Research Article

Audiological, Language and Multi-Detector Computed Tomography Evaluations in Children with Large Vestibular Aqueduct Syndrome

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Abstract
Purpose: To compare between Valvassori and Cincinnati criteria as regards their sensitivity to diagnose LVAS, To correlate between VA measurements made in the axial plane with those made in ٤٥° oblique reformate and determine cutoff criteria do diagnose LVA in the oblique ٤٥°, To evaluate language ability and hearing aid performance in children with LVAS and To correlate among subject profile, audiological findings, language ability and radiological findings in children with LVAS. **Materials and methods: The study group** included ٦١ children with LVAS diagnosed by MDCT were subjected to full audiometric assessment, language assessment and MDCT measurements of VA diameters in ٢ views axial and ٤٥° oblique reformate. **The control group** consisted of ١٠ children free from SNHL were subjected to MDCT measurements of VA diameters in ٢ views axial and ٤٥ ° oblique reformate. **Results:** ١٠٠٪ of children with LVA who had been included in our study had bilateral LVA as well as bilateral hearing loss. Degree of hearing loss ranged from mild to total. Air bone gap (ABG) was present in ٨٦ ears. Language age (LA) ranged from below ٢ years to fully developed language. No significant statistical correlation was found between VA size at the ٤ views and the difference between chronological (CA) and language age (LA) as an indicator of language ability. Many associated congenital anomalies were found in CT scans. The Cincinnati criteria are much more sensitive than the Valvassori criterion in diagnosis of LVA. The cutoff criteria for diagnosis of LVA in the oblique ٤٥° reformate are ٠.٨٧ mm at the midpoint and ١.١٦ mm at the operculum with ١٠٠٪ sensitivity and ١٠٠٪ specificity. **Conclusion:** LVAS is overlooked and missed as a cause of SNHL. We advocate the need to increase the awareness among both audiologists and radiologists about the syndrome and recommend using Cincinnati criteria instead of Valvassori criterion. We also recommend routine ٤٥°oblique reformate with the axial plane in children with SNHL suspected to have LVAS.

Keywords: Audiological, children, language, Aqueduct Syndrome

Introduction

Large vestibular aqueduct (LVA) is the most common imaging finding in children with sensorineural hearing loss (SNHL)[١,٢]. The earliest description of the LVA was made by Mondini in ١٩٧١[٣]. However, it was Valvassori and Clemis in ١٩٧٨[٤] who first described the association between LVA and SNHL and coined the term large vestibular aqueduct syndrome (LVAS) to describe this association. Since then, there were extensive studies describing the audiological profile and radiological findings in children with LVAS. The prevalence of LVAS is estimated to be as high as ١٥٪ of pediatric SNHL[٥]. Despite such high prevalence and presence of extensive studies, the syndrome is still overlooked and its diagnosis is often missed among radiologists and audiologists. Moreover, there are some points that deserve to be re-addressed as the radio-logical criteria to diagnose LVA and correlations among the audiological and radiological findings. In addition, there are other points that were not addressed before in LVAS research as language ability and hearing aid performance of children having the syndrome.

The initial size criterion for the diagnosis of an enlarged vestibular aqueduct was put forth by Valvassori and Clemis[٤] in their landmark paper in ١٩٧٨. In this report, the bony vestibular aqueduct (VA) was considered enlarged if it was greater than ١.٥ 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., > ١.٥ mm) to diagnose abnormally large VA. However, Boston et al.,[٦] suggested that the criteria for LVA to be revised to greater than ٠.٩ mm at the midpoint or greater than ١.٩ mm at the operculum. They referred to those criteria as Cincinnati criteria. One aim of the current work was to compare between Valvassori and Cincinnati criteria as regards their sensitivity to diagnose LVAS.

The ٤٥° 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 ٤٥° oblique image can be easily reformatted without any loss of resolution[٧]. Working on ٣٠ ears of normal children, Ozgen et al.,[٨] reported that the ٤٥° provides better visualization and more accurate measurement of the VA compared to the routine images in the axial plane. Ozgen et al.[٨] recommended VA measurement in the ٤٥° 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. Another aims of the current work were to correlate between VA measurements made in the axial plane with those made in ٤٥° oblique reformate and determine cutoff criteria do diagnose LVA in the oblique ٤٥°.

Rehabilitation of children with LVAS depends on the use of amplification device (hearing aids or cochlear implant). Determining the candidacy of children to either device depends essentially on their performance with the hearing aids and on their language ability. For our best knowledge, formal studies on the response to the hearing aid and language ability in children with LVAS are not available.

Patients and Methods

The study group included ٦١ children with SNHL proved by MDCT scan to have LVA. Forty children were referred for hearing assessment to the audiology unit, Minia University hospital in the period between ٣/٢٠١٠ and ٦/٢٠١٢. LVAS was suspected in those children based on the history and clinical presentation (see below) and referred to the radiology unit, Minia University hospital for CT scan assessment. Those ٤٠ children were recalled to be enrolled in the current study. In addition to those, ٢١ children newly diagnosed as having LVAS (from ٦/٢٠١٢ to ٦/٢٠١٣) were added to the study group. All children of the study group were enrolled in the current work after taken an oral consent from the parents and detailed explanation of the study procedure.

The control group consisted of ١٠ children with their age and sex distribution matched with the study group. They were free from SNHL and referred for CT scan for reason other than SNHL as CSOM. The purpose for including the control children was the computation of normative values of VA in children and to control for the specific method used in the current study to measure the VA.

Criteria that were used to suspect LVAS before referral for CT scan assessment were:

١- History of head trauma at the onset of hearing loss
٢- History of unexplained hearing deterioration either unilateral or bilateral

٣- Family history of siblings having LVAS
٥- Audiometric suspicions:

- Documented unilateral or bilateral hearing deterioration.
- Presence of air –bone gap in the frequencies (٥٠٠, ١٠٠٠ Hz) with normal middle ear pressure.
- Asymmetric SNHL.

Children in the study group were subjected to:

١**) <u>History taking:</u>** including full medical (prenatal, perinatal and post natal) and audiological history especially the description of hearing loss onset and progression in addition to history of head trauma and family history.

٢**) <u>Otoscopic examination</u>.**

٣**) <u>Audiological evaluation in the form of:</u>**

- Immittancemetry including tympanometry and acoustic reflex threshold recording at frequencies ٠.٥, ١, ٢ and ٤ KHz using impedance audiometer Zodiac ٩٠١.

- Conditioned play audiometry or conventional audiometry according the age and reliability of the children to assess the hearing threshold using audiometer amplaid model ٣٠٩ and sound treated room amplisilence. Speech reception threshold (SRT) and speech discrimination using Kinder Garden Arabic phonetically balanced words.

- Auditory Brain Stem Response (ABR) for children younger than ٣ years or older but failed to perform play audiometry: ABR was done to estimate hearing threshold using intelligent hearing system (IHS) with smart evoked potentials software version ٤.٥

- Aided response was done to assess the aided hearing threshold at frequencies ٠.٢٥, ٠.٥, ١, ٢ and ٤ kHz, using Madsen Astera audiometer and sound treated room amplisilence.

٤**) <u>MDCT Examination of the petrous bone:</u>**

CT scanning was done after a detailed explanation of the procedure to the parents and children with emphasis on the importance of refraining from movement throughout the procedure.

CT scan was performed using multi-detector CT scanning in the axial plane with a ١٦ channel multi detector CT scanner. We perform ٢ views axial and ٤٥ oblique reformate.

VA measurements were done at ٤ points: operculum in the axial plane (OA), midpoint in the axial plane (MPA), operculum in the ٤٥° oblique reformate (OOR) and midpoint in the ٤٥° oblique reformate (MOR) (Figure ١, ٢) .

CT images were interpreted in details to determine VA measurements and any associated congenital anomalies in ear as external auditory canal (EAC) atresia, ossicular abnormality, high jugular bulb, dehiscent jugular bulb, SCCD, vestibule-ocochlear anomalies and wide IAC.

For the control group, CT scan was performed and VA measurements were done at the ٤ measurement points. In addition, VA in the control ears was graded according to Ozgena et al [٨] as follow: nonvisualized (grade ٠), visualized with difficulty/very thin (grade ١), thin but visible (grade ٢), and well defined/easily traced (grade ٣).

٥**) <u>Language assessment:</u>**

Language assessment was done using the Arabic Language Test (Kotby et al., ١٩٩٥)[٩]

Results

The study group consisted of ٦١ patients. There were ٤١ females and ٢٠ males. Age of the study group ranged from ١.٤ years to ١٨ years, average was ٧.٧ years. History of head trauma was present in ١٦ patients (٢٦.٢٢٪), history of fever was present in ٥ patients (٨.١٩٪) and history of vertigo was present in ٦ patients (٩.٨٪). Hearing loss onset was since birth in ٣١ patients (٥٠.٨١٪), while in the remaining patients onset was ranged from ٠.٣ years to ١٠ years, average was ٢.٨ years, hearing loss was progressive in ١٧ patients (٢٧.٨٪); ٩ of them had history of trauma at the onset of progression (٤٧٪).

Positive family history of hearing loss was found in ٣٧ patients (٦٠.٦٥ %). LVA was present in ٦ patients with syndromic SNHL;

٣ of them were BOR syndrome, ١ was Treacher Collins syndrome, ١ was Wardenburg syndrome and the last was auditory neuropathy spectrum disorder (ANSD).

١٠٠٪ of children with LVA who had been included in our study had bilateral LVA as well as bilateral hearing loss. PTA was done to ٩٤ ears, average PT threshold of ٠.٥- ٤ kHz ranged from ٣١.٢٥ dB HL to ١١٦.٦٧ dB HL, mean was ٩٠.٣٦ dB HL (Figure ٣). Degree of hearing loss ranged from mild to total, ٥ ears (٥.٣٪) had mild hearing loss, ٥ ears (٥.٣٪) had moderate hearing loss, ٧ ears (٧.٤٤٪) had moderately severe hearing loss, ٢٢ ears (٢٣.٤٪) had severe hearing loss, ٥٥ ears (٥٨.٥٪) had profound hearing loss and ١ ear (١.٠٦٪) had total hearing loss (Figure ٤). Hearing loss was asymmetric in ٢٢ patients (٤٦.٨٪). This asymmetry in PTA average threshold ranged from ١٥ dB HL to ٨٢ dB HL, mean was ٣٢.٠٣١. Many audiometric configurations were found in children with LVAS (Figure ٥,٦). Flat configuration was the most common and was found in ٥٠ ears (٥٣٪), followed by high frequency sloping which was found in ٢٥ ears (٢٧٪).

Air bone gap (ABG) was present in ٨٦ ears (٩١.٤٪) (Figure ٦), all of them had ABG at ٥٠٠ Hz and ٣٦ of them (٣٦.٢٩٪) had ABG in both ٥٠٠ Hz and ١ KHz. Calculation of ABG was done after exclusion of children with type B tympanogram and other conductive pathologies (atresia and CSOM). At ٥٠٠ Hz, ABG ranged from ١٠ dB HL to ٧٠ dB HL, average was ٤٠.٠٧ dB HL.

Eleven children with LVAS (٤٨٪) had an aided threshold within the spectrum energy of speech vowels and constant. Language age (LA) ranged from below ٢ years to fully developed language. The majority of patients; ٢٠ patients (٥٧٪) had LA below ٢ years and ٣ patients (٨.٥٧٪) had fully developed language (Figure ٦٤). In between LA ranged from ٢.١١ years to ٦.٨ years, average was ٤.٥٨ years, median was ٤.٨٥ years and SD ١.٤٧ years. Chronological age (CA) ranged from ٢.١١ years to ١٤ years, average was ٧.٧ years, median was ٧.٦ years, mode was ٥ years and SD was ٣.٣٦ years. The difference between CA and LA ranged from ٠ year and ١٢.٩ years, average difference was ٤.٦ years, median was ٤ years, mode was ٤ years and SD was ٣.٣ years. No significant statistical correlation was found between VA size at the ٤ views and the difference between chronological (CA) and language age (LA) as an indicator of language ability.

In ٢٠ control ears, ٤ ears had grade ٠, ٨ ears had grade I, ٦ ears had grade II, ٢ ears had grade III. In study group ١٠٠٪ of children with LVA who had been included in our study had bilateral LVA. In ٦٨ ears in study group, ٤ measurements were taken; ٢ in axial view and ٢ in ٤٥° oblique reformate. In axial view, operculum measures ranged from ١.٢٤mm to ٥.٨٣mm, average was ٣.٤٣mm. In ٤٥° oblique reformate, operculum measures ranged from ١.١٦ mm to ٤.٢٧mm, average was ٢.٢٦٣ mm. Many associated congenital anomalies were found in CT scans. Absent modiolus was found in ٥٢ ears (٧٤.٢٨٪) with equality in distribution in both ears. Large vestibule was found in ٤٦ ears (٦٥.٧٪) also with equality of distribution in both ears.

١٠٠٪ (٦٨ ears) were diagnosed according to Cincinnati criteria (i.e., had VA diameter larger than ٠.٩mm at midpoint in the axial view and/or larger than ١.٩mm at operculum in the axial view. Using Valvassori criterion larger than ١.٥mm at midpoint in the axial view, only ٨٢٪ (٥٦ ears) were diagnosed as having LVA while ١٨٪ (١٢ ears) of them were missed (Figure ٧).

Statistical analysis was performed on VA measurement of the children with LVAS (٦٨ ears) using the Receiver Operating Characteristics (ROC) curve to specify cutoff criteria for diagnosis of LVA in the oblique ٤٥° reformate and to determine the sensitivity and specificity of this criteria. The ROC analysis revealed that cutoff criteria were ٠.٨٧ mm at the midpoint and ١.١٦ mm at the operculum with ١٠٠٪ sensitivity and ١٠٠٪ specificity.

Age of the children was not significantly correlated with diameter of the VA at any measurement points. No significant difference was found between the diameter of the VA in the right and left side at the ٤ measurement points. Statistical significant correlations were found among the ٤ measurements indicating that enlargement of the VA at one measurement point is associated with its enlargement at the other points. Worse threshold was associated with earlier hearing loss onset, longer hearing loss duration and larger ABG at ٠.٥ and ١ kHz. There was no significant difference between children with and children without severe cochlear deformities as regard difference between LA and CA.

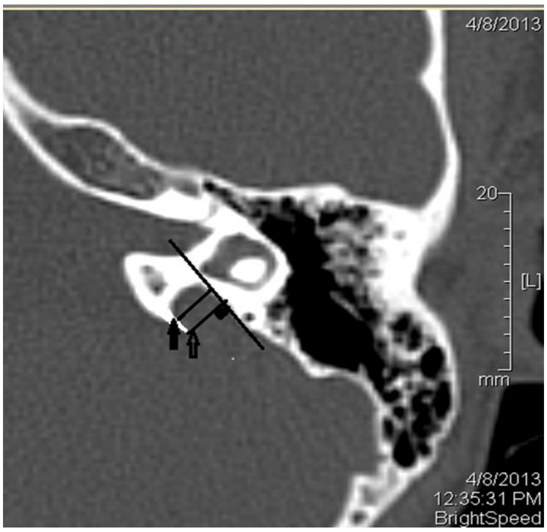

Figure ١: MDCT scan (axial view) shows method of measurement of midpoint (black filled arrow) and operculum (black hollow arrow)

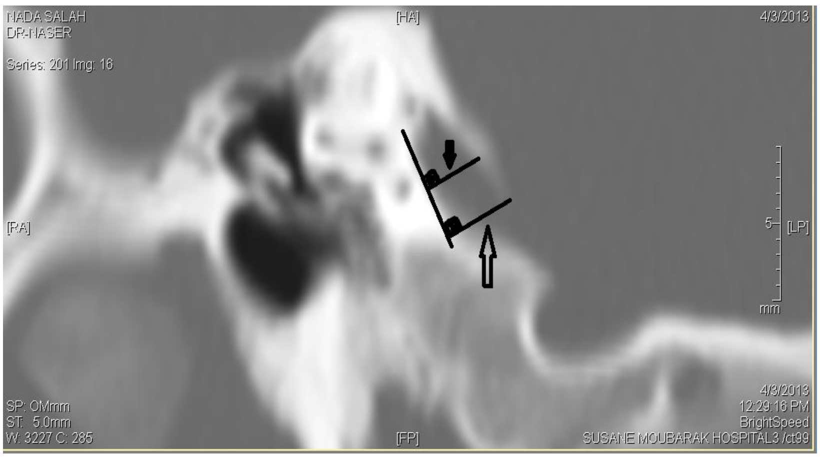

Figure ٢: MDCT (٤٥° oblique reformate) shows method of measurement of midpoint (black filled arrow) and operculum (black hollow arrow)

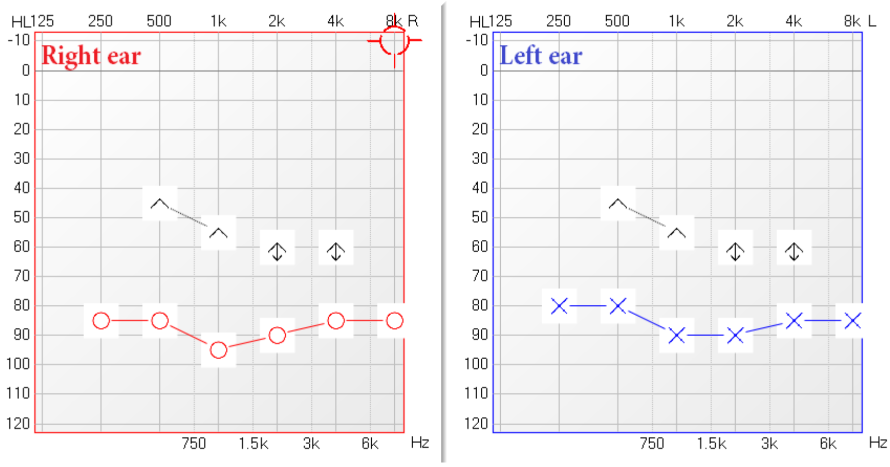

Figure ٣: Average audiometric threshold of children with LVAS

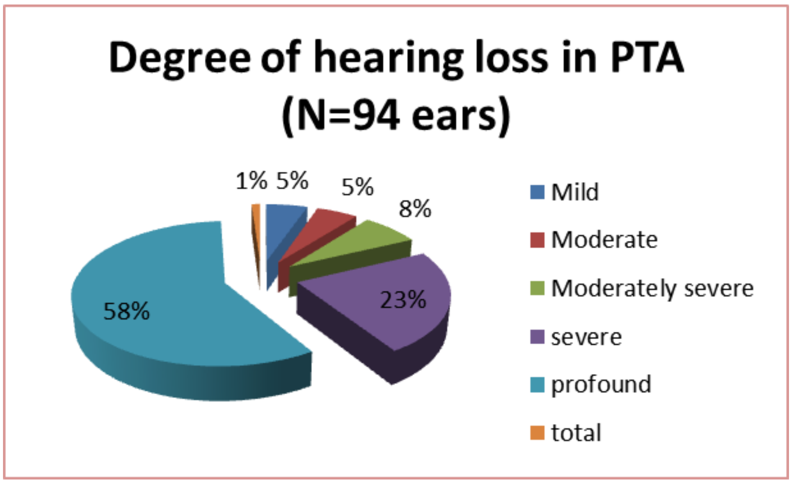

Figure ٤: Degrees of hearing loss in children with LVAS

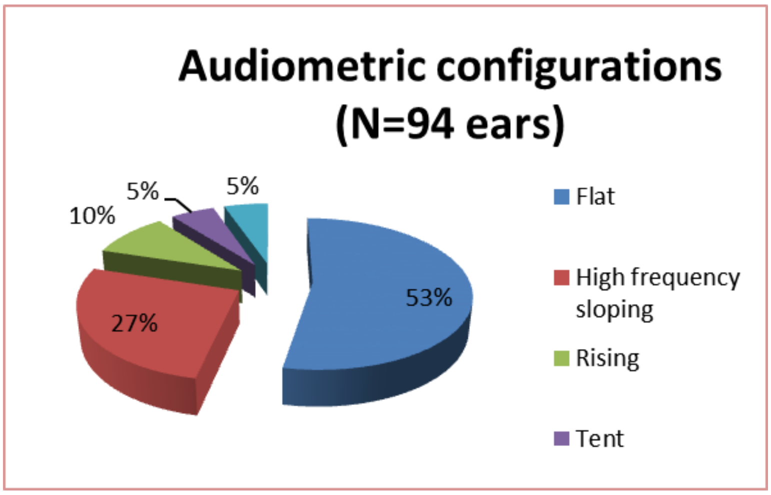

Figure ٥: Different audiometric configurations in children with LVAS

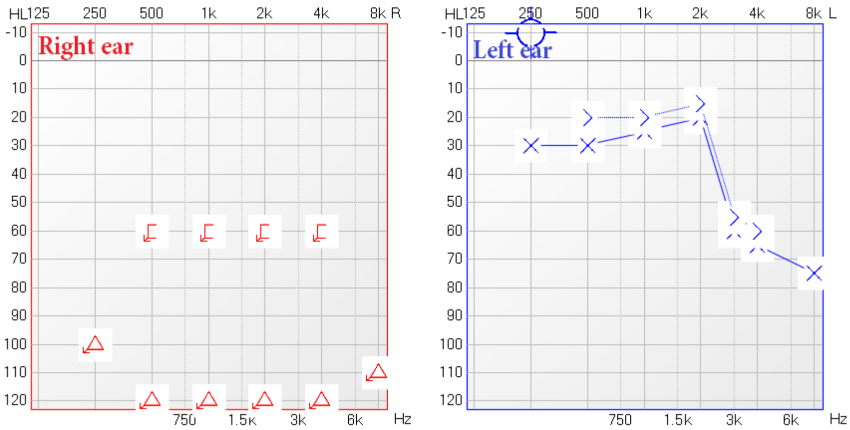

Figure ٦: PTA threshold of female child with LVAS showing asymmetric SNHL Right ear had total hearing loss and left ear had moderate high frequency sloping hearing loss. ABG presents in left ear.

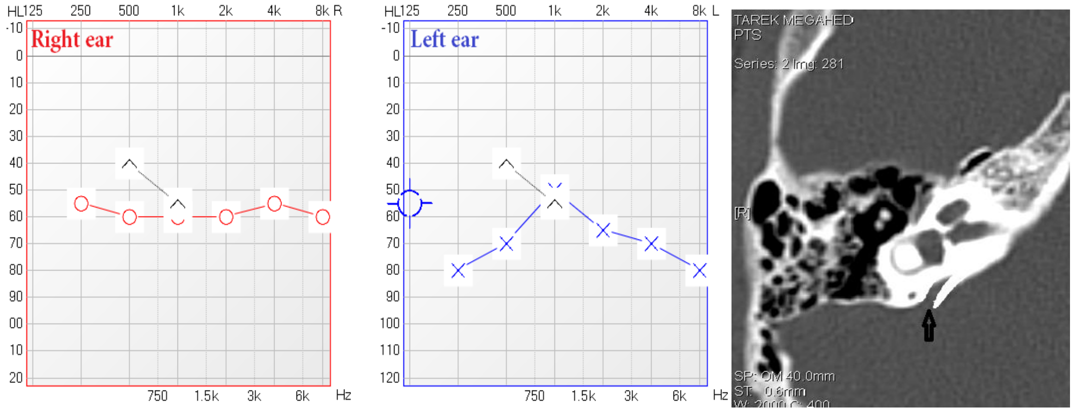

Figure ٧: left panel shows audiogram of child with LVAS. Right panel shows MDCT scan (axial view): an ear with LVA (black hollow arrow) according to Cincinnati criteria, which is considered normal according to Valvassori criterion

Discussion

Large vestibular aqueduct (LVA) is the most common imaging finding in individuals with SNHL dating to infancy or childhood[١٠، ١١]. The earliest description of the LVA was made by Mondini in ١٩٧١[١٢]. However, it was Valvassori and Clemis in ١٩٧٨[٤] who first described the association between LVA and SNHL. Despite such high prevalence and presence of extensive studies, the syndrome is still overlooked and its diagnosis is often missed among radiologists and audiologists. Moreover, there are some points that deserve to be re-addressed as the radiological criteria to diagnose LVA and correlations among the audiological and radiological findings.

Results of the current study agree with Dewan et al.,[١٣] that Cincinnati criterion is more sensitive than Valvassori criterion in diagnosis of LVA and advocate the use of Cincinnati criteria as alternative for the Valvassori criterion.

Performing statistical analysis on VA measurement of the children with LVAS (٦٨ ears) and using the Receiver Operating Characteristics (ROC) curve was done to specify cutoff criteria for diagnosis of LVA in the oblique ٤٥° reformate and to determine the sensitivity and specificity of this criteria. The ROC analysis revealed that cutoff criteria are ٠.٨٧ mm at the midpoint and ١.١٦ mm at the operculum with ١٠٠٪ sensitivity and ١٠٠٪ specificity.

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. . Rehabilitation of children with LVAS and its prognosis relay essentially on other factors as hearing sensitivity, response to hearing aids, and language ability.

Only two factors were significantly correlated with language delay: hearing loss duration and onset of the use of the hearing aids. More delay in language development was found in children with longer hearing loss duration and late onset of hearing aid use relative to the onset of hearing loss.

The overall results show that hearing aid can deliver speech signal within the normal speech spectrum to children with LVAS in more than ٤٠٪ of them. The rest of children may need assistance through assistive listening devices or even cochlear implant.

History of head trauma, even minor trauma, at the hearing loss onset or the onset hearing loss progression is an important factor in the clinical suspicion. Asymmetricity, progression and fluctuation of the hearing loss are common features in children with LVAS. Children with one or more of these features should be suspected for LVAS. The asymmetric feature of the hearing loss is also applied for children diagnosed through ABR. LVAS is considered the first differential diagnosis for infant and young children pressing with asymmetric ABR threshold in the absence of middle ear pathology.

In the absence of known middle ear pathology, presence of air bone gape in ٥٠٠ Hz or at ٥٠٠ and ١٠٠٠ Hz is another important factor to suspect LVAS. Children presenting with hearing loss with air bone gape at ٥٠٠ Hz or at ٥٠٠ and ١٠٠٠ Hz in the presence of intact acoustic reflex can be considered to have LVAS till establishment of the diagnosis through CT scan. Presences of features suggestive for the syndromes that are commonly associated with LVAS are clinical suspects for LVAS. Therefore, children presenting with hearing loss associated with thyroid dysfunction or thyroid enlargement as in Pendred syndrome or with cup-shaped auricle, pre-auricular fistula, neck fistula as in BOR syndrome should be suspected to LVAS.

The results indicate that hearing aids are not enough in rehabilitating children with profound degree of hearing loss. Cochlear implantation is strongly recommended for those children as early as possible to facilitate language development.

Fortunately, studies revealed good prognosis of cochlear implantation in children with LVAS in promoting language development.

Language assessment in children with LVA

For our best knowledge, formal language assessment in children with LVAS is not available in literature. In the current study, language assessment was performed in children with LVAS and results revealed marked delay in the language development in those children. Only ٨.٥٧٪ of children with LVAS had fully developed language while the vast majority (٩١.٤٣٪) had delay in language development. The difference (in years) between the chronological age (CA) and language are (LA) was taken as measure for the delay in language development. Language delay ranged between ٢ months and ١٢.٩ years with a mean delay of ٤.٦ years and SD of ٣.٣ years. Interestingly, there were no statis-tical significant correlations between language delay and factors that are expected to be associated with language delay as audiometric threshold, response to the hearing aid (i.e., aided response was within the normal speech spectrum or not) and language therapy (i.e., children who received vs. children who did not receive language therapy or children that were regular in language therapy vs. those that were irregular in language therapy). Only two factors were significantly correlated with language delay: hearing loss duration and onset of the use of the hearing aids). More delay in language development was found in children with longer hearing loss duration and late onset of hearing aid use relative to the onset of hearing loss.

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