Abnormalities of the vestibular aqueduct (EVA) is a significant cause of sensorineural hearing loss (SNHL) in infants and children. Multiple studies have verified that this anatomical malformation, even when not associated with other inner ear abnormalities, predisposes patients to significant hearing loss [1-2].

The vestibular aqueduct (VA) is a bony canal that traverses the otic capsule from the medial wall of the vestibule extending towards the posterior surface of the petrous bone. In addition to a vein and an artery, the vestibular aqueduct contains the endolymphatic duct which connects the endolymphatic sac with the vestibular labyrinth [. ] The anatomist Carlo Mondini was the first to describe the enlargement in the internal and external vestibular aqueduct aperture during an isolated dissection of the temporal bone in 1791 [4]; however, it wasn’t until 1978 that the clinical feature of hearing loss was linked to EVA by Valvassori and Clemis. Their work culminated in one of the largest radiologic assessments of this malformation. In addition to coining the term large vestibular aqueduct syndrome (LVAS), they also defined the radiographic parameters of the syndrome stating that LVAS describes an aqueduct with an anterio-posterior diameter of 1.5 mm or more on polytomography, and subsequently computed tomography [5]. Though subsequent studies have generated other criteria to define enlargement of the vestibular aqueduct, consensus around the definition by Valvassori and Clemis remains.

While it is known that HL is progressive in patients with syndrome EVA, such as those with a mutation in the SLC26A4 gene, little is known about hearing loss progression in patients with non-syndromic EVA. In addition, causes of progression of hearing loss in these patients have been poorly elucidated in current literature. Most studies have presented anecdotal evidence of triggers for hearing loss, including Valsalva, minor ear injury, scuba diving and even the common cold [6]. The importance of identifying patients with EVA who will progress to significant hearing loss cannot be over-emphasized. Such identification, if done early in the patient’s life, could lead to significant improvements in the clinical care, and management of these patients.

RESULTS

Females accounted for 62% of this cohort. 53 (56%) of the patients had bilateral EVA, while 21 (22%) and 20 (21%) of patients had left and right EVA, respectively. Notably 62/66(97%) of patients had a cochlear abnormality associated with their EVA. This number is similar to previously reported studies [7]. The median age at diagnosis (defined as time to diagnostic radiography) was 4.7 years (range 0.1 - 20.5 years). The median age at time of the first available audiogram was 11.4 years (0.5 - 20.8 years). The mean follow-up time was 8.6 years (0.15-20.8 years).

The baseline degree of hearing loss could be classified in 147 ears. HL was mild in 33 (22.4%) ears, moderate in 51 (34.7%) ears, severe in 30 (20.4%) and profound in 9 (6.1%). 13 ears had a conductive hearing loss, 17 (22.1%) had a sensorineural hearing loss, and 47 (61%) ears had a mixed hearing loss. The majority of air-bone gaps were detected at the lower frequencies in the baseline audiogram. However there was no significant correlation between the frequency of ABGs, the presence of a mutation in the SLC26A4, or laterality of EVA (P>0.05).

Individuals with a SLC26A4 mutation had a non-significant difference in presence of air-bone gaps, or the degree of HL, when compared with individuals without the mutation (p=0.1). In addition, in the absence of any SLC26A4 mutations individuals with bilateral EVA did not have a significantly different degree of hearing loss than individuals with unilateral EVA.

DISCUSSION

This study objective was to define hearing loss in patients with EVA as well as determine prognostic factors for its progression. While some studies have proposed that hearing in these patients progresses in a steady step-wise fashion [8], our cohort of patients did not experience a significant decline in hearing over an average of 8.6 years of follow-up, although they had frequent fluctuations. It is possible that previous studies were of a shorter duration, and identified fluctuations of hearing in these patients as actual decreases in hearing. This analysis may be limited by the small population of patients with a SLC26A4 mutation included in this study. The presence of mutations in the SLC26A4 gene did not significantly affect the progression of HL.

CONCLUSIONS

A significant change in hearing was not demonstrated in patients with unilateral or bilateral EVA during the mean follow-up time of 8.6 years. The presence of mutant SLC26A4 alleles did not have a significant association with the severity or type of hearing loss seen.

REFERENCES


CONTACT

Bunni Ajose-Popoola, B.A.
Harvard Medical School, 24 Shattuck St. Boston, MA, 02115
oaa5@hms.harvard.edu 925-207-7030