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The effect of bony cochlear nerve canal (BCNC) diameter on the degree of sensorineural hearing loss

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ABSTRACT

Purpose: This study investigated the correlation between the diameter of the bony cochlear nerve canal (BCNC), as determined by Temporal bone CT, and MRI findings of cochlear nerves (CN) in children with sensorineural hearing loss (SNHL).
Materials and methods: A prospective study design was followed. Radiological data (Temporal bone CT and MRI) of fifty children with sensorineural hearing loss (age <18 y) were included in the study. All patients (100 ears) underwent routine MRI protocol in addition to 3D CISS (3-D constructive interference in steady state).
Results: Based on CT findings, the BCNC was classified according to its diameter into three groups; group 1 (<1.4 mm), group 2 (1.4–2.0 mm), and group 3 (>2.0 mm). A significant difference between the three groups at degrees of SNHL ($p < 0.001$) was observed. Significant difference ($p < 0.001$) was also observed in the mean level of pure tone audiometry (PTA) average in group 1 compared to group 2. The CN was absent in 20 ears of group 1 CT results (29%), CN hypoplasia was noticed in 40 ears of group 1 CT (58%). However, CN was present in 9 ears of group 1 CT (13%), while in group 2 and 3, CN was present in 100% of the cases (27, and 4 ears, respectively, $p < 0.001$).
Conclusions: MRI and CT imaging are valuable in the diagnosis of SNHL in children. Moreover, with BCNC stenosis, there was a high probability of CN aplasia or hypoplasia.

1. Introduction

The bony cochlear nerve canal (BCNC) is recumbent between the cochlea at its base and the internal auditory canal (IAC) fundus [1]. It endues the nerve fibers from the cochlear nerve to the spiral ganglion [2]. The BCNC width is measured at its midportion between the inside edges of its bony walls. Therefore, narrowing of the BCNC might predispose to functional or anatomic disorders in the cochlear nerve. Indeed, a BCNC width smaller than 1.4 mm in the axial plane has been associated with cochlear nerve abnormalities [1] (see Figs. 1–22).

Computed tomography (CT) and magnetic resonance imaging (MRI) techniques are essential to the pre-operative preparation of patients. CT scanning establishes precise anatomical planning of the surgery detecting the bony frames of the external, middle, and inner ear and the anatomical forms that may affect surgery [3]. On the other hand, MRI is decisive in estimating the cochlear nerve and recognizing the inner ear, especially the labyrinthine part [4].

Recent imaging technology improvements enabled the identification of previously unidentified irregularities, such as the BCNC anomaly, the IAC anomaly [5], and other vestibular anomalies. Especially in the lack of cochlear malformation, BCNC anatomic stenosis may be considered as reason for congenital SNHL [6]. Therefore, the study conducted by Miyasaka and colleagues (2010) aimed to define which inner-ear malformations ordinarily escort BCNC and IAC stenosis and atresia [7]. More recently, a study by Tahir and coworkers (2017) estimated whether stenosis of the IAC and BCNC are consistent with deficiency of cochlear nerve, as investigated by MRI and CT [8].

2. Materials and methods

We performed a prospective study on fifty patients diagnosed with SNHL, chosen from the attendants of the outpatient ENT clinic at Minia University Hospital. Patients were selected after taking history, clinical assessment including full ENT examination and pure tone audiometry

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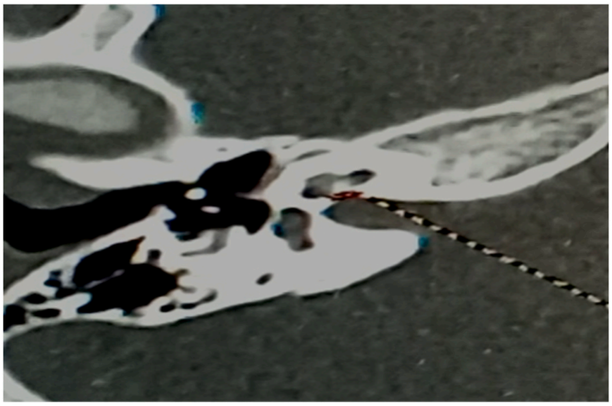

Fig. 1. Temporal bone CT (Axial section) of right ear, diameter of cochlear nerve canal 2.0 mm.

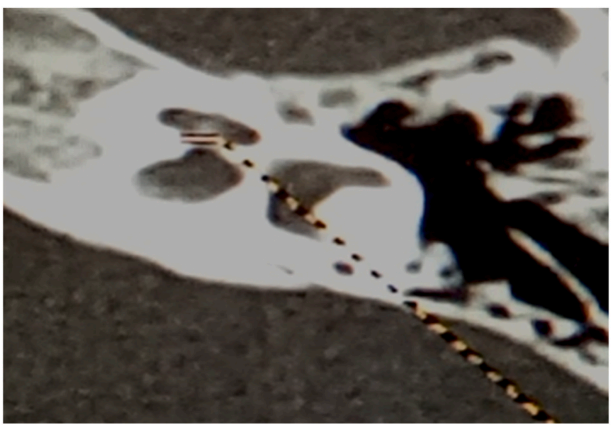

Fig. 2. Temporal bone CT (Axial section) of left ear, diameter of cochlear nerve canal 2.1 mm.

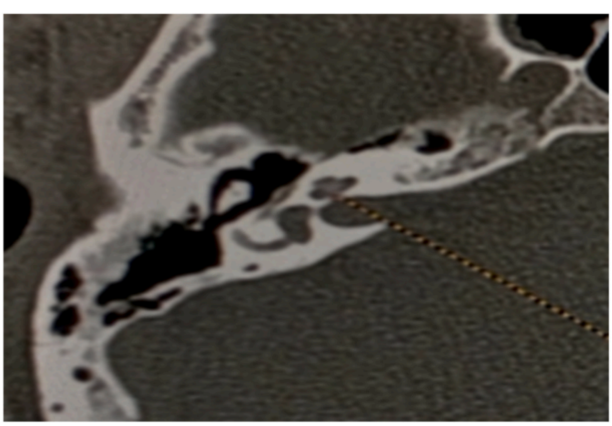

Fig. 3. Temporal bone CT (Axial section) of right ear, diameter of cochlear nerve canal 1.0 mm.

(PTA), then referred to the Radiology Department of Minia University Hospital for radiological assessment (Temporal bone CT and MRI).

The Committee for Medical Research Ethics approved the study. All patients' parents signed a written consent prior to being included in the research.

The following inclusion criteria were adopted.

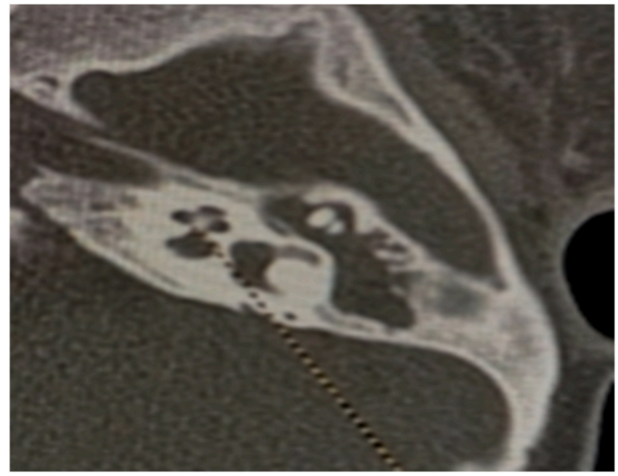

Fig. 4. Temporal bone CT (Axial section) of left ear, diameter of cochlear nerve canal 0.5 mm.

- Bilateral sensorineural hearing loss in children proved with audiometry PTA.
- Patients with age ≤18 years old.
- History suggestive of congenital SNHL, with no history of acquired causes as trauma or infection.

The exclusion criteria were as follows.

- General contraindication to MRI.
- Patients of age more than 18 years.
- Any case with mixed hearing loss.
- Any case with previous otological operations, cochlear implantation, ear trauma and ear infections.

2.1. All patients were subjected to

1. **Full history taking:**

   With special emphasis on clinical and family history and past history.

2. **Complete ENT examination.**
3. **Audiological evaluation:**

   That revealed bilateral sensorineural hearing loss.

4. **Imaging procedures:**

All patients underwent CT and conventional sagittal MRI of the petrous bone. The axial plane was performed in the same time with the infraorbitometal line. The BCNC diameter was investigated and its width at midportion, as previously reported, was assigned as the dimension between bony walls of BCNC at its inner margins.

According to the BCNC width, 50 children were assorted into three different groups: Group 1 (the BCNC width is < 1.4 mm, $n = 69$), Group 2 (the BCNC width is between 1.4 mm and 2.0 mm, $n = 27$), and Group 3 (the BCNC width is greater than 2.0 mm, $n = 4$).

3. Statistical analysis of data

Data were presented as the mean ± standard deviation (SD). Data were analyzed unsing the SPSS 20.0 software package. The differences were deemed significant at p-values less than 0.05.

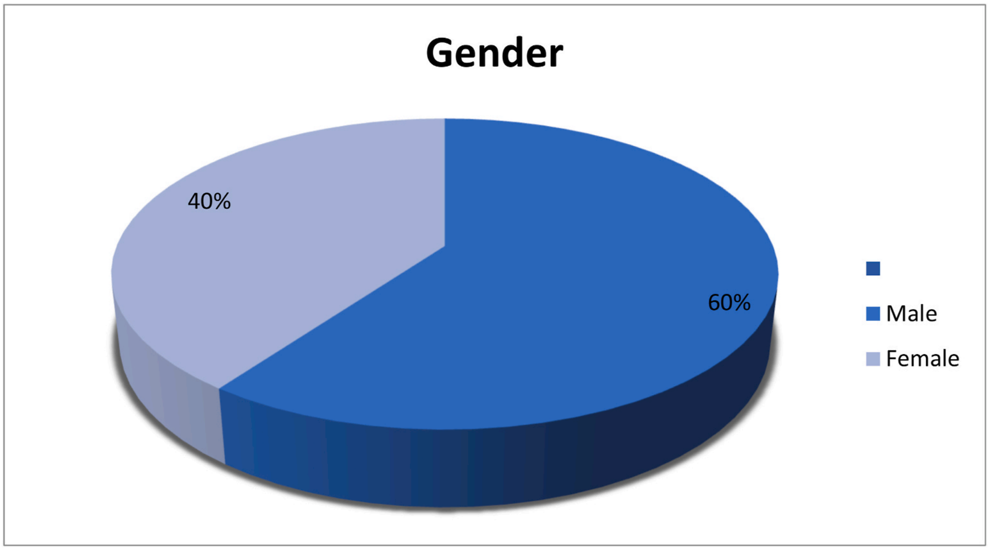

Fig. 5. Pie chart showing sex distribution among patients cohort.

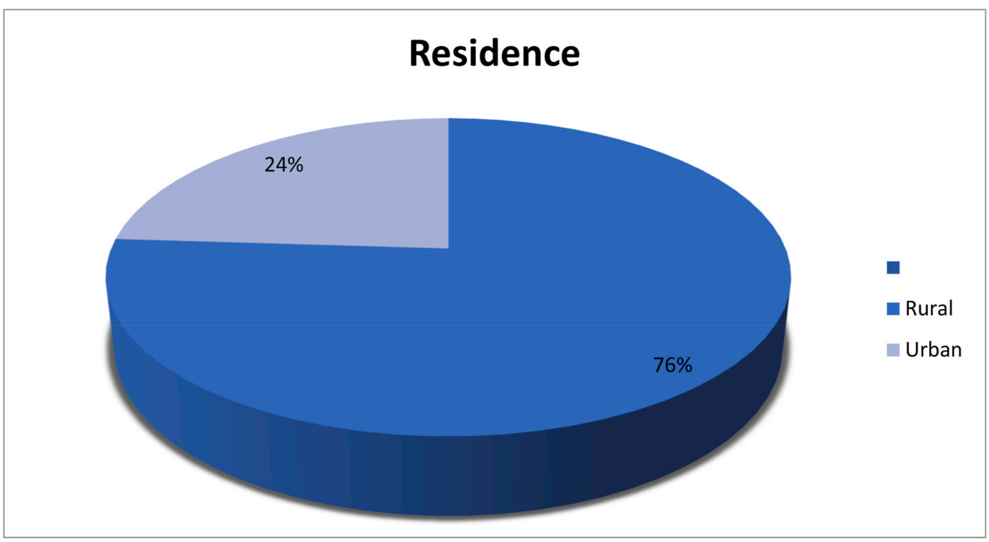

Fig. 6. Pie chart showing residence distribution among patients' cohort.

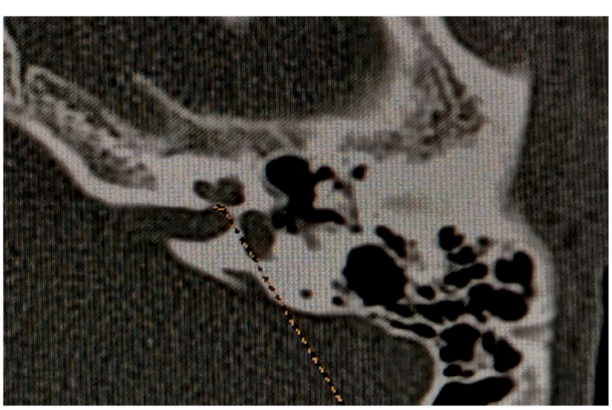

Fig. 7. Temporal bone CT (Axial section) of left ear, diameter of cochlear nerve canal 0.8 mm.

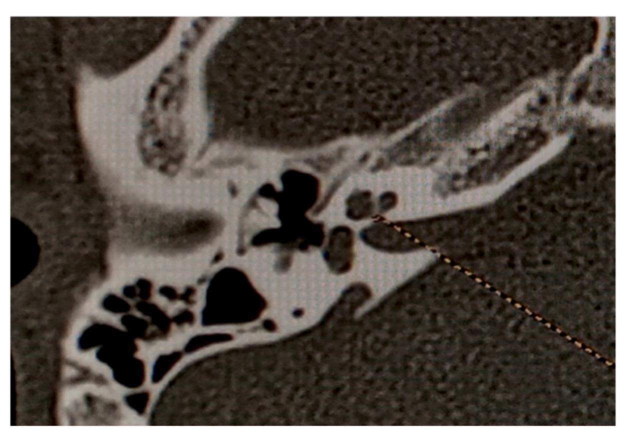

Fig. 8. Temporal bone CT (Axial section) of right ear, diameter of cochlear nerve canal 0.8 mm.

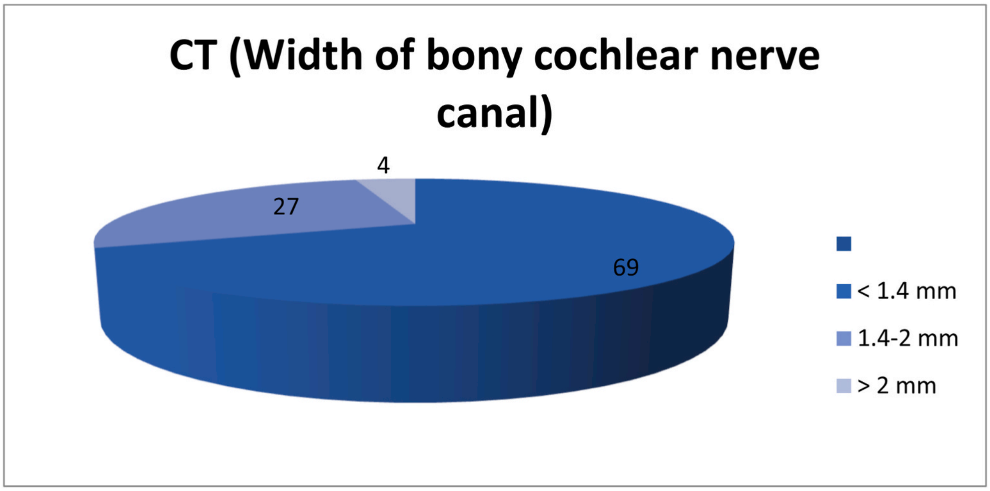

Fig. 9. Pie chart showing CT (width of bony cochlear nerve canal) among patients' cohort.

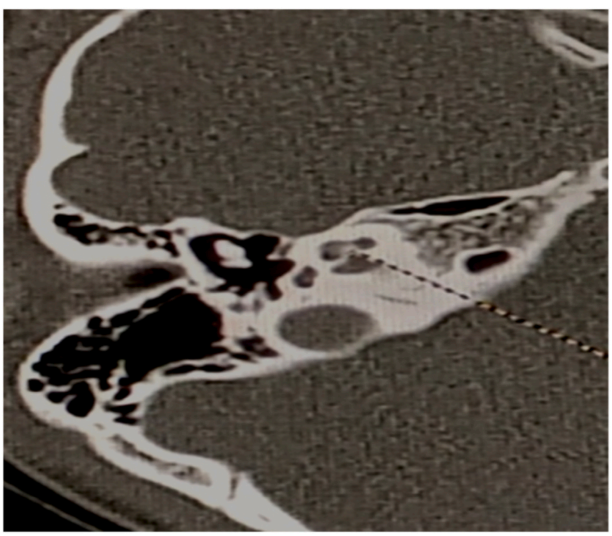

Fig. 10. Temporal bone CT (Axial section) of right ear, diameter of cochlear nerve canal 0.9 mm.

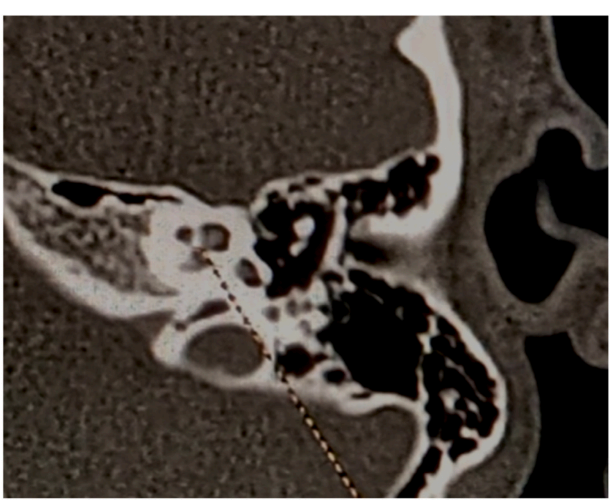

Fig. 11. Temporal bone CT (Axial section) of left ear, diameter of cochlear nerve canal is narrow about 0.8 mm.

4. Results

This study included a total of 50 patients (100 ears) comprising 30 males (60 ears; 60%) and 20 females (40 ears; 40%) diagnosed with bilateral sensorineural hearing loss (SNHL). Their ages ranged between 5 and 18 years (10.3 ± 3.9), as explained in Table 1. Patients were referred from the audiology unit at the ENT Department to the Radiology Department at Minia University Hospital.

4.1. Results of CT examination of the patient cohort

All patients enrolled in this study underwent CT and conventional sagittal MRI of petrous bone. The CNC diameters ranged from 0.3 mm to 2.5 mm, the mean diameter of CNC was 1.2 mm. Based on the BCNC width classification, the CT data revealed significant differences in the number of ears among the three groups. The results in Table 2 show that group 1 (<1.4 mm) comprised 69 ears (69%), group 2 criteria (1.4–2.0 mm) were established in 27 ears (27%), while group 3 (>2.0 mm) contained only 4 ears (4%). The BCNC width of all cases ranged from 0.3 to 2.5, with a mean of 1.2 and a SD of 0.4 (Table 2).

4.2. Results of audiological finding of the patients cohort

All the 50 patients underwent routine audiologic assessments. However, some cases were investigated by pure tone audiometry (PTA) and auditory brainstem response (ABR) testing. in some cases.

The results of the current study revealed that one ear (1%) had moderate sensorineural hearing loss, 18 ears (18%) with moderately severe sensorineural hearing loss, 30 ears (30%) with severe sensorineural hearing loss, and 51 ears (51%) with profound sensorineural hearing loss (Table 3).

In pure tone audiometry (PTA) of 100 ears at threshold 250 Hz range from (40–110) with mean 75.8 and SD ± 17.3, ears at threshold 500 Hz range from (50–120) with mean 85.3 and SD ± 17.7, ears at threshold 1000 Hz range from (55–120) with mean 91.1 and SD ± 17.5, ears at threshold 2000 Hz range from (55–120) with mean 97.7 and SD ± 17.6, ears at threshold 4000 Hz range from (50–120) with mean 101.1 and SD ± 17.4, at threshold 8000 Hz range from (60–120) with mean 100.1 and SD ± 14.6, PTA average range from (53.3–116.7) And mean 91.9 sd ± 15.4. as explained in Table 3.(see. Tables 4,5 and 6)

Significant difference was observed in the mean level of PTA threshold at 250 Hz in group 1 (<1.4 mm) if compared to group 2(1.4–2 mm) with p value (<0.001), Significant difference was also observed in the mean level of PTA threshold at 500 Hz in group 1 (<1.4 mm) if compared to group 2(1.4–2 mm) with p value (<0.001).

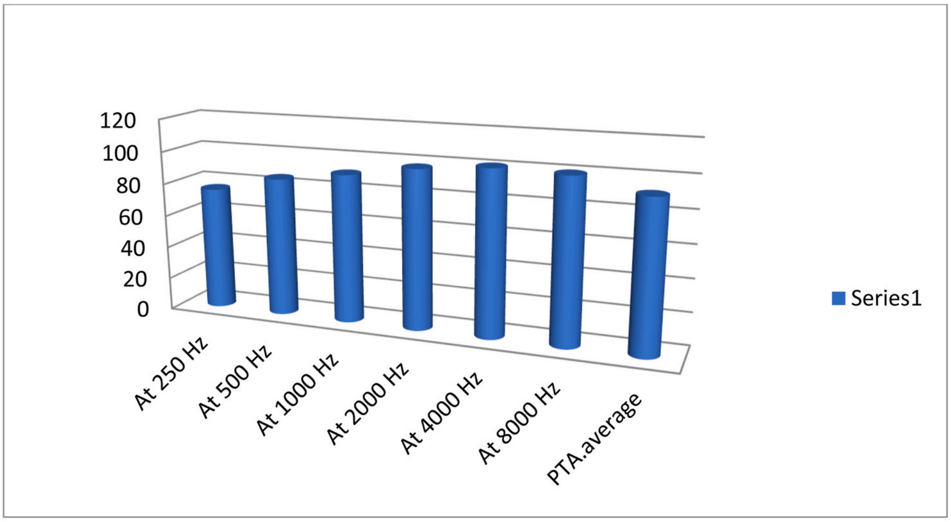

Fig. 12. Mean of pure tone audiometry at each threshold.fx13

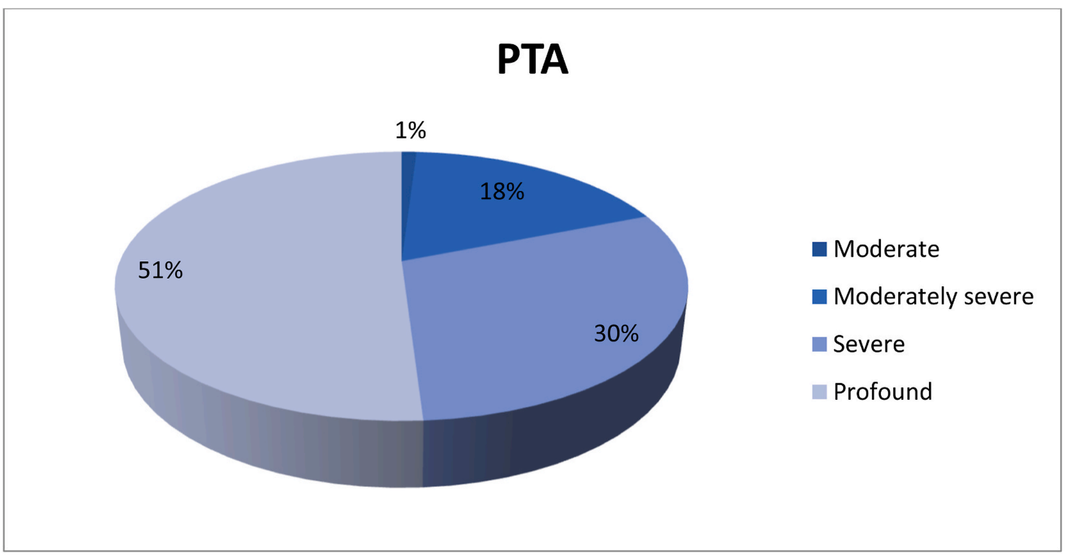

Fig. 13. Pie chart of degree of pure tone audiometry.

Significant difference was observed in the mean level of PTA threshold at 1000 Hz in group 1 (<1.4 mm) if compared to group 2 (1.4–2 mm) with p value (<0.001), Significant difference was also observed in the mean level of PTA threshold at 2000 Hz in group 1 (<1.4 mm) if compared to group 2(1.4–2 mm) with p value (<0.001).

There was significant difference in the mean level of PTA threshold at 4000 Hz in group 1 (<1.4 mm) if compared to group 2(1.4–2 mm) with p value (<0.001), There was significant difference in the mean level of PTA threshold at 8000 Hz in group 1 (<1.4 mm) if compared to group 2 (1.4–2 mm) with p value (<0.001).

There was significant difference ($P < 0.001$) in the mean level of PTA average in group 1 (<1.4 mm) if compared to group 2(1.4–2 mm). A significant difference was observed between the 3 groups of CNC diameter at degrees of SNHL with p value (<0.001).

Out of 69 ears in the BCNC diameter(<1.4 mm), 47 ears presenting (68.1%) group had profound hearing loss. 18 ears presenting (26.1%) out of 69 ears had severe hearing loss. 4 ears presenting (5.8%) out of 69 ears in BCNC stenosis group showed moderately severe hearing loss. In the same group, the pure tone average of 500, 1000, 2000, 4000 and 8000 Hz range from (62.5–116.7) [a] with mean 96.9 dB HL.

Out of 27 ears with BCNC diameter (1.4–2.0 mm),1 ear moderate hearing loss presenting (3.7%) 12 ears moderately severe hearing loss presenting (44.4%), 10 ears severe SNHL presenting (37%),4 ears profound SNHL presenting (14.8%). In the same group, the pure tone average of 500, 1000, 2000, 4000 and 8000 Hz range from (53.3–106.7) with mean 79.3 dB HL.

Out of 4 ears with BCNC diameter (>2.0 mm),2 ears moderately severe hearing loss presenting (50%), 2 ears severe sensorineural hearing loss presenting (50%). In the same group, the pure tone average of 500, 1000, 2000, 4000 and 8000 Hz range from (81.7–95) with mean 89.2 dB HL.

4.3. *Results of MRI of the patients cohort*

All 50 patients (100 ears) undergo MRI using the routine protocol in addition to 3D CISS (3-D constructive interference in steady state).

The CN of ears could be investigated by MRI. CN was absent in 20 ears of group 1 CT (<1.4 mm) presenting (29%), CN hypoplasia was noticed in 40 ears of group 1 CT (<1.4 mm) presenting (58%). CN was present in 9 ears of group 1CT (<1.4 mm) presenting (13%), in 27 ears of

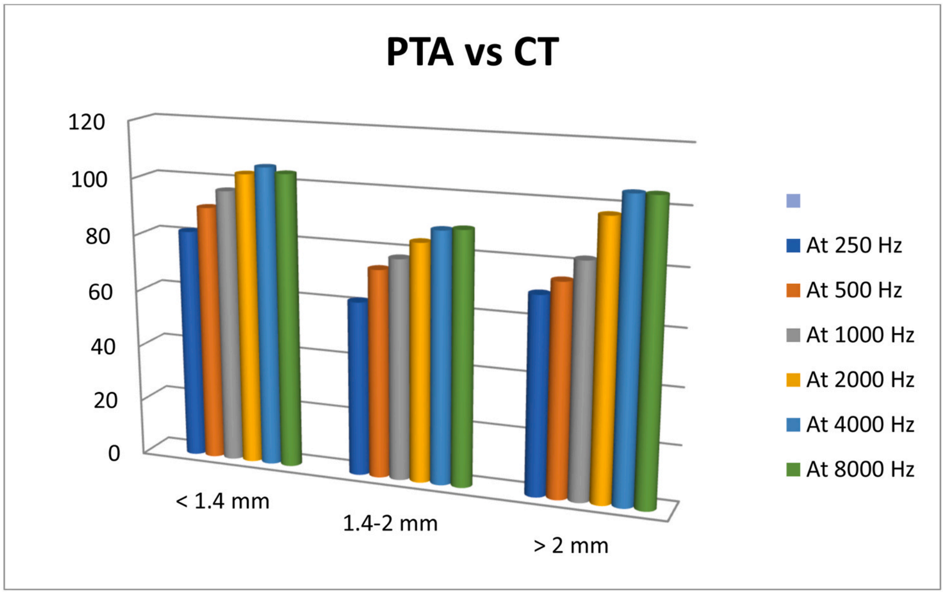

Fig. 14. Histogram of Distribution of CT and PTA findings among 50 patients with SNHL.

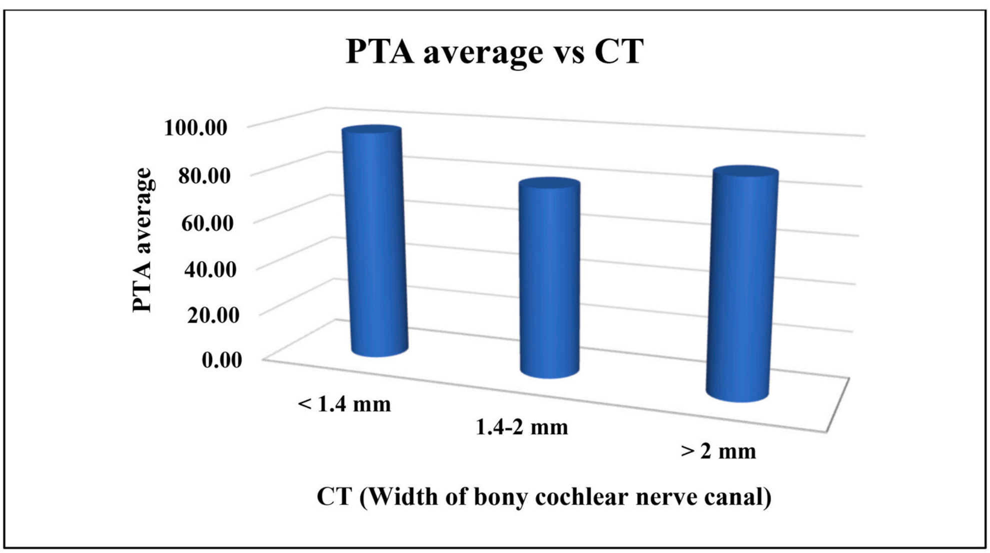

Fig. 15. Histogram of Distribution of CT and PTA average findings among 50 patients with SNHL.

group 2 CT (1.4–2.0 mm) presenting (100%) 0f them, and in group 3 CT (>2.0 mm) presenting 4 ears (100%) of them. With p value < 0.001.

Sixty Ears with aplasia or hypoplasia of CN were accompanied with stenosis of CNC (diameter <1.4 mm). The remaining 40 ears had a normal CN, 9 ears with CNC diameters >1.4 mm had normal CN.

Normal CN was observed in 27 ears, despite the CNC stenosis (<1.4 mm) in them. A smaller CNC was observed with CN aplasia or hypoplasia compared to that with a normal CN.

There was significant difference in group 1 of CNC diameter in CT (<1.4 mm) and CN in MRI if compared with 2 other groups and there is significance between CNC stenosis and CN hypoplasia/aplasia with p value (<0.001).

5. Discussion

In children and infants, different pathologic conditions can lead to SNHL [9]. Recent researchers have observed that dysfunction of cochlear nerve in children interprets about 10% of recently confirmed patients with SNHL [10].

The bony labyrinth is developed around the growing membranous labyrinth as a cartilaginous aggregation of mesenchyme. Its development begins in the fourth week of pregnancy, whereas the otic capsule expands between the eighth and sixteenth weeks and subsequently ossifies between the sixteenth and twenty-fourth weeks. It is substantial to know that the expansion of the bony labyrinth is subordinate to the right expansion of the vestibulocochlear nerve and the membranous labyrinth. [11].

CNC is known by different names, like, BCNC, cochlear fossette and cochlear aperture. Furthermore, it is unclear what is narrow vs normal width of CNC. But there are two clear points [1]: narrowing of CNC is usually accompanied with aplasia or hypoplasia of the CN, and [2] SNHL patients have narrower CNCs than those with normal hearing. Patients with profound SNHL have been evaluated in many previous studies considering CNC stenosis. Interestingly, Wilkins et al., 2012 revealed a broad range of level of hearing in patients with CNC stenosis, varying between sparingly severe hearing loss and profound hearing loss.[12].

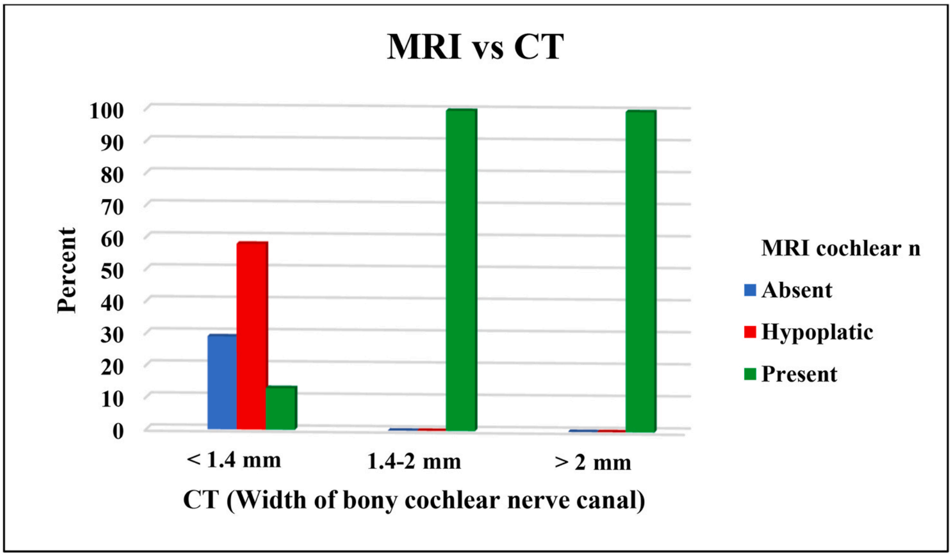

Fig. 16. Histogram of the relationship between CNC stenosis and CN hypoplasia/aplasia.

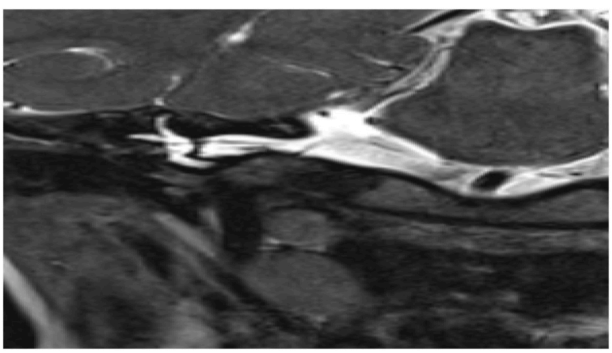

Fig. 17. Axial MRI image, the cochlear nerve is normal.

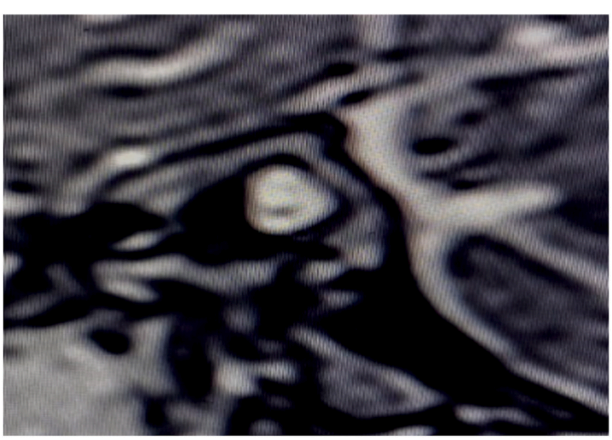

Fig. 18. Oblique sagittal MRI image, the cochlear nerve is normal.

Imaging exerts an essential role In the field of malformations of the inner ear.[13]. Conventionally, HRCT is the imaging technique of preference in their work-up [14]. However, for the soft-tissue abnormalities of the facial-vestibulocochlear nerves, MRI has priorities in investigating them [15], which are best investigated on 3-D CISS (3-D constructive interference in steady state) or 3-D FRFSE (3-D fast recovery fast spin-echo) images [16]. Therefore, in patients with inner ear

Fig. 19. Oblique sagittal MRI image, the cochlear nerve is hypoplasia.

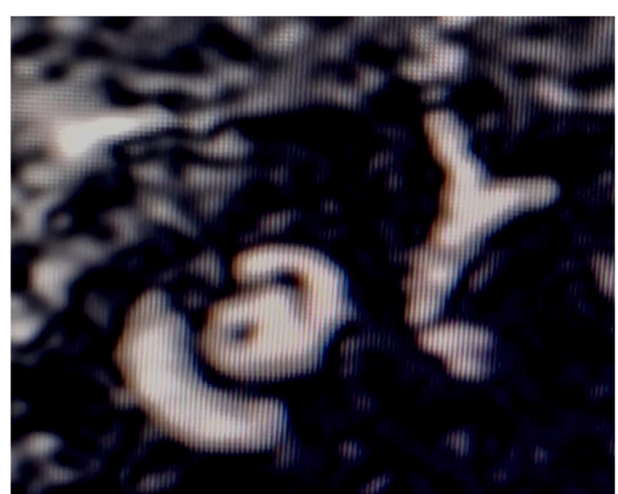

Fig. 20. Oblique sagittal MRI image, the cochlear nerve is hypoplasia.

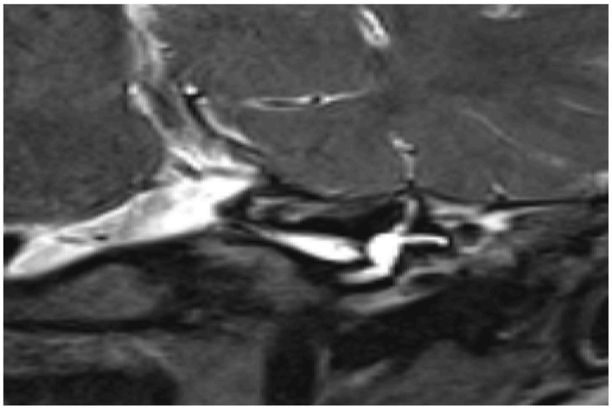

Fig. 21. axial MRI image, the cochlear nerve aplasia.

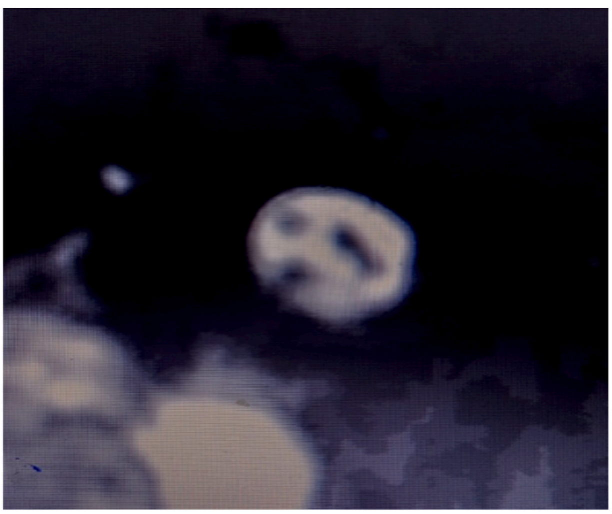

Fig. 22. Oblique sagittal MRI image, the cochlear nerve is hypoplastic.

Table 1
Demographic data of 50 patients with SNHL.

		Descriptive statistics, N = 50
Age	*Range*	[5–18]
	Mean ± SD	10.3 ± 3.9
Gender	*Male*	30(60%)
	Female	20(40%)
Residence	*Rural*	38(76%)
	Urban	12(24%)

Table 2
CT findings in 50 patients with 100 ears complaining of SNHL.

		Descriptive statistics N = 100
Width of bony cochlear nerve canal	*Range*	(0.3–2.5)
	Mean ± SD	1.2 ± 0.4
CT	< *1.4* mm	69(69%)
	1.4-2 mm	27(27%)
	> 2 mm	4(4%)

malformations, MRI is considered as the modality of choice to investigate abnormalities. Researchers have different opinions regarding the priorities of HRCT or MRI as a premier imaging technique for inner ear malformations [15].

Table 3
Pure tone audiometry in 50 patients (100 ears) with SNHL.

		Descriptive statistics N = 100
At 250 Hz	*Range*	(40–110)
	Mean ± SD	75.8 ± 17.3
At 500 Hz	*Range*	(50–120)
	Mean ± SD	85.3 ± 17.7
At 1000 Hz	*Range*	(55–120)
	Mean ± SD	91.1 ± 17.5
At 2000 Hz	*Range*	(55–120)
	Mean ± SD	97.7 ± 17.6
At 4000 Hz	*Range*	(50–120)
	Mean ± SD	101.1 ± 17.4
At 8000 Hz	*Range*	(60–120)
	Mean ± SD	100.1 ± 14.6
PTA average	*Range*	(53.3–116.7)
	Mean ± SD	91.9 ± 15.4
Hearing affection degree	Moderate	1(1%)
	Moderately severe	18(18%)
	Severe	30(30%)
	Profound	51(51%)

Table 4
Distribution of CT and PTA findings among 50 patients with SNHL.

		CT (Width of bony cochlear nerve canal)			P value
		<1.4 mm	1.4–2 mm	>2 mm	
		N = 69	N = 27	N = 4	
At 250 Hz	*Range*	(40–110) [a]	(40–90) [b]	(50–80)	***<0.001****
	Mean ± SD	81.5 ± 15.4	61.9 ± 14.2	70 ± 14.1	
At 500 Hz	*Range*	(50–120) [a]	(50–105) [b]	(60–80)	***<0.001****
	Mean ± SD	90.4 ± 16.7	73.7 ± 15.3	75 ± 10	
At 1000 Hz	*Range*	(60–120) [a]	(55–110) [b]	(70–90)	***<0.001****
	Mean ± SD	96.7 ± 15.6	78 ± 15.6	82.5 ± 9.6	
At 2000 Hz	*Range*	(70–120) [a]	(55–120) [b]	(90–100)	***<0.001****
	Mean ± SD	103 ± 15.2	84.1 ± 17.5	97.5 ± 5	
At 4000 Hz	*Range*	(60–120) [a]	(50–120) [b]	(100–110)	***<0.001****
	Mean ± SD	105.8 ± 14.9	88.7 ± 18.7	105 ± 5.8	
At 8000 Hz	*Range*	(70–120) [a]	(60–120) [b]	(100–110)	***<0.001****
	Mean ± SD	104 ± 12.1	89.4 ± 16.3	105 ± 5.8	

Between the three groups, One Way ANOVA test was performed for the parametric quantitative data, followed by post hoc LSD test between each two groups.
Superscripts with small different letters refer to significant difference between the two groups.
*: Significant level at P value < 0.0 1.

The CNC is reported as a sensitive and measurable index of malformation of the cochlear nerve. Furthermore, abnormalities of CNC may be considered as a kind of malformation of the cochlea [17]. Therefore, estimation of CNC diameters is worthy for investigating the aplasia or hypoplasia of the cochlear nerve [18].

Fatterpekar et al. [17] observed that congenital SNHL may result from BCNC stenosis. Additionally, they showed that the BCNC diameter was shorter compared to the control group. Also, compared to age-matched group, Kono [11], in the unilateral SNHL group, observed BCNC stenosis with diameter <1.7 mm (2 SD) suggesting hypoplasia. [5].

According to previous studies done by Stjernholm and Muren (2002) and Miyasaka et al. (2010), diameter of BCNC of 1.4 mm or 1.5 mm to was used to indicate hypoplasia, [7,19].

Profound hearing loss was associated with BCNC absence in patients. In addition, this study revealed a confirmed tendency as the hearing dB became worse and BCNC diameter was narrower. That is why, for

Table 5
Distribution of MDCT and PTA average findings among 50 patients with SNHL.

		CT (Width of bony cochlear nerve canal)			P value
		<1.4 mm	1.4–2 mm	>2 mm	
		N = 69	N = 27	N = 4	
PTA average	**Range**	(62.5–116.7) [a]	(53.3–106.7) [b]	(81.7–95)	***<0.001****
	Mean ± SD	96.9 ± 13.1	79.3 ± 14.7	89.2 ± 5.7	
Hearing affection degree	*Moderate*	0(0%) [a]	1(3.7%) [b]	0(0%) [b]	***<0.001****
	Moderately severe	4(5.8%)	12(44.4%)	2(50%)	
	Severe	18(26.1%)	10(37%)	2(50%)	
	Profound	47(68.1%)	4(14.8%)	0(0%)	

Between the three groups, One Way ANOVA test was performed for the parametric quantitative data, followed by post hoc LSD test between each two groups.
Fisher's exact test was performed for qualitative data between the groups.
Superscripts with small different letters refer to significant difference between the two groups.
*: $P < 0.05$.

Table 6
The correlation between CNC stenosis and CN aplasia/hypoplasia.

		CT (Width of bony cochlear nerve canal)			P value
		<1.4 mm	1.4–2 mm	>2 mm	
		N = 69	N = 27	N = 4	
MRI (cochlear nerve)	*Absent*	20(29%) [a]	0(0%) [b]	0(0%) [b]	***<0.001****
	Hypoplastic	40(58%)	0(0%)	0(0%)	
	Present	9 (13%)	27 (100%)	4 (100%)	

Between all groups, Fisher's exact test was performed for qualitative data.
Superscripts with small different letters refer to significant difference between the two groups.
*: $P < 0.05$.

estimating the cochlear nerve, a study proposed that an axial CT scan with MRI was adequate [5].

Wilkins et al. [12] was the first to examine the correlation between the grade of cochlear stenosis and the degree of hearing loss. That study observed an enormous ambit in CNC stenosis diameter, as well as an enormous ambit in hearing loss. They revealed that the smaller the axial measurement of the CNC, the higher the degree in hearing loss. In the current study, the outcomes of the audiologic evaluation also lend support to this theory. In addition, authors revealed a confirmed tendency as the hearing dB became worse when BCNC diameter was narrower.

However, in almost all conditions, CT can be quickly and easily used. In addition, CNC stenosis (diameter <1.5 mm) assisted in emphasizing the probably diagnosed CN aplasia or hypoplasia [20]. If cochlear malformation or CNC stenosis is observed on HRCT, further MRI might show CN hypoplasia or aplasia in these nominated children. MRI can emphasize the case of CN, whether there is hypoplasia or aplasia [7].

CN hypoplasia might be a cause for CNC stenosis [11]. It was reported that the probability of CN aberration should be taken into consideration if the diameter of CNC was <1.4 mm [19]. Furthermore, on CT investigations, patients with a narrow CNC were known to have hypoplasia of CN on MRI with specificity and sensitivity that reached about 88.9% [20]. In the same study, they stated that CN hypoplasia was observed on MRI in ears with CNC <1.5 mm on CT. In another study by other researchers, a diameter of CNC <1.7 mm indicated hypoplasia of CN, although no abnormalities of the cochlea was observed on CT [11].

Within the third week of pregnancy, the vestibulocochlear nerve develops. Additionally, around the ninth week of pregnancy, the vestibulocochlear nerve develops concurrently with the cartilaginous IAM [18,21]. Many researchers stated that the vestibulocochlear nerve's absence led to IAM stenosis or aplasia, while others reported that nerve expansion might lead to cartilage development [14].

In 2006, Adunka et al. [10] revealed that IAC hypoplasia was not usually accompanied with cochlear nerve aplasia. They also demonstrated the need for high-resolution CT and MRI in cases of profound SNHL.

This study did not depend on the size of internal auditory canal.

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Declaration of competing interest

No conflict of interest.

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