Literature Review

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Cervical vestibular evoked myogenic potential in children and adolescents with enlarged vestibular aqueduct: systematic review

Potencial miogênico evocado vestibular cervical em crianças e adolescentes com aqueduto vestibular alargado: revisão sistemática

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ABSTRACT

Purpose: To gather the parameters found in the cervical vestibular evoked myogenic potential (cVEMP) in children and adolescents with enlarged vestibular aqueduct syndrome (SAVA) and identify the possible changes, when compared to the values found in normal hearing people of the same age group. **Research strategy:** Systematic review registered in the PROSPERO database, prepared through a search in virtual databases, based on the selected keywords. **Selection criteria:** Included scientific articles available in full that reported the evaluation using cVEMP in the 0 and 18 years old group , with a diagnosis of SAVA, without restrictions of language and year of publication; Studies on patients with any disorder other than otoneurological ones and populations outside the proposed age range were excluded. **Results:** 984 records were identified from the search in the databases consulted and 5 articles were selected. In a total of 133 patients who underwent cVEMP, the presence of a response was observed in most cases, with no significant difference in latencies, but with an increase in amplitude and a decrease in cVEMP thresholds. **Conclusion:** The cVEMP test is recommended in the evaluation of children and adolescents with SAVA and the characteristics of increase in amplitude and decrease in thresholds can be used as clinical parameters in the identification of this syndrome, together with the patient's clinical history and imaging exams. However, it is essential to carry out more studies with the cVEMP test, also in children and adolescents with SAVA, to better standardize the values found, in order to make the correct diagnosis.

Keywords: Vestibular evoked myogenic potential; Child; Adolescent; Enlarged vestibular aqueduct; Vestibular evoked myogenic potential cervical

RESUMO

Objetivo: Reunir os parâmetros encontrados no potencial miogênico evocado vestibular cervical (cVEMP) em crianças e adolescentes com síndrome do aqueduto vestibular alargado (SAVA) e identificar as possíveis alterações, quando comparados aos valores encontrados em normo-ouvintes da mesma faixa etária. **Estratégia de pesquisa**: Revisão sistemática cadastrada na base PROSPERO, elaborada por meio de busca nos bancos de dados virtuais, a partir dos unitermos selecionados. **Critérios de seleção:** Incluídos artigos científicos disponíveis na íntegra que relataram a avaliação com o uso do cVEMP na faixa etária entre 0 e 18 anos, com diagnóstico de SAVA, sem restrição de idioma e ano de publicação; excluídos estudos em paciente com algum distúrbio, outras patologias otoneurológicas e população fora da faixa etária estimada. **Resultados:** Foram identificados 984 registros, a partir da pesquisa nas bases de dados consultadas e selecionados 5 artigos. Em um total de 133 pacientes que realizaram o cVEMP, foi observada presença de resposta na maioria dos casos, sem diferença significativa nas latências, mas com aumento na amplitude e diminuição nos limiares do cVEMP. **Conclusão:** O teste cVEMP é recomendado na avaliação de crianças e adolescentes com SAVA e as características de aumento na amplitude e diminuição nos limiares podem ser utilizadas como parâmetros clínicos na identificação da referida síndrome, juntamente com a história clínica do paciente e os exames de imagem. No entanto, é imprescindível a realização de mais estudos com o exame cVEMP, ainda, em crianças e adolescentes com SAVA, para a melhor padronização dos valores encontrados, a fim de efetivar o diagnóstico correto.

Palavras-chave: Potencial miogênico evocado vestibular; Criança; Adolescente; Aqueduto vestibular alargado; Potencial miogênico evocado vestibular cervical

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INTRODUCTION

The vestibular aqueduct (VA) is a bony canal that extends from the medial wall of the vestibule to an opening on the posterior surface of the petrous portion of the temporal bone. The endolymphatic duct and sac travel through it, filled with endolymph[1]. Enlarged vestibular aqueduct syndrome (EVA) is characterized by an increase in the connection between the endolymphatic sac and the vestibule. It is considered abnormal when there is a distance between the organs greater than 1.5 mm[2].

The clinical picture of EVA, in most situations, begins in childhood and has varieties in terms of its clinical characteristics. They may present hearing loss of different degrees (from moderate to severe), onsets (sudden or progressive), or types (mixed loss with a greater air-bone gap at low frequencies or sensorineural), associated or not with vertigo[3,4]. Therefore, early detection becomes important.

While some structures of the middle and inner ear are fully developed at birth, the vestibular aqueduct and endolymphatic sac are immature and small. Once the posterior cranial fossa expands, the VA and endolymphatic sac rapidly increase in size and reach maturity around 4 years of age[3]. Among the possible causes of the increase in this channel, interrupted development in embryonic life and/or poor postnatal development in early childhood are reported[5]. The diagnosis is made through imaging tests of the temporal bone, such as computed tomography and magnetic resonance imaging[6].

The endolymphatic duct extends from the junction of the utricular and saccular ducts, through the vestibular aqueduct, to end as an expanded portion that is the endolymphatic sac. The latter has the function of balancing the pressure between the vestibular system and the central nervous system, in addition to absorbing endolymph[7]. Some authors have identified a direct and proportional relationship between VA area and volume, using graphic reconstruction techniques. It is concluded that, if there is an enlargement of the VA, there is also an enlargement, at least, of the rough portion of the endolymphatic sac[8]. As a result of this ductal enlargement, there may be changes in the homeostasis of the endolymphatic circulation, with consequent damage to the cochlear neuroepithelium[9]. To evaluate this possible change in the functioning of the inner ear, which occurs with VA enlargement, the cervical vestibular evoked myogenic potential (cVEMP) is used.

cVEMP stands out among the possible tests adopted in the otoneurological evaluation of patients with enlarged vestibular aqueducts[10]. These are short-latency inhibitory response evoked potentials, recorded ipsilaterally, from the sternocleidomastoid (SCM), and provide information about the function of the saccule and inferior vestibular nerve[11]. Therefore, it is applied as a complementary exam in otoneurological disorders and the assessment of vertigo[12].

The cVEMP test is a complementary exam that provides information on the function of the saccule and lower portion of the vestibular nerve, regions that are not evaluated in traditional vestibular exams. Therefore, its application, together with other otoneurological tests proves to be effective in diagnosing vestibular pathologies. Furthermore, its use has numerous benefits, such as the fact that it is an objective, reliable, non-invasive, low-cost, easy-to-perform, fast, and discomfort-free examination for the EVA patient[12].

However, there is a lack of literature on otoneurological assessment in childhood and adolescence, which leads to difficulties in evidence-based clinical practice. In view of this, the present study was designed to gather the parameters found in the cVEMP in these age groups with enlarged vestibular aqueducts and identify possible changes. They are compared to the values found in normal-hearing individuals, to identify a pattern of responses and provide reference values that help diagnose such changes.

PURPOSE

To gather the parameters found in cVEMP in children and adolescents with enlarged vestibular aqueduct syndrome (EVA) and identify possible changes, when compared to values found in normal-hearing individuals in the same age group.

RESEARCH STRATEGY

This systematic review was carried out according to data from the PRISMA checklist (Preferred Reporting Items for Systematic Reviews and Meta-Analyses)[13]. Previously, a search was carried out in the Cochrane and PROSPERO databases, to be aware of the previous existence of a systematic review on the subject. With the negative response, the research protocol was prepared and registered on the PROSPERO platform under number CRD4202341302[14].

The research was carried out using the PICO strategy (P = Patients; I = Intervention; C = Comparison; O = Outcome), to prepare the following clinical question: "What parameters (O) are found in the cervical vestibular evoked myogenic potential (cVEMP) (I) in children and adolescents with enlarged vestibular aqueduct (P), compared to normal-hearing children (C)?".

SELECTION CRITERIA

For the selection of studies, the following inclusion criteria were defined: scientific articles available in full, which reported the evaluation using cervical vestibular evoked myogenic potential in the population aged between 0 and 18 years, diagnosed with enlarged vestibular aqueduct syndrome (EVA), with no restrictions on language and year of publication.

The exclusion criteria were: studies on patients with some disorder (physical/neurological/cognitive/orthopedic), with other otoneurological pathologies, population outside the estimated age range, case reports, editorials, monographs, books, chapters, and event summaries.

In cases where there was a discrepancy between the two reviewers, a third reviewer was consulted for the final review. The studies were collected in the following databases: Science Electronic Library Online (SciELO), PubMed, Latin American and Caribbean Literature in Health Sciences (LILACS, via VHL), Scopus, Web of Science, and Science Direct. The search in "gray literature" took place on the Google Scholar, BASE, and CORE platforms in March 2023.

The descriptors were selected using Health Sciences Descriptors (DeCS) and PubMed (MeSH). The search was performed using the Boolean operators "AND" and "OR" for a comprehensive search strategy. The keywords used were: "vestibular evoked myogenic potential", "child", "adolescent", "large vestibular aqueduct", "vestibular evoked myogenic potential cervical" and their respective terms in Portuguese.

DATA ANALYSIS

The study selection process occurred in two stages. In the first stage, two reviewers independently analyzed the titles and abstracts. Studies that did not meet the objectives of this review were excluded. In the next stage, the previously selected studies were subjected to full-text analysis, to verify whether the contents met the eligibility criteria and whether they answered the study's guiding question.

The analysis was carried out qualitatively since the sample was small and the methods adopted between the studies were heterogeneous. The selection was carried out by two reviewers, independently. The risk of bias was analyzed using the manual from the Joanna Briggs Institute – JBI[15], being considered high when the study obtained up to 49% of a "yes" score, medium when the study obtained a "yes" score of 50% to 69%, and low when the study obtained more than a 70% "yes" score.

RESULTS

Initially, 984 studies were identified in the 9 databases. Of these, 127 studies were duplicates and were therefore removed. After removing duplicate studies, 857 were analyzed using the title and abstract. After analyzing these, 825 studies were excluded, leaving 32 possibly eligible studies, which were then subjected to full reading. Of these, 27 studies did not meet the eligibility criteria and were excluded, therefore, 5 studies (0.51% of the initial sample total) were selected. The search and selection process for studies in this review is presented in a flowchart, as proposed by PRISMA (Figure 1).

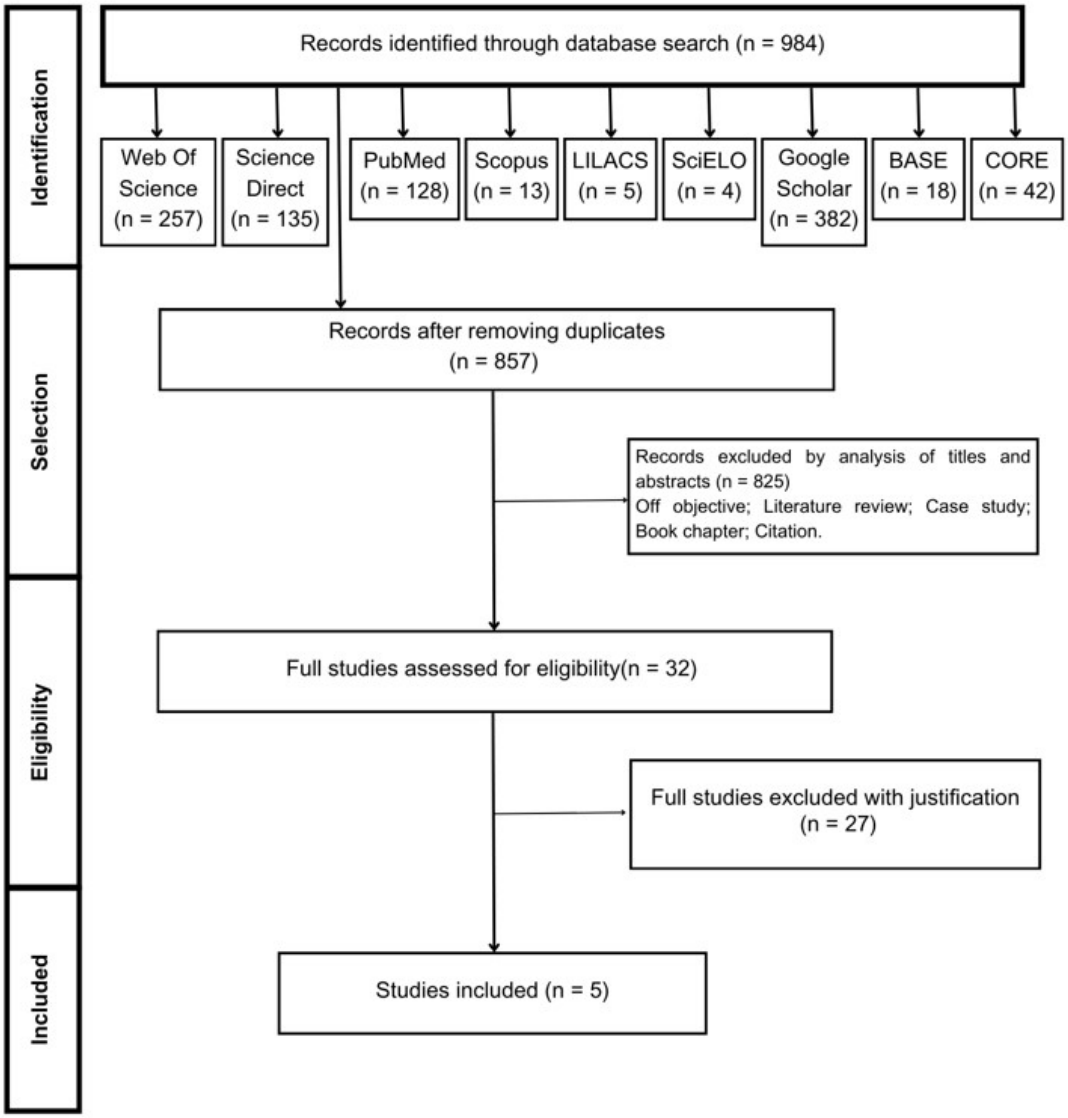

Figure 1. Bibliographic search flowchart and selection criteria adapted from Preferred Reporting Items for Systematic Reviews and Meta-Analyses

This systematic review included 5 studies[2,5,6,16,17] that mentioned the characteristics of the desired cervical vestibular evoked myogenic potential (cVEMP) parameters (P1 and N1 latencies and P1-N1 amplitude) within the pre-defined age class of patients with EVA.

Of the 5 studies included, 2[2,17] had numerical values of the investigated parameters. The other 3 studies[5,6,16] only presented variations in responses when analyzing the results. In all of them, a comparison was observed between patients with EVA and normal hearing patients.

Tables 1 and 2 exhibit data related to the identification of the selected studies, characteristics of the sample, description of the exam, stimulation and recording method, and cVEMP wave analysis process. They also show the results of the cVEMP parameters obtained in each study from the sample used in patients with EVA, compared to subjects without alteration of the aqueduct.

The values of P1/N1 latency (ms) and amplitude (μV), obtained when performing the exam with tone burst stimuli, at a frequency of 500 Hz and intensity of 100 dB nHL, reported in the observed studies, are shown in Table 3.

DISCUSSION

In the selected studies, 173 children and adolescents with the aforementioned syndrome were evaluated, of which 133 were subjected to cVEMP examination, with only one being excluded for not being within the pre-defined age range for this analysis. Among those with the syndrome, an age range between 3 and 16 years was observed[5,6,16,17], with a predominance of females (46% men and 54% women). In the literature, women are prevalent and there is no explanation for this event. However, this difference does not extend to the occurrence of hearing losses[18].

The majority of those with the syndrome presented sensorineural hearing loss, ranging from mild to profound, and with the presence of associated vestibular symptoms, such as vertigo, imbalance, or delayed neuropsychomotor development[17]. Concerning vestibular manifestations, these were characterized by a lower incidence and late-onset when compared to cochlear symptoms, which represents a system that is less vulnerable to mechanical or chemical damage[19].

Furthermore, possible causes for the occurrence of hearing loss in patients with enlarged VA include increased pressure in the endolymphatic fluid that causes damage to hair cells in the cochlea, a change considered similar to Ménière's disease, cochlear dysplasia (Mondini) and genetic mutations[6]. They may be associated with other vestibular anomalies, such as increased isthmus diameter and enlarged dysplastic vestibule[20].

It was possible to observe that the syndrome can occur unilaterally or bilaterally. In the selected studies[5,6,16], there was a predominance of bilateral diagnosis (63%), an occurrence that can be explained by a probable genetic inheritance. In some cases, it is related to still unknown etiological factors[1]. However, some authors observed a relationship between enlarged VA and the SLC26A4 gene mutation, especially in bilateral cases, concluding a possible difference in the origin of the enlargement of this aqueduct between unilateral and bilateral cases[21-23].

In cases of unilateral EVA, there was no significant difference in the predominance of occurrence in one ear in relation to the other (19 occurrences only in the right ear and 20 only in the left ear). It was observed that in some cases the increase in the VA was not isolated, but rather associated with other cochlear malformations, such as Mondini dysplasia and dilated vestibule, in addition to a history of tympanostomy with tube placement[6].

Normal-hearing children presented the following mean values in the cVEMP analysis parameters, when performed with 500 Hz tone burst stimuli and with intensities between 95 and 130 dB nHL: P1 latency between 11.9 and 16.13 ms (standard deviation [SD] between 0.9 and 2.12 ms); N1 latency between 17.6 and 24.78 ms (SD between 1.4 and 2.77 ms) and P1-N1 amplitude of 6.0 μV (± 1.2). The adolescents presented the following mean values: P1 latency between 12.7 and 17.26 ms (SD between 12.7 and 24.78 ms) and P1-N1 amplitude between 1.65 and 6.3 μV (SD between 0.65 and 1.6 μV), in accordance with the literature[12,24-26].

The latencies of the cVEMP components (P1, N1) depend greatly on the stimulus design (click or tone burst) and the applied frequency[27-29]. The surface electrodes measure a biphasic potential, labeled PI and NII (or P13 and N23) for positive and negative deflection, during tonic muscle contraction[30].

In relation to the selected studies, standardization was identified in the performance of cVEMP with the tone burst type stimulus to record the potential with a predominance of the frequency of 500 Hz. The low-frequency pattern results from more homogeneous responses, higher response rates, and amplitudes. Therefore, the frequency of 500 Hz is more effective[31].

Furthermore, studies show that cVEMP presents better responses to low-frequency stimuli. Tone burst stimuli at frequencies equal to or lower than 1,000 Hz demonstrate better wave definition and greater amplitude of responses than those evoked by click stimuli[32-34], with the frequency of 500 Hz being the most used clinically[32,35].

Regarding the stimulation mode, all studies used airway stimulation. This allows the standardization of findings, since airway stimuli are specific in stimulating saccular responses, and are more clinically used for capturing cVEMP than bone or galvanic stimuli[36].

cVEMP responses may be absent or present, characterizing whether or not there is a functional change in the otolith organs and/or the vestibular nerve. Of the total number of patients analyzed, only nine presented an absence of waves[2,5,6]. Of these, in the clinical history, there were reports of sudden hearing loss and complaints, such as dizziness, before the exam[5,6]. Thus, sudden losses with complaints of dizziness may be indicative of EVA, which makes the recommendation of cVEMP important. However, due to the lack of responses occurring in a small number of patients with EVA, it can be inferred that this is not a characteristic directly linked to the syndrome, but rather to other factors that may be associated with it, such as, for example, a saccular functional loss[5,6].

Regarding latency, the time that elapses from acoustic stimulation until the appearance of the most positive or negative value of the waves[37], most studies did not report changes in this parameter, except for one study[2], which it related to the fact that EVA is characterized by a peripheral alteration. The study, when analyzing a group of adults with the syndrome, observed higher N1 and lower P1 latencies, compared to their healthy controls, different from the comparison that occurred between children, when they did not find significant differences within their age group[2].

Table 1. Description of study characteristics

ID	Sample	Exam description	Stimuli and records	Wave analysis
Zhou et al.[6]	54 patients (22 male and 32 female) with EVA, age range between 2 and 16 years and an average of 7 years.	Not reported by the authors.	The acoustic stimulus used was a 500 Hz tone burst, intensity of 90 dB nHL, through ER-3A insert headphones.	All patients with EVA had hearing loss, although the degree and configuration of hearing loss varied considerably. The VEMP test was performed on 14 patients with EVA; In all ears with the syndrome, except one, increased test responses were found: decreased thresholds in ears with EVA in relation to normal and increased amplitude.
Zhou & Gopen[5]	A total of 25 cases (37 ears), 13 with unilateral EVA and 12 with bilateral EVA, were included for analysis. The average age of these patients was 8.2 years.	Sitting, the patient was asked to turn the head to the side contralateral to the tested ear.	Electrodes were placed on the SCM muscle. VEMP responses were obtained using acoustic tone burst stimuli of 250 and 500 Hz, through insert headphones. The lowest stimulus intensity at which a clear and repeatable biphasic wave (P1/N1) was observed was recorded as the VEMP threshold. If no repeatable response was found, the VEMP was considered absent. VEMP amplitude and P1/N1 latencies were measured at the stimulus level of 90 dB nHL.	Characteristics of VEMP in EVA include lower thresholds and higher amplitudes. The abnormally low VEMP threshold suggested a "third window" effect in this pathological condition. The unilateral absence of VEMP responses in children with EVA may be indicative of peripheral vestibular dysfunction. VEMP testing is recommended in the evaluation of children with EVA.
Yang et al.[16]	27 patients diagnosed with EVA, 37% with unilateral involvement and 63% bilateral. The age range varied from 3 to 12 years old.	To perform the VEMP, the SmartEP analyzer from Intelligent Hearing Systems (Miami, FL) was used. Children were instructed to contract the SCM by rotating the head in a sitting or lying supine position at a 20-degree angle and contract the SCM by elevating the head.	The surface electrodes were placed in the following positions: active on the SCM muscle, reference on the sternoclavicular junction and grounding on the forehead. The acoustic stimulus used was a 500 Hz tone burst, intensity of 107 dB nHL, through an ER-3A insert earphone at a rate of 5.1.	15% of patients had abnormal cVEMP results. There was no statistically significant correlation between absolute cVEMP thresholds and amplitudes and age, clinical symptoms, audiometric or radiographic findings. All analyses were performed using SAS for Windows 9.3 (SAS Institute Inc., Cary, NC).
Zhang et al.[2]	29 patients diagnosed with enlarged vestibular aqueduct (EVA) syndrome, 23 children (3 to 12 years old) and 6 adults (15 to 33 years old)	To perform the VEMP, the Otometrics (Taastrup, Denmark) ICS Chartr EP analyzer was used. The patient remained lying in the supine position and was instructed to raise their head 30° when listening to the sound in the headphones.	The surface electrodes were placed in the following positions: active on the SCM muscle bilaterally, reference above the sternoclavicular joint, and grounding on the midline of the forehead. The acoustic stimulus used was a 500 Hz tone burst. VEMP thresholds were recorded as lower stimulus intensities that could cause a repeatable and clear biphasic wave.	VEMP parameters were recorded from the elicited short-latency reproducible biphasic waveform. The Pearson chi-square test or Fisher's exact test were used to compare response rates. The t-test was used to compare thresholds, amplitudes, P1 and N1 latencies and latencies between peaks. Collected data were analyzed with IBM SPSS Statistics version 19.0.0 (IBM SPSS Statistics, Armonk, NY).
Liu et al.[17]	54 children, 44 with enlarged vestibular aqueduct syndrome as the study group and 10 normal-hearing children as the control group. Patients over 14 years of age were excluded from the study.	To perform the cVEMP, the Eclipse equipment (Interacoustics A/S, Denmark) was used. The cVEMP test was performed with patients sitting and instructed to turn their head to the side contralateral to the stimulus, both via air and bone conduction .	The surface electrodes were placed in the following positions: active on the manubrium of the sternum, reference on the superior position of the sternocleidomastoid muscle (SCM) and grounding on the forehead. Air conduction: the type of stimulus used was 500 Hz tone burst and the stimulation rate was 5.1/s, with an initial intensity of 100 dB nHL with steps of 5 or 10 dB to elicit the threshold. The high-pass filter was 10 Hz and the low-pass filter was 750 Hz. The recording window was defined from 20 to 80 ms. 200 stimuli were used and the P wave was set to the upward direction. Bone conduction: B-81 bone transducer, mastoid placement. Initial stimulation intensity of 70 dB nHL and adjusted to 5 or 10 dB.	cVEMP thresholds by air conduction were categorized as "normal" if they were 70 to 80 dB, as "low" if they were 65 dB or less, and as "elevated" if they were greater than 90 dB nHL. Bone conduction cVEMP thresholds were categorized as "low" if they were 25 dB nHL or less, and "high" if they were equal to or greater than 45 dB nHL. The Wilcoxon test was applied to analyze the difference in VEMP parameters (P1, N1, P1-N1 latency, amplitude and threshold). The analyses were performed using SPSS 17.0 software (IBM, New York, USA, 2020).

Subtitle: ID = identification; SCM = Sternocleidomastoid; EVA = Enlarged vestibular aqueduct syndrome; VEMP = Vestibular evoked myogenic potential; cVEMP = Cervical vestibular evoked myogenic potential; dB nHL = decibels normalized hearing level; Hz = Hertz

Table 2. Results of cervical vestibular evoked myogenic potential parameters in selected studies

ID	Sample	Age	Response	Thresholds	Amplitude	P1/N1 Latency
Zhou et al.[6]	14 patients with EVA	2 to 16 years old	Present in 93% (13/14)	Low in 93% (13/14)	Increase	Not reported in the study
Zhou & Gopen[5]	24 patients with EVA	3 to 16 years old	Present in 92% (22/24)	Low in 92% (22/24)	Increase	No significant difference
Yang et al.[16]	27 patients with EVA	3 to 12 years old	Present in 85%	No significant difference	No significant difference	No significant difference
Zhang et al.[2]	23 patients with EVA	3 to 12 years old	Present in 86.95%	No significant difference	No significant difference	No significant difference
Liu et al.[17]	44 patients with EVA	Not reported in the study	Not reported in the study	Low in 4.5% (2/44)	Increase	No significant difference

Subtitle: ID = Identification; EVA = Enlarged vestibular aqueduct syndrome

Table 3. Values of P1 and N1 latencies and P1-N1 amplitude via air conduction

ID	Group	P1 latency (ms)	N1 latency (ms)	Amplitude (μV)
Zhang et al.[2]	Control	15.05 ms (± 0.96)	21.68 ms (± 1.33)	379.84 μV (± 178.69)
	Test	14.60 ms (± 1.86)	20.94 ms (± 2.47)	344.57 μV (± 210.33)
Liu et al.[17]	Control	12.74 ms (± 0.94)	21.43 ms (± 1.20)	70.53 μV (± 18.75)
	Test	12.86 ms (± 0.92)	21.96 ms (± 2.21)	98.11 μV (± 51.65)

Subtitle: ms = milliseconds; μV = microvolts; ± = standard deviation

A possible limitation for comparing this result may be the lack of mention, by the selected studies, of the degree of VA of the patients tested, which can vary from I to V[24].

In the analysis of cVEMP amplitude, an increase in this parameter was observed in three studies[5,6,17], while in two, there was no significant difference[2,16]. Some authors characterize EVA as a third window injury[5], which refers to an additional opening for the inner ear. Thus, it is expected that there will be an increase in cell stimulation, since there is a decrease in the impedance of the tympanic-vestibular system, therefore requiring lower intensities to mediate cVEMP responses[2]. There is also the possibility of pneumatization of the temporal bone, given that under these conditions the loss is mostly sensorineural and bilateral[38]. However, this situation was not mentioned in the selected studies.

Regarding the threshold, it was possible to identify three studies with cVEMP findings with reduced values[5,6,17] and two studies without significant differences[2,16]. In these last two cases, most likely due to the sample size, which was smaller compared to other studies.

Thus, when sound pressure is transmitted to the vestibule of children with EVA, due to the possibility of deviation of the third window, the vibration received by the saccule and utricle is greater than in children without the syndrome. Therefore, the cVEMP amplitude of children with EVA, under the same stimulation intensity, is greater than in normal people, and the lower stimulation intensity can elicit cVEMP[17], that is, lower intensities are needed to record waves in the cVEMP due to possible hyperstimulation of the system.

Finally, as limitations found in this study, it was possible to observe a reduced number of findings in the literature that address the values of each cVEMP parameter, making it not possible to carry out a quantitative study within the age range of children and adolescents with EVA. Therefore, new studies, with a larger number of participants, should be conducted to acquire more detailed information on cVEMP in patients with this syndrome.

CONCLUSION

cVEMP responses in patients aged between 3 and 16 years, diagnosed with EVA, showed increased amplitudes and decreased thresholds, suggestive of a third window lesion in this condition.

The cVEMP test is recommended in the evaluation of children and adolescents with EVA and the characteristics highlighted in this study can be used as clinical parameters in identifying the syndrome, together with the patient's clinical history and imaging exams. However, it is still necessary to carry out more studies using the cVEMP test in children and adolescents with enlarged VA, to better standardize the values found and early detection of the disease.

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