

# Large Vestibular Aqueduct Syndrome: Parents' and Audiologists' Perspectives

DOI: 10.3766/jaaa.17031

James Connor Sullivan\*

Carole E. Johnson\*

Jeffrey L. Danhauer†

Anna Marie Jilla\*

Kristin R. Sanchez\*

## Abstract

**Background:** Large vestibular aqueduct syndrome (LVAS) is an auditory disorder that is difficult to diagnose and manage; it is confirmed when the vestibular aqueduct is  $>1.5$  mm in diameter. Diagnosis of LVAS in children can devastate parents and challenge healthcare professionals who serve these patients and their families.

**Purpose:** This study surveyed parents of children with LVAS about their knowledge of and experiences with LVAS and their attitudes about the support provided to them by healthcare professionals. This study also surveyed audiologists about their knowledge of and experiences with LVAS and their level of confidence in serving families with children having this disorder.

**Research Design:** Cross-sectional survey.

**Study Sample:** 100 parents, mostly mothers, and 144 audiologists responded to invitations to participate in surveys designed to elicit information about their knowledge of, experiences with, and attitudes toward LVAS.

**Data Collection and Analysis:** Invitations via links to participate in a survey on [surveymonkey.com](https://www.surveymonkey.com) were posted in LVAS support group pages on [Facebook.com](https://www.facebook.com) for parents and sent to audiologists randomly selected from the American Academy of Audiology Membership Directory. Descriptive statistics were used to analyze trends in parents' and audiologists' responses.

**Results:** A response rate could not be obtained for the parents' survey because it was impossible to know how many parents actually viewed the invitation to participate via [Facebook.com](https://www.facebook.com). The response rate for the audiologists' survey was 10%. Most of the parents reported that their children had clinical trajectories similar to those of cases reported in the literature, and said they needed more information from their healthcare providers, especially pediatricians and primary care physicians. Most of the audiologists felt confident in their knowledge of and/or skills in aiding in the diagnosis and/or treatment of LVAS, except for issues surrounding cochlear implants. Audiologists were interested in obtaining continuing education about LVAS from multiple sources.

**Conclusions:** Parents of children having LVAS need greater support from their healthcare providers, who in turn need additional information on the topic and should collaborate for supportive and appropriate interprofessional care.

**Key Words:** audiologists, children, cochlear implants, counseling, large vestibular aqueduct syndrome, parents, physicians, sensorineural hearing loss

**Abbreviations:** ANSD = auditory neuropathy spectrum disorders; LVAS = large vestibular aqueduct syndrome; PCP = primary care physician; SNHL = sensorineural hearing loss

---

\*Department of Communication Sciences and Disorders, Hearing Evaluation, Rehabilitation, and Outcomes Laboratory, Oklahoma City, OK; †Department of Speech and Hearing Sciences, University of California Santa Barbara, Santa Barbara, CA

Corresponding author: Carole E. Johnson, Department of Communication Sciences and Disorders, Hearing Evaluation, Rehabilitation, and Outcomes Laboratory, Oklahoma City, OK 73117; Email: [earstoyou@aol.com](mailto:earstoyou@aol.com)

Portions of this article were presented at 2015 American Auditory Society Scientific and Technical Conference, March 2–4, and at the XXXIII World Congress of Audiology, September 21–23, 2016, Vancouver, BC, Canada.

## INTRODUCTION

**L**arge vestibular aqueduct syndrome (LVAS) is an auditory disorder that is difficult to diagnose and manage (Gopen et al, 2011) because its presentation of symptoms and prognosis are highly variable across patients. This variability may create uneasiness in patients and their family members and uncertainty in healthcare professionals regarding the diagnosis and treatment of this disorder. LVAS accounts for ~5–15% of all cases of pediatric hearing loss (National Institute on Deafness and other Communication Disorders, 2015). An enlargement of the vestibular aqueduct in the inner ear that can lead to hearing loss of varying types and degrees and can also result in vestibular deficits (Gopen et al, 2011). The vestibular aqueduct is an opening and extends to the petrous portion of the temporal bone, providing a passageway for a vein and the endolymphatic duct. According to Valvassori and Clemis (1978), who first published on the clinical significance of LVAS, this disorder is diagnosed when the vestibular aqueduct is >1.5 mm in diameter at the midpoint as verified from computerized axial tomography and magnetic resonance imaging. However, the more sensitive and conservative Cincinnati criterion (Vijayasekaran et al, 2007) states that LVAS is diagnosed when the aqueduct is >0.9 mm at the midpoint or when the operculum is >1.9 mm.

Mutations in the SLC26A4 gene are involved in the development of nonsyndromic LVAS and are associated with Pendred syndrome (e.g., Yang et al, 2007). LVAS is associated with progressive, fluctuating, asymmetrical sensorineural hearing loss (SNHL) and/or mixed hearing loss (Lai and Shiao, 2004; Merchant et al, 2007) that may commence with a minor head injury (Noordman et al, 2015). Patients with LVAS may initially present with normal hearing or mild hearing losses that have a high probability of worsening to the severe-to-profound range. Patients having LVAS also experience vestibular abnormalities that range from a slight sense of imbalance to episodic vertigo (Manzari, 2008). Possible ways to manage LVAS have included the initial use of hearing aids until hearing sensitivity decreases to the severe-to-profound range (e.g., Clark and Roeser, 2005), hyperbaric oxygenation (Shilton et al, 2014), corticosteroid therapy (Lin et al, 2005), and cochlear implants (e.g., Mey et al, 2016). Cochlear implantation is a relatively safe (Asma et al, 2010) and effective (Miyamoto et al, 2002) intervention for children with LVAS, but the best time to perform the surgery is not always evident because of the highly variable progression of hearing loss across patients (Ko et al, 2013) and the procedure is not without risk (Wan et al, 2009). Uncertainty about the diagnostic criteria for LVAS, prognosis, and recommended timelines for cochlear implantation

may complicate the identification, diagnosis, and management of patients with LVAS.

Parents of children with LVAS may find the uncertainty surrounding this disorder unsettling, as can also occur with auditory disorders such as auditory neuropathy spectrum disorder (ANSD). For example, Stroebel and Swanepoel (2014) compared parents' experiences with the diagnostic and intervention processes of children with ANSD to those of parents of children with SNHL. Parents of children with ANSD had poorer understanding of, and greater uncertainty about, the disorder, and received conflicting information from healthcare professionals who did not give them enough time to ask questions than parents of children with SNHL. It is possible that parents of children with LVAS have had similar experiences with healthcare professionals. Parents who do not get appropriate support from their healthcare professionals may have to rely on the array of support sources for people with hearing loss, in particular those with cochlear implants, which are available on social media (Saxena et al, 2015). Stroebel and Swanepoel (2014) recommended that healthcare professionals should provide ongoing consultations and structured timelines throughout the diagnostic and rehabilitative processes to reduce uncertainty for parents of children with ANSD; these recommendations may also be helpful for families of children with LVAS. Audiologists should play a key role in this process, which requires knowledge of LVAS and appropriate medical referrals for radiographic studies to confirm its presence. However, some audiologists may be uncomfortable in their knowledge of LVAS and unsure about what recommendations to make.

Given the lack of evidence surrounding parent and audiologist knowledge and experiences with LVAS, the present study was designed to survey (a) parents of children with LVAS about their knowledge of and experiences with LVAS, and their attitudes regarding the support provided to them by healthcare professionals; and (b) audiologists about their knowledge of, and experiences with, LVAS and their level of confidence in serving families with children. The results of this study will help identify the needs of families with children with LVAS and of pre-professionals and professionals, and for continuing education and experiences of audiologists.

## METHOD

### Questionnaires

An exhaustive search of the literature revealed that no questionnaires addressing the needs of this study existed in the literature. Thus, we designed separate questionnaires for the parents' and audiologists' surveys using guidelines provided by Cummings et al (2013).

### Parents' Survey

We designed the 22-item *Parents of Children with LVAS Questionnaire* shown in Appendix A. Questions 1–18 were closed-response items and the remainder solicited parents' free responses. The questionnaire was developed using input from parents and colleagues in addition to using items used in previous surveys of parents (e.g., Danhauer et al, 2015). These 22 items were selected to explore the journeys of parents of children with LVAS and to inquire about critical milestones in the diagnosis and management of this disorder. Items were also included that would capture both parents' and their children's experiences. Earlier versions of the questionnaire, initially with 27 items, were constructed and pilot-tested on a small focus group consisting of three parents and two pediatric audiologists, which resulted in the final 22-item form. Five items were deleted because of redundancy or lack of relevance. Many of the items provided a "Not Applicable" option to account for situations in which parents did not have a particular experience involving their child with LVAS. Inclusion of this response should decrease the likelihood of parents responding negatively to a question simply because it was not relevant. The questionnaire elicited information about demographics, characteristics of the child's hearing loss, and specifics on the diagnosis/treatment of LVAS, in addition to families' experiences with healthcare providers. The questionnaire was designed to be completed in <10 min to maximize the likelihood of having families complete the survey.

We contacted the leaders of three LVAS support groups on Facebook.com and obtained permission to join and invite their members to participate in the survey. An invitation to participate in the study via a link to the questionnaire was posted on LVAS support groups via Facebook.com on September 25, 2014, and October 26, 2014. Descriptive statistics revealed trends in participants' responses. For the free-response items, the investigators individually coded parents' statements into categories, which were then prioritized by frequency of occurrence using a consensus approach.

### Audiologists' Survey

We designed the 18-item *LVAS Questionnaire for Audiologists* shown in Appendix B. The questionnaire was designed using similar steps described earlier for the parents' survey and elicited information about audiologists' demographics and their knowledge of, experiences with, and attitudes toward LVAS and treating patients with the disorder and their families. Three audiologists provided feedback on earlier versions of a 22-item survey; four items were eliminated because of redundancy and/or lack of relevance. Earlier versions

of the questionnaire were constructed and pilot tested on small focus groups of audiologists, which resulted in the final 18-item form. The audiologists were mid-career clinicians who had worked in a variety of settings; no efforts were made to control for experience with LVAS or work settings. Again, this questionnaire was designed to be completed in <10 min. An invitation to participate was emailed twice to 1,511 randomly selected audiologists listed in the American Academy of Audiology Membership Directory in November 2014. The email provided a link to [surveymonkey.com](http://surveymonkey.com) and audiologists self-selected their participation. This study was approved by the University of Oklahoma Health Sciences Center Institutional Review Board (OUHSC IRB 4629).

## RESULTS AND DISCUSSION

As with most surveys, it was difficult to determine details about the respondents who self-selected their participation in the study and whether they differed from those who elected not to complete the questionnaire. For example, it was not known if parents who responded were representative of their peers with children with LVAS. Similarly, details about the depth of knowledge about LVAS, interest in LVAS, or experiences with patients with LVAS and their families were not known for the audiologists who chose to participate, or whether they differed from those who did not respond.

Each finding is presented and discussed here in reference to items on the two surveys: items for parents are indicated by "P" with results shown in Appendix A and those for audiologists are referred to by "A" with results shown in Appendix B. For example, discussion of findings from the first item on the parent survey would be indicated by "P1."

### Parents' Survey

#### *Demographics and Experiences with LVAS*

The participants' data are reported here according to the numbers and the percent replying out of the total sample that responded to each item on the questionnaire. The percentages are rounded for ease of presentation. The maximum possible number of responses was 100, but in some cases, the number was smaller if participants failed to provide a response, or larger if the item allowed for more than one option. Although 100 parents responded, it was impossible to calculate the response rate because it was unknown as to how many parents actually saw the invitation to complete the survey. Respondents were predominantly mothers (95%; 95/100; P2), of which 63% (63/100; P3) were reportedly between 35 and 44 years of age.

Most children were reportedly diagnosed with hearing loss (86%; 76/88; P4) and LVAS (77%; 69/90; P6) by 5 years of age. The most common type of hearing loss reported was SNHL (73%; 66/90; P5); a few parents reported conductive (3%; 3/90; P5), mixed losses (14%; 13/90; P5) or were unsure (9%; 8/90; P5). The reported degree of hearing loss from the respondents in this study varied from normal to profound, with ~40% moderate, 20% severe, and 17% profound classifications (P8/9). These responses reflected the diversity of fluctuating and progressive hearing loss reported in the LVAS population (P10) (Lai and Shiao, 2004; Merchant et al, 2007). In addition, parents of children with LVAS need to be aware that their child's hearing loss has a greater likelihood of fluctuating and/or progressing than their peers with SNHL without LVAS. Indeed, parents with children who have LVAS in addition to a history of otitis media may need to know that low-frequency air-bone gaps may be due to LVAS and not an ear infection. Indeed, the vestibular aqueduct creates a third moveable window that may redirect air-conducted sound away from the cochlea, elevating air-conduction thresholds while simultaneously decreasing the impedance between the oval and round windows and decreasing bone-conduction thresholds. Zhou et al (2008) called air-bone gaps in LVAS patients the "third" labyrinthine window syndrome when observed in the presence of normal tympanometry, acoustic reflexes, and low-threshold vestibular evoked myogenic potentials.

Most of the parents reported that their child's LVAS was bilateral (82%; 74/90; P7). Figure 1 shows that parents reported that their child's LVAS was associated with hereditary (19%; 16/85; P10), fluctuating (48%; 39/82; P10), progressive (49%; 41/84; P10), and sudden (40%; 34/84; P10) hearing loss, but many parents were unsure. For example, some parents were unsure about whether their child's LVAS was hered-

itary (29%; 25/85; P10), fluctuating (13%; 11/82; P10), or progressive (23%; 19/84; P10).

These results agreed with the findings of Mori et al (2008) who conducted a retrospective chart review in a tertiary pediatric center in addition to a systematic review of the literature and combined data for 310 ears with LVAS with an average follow-up time of 4 years. They found that bilateral LVAS was six times more prevalent than unilateral cases. Moreover, they found that the hearing loss was stable in 67% of their patients' ears and progressive in 33%. Fluctuating hearing loss was found in 34% of stable ears, meaning that thresholds changed, but did not become more severe in degree, and in 50% of those with progressive hearing loss, or those that became consistently worse over time.

It is noteworthy that 41% (34/84; P10) of the parents associated their child's LVAS with "sudden hearing loss" and 28% (23/82; P10) with "head trauma," which agrees with the results of a systematic review of the literature with meta-analysis that found that one-third of patients with confirmed LVAS had a sudden drop in hearing thresholds due to head trauma (Noordman et al, 2015). Moreover, Noordman et al (2015) stated that patients with LVAS most at risk for threshold shifts after head trauma are those with preexisting fluctuating hearing loss. Furthermore, they recommended that parents should consider having their children with LVAS avoid activities that would place them at risk for head trauma (e.g., contact sports) and that they should consider the use of a helmet when their children are involved in activities such as ice-skating and skateboarding.

Regarding communication options, parents' responses indicated that most children's communication mode was aural/oral (94%; 83/88; P11), followed by Total Communication (18%; 13/74; P11), and manual communication or sign language (5%; 4/73; P11).

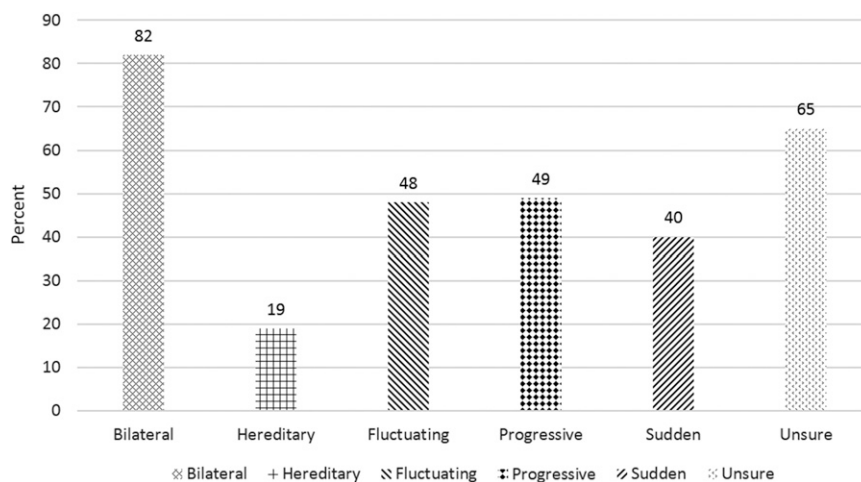


Figure 1. Characteristics of children's hearing losses. Reported characteristics of children's hearing losses from the parent survey.

Regarding technology, averaging across ears, 65% (56/86; P12) of the children wore digital hearing aids and 52% (45/86; P12) used FM systems. Interestingly, 21% (19/90; P12) used cochlear implants. The group data failed to reveal just how many children had unilateral or bilateral cochlear implants, or had received bimodal stimulation (i.e., wearing a hearing aid on the ear opposite to the cochlear implant). Thus, the individual data were analyzed, which showed that ~58% (11/19; P12) of cochlear implant users were bilaterally implanted, 37% (7/19) used bimodal stimulation; and only one child (5%; 1/19; P12) used a cochlear implant on one side, without any technology used in the opposite ear.

Generally, the parents in the present study reportedly felt that they had received adequate insurance or third-party coverage for items such as audiology [85%; 71/(90 – 8 Not Applicable responses); P13], primary care physician (PCP) [91%; 75/(88 – 6 Not Applicable responses); P13], pediatrician [93%; 77/(90 – 7 Not Applicable responses); P13], and otolaryngology [84%; 70/(88 – 5 Not Applicable responses); P13] visits. In the previous sentence, we subtracted the number of parents responding to “Not Applicable” from the denominator in calculations to obtain valid percentages of parents’ views on adequacy of coverage. Only 46% [38/(90 – 8 Not Applicable responses); P13] and 59% [20/(88 – 54 Not Applicable responses); P13] of the parents said they had received adequate support from insurers and other third-party payers for hearing aids and cochlear implants, respectively. Some parents may have interpreted the P13 survey item in two different ways. Some parents may have read the item and responded as to whether or not they had insurance instead of what they thought about the adequacy of the coverage that was provided.

### ***Experiences with Audiologists and Other Healthcare Professionals***

The survey allowed parents to rate how effective and helpful services and technologies were in managing their child’s LVAS. However, some of the services and technologies rated by parents for managing LVAS were not treatments for this condition. For example, 82% [73/(90 – 1 Not Applicable responses); P14] and 75% [64/(90 – 5 Not Applicable responses); P14] rated hearing evaluations and hearing aid evaluations, respectively, as being helpful in the treatment of LVAS, although they are not interventions for this condition. However, because the majority rated these two services as effective and helpful in treating LVAS, they may have felt that hearing evaluation and hearing aid evaluations contributed positively toward efforts in managing this disorder. Alternatively, 75% [63/(90 – 6 Not Applicable Responses); P14] and 73% [19/(90 – 64 Not

Applicable Responses); P14] rated hearing aids and cochlear implants, respectively, positively in managing their child’s LVAS. However, ~20–25% of parents were either neutral or did not believe that these audiologic services and technologies were helpful in the treatment of their child’s LVAS. It was difficult to determine the reasons for the ratings by the parents, but perhaps they may have been due to their frustration with and uncertainty about the audiometric findings associated with this disorder. For example, we hypothesize that parents may have been required to take their child to more hearing and hearing aid evaluations than they perceived were necessary. Consistent with the point made earlier, some parents may have reported that technologies and services were not helpful in managing LVAS because they did not view some of them as treatments. Similarly, we hypothesize that they may have had similar feelings about hearing aids which, even when fit appropriately, may have needed to be reprogrammed more than once to accommodate their child’s fluctuating and/or progressive losses.

Generally, parents of children with LVAS also interface with other healthcare professionals. For example, 73% [55/(90 – 15 Not Applicable responses); P14] and 70% [45/(90 – 26); P14] of the parents in the present study, respectively, reported that speech–language evaluations and speech–language therapy were helpful. Again, ~20–25% (P14) of parents were either neutral or did not believe that speech services were helpful in the treatment of their children’s LVAS. When queried about how helpful visits to PCPs and pediatricians were in the effective treatment of their child’s LVAS, only 36% [28/(90 – 13 Not Applicable responses); P14] and 39% [31/(90 – 10); P14] of the parents reported that visits to their PCPs and pediatricians, respectively, were helpful. Moreover, only ~13% [10/(89 – 13); P15], and 11% [9/(90 – 7); P15] of the parents respectively believed that their pediatricians and PCPs were well trained to treat children with LVAS.

The parents’ answers to the free-response questions inquiring how their experiences could have been improved with the pediatrician and/or PCP who diagnosed/treated their child’s LVAS provided support for these findings (P19 and P20). More than half of the free responses indicated that these healthcare professionals needed education about LVAS and Pendred syndrome. For example, one parent commented that the pediatrician was 90% (P19) certain that their daughter’s hearing loss was caused by fluid accumulation, which could be cured with ventilation tubes. Additionally, the parents believed that their pediatricians and PCPs (P19 and P20) needed more information about hearing aids and cochlear implants. One parent commented, “I cannot tell you how many times we’ve had to explain the difference between hearing aids and cochlear implants” to their child’s physician. This type of response was not

surprising and agreed with previous research that has similarly shown that some pediatricians and PCPs demonstrated deficits in current and accurate knowledge about cochlear implants (Carron et al, 2006; Mathews et al, 2009).

Generally, parents reported better experiences with and impressions of their otolaryngologists and audiologists than with their pediatricians and PCPs. For example, ~79% and 81% of the parents surveyed believed that their otolaryngologists (P15) and audiologists (P15), respectively, were well trained in treating children with LVAS. Nevertheless, as noted for pediatricians and PCPs, the parents' most frequent answer to the free-response questions regarding how their experiences could have been improved with the otolaryngologist and/or audiologist who diagnosed/treated their child's LVAS was that the professionals also needed more education. Although most of the parents agreed that audiologists were well trained in treating children with LVAS, the authors thought that it would be important to survey audiologists themselves about their knowledge of, experiences with, and attitudes toward serving children with LVAS and their families, which leads to the second survey in the present study.

## Audiologists' Survey

### *Demographics, Knowledge of, and Experiences with LVAS*

Similar to the parents' survey, the audiologists' survey data in the present study are reported here according to the percent and numbers replying out of the total sample that responded to each item on the questionnaire. Of the 1,511 audiologists who were invited to participate in this survey, 22 were "return-to-sender" and 18 were "out-of-office" responses, leaving 1,471 audiologists who could have read the invitation. Of the 1,471 invited to participate, 163 responded, but 17 students and two retirees were removed, leaving 144 participants. Thus, the response rate was ~10% (144/1,471). Low return rates in studies surveying healthcare professionals, such as physicians, have been discussed in the medical literature (e.g., Templeton et al, 1997; Kaner et al, 1998; Pirota et al, 1999; Hocking et al, 2006). Templeton et al (1997) suggested that a relatively low response does not necessarily affect the validity of the data collected. Furthermore, a low response rate does not necessarily mean that the sample is not representative of the greater population of audiologists, considering the way in which they were randomly selected from a directory. Reasons for not responding to the invitation to participate in the survey may have included paucity of time, lack of interest, inexperience with LVAS, and possibly not having received the invitation in the first place. Thus, 144 audiologists' responses were

analyzed and the maximum possible number of responses for each item on the questionnaire was 144; in some cases, the number was smaller if participants failed to provide a response. Percent of respondents was calculated and rounded for each item.

Respondents were predominantly women (86%; 126/144; A3), which is consistent with statistics from other surveys completed for the profession (ASHA, 2014). Using the filtering feature on surveymonkey.com to remove the retired audiologists from the sample, we found that most (65%; 94/144; A4) of the respondents in the present study were between 35 and 64 years of age, had an AuD degree (84%; 120/143; A2), and had worked primarily in a hospital (28%; 41/144; A5), otolaryngology clinic (25%; 36/144; A5), or private practice (24%; 34/144; A5) setting.

Approximately 6% (9/143; A6) of the respondents reported that they did not know or were not sure what LVAS was. Furthermore, 44% (61/140; A14) of the audiologists were unaware that LVAS accounted for between 5% and 15% (NIDCD, 2015) of all cases of pediatric SNHL, and 32% (46/143; A12) were unsure about the age at which LVAS was most commonly diagnosed. However, most of the audiologists knew that many LVAS patients are known to experience balance disorders (82%; 115/141; A11), and possible fluctuating (82%; 116/142; A11), progressive (88%; 125/142; A11), and/or sudden (80%; 112/139; A11) SNHL (92%; 129/141; A11). However, there was less certainty about whether patients with LVAS were known to experience hereditary (44%; 59/133; A11) or conductive hearing losses (54%; 74/136; A11). It is important for audiologists to know that LVAS may be caused by mutations in the SLC26A4 gene that is involved in the development of nonsyndromic LVAS, and is associated with Pendred syndrome (e.g., Yang et al, 2007) so that referrals for genetic counseling can be made. Our findings indicate that less than half (42%; 58/139; A13) knew that LVAS was associated with Pendred syndrome. Moreover, it is important for audiologists to know that patients with LVAS may have air-bone gaps, present on their audiometric threshold tests, but normal middle ear function. Indeed, air-bone gaps in the presence of normal tympanometry, presence of acoustic reflexes, and low threshold vestibular evoked myogenic potentials may signal a need for referral to an otologist for an evaluation for LVAS or other "third-window" lesions such as superior canal dehiscence (e.g., Zhou et al, 2008; Sone et al, 2016). In addition, although more than half of the audiologists knew that patients with LVAS have been known to experience tinnitus (65%; 91/140; A11), <30% (A11) knew that perilymphatic fistulas (e.g., Belenky et al, 1993) could also occur with this disorder. It is surprising that 63% (89/144; A7) of the audiologists reported that they had diagnosed or treated between 0 and 3 patients with LVAS in the past 5 years, considering

that LVAS accounts for 5–15% of all pediatric SNHL cases (NIDCD, 2015). For the LVAS patients whom the audiologists had seen, audiometric results (84%; 98/117; A8), computerized axial tomography (87%; 100/115; A8), and magnetic resonance imaging (69%; 73/106; A8) were key to the diagnosis of the disorder.

### ***Confidence in Knowledge of and Skills for Serving Patients with LVAS and Their Families***

Most of the audiologists were comfortable with their knowledge and skills concerning the audiologic diagnosis of LVAS (65%; 91/140; A9), treatment with hearing aids (73%; 102/140; A9), counseling patients and their families (69%; 96/140; A9), and making appropriate referrals (80%; 112/140; A9). However, only about 40% (56/139; A9) were confident about treatment with cochlear implants, which corresponds with the current controversy about when and how to implant children with LVAS. For example, many audiologists believe that the time to implant an ear with LVAS is when audiometric thresholds progress to the profound range and phonetically balanced word list word recognition scores fall <40%. Indeed, Ko et al (2013) found high speech perception and intelligibility performance in patients with LVAS when tested at 5 years postimplantation, regardless of when they were implanted. Moreover, there has been some concern over the use of sequential cochlear implantation in LVAS patients with incomplete partition malformations. Manzoor et al (2016) found that sequential cochlear implantation can be performed safely with patients with LVAS and incomplete partition malformations and that these children do as well as their peers without these anomalies. Moreover, the recommendation to implant a child with LVAS is also mediated by cochlear implant candidacy. Indeed, children with LVAS may not always be candidates for implantation or may have difficulty with insurance coverage when it comes to sequential implantation.

Most of the audiologists in the present study said they had learned about LVAS via clinical practice (74%; 104/139; A10), the Internet (51%; 64/125; A10), continuing education opportunities (51%; 66/129; A10), and graduate school (53%; 71/134; A10). Unfortunately, because question 5 stated “Check all that apply,” it is impossible to determine what the primary source of learning about LVAS was for the audiologists who obtained information from a variety of sources. It is not surprising that only about 20% (29/145; A16) of the audiologists believed that their graduate program had prepared them to help diagnose and treat LVAS, considering it is a disorder for which the etiology, diagnosis, and best treatment options have not been fully determined (Gopen et al, 2011). Most of the audiologists reported wanting to receive continuing education on LVAS via professional meetings (79%; 110/140; A17), the Internet (79%; 110/139; A17),

journal articles (83%; 116/140; A17), knowledgeable physicians (72%; 100/138; A17) colleagues (84%; 116/139; A17), and patients with LVAS and their families (66%; 92/139; A17).

When asked to provide their degree of agreement to the statement “Most pediatricians are knowledgeable about (LVAS),” only 4% of the audiologists (6/142; A18) either moderately or strongly agreed, which is consistent with the attitudes reported by the parents of children with LVAS and indicates a need for physicians’ continuing education on this topic. This is important because pediatricians serve as children’s Medical Homes and are supposed to coordinate early hearing detection and intervention efforts (Joint Committee on Infant Hearing of the American Academy of Pediatrics, 2013). Approximately 21% (30/143; A18) and 48% (69/143; A18), more than two-thirds (69%; 100/145; A18), of these audiologists either moderately or strongly agreed, respectively, that most otolaryngologists are knowledgeable about LVAS; and 3% (5/143; A18) and 44% (63/143; A18) of these audiologists either moderately or strongly agreed (69/145; A18) that their peers were knowledgeable about LVAS. Indeed, both parents and audiologists indicated that continuing education and consensus regarding the diagnosis and treatment of LVAS are needed within and across healthcare disciplines.

### **SUMMARY**

**L**VAS is a complex auditory disorder which is difficult to diagnose and manage. The results of the surveys presented here indicated that parents of children with LVAS undergo experiences not unlike those of parents of children with ANSD. Recall that Stroebel and Swanepoel (2014) found that parents of children with ANSD had poorer understanding of the disorder and greater uncertainty, and received conflicting information from healthcare professionals who did not give them enough time to ask questions compared with parents of children with SNHL. From parents’ and audiologists’ responses to our surveys, it seems that audiologists, pediatricians, PCPs, otolaryngologists, and speech-language pathologists are perceived as requiring additional and up-to-date knowledge and skills for serving patients with LVAS and their families.

Each healthcare professional plays a pivotal and unique role in the diagnosis, management, and support of patients with LVAS and their families. Indeed, audiologists are the hearing healthcare experts and should be the healthcare professional to explain possible treatment options for LVAS. Similarly, otolaryngologists need to have knowledge about the aspects of LVAS for the medical diagnosis and treatment of the disorder. Pediatricians and PCPs need to know to refer children who present with hearing loss and/or dizziness to otolaryngologists and/or audiologists for further evaluation.

Finally, speech-language pathologists need to know how to rehabilitate children with LVAS and be aware of how treatment needs may change as the condition progresses, for example. In addition, speech-language pathologists may be the first to notice changes in hearing sensitivity, warranting a referral for an audiologic evaluation. Indeed, an interprofessional approach is needed to serve children with LVAS and their families. Professional organizations should work together to develop consensus statements on the best practices for the diagnosis and treatment of patients with LVAS.

## CONCLUSION

A number of parents did feel positive about some of their interactions with healthcare providers and the information that they received about their child's LVAS, but additional support and explanation of ways to manage LVAS from their healthcare providers would be beneficial. In addition, healthcare providers who also need additional information on the topic and should collaborate for supportive and appropriate interprofessional care. In particular, physicians and audiologists, from whom parents seek guidance and support, appear to have limited knowledge and experience with LVAS. More continuing education is needed on LVAS across relevant healthcare professions. Audiologists should seek every opportunity to educate physicians on the topic of LVAS, when appropriate, by providing the literature with the audiologic report which could foster a team approach in managing children with LVAS.

## REFERENCES

American Speech-Language-Hearing Association (ASHA). (2014) *2014 Audiology Survey*. Rockville, MD: ASHA.

Asma A, Anouk H, Luc VH, Brokx JP, Cila U, Van De Heyning P. (2010) Therapeutic approach in managing patients with large vestibular aqueduct syndrome (LVAS). *Int J Pediatr Otorhinolaryngol* 74:474–481.

Belenky WM, Madgy DN, Leider JS, Becker CJ, Hotaling AJ. (1993) The enlarged vestibular aqueduct syndrome (EVA syndrome). *Ear Nose Throat J* 72:746–751.

Carron JD, Moore RB, Dhaliwal AS. (2006) Perceptions of pediatric primary care physicians on congenital hearing loss and cochlear implantation. *J Miss State Med Assoc* 47:35–41.

Clark JL, Roeser RJ. (2005) Large vestibular aqueduct syndrome: a case study. *J Am Acad Audiol* 16:822–828.

Cummings SR, Kohn MA, Hulley SB. (2013) Designing questionnaires, interviews, and online surveys. In: Hulley SB, Cummings SR, Browner WS, Grady DG, Newman TB, eds. *Designing Clinical Research*, 4th ed. Philadelphia, PA: Lippincott, Williams, & Wilkins, 223–236.

Danhauer JL, Johnson CE, Baker JA, Ryu JA, Smith RA, Umeda CJ. (2015) Will parents participate in and comply with programs and regimens using xylitol for preventing acute otitis media in their children? *Lang Speech Hear Serv Sch* 46:127–140.

Gopen Q, Zhou G, Whittemore K, Kenna M. (2011) Enlarged vestibular aqueduct: review of controversial aspects. *Laryngoscope* 121:1971–1978.

Hocking JS, Lim MS, Read T, Hellard M. (2006) Postal surveys of physicians gave superior response rates over telephone interviews in a randomized trial. *J Clin Epidemiol* 59:521–524.

Joint Committee on Infant Hearing of the American Academy of Pediatrics; Muse C, Harrison J, Yoshinaga-Itano C, Grimes A, Brookhouser PE, Epstein S, Buchman C, Mehl A, Vohr B, Moeller MP, Martin P, Benedict BS, Scoggins B, Crace J, King M, Sette A, Martin B. (2013) Supplement to the JCIH 2007 position statement: principles and guidelines for early intervention after confirmation that a child is deaf or hard of hearing. *Pediatrics* 131:e1324–e1349.

Kaner EF, Haighton CA, McAvoy BR. (1998) 'So much post, so busy with practice—so, no time!': a telephone survey of general practitioners' reasons for not participating in postal questionnaire surveys. *Br J Gen Pract* 48:1067–1069.

Ko HC, Liu TC, Lee LA, Chao WC, Tsou YT, Ng SH, Wu CM. (2013) Timing of surgical intervention with cochlear implant in patients with large vestibular aqueduct syndrome. *PLoS One* 8:e81568.

Lai CC, Shiao AS. (2004) Chronological changes of hearing in pediatric patients with large vestibular aqueduct syndrome. *Laryngoscope* 114:832–838.

Lin CY, Lin SL, Kao CC, Wu JL. (2005) The remediation of hearing deterioration in children with large vestibular aqueduct syndrome. *Auris Nasus Larynx* 32:99–105.

Manzari L. (2008) Enlarged vestibular aqueduct (EVA) related with recurrent benign paroxysmal positional vertigo (BPPV). *Med Hypotheses* 70:61–65.

Manzoor NF, Wick CC, Wahba M, Gupta A, Piper R, Murray GS, Otteson T, Megerian CA, Semaan MT. (2016) Bilateral sequential cochlear implantation in patients with enlarged vestibular aqueduct (EVA) syndrome. *Otol Neurotol* 37:e96–e103.

Mathews MR, Johnson CE, Danhauer JL. (2009) Pediatricians' knowledge of, experience with, and comfort levels for cochlear implants in children. *Am J Audiol* 18:129–143.

Merchant SN, Nakajima HH, Halpin C, Nadol JB, Jr, Lee DJ, Innis WP, Curtin H, Rosowski JJ. (2007) Clinical investigation and mechanism of air-bone gaps in large vestibular aqueduct syndrome. *Ann Otol Rhinol Laryngol* 116:532–541.

Mey K, Bille M, Caye-Thomasen P. (2016) Cochlear implantation in Pendred syndrome and non-syndromic enlarged vestibular aqueduct - clinical challenges, surgical results, and complications. *Acta Otolaryngol* 136:1064–1068.

Miyamoto RT, Bichey BG, Wynne MK, Kirk KI. (2002) Cochlear implantation with large vestibular aqueduct syndrome. *Laryngoscope* 112:1178–1182.

Mori T, Westerberg BD, Atashband S, Kozak FK. (2008) Natural history of hearing loss in children with enlarged vestibular aqueduct syndrome. *J Otolaryngol Head Neck Surg* 37:112–118.

National Institute of Deafness and Other Communication Disorders (NIDCD). (2015) Enlarged vestibular aqueducts and childhood hearing loss. <https://www.nidcd.nih.gov/health/enlarged-vestibular-aqueducts-and-childhood-hearing-loss>

Noordman BJ, van Beeck Calkoen E, Witte B, Goverts T, Hensen E, Merkus P. (2015) Prognostic factors for sudden drops in hearing level



- after minor head injury in patients with an enlarged vestibular aqueduct: a meta-analysis. *Otol Neurotol* 36:4–11.
- Pirotta M, Gunn J, Farish S, Karabatsos G. (1999) Primer post-card improves postal survey response rates. *Aust N Z J Public Health* 23:196–197.
- Saxena RC, Lehmann AE, Hight AE, Darrow K, Remenschneider A, Kozin ED, Lee DJ. (2015) Social media utilization in the cochlear implant community. *J Am Acad Audiol* 26:197–204.
- Shilton H, Hodgson M, Burgess G. (2014) Hyperbaric oxygen therapy for sudden sensorineural hearing loss in large vestibular aqueduct syndrome. *J Laryngol Otol* 128(1, Suppl):S50–S54.
- Sone M, Yoshida T, Morimoto K, Teranishi M, Nakashima T, Naganawa S. (2016) Endolymphatic hydrops in superior canal dehiscence and large vestibular aqueduct syndromes. *Laryngoscope* 126:1446–1450.
- Stroebel D, Swanepoel de W. (2014) Does parental experience of the diagnosis and intervention process differ for children with auditory neuropathy? *Int J Pediatr Otorhinolaryngol* 78:2210–2215.
- Templeton L, Deehan A, Taylor C, Drummond C, Strang J. (1997) Surveying general practitioners: does a low response rate matter? *Br J Gen Pract* 47:91–94.
- Valvassori GE, Clemis JD. (1978) The large vestibular aqueduct syndrome. *Laryngoscope* 88:723–728.
- Vijayasekaran S, Halsted MJ, Boston M, Meinzen-Derr J, Bardo DM, Greinwald J, Benton C. (2007) When is the vestibular aqueduct enlarged? A statistical analysis of the normative distribution of vestibular aqueduct size. *AJNR Am J Neuroradiol* 28:1133–1138.
- Wan LC, Guo MH, Qian YH, Liu SX, Zhang HZ, Chen SJ, Chen H, Gong J. (2009) Cochlear implant in patients with congenital malformation of the inner ear. *Nan Fang Yi Ke Da Xue Xue Bao* 29:2118–2121.
- Yang T, Vidarsson H, Rodrigo-Blomqvist S, Rosengren SS, Enerback S, Smith RJ. (2007) Transcriptional control of SLC26A4 is involved in Pendred syndrome and nonsyndromic enlargement of vestibular aqueduct (DFNB4). *Am J Hum Genet* 80:1055–1063.
- Zhou G, Gopen Q, Kenna MA. (2008) Delineating the hearing loss in children with enlarged vestibular aqueduct. *Laryngoscope* 118:2062–2066.

**APPENDIX A: Parents' Responses on the Parent of Children with LVAS Questionnaire**

1. This questionnaire is designed for parents of patients with LVAS. If you are a parent of a person with LVAS, we would appreciate it if you could answer this brief survey to the best of your ability. Thank you for your participation. Are you a parent of a person with LVAS?

Answer Options	Response Percent	Response Count
Yes	100.0	100
No	0.0	0
I am not sure	0.0	0

2. Are you

Answer Options	Response Percent	Response Count
Male	5.0	5
Female	95.0	95

3. What is your age?

Answer Options (years)	Response Percent	Response Count
18–24	1.0	1
25–34	17.0	17
35–44	63.0	63
45–54	17.0	17
55–64	2.0	2
65–74	0.0	0
>75	0.0	0

4. How old was your child when he/she was first diagnosed with a hearing loss?

Answer Options (years)	Response Percent	Response Count
1–5	86.4	76
6–10	10.2	9
11–15	3.4	3
16–20	0.0	0
21–25	0.0	0
26–30	0.0	0
31–35	0.0	0
36–40	0.0	0
>40	0.0	0

5. What type of hearing loss does your child have?

Answer Options	Response Percent	Response Count
Sensorineural	73.3	66
Conductive	3.3	3
Mixed	14.4	13
I am not sure	8.9	8

6. How old was your child when he/she was diagnosed with LVAS?

Answer Options (years)	Response Percent	Response Count
1–5	76.7	69
6–10	16.7	15
11–15	5.6	5
16–20	1.1	1
21–25	0.0	0

**APPENDIX A. Continued**

26-30	0.0	0
31-35	0.0	0
36-40	0.0	0
>40	0.0	0

7. In which ear is your child's LVAS?

Answer Options	Response	Response
	Percent	Count
Right	12.2	11
Left	4.4	4
Both	82.2	74
I am not sure	1.1	1

8. What was the degree of your child's hearing loss when he/she was initially diagnosed with LVAS?

Answer Options	No Loss	Mild	Moderate	Severe	Profound	I Am Not	Response
						Sure	Count
Right ear	8	12	34	21	14	0	89
Left ear	7	12	35	16	16	0	86

9. What is the degree of your child's hearing loss presently?

Answer Options	No Loss	Mild	Moderate	Severe	Profound	I Am Not	Response
						Sure	Count
Right ear	4	3	27	32	23	0	89
Left ear	5	9	24	22	24	0	84

10. Indicate whether your child's LVAS was associated with each of the following

Answer Options	Yes	No	Response	
			I Am Not Sure	Count
Hereditary hearing loss	16	44	25	85
Fluctuating hearing loss	39	32	11	82
Progressive hearing loss	41	24	19	84
Sudden hearing loss	34	34	16	84
Head trauma	23	39	20	82
Barometric changes	5	49	26	80
Other (please specify)				15

11. Indicate whether each of the following is your child's preferred method of communicating

Answer Options	Yes	No	Response
			Count
Manual (sign language)	4	69	73
Aural/oral (speech)	83	5	88
Total communication (speech and sign language)	13	61	74
Written	16	53	69
Another form of communication (please specify)			3

12. Indicate which of the following technologies your child has used as a treatment option for his/her LVAS (check all that apply)

Answer Options	Analog	Digital	Bone-	Cochlear	FM	None of	Response
	Hearing	Hearing	anchored				Count
	Aid(s)	Aid(s)	Hearing	Implant(s)	System(s)	These	
Right ear	9	61	1	14	48	7	88
Left ear	4	51	1	16	42	16	85
Other (please specify)							5

This document was downloaded for personal use only. Unauthorized distribution is strictly prohibited.

APPENDIX A. Continued

13. Indicate whether your child had insurance coverage that you believe adequately paid for each of the following services and technologies

Answer Options			I Am	Not	Response
	Yes	No	Not Sure	Applicable	Count
Hearing evaluations	71	11	1	7	90
Hearing aid evaluations	56	20	5	9	90
Hearing aids	38	42	2	8	90
Cochlear implant(s)	20	5	9	54	88
PCP visits	75	6	1	6	88
Pediatrician visits	77	5	1	7	90
Ear, nose, and throat physician visits	70	12	1	5	88
MRI(s)	52	12	2	22	88
CT scan(s)	63	9	0	17	89
Speech/language evaluations	47	18	7	17	89
Speech therapy	39	19	8	23	89
Other (please specify)					9

14. Indicate how effective that you believe that each of the following services and technologies was as a treatment for your child's LVAS

Answer Options	Very		Neutral	Moderately		Not	Response
	Unhelpful	Unhelpful		Helpful	Helpful		Applicable
Hearing evaluations	9	2	5	5	68	1	90
Hearing aid evaluations	11	1	9	7	57	5	90
Hearing aids	12	3	6	4	59	6	90
Cochlear implant(s)	2	0	5	0	19	64	90
PCP visits	14	7	28	10	18	13	90
Pediatrician visits	16	6	27	12	19	10	90
Ear, nose, and throat physician visits	5	8	12	21	38	6	90
MRI(s)	4	2	5	7	43	29	90
CT scan(s)	8	2	7	11	43	19	90
Speech/language evaluations	7	2	11	15	40	15	90
Speech therapy	7	2	10	6	39	26	90
Other (please specify)							2

15. Indicate how well trained that you believe that each of your child's following professionals was for treating patients with LVAS

Answer Options	Moderately		Neutral	Moderately		Not	Response
	Very Well	Well		Poorly	Poorly		Applicable
Audiologist(s)	46	26	9	5	3	0	89
Ear, nose, and throat specialist(s)	45	26	11	2	4	2	90
PCP(s)	10	6	19	12	29	13	89
Pediatrician(s)	9	9	23	15	27	7	90
Speech/language therapist(s)	19	21	18	11	3	18	90
Other (please specify)							4

16. Indicate how well that you believe that you are able to explain your child's LVAS to each of your following

Answer Options	Moderately		Neutral	Moderately		Not	Response
	Very Well	Well		Poorly	Poorly		Applicable
Family	33	44	10	2	1	0	90
Peers/friends	26	46	15	1	2	0	90
PCPs	25	47	16	1	1	0	90
Coworkers	20	36	13	4	3	13	89
Other (please specify)							0

This document was downloaded for personal use only. Unauthorized distribution is strictly prohibited.

**APPENDIX A. Continued**

17. Indicate whether your child has experienced each of the following

Answer Options	Yes	No	I Am Not Sure	Response
				Count
Vision issues	20	58	12	90
Balance issues	49	30	10	89
Falling	41	44	5	90
Head trauma	34	45	10	89
Headaches	32	42	16	90
Migraines	8	60	20	88
Migraines with aura (sensory signs/symptoms such as light flashes, blind spots, or tingling in the hands or face)	3	65	21	89
Sensitivity to light	19	53	17	89
Other (please specify)				9

18. Indicate how you believe that your experience with the ear, nose, and throat (ENT) specialist(s) who diagnosed/treated your child's LVAS could be improved.

Answer Options	Response Count
	82

19. Indicate how you believe that your experience with the pediatrician(s) who diagnosed/treated your child's LVAS could be improved.

Answer Options	Response Count
	82

20. Indicate how you believe that your experience with the PCP(s) who diagnosed/treated your child's LVAS could be improved.

Answer Options	Response Count
	82

21. Indicate how you believe that your experience with the audiologist(s) who diagnosed/treated your child's LVAS could be improved.

Answer Options	Response Count
	82

22. What is the one piece of advice that you would give to a parent of a child who was just diagnosed with LVAS?

Answer Options	Response Count
	82

This document was downloaded for personal use only. Unauthorized distribution is strictly prohibited.

**APPENDIX B: Audiologists' Responses to the LVAS Questionnaire for Audiologists****1. Are you an audiologist that has seen patients with LVAS?**

Answer Options	Response	Response
	Percent	Count
Yes	75.5	108
No	18.8	27
I am not sure	5.6	8

**2. I am**

Answer Options	Response	Response
	Percent	Count
An AuD-level audiologist who graduated from a residential program within the last 5 years (i.e., not before 2006)	31.2	45
An AuD student and have completed at least the first year of a residential program (i.e., second, third, or fourth year student)	0.0	0
An MA-level audiologist	9.0	13
An AuD-level audiologist with a degree from a distance-learning program	43.1	62
A PhD-level audiologist	7.6	11
A PhD/AuD-level audiologist	9.0	13
None of the above	0.0	0

**3. I am**

Answer Options	Response	Response
	Percent	Count
Male	11.9	17
Female	88.1	126
Answered question		143
Skipped question		1

**4. What is your age?**

Answer Options (years)	Response	Response
	Percent	Count
18–24	0	0
25–34	33.3	48
35–44	27.1	39
45–54	18.7	27
55–64	18.7	27
65–74	2.1	3
>75	0	0

**5. My primary work environment is**

Answer Options	Response	Response
	Percent	Count
Private practice	23.6	34
Hospital	28.5	41
Physician's office	2.7	4
ENT clinic	25.0	36
College/university	5.5	8
Primary or secondary school	8.3	12
Manufacturing facility	3.4	5
Military	0	0
Retired	0	0
Other	9.7	14

## APPENDIX B. Continued

6. Do you know what LVAS is?

Answer Options	Response	Response
	Percent	Count
Yes	93.7	134
No	4.2	6
I am not sure	2.1	3

7. How many patients with LVAS have you personally seen for diagnosis and/or treatment as an audiologist in the past 5 years?

Answer Options	Response	Response
	Percent	Count
0	24.6	35
1-3	38.0	54
4-5	16.2	23
6-10	12.7	18
11-15	2.1	3
>15	6.3	9

8. For the patients that you have seen with LVAS, indicate whether each of the following was used as the basis of the diagnosis

Answer Options	Yes	No	I Am Not		Response Count
			Sure	Response	
I have seen patients with LVAS	98	22	12	132	
Audiogram	98	8	11	117	
ABR	32	48	17	97	
ENG	18	51	19	88	
CT scan	100	4	11	115	
Magnetic resonance imaging	73	11	22	106	
Other (please specify)					13

9. Indicate how comfortable you are with your knowledge and skills concerning each of the following regarding LVAS

Answer Options	Very	Somewhat	Neutral	Somewhat	Very	Response
	Comfortable	Comfortable		Uncomfortable	Uncomfortable	Count
Audiologic diagnosis	66	31	15	10	18	140
Treatment via hearing aids	62	40	20	5	13	140
Treatment via cochlear implants	32	24	28	18	37	139
Counseling patients/families	62	34	14	15	15	140
Making appropriate referrals	84	28	9	8	11	140

10. Indicate whether you mainly learned about LVAS via each of the following

Answer Options	Yes	No	Response
			Count
Graduate school	71	63	134
Postgraduate training	51	78	129
Clinical practice	104	35	139
Continuing education	66	63	129
Internet	64	61	125
Training by colleagues in your work setting	58	68	126
Word of mouth	26	95	121
Other (please specify)			11

**APPENDIX B. Continued**

11. Indicate whether each of the following is known to be experienced by patients with LVAS

Answer Options	Yes	No	I Am Not	
			Sure	Response Count
Hereditary hearing loss	59	23	51	133
Fluctuating hearing loss	116	6	20	142
Conductive hearing loss	74	39	23	136
SNHL	129	1	11	141
Progressive hearing loss	125	0	17	142
Sudden hearing loss	112	12	15	139
Balance disorders	115	3	23	141
Vision difficulties	19	31	84	134
Migraine headaches	26	21	89	136
Tinnitus	91	3	44	138
Nausea/vomiting	58	16	64	138
Missing bony modiolus	28	14	92	134
Perilymphatic fistula	41	21	75	137
Other (please specify)				4

12. Indicate the age that you believe that LVAS is most commonly diagnosed

Answer Options	Response Percent	Response Count
0–5 months	0.0	0
6–11 months	6.0	8
1–2 years	12.6	18
3–4 years	14.7	21
5–6 years	14.0	20
7–8 years	9.8	14
9–10 years	4.2	6
>10 years	7.0	10
I am not sure	32.2	46

13. Based on your experience, indicate whether each of the following is true about patients with LVAS

Answer Options	Yes	No	I Am Not	
			Sure	Response Count
Hearing sensitivity will stay the same	6	118	15	139
Hearing sensitivity will get better	21	103	16	140
Hearing sensitivity will get worse	123	2	16	141
LVAS is autosomally inherited	33	10	96	139
LVAS is genetically related to Pendred syndrome	58	6	75	139
LVAS affects males more frequently than females	20	19	100	139

14. Indicate what percentage of pediatric cases of SNHL may be attributed to LVAS

Answer Options	Response Percent	Response Count
<15%	43.6	61
>16%	12.9	18
I am not sure	43.5	61



## APPENDIX B. Continued

15. Based on your experience, indicate whether patients with LVAS usually perform well with each of the following

Answer Options	Yes	No	I Am Not	
			Sure	Response Count
Manual communication (sign language)	53	17	64	134
Oral/aural communication	103	1	35	139
Total communication	73	11	53	137
Cued speech	47	14	74	135
Traditional hearing aids	102	8	30	140
Bone-anchored hearing aid	22	56	55	133
Unilateral cochlear implants	74	3	60	137
Bilateral cochlear implants	72	1	63	136
Bimodal stimulation (i.e., hearing aid on one side and cochlear implant on the other)	74	2	61	137

16. Do you believe that your graduate program prepared you adequately to help diagnose and treat patients with LVAS?

Answer Options	Response Percent	Response Count
Yes	16.1	23
No	73.4	105
I am not sure	10.5	15

17. Indicate whether you would like to receive additional continuing education information about LVAS via each of the following

Answer Options	Yes	No	I Am Not	
			Sure	Response Count
Professional meetings	110	25	5	140
Internet	110	21	8	139
Journal articles	116	21	3	140
Physicians with expertise on this topic	100	31	7	138
Audiologists with expertise on this topic	116	19	4	139
Patients/families living with LVAS	92	33	14	139
Other (please specify)				7

18. Please indicate your degree of agreement with the following statements

Answer Options	Strongly Agree	Moderately			Strongly Disagree	Response Count
		Agree	Neutral	Disagree		
"Most pediatricians are knowledgeable about LVAS"	2	4	29	58	49	142
"Most otolaryngologists (ENTs) are knowledgeable about LVAS"	30	69	29	12	3	143
"Most audiologists are knowledgeable about LVAS"	5	63	34	37	4	143