Miniature cochlea: a study of radiological measurements and its implications during the cochlear implant surgery

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Received 5 March 2016
Accepted 31 March 2016

The Egyptian Journal of Otolaryngology
2016, 32:170–177

Hypothesis
Approximately 25% of patients with congenital sensorineural or mixed hearing loss show bony inner ear malformations on computed tomography (CT) of the temporal bone, with significantly smaller cochlear height (CH) compared with normal-hearing patients. The miniature cochlea has an apparently normal radiological appearance and could be missed if proper measurements are not taken. Inner ear measurements not only aid in the duplication of radiologically diagnosed inner ear malformation but can also provide additional information about which specific part is abnormal.
Aims
The aims of the present study were to establish the normal measurements of the height and width of fully developed Egyptian cochleae using high-resolution CT scans of the temporal bone in normal individuals, and to predict the characteristic radiologic sings and measurements of miniature cochlea and its implication during cochlear implant surgery.
Study design
The authors conducted a prospective, comparative study.
Patients and methods
The study comprised three groups: the control group, which comprised 50 cochleae with normal hearing and negative history for head trauma or ear surgery; the patients group, which comprised candidates for cochlear implanting with a negative history for meningitis or head trauma, and was further subclassified into group A, which comprised 22 cochleae with CH small but not less than 2 SD from the controls, and group B, which comprised nine cochleae with CH less than 2 SD from the controls. High-resolution 64-slice CT scan of 1 mm slice thickness was obtained; in addition, CH, basal turn height and width, upper turn height and width, and oval window–round window distance were estimated according to well-established definitions in the literature.
Results
CH in the control group ranged between 4.8 and 6.9 mm, with a mean of 6.1 ± 0.29 mm, whereas, in group A, CH showed a mean of 5.9 ± 0.34 mm, which was statistically significant compared with the control group. In group B, CH showed a mean of 3.8 ± 0.31 mm, which was also statistically significant compared with the other two groups. The basal turn width, upper turn width, and oval window to the round window distance – all showed statistically significant difference when compared within the three groups. In contrast, both basal turn height and upper turn height showed no significant difference when compared within the three groups.
Conclusion
In cochlear implant surgery, the detailed and good radiological analysis aids in a safe, effective, and well-planned surgery. The development of standardized measurements to complement visual inspection improved the diagnostic accuracy and helped in the subclassification of hypoplastic cochleae. We found CH to be the most essential differentiating measurement in the subclassification and diagnosis of hypoplastic and dwarf cochleae. If dwarf cochlea is radiologically diagnosed, special surgical modifications regarding electrode length and site of cochleostomy during cochlear implanting should be applied.

Keywords:
cochlear height, cochlear implant, cochlear measurements, dwarf cochlea, hypoplastic cochlea, miniature cochlea

Egypt J Otolaryngol 32:170–177

1012-5574

Introduction

The precise knowledge of the normal anatomy and the anatomic variations of the normal human cochleae are one of the most essential basic skills for a cochlear

implant surgeon to have so as to have a significant effect on improving surgical outcome. Approximately 25% of the patients with congenital sensorineural or mixed hearing loss show bony inner ear malformations on computed tomography (CT) of the temporal bone, with significantly lesser cochlear height (CH) compared with normal-hearing patients [1,2].

Little information can be found in the literature about miniature cochlea (dwarf cochlea); it was usually included in descriptions that were broader in scope – 'hypoplastic cochlea'. Early on, it was described by Schuknecht[3] as a two-and-half turns cochlea only 20 mm or less in length, with well-developed ducts. Other researchers reported associated minor abnormalities in the semicircular canals, and/or vestibular aqueduct, with incidence ranging between 7% [4] and 36% [5]. The most accepted theory of miniature cochlea development was the arrest of development after the 6-week intrauterine life.

More recently, a modified classification system for cochlear malformations based upon radiographic findings has gained popularity (Table 1) [4].

Strong evidence exists in the literature to prove that there is no cochlear growth postnatally, and that CH is smaller in patients with developmental sensorineural or mixed hearing loss than in patients with normal hearing. Some researchers proved that CH varies according to sex, with males having slightly greater measurements than do females. Thus, normative sex-specific CH measurements can be used across all ages to diagnose cochlear hypoplasia using 2 SDs below the mean; CH that is below 4.48 mm in males and 4.25 mm in females is a useful threshold to define cochlear hypoplasia resulting in hearing loss [6].

The anomalies of the cochlea that can be visualized by using CT are related to its morphology, size, patency, and extent of the lumen, the modiolus, and the bony partitions between the coils [7]. Severe malformations can be easily diagnosed by visual inspection of the radiographs, but this is often inadequate to diagnose subtle abnormalities, which are highly dependent on the experience of the examiner. The development of normative radiographic measurements has proved to be invaluable in the evaluation of the disease process in many parts of the body; consequently, the establishment of normative data for the dimensions of inner ear structures using high-resolution computed tomography (HRCT) imaging allow for consistent and accurate assessment of patients of all ages [2]. Each passing day, a detailed radiological anatomical measurement about dwarf cochlea for specific races is being added to the literature. Fernando *et al*. [8] provided detailed measurement of the Filipino's cochlea, which was significantly different from that of other types.

Objectives

The aims of the present study were to establish normal measurements of the height and the width of the fully developed cochleae using HRCT scans of the temporal bone in normal individuals, and to predict the characteristic radiologic sings and measurements of miniature cochlea and its implication during cochlear implant surgery.

Patients and methods

This prospective, comparative study was conducted between March 2010 and April 2014 in Egypt. Approval for the study was obtained from the ethical committee. Parents of the participants signed a detailed informed consent.

This study was stratified as follows:

(1) The control group, in which 25 individuals (50 cochleae) were selected from persons with normal hearing, normal audiologic findings, without

Table 1 Radiologic classification of cochlear malformations

Michel deformity	Complete absence of all cochlear and vestibular structures.
Cochlear aplasia	Complete absence of the cochlea.
Common cavity deformity	A cystic cavity representing the cochlea and vestibule without showing any differentiation into cochlea and vestibule.
Cochlear hypoplasia	The cochlea and vestibule are separate from each other but their dimensions are smaller than normal.
Incomplete partition type I (IP-I)	The cochlea is lacking the entire modiolus and cribriform area, resulting in a cystic appearance.
Incomplete partition type II (IP-II)	Also known as Mondini malformation. The cochlea consists of 1.5 turns, in which the middle and apical turns coalese to form a cystic apex, accompanied by detailed vestibule and enlarged VA.
Incomplete partition type III (IP-III)	Bulbous fundus and basal turn of the cochlea, absence of the lamina cribrosa.

history of severe head trauma or ear surgery who were candidates for skull CT for other reasons.
(2) The patients group, for which patients were selected from candidates for cochlear implant surgery, without visible inner ear malformation in their CTs, apparently normal cochlear morphology, and negative history of meningitis or severe head trauma (only the implanted ears were included in statistics).

During the study and after radiological measurements of the CH, this group was divided into two subgroups according to the definitions proposed by Mori and Chang [6].

(1) Group A, which included 22 patients (22 cochleae), with CH not less than 2 SD from the controls.
(2) Group B, which included nine patients (nine cochleae), with CH less than 2 SD of the controls (miniature cochlea).

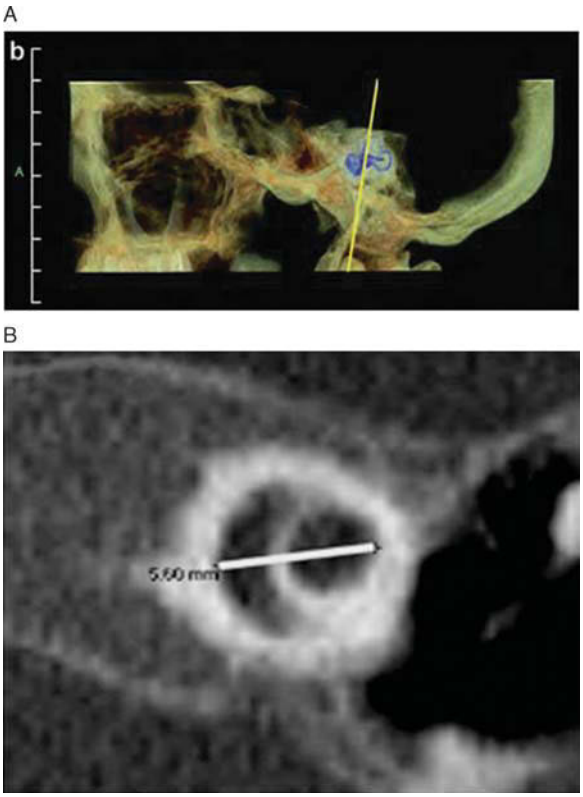
CT images were acquired using 64-detector HRCT of the temporal bone; parameters were fixed as follows: window width of 4000, window level 200, 120 kv, 250 mAs, and section thickness of 1 mm. The acquired images were uploaded to picture archiving and communications system for analyses, calibration, and measurements. Analyses were carried out by a radiologist, and double checked by other authors.

Radiological planes and measurements were taken according to the definitions provided by Purcell *et al.* [2], Mori and Chang [6], Giesemann *et al.* [7], Escude *et al.* [9], and Lane and Write [10].

CH was defined as the measurement from the midpoint of the basal turn to the midpoint of the apical turn, taken perpendicular to the axes of the cochlear lumens in its widest view in the coronal plane (Fig. 1); it was measured on the cut with the maximum height that included the basal and apical turns. In patients with multiple HRCTs for any reason, the interscan difference regarding the CH was estimated and analyzed, and error factor was considered.

The basal turn height (BTH) and upper turn height (UTH) (the rest of cochlear turns other than basal one) were measured as the maximum vertical distance within the cochlear lumen in each corresponding turn, in the mid-modular plane in axial HRCT (Fig. 2b).

Figure 1



(a) The axis of the coronal plane projection (yellow line) for the temporal bone (perpendicular to the plane of the lateral semicircular canal) [10]. (b) Cochlear height measured from the midpoint of the basal turn to the upper turn in coronal section high-resolution computed tomography [6].

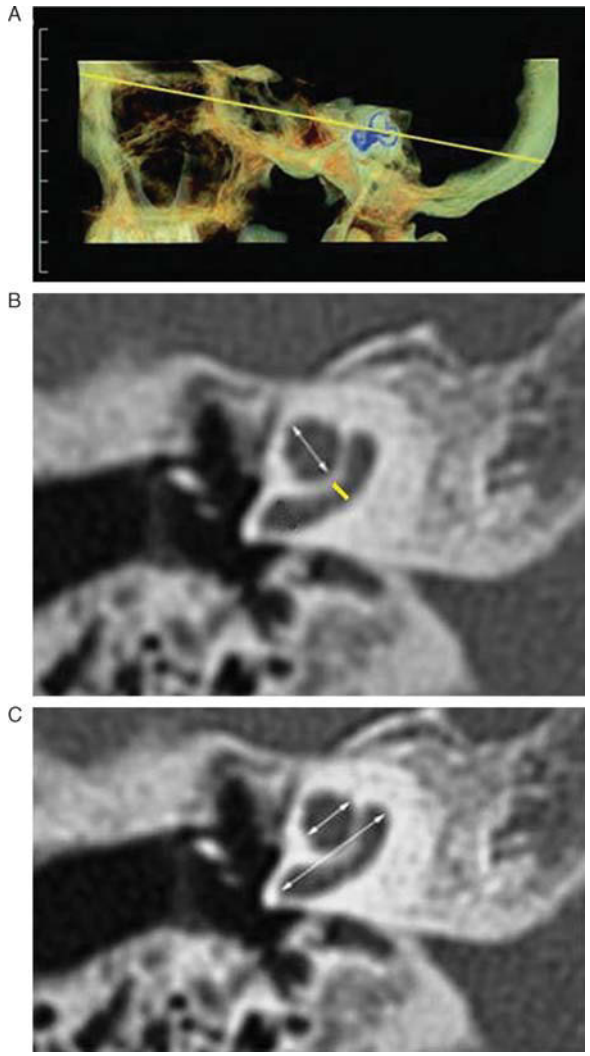
Basal turn width (BTW) was defined as the simple straight measurement extending from the center of the round window – through the cochlear lumen – to the opposite wall of the basal turn in its widest view in the axial plane (Fig. 2c); this reflects the cochlear width.

Upper turn width (UTW) was measured as the maximum horizontal distance between the cochlear walls of the upper turn in the same section (Fig. 2c).

The distance from the center of the oval window to the center of the round window (OW–RW) was measured radiologically in the oblique sagittal reconstruction through the turns of the cochlea, and also checked during cochlear implant surgeries using ear-micro instruments for the patients group (Fig. 3).

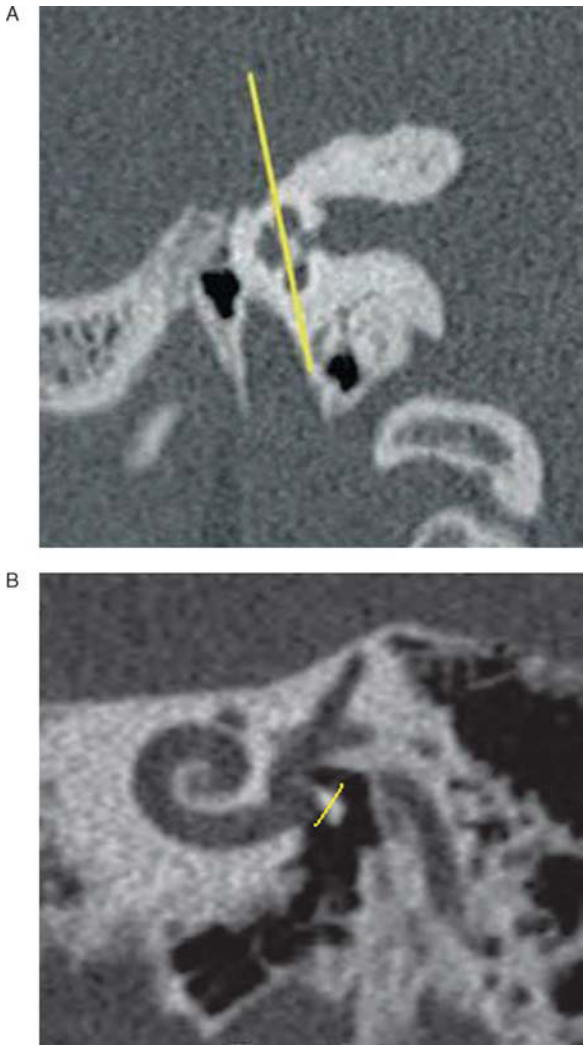
In the case of the patients group, special notes were taken during cochlear implant surgery, regarding OW–RW distance; the cochleostomy site, angle, and depth of electrode insertion; aberration from

Figure 2



(a) The axis of axial plane projection (yellow line) for the temporal bone (in the plane of the lateral semicircular canal) [10]. (b) Basal turn height and upper turn height measured in the mid-modular axis from the midpoint of the basal turn, and from the midpoint of the upper turn in each corresponding lumen, axial plane high-resolution computed tomography (HRCT). BTH (yellow line) & UTH (double headed arrow) measured in the mid-modilar axis from midpoint of the basal turn, and from the midpoint of the upper turn in each corresponding lumen, axial plane HRCT. (c) Basal turn width and upper turn width measured in the longest axis of the basal turn and upper turn lumens, axial plane HRCT [7]. BTW (lower long arrow) & UTW (upper short arrow) measured in the longest axis of the basal turn and upper turn lumens, axial plane HRCT.

Figure 3



(a) Oblique coronal reference images demonstrating the multiplanner reconstruction plane (yellow lines) required for the oblique sagittal views through the turns of the cochlea. The angles of reconstruction are slight modifications of the Stenvers plane [10]. (b) Basal turn where the round window–oval window distance could be measured [7].

normal anatomy; and expected complications. All this clinical data were to be correlated with received information from the radiological study.

Statistical analysis

The SPSS program (version 16; SPSS Inc., Chicago, Illinois, USA) was used for analysis. Quantitative variables were described as means ± SD. One-way ANOVA test was used for comparison between different groups using least significant difference to find subgroups' significance.

Results

This study involved 56 individuals, selected according to special criteria, with all patients between 2 and 6 years of age (mean: 3.8 years).

CH in the control group ranged between 4.8 and 6.9 mm, with a mean of 6.1 ± 0.29 mm. In group A, CH showed a mean of 5.9 ± 0.34 mm, which was statistically significant compared with the control group. In group B, CH showed a mean of 3.8 ± 0.31 mm, which was also statistically significant compared with the other two groups. The BTW, UTW, the OW–RW distance – all showed statistically significant difference when compared within the three groups. In

contrast, both BTH and UTH showed no significant difference when compared within the three groups (Table 2 and Figs. 4–6).

Interscan difference regarding CH for patients with multiple HRCTs ranged between 0.25 and 0.6 mm, with a mean value of 0.4 ± 1.2 mm, which was not statistically significant.

Cochleostomy was carried out above the round window (in a plane between the oval window and the round window) in all nine cochleae of group B (Fig. 7). Otherwise, the standard surgical cochleostomy (antroinferior to round window niche) was conducted in group A, without detectable odd presentations or complications (Fig. 8).

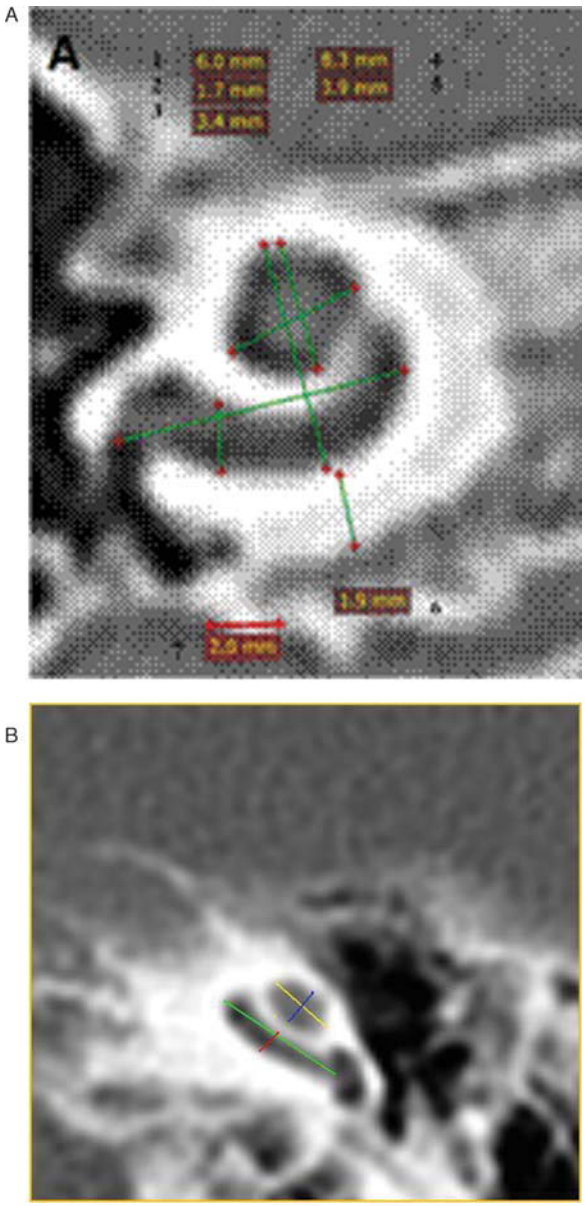
Discussion

Cochlear malformations are well-established causes of congenital sensori-neural hearing loss (SNHL) in children. Cochlear anomalies such as common cavity deformity, cochlear aplasia, cochlear hypoplasia, and incomplete partition (classic Mondini deformity) are thought to result from the premature arrest in cochlear development during various stages of gestation [11].

Even today, little information is available on these cochleae of significantly reduced size. Although there are frequent references to them as hypoplastic cochleae, many studies either classify them under Mondini malformation or do not mention this entity at all [12].

The present study introduced cochleae significantly less than the normal size, condition known as dwarf cochlea (miniature cochlea). These abnormal cochleae of reduced height generally have cochlear ducts and shortened heights [13].

Figure 4



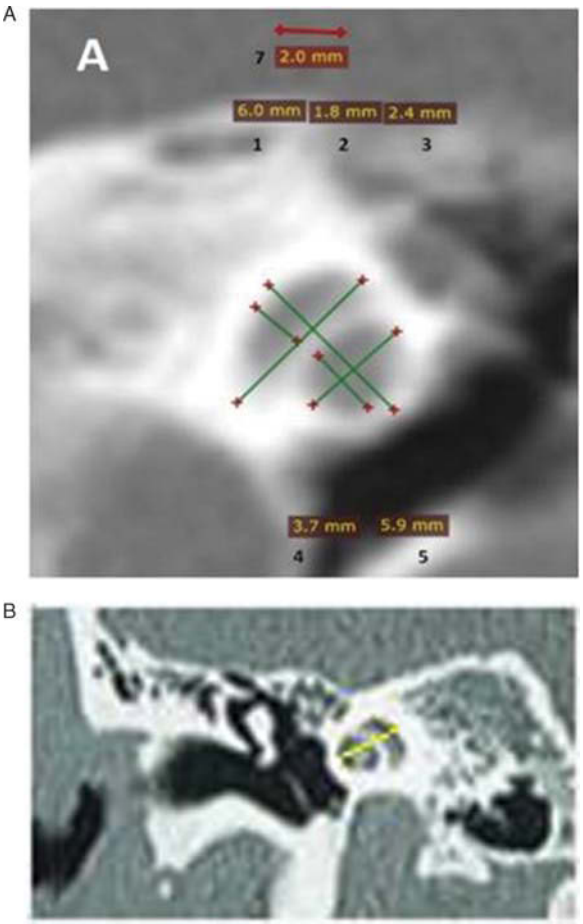
Axial computed tomography of the cochlea showing measurements in (a) normal cochlea, (b) miniature cochlea showing basal turn height (red), height of upper turns (blue), width of the basal turn (green), and width of the upper turns (yellow).

Table 2 Computed tomography measurements in all groups

Measures	Control group ($n=50$) (control)	SNHL group ($n=22$) (group A)	Dwarf group ($n=9$) (group B)	P-value
CH	6.1 ± 0.29	5.9 ± 0.34	3.8 ± 0.31	0.003
BTH	2.2 ± 0.21	2.1 ± 0.23	1.8 ± 0.11	0.34 (NS)
UTH	3.1 ± 0.23	2.8 ± 0.33	1.9 ± 0.12	0.12 (NS)
BTW	8.2 ± 0.38	7.8 ± 0.52	6.1 ± 0.41	0.03 (control vs. dwarf, $P=0.01$)
UTW	4.1 ± 0.27	3.7 ± 0.43	2.8 ± 0.49	0.002 (control vs. dwarf, $P=0.03$)
OW–RW	1.9 ± 0.35	1.78 ± 0.38	1.3 ± 0.12	0.02 (control vs. dwarf, $P=0.003$)

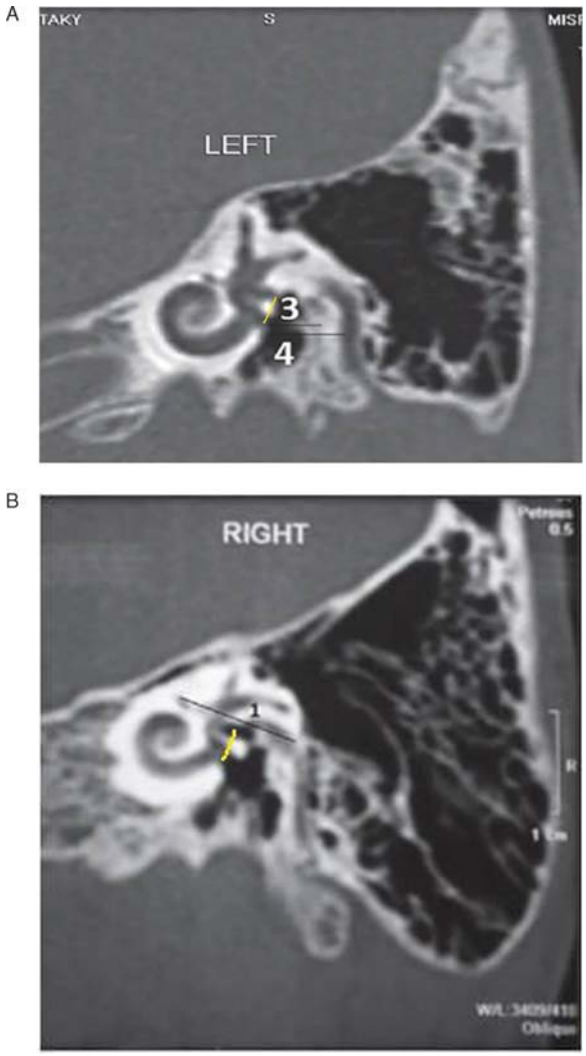
All measurements were in mm.
BTH, basal turn height; BTW, basal turn width; CH, cochlear height; OW–RW, oval window–round window distance; UTH, upper turn height; UTW, upper turn width.

Figure 5



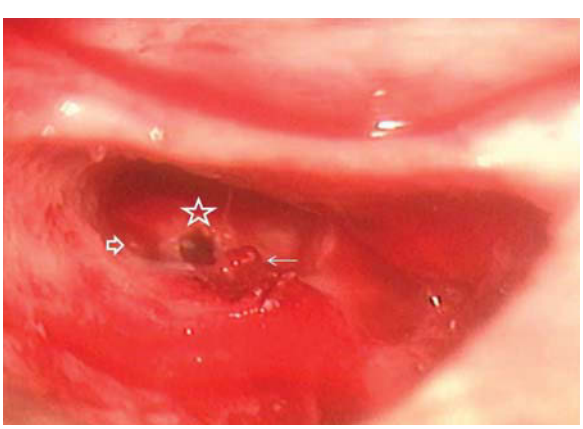
Coronal computed tomography of the cochlea showing measurements in (a) normal cochlea, (b) miniature cochlea, representing cochlear height (yellow).

Figure 6



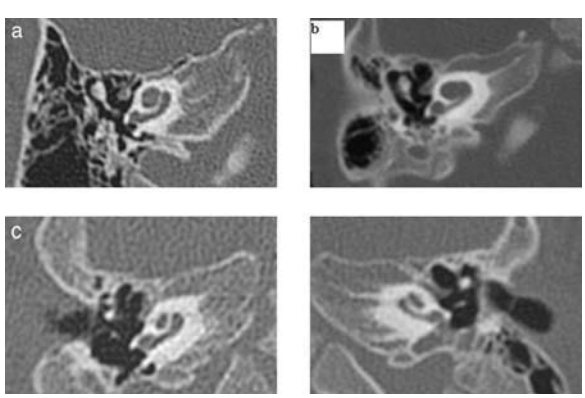
Oval window–round window distance distance (yellow) in oblique sagittal views (a) normal cochlea, (b) dwarf cochlea.

Figure 7



Intraoperative view of a left-sided miniature cochlea showing cochleostomy (fx1), round window niche (fx2), and stapes head (←).

Figure 8



Appearance of a hypoplastic cochlea in computed tomography normal cochlea for comparison (a) and different patients with cochlear hypoplasia (c, d). An incomplete partition (b) for differentiation [7].

In contrast to the hypoplastic cochlea with an evident radiological appearance, the miniature cochlea has an apparent normal radiological appearance and could be missed if proper measurements are not taken. Subtle abnormalities of the inner ear, such as cochlear hypoplasia and semicircular canal dysplasia (which account for the majority of inner ear malformations associated with sensorineural hearing loss), were often missed on the CT of the temporal bone due to radiologic inexperience of the clinician and absence of normative data to aid in diagnosis [2].

Recent studies have sought to increase the number of radiographically recognizable causes of congenital sensorineural hearing loss, because historically only 20–39% of children with congenital sensorineural hearing loss have an associated radiographic abnormality. Inner ear measurements not only aid in the duplication of radiologically diagnosed inner ear malformation but can also provide additional information about which specific part is abnormal [14,15].

This present study demonstrated the mean CH of 4.8–6.9 mm in the control group, which was near the values reported by Mori and Chang [6] (4.4–6.2 mm), but was slightly different from Filipino's CH (3.3–5.2 mm) [8]. Group A (sensorineural hearing loss) showed significant difference in CH compared with the control group, which was in agreement with the reports that patients with sensorineural and mixed hearing loss demonstrate significantly smaller CHs compared with normal-hearing patients [1]. In group B (patients with miniature cochlea), the CH was found both significantly different and less than 2 SD from the control group, which was in line with the results of other reports [6,7].

In addition, we reported a mean of 0.4 mm interscan variation regarding CH, which was less than that reported by Mori and Chang [6] (0.7 mm). This interscan difference was attributed to the differences in CT scan orientation or image quality. We reduced this phenomenon by double checking each value, standardizing the section images, reference points, and plane to be examined, and, lastly, by examining only high-quality images.

Our study demonstrated that both BTW and UTW (which together crudely reflects the cochlear duct length) were significantly reduced in patients with miniature cochlea compared with controls, which was logically anticipated, and confirmed the validity of the definition of a miniature cochlea [10].

In contrast to the results of Giesemann *et al*. [7], the current study showed no significant differences regarding BTH and UTH among the three groups. This is could be attributed to heterogeneity of this hypoplastic cochlea group, small sample size, and the wide range of deformities incorporated. On the other hand, this indifference was in line with the hypothesis that dwarf cochlea is a miniature one with short patent ducts.

An important finding was that the OW–RW distance was statistically significantly shorter in patients with miniature cochlea (those in group B) compared with controls. This finding was not reported before; thus, this finding and the postulated short BTW had a significant reflection on the site of cochleostomy, which was performed above the round window (in a plane between the oval window and the round window) in the last seven cochleae of the group B, as it was more anatomically, allowed for an easier array introduction, and helped to avoid array hooking with the round window. In addition, short electrode array was used in these last seven cases to match the reduced cochlear duct length and achieve full insertion after the first two cases, where we had inserted only 10 out of the classical 22 electrodes.

Finally, we could define miniature Egyptian dwarf cochlea as a morphologically normal cochlea with less than two and a half turns, patent ducts, and with significantly reduced CH, BTW, UTW, and OW–RW distance in HRCTs. More clinical studies are needed to correlate this radiological diagnosis to clinical audiological tests, and to other cochlear abnormalities, which remains a challenge that we hope will be addressed in near future.

Conclusion

In cochlear implant surgery, the detailed and good radiological analysis aids in a safe, effective, and well-planned surgery. The development of standardized measurements to complement visual inspection improved the diagnostic accuracy and helped in the subclassification of hypoplastic cochleae. We found CH to be the most essential differentiating measurement in the subclassification and diagnosis of hypoplastic and dwarf cochleae. If dwarf cochlea is radiologically diagnosed, special surgical modifications regarding electrode length and site of cochleostomy during cochlear implanting should be applied. The rapidly evolving micronavigation technology and its application on large-scale studies will be of great help in understanding and managing such a clinical entity.

Conflicts of interest

There are no conflicts of interest.

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