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Investigation of Third Window Symptoms in Patients With Enlarged Vestibular Aqueduct: A Pilot Study

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Abstract

Objective. To assess if patients with enlarged vestibular aqueduct (EVA) experience symptoms characteristic of other third mobile window disorders such as superior semicircular canal dehiscence syndrome (SCDS).

Study design. Cross-sectional study.

Setting. Tertiary care center.

Methods. Adult and pediatric patients with EVA were screened. Patients with additional middle or inner ear pathology were excluded. The included patients and parents of pediatric patients were asked to complete a survey regarding symptoms, including autophony, hyperacusis, and sound- and pressure induced dizziness.

Results. Of the 121 patients who met inclusion criteria, 36 patients and parents completed the questionnaire (15 children and 21 adults). Adult EVA patients appeared to suffer from similar rates of hyperacusis (67%), autophony (52%), pressure induced dizziness (38%), ear fullness (62%), and spontaneous dizziness or vertigo (67%) compared to reported rates among SCDS patients. Children with EVA also appeared to have similar rates of hyperacusis (67%), ear fullness (47%), and spontaneous dizziness or vertigo (40%), while pressure induced dizziness (13%) and autophony (20%) seem less common in children and sound induced dizziness appears less common in both children and adults (13% and 14% respectively).

Conclusion. This is the first study investigating the prevalence of typical third-window symptoms in patients with EVA. We found an overlap in the prevalence of several specific and nonspecific third window symptoms for patients with EVA and SCDS.

Keywords

enlarged vestibular aqueduct, symptoms, third window

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The vestibular aqueduct connects the membranous labyrinth to the endolymphatic sac and duct, which are thought to play a role in the absorption and secretion of endolymph.[1-3] Enlargement of the vestibular aqueduct was first described as part of the Mondini malformation, which also includes incomplete partitioning of the cochlea and an enlarged vestibule.[4] Enlarged vestibular aqueduct (EVA) syndrome was later described by Valvassori and Clemis in 1978 in association with hearing loss and vestibular impairment. EVA occurs both with and without other inner ear abnormalities and as part of other syndromes (including Pendred, branchio-oto-renal, and Waardenburg syndromes) or as an isolated abnormality.[5-8] EVA is diagnosed using computer tomography (CT) or magnetic resonance imaging (MRI) of the temporal bone, and several radiographic size criteria have been proposed.[5,9-15] EVA is the most commonly found radiographic abnormality in children with hearing loss, with a prevalence of 1.4% among children with SNHL.[16]

EVA is considered a third mobile window disorder. The term "third window" was first introduced in the 1950s and describes paths of abnormal pressure transmission within the inner ear.[17] The oval and round windows are the first and second inner ear windows. In a physiologic two-window system, oval window motion creates a pressure wave in the inner ear and equal motion of the round window. Although additional small openings

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into the inner ear occur at the cochlear aqueduct, vestibular aqueduct, and foramina that allow passage of blood vessels and nerves, these are not thought to transmit pressure adequately to alter inner ear mechanics.[18-20] However, several pathologic third window conditions have been described, including superior, posterior, and lateral semicircular canal dehiscence syndrome and enlarged vestibular aqueduct syndrome. In the presence of a pathologic third window, the energy created by oval window motion follows the path of least resistance. It is partially shunted towards the third window, leading to a decreased pressure difference across the basilar membrane in the cochlea and increased energy transmission to the vestibular organs.[21] Auditory and vestibular symptoms and testing abnormalities have been described in these third window syndromes. Superior semicircular canal dehiscence syndrome (SCDS) is a well-studied third window syndrome. SCDS patients suffer from a specific set of symptoms, including autophony, hyperacusis, sound- and pressure-induced vertigo, as well as abnormalities on audiometric (low-frequency air-bone gaps [ABGs]) and vestibular testing (low thresholds and high amplitudes on vestibular evoked myogenic potential [VEMP] tests).[22,23]

Symptomatology in EVA is less well-studied. For example, a review of 18 studies and 475 patients reports that vestibular symptoms were present in 0% to 100% of patients and included vertigo, imbalance, and gross motor delays.[24] The existing literature does not provide further details about the nature of these symptoms and whether patients suffer from symptoms seen in other third-window syndromes such as SCDS. There are reports of patients with EVA having low-frequency ABGs similar to SCDS patients.[24] Although few studies have reported on VEMP testing in EVA patients, low VEMP thresholds or high amplitudes as seen in SCDS have been described, with one study reporting abnormally low cervical VEMP (cVEMP) thresholds in 92% of pediatric EVA ears.[25-27] In addition, diminished response to caloric testing has also been described in patients with EVA, while video head impulse tests are typically normal.[28]

This pilot study aims to assess the nature and prevalence of audiologic and vestibular symptoms, including symptoms specific to other third-window syndromes, among patients with EVA.

Methods

Approval for this study was obtained from the Institutional Review Board at Johns Hopkins University (IRB00371611). Patients with EVA previously seen at our institution between January 1, 2002 and December 31, 2023 were identified by searching our electronic medical record system. The following search terms and ICD codes were used: "enlarged vestibular aqueduct" and "dilation of vestibular aqueduct" (no associated ICD-10 code), "anomalies of inner ear" (ICD-9 744.05), and "congenital anomaly of inner ear" (ICD-10 Q16.5). Charts of identified patients were then screened, and patients of all ages who were found to have unilateral or bilateral EVA based on their otolaryngologist's clinic note or imaging report were included. Exclusion criteria were absence of EVA, presence of other inner ear malformations (including cochlear malformations), presence of other otologic disease (including cholesteatoma, otosclerosis, vestibular neuritis, Meniere's disease, vestibular schwannoma, superior semicircular canal dehiscence syndrome), patients without contact information, patients whose preferred language is not English or American Sign Language, and patients who the providers on our IRB did not see.

Data collection included age, sex, race/ethnicity, insurance type (private, public, military, or none), EVA sidedness, presence of comorbidities (including syndromes, migraine, autism, developmental delay, and other), history of cochlear implantation, and audiometric findings.

Adult patients (≥18 years old) and parents of pediatric patients (<18 years old) were contacted and asked to fill out a survey (Supplemental File SA, available online) regarding their symptoms after informed consent was obtained. The survey included questions regarding the presence or absence of the following symptoms: subjective hearing loss, aural fullness, autophony, hyperacusis, tinnitus, pulsatile tinnitus, vertigo, poor balance, sound-, pressure-, and exercise induced dizziness, and nystagmus. Patients with vertigo were asked to categorize symptoms as constant or episodic and to indicate the frequency and duration of episodes if present. Patients were asked if they had a history of head trauma resulting in dizziness or hearing loss, concern for gross motor function or balance, if they considered themselves to be clumsy, and if they had received physical therapy for balance issues. Adult patients were asked to complete the autophony index and the questionnaire on hypersensitivity to sound. The autophony index is a 26-item questionnaire asking the patient to rate how bothersome their autophony symptoms are on a scale from 0 (*not at all*) to 4 (*almost all of the time*), resulting in a total score ranging from 0 to 104.[29] The questionnaire on hypersensitivity to sound is a 15-item questionnaire asking patients to rate statements on distress related to hyperacusis on a scale from 0 (*never*) to 3 (*always*), resulting in a total score ranging from 0 to 45. To determine the level of distress hyperacusis causes the total score has been categorized into light (0-9), moderate (10-15), serious (16-23), and severe (24-45). Patients are also asked to rate their hypersensitivity to sound from 0 (*no problem*) to 10 (*severe problem*).[30,31] Parents of pediatric patients were asked if their child ever suffered from head tilt with or without vomiting as a baby or toddler and the age at which they achieved developmental milestones. Independent sitting later than 9 months, independent standing later than 17 months, independent walking later

than 22 months, and riding a bike on 2 wheels later than 7 years were considered gross motor delays.

Statistical Analyses

Fisher's exact tests were used to compare demographics for the group of patients who met inclusion criteria and the patients who responded to the survey, to compare the prevalence of auditory and vestibular symptoms in adults versus children and to compare symptoms between patients with and without cochlear implants. Bonferroni adjustments were used to correct for multiple comparisons. A $P < .005$ ($= 0.05/10$) was considered statistically significant for demographics. For auditory symptoms, a $P < 0.008$ ($= 0.05/6$) and for vestibular symptoms, a $P < .007$ ($= 0.05/7$) were considered statistically significant.

Results

Results Among EVA Cohort

Demographics

One hundred twenty-one patients (77 adults and 44 children) met the inclusion criteria. Most patients (73%) had bilateral EVA, while 11% had left-sided and 16% had right-sided EVA. The average age was 25 years (range 2-75; adult $u = 32$, range 18-75; pediatric $u = 12$, range 2-17), and 42% were male. The majority of patients were white (73%), non-Hispanic (84%), and had private insurance (64%; **Table 1**). Syndromic diagnoses included type 2 Usher syndrome (1 adult), Pendred syndrome (1 child and 1 adult), and Turner syndrome (1 child). However, the records for most patients did not mention genetic evaluation and testing. Comorbidities included migraine (2 children and 19 adults), autism (1 adult), and developmental delay (4 children and 3 adults).

The majority of patients had sensorineural hearing loss (69.5%) or mixed hearing loss (26%), while the presence of a pure conductive loss (1.5%) and the absence of hearing loss (1.5%) were rare. Progressive or fluctuating hearing loss occurred in 58% of patients. Over half of all patients (56%) underwent cochlear implantation (**Table 1**).

Results Among Surveyed Cohort

Demographics

Thirty-six patients and parents completed the questionnaire (29.8%), including 15 children (average age 11 years, range 2-17, 95% confidence interval [CI] 9-13) and 21 adults (average age 37 years, range 18-75, 95% CI 29-45). The majority (72%) had bilateral EVA, and 20% of children and 47.6% of adults had undergone cochlear implantation. There were no differences in demographic variables between the entire cohort of patients with EVA and the population that responded to the survey (P values ranged from .079 to 1.00, **Table 1**).

Auditory Symptoms

All 36 patients who completed the survey had hearing loss, which was progressive or fluctuating (64%) and sensorineural (50%) or mixed (36%) in the majority of patients. Among adults, tinnitus (90%), aural fullness (62%), and hyperacusis (67%) were common symptoms, while hyperacusis was common among children (67%; **Figure 1**). Tinnitus was the only symptom significantly more common in adults (90%) than children (47%; $P = .007$). Autophony index (average: 3.0; 95% CI 0.4-5.7) and hyperacusis scores (average: 9.2; 95% CI 5.6-12.8) were low among adult patients.

Vestibular Symptoms

About half of all patients reported experiencing vertigo (56%) or poor balance (53%; **Figure 2**). In all patients who suffered from vertigo, this symptom was episodic (n = 20), and the duration was most commonly less than 5 minutes (n = 12), followed by less than 24 hours (n = 4), and up to 2 days (n = 1). The duration of vertigo could not be determined in 3 patients. Six out of the 20 patients (30%, all adults) with vertigo also had migraine, while none of the patients without vertigo had migraine. Sound induced dizziness was uncommon in both children (13%) and adults (14%; $P = 1.00$). Pressure induced dizziness was reported by 38% of adults and 13% of children ($P = .14$), and exercise induced dizziness was reported by 43% of adults and 13% of children ($P = .08$; **Figure 2**).

Most pediatric patients with EVA had at least one delayed gross motor milestone (73%). Episodic head tilt as an infant or toddler was reported in 20% of children. 40% of children and 43% of adults underwent vestibular physical therapy. A history of head trauma causing hearing loss, dizziness, or loss of consciousness requiring formal medical evaluation was uncommon in both children (7%) and adults (33%; $P = .10$). The presence of a CI did not affect the prevalence of any symptoms (P-values ranged from 0.17 to 1.00).

We compared the prevalence of symptoms among patients with EVA included in the current study to published cohorts of patients with SCDS in **Table 2**. Among patients with EVA in this study, two-thirds of both children and adults suffered from hyperacusis, which is similar to what has been reported for patients with SCDS (4.9%-73.2%).[23,29,35,36] Autophony was reported by about half of adults, which is similar to patients with SCDS (42.5%-82.8%), while it was less commonly reported among children with EVA (20%). The severity of autophony reported by adults with EVA, as quantified by the autophony index (average: 3.0; 95% CI 0.4-5.7) was lower compared to SCDS patients (autophony index average: 42, SD: 27).[29] Hyperacusis experienced by adult EVA patients was also less severe (average: 9.2; 95% CI 5.6-12.8) compared to patients with hyperacusis undergoing round and oval window reinforcements for this symptom (average: 29.6, SD: 6, respectively).[29,37] The

Table 1. Demographics of All Screened Patients

	Pediatric N (%) (total n = 44)	Adult N (%) (total n = 77)	Total N (%) (total n = 121)	Survey pediatric N (%) (total n = 15)	Survey adult N (%) (total n = 21)	Survey total N (%) (total n = 36)	*P* value[a]
Gender							1.000
Male	19 (43%)	32 (42%)	51 (42%)	6 (40%)	9 (43%)	15 (42%)	
Female	25 (57%)	45 (58%)	70 (58%)	9 (60%)	12 (57%)	21 (58%)	
Race							.519
American Indian or Alaska Native	0	1 (1%)	1 (1%)	0	0	0	
Asian	3 (7%)	0	3 (2%)	0	0	0	
Black or African American	8 (18%)	11 (14%)	19 (16%)	1 (6.5%)	1 (5%)	2 (6%)	
White	30 (68%)	59 (77%)	89 (73%)	13 (87%)	18 (85.5%)	31 (86%)	
Other	3 (7%)	4 (5%)	7 (6%)	1 (6.5%)	2 (9.5%)	3 (8%)	
Unknown	0	2 (3%)	2 (2%)	0	0	0	
Ethnicity							1.000
Hispanic or Latino	4 (9%)	2 (3%)	6 (5%)	1 (7%)	1 (5%)	2 (6%)	
Not Hispanic or latino	37 (84%)	65 (84%)	102 (84%)	13 (86%)	17 (81%)	30 (83%)	
Not available	3 (7%)	10 (13%)	13 (11%)	1 (7%)	3 (14%)	4 (11%)	
Insurance							.964
Private	26 (59%)	51 (66%)	77 (64%)	10 (67%)	14 (67%)	24 (66.7%)	
Public	14 (32%)	18 (23%)	32 (26%)	2 (13.5%)	7 (33%)	9 (25%)	
Military	1 (2%)	2 (3%)	3 (2%)	1 (6.5%)	0	1 (2.8%)	
Not insured	2 (5%)	0	2 (2%)	1 (6.5%)	0	1 (2.8%)	
Unknown	1 (2%)	6 (8%)	7 (6%)	1 (6.5%)	0	1 (2.8%)	
EVA							.905
Left	8 (18%)	5 (6%)	13 (11%)	2 (13%)	3 (14%)	5 (14%)	
Right	10 (23%)	9 (12%)	19 (16%)	3 (20%)	2 (10%)	5 (14%)	
Bilateral	26 (59%)	63 (82%)	89 (73%)	10 (67%)	16 (76%)	26 (72%)	
Congenital hearing loss	14 (32%)	34 (44%)	48 (40%)	3 (20%)	7 (33%)	10 (28%)	.392
Progressive or fluctuating hearing loss	11 (25%)	59 (77%)	70 (58%)	7 (47%)	16 (76%)	23 (64%)	.742
Hearing loss type							.079
Sensorineural	30 (68%)	54 (70%)	84 (69.5%)	7 (47%)	11 (52%)	18 (50%)	
Mixed	13 (30%)	18 (23%)	31 (26%)	7 (47%)	6 (28%)	13 (36%)	
Conductive	0	2 (3%)	2 (1.5%)	0	1 (5%)	1 (3%)	
No hearing loss	1 (2%)	1 (1%)	2 (1.5%)	0	1 (5%)	1 (3%)	
Not available	0	2 (3%)	2 (1.5%)	1 (7%)	2 (10%)	3 (8%)	
Cochlear implant	17 (39%)	51 (66%)	68 (56%)	3 (20%)	10 (48%)	13 (36%)	.216
Left	6 (35%)	16 (31%)	22 (32%)	1 (33.3%)	2 (20%)	3 (23%)	
Right	3 (18%)	18 (35%)	21 (31%)	1 (33.3%)	4 (40%)	5 (38.5%)	
Bilateral	8 (47%)	17 (33%)	25 (37%)	1 (33.3%)	4 (40%)	5 (38.5%)	
Comorbidity							
Syndrome	2 (5%)	2 (3%)	4 (3%)	1 (7%)	0	1 (3%)	1.000
Migraine	2 (5%)	18 (23%)	20 (17%)	0	6 (29%)	6 (17%)	1.000
Neurodevelopmental[b]	4 (9%)	3 (4%)	7 (6%)	2 (13%)	1 (5%)	3 (8%)	.697

[a]Comparing the total (pre-survey) group of patients to the group of patients that filled out the survey.
[b]Includes autism and developmental delay.

prevalence of pressure induced dizziness among adult EVA patients was 38%, which is on the lower end of the previously reported range for SCDS patients (37.4%-72.4%). Sound induced dizziness was less common in both adult and pediatric patients with EVA (<15%) compared to patients with SCDS (42.7%-82.8%).[23,35,36] The prevalence of other less specific symptoms, such as ear fullness and spontaneous dizziness or vertigo, in both adult (62% and 67% respectively) and pediatric (47% and 40% respectively) patients with EVA was also similar to patients with SCDS (24.6%-93.1% and 51%-89.7% respectively). Statistical analyses could be performed

between adult SCDS patients and adult EVA patients from the current study using Fisher's exact tests with a Bonferroni adjusted P value of .006 (=0.05/9) and showed that hearing loss was more common in the EVA group ($P < .001$), while sound induced dizziness was more common in the SCDS group ($P < .001$) (**Table 3**).[23,36]

Discussion

This pilot study reports auditory and vestibular symptoms among a cohort of patients with EVA and compares the prevalence of symptoms among adult and pediatric patients. Rates of hearing loss, vertigo, delayed gross motor milestones, and head trauma resulting in dizziness or hearing loss were similar to previously reported EVA cohorts.[24,38] Patients were surveyed about symptoms typically reported among patients with other third window syndromes such as SCDS. We found that adult EVA patients experience symptoms similar to those seen in patients with SCDS, including aural fullness, hyperacusis, autophony, and pressure induced dizziness.[23,29,35,36] Among adult patients, hearing loss appears more common in EVA patients ($P < .001$) while sound induced dizziness seems more common in SCDS ($P < .001$; **Tables 2** and **3**). To the authors' knowledge, this is the first study to investigate the presence of third window symptoms in patients with EVA.

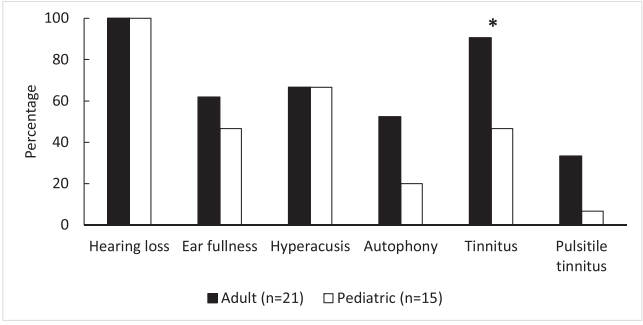

Figure 1. Prevalence of auditory symptoms displayed as percentages (*y*-axis) for adult (black bars) and pediatric (white bars) patients. *Tinnitus was significantly more common in adults compared to children ($P = .007$).

This study shows an overlap in symptomatology between patients with EVA and SCDS. Although both fall under the umbrella of third mobile window disorders, they are distinct pathologies. Most patients with EVA in our study had SNHL, and just over one-third had undergone cochlear implantation. Though the pathophysiology of SNHL in patients with EVA is incompletely understood, several genetic mutations have been linked to EVA and are thought to play a role in developing SNHL in this patient population.[24] In contrast, SCDS patients do not typically have SNHL. Patients with EVA also suffer from vestibular hypofunction, which has been theorized to occur due to an endolymphatic osmotic and chemical imbalance that leads to degeneration of vestibular hair cells. In contrast, the vestibular system is overstimulated in patients with SCDS, but degeneration or changes to end organs or hair cells have not been reported.[39-42] The pathologic inner ear changes resulting in SNHL and vestibular hypofunction among patients with EVA may confound the reporting of third window-type symptoms and contribute to differences in reported symptoms compared to patients with SCDS. For example, higher hearing loss and tinnitus rates among patients with EVA would be expected compared to patients with SCDS. Conversely, pathophysiologic changes in the cochlea and vestibular system among patients with EVA, which can be progressive, may mask other auditory and vestibular third window symptoms. In addition, the presence of a cochlear implant affects cochlear mechanics and may affect symptomatology. In the current study, it is unclear if patients with cochlear implants experienced differences in symptoms with their implant turned on or off and if patients had residual hearing after cochlear implantation, which could also

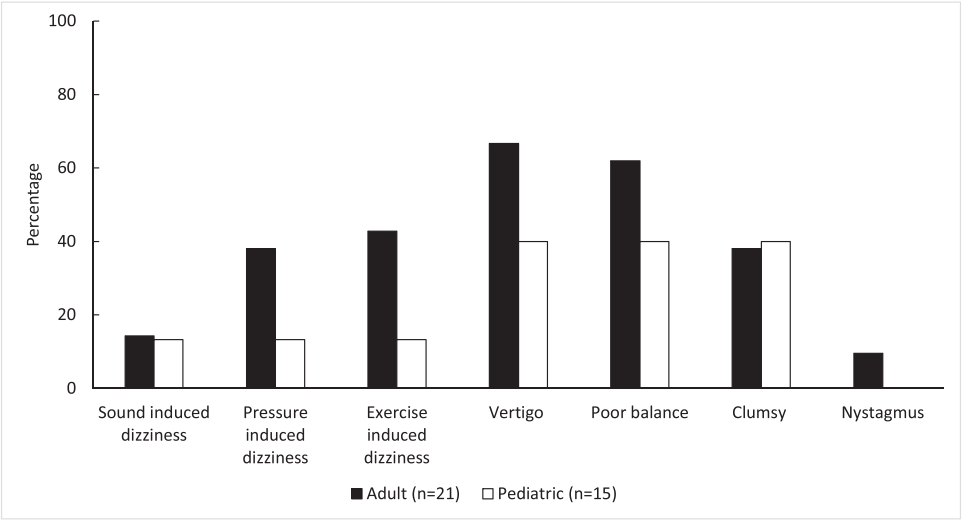

Figure 2. Prevalence of vestibular symptoms displayed as percentages (*y*-axis) for adult (black bars) and pediatric (white bars) patients.

Table 2. Comparing Prevalence of Symptoms in Patients With Semicircular Canal Dehiscence (SCDS) Syndrome and Enlarged Vestibular Aqueduct

Symptoms in SCDS patients	Naert et al. 2018 (n = 431, retrospective)[32]	Naert et al. 2020 (n = 29, prospective)[33,34]	Noij et al. 2018 (n = 98)[34]	Current study, adults with EVA (n = 21)	Current study, children with EVA (n = 15)
Hearing loss	39.9%	69%	57.5%	100%	100%
Ear fullness	24.6%	93.1%	78.2%	62%	47%
Hyperacusis	4.9%	58.6%	73.2%	67%	67%
Autophony	42.5%	82.8%	76.5%	52%	20%
Nonpulsatile tinnitus	17.2%	69%	61.6%	90%	47%
Pulsatile tinnitus	0.5%	62.1%	-	33%	7%
Dizziness or vertigo	51%	89.7%	80.4%	67%	40%
Sound induced dizziness	42.7%	82.8%	62.0%	14%	13%
Pressure induced dizziness	37.4%	72.4%	64.6%	38%	13%

Table 3. *P* Values for the Comparison of Symptom Prevalence Between Adult EVA and SCDS Patients

Comparing symptom prevalence between adult EVA and SCDS[22,35] patients	*P* values[a]
Hearing loss	<.001*
Ear fullness	.077
Hyperacusis	.802
Autophony	.027
Nonpulsatile tinnitus	.012
Pulsatile tinnitus	.085
Dizziness or vertigo	.133
Sound induced dizziness	<.001*
Pressure induced dizziness	.025

Abbreviations: EBA, enlarged vestibular aqueduct; SCDS, Semicircular Canal Dehiscence.
[a]Bonferroni adjusted *P* value for statistical significance is .006 (.05/9).
*Indicates statistical significance.

affect symptoms. Prospective studies are needed to determine whether there are differences in third window symptoms among patients with EVA with different hearing and vestibular impairment levels before and after cochlear implantation.

Surgical techniques are effective in treating symptoms caused by several third-window syndromes. Semicircular canal plugging is effective in treating symptoms of SCDS. Similarly, perilymphatic fistulas may be treated by plugging the defect, and cochlea-facial nerve dehiscence has been treated with round window reinforcement.[43-47] Although attempts have been made to surgically treat EVA with various endolymphatic sac procedures, including shunting, occlusion, and obliteration, none have been effective.[24,46] Though there are no current treatment options to address third window symptoms in EVA, an improved understanding of their prevalence is important as this could guide future treatment paradigms.

The small sample size of this study and heterogeneity of CT imaging, which was performed at various institutions using different protocols, precluded assessment of the relationship between EVA size and third window symptoms. Previous studies investigating EVA size have not found a correlation with hearing loss or vertigo.[48,49] Future studies may determine whether EVA size correlates with third window-specific symptomatology. Notably, the dehiscence size in patients with SCDS has not been shown to correlate with the severity of autophony or dizziness.[29,47] In addition, in the current study no information was available regarding the degree of hearing loss at the time of the survey, which precluded analyses of relationship between hearing loss severity and symptomatology. In future studies it would be valuable to combine obtaining survey responses with an audiology appointment.

Our study was limited by a 30% response rate among our cohort, which resulted in a small sample size of adult and pediatric patients. As shown by Naert et al, large differences in symptom reporting can be seen in patients who are prospectively (prior to any kind of intervention) versus retrospectively questioned about their symptoms and although there were no demographic differences in the entire sample of EVA patients and the survey participants, there is risk for a selection bias as patients with symptoms may be more likely to respond to a study participation request, which would result in an overestimation of symptom prevalence.[36] Therefore, a large, prospective, and ideally multi-institutional study inquiring about third window symptoms in EVA patients is warranted to better understand symptomatology in this patient population. In addition, there was a wide range of developmental stages in the pediatric group included in

this study and the questionnaires in the pediatric population were in part parent-reported, which adds to the difficulty in interpretation of these results. Especially in young children, the presence of symptoms such as tinnitus, autophony, and pressure induced dizziness may be difficult to assess, which could have affected the actual prevalence of these symptoms. Few patients in this study underwent vestibular testing. Although reports in small groups of EVA patients have found VEMP results similar to findings in SCDS, a larger prospective study is needed to assess vestibular testing outcomes in patients with EVA and how they compare to patients with SCDS.[24]

Conclusion

This pilot study is the first study investigating the prevalence of symptoms typical of other third-window disorders among patients with EVA. There is overlap in the prevalence of several specific and nonspecific third window symptoms among patients with EVA and SCDS, with adult EVA patients suffering from similar rates of hyperacusis, autophony, pressure induced dizziness, ear fullness, and spontaneous vertigo. Children with EVA had similar rates of hyperacusis, ear fullness, and spontaneous vertigo, while pressure induced dizziness and autophony were less commonly reported in children, and sound induced dizziness was less commonly reported in pediatric and adult EVA patients compared to SCDS patients. Future research, including prospective studies, may allow for better identification and management of symptoms among patients with EVA.

Author Contributions

Kimberley S. Noij, substantial contributions to conception and design, conduct, analysis and presentation of research; **Emily Y. Huang**, substantial contributions to conception and design, conduct, analysis and presentation of research; **Nadia L. Samaha**, substantial contributions to conduct, analysis and presentation of research; **Jonathan M. Walsh**, substantial contributions to conception and design, conduct, analysis and presentation of research; **Bryan K. Ward**, substantial contributions to conduct, analysis and presentation of research; **John P. Carey**, substantial contributions to conduct, analysis and presentation of research; **Carolyn M. Jenks**, substantial contributions to conception and design, conduct, analysis and presentation of research.

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Supplemental Material

Additional supporting information is available in the online version of the article.

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