Introduction

Pediatric sensorineural hearing loss (SNHL) is a common clinical problem that carries great risk for speech, social, and cognitive dysfunction. Clinically significant congenital SNHL is present in 1,300 children by age 4. Enlarged vestibular aqueduct (EVA) is the most common congenital inner ear finding associated with congenital or childhood onset HL. Patients with EVA have a marked tendency for deterioration in hearing, with progression seen in up to 46% of patients. There is weak evidence to suggest that HL seen in some patients is linked to minor head trauma and from this is generally recommended that children with EVA avoid contact sports to prevent progression. However, there is a paucity of data to support this conclusion. Standard practice is to obtain imaging in cases of congenital or childhood-onset SNHL, among other reasons, to rule out EVA, despite the risks associated with radiation exposure and general anesthesia in pediatric patients. Therefore, we attempt to better delineate the progression of HL with EVA and its association with trauma. Additionally, we analyze the cost of obtaining early and late imaging studies.

Methods

Systematic Review
Published search was performed. Exclusion criteria were as follows:
1. Any study with < 5 patients; 2. Any study without audiometric data; 3. Any study without information on progression of hearing loss; 4. Any non-human study; 5. All review papers; 6. Any study reported in language other than English. The remaining articles were subject to full-length review and individual-level data extracted for meta-analysis (Figure 1). In addition, the reference lists for each included article were reviewed and no other articles fulfilling inclusion criteria were noted.

Meta-analysis
Data were converted to ears for meta-analysis. Data were extracted in 6 categories +/- progression, +/- unknown etiology. Information of definition or progression and definition of head trauma also included. Study-level data was combined to generate percentages for comparison. Performance, reporting, and analysis of this systematic review and meta-analysis has been done in accordance with PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) guidelines.

Results

Two studies reported full data on head trauma and progression in subjects with EVA. Aggregate data (n=72) did not show any statistically significant association between progression and head trauma (Figure 2).

Discussion

By itself, EVA portends a high risk of progressive hearing loss, with 41.3% of patients in this meta-analysis experiencing progressive hearing loss. In the only two studies where history of head trauma was assessed and reported for all subjects, there was no association between head trauma and progressive hearing loss. The typical reason for obtaining early imaging - to identify EVA - which could change management and counseling regarding head trauma, is not justified. Therefore, we recommend delayed MRI after Connexin 26 and CMV newborn bloodspot testing to eliminate unnecessary radiation and anesthesia exposure and increase the diagnostic efficiency of imaging.

Conclusions

- There is a 41.3% risk of progressive hearing loss in EVA.
- The association between head trauma and progressive hearing loss is not supported by the literature.
- When possible, delayed, non-sedated MRI should be used to evaluate these patients, as earlier diagnostic testing does not significantly change management.

References