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Middle ear volume as an adjunct measure in congenital aural atresia☆

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ABSTRACT

Objective: To examine middle ear volume in patients with aural atresia and investigate the role of middle ear volume as an adjunct measure in determining surgical candidacy.
Methods: We performed a retrospective review of children with aural atresia in a tertiary academic pediatric otolaryngology practice. High resolution multiplanar CT scans of the temporal bones were analyzed for middle ear volume and staged according to existing clinical grading scales. Atretic ears were compared to the nonatretic ears of the same patient as well as to ears of a control population.
Results: The average age of patients at the time CT was performed was 4.7 years (range <0.1–13.8 years). The average middle ear volume of the atretic ears was 0.34 cc compared to an average of 0.51 cc for the nonatretic ears. The mean ratio of the atretic to nonatretic volume was 0.67. In patients who underwent serial scans, no statistically significant difference in rates of growth existed between atretic and nonatretic ears. Finally, measures of middle ear volume correlated well with clinical grading scales.
Conclusions: Both middle ear volume and the ratio of the atretic volume to nonatretic volume serve as useful adjunct measurements in determining surgical candidacy. The practitioner may be better able to assess surgical candidacy by supplementing classic atresia classification systems with middle ear volume measurements.

1. Introduction

Congenital aural atresia (CAA) describes the condition of an embryologically malformed external auditory canal. The widely accepted incidence of aural atresia is one in 10,000 to 20,000 live births [1]. The incidence of unilateral atresia is roughly three times greater than bilateral atresia, and there is a slight male predominance. CAA is often associated with auricular and middle ear deformities, and there is a correlation between the degree of external ear deformity and middle ear deformity [2]. The etiology is multifactorial with genetic components, exogenous environmental factors, and sporadic developmental insults all thought to contribute to CAA. Terminal deletions of chromosome 18q are frequently associated with development of CAA [3,4].

A significant conductive hearing loss resulting in an air-bone gap of 40 to 50 dB typically accompanies CAA [5]. The goal of surgical intervention for CAA is to correct this conductive hearing loss and bestow serviceable hearing upon the affected ear. Additional motivation in most instances is the desire to avoid using a hearing aid. Not all patients who possess aural atresia are suitable candidates for atresia repair, as more severe anomalies defy operative intervention.

High resolution CT (HRCT) scanning is an essential tool in the preoperative evaluation of patients to determine their candidacy for surgical intervention. The Jahrsdoerfer scale (JS) provides a numerical grade of the severity of CAA based on anatomic features assessed clinically and via HRCT [6]. Recently, a score of seven or higher on the JS has been promoted as a threshold to establish suitable candidacy for surgical repair of CAA [7].

Criteria for assigning points on the JS were stated by Yeakley and Jahrsdoerfer, though many of the criteria are quite vague [8]. The subjective nature of assigning points for stapes development, course of the facial nerve, and mastoid aeration are obvious. Precise measurements are listed for middle ear space and round window diameter, though variability with plane of imaging is not discussed. This is significant since others have identified middle ear volume as an important factor in determining the outcome of aural atresia surgery [7,9]. Previous studies have used representative cross-sectional area measurements or volume estimates to demonstrate that middle ear volumes in atretic ears are smaller than developmentally normal ears [10,11]. The current volumetric study of the middle ear space in atretic ears proposes to identify an objective threshold value for middle ear volume when assigning a point in the JS and in relation to surgical candidacy.

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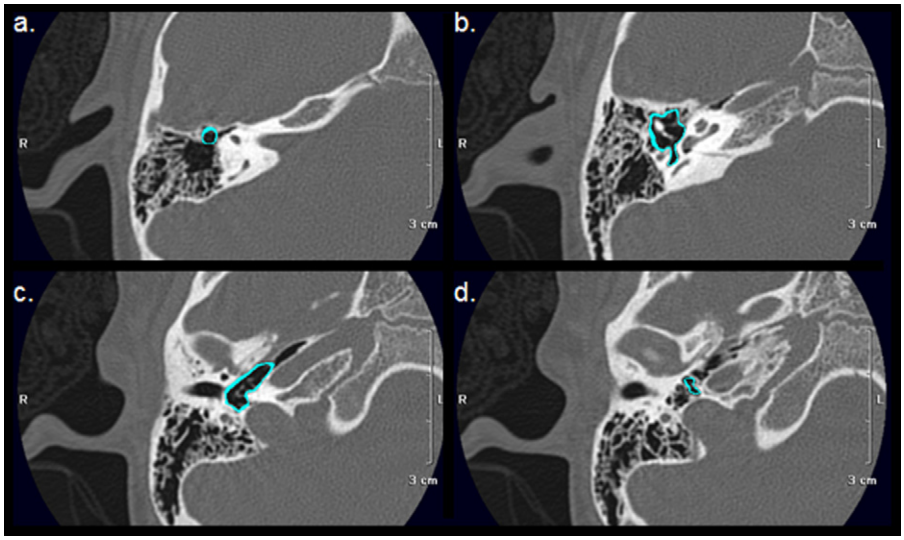

Fig. 1. Representative slices from HRCT scans of the temporal bones in a patient with aural atresia. The area outlined in blue has been drawn by the authors and is the space used by the software to calculate the volume of the middle ear. Sections at (a) the epitympanic space above the malleoincudal joint, (b) the promontory, (c) the Eustachian tube, and (d) the hypotympanum are shown. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of the article.)

2. Materials and methods

This retrospective review was approved by the Baylor College of Medicine Institutional Review Board. Methodology for assigning points in the Jahrsdoerfer scale follows prior published criteria with the following variations [8]. For the oval and round windows, a point is assigned unless the window is absent, significantly narrowed or obstructed by adjacent anatomical structures. The stapes must be seen well (crurae visible) for 2 points, and present in some form for 1 point. The facial nerve is given 1 point unless preoperative dysfunction is evident or it lies over the oval window. Mastoid aeration extending well beyond the labyrinth (complete aeration of epitympanum and antrum with at least some extension into the mastoid) is given one point. Surgical candidacy in this cohort required a minimum JS score of 6 (excluding the point for middle ear aeration as described below) and required a normal oval window, round window and facial nerve.

Middle ear aeration was not included in initial scoring calculations to allow comparison of two different methods for assessing middle ear aeration: linear measurement of middle ear depth, and volumetric measurement. In the linear method a point for middle ear aeration is assigned by measuring the distance from the promontory to the atresia plate with 3 mm being the minimal acceptable value [8]. In this study, the distance is analyzed in the coronal plane with the intent of recording the maximal distance.

Volumetric measurements were calculated using Vitrea 2 (version 3.7) imaging software (Vital Imaging, Plymouth, MN). Bone windows of dedicated temporal bone HRCT images (slice thickness was 0.63 mm) were analyzed in all cases. The boundaries of the middle ear space were outlined on axial cuts and were set laterally at the tympanic membrane (nonatretic ears) or the atresia plate (atretic ears), medially at the bony labyrinth, superiorly at the epitympanic space surrounding the malleoincudal joint, inferiorly at the last cut that distinct aeration was visible below the basal turn of the cochlea, anteriorly at the Eustachian tube opening and carotid artery, and posteriorly at the anterior border of the antrum (Fig. 1). The imaging software constructs a three-dimensional space from the outlined areas of interest on the axial cuts and calculates the volume of the space in cubic centimeters, up to the hundredths decimal place. Since bony windows were used, the volume estimate was not altered by fluid in the middle ear. A subset of films was analyzed in both the axial and coronal planes. Once it was established that values obtained in both methods were consistently within 5% of each other, we only performed axial plane measurements. Standard parametric statistical methods were used to calculate the average values and standard deviations listed in the results. The sum of least squares method was used for linear regression data displayed in Fig. 2.

3. Results

The study population consisted of 42 patients with unilateral atresia and no prior surgery and 3 patients with bilateral atresia, none of whom had surgery. Serial scans were available for 11 of these individuals. Of those with unilateral atresia, 53% were on the right side and 58% of the patients were male. Two of the three patients with bilateral atresia were male. Average age at time of imaging was 4.7 years (range <0.1–13.8 years). A control group consisted of 29 individuals having a CT scan as part of their evaluation for cochlear implantation. Six of these had repeated scans for a total of 35 scans. Average age of the controls was 4.4 years (<0.5–14.8 years).

In patients with unilateral atresia, average volume of the atretic ear was 0.34 cc (standard deviation 0.14 cc, range 0.01–0.68 cc), while the volume of the nonatretic ear averaged 0.51 cc (standard

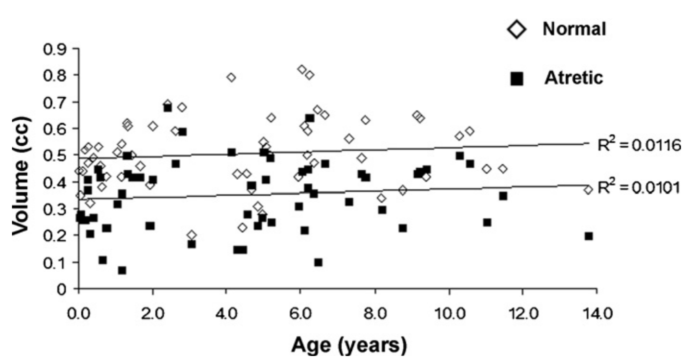

Fig. 2. Normal and atretic middle ears grow at the same rate during child development. Normal (open diamonds) and atretic (squares) ear volumes are plotted with respect to age. Regression lines were drawn demonstrating both normal and atretic ears grow minimally over time and essentially at the same rate as each other. The regression line for normal ears is above that for atretic ears.

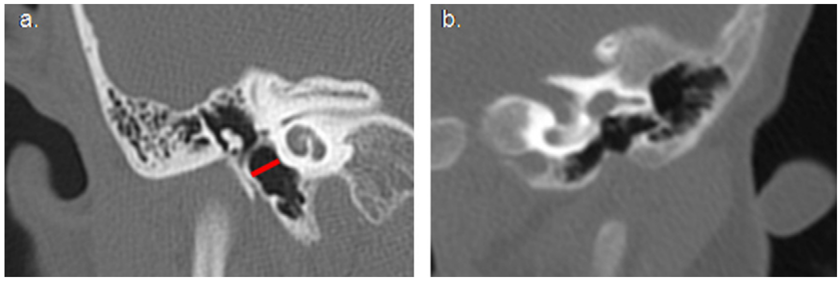

Fig. 3. Linear measurements of middle ear depth from coronal HRCT scans are variable. (a) The representative distance from the promontory to the atretic plate used to estimate the middle ear volume by Yeakley and Jahrsdoerfer [8] is drawn in red. (b) Variability in the contour of the lateral bone increases difficulty in measuring middle ear depth. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of the article.)

deviation 0.14 cc, range 0.23–0.82 cc) ($p < 0.001$). The volume of the atretic ear averaged 67% (standard deviation 21%) of the nonatretic ear with a wide range of 3–98%. The average volume in control subjects was no different from that of the nonatretic ears at 0.48 cc (standard deviation 0.09 cc, range 0.29–0.71 cc) ($p > 0.5$).

There was no significant correlation between middle ear volume and age in either atretic or nonatretic ears (Fig. 2). This is consistent with prior observations that middle ear volume is largely determined at the time of birth [12]. We also assesed change in middle ear volume over time in individuals with serial CT scans. Change in middle ear volume was divided by the time interval between scans to determine the annual rate of change. Eight individuals with unilateral atresia and six control subjects had serial scans. The average interval between scans was 2.9 years (range 0.5–6.1 years). The average age at the first and second scan was 1.7 years old and 4.6 years old, respectively. The atretic ears demonstrated an average rate of change of 0.02 cc/year (standard deviation 0.04 cc/year) versus 0.028 cc/year (standard deviation 0.03 cc/year) in nonatretic ears and 0.02 cc/year in control ears ($p > 0.5$).

Of the 42 cases of unilateral atresia, 19 were deemed surgical candidates and 23 were excluded from atresia repair. The candidate ear average JS score (excluding middle ear aeration) was 7.6 versus 3.6 in noncandidate ears ($p < 0.001$). The noncandidate group included 6 abnormal oval windows, 2 abnormal round windows and 9 abnormal facial nerves. In addition, a two point stapes was not observed in any noncandidate ear.

Measurement of the distance from the promontory to the atresia plate laterally was highly variable. Contributing factors include curvature of the promontory, irregularity of bone lateral to the middle ear space, and plane of imaging (Fig. 3). Variability of the measurement in adjacent sections averaged 0.3 mm anteriorly and 0.5 mm posteriorly to the chosen image (range 0–1.0 mm). Maximal distance of 3 mm or more was seen in 32 of 42 cases, (average 3.4 mm; range 0.5–5.8 mm).

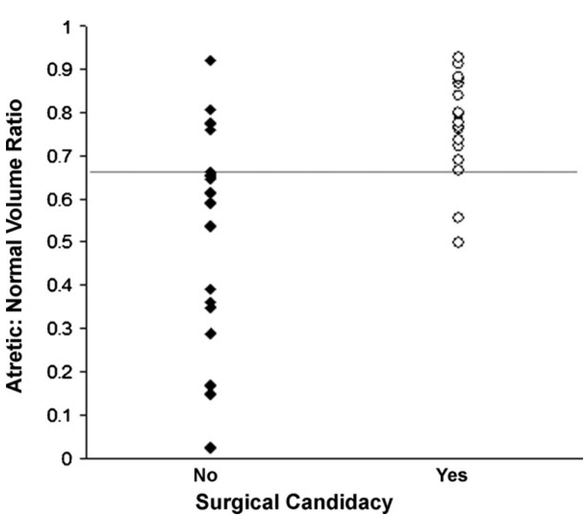

Fig. 4. Surgical candidacy is predicted by the ratio of the middle ear aeration in the atretic ear to that of the normal ear in an individual. The volume ratios for surgical candidates (open circles) and non-candidates (diamonds) are shown. A line is drawn at 0.67 to represent our proposed threshold for surgical candidacy.

Atretic ears deemed operative candidates differed from noncandidates using each of the measurements for assessing middle ear volume. The average maximal distance from the promonotry to bone laterally was 3.9 mm in candidates and 2.9 mm in noncandidates ($p < 0.001$). A significant difference in mean volume was observed between candidate ears (0.43 cc) versus noncandidates (0.28 cc, $p = 0.001$). The mean volume ratio between the atretic ear and the nonatretic ear was 0.79 in candidates and 0.55 in noncandidates. All three of these measurements correlated well with the JS score excluding aeration (volume, $r = 0.65$; distance, $r = 0.67$; volume ratio, $r = 0.70$). Thus, these measurements have the capability to serve as independent predictors of JS score, and overall surgical candidacy.

In an effort to define parameters for assigning a point for middle ear aeration, we compared the distance measurement with volume and volume ratios exceeding the mean in our sample of atresia patients. Using a distance of greater than 3 mm from promontory to bone laterally as a predictor of surgical candidacy yields a sensitivity of 100%, but a specificity of only 43%, and a positive predictive value of 59%. A middle ear volume of 0.35 cc or greater was found to predict surgical candidacy with a sensitivity of 79%, specificity of 61%, positive predictive value of 63% and negative predictive value of 78%. The volume ratio of the atretic ear to the nonatretic ear for a given individual of 0.67 or greater also proved reliable with a sensitivity of 89%, specificity of 74%, positive predictive value of 74%, and negative predictive value of 89% (Fig. 4).

4. Discussion

The Jahrsdoerfer grading scale is valuable as a guide for the systematic interpretation of temporal bone images in aural atresia patients. However, the parameters for scoring have not been stated distinctly. This is further confounded by repeated statements by the originators that a perfect score of 10 is never assigned and the notion that half points are used [7]. If the user arbitrarily decides what constitutes a point for each structure on the scale, it creates difficulty in comparing inter-institutional results. For instance, the distribution of scores for surgical candidates in this series differs dramatically from other published series (Table 1) [6,7]. This study intends to remove some ambiguity by defining a middle ear volume threshold for surgical candidacy.

A previous report examining components of the JS found that middle ear aeration had the most significant correlation with hearing outcome; however, the method for assigning points for aeration was not reviewed [7]. Our measurements support the impression that greater volume is associated with operative

Table 1
Distribution of JS scores in operative candidates.

JS score	UTH [6] 1985–91	UVA [7] 1996–2006	Current
5	3	1	0
6	17	10	0
7	29	49	3
8	29	53	5
9	8	3	7
10	0	0	4
Total	66	105	19

JS score, Jahrsdoerfer scale score; UTH, University of Texas–Houston; UVA, University of Virginia.

candidacy. In theory, greater volume allows increased distance from the edges of the ossicular mass to the bone of the neo-canal resulting in larger area of the tympanic membrane. Since the lever mechanism is absent in anomalous ossicles, a large tympanic membrane:footplate area ratio is essential for optimizing hearing outcomes.

Our results are in accordance with previous volume estimates of middle ear volume. Ikui and colleagues estimated normal middle ear volumes to be 0.45 cc in infants and 0.61 cc in adults [12]. Our slightly smaller volumes may be explained by differences in technique. We were able to find only one study that estimated middle ear volume in patients with aural atresia [10]. These authors demonstrate an average mesotympanic volume of 0.073 cc in atretic ears and 0.134 cc in non-atretic ears. These results are markedly different from ours and are the result of Oliver et al. calculating a hypothetical middle ear volume derived from a series of linear measures. The five fold difference in average volume of atretic ears between the two studies is more surprising when considering the series of Oliver et al. is limited to surgical candidates and does not examine individuals excluded from surgery.

While each of the methods we examined for assessing middle ear volume proved useful, each method is subject to measurement error and has its own drawbacks. These can be categorized into image acquisition and display, operator dependent measurement technique, mathematical calculations, and accessibility and cost of software. The plane of image acquisition is relevant when a linear measurement is made in two dimensions. The exact measure is made when the plane of imaging is exactly parallel to the desired parameter. Slight rotations of the plane of imaging create distortions that can lengthen or shorten the distance of interest. Three dimensional techniques utilizing all images through the structure of interest will minimize this effect, with only the top and bottom most images being subject to distortion effects. Operator dependent measurement effect for linear measures is limited to angulation of the line of interest. When tracing complex structures, there is greater chance for deviation from the desired shape with increasing distances. If volume is the measure of interest, this effect is minimized as deviations from exact are equally likely to include or exclude volume. Mathematical calculations and extrapolation to the measure (volume) of interest are dependent upon above operator dependent measurements. Simple cylindrical or cubic representations of complex shapes based on linear measures are least likely to reflect true volume. Software allowing 3D volume calculations from serial area measurements enhances the accuracy of measuring volume of an irregular shape. However, an area based measurement as in the present study will include the volume of the ossicles within the measured volume. The most accurate method for measuring volume is a calculation based on pixel density. In this manner, specification of a desired density can further improve the accuracy when measuring highly complex shapes of interest [13]. Nonetheless, there will be certain arbitrary points at which boundaries are drawn between the middle ear and mastoid or middle ear and Eustachian tube. Finally, access to desired software programs, cost of purchase, and increased time to perform measurements cannot be ignored when defining optimal measurement method, though these barriers are expected to become less formidable with time.

This study demonstrates that increasing middle ear volume is correlated with higher scores on a nine point JS (excluding middle ear volume). Others have demonstrated the importance of volume on hearing outcomes. We would like to specify a volume measure that would be required for surgical candidacy, though this necessitates correlation with surgical outcomes, which was not a focus of this study. Thus we are limited to proposing theoretical thresholds for volume in establishing surgical candidacy. Our data suggests that volume that is 70% of the normal average (approximately 0.35 cc) and/or a volume ratio of greater than 67% for the atretic ear:nonatretic ear will be the minimum acceptable volume criteria. Further work using this proposed threshold and correlating it with surgical outcomes will be necessary as validation of the current proposal.

We did not find a correlation between middle ear volume and age. Furthermore, our measures of the rate of change in middle ear volume with time were small. Previous work has shown that the volume of the middle ear space is largely determined at birth, but that the total volume does seem to expand with age [12]. This is largely due to expansion of the epitympanic space in adults compared to infants. Our growth rate of approximately 0.02 cc/year would be consistent with that found by Ikui and colleagues. The fact that we do not see the same correlation of middle ear volume with age as this previous study may be due to differences in our methods of measuring the middle ear space. We limited our epitympanic boundary to the region superior to and surrounding the ossicles. This means of measurement would largely eliminate any area of the epitympanum posteriorly as it connects to the antrum. Since the mastoid and antrum undergo the most change in volume postnatally, much of the epitympanic increase in volume seen previously may be due to this posterior aspect. Our measurements, which do not include this portion, would thus not reflect an increase in posterior epiptympanic and antral aeration.

Determining surgical candidacy in aural atresia patients is difficult and honed through experience in evaluating and managing these patients. Properly performed canal reconstruction can produce a postoperative SRT of <30 dB in the majority of patients. However, the aided thresholds achieved with contemporary bone conduction devices are often better, easier to achieve, and more durable [5]. Thus, criteria for surgical candidacy for atresia repair should be refined, offering surgery only in the most optimal cases. Certain anatomical abnormalities mandate exclusion, specifically, oval and round window anomalies. Operative repair of oval window anomalies is challenging and results are marginal at best [14]. There are no descriptions of successful repair of round window aplasia. Other anomalies such as stapes fixation and facial nerve anomalies can be addressed surgically, though not without significant risk [14]. Facial nerve dysfunction in CAA patients is also correlated with inner ear anomalies and severe middle ear malformation [15]. Defined volume measures associated with exclusion of uncorrectable deformities is proposed to improve candidate selection.

5. Conclusions

On average, middle ear volumes of atretic ears average 0.35 ml and are approximately two-thirds the volume of the nonatretic middle ears of the same individual. Both middle ear volume and the ratio of the atretic volume to nonatretic volume serve as useful adjunct measurements in determining surgical candidacy. Finally, the middle ear volume does not significantly change with serial measurement.

Conflict of interest statement

The authors (J.T.V, J.S.O, and A.J.O) declare that they do not have conflicts of interest, financial, intellectual, academic, personal, or otherwise. This study was not sponsored by any outside institution or corporation. Jeffrey T. Vrabec had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

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References

[1] R.A. Jahrsdoerfer, Congenital atresia of the ear, Laryngoscope 88 (September (9 Pt 3 Suppl 13)) (1978) 1–48.
[2] S.E. Kountakis, E. Helidonis, R.A. Jahrsdoerfer, Microtia grade as an indicator of middle ear development in aural atresia, Arch. Otolaryngol. Head Neck Surg. 121 (August (8)) (1995) 885–886.
[3] A. Dostal, J. Nemeckova, R. Gaillyova, V. Vranova, D. Zezulkova, M. Lejska, et al., Identification of 2.3 Mb gene locus for congenital aural atresia in 18q22.3 deletion: a case report analyzed by comparative genomic hybridization, Otol. Neurotol. 27 (April (3)) (2006) 427–432.
[4] P.E. Kelley, M.A. Scholes, Microtia and congenital aural atresia, Otolaryngol. Clin. North Am. 40 (February (1)) (2007) 61–80.
[5] A. De la Cruz, K.B. Teufert, Congenital aural atresia surgery: long-term results, Otolaryngol. Head Neck Surg. 129 (July (1)) (2003) 121–127.
[6] R.A. Jahrsdoerfer, J.W. Yeakley, E.A. Aguilar, R.R. Cole, L.C. Gray, Grading system for the selection of patients with congenital aural atresia, Am. J. Otol. 13 (January (1)) (1992) 6–12.
[7] D.C. Shonka Jr., W.J. Livingston 3rd, B.W. Kesser, The Jahrsdoerfer grading scale in surgery to repair congenital aural atresia, Arch. Otolaryngol. Head Neck Surg. 134 (August (8)) (2008) 873–877.
[8] J.W. Yeakley, R.A. Jahrsdoerfer, CT evaluation of congenital aural atresia: what the radiologist and surgeon need to know, J. Comput. Assist. Tomogr. 20 (September–October (5)) (1996) 724–731.
[9] P.R. Lambert, Major congenital ear malformations: surgical management and results, Ann. Otol. Rhinol. Laryngol. 97 (November–December (6 Pt 1)) (1988) 641–649.
[10] E.R. Oliver, P.R. Lambert, Z. Rumboldt, F.S. Lee, A. Agarwal, Middle ear dimensions in congenital aural atresia and hearing outcomes after atresiaplasty, Otol. Neurotol. 31 (August (6)) (2010) 946–953.
[11] M. Tasar, S. Yetiser, D. Yildirim, U. Bozlar, M.A. Tasar, M. Saglam, et al., Preoperative evaluation of the congenital aural atresia on computed tomography; an analysis of the severity of the deformity of the middle ear and mastoid, Eur. J. Radiol. 62 (April (1)) (2007) 97–105.
[12] A. Ikui, I. Sando, S. Haginomori, M. Sudo, Postnatal development of the tympanic cavity: a computer-aided reconstruction and measurement study, Acta Otolaryngol. 120 (March (3)) (2000) 375–379.
[13] J.T. Vrabec, S.W. Champion, J.D. Gomez, R.F. Johnson Jr., G. Chaljub, 3D CT imaging method for measuring temporal bone aeration, Acta Otolaryngol. 122 (December (8)) (2002) 831–835.
[14] R.A. Jahrsdoerfer, P.R. Lambert, Facial nerve injury in congenital aural atresia surgery, Am. J. Otol. 19 (May (3)) (1998) 283–287.
[15] J.T. Vrabec, J.W. Lin, Inner ear anomalies in congenital aural atresia, Otol. Neurotol. 31 (December (9)) (2010) 1421–1426.