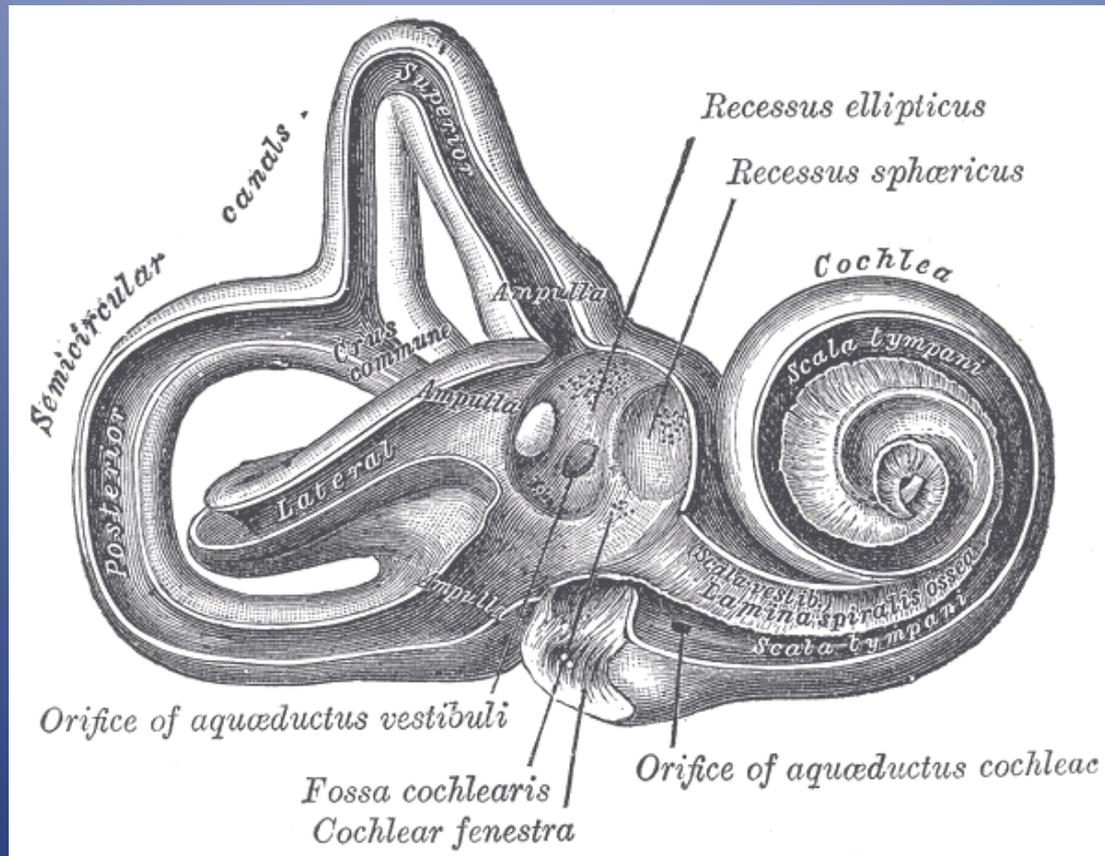


Enlarged Vestibular Aqueduct and Pediatric Hearing Loss



Pediatric Hearing Loss

- In the U.S., an estimated 2.7 per 1000 children are diagnosed with hearing loss before age **five** (Morton & Nance, 2006).
- Early identification of hearing loss (≤ 6 months of age) helps maintain age-appropriate language, social, and emotional development **regardless of mode of communication** (Yoshinaga-Itano et al., 1998)

Risk Factors for Pediatric Hearing Loss

- The Joint Committee on Infant Hearing (JCIH) recommends a list of risk factors to encourage audiological monitoring of children even if they pass newborn hearing screening.
- These risk factors include:
 - Caregiver concern
 - Family history of hearing loss
 - NICU stay > 5 days
 - In-utero infections
 - Craniofacial anomalies
 - Certain syndromes and associated physical findings
 - Neurodegenerative disorders
 - Postnatal infections
 - Head trauma
 - Chemotherapy



Enlarged Vestibular Aqueduct

- The vestibular aqueduct contains the endolymphatic duct and sac.
- Enlarged Vestibular Aqueduct (EVA) is a congenital anomaly in which the vestibular aqueduct is larger than 1.5 mm in diameter (www.nidcd.nih.gov/health/hearing/eva.htm).
- Can only be diagnosed by imaging such as CT scan or MRI.
- Is diagnosed in 12% of children with hearing loss by the age of four (Morton & Nance, 2006).
- Is commonly seen in patients with Pendred Syndrome (Luxon et al., 2003) or nonsyndromic mutations of SLC26A4 (Albert et al., 2006).

EVA and Hearing Loss

- 40% of children with EVA will develop profound SNHL (Mafong et al., 2002).
- Hearing loss is commonly progressive and/or fluctuating, and can be late-onset (Madden et al., 2003; Zalzal et al., 1995).
- Seen with other inner ear malformations in 41-88% of cases (Atkin et al., 2009).

Unilateral vs. Bilateral EVA

- Less is known about how EVA differs in cases with unilateral and bilateral hearing loss
- Researchers have found that patients can have bilateral EVA but unilateral hearing loss. EVA can affect each ear differently even within the same patient (Atkin et al., 2009; Bamiou et al., 1999).

The Current Study



- This project examined the etiology of hearing loss in the pediatric population at the Lions Children's Hearing Center
 - 102 families provided consent to be in the database
 - Reviewed electronic medical record to abstract data about hearing thresholds, progression, risk factors, age of identification, and results from CT scan and genetic tests.
- Specifically, this thesis focuses on the presence of EVA within this group

Research Questions

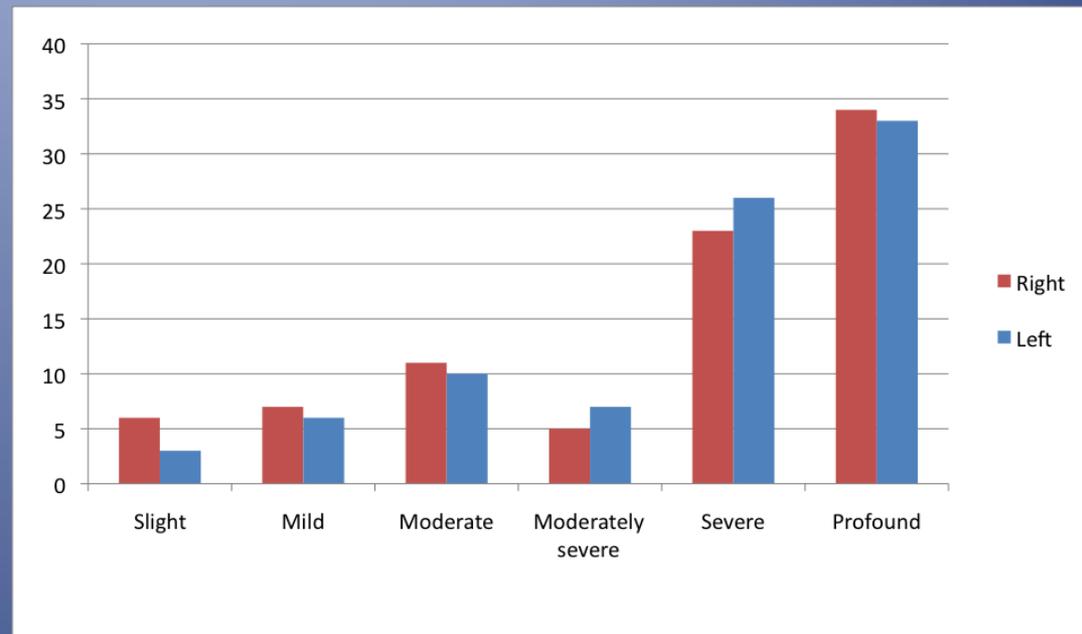
- What is the incidence of EVA in the cohort with unilateral hearing loss vs. bilateral hearing loss?
- What is the age of identification of hearing loss in the two groups (i.e., unilateral vs. bilateral hearing loss) with EVA?

Hearing Thresholds of the Study Group

- Of the initial study group, 97 children had hearing loss
 - 85 had bilateral loss
 - 12 had unilateral loss

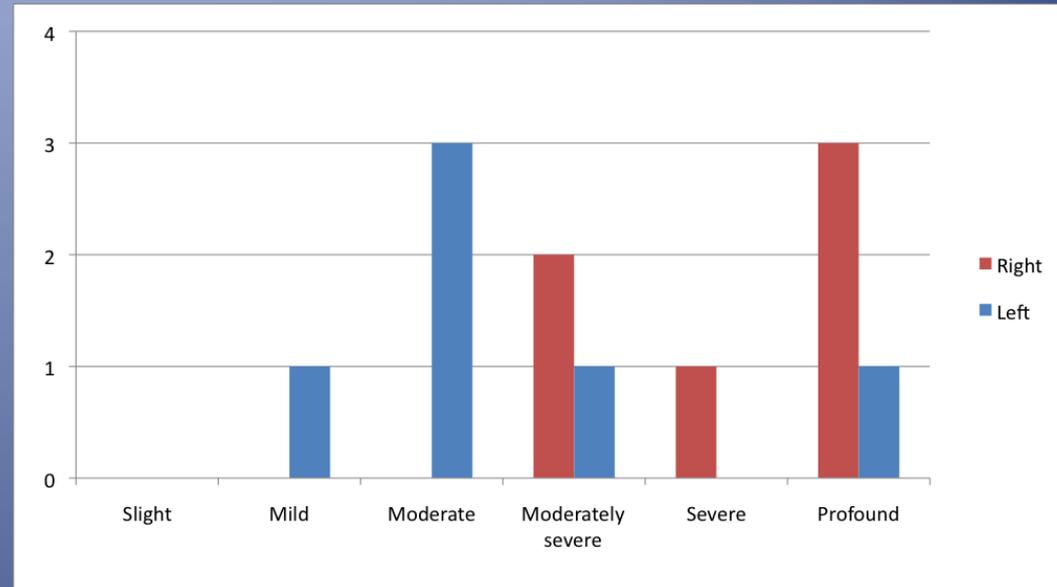
Bilateral hearing loss

- 85 children
- Majority (75%) severe or profound loss
- 80 SNHL, 2 conductive, 3 mixed
- 3 with auditory neuropathy/dys-synchrony
- 35% progressive

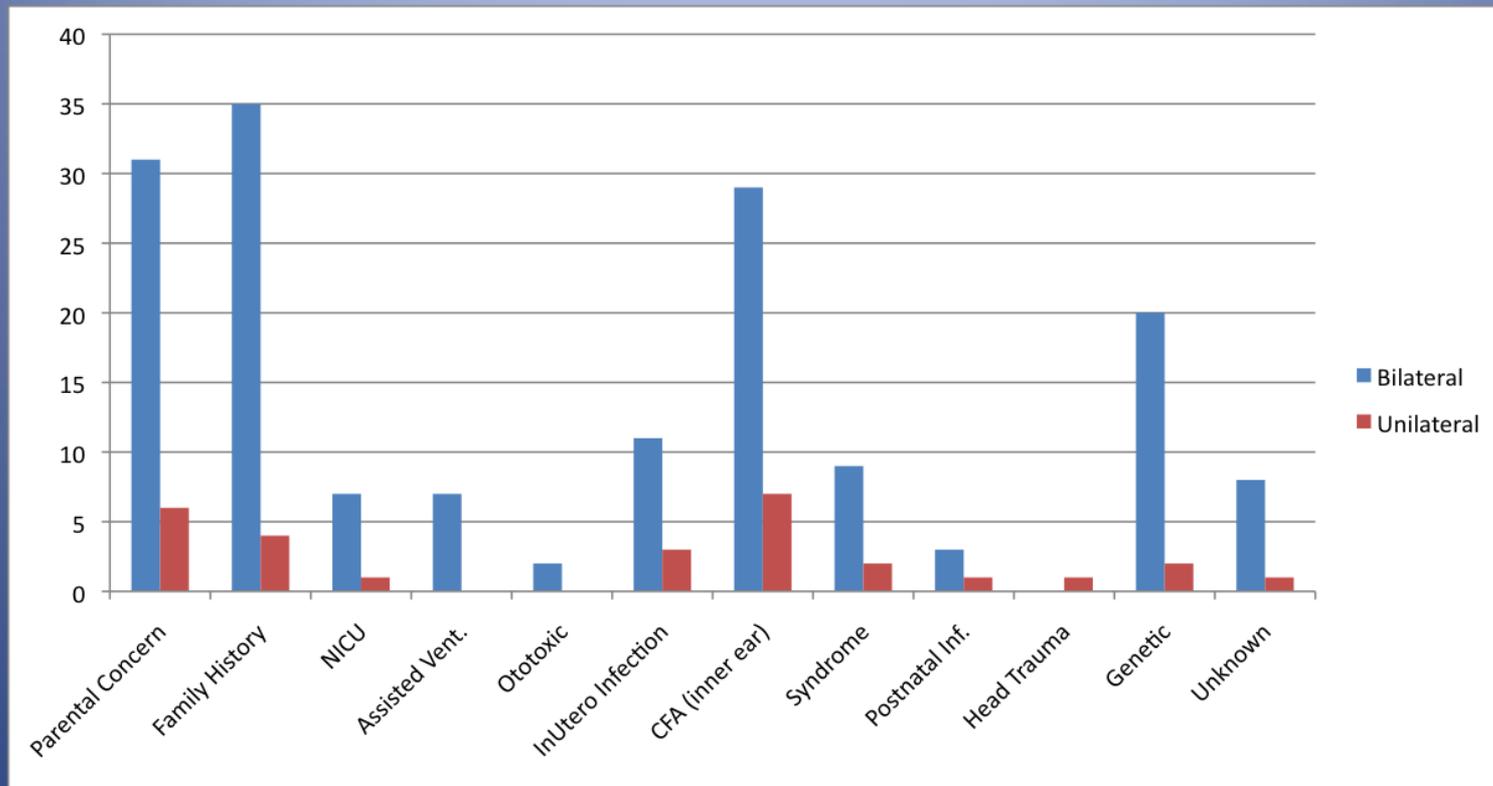


Unilateral hearing loss

- 12 children
- 5 moderate/moderately-severe, 5 severe/profound
- Thresholds generally worse for right-sided losses
- 7 SNHL, 1 conductive, 4 mixed
- 1 auditory neuropathy/dys-synchrony
- Progressive for 2 children (17%)

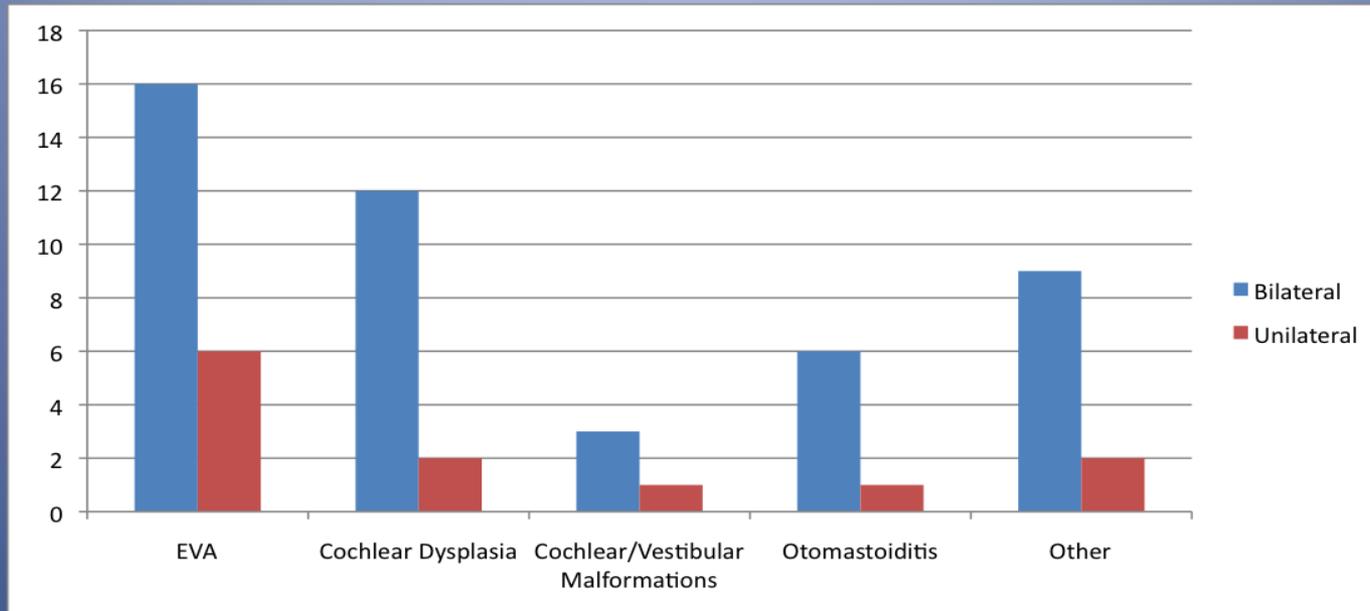


JCIH Risk Factors



- At least one risk factor was identified in all but 11 children
- Some children had more than one risk factor
- Two cases had incomplete documentation of medical history

Temporal Bone Anomalies and EVA

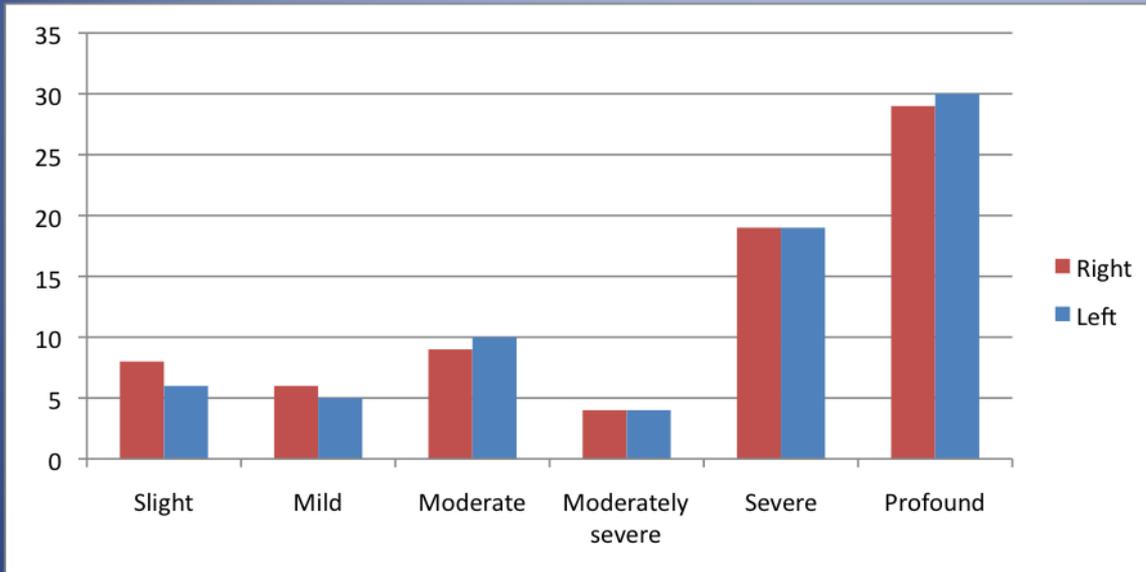


- Cranio-facial malformation identified in 39 (40%)
- EVA was isolated finding for nine children
- 13 children with EVA had at least one other malformation
 - Most common: cochlear dysplasia (12, 54.5%)

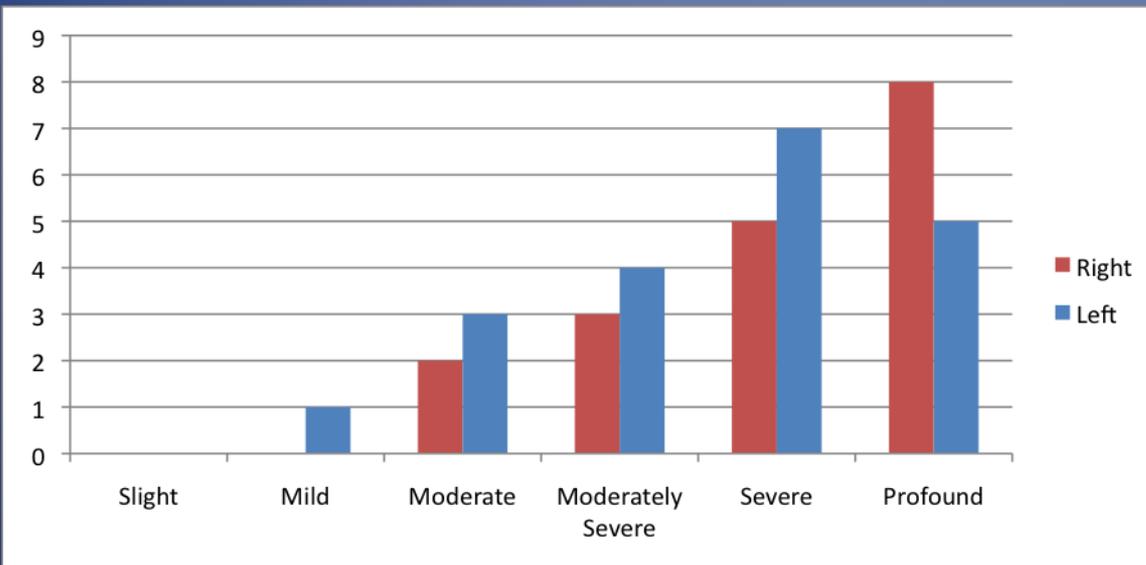
Hearing Loss with EVA

- Majority with EVA (64%) had severe or profound hearing loss
- Similar distribution but more concentrated in severe/profound loss compared to children without EVA
- 45% with EVA progressive, compared to 29% without

Hearing Loss with EVA



Hearing thresholds no EVA



Hearing thresholds with EVA

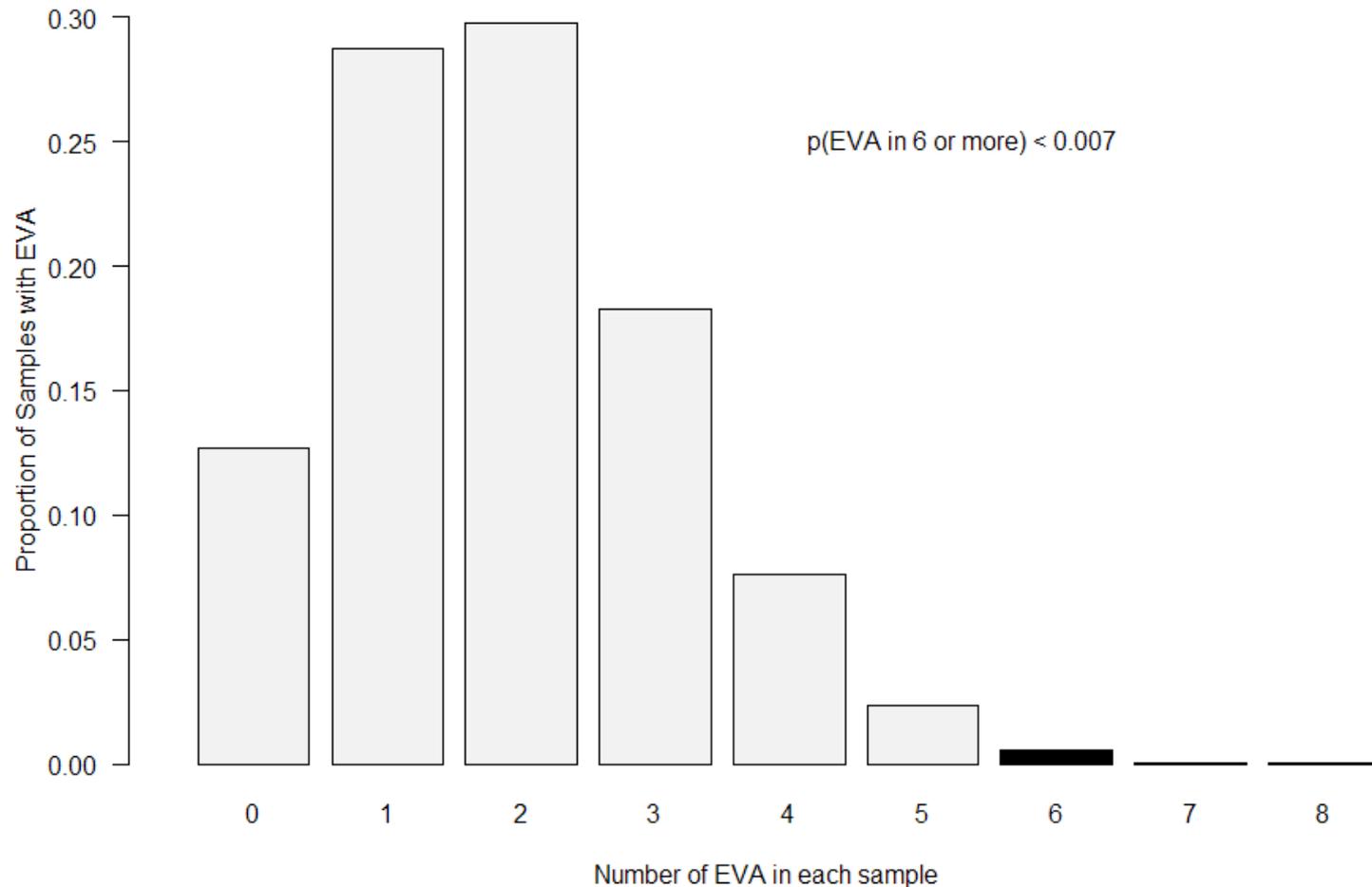
Bilateral and Unilateral hearing loss and EVA

- EVA was seen in 50% of children with unilateral hearing loss (6/12) versus 18.8% (16/85) of children with bilateral hearing loss
- EVA is reportedly twice as likely to cause bilateral than unilateral hearing loss (Tharpe & Sladen, 2008)
 - In our sample population, 2.6 times more bilateral than unilateral hearing loss with EVA
 - But, hearing loss in general is twice as likely to be bilateral (Lieu, 2010; Morton & Nance, 2006)
 - Unclear if rate of EVA is truly more commonly bilateral or if simply differs in accordance to general rates of each

Bilateral and Unilateral hearing loss and EVA

- We performed a jackknife re-sampling procedure to compare rate of EVA for children with bilateral and unilateral hearing loss
 - Compares small group to large group by taking repeated samples from larger group and comparing possible outcomes
- 0.007% probability that random sample of 12 children with bilateral loss would have 6 cases of EVA
- EVA is significantly more common in our group with unilateral loss

Bilateral and Unilateral EVA

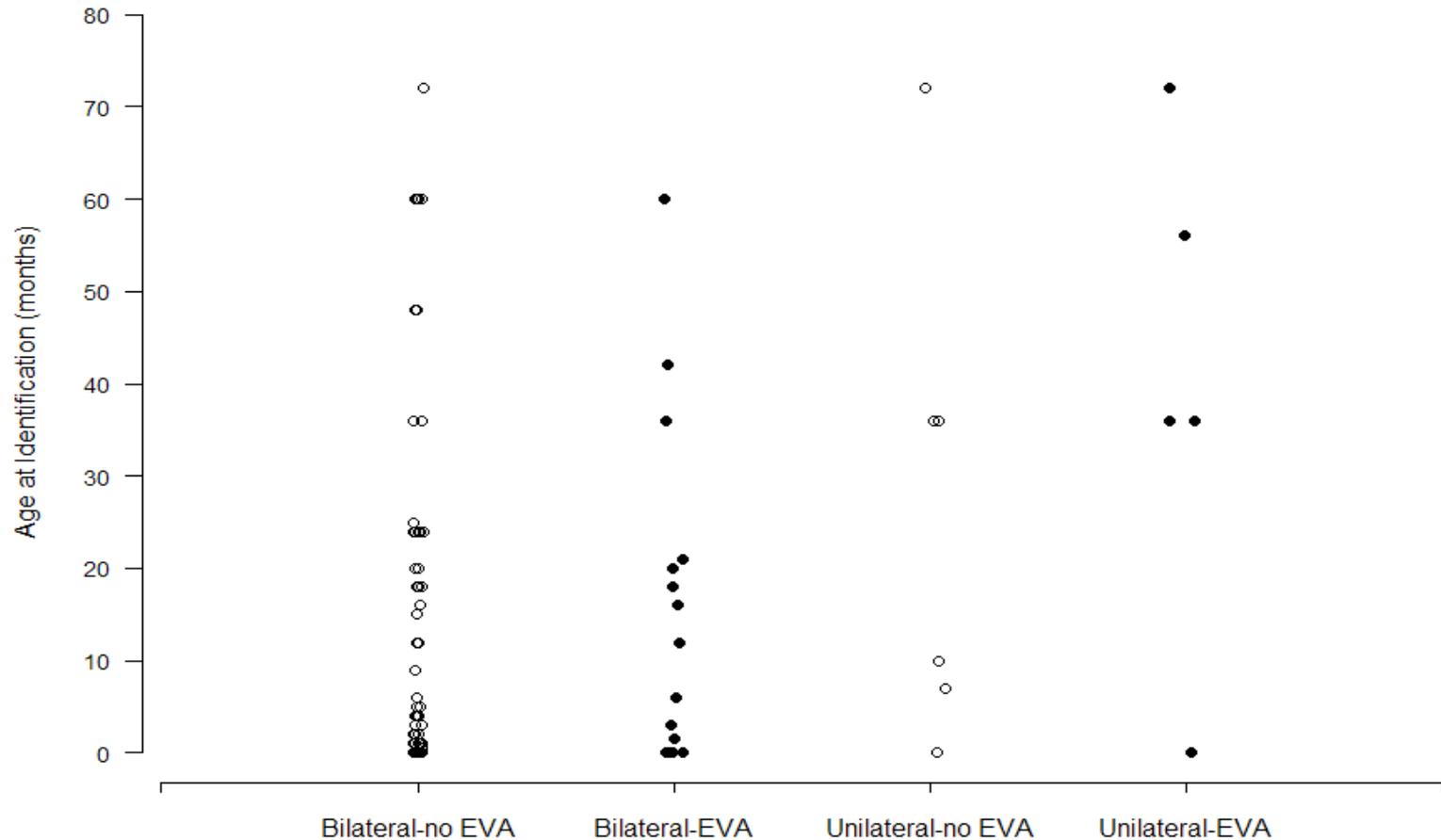


Jackknife re-sampling results

Age of Identification of Hearing Loss

- Examined the age of identification for children with and without EVA for both bilateral and unilateral hearing loss.
- As EVA is commonly progressive and/or late-onset, hearing loss may not be present when newborn hearing screening is performed.
- 50% of children with unilateral loss without EVA were identified at around 10 months of age
- Children with unilateral loss with EVA all identified at 36 months or later, except one patient with microtia.
- Some of these children were born before newborn hearing screening programs were mandated in this state

Age of Identification of Hearing Loss



Distribution of age of identification (months) per group. Data are jittered slightly for clarity.

Conclusion and Recommendations

- EVA was the most common cause of hearing loss for children in this database
- Hearing loss with EVA was most commonly severe or profound, and progressive for 45% of cases
- Statistically significant higher rate of EVA for children with unilateral hearing loss in this sample population

Conclusion and Recommendations

- Children with EVA, especially EVA and unilateral hearing loss, were identified later than other groups.
- Possible influence of progression and/or late-onset loss
 - Progression of loss with EVA is not uniform and can develop at any age (Zalzal et al., 1995)
 - It is unknown how long a loss might have been present before identification.
- It is possible that regular hearing screening using physiologically-based procedures during well-baby checks will improve identification rates and lower the age of identification

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