Enlarged Vestibular Aqueduct: Hearing Progression and Outcomes in Cochlear Implant Patients

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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)[1]. 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 operculum[2-4]. It has been associated with up to 15% of all pediatric causes of sensorineural hearing loss (SNHL) and the bilateral:unilateral ratio is approximately 2:1[4,5]. A 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 audiologic observation to identify any hearing loss and its progression. Audiologic intervention in this population varies depending on the severity of the hearing loss and ranges from traditional amplification to cochlear implantation. Cochlear implantation (CI) has been shown to be of optimal hearing benefit in these patients with severe-profound sensorineural hearing loss[6,7]. While there are existing studies that describe the incidence of progression in this population, data regarding the amount of progression are lacking. Characterizing the prevalence of cochlear implant candidacy in children with EVA provides information to clinical care providers to guide counseling and intervention decisions.

Aim

This study aims to investigate the rate of hearing loss progression and the incidence of cochlear implant candidacy in children with EVA.

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.

The 259 ears were considered the "Full cohort." Of this full cohort, 160 ears met candidacy at the time of presentation to the center. The remaining 99 ears were reviewed separately as a "Potential progression cohort."

Results

Full Cohort

- 196 ears (76%) met candidacy prior to age 13.
- 131 ears (51%) actually received implants in childhood. This is only 67% of the potential candidate pool, indicating that this population may be underserved.
- Figure 1 illustrates the percentage of children with unaided thresholds that could still potentially be appropriately fit with a hearing aid by age in years.
    - Less than 30% of the participants had a PTA <75 dB HL by the age of 12.
- The mean age of implantation for the total EVA group was 7.22 years.
    - In general, speech perception outcomes in this population are good, with a mean highest CNC word score of 76.22%. Scores above 70% were achieved by 75% of the participants (Figure 2).

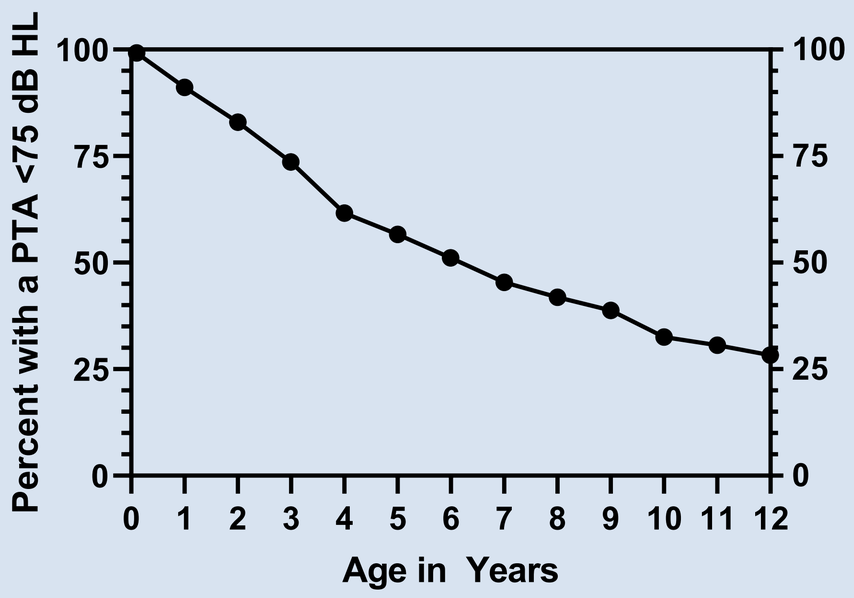

Figure 1: Percentage of children not yet meeting implant candidacy by age within the full cohort.

- Of the full cohort, a total of 62 ears that met candidacy for CI were not implanted.
- 45 were evaluated in clinic and of those, a CI was recommended for 35 (78%). The reason for not implanting in 34 (97%) of cases was parent/child decision. The one remaining was an insurance denial of bilateral implantation.

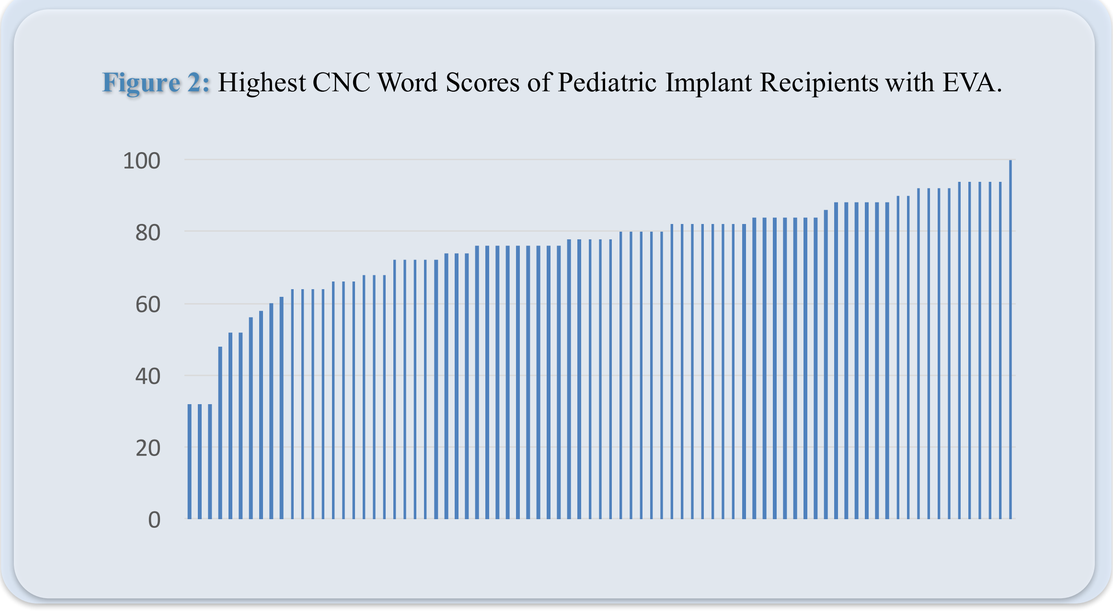

Figure 2: Highest CNC Word Scores of Pediatric Implant Recipients with EVA.

Potential progression cohort.

- 49% of this group experienced a PTA shift of ≥ 15dB HL prior to the age of 13.
- 37% of the potential progression cohort reached candidacy prior to their teen years. Available data indicated that an additional 5 ears experienced further progression and implanted as teens for a total of 42% of the progression cohort receiving cochlear implants as children thus far.

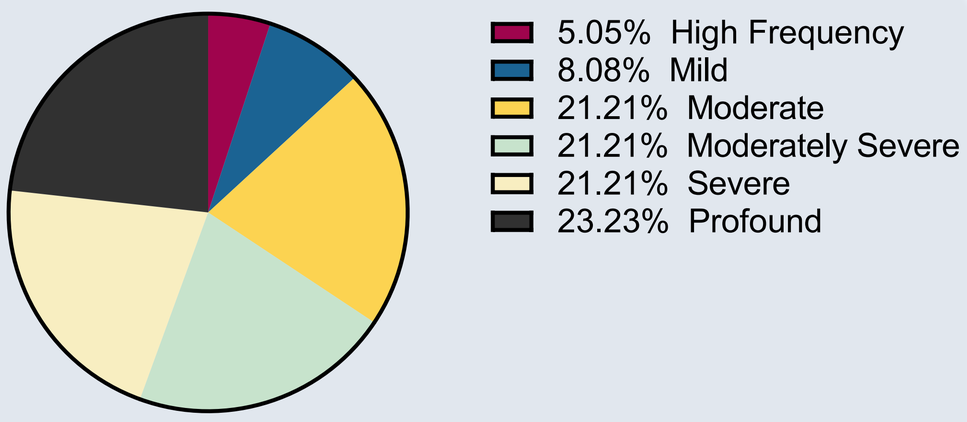

Figure 3: PTA Range of Potential Progression Cohort by Age 12.

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.

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Disclosure(s)

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- EMERSE was utilized to find patients in EPIC diagnosed with EVA (see separate citation in References section).