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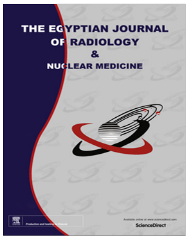

ORIGINAL ARTICLE

Multi-detector computed tomography (MDCT) and audiological criteria to diagnose large vestibular aqueduct syndrome☆

Mohamed M. El-Badry [a], **Nasr Mohamed M. Osman** [b,*], **Haytham Mamdouh Mohamed** [c], **Fatma M. Refat** [d]

[a] *Otolaryngology Department, Audiology Unit, El Minia University, El Minia, Egypt*
[b] *Radiology Department, El Minia University, El Minia, Egypt*
[c] *Otolaryngology Department, Phoniatric Unit, El Minia University, El Minia, Egypt*
[d] *Otolaryngology Department, Audiology Unit, El Minia University, El Minia, Egypt*

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KEYWORDS

LVAS;
45° oblique reformats;
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Abstract *Objective:* The purpose of this study was to compare between Valvassori and Cincinnati criteria and to correlate between vestibular aqueduct measurements made in axial plane with those made in 45° oblique reformats.

Patients and methods: The study group included 61 children with LVAS. All participants were subjected to full Audiological evaluation and MDCT scanning in axial plane. The axial data were transferred to workstation for post-processing with multiplanar reformatting software in order to obtain the 45° oblique reformats. Vestibular aqueduct measurements were made at 4 points: midpoint and operculum in both the axial plane and 45° oblique reformats.

Results: 100% (122 ears) were diagnosed according to Cincinnati criteria, while 81% (99 ears) of children with LVAS fit Valvassori criterion, and 19% were (23 ears) missed. There were statistically significant correlations among the diameters of the VA in the axial plane (midpoint and operculum) and their counterparts in the 45° oblique reformats. Values equal to or greater than 1.2 mm in the midpoint and 1.3 mm in the operculum are proposed to be the criteria to diagnose LVA in the 45° oblique reformats.

Conclusion: Cincinnati criteria are more sensitive than Valvassori criteria in the diagnosis of LVAS.

Abbreviations: VA, vestibular aqueduct; LVA, large vestibular aqueduct; LVAS, large vestibular aqueduct syndrome; SNHL, sensorineural hearing loss; LSC, lateral semicircular canal; SSC, superior semicircular canal; IAC, internal auditory canal

[*] Corresponding author. Tel.: +20 1005039104.

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1. Introduction

Large vestibular aqueduct syndrome (LVAS) is a congenital malformation of the temporal bone characterized by the presence of an abnormally large vestibular aqueduct (VA) and sensorineural hearing loss (SNHL) started from childhood (1). The prevalence of LVAS is estimated to be as high as 15% of pediatric SNHL (2). Generally, it is associated with fluctuating and progressive SNHL; often with sudden onset or progression secondary to minor head trauma, or large sudden shifts of barometric pressure (3,4). Although LVA is considered the most common imaging finding in children with SNHL (5,6) and despite the presence of extensive studies about LVAS, the syndrome is still overlooked and its diagnosis is often missed among radiologist and audiologist. The main objective of the current work was to increase the sensitivity of the radiological diagnosis of LVAS. The earliest description of the LVA was made by Mondini in 1971 (7). However, it was Valvassori and Clemis in 1978 (8) who first described the association between LVA and SNHL and referred to this association as LVAS. The initial size criterion for the diagnosis of LVA was put forth by Valvassori and Clemis (8) in their landmark paper in 1978. In this report, VA was considered enlarged if it was greater than 1.5 mm at the midpoint of its course from the vestibule to the posterior cranial fossa. Most authors have continued to measure VA at the same midpoint and use the Valvassori criterion (i.e., >1.5 mm) to diagnose abnormally large VA. However, Boston et al. in 2007 (9) suggested more sensitive criteria to diagnose LVA (greater than 0.9 mm at the midpoint or greater than 1.9 mm at the operculum). They referred to those criteria as Cincinnati criteria. Studies comparing both criteria are quite few (10,11). One aim of the current work was to compare between Valvassori and Cincinnati criteria as regards their sensitivity to diagnose LVAS.

The 45° oblique plane provides the best view of the VA on tomograms of temporal bone (12). However, this plane requires very difficult head positioning on CT scanner. Therefore, most of the reported data on the CT imaging of the VA were based on measurements obtained on routine axial sections. Currently, with the widespread use of multidetector spiral CT scanners, the 45° oblique image can be easily reformatted without any loss of resolution (12). Ozgen et al. (12) reported that the 45° provides better visualization and more accurate measurement of the VA compared to the routine images in the axial plane. They recommended VA measurement in the 45° oblique reformats in borderline cases; however, they did not specify cutoff measurements between the normal and abnormally large VA as in the case of the axial plane. Other aims of the current work were to measure VA diameter in the 45° oblique reformats in children with LVAS, to correlate between VA measurements made in the axial plane with those made in 45° oblique reformats and determine cutoff criteria to diagnose LVA in the 45° oblique reformats.

2. Aim of the work

The aim of this study was to compare between Valvassori and Cincinnati criteria as regards their sensitivity to diagnose LVAS and to correlate between VA measurements made in the axial plane with those made in 45° oblique reformats and determine cutoff criteria to diagnose LVA in the oblique 45°.

3. Patients and methods

This study was approved by the ethics committee of our institution.

3.1. Patients

The study group included 61 children with SNHL and LVA according to Cincinnati criteria (greater than 0.9 mm at the midpoint or greater than 1.9 mm at the operculum). There was a control group consisted of 25 children with their age and sex distribution statistically matched with the study group. They were free from SNHL and referred for CT scan for reasons other than SNHL as chronic suppurative otitis media. The purpose for including the control children was the computation of normative values of VA in children. All children of the study group were enrolled in the current work after taken a written consent from the parents following detailed explanation of the study procedure.

All patients were referred to the audiology department in the period from 3/2010 to 6/2013. LVAS was suspected in those children based on the history and clinical presentation. All patients referred to the Radiology department for MDCT petrous bone examination.

Children in the study group were 41 females and 20 males ranged from 1.4 years to 18 years with mean age 7.7 ± 3.8 years. History of head trauma was present in 16 patients (26.22%), history of fever was present in 5 patients (8.19%) and history of vertigo was present in 6 patients (9.8%). Hearing loss onset was since birth in 31 patients (50.81%), while in the remaining patients onset was ranged from 0.3 years to 10 years, with mean 2.8 ± 2.1 years. Duration of hearing loss ranged from 0.4 years to 18 years with mean 6.35 ± 4 years. Hearing loss was progressive in 17 patients (27.8%); 9 of them had history of trauma at the onset of progression.

3.2. Methods

Children in this study group were subjected to the following:

(1) **History taking**: including full medical history (prenatal, perinatal and post-natal history), audiological history especially the description of hearing loss onset and progression and history of head trauma, and family history.

(2) **Otoscopic examination.**

(3) **Audiological evaluation** in the form of the following:
   - Immittancemetry to assess middle ear function. Immittancemetry included tympanometry and acoustic reflex threshold recording at frequencies 500, 1000, 2000 and 4000 Hz using middle ear analyzer Zodiac 901. Children with clear conductive pathologies as otitis media were excluded.
   - Hearing assessment: According to the age and reliability of the child, hearing assessment was done through conditioned play audiometry, conventional audiometry, or auditory brainstem response (ABR)

testing. Audiometry was performed using audiometer Amplaid model 309 and sound treated room Amplisilence. Air conduction threshold was measured at frequencies 250, 500, 1000, 2000, 4000 and 8000 Hz and bone conduction threshold was measured at frequencies 500, 1000, 2000 and 4000 Hz. The degree of hearing loss was calculated for each ear based on the average air conduction thresholds at 500 Hz, 1000 Hz, 2000 Hz, and 4000 Hz. Average air conduction threshold $>20 \leqslant 40$ dB HL was considered of mild degree; $>40 \leqslant 55$ was considered of moderate degree; $>55 \leqslant 70$ was considered of moderately severe degree; $>70 \leqslant 90$ was considered of severe degree; >90 was considered of profound degree (13). When there was no measurable hearing at the maximum level of the audiometer (120 dB HL), hearing loss is considered total. Air bone gape (ABG) was calculated as the difference between air conduction threshold and bone conduction thresholds at each of the frequencies 500 Hz, 1000 Hz, 2000 Hz, and 4000 Hz. Exclusion of children with clear conductive pathologies as otitis media maximized the likelihood that the presence of ABG was attributed to the LVAS and not to the conductive pathology. Auditory Brain Stem Response testing was done for children younger than 3 years or children older than 3 years but failed to perform play audiometry. The test was done using intelligent hearing system (IHS) with smart evoked potentials software version 4.5.

(4) **Language assessment**: Language assessment was done using the Arabic Language Test (14). The test included 7 items, which are attention of the child by observation, receptive part of the semantics, expressive part of the semantics, receptive part of the syntax, expressive part of syntax, pragmatics, and prosody.

(5) **MDCT examination of the petrous bone**: Using multidetector CT scanner 16 channel (GE CT/Bright speed Elite scanner, General Electric Medical System, USA).

For both the study group and control group, VA measurements were made at 4 points: midpoint in the axial plane (MPA), operculum in the axial plane (OA), midpoint in the 45° oblique reformats (MOR) and operculum in the 45° oblique reformats (OOR) according to Ozgen et al. and Vijayasekaran et al. (12,15) methods. The midpoint was defined as the halfway between the external aperture and common crus, and a line was drawn in the halfway parallel to the line of the operculum. In the operculum point, VA width was measured by drawing a line from the operculum edge anterolaterally to form 90° angle with the posterior wall of the petrous bone. Figs. 1 and 2 display the method used to measure VA in the axial and 45° oblique reformats. Sometimes 90° could not be achieved when the contour of the operculum and petrous wall is J-shaped. In such instances, 70° or 80° was acceptable. For the control group, VA was graded according to Ozgen et al. (12) criteria for normal VA. They measured VA both in the axial view and in 45° oblique reformats. They categorized the normal VA into 4 grades: nonvisualized (grade 0), visualized with difficulty/very thin (grade I), thin but visible (grade II), and well defined/easily traced (grade III). Only grades II and III are measurable.

CT technique

CT examination was performed using multi-detector CT scanner 16 channel. The images were obtained with 0.5 mm collimation, 0.5 mm thickness, 320 mAs, and 120 kVp. The obtained data were reconstructed in the axial plane using high resolution bone algorithm (extended window sitting at around 4000HU and window level around 300HU) with 0.5 mm section thickness, .05 mm increments, and a FOV of 100, with a matrix size of 512 × 512. At this collimation, an isotropic voxel (which measure 0.5 mm per side) was obtained. The axial data were then transferred to a separate workstation for post-processing, with a commercially available 3D reformatting software (Baxara 3D). A related digital radiograph (scout view) was obtained and sections were performed parallel to the anthropologic line (plane intersecting the inferior orbital rim and the superior margin of the external auditory canal). Sections were taken at 1 mm increments beginning at the level of the floor of hypotympanum and jugular fossa extending to the level of arcuate eminence using line for localization. From it, the axial images were obtained directly and 45° oblique reformats were reconstructed. To obtain the 45° oblique reformats images, the sections were done using the axial image

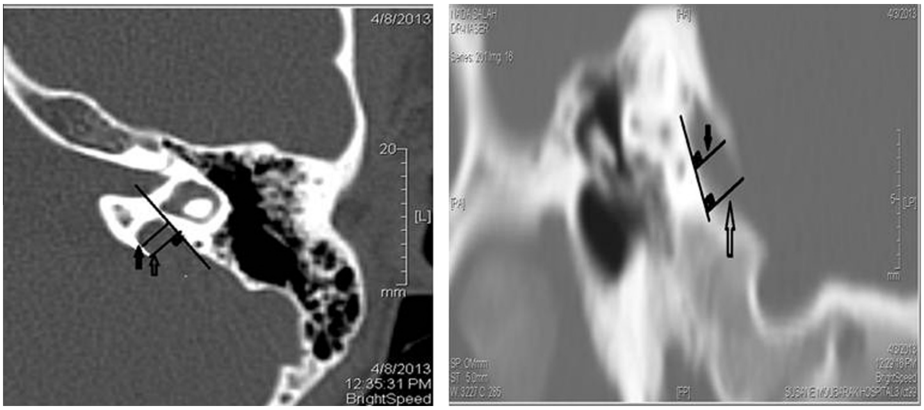

Fig. 1 MDCT scan shows method of VA measurement at the midpoint (black filled arrow) and the operculum (black hollow arrow). The right image in axial cuts and left for the 45° oblique reformats.

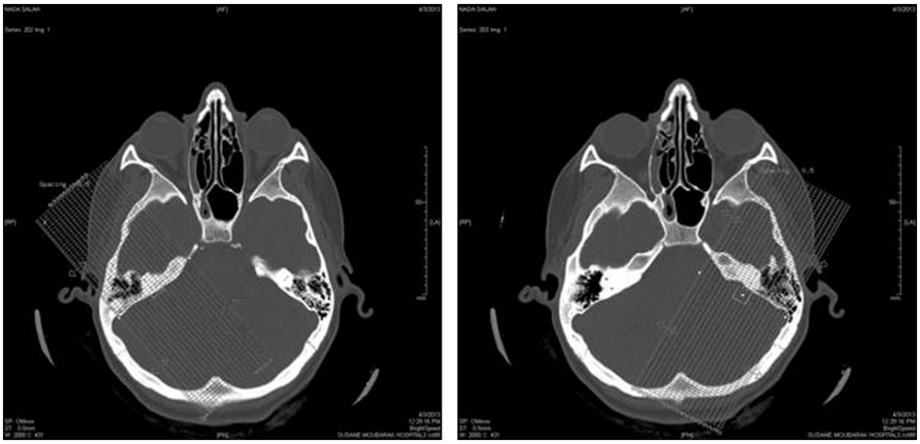

Fig. 2 Planes to obtain the 45° oblique reformats images based on the axial scout images taken parallel to the SSC.

Fig. 3 MDCT scan (axial CT section). Upper image (axial epitympanic level) shows normal VA and lower image (axial LSC level) shows LVA (arrows point to VA).

localizer, and the images were acquired perpendicular to the petrous pyramid with 0.5-mm thickness Fig. 2. Because the superior semicircular canal (SSC) is located perpendicular to the longitudinal axis of the petrous bone, the 45° oblique reformats from an axial scout image was determined to be parallel to the plane of the SSC. Electronic calipers were used for all measurements and the calipers were placed at the last point which demonstrated bone attenuation by visual inspection of each wall of the aqueduct. The scans were monitored to avoid obtaining more slice than necessary. Fig. 3 shows normal VA and LVA in axial plane, and Fig. 4 shows normal VA and LVA in 45° oblique reformats.

Fig. 4 MDCT scan (45° oblique reformats). Upper image shows normal VA and lower image shows LVA (arrows point to VA).

Table 1 Vestibular aqueduct measurements in the control group.

	Minimum (mm)	Maximum (mm)	Mean (mm)	SD (mm)	Mean ± 2.5 SD (mm)
Midpoint axial plane ($N = 16$ ears)	0.5	0.88	0.61	0.09	0.84
Operculum axial plane ($N = 16$ ears)	0.99	1.84	1.23	0.23	1.81
Midpoint 45° oblique reformats (16 ears)	0.54	0.78	0.62	0.16	1.02
Operculum 45° oblique reformats ($N = 16$ ears)	0.65	0.91	0.73	0.15	1.11

4. Results

4.1. Audiological evaluation

Hearing loss was bilateral in all children of the study group. Hearing sensitivity was measured by audiometry in 47 children and was estimated by ABR testing in 14 children. The degree of hearing loss ranged from mild to total. The most common degree was the profound degree (67 ears; 54.9%), followed the severe degree (29 ears; 23.8%). Ten ears (8.2%) had moderately severe degree, seven ears (5.7%) had moderate hearing loss, 7 ears (5.7%) had mild hearing loss, and 2 ears (1.6%) had total hearing loss. For children who performed audiometry, the ABG was present in 86 ears (91.5%); all of them had ABG at 500 Hz and 36 of them (38.3%) had ABG in both 500 Hz and 1 kHz.

4.2. CT scan findings

4.2.1. VA measurements

Figs. 3 and 4 show the VA in one of the control children and one of children with LVAS in the axial view and 45 oblique reformats.

4.2.1.1. Control group. In 50 control ears, 10 ears (20%) had grade 0 (non-visualized), 20 ears (40%) had grade I (visualized with difficulty/very thin), 15 ears (30%) had grade II (thin but visible and might be measured), and 5 ears (10%) had grade III (well defined/easily traced and can be measured). Table 1 shows the minimum, maximum, mean, SD, and the mean ± 2.5 SD of VA diameters for control ears that could be measured in both the axial view and 45° oblique reformats (Table 1).

4.2.1.2. Study group. All children of the study group had bilateral LVA. Table 2 shows the minimum, maximum, mean, and SD of VA diameters of children with LVAS in both the axial plane and 45° oblique reformats. There were statistically significant correlations among the diameters of the VA in the axial plane (both in the midpoint and operculum) and their counterparts in the 45° oblique reformats (r value = 0.81 and p value = 0.001 for the correlation in the midpoint; r value = 0.78 and p value = 0.001 for the correlation in the midpoint).

4.2.2. Valvassori versus Cincinnati criteria for diagnosis of LVAS

The maximum VA diameters in the axial plain in the control children were less than Cincinnati criteria (i.e., less than 0.9 mm at midpoint and less than 1.9 mm at operculum). The values of Cincinnati criteria were less than the mean and 2.5 SD of the VA diameters of the control children at both midpoint and the operculum. 100% (122 ears) were diagnosed according to Cincinnati criteria (i.e., VA diameter larger than 0.9 mm at midpoint and larger than 1.9 mm at operculum). Using Valvassori criterion (i.e. VA diameter larger than 1.5 mm at midpoint in the axial plane), only 81% (99 ears) of children with LVAS were identified, while 19% (23 ears) were missed. Fig. 3 shows the audiogram of a child with LVAS with clinical suspicion for the syndrome (asymmetric hearing loss and the presence of air–bone gap without other indefinable causes for SNHL). The figure also shows the CT scan petrous bone of the child. The diameter of the VA is considered normal according to Valvassori criterion and abnormally large according to Cincinnati criteria.

Table 2 Vestibular aqueduct measurements in the patient group.

	Minimum (mm)	Maximum (mm)	Mean (mm)	SD (mm)
Midpoint axial plane	0.9	4.45	2.34	0.83
Operculum axial plane	1.88	5.83	3.43	1.15
Midpoint 45° oblique reformats	0.8	3.97	2.23	0.74
Operculum 45° oblique reformats	1.16	4.27	2.26	0.78

4.2.3. Criteria to diagnose LVA in the 45° oblique reformats

The fifth percentile of the VA diameter in children with LVAS in the 45° oblique reformats was 1.18 mm in the midpoint and 1.29 mm in the operculum. Statistical analysis using the Receiver Operating Characteristics (ROC) was performed to determine the sensitivity and the specificity of these values as cutoff criteria in the diagnosis of LVA in the oblique 45° reformats. The ROC analysis revealed sensitivity of 98% and a specificity of 100% of these cutoff values in each of midpoint and operculum measurements. We suggest these values (i.e., 1.18 mm in the midpoint and 1.29 in the operculum) to be the cutoff criteria to diagnose LVA in the 45° oblique reformats (see Fig. 5).

4.2.4. Associated congenital anomalies

Table 3 lists the associated congenital anomalies found in CT scans of children with LVAS in the current study. Absent modiolus was the most common associated anomaly. It was found in 90 ears (73.8%) with equal distribution between both ears. Large vestibule was the second common associated congenital anomalies and it was found in 78 ears (63.9) also with equal distribution between both ears. (Figs. 6–8) show some of the associated congenital anomalies in children with LVAS.

4.2.5. Associated syndromes

In the current study, 3 children had the criteria suggestive for Branchio-Oto-Renal syndrome (BOR). Fig. 9 shows a child with some of these features with CT showing the abnormally large VA.

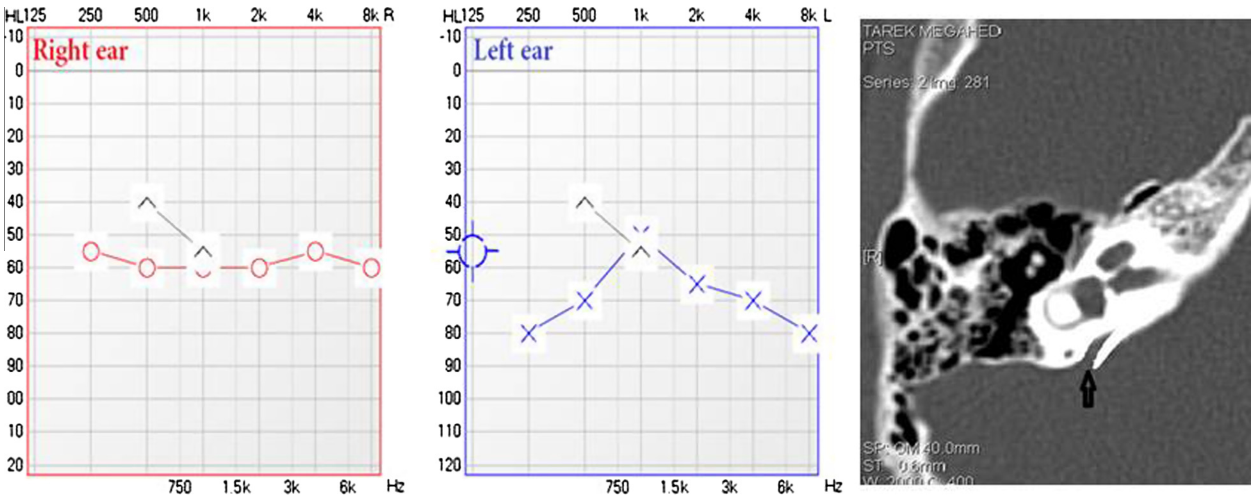

Fig. 5 The audiogram (right and left ears) of child with LVAS. Axial MDCT scan LSC level for the same patient. Black hollow arrow refers to VA which is considered large according to Cincinnati criteria, but normal according to Valvassori criterion.

Table 3 Associated congenital anomalies in children with large vestibular aqueduct syndrome.

Congenital anomaly	Right ear	Left ear	Total
Absent modiolus	45	45	90 (73.8%)
Large vestibule	38	38	78 (63.9%)
High jugular bulb	35	11	46 (37.7%)
Hypoplastic lateral semicircular canal	5	5	10 (8.2%)
Highly deformed cochlea	4	4	8 (6.6%)
Large internal auditory canal	3	3	6 (4.9%)
Dehiscent superior semicircular canal	4	2	6 (4.9%)
Absent posterior semicircular canal	2	2	4 (3.3%)
Cystic cochlea	2	2	4 (3.3%)
Small vestibule	2	2	4 (3.3%)
Laterally displaced ossicles	2	2	4 (3.3%)
Dehiscent Jugular bulb	2	0	2 (1.6%)

5. Discussion

Despite the presence of extensive studies about LVAS, the syndrome is still overlooked and its diagnosis is often missed among radiologist and audiologist. In the current study, the time lag between diagnosing the child as having SNHL and its radiological diagnosis as having LVAS was up to 9 years, with mean of 2.27 ± 2.34 years. This time lag to establish the etiological diagnosis reflects how frequent the LVAS is overlooked and missed as a cause of SNHL and advocates the need to increase the awareness among both audiologists and radiologists about the syndrome. The main objective of the current work was to increase the sensitivity of the radiological diagnosis of the syndrome. The specific aims were to compare between the two famous criteria to diagnose LVA (i.e., Valvassori and Cincinnati), to correlate between VA measurements in the axial view and those in 45° oblique reformats in children with LVAS, and to define radiological criteria to diagnose LVA in the 45° oblique reformats.

5.1. Valvassori criterion vs. Cincinnati criteria

The most popular criterion for the diagnosis of an enlarged vestibular aqueduct was put forth by Valvassori and Clemis in 1978 (8). They considered the VA abnormally enlarged if it was greater than 1.5 mm at the midpoint. This criterion is referred to as Valvassori criterion. Boston et al. (9) suggested more sensitive criteria for LVA (greater than 0.9 mm at the midpoint or greater than 1.9 mm at the operculum). They referred to them as Cincinnati criteria. Their suggestion for the new criteria was based on their findings that 95th percentile of the VA in normal children was 0.9 mm at the midpoint and 1.9 mm at the operculum (12). Studies compared between Valvassori criteria and Cincinnati criteria are scarce. Dewan et al. (13) compared between both criteria by reviewing data from 130 cochlear implant surgeries and found that Cincinnati criteria diagnosed 70 ears as LVA. These ears were diagnosed as normal using the Valvassori criteria. These results indicate that the Cincinnati criteria are much more sensitive than the Valvassori criterion in the diagnosis of LVA.

In the current study, we reached the same conclusion of Dewan et al. (13) by comparing VA measurements in children with LVAS to those in control children. The maximum VA diameters in the control group (0.88 mm at the midpoint and 1.84 mm at the operculum) were less than cutoff values of the Cincinnati criteria. Moreover, the values of Cincinnati criteria were less than the mean and 2.5 SD of the VA diameters of the control children in both the midpoint and the operculum (Table 1). This indicates sensitivity more than 98% of these values to diagnose LVA. On the other hand, only 81% of ears of children with LVAS (99 ears) fitted Valvassori criterion, while 19% (23 ears) were missed. Our results agree with Dewan et al. (13) that Cincinnati criterion is more sensitive than Valvassori criterion in diagnosis of LVA. We recommend

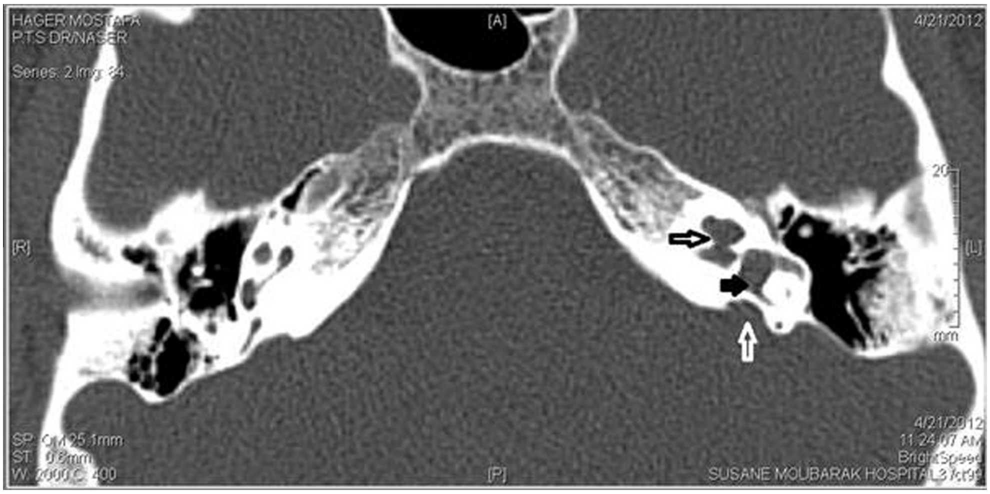

Fig. 6 Axial MDCT scan midtympanic level shows deficient modiolus (black hollow arrow) representing one of the more common inner ear anomalies associated with, large vestibule (black filled arrow) and LVA (white hollow arrow).

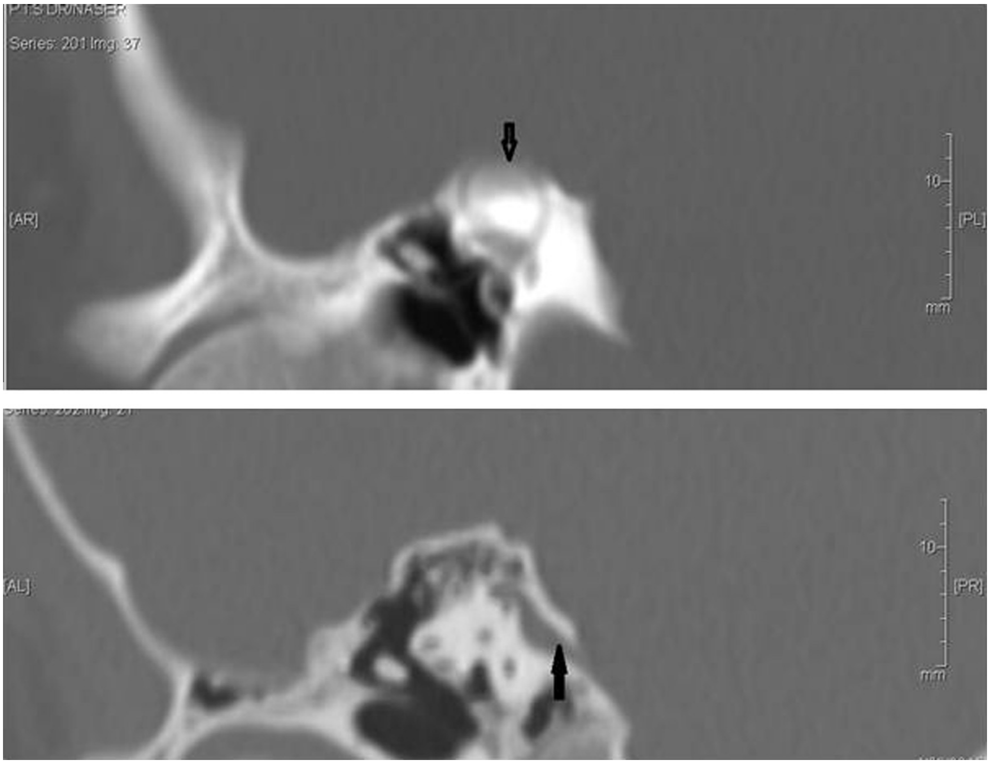

Fig. 7 MDCT scan (45° oblique reformats). Upper image shows dehiscent of the superior semicircular canal (black hollow arrow) associated with LVA. Lower image shows LVA (black filled arrow) of the same patient.

the application of Cincinnati criteria instead of the Valvassori criterion in the diagnosis of LVA as using Valvassori criteria will miss a considerable percentage of children with LVAS (19% in the current study).

It was our clinical observations that there are many children with clinical suspicion of LVAS in the absence of other identifiable causes of SNHL. The diameter of VA was considered normal using the Valvassori criterion. Using Cincinnati criteria established the diagnosis of LVAS in those children. Fig. 3 in the current study shows the audiogram and CT scan of one of these children. The asymmetric hearing loss and the presence of air–bone gap without other indefinable causes for SNHL are some of clinical features suggestive for LVAS. The VA is considered normal using the Valvassori criterion, but it is abnormally large according to the Cincinnati criteria. Therefore, using Cincinnati criteria instead of Valvassori criterion will increase the identification of more cases with LVAS and identify the cause of SNHL in subset of children in whom the cause of SNHL is thought to be unknown. Determining the cause of SNHL in the cases of LVAS has very important clinical implications related to the management of this syndrome. Avoidance of head trauma, and some kind of sports and activities that predispose to hearing loss progression in children with LVAS is an important point in the management to prevent hearing loss progression. The progressive nature of this hearing disorder must be considered in the selection of suitable hearing aid parameters. In addition, because of the progressive nature of hearing loss in LVAS, children with this syndrome are potential candidates for cochlear implantation.

5.2. CT scan axial plane vs. 45° oblique reformats

Working on 30 ears of normal children, Ozgen et al. (12) reported that the 45° oblique reformats provides better visualization and more accurate measurement of the VA compared to the routine images in the axial plane. Ozgen et al. (12) recommended VA measurement in the 45° oblique reformats in borderline cases; however, they did not measure VA diameter in children with LVAS and did not specify cutoff criteria between the normal and abnormally large VA. In the current study, the VA was measured at the midpoint and the operculum both in the axial plane and in the 45° oblique reformats. There was statistically significant correlation among the diameters of the VA in the axial view (both in the midpoint and in the operculum) and their counterparts in the 45° oblique reformats. The results indicate the validity of VA measurements in the 45° oblique reformats in the diagnosis of LVAS.

The upper limits of normal VA measurements found by Ozgen et al. (12) in the 45° oblique reformats were 0.73 mm at the midpoint and 0.85 mm at the operculum. In the current study, the upper limit of VA in the control children was 0.78 mm in the midpoint and 0.91 mm in the operculum. These measurements are very close to those obtained by Ozgen et al. (12). These values are less than the minimum diameter found in children with LVAS participated in the current study. The fifth percentile of the VA of children with LVAS in 45 oblique reformats was 1.18 mm and 1.29 mm for the midpoint and the operculum respectively. The ROC analysis revealed sensitivity of 98% and specificity of 100% of these values as cutoff values in each of midpoint and operculum measurements. We suggest

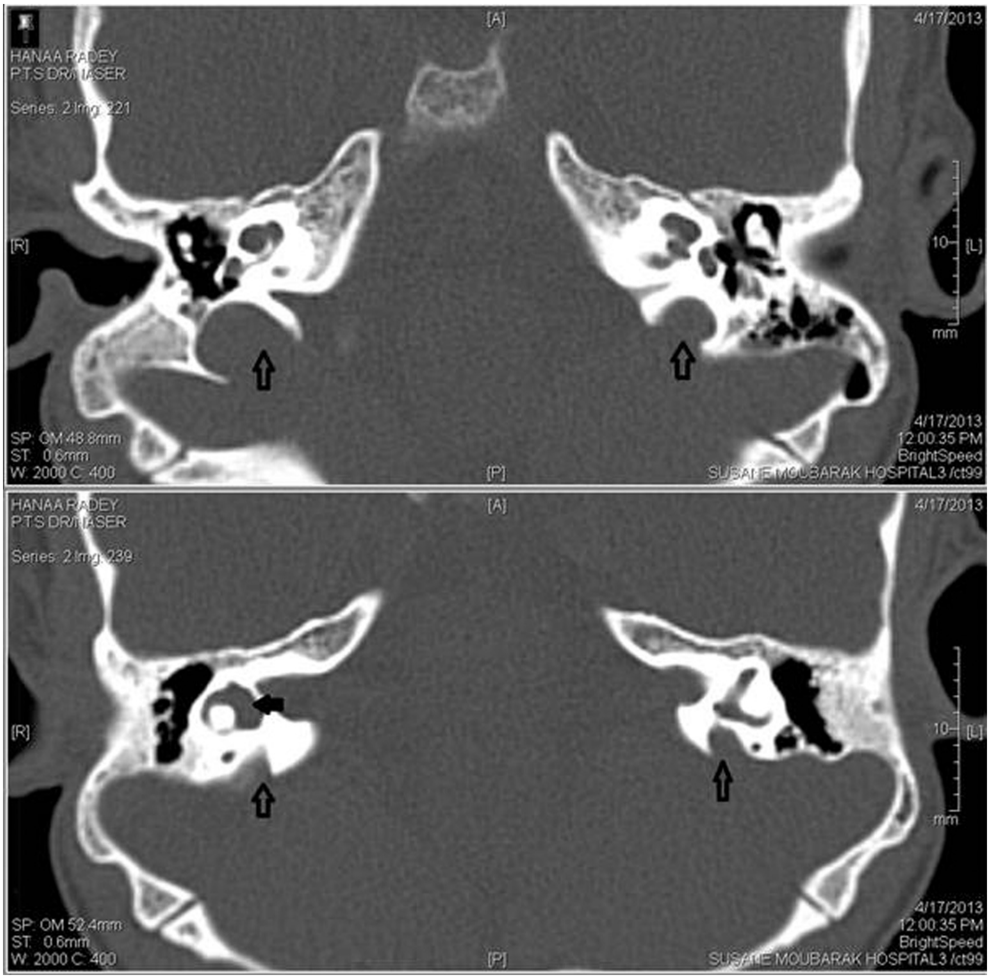

Fig. 8 Axial MDCT. Upper cut is axial epitympanic level image showing bilateral high riding jugular bulb (hollow arrow), lower cut is axial IAC level image showing bilateral LVA (hollow arrows). Large vestibule (filled arrows) seen at the right side of the same patient.

values equal to or greater than 1.2 mm in the midpoint and 1.3 mm in the operculum to be the criteria to diagnose LVA in the 45° oblique reformats.

5.3. Correlations among radiological findings, subject characteristics, and audiological findings

No significant correlations were found between the VA diameters at the axial or 45° oblique and each of the following: children age, children sex, history of trauma, history of vertigo, history of fever, history of hearing loss progression, positive family history, hearing loss onset, hearing loss duration, presence of air–bone gab, and language ability. Results indicate that none of the previously mentioned items can predict that size of VA in either the axial view or 45° oblique reformats.

Most studies on LVAS have not shown correlation between hearing loss severity and duct size (16–20). Similar finding was found in the current study whereas the average audiometric thresholds of children with LVAS did not correlate with VA size at any of the measurement points. Fig. 10 displays such result. The figure shows the audiograms of a child with LVAS with marked asymmetry in the hearing sensitivity. However, there was not much difference in the VA size between the 2 ears. No specific explanation was reported in the literature for the absence of correlation between the radiological size of the VA and audiometric threshold. Overall, results of the current study and other studies demonstrate that the audiometric threshold is not a predictor for the VA size similar to the subject characteristic and other audiological findings. Moreover, results indicate that absolute size of VA is not an important factor in the rehabilitation process of children with LVAS. Rather, VA size is essential and critical factor in the diagnosis and identifying the cause of SNHL.

The incidence of inner ear anomalies associated with LVA was between 41% (21) and 55% (22). Modiolar deficiency was the most common anomalies found (23). In current studies, several inner ear anomalies were found. Table 3 lists these anomalies and their percentages. Figs. 6–8 show examples of these anomalies. Consistent with the literature, the most common anomaly was modiolar deficiency.

6. Conclusion and recommendation

Cincinnati criteria are more sensitive than Valvassori criteria in the diagnosis of LVAS. We recommend the application of

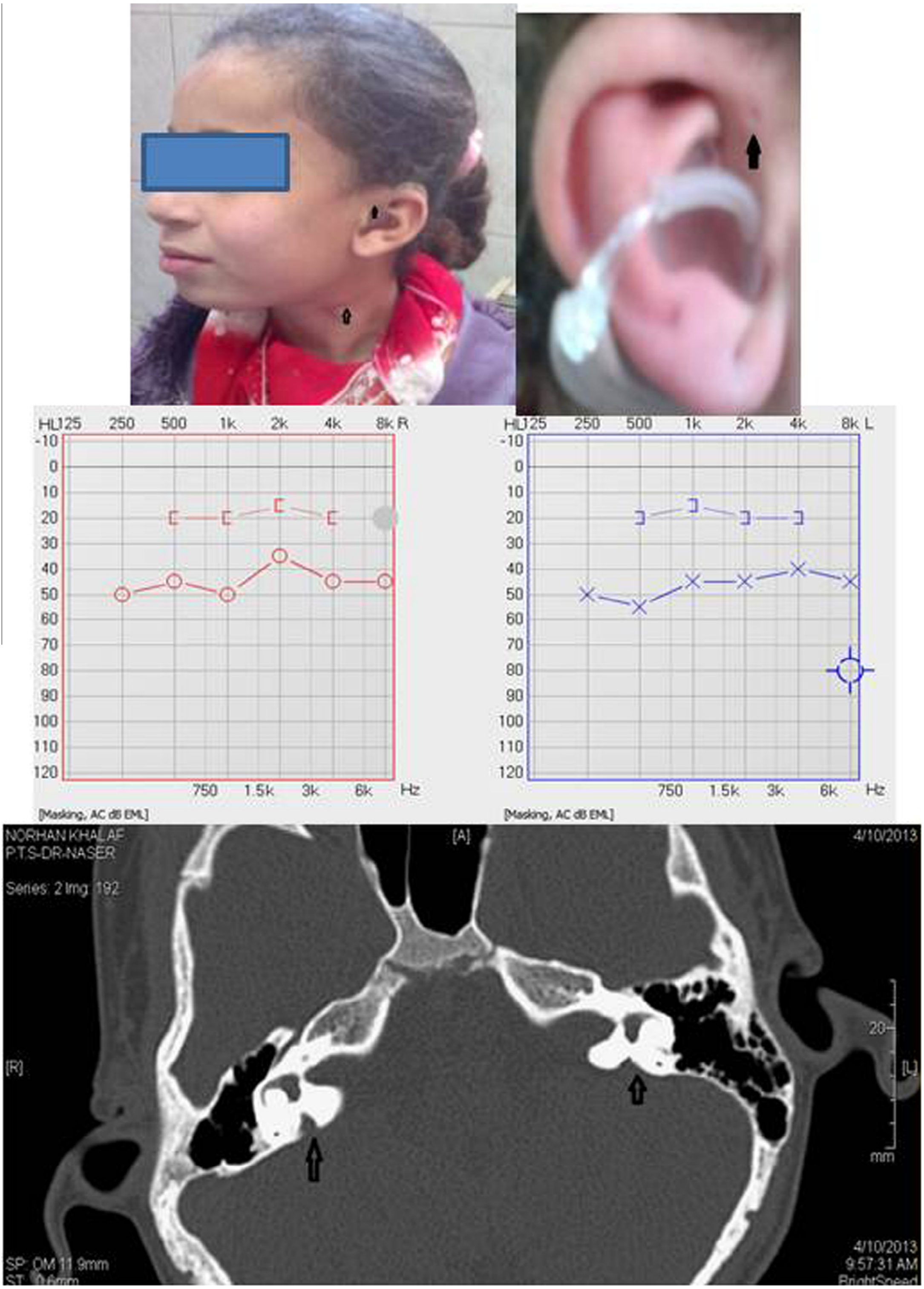

Fig. 9 Shown the Branchio Oto Renal syndrome. Patient face showing cup shaped auricle, branchial fistula (black hollow arrow) and pre-auricular fistula (black filled arrow). Middle image shows right and left audiograms. Axial MDCT scan showing bilateral LVA (Black hollow arrows).

Cincinnati criteria instead of Valvassori criteria. In children with LVAS, there are good correlations in the VA diameters between the routine axial plane and 45° oblique reformats. The early confirmed diagnosis of LVAS is important in patient management by hearing aids and cochlear implantation to facilitate language development.

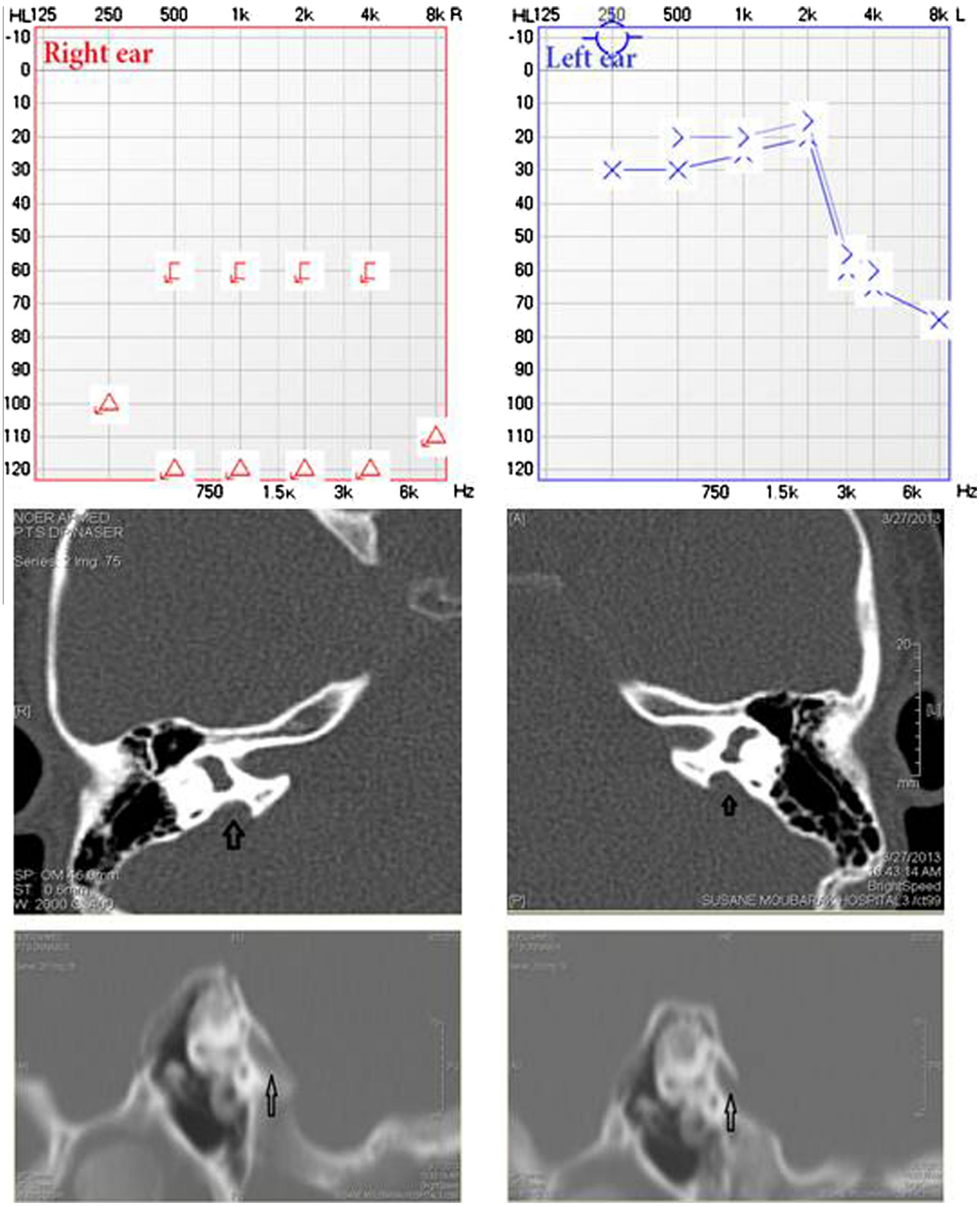

Fig. 10 Upper image shows the audiogram of a child with LVAS. Right ear had total hearing Loss and left ear had moderate hearing Loss. MDCT scan in axial IAC level and 45° oblique reformats of the same patient. Arrows denote the LVA. Although there was a large difference in the audiometric thresholds between right and left ear, there was a small difference between VA diameters. Measurements of the VA are as follows: axial view (right midpoint = 2.76 mm; left midpoint = 3 mm; right operculum = 4.53 mm; left operculum = 4.17 mm). 45° oblique reformats (right midpoint = 3.14 mm; left midpoint = 2.82 mm; right operculum = 3.13 mm; left operculum = 2.81 mm).

Conflict of interest

No conflict of interest statement.

Authors' contribution

All authors have appraised the article and actively contributed in the work.

Nasr Mohamed M. Osman: Data collection, technique, CT interpretation, revision and final editing.

Mohamed Mohamed El Badery: Patients selection, data collection and audiological examination.

Hytham M. Mohammed: Patients selection, and phoniatrics examination.

Fatma M. Refat: Audiological examination.

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