Background

Enlarged vestibular aqueduct (EVA) is the most common congenital malformation of the inner ear. It is diagnosed radiographically and can account for up to 32% of the hearing loss in children who undergo temporal bone computed tomography (CT). First described by Valvassori and Clemis in 1978, EVA was defined as >1.5mm dilation at the midpoint of the vestibular aqueduct or ≥2mm at the opisthocranion1. It has been associated with up to 15% of all pediatric causes of sensorineural hearing loss (SNHL) and the unilateral/bilateral ratio is approximately 2:1.4. Clinically critical aspect of EVA is its association with hearing loss progression. Deterioration of hearing, frequently concomitant with EVA, is a critical aspect of this disease process that warrants continued investigation and careful surveillance so as to limit its detrimental effects.

Thus, those who are diagnosed with EVA merit close investigation and careful surveillance so as to limit its detrimental effects.

Methods

Electronic medical records were reviewed using EMERSE to locate 172 patients (343 individual ears) with EVA who were under the age of 18 at the time of diagnosis.

• Each ear was treated as a separate subject.
• All diagnoses of EVA were made via radiographic imaging.

Exclusion criteria included:
• Cochlear malformations more severe than a Mondini/Incomplete Partition (IP-2) type cochlea.
• Any subject with a diagnosis of cochlear nerve deficiency.
• Any adult EVA subject who did not have childhood audiograms available to assess hearing loss progression over time.
• Lack of acoustic hearing history through the age of 12 or cochlear implantation.

A total of 259 ears remained in the analysis following review for exclusion. Serial audiometric data and CI speech perception outcomes were studied for each individual ear.

An individual ear was considered to have met CI candidacy criteria once the 3 frequency pure tone average (PTA) met or exceeded 75 dB HL.

Conclusions

Approximately 62% of Full EVA Cohort met audiometric criteria for implantation at the time of presentation to our cochlear implant center. Of the remaining subjects over 1/3 ultimately progressed to meet audiometric criteria for cochlear implantation before the age of 13. With a total of 78% of ears reaching severe-to-profound thresholds prior to the teen years, this strongly suggests that patients with EVA who are identified with hearing loss in childhood should be thoroughly counseled on the likelihood of progression and closely monitored for cochlear implant candidacy. The majority of the EVA population that reach cochlear implant candidacy do so prior to reaching adulthood.

References