CLINICAL, AUDIOLOGICAL AND RADIOLOGICAL FEATURES OF BILATERAL ENLARGED VESTIBULAR AQUEDUCTS SYNDROME : CASE SERIES

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ABSTRACT

Background: Enlarge Vestibular Aqueducts (EVA) is a vestibular aqueduct dilation bigger than 1 mm which can cause progressive hearing loss. EVA diagnosis regards clinical features, audiological examination, CT Scan and/or MRI. **Objectives**: Describing the knowledge about clinical features, audiological and radiological examinations and management of 3 EVA patients. **Case Report**: Three patients were diagnosed with bilateral progressive hearing loss. The first patient, a 12-year-old boy with a chief complaint of decreased hearing in both ears since 3 years had PTA examination showing Severe MHL (86.25 dB) in the right ear and moderate MHL (55.75 dB) in the left ear. The CT scan showed 2.5 mm dilation in vestibular aqueduct of the right ear and 2.44 mm in the left ear. The second patient, a 12-year-old boy complained of having experienced progressive hearing loss in both ears for 7 years. PTA examination showed moderate Severe SNHL 65 dB in the right ear and 63.75 dB in the left ear and 2.8 mm enlarged aqueduct in the right ear and the left ear. The third patient, an 18-year-old woman had experienced hearing loss for 3 years. PTA examination resulted in moderate Severe SNHL 57.5 dB in the right ear and 56.25 dB in the left ear. The CT scan found aqueductal dilation of 1.5 mm in the right ear and 1.6 mm in the left ear. Three patients were given hearing aids and follow-up examination every 3 months to evaluate the hearing status, progression and effectiveness of hearing aids. hearing aids interventions were found quite effective in 2 patients, and it was regarded less optimal in 1 patient. The three patients were taught about the management of cochlear implants (CI) if hearing aids no longer effective. **Methods**: Relevant literatures were collected from Pubmed database using certain keywords and predetermined inclusion criteria. **Results**: Four articles relevant to this case series were obtained. **Conclusion**: EVA is a disorder of the inner ear that causes progressive sensorineural hearing loss. Correct diagnosis can be made based on careful analysis of the medical history, audiometric examination, CT scan and or MRI in order to determine appropriate management and anticipate progressive symptoms.

Keywords: Enlarge Vestibular Aqueducts (EVA), Hearing Loss, Hearing Aids.

INTRODUCTION

Enlarged Vestibular Aqueducts (EVA) is a condition characterized by the dilation of the vestibular aqueduct, which exceeds a measurement of 1 millimeter. This dilation is typically accompanied by the enlargement of the endolymphatic ducts and sacs. The onset of hearing loss associated with EVA commonly occurs during childhood, and the severity of hearing impairment often worsens over time. Factors such as head trauma, strenuous exercise, or upper respiratory infections are frequently linked to the progression of hearing loss in individuals with EVA. In order to diagnose hearing loss in these cases, healthcare professionals consider a combination of clinical features, audiological examinations, and imaging techniques such as CT scans or MRI. This diagnostic process is particularly important when evaluating children, as they frequently experience progressive unilateral or bilateral hearing loss. Therefore, it is crucial to conduct thorough examinations to determine the specific types of sensorineural deafness present in these individuals.[1, 2]

Enlarged Vestibular Aqueducts (EVA) can result in varying degrees of hearing loss severity among affected individuals. However, it is commonly observed that patients with EVA tend to experience a significant progression of hearing loss, often categorized as severe or even very severe. Diagnostic imaging techniques, such as CT scans, are frequently employed to visualize the enlargement of the vestibular aqueduct on one or both sides of the affected individuals.[3]

Beyond the impact on hearing, patients with EVA may also encounter vestibular disorders, further complicating their overall condition. These additional challenges can manifest as difficulties with balance, spatial orientation, and coordination, which can significantly affect their quality of life.[4]

The medical management of individuals with EVA primarily revolves around addressing their hearing loss based on its degree of severity. One of the common treatment options involves the use of hearing aids, which can help amplify sounds and improve the patient's ability to perceive auditory stimuli. However, in cases where the hearing loss is particularly severe or very severe, more advanced interventions may be necessary.[4]

Cochlear implants (CIs) are often recommended for patients with EVA who experience significant hearing impairment. By utilizing this advanced technology, individuals with severe or very severe hearing loss due to EVA can regain a degree of auditory function and potentially enhance their overall communication abilities.[5]

The decision to pursue cochlear implantation is typically made after a comprehensive evaluation by a team of healthcare professionals, including otolaryngologists, audiologists, and speech-language pathologists. These

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experts carefully assess the patient's medical history, audiological test results, and functional communication abilities to determine the most appropriate course of action.[5]

Furthermore, ongoing follow-up care is crucial for individuals with EVA who have undergone cochlear implantation. Regular audiological assessments, mapping sessions, and speech therapy sessions are typically recommended to optimize the performance and functionality of the cochlear implant, ensuring that the patient continues to receive the maximum benefit from this intervention.[6,7]

In conclusion, Enlarged Vestibular Aqueducts (EVA) can result in a wide range of hearing loss severity, with most patients experiencing significant progression of hearing impairment. Alongside hearing deficits, patients with EVA may also face vestibular disorders, which can further complicate their overall health. The management of EVA primarily revolves around addressing hearing loss through various means, including the use of hearing aids and, in severe cases, cochlear implants. These interventions require careful evaluation, specialized care, and ongoing follow-up to optimize the patient's hearing outcomes and enhance their overall quality of life. [6,7]

EVA generally occurs during childhood that it should be carefully intervened considering the importance of communication in children's development and education. [6]

This case report was written to share knowledge regarding EVA in relation to the use of clinical-based approach and proper management to determine accurate interventions against hearing loss due to EVA

CASE REPORT 1

A 12-year-old male patient was referred to the Ear, Nose, and Throat (ENT) clinic at Wahidin Sudirohusodo Hospital with significant complaints of hearing loss persisting for the past three years. The hearing loss experienced by the patient had been progressively worsening in both ears and was accompanied by a persistent ringing sensation known as tinnitus. Upon conducting an otoscopy examination, no abnormalities were observed in the outer ear or the tympanic membrane, suggesting that the cause of the hearing loss might lie deeper within the auditory system.

To further investigate the extent of the hearing loss, an audiometry examination was conducted. The results revealed a severe Mixed Hearing Loss (MHL) with a threshold of 86.25 decibels in the right ear and a moderate MHL with a threshold of 55.75 decibels in the left ear (as illustrated in Figure 1). These findings indicated that the patient's ability to perceive sound was significantly impaired in both ears, with the right ear experiencing a more pronounced level of hearing loss compared to the left ear.

In order to gain a more comprehensive understanding of the underlying cause of the patient's hearing impairment, a CT scan of the temporal bone was performed. The results of the CT scan revealed the presence of aqueductal dilatation, which is the enlargement of the vestibular aqueduct, measuring 2.5 millimeters in the right ear and 2.4 millimeters in the left ear (as depicted in Figure 2). This finding indicated that the vestibular aqueduct, a crucial component of the inner ear responsible for maintaining balance and hearing functions, was abnormally enlarged in both ears.

The combination of the clinical presentation, audiometry results, and CT scan findings strongly suggested that the patient's hearing loss was associated with Enlarged Vestibular Aqueducts (EVA). EVA is a condition characterized by the dilation of the vestibular aqueduct, often leading to progressive hearing loss over time. In this particular case, the EVA was bilateral, affecting both the right and left ears. The presence of tinnitus further indicated the involvement of the auditory system.

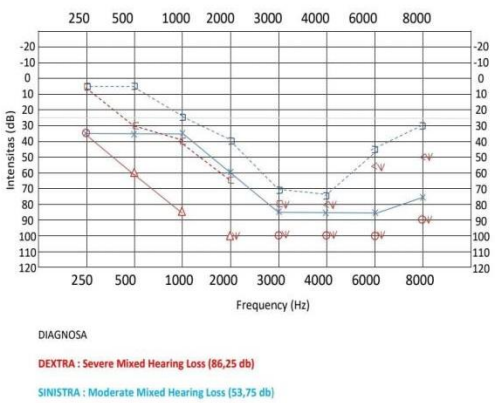

Figure 1. Patient 1 Audiogram

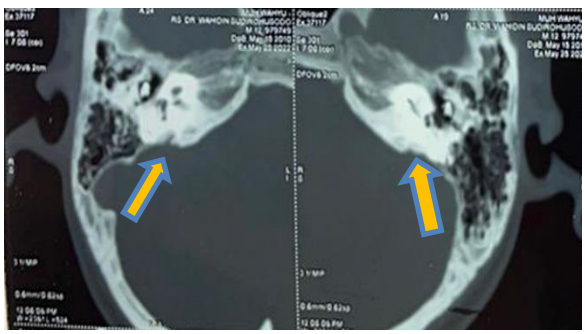

Figure 2. Temporal Bone CT Scan

The patients were given hearing aids hearing aids and were required to have follow up examination every three months for the evaluation of the hearing status, hearing loss progressiveness and the effectiveness of the hearing aids.

CASE REPORT 2

A 12-year-old male patient sought medical assistance at the ENT clinic of Wahidin Sudirohusodo Hospital as he had been grappling with progressively worsening hearing loss for a period of 7 years in both ears. During the examination of the outer ear and tympanic membrane, known as otoscopy, no abnormalities were detected. However, the subsequent audiometry examination yielded concerning

results. The patient exhibited a moderate to severe Sensorineural Hearing Loss (SNHL) with a threshold of 65 decibels in the right ear and 63.75 decibels in the left ear (as depicted in Figure 3). These findings indicated a significant impairment in the patient's ability to perceive and interpret sounds, affecting both ears.

To delve deeper into the root cause of the hearing loss, a CT scan of the temporal bone was conducted. The obtained images revealed enlarged aqueducts measuring 2.8 millimeters on both the right and left sides. These findings highlighted the abnormal dilation of the vestibular aqueduct, a crucial component of the inner ear responsible for maintaining balance and hearing functions.

Based on the clinical presentation, along with the audiometry and CT scan results, it can be inferred that the patient is suffering from a condition known as Enlarged Vestibular Aqueducts (EVA). EVA is characterized by the enlargement of the vestibular aqueduct, often leading to progressive hearing loss over time. In this particular case, both ears were affected, as evidenced by the similar measurements of the enlarged aqueducts on the CT scan images.

It is important to emphasize that EVA is a complex condition that necessitates a comprehensive approach to its management. The presented case underscores the significance of conducting further evaluations and formulating an appropriate treatment plan, taking into account factors such as the severity of the hearing loss, its impact on the patient's daily life, and potential interventions to mitigate the hearing impairment. Regular monitoring and follow-up appointments will be crucial to assess the progression of the hearing loss and determine the most suitable interventions, which may involve the utilization of hearing aids, cochlear implants, or other specialized treatments tailored to meet the specific needs of the individual patient.

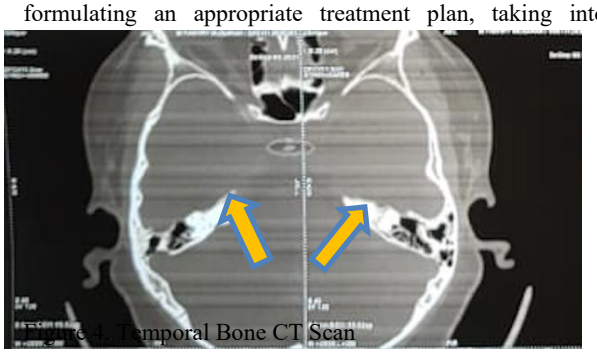

Figure 4. Temporal Bone CT Scan

The hearing aids management was regarded helpful in patient 2. The patient was required to have follow-up examination in 3-months to evaluate the hearing status, progression and effectiveness of hearing aids.

CASE REPORT 3

A young woman, aged 18, presented with a concerning case of progressive hearing loss that had been ongoing for a duration of 3 years, affecting both of her ears. Upon conducting an audiometry examination, the results revealed a moderate Sensorineural Hearing Loss (SNHL) with thresholds of 57.5 decibels in the right ear and 56.25 decibels in the left ear (as illustrated in Figure 5). These findings indicated a noticeable impairment in the patient's ability to hear and comprehend sounds in both ears.

To further investigate the underlying cause of the hearing loss, a CT scan of the temporal bone was performed. The images obtained from the CT scan demonstrated a dilatation of the aqueducts, measuring 1.5 millimeters in the right ear and 1.6 millimeters in the left ear (as depicted in Figure 6). These findings indicated an abnormal enlargement of the vestibular aqueducts, which

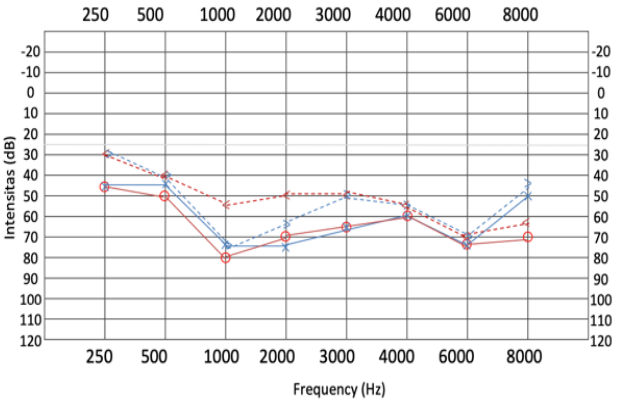

Figure3.Audiogram

are critical structures within the inner ear responsible for maintaining balance and hearing functions.

Considering the clinical presentation, along with the audiometry and CT scan results, it can be inferred that the patient is experiencing a condition known as Enlarged Vestibular Aqueducts (EVA). In this particular case, both ears were affected, as evidenced by the similar measurements of aqueduct dilatation on the CT scan images.

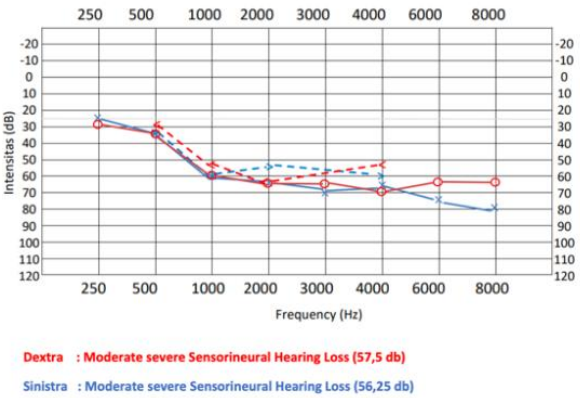

Figure 5. Case 3 Audiogram

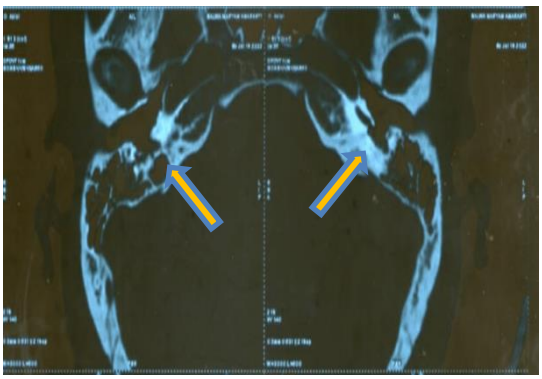

Figure 6. Temporal Bone CT Scan Image

Hearing aids were regarded optimal in improving the hearing ability in both ears.

CLINICAL QUESTION

How is the proper diagnosis and management of *Enlarged Vestibular Aqueducts?*

METHOD

Relevant literatures were collected based on the predetermined keywords and inclusion criteria from Pubmed database on July 20th, 2022. The inclusion criteria are: 1) Keyword match : Enlarged Vestibular Aqueducts, 2) Article published within the last 10 years, 3) full text available. Whereas, article reviews were excluded. Initially, 122 journal articles were obtained to be sorted out. There were 4 literatures relevant to this case report included. (Figure 7).

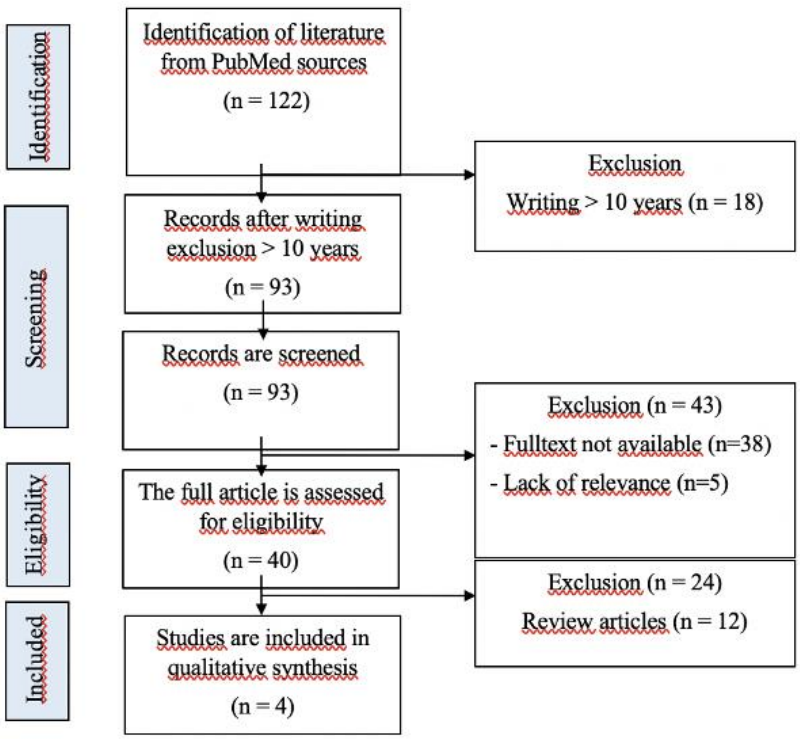

Figure 7. Term search and selection of articles (PRISMA Flowchart)

RESULTS

Four articles were selected as they have high relevance regarding clinical review of the clinical question proposed in this research. The critical assessment is explained in Table 1.

No	Author	Publication Type	Population/ Problem	Intervention/ Index/ Indicator	Comparison	Outome
1.	Cedric pritchett *et al. 2015*	Retrospective Research	55 EVA cases from 1991-2013	Cochlea implantation and speech perception	-	The highest improvement in speech perception occurred in the first 12 months after cochlea implantation.
2.	Ruijie Wang et al. 2021	Retrospective Research	16 EVA patients and 16 typical children	Cochlea Implantation	Comparing the vestibular characteristics in children with EVA undergoing cochlea implantation and children with normal cochlea	Despite the disrupted otolith function, slight improvement was found in the function of three semicircular canals after the implantation

	Population / Patients / Problem	Intervention / Index / Indicator	Comparison	Outcome
Serial cases	3 patients (2021– 2022)	Hearing Aids	-	2 cases showed optimal hearing aids outcome and 1 patients with non-optimal outcome

Table 2. Our Serial Cases

DISCUSSIONS

The onset of hearing loss in the three cases occurred during childhood and adolescence, aligning with the findings of Cedric Pritchett et al., who identified 55 cases of Enlarged Vestibular Aqueducts (EVA) in children. Similarly, Rabindra Pradhananga et al. reported three cases of children aged 2-4 years who were diagnosed with EVA and subsequently received cochlear implants (CI) to manage their hearing loss.[4,8]

In our case report, the three patients experienced a progressive decline in their hearing abilities over a period of 3-7 years. Notably, the second patient demonstrated a significant worsening of hearing loss from a moderate to a severe degree within a span of just 2 years. However, the first and third patients did not undergo early Pure Tone Audiometry (PTA) testing, following the recommendation proposed by Claros P et al.[9]

The CT scan images of the temporal bone revealed varying degrees of vestibular aqueduct dilation in the three patients. The first patient exhibited dilations of 2.5 mm (right ear) and 2.4 mm (left ear). In the second patient, the dilatation was observed on both the right and left sides, measuring 2.8 mm. The third patient presented with dilations of 1.5 mm (right ear) and 1.6 mm (left ear). These findings conform to the criteria set by Ruijie Wang et al., where EVA diagnosis is based on vestibular aqueduct enlargement greater than 1 mm from the midpoint between the posterior cranial fossa and the vestibule of the inner ear. In a retrospective study involving 32 children, 16 of them demonstrated bilateral Enlarged Vestibular Aqueducts on CT scans and MRI.[3,6]

Cedric Pritchett et al. have established specific criteria for diagnosing EVA, considering vestibular aqueduct enlargement of greater than 2 mm in the operculum or greater than 1 mm at the operculum midpoint on axial CT scans.[9]

Cochlear implants (CI) have been widely used in managing patients with EVA. In our case, hearing aids were initially employed as a treatment option. Two patients reported positive outcomes with the use of hearing aids, while one patient did not find them to be optimally effective in improving their hearing. All three patients were informed

about the possibility of receiving cochlear implants. However, the first patient faced socioeconomic constraints that made it difficult to proceed with CI installation. Conversely, the second and third patients expressed a willingness to undergo CI surgery if their hearing aids were no longer effective in improving their hearing abilities.

CONCLUSIONS

EVA is a disorder of the inner ear indicated by progressive hearing loss effect that often occurs in school-age children. Concerning the importance of hearing ability in the children development and communication, comprehensive measures should be considered in making EVA diagnosis in order to determine immediate and accurate interventions.

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