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Measurements of normal inner ear on computed tomography in children with congenital sensorineural hearing loss

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Abstract The objective of this study is to use standardized measurements of the inner ear to see whether there are subtle bony malformations in children with congenital sensorineural hearing loss (SNHL) whose temporal bone computed tomography (CT) are grossly normal. The study includes 45 ears with congenital SNHL and grossly normal temporal bone CT scans and 45 ears with normal inner ear structures and normal hearing. Standardized measurements of the inner ear structures were made on axial temporal bone CT scans. Student's *t* test was performed to compare the measurements of the two groups. There were significant differences in the measurements of the bony island width of the superior semicircular canal, bony island width of the lateral semicircular canal and maximal height of cochlea between two groups ($P < 0.05$). In conclusion, standardized measurements of bony labyrinth of inner ear on temporal bone CT can identify subtle abnormalities of inner ear in patients with congenital SNHL having grossly normal radiological images.

Keywords Inner ear · Congenital sensorineural hearing loss · Computed tomography · Bony labyrinth · Semicircular canal · Cochlea

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Introduction

The advent of high-resolution computed tomography (CT) scanning in the 1980s has revolutionized diagnostic imaging of the temporal bone. Temporal bone CT scan is useful for detecting malformations of the inner ear, such as labyrynthine anomalies, cochlear anomalies, enlargement of the vestibular aqueduct, and narrow internal auditory canal (IAC) [1, 2].

Inner ear malformations of the bony labyrinth are present in 20–25% of patients with congenital sensorineural hearing loss (SNHL) undergoing temporal bone CT [1, 3]. Identification of obvious morphogenetic malformations such as common cavity deformity, Mondini deformity, and Michel aplasia is not difficult. However, nearly one-third of less severe dysplasias may be missed by simple visual inspection of the radiological images [4, 5]. Recognition of these less severe dysplasias is dependent on the experience of the clinician.

The limitations of visual inspection and the reliance on clinical experience have been partially overcome by the development of radiographic measurements [6]. For

example, measurement of the vestibular aqueduct is crucial in establishing the diagnosis of large vestibular aqueduct (LVA) syndrome [7]. Since the inner ear does not change in size after birth, using PACS (picture archiving and communications systems) with precise digital measurements can help us with standardized measurements of bony labyrinth [6]. In this study, we use standardized measurements of the inner ear to see if there are subtle bony malformations in patients with congenital SNHL whose temporal bone CT are grossly normal.

Materials and methods

We retrospectively reviewed the high-resolution CT scans of 45 ears in 27 patients with congenital SNHL and normal radiological images reported by radiologists and 45 ears in 41 patients with normal inner ear structures and normal hearing with Institutional Review Board approval. The medical records and the pure-tone audiometric results were also reviewed. Patients with cochlear anomalies, labyrinthine anomalies, aqueductal anomalies, and IAC anomalies were excluded from the SNHL group. Children with prenatal or perinatal infection, such as CMV, HSV, measles, and so on, were also excluded in the study. The control subjects underwent CT for the evaluation of cholesteatoma, or mastoiditis or middle ear ossicular chain problem or external ear disease. The CT scans of the SNHL group were performed between July 2005 and July 2007 and those of the control group were performed between January 2003 and August 2008.

The studies consisted of contiguous 2-mm scans of the temporal bone in the axial planes. Measurements were taken on PACS terminals with electronic calipers under magnification. Ten measurements in the axial plane were obtained, including canal lumen width and bony island width of superior semicircular canal (SSCC) and lateral semicircular canal (LSCC), canal lumen width and inferior

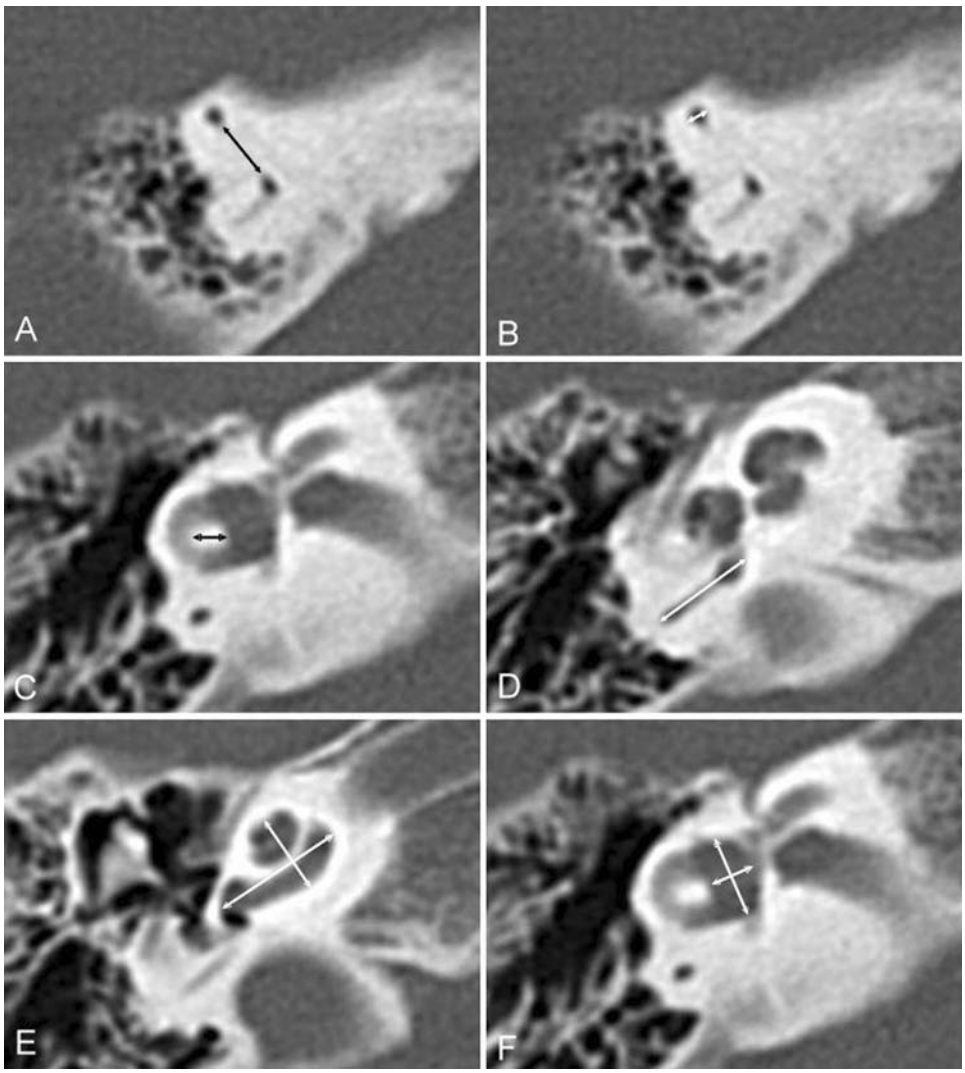

Fig. 1 Inner ear measurements made on axial temporal bone CT scan. **a** SSCC bony island width, **b** SCC canal lumen width, **c** LSCC bony island width, **d** PSCC inferior limb length, **e** Maximal height and length of cochlea, **f** Maximal width and length of vestibule

limb length of posterior semicircular canal (PSCC), maximal height and length of cochlea, and maximal width and length of vestibule (Fig. 1).

Statistical comparisons of two-group data by Student's *t* test were analyzed using SPSS 12.0 software. $P < 0.05$ was considered statistically significant.

Results

Twenty-seven patients (45 ears) with SNHL were included in this study. Forty-one patients (45 ears) with at least one normal hearing ear underwent CT scanning for various otologic diseases: cholesteatoma, mastoiditis, external canal polyp, and ossicular chain problem. All inner ear structures were assessed as normal by radiologists.

The average age of SNHL group (13 males, 14 females) was 5.37 years (range 1–14 years). The average age of control group (27 males, 14 females) was 8.63 years (range 1–16 years). Pure-tone audiometric testing revealed that 75.5% of the SNHL group had moderately severe to profound hearing loss (hearing threshold greater than 55 dB). The means of hearing thresholds of three-frequency (500, 1,000, 2,000 Hz) pure-tone average in the SNHL group is 75.1 dB.

We used the 45 ears with normal hearing and normal inner ear structures as the control group to generate normative data. There were no statistical differences in inner ear measurements between male and female ($P > 0.05$, data not shown), also no statistical correlation between inner ear measurements and the age ($P > 0.05$, data not shown) in the control group. Both the mean value and the standard deviation for each measurement of the inner ear were shown on Table 1. Comparison between the measurements of the SNHL group and the control group were done using Student's *t* test. *P* values were chosen according to the Levene's test for equality of variances.

We found significant differences in the measurements of the bony island width of SSCC, bony island width of LSCC and maximal height of cochlea between patients with and without congenital SNHL ($P < 0.05$). There were no significant differences between two groups in the measurements of the canal lumen width of SSCC, LSCC, and PSCC, inferior limb length of PSCC, maximal length of cochlea, and maximal width and length of vestibule (Table 1).

The bony island width of SSCC in the SNHL group was smaller than in the control group (4.79 vs. 5.06 mm), so as the bony island width of LSCC (3.41 vs. 3.64 mm). On the contrary, the maximal height of cochlea in the SNHL group was larger than in the control group (4.79 vs. 4.46 mm). In addition, we also found that there were more cases with central lucency of bony island of LSCC in the congenital SNHL group than in the control group (23 ears vs. 4 ears) and statistical comparisons of two-group data using χ^2 test showed significant difference ($P < 0.05$) (data not shown) (Fig. 2).

Table 1 Measurements of inner ear in congenital SNHL group and normal group

	SNHL		Control		*t* test
	Mean (mm)	SD (mm)	Mean (mm)	SD (mm)	*P* value
SSCC					
Canal lumen	1.29	0.11	1.31	0.10	0.56
Bony width	4.79	0.43	5.06	0.31	0.001*
LSCC					
Canal lumen	1.38	0.21	1.38	0.11	0.92
Bony width	3.41	0.46	3.64	0.38	0.01*
PSCC					
Canal lumen	1.31	0.14	1.36	0.11	0.08
Inf limb length	6.92	0.56	6.89	0.61	0.83
Cochlea					
Height	4.79	0.45	4.46	0.26	<0.001*
Length	8.71	0.29	8.67	0.32	0.5
Vestibule					
Width	3.16	0.30	3.17	0.22	0.85
Length	5.86	0.35	5.95	0.22	0.15

SSCC superior semicircular canal, *LSCC* lateral semicircular canal, *PSCC* posterior semicircular canal

*Unequal variance

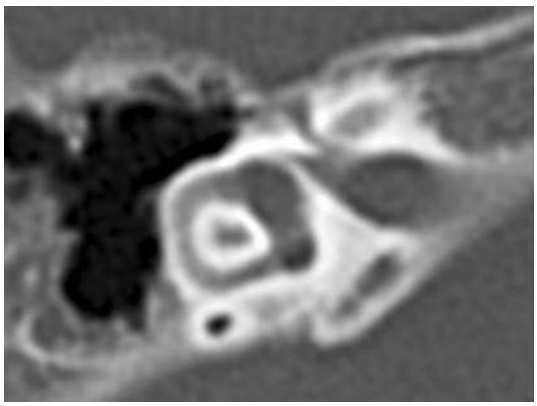

Fig. 2 Central lucency of bony island of LSCC

Discussion

The childhood SNHL is diagnosed by otologic, audiologic, and physical examination, family history and image testings. Among the image examinations, temporal bone CT scanning provides structural analysis of the external and

middle ear, as well as bony labyrinth of the inner ear. However, only about 25% of patients with congenital SNHL will have obvious bony inner ear malformation on temporal bone CT scans [1, 3]. Subtle abnormalities of the inner ear such as cochlear hypoplasia and semicircular canal dysplasia may be missed due to the inexperience of the clinician and absence of normative date to aid in diagnosis [4, 5].

Except the IAC, there is little variability in the dimensions of the human inner ear [4]. In this study, we use standardized measurements to evaluate 10 different dimensions of inner ear structures. Our data show that there are mild but measurable differences in cochlear and vestibular dimensions in patients with congenital SNHL and visually normal inner ear structure on temporal bone CT scans. Significant differences in the measurements of the bony island width of SSCC, the bony island width of LSCC and maximal height of cochlea between patients with and without congenital SNHL were found ($P < 0.05$).

We also found that there are more cases with central lucency of bony island of LSCC in congenital SNHL group than in control group (23 ears vs. 4 ears) with statistically significant difference. The most common inner ear malformations involve LVA, LSCC, and cochlea [6, 8]. Normal development of semicircular canal (SCC) involves formation of the bony island in the center of the canals [5]. As in our study, incomplete central bony island formation of LSCC with radiologically central bony lucency is more commonly found in patients with SNHL.

In Purcell et al. study, they found there are smaller basal turn width of cochlea and larger bony width of SSCC and LSCC with statistical significance on temporal bone CT in patients with congenital SNHL and radiologically undetectable abnormalities [5]. Moreover, they used 21 different measurements in their study [5]. In our study, we use only 10 measurements and found that they are sufficient to identify subtle bony malformation of the inner ear. However, our data showed larger vertical height of cochlea and smaller bony width of SSCC and LSCC in patients with congenital SNHL. Statistical differences are found in the measurements of vertical height of cochlea and bony width of SSCC and LSCC. Racial difference may be the possible reason to explain the different measurement results. Enrollment of more cases and further molecular genetic study may help to elucidate this question.

In Johnson et al. study, approximately 50% of patients with bilateral, symmetric inner ear malformation had asymmetric hearing loss [4]. Lack of correlation between radiological imaging and audiological findings was found [4]. Although an essential interrelation does exist between the otic epithelium (membranous labyrinth) and periotic mesenchyme (bony labyrinth), inner ear malformations can present with variable levels of abnormalities in the bony and membranous labyrinth [9]. Classic temporal bone dissections by Johnsson show that inner ear malformations may be isolated to either the bony or membranous labyrinth [10]. Although temporal bone CT scan is a convenient and cheaper tool for evaluating bony labyrinth, advances in magnetic resonance imaging technology offer further resolution as well as molecular genetic analysis giving more information in cases with congenital SNHL.

Conclusion

Standardized measurements of bony labyrinth of inner ear on temporal bone CT can help us to identify subtle abnormalities of inner ear in patients with congenital SNHL who have normal radiological inner ear structure by visual inspection.

Conflict of interest statement The authors declare that they have no conflict of interest.

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