Prognostic Factors for Sudden Drops in Hearing Level After Minor Head Injury in Patients With an Enlarged Vestibular Aqueduct: A Meta-analysis

Bo Jan Noordman, Eveline van Beeck Calkoen, Birgit Witte, Theo Goverts, Erik Hensen, and Paul Merkus

Department of Otorhinolaryngology and Head and Neck Surgery, The EMGO Institute for Health and Care Research VU University Medical Center, Amsterdam, The Netherlands

Objective: To identify factors associated with sudden drops in hearing level after minor head trauma in patients with an enlarged vestibular aqueduct (EVA).

Methods: A systematic review of the literature on sudden drops in hearing level after minor head trauma in patients with an EVA was conducted. The studies were retrieved from Embase, PubMed, CINAHL, and Cochrane and critically appraised using predefined criteria. Data on all described parameters were collected, and their relation with sudden drops after minor head trauma was statistically analyzed.

Results: Pooled data of 31 articles included 179 patients with 351 EVAs. Drops in hearing level after minor head trauma were experienced by 34% of the patients. We found a significant association between sudden deterioration of hearing after minor head trauma and preexisting fluctuating hearing loss (HL) (odds ratio, 8.6; p < 0.001; 95% confidence interval, 3.9–19.3). The

The vestibular aqueduct (VA) is a bony channel in the temporal bone that courses from the posterior cranial fossa to the medial wall of the vestibule. It contains the endolymphatic duct, which connects the endolymphatic sac with the vestibule. The normal width of the VA has been described to be less than 0.9 mm at its midpoint or less than 1.9 mm at the operculum (1). The operculum is a variable projection of bone on the posterior face of the petrous bone that outlines the opening of the VA (2).

An enlarged VA (EVA) is a common finding in children with congenital hearing loss (HL) (3,4). About 10% of all children with significant permanent HL have an

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diameter of the VA, type of preexisting HL, severity of HL, preexisting progressive HL, and the diagnosis Pendred syndrome were not significantly associated with sudden drops in hearing levels after head trauma.

Conclusion: Only one-third of the patients with a proven EVA experienced sudden drops in hearing level because of head trauma. There is a significant association between preexisting fluctuating HL and the chance of sudden drops in hearing level caused by trauma. Stringent lifestyle advices, like avoiding activities with a risk of minor head trauma such as contact sports, might be restricted to patients with a fluctuating HL and those with a history of sudden drops on minor head trauma. **Key Words:** Congenital sensorineural hearing loss—Enlarged vestibular aqueduct—Head trauma—Pediatric—Pendred—Sudden drops.

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EVA, making this the most frequent morphogenetic abnormality in these children (5–7). Different criteria for the diagnosis EVA are found in the literature. Originally, the VA was considered enlarged if it was greater than 1.5 mm at the midpoint (8). Other investigators define EVA as a diameter greater than 2 mm at the midpoint or a diameter greater than 4 mm at the operculum (9–12).

In the majority of EVA patients, computed tomography or magnetic resonance imaging scans reveal additional inner ear anomalies, mostly a Type 2 incomplete partition of the cochlea (IP2), previously known as Mondini malformation (13–15). The IP2 or classical Mondini malformation consists of a triad: an EVA, a dilated vestibule, and a dysplastic cochlea, with 1.5 turns caused by a cystic cochlear apex with a normal basal turn. Combinations of EVA with other isolated malformations are also possible, such as an abnormally large vestibule, enlargement of semicircular canals, or a hypoplastic cochlea (8,16–18). Furthermore, EVA is associated with defects in thyroid iodine organification caused by mutations in the *SLC26A4* (PDS)

Address correspondence and reprint requests to Paul Merkus, M.D., Ph.D., Department of Otorhinolaryngology and Head and Neck Surgery, The EMGO Institute for Health and Care Research VU University Medical Center, KNO 1d-114, P. O. Box 7057 1007 MB, Amsterdam, The Netherlands; E-mail: P.Merkus@VUmc.nl, b.j.noordman@gmail.com

gene, resulting in hypothyroidism and goiter (19,20). The combination of EVA and thyroid dysfunction is called Pendred syndrome, which is an autosomal recessive disorder (21). Other associated syndromes are distal renal tubular acidosis (5,6), branchio-oto-renal syndrome (22), and Waardenburg's syndrome (7).

HL in EVA patients is predominantly sensorineural. A conductive component, explained by a third window effect, may be observed at the lower frequencies, and fluctuations in hearing level and progression of HL are frequent (9,23,24).

A sudden drop in hearing level triggered by minor head injuries, barotrauma, or noise trauma is a well-known feature of EVA patients. The reported risk of sudden drops on minor head trauma, noise trauma, or barotrauma in patients with EVA is highly variable (3%–80%) (10,25). Because of the risk of deterioration of hearing on these events, some clinicians recommend all EVA patients to avoid activities such as contact sports and scuba diving or to wear helmets (26–28). This policy can be quite restricting, especially in young children. Because only a minority of EVA patients experience HL on head trauma or barotrauma, we conducted the present meta-analysis to define prognostic factors for sudden HL and identify subgroups of EVA patients that would benefit from these recommendations and, more importantly, subgroups that would not.

MATERIALS AND METHODS

Search Strategy and Study Selection

A systematic literature search was performed on EVA in PubMed, Embase, CINAHL, and the Cochrane Library databases from the inception of the databases to July 2, 2013, using the search term enlarged vestibular aqueduct and its synonyms in the title and abstract fields. A complete overview of the search terms is shown in Table 1. To increase the yield of relevant studies, the reference lists of all identified articles were screened, and related publications were searched in Web of Science. Two reviewers (P. M., B. N.) independently screened titles and abstracts of the retrieved publications. Discrepancies between the reviewers in the assessment of the articles were resolved by consensus discussion. All articles on Pendred syndrome, Mondini/IP2-type malformation, EVA, and/or large VA syndrome in which hearing was reported were selected. The full text of these eligible studies was screened for a more detailed selection. Studies had to meet all of the following criteria to be included in this meta-analysis: the described patients have a radiographically proven EVA in at least one ear and sudden drops in hearing after head trauma, barotrauma, or noise trauma is reported. Furthermore, at least one of the following was described: the existence of Pendred syndrome, fluctuating HL or progressive HL, the degree of HL in dB or classification, the type of HL, the mid-diameter or size of the operculum of the VA in millimeters, and/or the occurrence of vestibular symptoms. Parameters had to be described separately per patient and/or per ear. The criteria for the diagnosis of an EVA were used as defined by the authors of the included studies.

TABLE 1. Systematic search for studies on enlarged vestibular aqueduct (date of search: July 2, 2013)

Database	Search	Hits		
PubMed	(Large[Title/Abstract] AND vestibular[Title/Abstract] AND aqueduct[Title/Abstract]) OR (Large[Title/Abstract] AND vestibular[Title/Abstract] AND aqueducts[Title/Abstract]) OR (Enlarged[Title/Abstract] AND vestibular[Title/Abstract] AND aqueducts[Title/Abstract]) OR (Enlarged[Title/Abstract] AND vestibular[Title/Abstract] AND aqueducts[Title/Abstract]) OR (Wide[Title/Abstract] AND vestibular[Title/Abstract] AND aqueducts[Title/Abstract]) OR (Wide[Title/Abstract] AND vestibular[Title/Abstract] AND aqueducts[Title/Abstract]) OR (Wide[Title/Abstract] AND vestibular[Title/Abstract] AND aqueducts[Title/Abstract]) OR (Enlargement[Title/Abstract] AND vestibular[Title/Abstract] AND aqueducts[Title/Abstract]) OR (Enlargement[Title/Abstract] AND vestibular[Title/Abstract] AND aqueducts[Title/Abstract]) OR (Enlargement[Title/Abstract] AND vestibular[Title/Abstract] AND aqueducts[Title/Abstract]) OR (Widened[Title/Abstract] vestibular[Title/Abstract] aqueduct[Title/Abstract]] OR (Widened[Title/Abstract] vestibular[Title/Abstract] aqueducts[Title/Abstract]] OR			
Embase	 (Large:ab,ti AND vestibular:ab,ti AND aqueduct:ab,ti) OR (Large:ab,ti AND vestibular:ab,ti AND aqueduct:ab,ti) OR (Enlarged:ab,ti AND vestibular:ab,ti AND aqueduct:ab,ti) OR (Enlarged:ab,ti AND vestibular:ab,ti AND aqueduct:ab,ti) OR (Wide:ab,ti AND vestibular:ab,ti AND aqueduct:ab,ti) OR (Wide:ab,ti AND vestibular:ab,ti AND aqueduct:ab,ti) OR (Enlargement:ab,ti AND vestibular:ab,ti AND aqueduct:ab,ti) OR (Enlargement:ab,ti AND vestibular:ab,ti AND aqueduct:ab,ti) OR (Enlargement:ab,ti AND vestibular:ab,ti AND aqueduct:ab,ti) OR (Wide:ab,ti AND aqueduct:ab,ti) OR (Wide:ab,ti vestibular:ab,ti aqueduct:ab,ti) OR (Wide:ab,ti vestibular:ab,ti aqueduct:ab,ti) 	421		
CINAHL	 TI (large AND vestibular AND (aqueduct OR aqueducts.a), i) OR TI (enlarged AND vestibular AND (aqueduct OR aqueducts)) OR TI (wide AND vestibular AND (aqueduct OR aqueducts)) OR TI (enlargement AND vestibular AND (aqueduct OR aqueducts)) OR TI (wide AND vestibular AND (aqueduct OR aqueducts)) OR TI (aqueduct OR aqueducts)) OR AB (large AND vestibular AND (aqueduct OR aqueducts)) OR AB (enlarged AND vestibular AND (aqueduct OR aqueducts)) OR AB (enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (wide enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (wide enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (wide enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (wide enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (wide enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (wide enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (wide enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (wide enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (wide enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (wide enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (wide enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (wide enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (wide enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (wide enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (wide enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (wide enlargement AND vestibular AND (aqueduct OR aqueducts)) OR AB (wide enlargement AND vestibular AND (aqueduct OR aqu	60		
Cochrane	 No. 1 [in "record title"] (large AND vestibular AND (aqueduct OR aqueducts)) OR (enlarged AND vestibular AND (aqueduct OR aqueducts)) OR (wide AND vestibular AND (aqueduct OR aqueducts)) OR (enlargement AND vestibular AND (aqueduct OR aqueducts)) OR (Widened AND vestibular AND (aqueduct OR aqueducts)) No. 2 [in "abstract"] (large AND vestibular AND (aqueduct OR aqueducts)) OR (enlarged AND vestibular AND (aqueduct OR aqueducts)) OR (aqueduct OR aqueducts)) OR (enlargement AND vestibular AND (aqueduct OR aqueducts)) No. 2 [in "abstract"] (large AND vestibular AND (aqueduct OR aqueducts)) OR (wide AND vestibular AND (aqueduct OR aqueducts)) OR (aqueduct OR aqueducts)) OR (enlargement AND vestibular AND (aqueduct OR aqueducts)) OR (Widened AND vestibular AND (aqueduct OR aqueducts)) No. 1 OR No. 2 	4		

Pendred syndrome was classified as an EVA with at least one of the following concurrent findings: a positive perchlorate test, a positive genetic analysis (two mutant SLC26A4 [PDS] alleles), or hypothyroidism in combination with goiter. Hearing loss was classified as no HL (<20 dB), mild HL (20-40 dB), moderate HL (41-70 dB), severe HL (71-95 dB), and profound HL (>95 dB), as described earlier by Martini and Mazzoli (29). Hearing level was evaluated with a pure-tone average of 500, 1,000, and 2,000 Hz. When audiologic data were not shown, the classification of HL as used by the authors was used. Progressive hearing loss was, pragmatically, defined as a deterioration of more than 10 dB at two or more frequencies or a deterioration of 15 dB at one or more frequencies in 1 year. Fluctuating hearing loss was defined as a 10-dB improvement in hearing in two or more frequencies or a 15-dB improvement at one frequency. When audiologic data were not shown, the definitions for progressive and fluctuating hearing loss as used by the authors were used. In the absence of information on whether the fluctuations/progression in HL or the presented level of HL preceded the sudden drops in hearing level or vice versa, fluctuating HL, progressive HL, and level of HL were classified as preexisting. The complete selection process is presented in Figure 1.

Data Collection

The data from all patients included in the selected studies were pooled. A standardized data collection form was developed, pilot tested, and refined. Information on the following themes was extracted for each patient (and when possible for each ear) that was evaluated in the included studies: Pendred syndrome, preexistent fluctuations in HL, preexistent progression of HL, degree of HL, mid-diameter of the VA, size of the operculum, preexistent vestibular symptoms, and type of HL (Table 2). Only studies with data on sudden deterioration of the hearing of individual patients were selected. If information on sudden drops of hearing was absent for one of the patients,

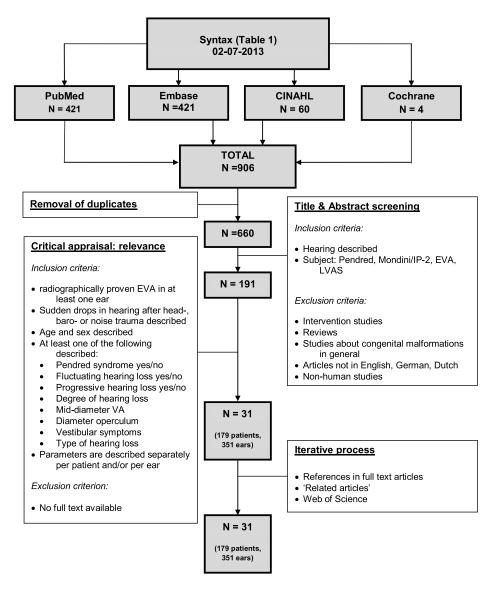


FIG. 1. Flowchart for the selection process of studies on sudden drops in hearing level in EVA patients. LVAS indicates large vestibular aqueduct syndrome; IP-2, type 2 incomplete partition.

Variable		n/N	OR	p value	95% CI
Pendred syndrome	No	6/11 patients	1.0	0.197	
	Yes	13/40 patients	0.40		0.10-1.6
Fluctuating hearing loss	No	6/85 ears	1.0	< 0.001	
	Yes	44/86 ears	8.6		3.9-19.3
Progressive hearing loss	No	20/75 ears	1.0	0.606	
	Yes	44/148 ears	1.2		0.63-2.2
Degree of hearing loss	No	3/12 ears	1.0	0.497	
	Mild	3/10 ears	1.3		0.20-8.8
	Moderate	16/41 ears	2.3		0.53-10.1
	Severe	21/70 ears	1.4		0.34-5.7
	Profound	46/118 ears	2.2		0.56-8.9
Mid diameter VA	≤2.0 mm	5/31 ears	1.0	0.446	
	2.01-2.50 mm	6/18 ears	2.5		0.57-10.5
	2.51-3.00 mm	7/19 ears	3.4		0.73-16.0
	>3.0 mm	4/18 ears	2.0		0.42-9.6
Size operculum	≤7.00 mm	2/21 ears	1.0	0.134	
*	>7.00 mm	4/13 ears	4.6		0.63-33.5
Vestibular symptoms	No	7/22 patients	1.0	0.612	
- A	Yes	27/70 patients	1.3		0.45-3.9
Type of hearing loss	SNHL	7/24 ears	1.0	0.259	
· · · ·	Mixed	29/79 ears	1.8		0.64-5.3

TABLE 2. Logistic regression analysis of precipitating factors related with sudden hearing drops

n indicates the number of patients or ears with sudden hearing drops; N, the number of patients or ears that were subtracted from the literature that fulfilled the criteria to be analyzed on this specific variable; OR, odds ratio; 95% CI, 95% confidence interval; VA, vestibular aqueduct.

whereas the presence of drops of hearing was reported for other patients within the same study, the patient was classified as having *no drops in hearing*. The parameters *Pendred syndrome* and *vestibular symptoms or abnormalities* were analyzed *per patient*. The anatomic aspects or aspects of hearing were analyzed *per ear*. In case of a radiologically proven bilateral EVA and missing information on lateralization of one of these parameters, the concerning parameter was classified as *bilateral*. Two authors independently extracted data from the included studies (P. M., B. N.). Any discrepancies were resolved by consensus discussion.

Statistical Analysis

The results were calculated using logistic regression analysis and are expressed as odds ratio (OR), p value, and 95% confidence interval (CI). All results were corrected for age and sex using logistic regression analysis. Therefore, only patients with available data on age and sex were included in the statistical analyses. All statistical analyses were performed using the Statistical Package for the Social Sciences software version 20.0 (IBM Corp., Armonk, NY, USA). A value of p < 0.05 was considered statistically significant.

RESULTS

Search Results

Figure 1 shows the flowchart of our search. We identified 906 records from the database search (search date, July 2, 2013). After removing the duplicates, 660 unique articles remained. A total of 191 articles concerning hearing in EVA patients were identified. Full text screening using the inclusion and exclusion criteria as previously described resulted in the selection of 31 articles, describing 179 patients with 351 ears with EVA (6,24,30–58).

Definition of EVA

Definitions for EVA used by the included studies are shown in Table 3. Fifteen of the included studies (48%)

TABLE 3.	Definition	of EVA
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Definition	Studies
>1.5 mm measured halfway between the common crus and the medial aspect of the	n = 14 (6,24,31,32,34,35,
opening of the operculum on the posterior wall of the temporal bone (8) .	38,39,46,48-51,55,57)
Explanation of the <i>mid duct measurement</i> is given in Figure 2.	
Definition not mentioned	n = 10 (30, 33, 36, 37,
	42,43,45,47,52,56)
>2.0 mm, by measurement of a line extending between the two lips of the external aperture (11).	n = 2 (40,41)
Diameter >2 mm, based on the work of Wilbrand et al. (59) who described the diameter of the VA on normal temporal bones as $0.4-1.0$ mm.	n = 1 (53)
Aperture larger than 4 mm and a distance between the vestibule and the traceable	n = 1 (44)
outline of the VA of less than 1 mm on high-resolution CT scans (12).	
>1.5 mm at the midpoint between the common crus and the external operculum or 2.0 mm at the operculum itself	n = 1 (48)
Multiple definitions mentioned. "Cases fulfill all definitions."	n = 1 (54)

The definition is mentioned in the left column, and the studies using this definition are listed in the right column.

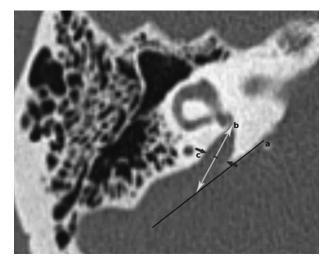


FIG. 2. Vestibular aqueduct: mid duct diameter measurement. Example to explain how to measure an enlarged vestibular aqueduct at the mid duct diameter. *a*, create a virtual line (*black line*) of the posterior cranial fossa dura; the duct cannot be measured beyond this point. *b*, create in the middle of the duct a line between the most anteromedial point and the posterolateral point. *c*, in the middle of this line (*cross mark*) is the best position to measure the diameter of the vestibular aqueduct in millimeters. In this case, between the *two arrows*.

use the definition as presented by Valvassori and Clemis (8) in 1978; a diameter greater than 1.5 mm halfway between the common crus and the medial aspect of the opening of the operculum on the posterior wall of the temporal bone. How to measure the EVA is explained in Figure 2. In 10 (32%) of the studies, no clear definition is presented. Other definitions found in this study were those described by Levenson et al., Willbrand et al., and Okumura et al. (Table 3) (11,12,59).

Statistical Analysis

To find factors associated with sudden drops in hearing level after minor head trauma, noise trauma, or barotrauma, we analyzed the presence of sudden drops and its association with Pendred syndrome, preexisting fluctuating HL, preexisting progressive HL, the preexisting severity of HL, the mid-diameter VA, the diameter of the operculum, vestibular symptoms, and the type of HL. All parameters will be discussed separately below and are shown in Table 2.

Sudden Drops in Hearing

In 61 (34%) of 179 patients, sudden drops in hearing after minor head trauma, barotrauma, or noise trauma in at least one ear were reported. Of all 351 ears with EVA, 108 (30%) were described to suffer from sudden drops. For most patients, minor head trauma was reported as a trigger for sudden HL; other reported triggers are barotrauma, noise trauma, and upper respiratory tract infection.

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Pendred Syndrome

Data on Pendred syndrome, as diagnosed by the perchlorate test, genetic analysis, and/or the presence of hypothyroidism in combination with goiter, was available in 51 (28%) of all included patients. Of these 51 patients, 40 (78%) were classified to suffer from Pendred syndrome. No significant relationship between Pendred syndrome and the presence or absence of sudden drops was found (OR, 0.40; p = 0.197; 95% CI, 0.10–1.6).

Fluctuating HL

Information on preexistent fluctuating HL was available in 171 (49%) of all included ears. Fluctuating HL was found in 86 of these ears (50%). Detailed audiometric data (including tympanometry and bone conduction measures) were not provided in 8 (38%) of 23 studies. Therefore, fluctuating and progressive HL as reported in these studies may be caused by fluctuating sensorineural thresholds or fluctuating middle ear function, for example, based on otitis media with effusion. Otitis media with effusion might even obscure fluctuations in sensorineural thresholds in some cases. Our analysis showed a significant association between preexistent fluctuating HL and sudden drops (OR, 8.6; *p* < 0.001; 95% CI, 3.9–19.3). This OR represents the ratio of the odds that a patient with fluctuating HL will experience sudden drops in hearing compared with the odds of a patient with no history of fluctuating HL. Thus, the odds for experiencing a sudden drop in hearing is 8.6 times higher in patients with preexistent fluctuating HL compared with patients without preexistent fluctuating HL.

Progressive HL

Data on progression of HL were available in 223 (64%) of all included ears. Of those ears, 148 (67%) were found to have progressive HL before the head trauma. No significant relationship between preexistent progressive HL and sudden drops in hearing level after minor head trauma was found (OR, 1.2; p = 0.606; 95% CI, 0.63–2.2).

Degree of HL

The preexistent degree of HL was described in 251 (72%) of all included ears. In the majority (20/26, 77%) of studies with data on the degree of HL, hearing level was evaluated using a pure-tone average of 500, 1,000, and 2,000 Hz. The other studies used the mean hearing level of all frequencies (250, 500, 1,000, 2,000, 4,000, and 8,000 Hz) or speech reception threshold. HL was classified as no HL (n = 12; 5%), mild HL (n = 10; 4%), moderate HL (n = 41; 16%), severe HL (n = 70; 28%), and profound HL (n = 118; 47%) (29). The odds for developing sudden HL were calculated for each subgroup and, for the OR, no HL was set as the reference group. No significant relation was found between the degree of HL and the incidence of sudden drops in hearing level after minor head trauma (mild HL: OR, 1.3; 95% CI, 0.20-8.8; moderate HL: OR, 2.3; 95% CI, 0.53-10.1; severe HL: OR, 1.4; 95% CI, 0.34–5.7; profound HL: OR, 2.2; 95% CI, 0.56-8.9; overall p = 0.497).

Mid-diameter VA

The diameter of the midpoint of the VA was presented in 86 (25%) of all included ears. In the absence of a linear relationship between the mid-diameter of the VA and sudden drops in hearing level (data not shown), VA midpoint diameter was classified in four categories (\leq 2.00 mm, 2.01–2.50 mm, 2.51–3.00 mm, and >3.00 mm). Evaluation of the relationship between the midpoint diameter of the VA and the presence or absence of sudden drops did not show a significant association (overall *p* = 0.446; ORs were calculated compared with the \leq 2.0-mm group); 2.01 to 2.50 mm: OR, 2.5 (95% CI, 0.57–10.5); 2.51 to 3.00 mm: OR, 3.4 (95% CI, 0.73–16.0); and greater than 3.0 mm: OR, 2.0 (95% CI, 0.42–9.6).

Size of the Operculum

The size of the operculum of the VA was described in 34 of all included ears (9%). No linear relationship between the size of the operculum and sudden drops was found (data not shown). Therefore, operculum diameter was classified in two categories (\leq 7.00 mm and >7.00 mm). No significant association was found between sudden drops and the size of the operculum (OR, 4.6; *p* = 0.134; 95% CI, 0.63–33.5).

Vestibular Symptoms or Abnormalities

In 92 (51%) of all included patients, the occurrence of vestibular symptoms or abnormalities was described. Vestibular symptoms were found in 70 (76%) of these patients. Further analysis did no not show a significant relationship between preexisting vestibular symptoms or abnormalities and the occurrence of sudden drops in hearing level on minor head trauma (OR, 1.3; p = 0.612; 95% CI, 0.45–3.9).

Type of HL

Data on the type of HL were available for 108 ears (31%). The predominant type was mixed HL (n = 79, 73%). Sensorineural HL was found in 24 ears (22%), no HL in 5 ears (5%), and none of the included ears showed a pure conductive HL. Our analysis did not reveal a significant association between type of HL and sudden drops of hearing (OR, 1.8; p = 0.259; 95% CI, 0.64–5.3).

DISCUSSION

The objective of this article is to provide EVA patients with evidence-based lifestyle advice and to identify cases in which there is a need for restricting activities to avoid hearing deterioration. Previous studies show a wide variability in the percentage of EVA patients that develop sudden drops in hearing levels on minor head trauma, noise trauma, or barotrauma (3%-80%) (10,25). In this meta-analysis, we found that approximately one-third of all EVA patients experience deteriorations of hearing after head trauma and, therefore, the majority of EVA patients do not. Similar percentages on sudden drops in hearing level were found in a previous study on long-term follow-up of HL in 27 patients with EVA (33%) (27). Unfortunately, because of missing data on individual patients, this study could not be included in our analysis.

In this meta-analysis, we found that the occurrence of sudden drops in hearing level after minor head trauma, barotrauma, or noise trauma in EVA patients is significantly associated with preexisting fluctuating HL. The other studied parameters (Pendred syndrome, the severity of HL, the diameter of the VA, vestibular symptoms or abnormalities, and type of HL) did not show a significant relationship with deterioration of hearing after head trauma. In contrast to the report of Colvin et al. (27), we also found no association between preexistent progressive HL and sudden drops in hearing after head trauma. Possible explanations for this discrepancy are the difference in definition of EVA and the relatively small number of included EVA patients (n = 27) in the study by Colvin et al. (27).

The findings of this review may have important implications for the counselling of EVA patients and their parents. It seems unnecessary to recommend all EVA patients to restrain from activities such as contact sports and scuba diving or recommend young children to wear helmets (26-28). Especially for children, these restrictions may have far-reaching social consequences. This study provides a rationale for the limitation of these lifestyle advices to the following subgroups of EVA patients: first, the group who presents themselves with a history of sudden drops in hearing level after minor head trauma, barotrauma, or noise trauma. The second group consists of EVA patients who are characterized by spontaneous preexistent fluctuations in hearing level. These fluctuations can only be recognized when audiometric assessment is appropriate and follow-up has been long enough to have several documented episodes of fluctuations in HL (without trauma). We therefore recommend accurate age-appropriate and frequent audiologic assessment of children with EVA, distinguishing sensorineural and conductive components.

To our knowledge, this is the first study to review the occurrence of sudden drops of hearing level in EVA patients on head trauma in a systematic way. The obvious benefit of this approach is the large number of EVA patients included in the analysis. Our study has also a number of limitations. First, because of the number of parameters included in this review and the fact that not all parameters were studied in each of the individual studies, there is a substantial amount of missing data. This explains the different patient numbers included in the analyses of the different parameters. Second, most included studies only describe the presence of sudden drops in hearing level, whereas the absence of sudden drops is not described in most patients. To overcome this, we have included only studies reporting on patients with sudden drops on minor head trauma, noise trauma, or barotrauma. Within these studies, we have classified patients who were not reported to have experienced sudden deterioration of hearing as patients with stable hearing after head trauma. Third, because of the retrospective study design,

it was not always possible to determine if the fluctuations in HL preceded the sudden drop or vice versa. A sudden loss of hearing on head trauma may be permanent or reversible. In case of a reversible event, one might argue that it is an expression of fluctuating HL, thereby explaining the association with preexisting fluctuating HL. However, the deterioration in hearing level is often permanent or not completely reversible after head trauma. In these cases, an association with preexisting progressive HL might be expected, but we did not find such an association. Finally, our analyses may be affected by the use of different definitions for EVA by the included studies. We expect this effect to be negligible because the differences between the various definitions are marginal (Table 3). This holds also for the definitions of the variables included, especially fluctuations in hearing loss and progression of hearing loss. To improve the comparability between patients and studies in the future, a uniform definition is necessary. We prefer the initial definition for EVA, the size criterion as put forth by Valvassori and Clemis (8) in 1978 (>1.5 mm measured halfway between the common crus and the medial aspect of the opening of the operculum on the posterior wall of the temporal bone). It is a frequently used straightforward definition that is easily applicable in all axial high-resolution computed tomography scans of the temporal bone with a slice thickness of 1 mm or less. If the length of the VA to the posterior fossa is short, it can be hard to measure the mid duct diameter. In these cases, the operculum measurement can be of help as an alternative criterion.

Even with these limitations, our results suggest a strong association between preexistent fluctuating HL and sudden deterioration of hearing on head trauma in this systemic review. It is a prognostic factor that should be further explored in future prospective research on EVA patients.

CONCLUSION

The majority of patients with a proven EVA do not experience sudden drops in hearing level caused by (minor) head trauma. This meta-analysis shows that only approximately one-third of the reported EVA patients are affected. Patients who experience fluctuations in their hearing level before the minor head trauma, noise trauma, or barotrauma have a significantly increased risk of hearing deterioration on this event. We found no other significant prognostic factors. Therefore, recommendations such as the use of helmets by young EVA patients and restricting activities such as contact sports might be reserved for EVA patients with preexistent fluctuating HL or patients with a history of sudden perception loss after head trauma. It is therefore essential to closely follow the progression of HL and the way in which the progression of HL takes place in all EVA patients.

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