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Evaluation of the radiological criteria to diagnose large vestibular aqueduct syndrome

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ABSTRACT

Objective: The main objective of the current work is to increase the sensitivity of the radiological diagnosis of the large vestibular aqueduct syndrome (LVAS). The specific aims were to compare between the two famous criteria to diagnose large vestibular aqueduct (LVA), (i.e., Valvassori and Cincinnati), to correlate between vestibular aqueduct (VA) measurements in the axial view and those in 45° oblique reformate in children with LVAS, and to define radiological criteria to diagnose LVA in the 45° oblique reformate.

Methods: The study group included 61 children with LVAS according to Cincinnati criteria (greater than 0.9 mm at the midpoint or greater than 1.9 mm at the operculum in the axial view). All participants were subjected to full Audiological evaluation and CT scanning in axial plane. The axial data were then transferred to workstation for post-processing with 3D reformatting software (Baxara 3D) in order to obtain the 45° oblique reformates. VA measurements were done at 4 points: midpoint and operculum in both the axial plane and the 45° oblique reformate.

Results: Only 81% of ears of children with LVAS (99 ears) fit Valvassori criterion (i.e., larger than 1.5 mm at midpoint), while 19% (23 ears) of them were missed. There were statistically significant correlations among the diameters of the VA in the axial view (both in the midpoint and operculum) and their counterparts in the 45° oblique reformate. 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 reformate. Finally, no significant correlations were found between the degree of hearing loss and VA diameters at the axial or 45° oblique reformate.

Conclusion: Cincinnati criteria are more sensitive than Valvassori criterin in the diagnosis of LVAS. We recommend the application of Cincinnati criteria instead of Valvassori criteria in order not miss cases with LVAS. Measurement of VA in the 45° oblique reformate is a reliable method to diagnose LVA. Criteria to diagnose LVA in the 45° oblique reformate were proposed.

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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 is to increase the sensitivity of the radiological diagnosis of LVAS.

The earliest description of the LVA was made by Mundini in 1791 [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

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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]. 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. 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 [11]. 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 reformate 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 reformate in children with LVAS, to correlate between VA measurements made in the axial plane with those made in 45° oblique reformate in those children and determine cutoff criteria do diagnose LVA in the 45° oblique reformate.

2. Subjects and methods

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 that 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.

Children in the study group were 41 females and 20 males. Age of them ranged from 1.4 years to 18 years with mean 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 ± 4years. Hearing loss was progressive in 17 patients (27.8%); 9 of them had history of trauma at the onset of progression.

Children in the study group were subjected to:

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:
   - 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 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 $>20 \leq 40$ dB HL was considered of mild degree; $>40 \leq 55$ was considered of moderate degree; $>55 \leq 70$ was considered of moderately severe degree; $>70 \leq 90$ was considered of severe degree; >90 was considered of profound degree. 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 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. The stimuli were 100 μs alternating click delivered through insert ear phone at intensity level of 90 dB nHL and in 10 dB decrement till abolishment of the response. Repetition rate of the stimuli was 31p/s. Electrode montage was high forehead to ipsilateral mastoid. The common electrode was placed on the contralateral mastoid. The response was filtered between 100 and 3000 Hz, amplified 100,000 times, recorded over 10.24 ms time window, and 2000 sweeps were averaged for each run. ABR testing for each child was carried under chloral hydrate.
4) Language assessment: language assessment was done using the Arabic Language Test [13]. 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) Multi-detector computed tomographic (MDCT) examination of the petrous bone:

   For both the study group and control group, VA measurements were done at 4 points: midpoint in the axial plane (MPA), operculum in the axial plane (OA), midpoint in the 45° oblique reformate (MOR) and operculum in the 45° oblique reformate (OOR). Ozgena et al. and Vijayasekaran et al. [12,14] methods were used in the measurement at the 4 points. The midpoint was defined as the half way between the external aperture and common crus, a line was drawn in the half way 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 90̊ 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 reformate. 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

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