1.77)**

On axial and sagittal-oblique T2-MRI images, the diameter of the cochlear nerve was measured in all patients. The values ranged from 0 to 1.15mm, with an average value for the diameter of 0.88±0.3mm. In half of the patients, this diameter was smaller than 0.97 mm (Table 3).

Deficiency of the cochlear canal was diagnosed in 4 (18.18%) ears, that is, 2 (18.18%) patients on the right and left ear. One patient had bilateral aplasia and the other had unilateral aplasia and unilateral hypoplasia.

Out of a total of 22 ears analyzed, 4 had a cochlear nerve deficiency and 18 had a normal cochlear nerve. On HRCT, 7 positive findings were obtained, 4 true positive and 3 false positive findings, 15 negative findings were obtained, all true negative.

Graph 1.

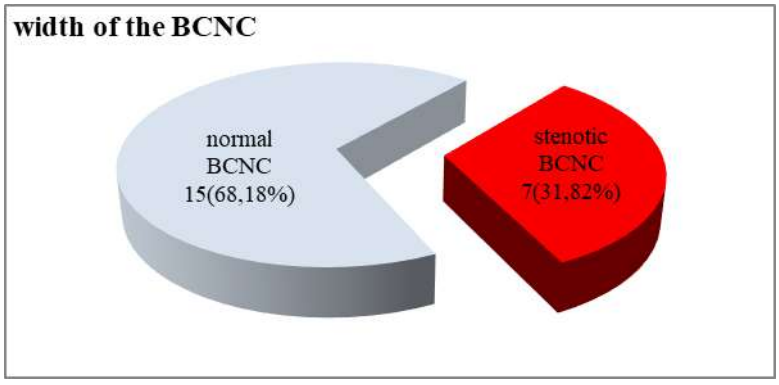


Table 4.

CT	MRI		total
	status of the cochlear nerve		
BCNC width in mm	deficient	normal	
stenotic	4	3	7
normal	0	15	15
total	4	18	22

STATISTICAL ANALYSIS

The statistical analysis of the data obtained from the research was done in the statistical program SPSS 23.0. Categorical (attributive) variables are shown with absolute and relative numbers. Numerical (quantitative) variables are shown with average, standard deviation, minimum and maximum values, median value and interquartile range. The validity of CT in determining the status of the cochlear nerve was analyzed by determining the diagnostic performance (sensitivity, specificity, and global accuracy) of the BCNC width as a radiological marker compared with MR as the gold standard.

DISCUSSION

Cochlear nerve deficiency is one of the common causes of hearing loss, and children with bilateral, severe sensorineural hearing loss who have cochlear nerve deficiency are thought to develop poorer auditory-verbal performance after cochlear implantation compared to those without. A deficiency of the cochlear nerve is considered the condition when we have quite thin and a cochlear nerve with a reduced number of nerve fibers, known as hypoplasia. Or it is also when the cochlear nerve is completely absent, a condition defined as aplasia. According to the literature, this condition of cochlear nerve deficiency occurs in as many as 15.4% of children with congenital deafness [11]. The size of the cochlear nerve is associated with the population of cells in the spiral ganglion. Therefore, the determination of nerve caliber may be useful in predicting the outcome of CI.

Radiodiagnosis, which is a standard procedure in the workup of these patients, allows preoperative evaluation of the auditory nerve. An optimal radio-diagnostic protocol has not yet been defined, as both CT and MRI are methods that are used for that purpose and complement each other. CT is the historical and current method of choice in the diagnostic treatment of patients before implantation in a large number of institutions around the world which deal with this issue. On the other hand, MR has an advantage as a diagnostic method for detecting soft tissue anomalies, especially when it comes to imaging and evaluating the vestibulocochlear nerve. Thus, MRI is accepted as the gold standard for determining the status of the cochlear nerve, but opinion varies regarding the advantages of CT versus MRI as the initial modality for evaluating a candidate for a cochlear implant. In conditions where MR cannot be performed for technical or other reasons (it is a procedure that takes longer and is usually performed with sedation in pediatric patients) high-resolution CT can be of great benefit in assessing the status of the cochlear nerve.

Jackler et al. [14] as well as Shelton et al. [15] suggested that the presence of a narrow IAC on high-resolution CT is indicative for CN aplasia. The vestibulocochlear nerve begins to develop at approximately week 3 of gestation. At the 9th week, the IAC forms with the accumulation of cartilage around the developing nerve. Therefore, the IAC cannot form in the absence of the nerve [5, 16]. In addition, some authors have reported cases of CN aplasia with normal IAC dimensions [17, 18]. The causes of acquired CN deficiency are complex. CN deficiency may result from degeneration of nerve fibers in the IAC following cochlear injury (e.g., vascular, traumatic, compressive, or inflammatory injury). Therefore, findings of a normal-sized IAC with CN deficiency suggest an acquired cause of SNHL [6]. So, IAC width assessment alone is not a sufficient indicator of cochlear nerve assessment.

Fatterpekar et al. [7] reported that BCNC hypoplasia is a possible cause of congenital SNHL. Since then, some authors have reported a relationship between BCNC stenosis and CN hypoplasia [17, 6]. The reason for narrow BCNC is unclear. The BCNC is thought to be formed at the same time as the IAC and requires stimulation for normal development. Since the IAC is formed around the nerve fibers of the VIII-vestibulocochlear nerve, stenosis of the IAC and BCNC is likely due to the absence of normal development of the cochlear nerve [19] and stenosis of the BCNC is secondary to DCN. Fatterpekar first performed BCNC measurements using CT. According to him, the average value for canal width in patients without sensorineural hearing loss is 2.13 ± 0.44 and is significantly higher than that in patients with severe, bilateral sensorineural hearing loss (1.82 ± 0.24) [7]. Stjernholm & Muren performed measurements on silicone molds of 117 temporal bones, and in 16 of them they compared the dimensions measured in this way with the dimensions measured on the corresponding CT images of these samples. According to the results of their measurements, BCNC with a width (< 1.4 mm) is considered to be an extremely

narrow channel, and in that case the possibility of CN abnormality should be considered.

In a previous study by the author of this paper , a correlation was made between the width of the BCNC in children with bilateral severe sensorineural hearing loss compared to children who have normal hearing. It was obtained that the average value of the width of the BCNC canal in children with severe bilateral sensorineural hearing reduction is 1.5± 0.3mm and is statistically significantly lower by $p<0.05$ (t-test=6.62912, p=0.000000), compared to the average value of BCNC (2.1±0.3) mm in patients with normal hearing. The average value of 1.5mm is taken as the borderline width and all values below this will be defined as stenotic or stenotic canal [20].

In this current study, which is a logical follow-up to the previous one, a correlation was made between CT and MRI findings, i.e., a correlation between BCNC stenosis and the existence of cochlear nerve deficiency. BCNC diameter ranged from 0.1mm to 2.33mm with a mean value of 1.46±0.6mm. Stenotic bone canal, BCNC ≤1.5 mm was measured in 7 ears (31.82%), 3 (27.27%) on the right and 4 (36.36%) on the left ear (Graph 1). In three cases it was bilateral stenosis and in one it was unilateral. Cochlear canal deficiency was diagnosed in 4 (18.18%) ears or 2 (18.18%) patients. One patient had bilateral aplasia and the other had unilateral aplasia and unilateral hypoplasia.

From the obtained measurements in all patients with CND, a stenotic bone canal was recorded on CT. Patients with stenotic BCNC on CT were diagnosed with CN deficiency on MRI with 100% sensitivity and 83.33% specificity and a global accuracy of 86.36%. These data agree with the data obtained in the literature. According to a report by Komatzubara et al. in patients who had narrow BCNC on CT, cochlear nerve dysplasia was diagnosed on MR with 88.9% specificity and 88.9% sensitivity. According to the same authors, in patients who have BCNC ≤1.5 mm, it is very likely that cochlear nerve deficit will be demonstrated on MR [21].

Based on these findings, we recommend CT for the initial screening of candidates for cochlear implantation, and the finding of BCNC stenosis can be used as a selective criterion for children with BSHL who should be referred for further evaluation with MRI.

In a report by Casselman et al. [16], CN hypoplasia or aplasia was detected with or without labyrinth abnormalities. The incidence of children with BSHL showing inner ear malformations on CT ranges from 20% to 30% [12, 13]. In this study, malformations of the inner ear were present in 3 of 11 cases (27%) (Table 5). The children had a variety of inner ear malformations. CND was recorded in two out of three children with inner ear malformations (Table 1). These results were similar to those of previous reports [12, 13].

CONCLUSION

An optimal radio diagnostic protocol has not yet been defined. CT is a readily available method of choice in the diagnostic work-up of patients before implantation in a large number of institutions around the world. But since with this method cannot directly show the cochlear nerve, a

Results

Statistic	Value	95% CI
Sensitivity	100.00%	39.76% to 100.00%
Specificity	83.33%	58.58% to 96.42%
Positive Likelihood Ratio	6.00	2.14 to 16.86
Negative Likelihood Ratio	0.00	
Accuracy (*)	86.36%	65.09% to 97.09%

Table 5.

	inner ear malformation		
	right **n (%)**	left **n (%)**	total **n (%)**
Michel deformity	1 (9.09)	1 (9.09)	2(9.09)
IP2	2 (18.18)	2 (18.18)	4(18.18)
normal cochlea	8 (72.73)	8 (72.73)	16 (72.73)

narrowed BCNC on CT can be an indicator for the selection of children with sensorineural hearing loss who will have to be referred to MRI in order to definitively assess the status of the cochlear nerve.

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Резиме

ШИРИНАТА НА КОСКЕНИОТ КАНАЛ ОД КОХЛЕАРНИОТ НЕРВ КАКО ИНДИРЕКТЕН МАРКЕР ЗА ПРЕДВИДУВАЊЕ НА СТАТУСОТ НА СЛУШНИОТ НЕРВ КАЈ ДЕЦА СО ПРЕЛИНГВАЛНА ГЛУВОСТ

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Цели: Присуството на функционален кохлеарен нерв е клучно прашање во предоперативната евалуација на педијатриските кандидати за кохлеарни импланти. Корелациите меѓу недостигот на кохлеарен нерв и коскени абнормалности на лавиринтот или коскениот канал на кохлеарниот нерв сè уште не се добро разјаснети. Целта на оваа студија беше да се утврди дали ширината на коскениот канал од кохлеарниот нерв (BCNC) може да послужи како сигурен предиктивен фактор за постоење дефицит на кохлеарен нерв.

Материјали и методи: Во оваа студија беа вклучени вкупно 11 деца со дијагноза на прелингвално, тешко сензоневрално оштетување на слухот, кај кои беше поставена индикација за вградување кохлеарен имплант. Кај сите пациенти во состав на предоперативниот протокол е реализирана КТ со висока резолуција и МР на пирамиди по протокол. Пресеците во аксијална рамнина од снимките на КТ на пирамиди, односно нивните реконструкции реализирани на растојание од 0,6 mm, ќе се искористат за мерење на ширината на коскениот канал од кохлеарниот нерв (BCNC), а кохлеарните нерви ќе бидат евалуирани на аксијални и сагиталнокоси Т2 – МРИ-снимки и класифицирани како нормален, хипопластичен или апластичен. Ретроспективно ќе бидат разгледани два фактора: евалуација на присуството на аномалии на внатрешното уво и односот меѓу стенозата на BCNC и постоењето дефицит на кохлеарниот нерв.

Резултати: Од вкупно анализираните 22 темпорални коски (22 уши кај 11 пациенти), малформации на внатрешно уво беа детектирани кај 6 уши од тројца пациенти (27,27 %). Кај сите три деца се работеше за билатерано постоење малформација, и тоа кај едно стануваше збор за Michel deformity, а кај две за IP2 (incomplete partition 2). Дијаметарот на BCNC се движеше во ранг од 0,1 mm до 2,33 mm, со просечна вредност од 1,46 ± 0,6 mm. Дефицит на кохлеарниот нерв беше евидентиран кај 4 од 22 уши и сите беа асоцирани со стеноза коскениот канал од кохлеарниот нерв. Кај вкупно три уши со стенотичен канал добивме уреден наод за кохлеарниот нерв на МР.

Заклучок: Децата со стеноза на BCNC имаат висока инциденца на ДКН. Стеснетиот BCNC на КТ може да биде индикатор за селекција на децата со сензоневрално губење на слухот, кои дополнително ќе треба да се упатат на МРИ со цел дефинитивна процена на статусот на кохлеарниот нерв.

Клучни зборови: канал на кохлеарниот нерв, КТ на темпорална коска, тешка сензоневрална редукција на слухот, деца, МР, дефицит на кохлеарниот нерв