Evaluating Common Cavity Cochlear Deformities Using CT Images and 3D Reconstruction

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Objectives: The aim of this study is to compare the common cavity (CC) with the normal anatomy inner ear in order to evaluate whether the cavity is representing both the cochlear and the vestibular parts of the inner ear and to revisit CC deformity from a three-dimensional (3D) perspective.

Methods: High-resolution computed tomography image datasets of 17 temporal bones initially identified as CC were evaluated with 3D reconstruction and multiplanar image analysis using a free available software for 3D segmentation of the inner ear. All 3D images of CC were compared to a normal inner ear. Maximum and minimum diameter of the CC were correlated with the circumference of the CC in an axial plane.

Results: In 13 cases (76%), CC represented only the vestibular part of the inner ear and did not represent CC as defined here and by Sennaroglu, Kono, and Khan. True CC was correctly diagnosed in only one case (6%). In three cases (18%), a rudimentary part of the cochlear portion could be identified. The axes’ length of the elliptical cavity showed a strong positive linear relation to the circumference of the cavity (long axis: $r = 0.94$; short axis: $r = 0.68$; $P = 0.0029$).

Conclusion: This study supports the assumption that many reported CC cases only represent the vestibular part of the inner ear and are therefore cases of cochlear aplasia. 3D segmentation and systematic analysis of CT-imaging add clinical value to the comprehension of the morphology of the anatomical structures of the inner ear.

Key Words: Common cavity, cavity size, circumference, cochlear aplasia.

Level of Evidence: 2C

INTRODUCTION

Human inner ear malformation is a special topic that needs to be well understood before contemplating cochlear implantation (CI) for remediation of sensorineural hearing loss (SNHL). The incidence rate of radiographically demonstrable inner ear malformation cases is reported in 20% to 30% of subjects with SNHL.1–3

Three types of single cavity inner ear malformations have been identified:

1. Primitive otocyst (Michel’s otocyst deformity): A single cyst is identified in the temporal bone. It is unsegmented, round, and fluid-filled. The internal auditory canal (IAC) and cochlear nerve are absent.

2. Cochlear aplasia (CA): The vestibular portion of the inner ear is present as a variably shaped cystic space posterior to the IAC, which enters its anterior end. A radiographically identifiable cochlear nerve is absent. All or portions of the semicircular canals may be present (or not) and are often dysmorphic (usually widened).

3. Common cavity (CC) deformity: a large, ovoid single cavity, either with or without rudimentary semicircular canals, is seen extending both anterior and posterior to the IAC, which enters the midportion of the cavity. A wide opening between the vestibular portion (posterior to the IAC) and the cochlear portion (anterior to the IAC) fuse the two cavities. A cochlear nerve is typically identifiable. Histologically, portions of the cochlear duct and spiral ganglion cells have been identified.4

Although anatomical malformation may occur in any specific portion of the inner ear, the term CC is used when both the cochlear and the vestibular portion fuse to form an empty cavity either with or without rudimentary semicircular canals. However, the definition of CC is not uniformly used in the literature.2,5 and different variations of this type of inner ear malformation (IEM) are mentioned.4,6

Primitive otocyst, once understood, is easily identified and will not be discussed further.

The CC malformation was first described by Edward Cock in 1838 as a disruption in the differentiation of
inner ear structures during the fourth and fifth week of gestation. It is characterized by an empty cochlea that exhibits a wide basal turn. In recent classifications, the IAC has to enter the center of the CC to differentiate this IEM from dilated vestibular cavity with simultaneous CA. Clinical tomography imaging alone may be hard to interpret concerning these criteria. Computed tomography (CT) and magnetic resonance imaging (MRI) do not capture every case of malformation and an underestimation is assumed, especially when the resolution of tomography is poor.

Different radiological phenotypes are being discussed. The size of the CC varies in previous studies and the limit to other malformations such as incomplete partition type 1 (IP-1) is not clearly defined. Although CI is considered a safe and effective treatment option for children with CC, these patients suffer from poorer and more slowly developing hearing performance compared to children with SNHL and regularly formed cochlea or other IEM. Although reports on the audiological performance of CC patients implanted with CI vary to a considerable extent, it is unclear how many of these subjects had a true CC deformity with a primitive cochlea and how many had CA with only a large, dilated vestibule. Because CA cases typically exhibit abnormal cochleovestibular nerves on imaging, the results are predictably poorer than in CC, where there is a normal-appearing cochleovestibular nerve on imaging, and a portion of the cochlear duct and spiral ganglion cells have been identified histologically. The issue of mistaking CA with an enlarged vestibule with a true CC deformity previously has been noted. The misidentification of many of the cases in the series we report in this study may partially account for the variable outcomes reported for CC deformities.

Thus, distinguishing the two malformations has significant implications for hearing prognosis and preoperative counseling.

We assume that 3D reconstruction facilitates the evaluation of the exact anatomical relationship between the CC and the IAC.

If a cochlear portion is present, it should extend anteriorly to an extended line parallel to the posterior limit of the IAC. If the cavity lies entirely posterior to this same line, it likely represents only a dilated vestibule.

METHODS

CT Datasets
In this case control study, preoperative high-resolution spiral CT image data sets with a slice thickness between 0.635 and 1 mm of human temporal bones initially classified as CC were analyzed. The images originate from several international centers and were sent to the authors, requesting a consultation concerning the correct electrode choice.

A CT dataset of a healthy patient with no prior ear surgery who received a temporal bone CT to exclude a temporal bone fracture was used as anatomical reference. All CT datasets were anonymized prior image analyses.

The study was approved by the local ethics committee [A2019-0201]. Due to the use of anonymized data, written informed consent was waived.

Image Analyses
The CT datasets were reconstructed using 3D slicer 4.11.0 (https://www.slicer.org), an open source software for the analysis and visualization of medical tomography imaging. Segmentation of the inner ear was performed using threshold analysis, and a 3D model of the inner ear was reconstructed. Furthermore, multiplanar slices were reconstructed in the axial plane (0.625–1 mm), as described elsewhere. The 3D model was orientated in relationship to the longitudinal axis of the IAC. Relationship of the CC to the longitudinal axis of the IAC and the medial border of the vestibular part of the inner ear was evaluated using a line extended along the posterior limit of the IAC through the lateral temporal bone. The cochlear portion of the CC was considered present if the CC was located anterior and was rated as absent if the CC was posterior to the IAC, respectively.

In order to measure the maximal cavity size, the slice presenting the longest length of both the long and the short axis of the cavity was selected (Fig. 1). The values were generated in the individual image slice that gave the greatest distance for either the short or the long axis.

The circumference of the elliptical cavity was calculated using the equation \(2\pi \sqrt{(a^2 + b^2)/(2)}\).

Statistical Analysis
All statistical tests were selected before data collection. Statistical analyses were performed using Prism version 8 (GraphPad Software, La Jolla, CA). The significance level was set to \(P < .05\).

The assumption of normality was tested using the D’Agostino-Pearson, Shapiro–Wilk, and Kolmogorov–Smirnov tests. Continuous variables are presented as absolute numbers and percentages or means and standard deviation, if not indicated otherwise. Correlations were calculated using Spearman rank test.

Fig. 1. Procedure of measurements. The long and the short axis lengths of the cavity are measured in the axial view. The individual values are generated in the image slice, which gives the maximum distance for either the short or the long axis.
RESULTS

All CT datasets could be segmented successfully, and a 3D model of the inner ear was reconstructed (Fig. 2).

Although all CT datasets were classified as CC, after reviewing the 3D reconstructions this diagnosis could
only be confirmed in one case (6%; case 10) (Fig. 3), with the cavity extending anteriorly to the IAC. In three cases, a rudimentary part of the cochlear portion could be identified (18%; case 7, 11, and 12) (Fig. 2). In 13 cases (76%), however, 3D reconstruction demonstrated the absence of the cochlear portion and the cavity only representing the dilated vestibular portion of the ear.

The circumference of the CC demonstrated a strong linear relation with the length of the long axis \( (r = 0.94; P < .0001; R^2 = 0.88) \) and the short axis \( (r = 0.68; P = .0029; R^2 = 0.46) \), respectively (Fig. 4). No linear correlation \( (r = 0.38; P = .13; R^2 = 0.14) \) was found between the length of the long and the short axis of the cavity.

**DISCUSSION**

**Main Findings**

The success rate in restoring the hearing to patients with IEM is rather low compared to patients with other causes of SHNL, and the most successful groups of patients with IEM are IP-II and enlarged vestibular aqueduct. Therefore, exact classification of IEM is mandatory prior surgery.

However, using standard CT and MRI imaging planes, this could be difficult, and false classification of the IEM is assumed, especially with low spatial resolution scans. Although MRI is the gold standard for the identification of the cochlear nerve (CN), it is discussed to be prone to errors. The absence of a clearly visible CN is not appropriately distinguishing between CC and CA because CA may present with a hypoplastic nerve, and CC usually is associated with a common cochleovestibular nerve (CVN). Batuk et al. report a case series of 12 children who received auditory brainstem implant after poor hearing outcomes with CI alone. They found a common CVN in cases of CC and report one case of CA with a hypoplastic CN. Monsanto et al. found a hypoplastic CN in 66% of their cases with CA. Furthermore, Adunka et al. emphasize that, even in cases with a radiologically present CN, cochlear stimulation may reveal functional CN deficiency, and Birman et al. report children with functional hearing after cochlear implantation even when preoperative MRI did not reveal a nerve inside the IAC. Consequently, the preoperative distinction between CA and CC and a prediction of the hearing outcome by the nerve status alone is not reliable. Thus, it was one aim of this study to introduce an additional opportunity to further distinguish between CC and CA because the nerve status inside the IAC may not adequately predict the functional outcome of CI.

3D segmentation facilitates the orientation and projection of the inner ear position and helps adjust it to the line extended along the IAC, as described in the Methods section. Applying the method using CT alone may be challenging because the exact cochlear position only results from analyzing all different views and planes. In this regard, 3D segmentation adds value to the diagnostic tools, as seen in Figure 5. This study demonstrated that 3D reconstruction of standard CT examinations alters the diagnosis of CC in 16 of 17 cases. In 13 cases, 3D reconstruction demonstrated severe CA. This is in accordance with previous results by Giesemann et al.

The misinterpretation of severe CA as CC based on standard imaging planes may be one reason for highly variable audiological performance of patients with CC.

This issue is supported by findings from Yamazaki et al, in which the electrically evoked auditory brainstem response as tested via the CI electrode suggested that the auditory neural elements are mainly distributed to the anteroinferior part of the single cavity, that is, the cochlear portion, whereas in cases of CA only vestibular parts are histologically present. Structured reports on hearing outcomes after cochlear implantation in CC cases are rare, and varying results are reported. Compared to other malformations, CC cases tend to have poorer hearing outcomes.

Presuming the IAC enters the center of the cavity in genuine cases of CC, our data support these studies with the assumption that a majority of CC cases reported in this study were misdiagnosed as CC. The large portion that has been interpreted as CC was found to actually represent the vestibular portion only and not the cochlear portion.

This is supported by the results of previous studies. For this reason, Giesemann et al. proposed a subclassification system of CA to improve outcome prediction based on preoperative imaging.

Although it can be challenging to compile series of 2D image slices to bring a 3D representation of the anatomical structures, 3D segmentation of the inner ear facilitates the visualization of the CC toward the IAC and can add value to the correct diagnosis and treatment. The effort is manageable low with an average time of 10 additional minutes during the preoperative image evaluation.

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Fig. 5. Illustrating the corresponding axial computed tomography slice to the 3D reconstructed cochlea in one case with cochlear aplasia (sample 4). [Color figure can be viewed in the online issue, which is available at www.laryngoscope.com.]
As a sidenote, the assumed CC was further analyzed concerning its size. In cases that present inconclusive anatomy or hypoplastic CN, the decision between ABI and CI is reported to be challenging, and as a first-approach CI is still recommended. In patients with CC, the neural elements have been described as present along the medial wall of the CC. As a consequence, approaches have focused on placing the electrode along the medial wall. With the established formula, calculation of the length of the electrode is based on the short and the long axis diameter. We could demonstrate that the latter has a stronger correlation.

This important finding of this study can be applied clinically and supports former studies in which the use of straight electrodes was recommended. The variable sizes of the cavities reported in this study convey that no malformation is similar in size or shape compared to the other. Although choosing the CI electrode type is highly subjective and mainly depends on the operating surgeon, a suitable choice of electrode needs to be discussed and may be adapted to the circumference of the CC. Especially in CC cases, when further identification of the development in the basal turn of the cochlear or the semicircular canals of the vestibular portion is of interest, 3D segmented imaging of the inner ear is one step toward better comprehension of the morphology of the inner ear.

**Limitations**

The present study has some limitations. The study sample size is small because the prevalence of CC itself is low. For this reason, statistical analyses are limited due to the small number of subjects. The anonymously obtained data are assumed to be provided due to the subject’s poor audiological outcomes after surgery. We assume that this explains the high rate of absent cochlear portion in this group. In cases in which the diagnosis is made correctly and nerve fibers can be expected inside the cavity, the chance of having a postoperative hearing impression with a CI is more likely. Another limitation is the variability in the slice thickness of the individual datasets, and that the corresponding MRI is missing. However, because this study was primarily designed as an exploratory approach, we showed that 3D segmentation and cavity measurements were achievable in each case and complement standard CT scans. Although applying the method presented in this study may not prevent every case of unsatisfying audiological results and does not replace careful pre- and postoperative audiometry and electrophysiological testing, we believe that improved understanding of the anatomy of malformations helps increase the quality of patient treatment, and 3D reconstruction of the inner ear increases diagnostic accuracy for the differentiation between CC and CA prior CI surgery.

**Conclusion**

This study reports a high rate of misdiagnosed cases of CC. In case of IEM, 3D reconstruction of the inner ear increases diagnostic accuracy for the differentiation between CC and CA prior CI surgery and complements routine CT and MRI investigations.

In order to systematically investigate the outcome of inconclusive cases, we encourage future studies that retrospectively apply the method of this study and correlate it to the hearing performance of CC cases.

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