An Analysis of Risk Factors in Unilateral versus Bilateral Hearing Loss Identified Through Universal Newborn Hearing Screening in the Commonwealth of Virginia.

Otolaryngology – Head and Neck Surgery

Eric N. Appelbaum, MD[1]; Michael F. Armstrong, BS [1]; Derek Chapman, PhD[2]; Elizabeth Musser PhD[3], Kelley M. Dodson, MD[1]

Virginia Commonwealth University [1]Department of Otolaryngology – Head and Neck Surgery [2]Department of Epidemiology

[3]Virginia Department of Health, Richmond, VA

Abstract

Objective: To compare the incidence of Joint Committee on Infant Hearing (JCIH) risk factors in children with unilateral hearing loss (UHL) to bilateral hearing loss (BHL).

Methods: Retrospective review of children with confirmed hearing loss identified through universal newborn hearing screening (UNHS) in Virginia from 2010—2014.

Summary of Results: Over the 5-year study period, 1,004 children (0.20% of all births) developed a confirmed hearing loss, with 544 children (51%) having at least one JCIH risk factor. Overall, 18% of children with confirmed hearing loss initially passed UNHS. Of all children with risk factors, 226 (42%) demonstrated UHL and 318 (58%) had BHL. The most common risk factors for UHL were neonatal indicators (69%), craniofacial anomalies (30%), stigmata of HL syndromes and family history (14% each). The most common risk factors in BHL were neonatal indicators (49%), family history (27%), stigmata of HL syndromes (19%), and craniofacial anomalies (16%). Children with the risk factor of positive family history were more likely to have BHL, while those with craniofacial anomalies were more likely to have UHL ($p<0.001$).

Conclusion: Neonatal indicators were the most common identified risk factor in both UHL and BHL populations. Children with UHL were significantly more likely to have craniofacial anomalies, while children with BHL were more likely to have a family history of hearing loss. Further studies assessing the etiology underlying the hearing loss and risk factor associations are warranted.

Introduction

Congenital hearing loss has a prevalence ranging from 0.97 to 2.0 per 1,000 newborns.[1-3] Unilateral hearing loss (UHL) is more prevalent than bilateral hearing loss among children ages 6-19.[4] Children with UHL often went undiagnosed and unnoticed in children until they reached elementary school. Universal newborn hearing screening (UNHS) was adopted to identify children with hearing loss at an early age. Early detection allows for early intervention, which is shown to play a key role in cognitive and verbal development.

Research has shown that as many as 22% to 35% of children with hearing loss fail at least one grade.[5,6] Additionally, up to 20% are identified as having behavioral ADHD-type problems.[7,8] Progression of hearing loss has been documented in up to 33% of children with hearing loss emphasizing need for comprehensive early detection and intervention strategies.[9]

In 2007, the Joint Committee on Infant Hearing (JCIH) released an updated position statement, regarding benchmark ages and guidelines for hearing screening, audiological evaluation, and intervention as well as identifying risk factors for congenital hearing loss (**Table 1**).[14]

Previous work in this department compared the incidence of JCIH risk factors and co-morbid birth defects occurring in children born between January 1, 2002 and December 31, 2010 with UHL or BHL. In UHL, craniofacial anomalies were the most commonly reported risk factor. In BHL, family history of hearing loss was the most frequent risk factor. Cardiovascular anomalies were the most common co-occurring birth defects in infants with either UHL or BHL.[16]

Study objectives: to analyze and compare the JCIH risk factor profiles of infants with UHL or BHL using updated, population based data.

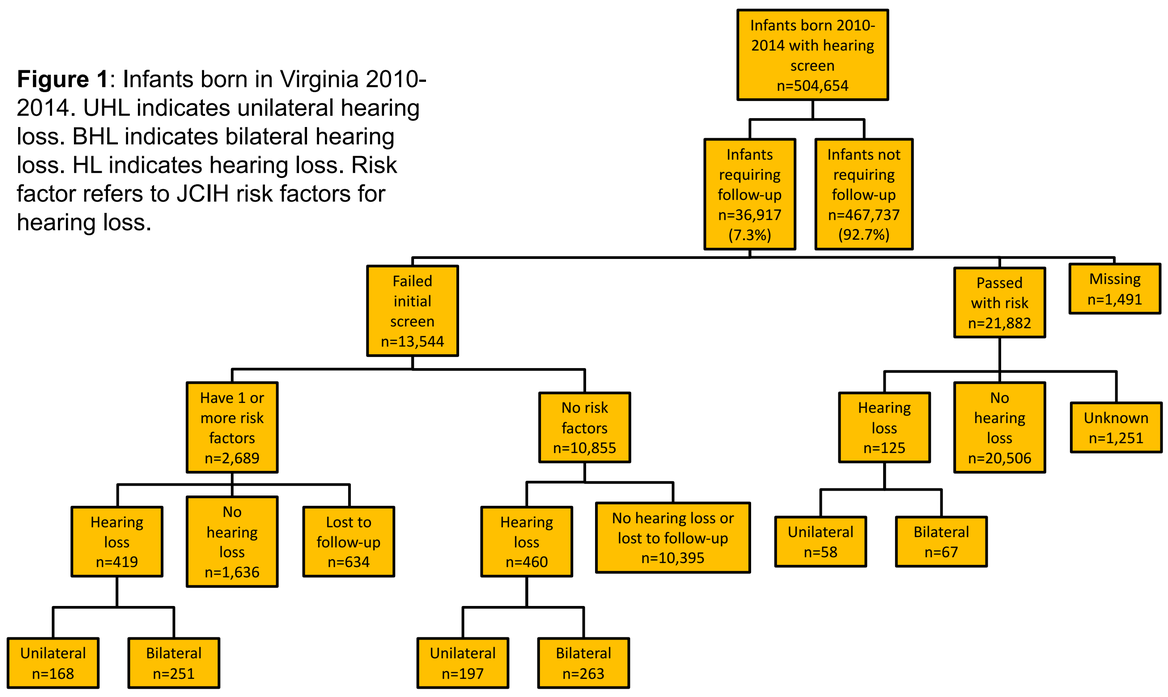

Figure 1: Infants born in Virginia 2010-2014. UHL indicates unilateral hearing loss. BHL indicates bilateral hearing loss. HL indicates hearing loss. Risk factor refers to JCIH risk factors for hearing loss.

Methods and Materials

Data was extracted regarding newborn hearing screening and confirmatory diagnoses from the Virginia Early Hearing Detection and Intervention (VEHDI) program database for children born between January 1, 2010 and December 31, 2014. All newborns with confirmed BHL, including those newborns who passed their initial hearing screening and required follow up due to a known risk factor were analyzed.

Children were grouped into categories regarding status of their newborn hearing screen, confirmatory test outcome, and risk factor status. Hearing loss defined as having one of the following ICD-9-CM;14 codes reported in both ears at a follow-up assessment by a licensed audiologist: 389.0 (conductive hearing loss), 389.1 (sensorineural hearing loss), 389.2 (mixed hearing loss), and 389.9 (undetermined hearing loss).

JCIH risk factors categorized as follows: (a) craniofacial anomaly, (b) family history of permanent childhood hearing loss, (c) head trauma requiring hospitalization, (d)in utero infections such as CMV, herpes, rubella, syphilis, and toxoplasmosis, (e) neonatal indicators for hearing loss such as neonatal care of more than 5 days or ECMO, assisted ventilation, exposure to ototoxic medications (gentamycin or tobramycin) or loop diuretics (furosemide), and hyperbilirubinemia that requires exchange transfusion, and (f) stigmata of syndrome with known hearing loss.

Results

- 504,654 infants underwent UNHS 2010—2014 in Virginia
- 36,917 (7.3%) infants required follow up due to failed screen or presence of risk factors
- 1004 (0.2%) infants with diagnosed hearing loss
- 581 (0.012%) with bilateral hearing loss, 423 (0.008%) with unilateral hearing loss
- 13,544 (2.7%) failed initial UNHS, with 3.1% developing hearing loss
- At least 1 JCIH risk factor was identified in 168 (6.2%) infants with confirmed UHL and 251 (9.3%) infants with confirmed UHL.
  - The most common risk factors for UHL were neonatal indicators (69%), craniofacial anomalies (30%), stigmata of HL syndromes and family history (14% each).
  - The most common risk factors in BHL were neonatal indicators (49%), family history (27%), stigmata of HL syndromes (19%), and craniofacial anomalies (16%). (**Table 1**)
- Rates of family history of hearing loss were significantly higher in infants with confirmed BHL, with rates of craniofacial anomalies significantly higher in infants with UHL, $p < 0.001$.

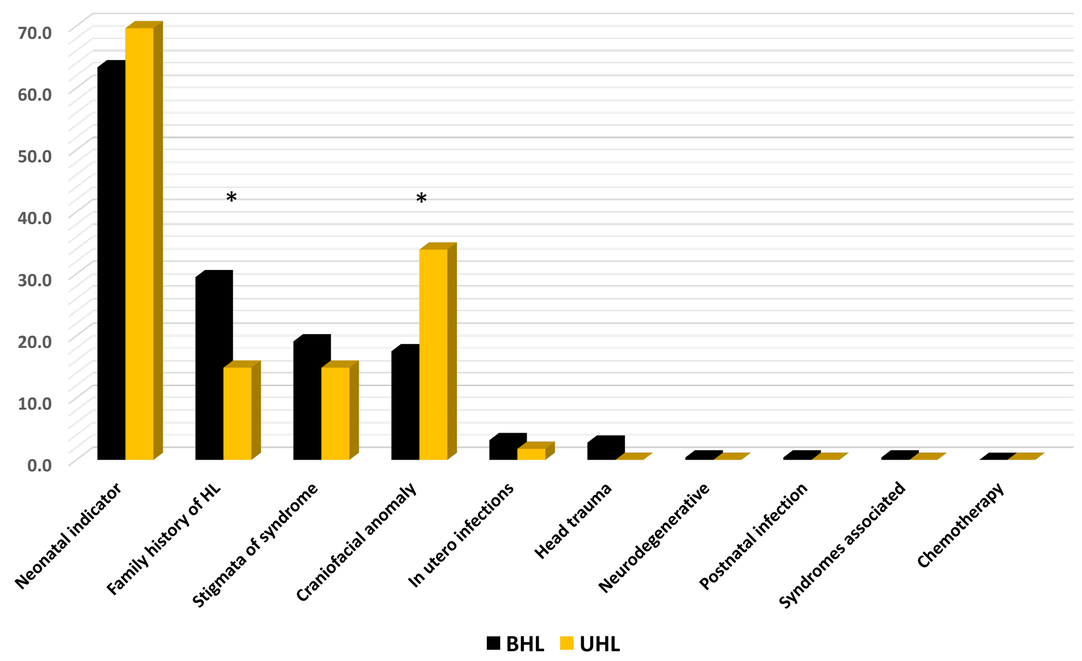

Figure 2. JCIH Risk Factors present in infants diagnosed with BHL vs. UHL. Graph comparing rates of JCIH risk factors present among infants undergoing UNHS In the Commonwealth of Virginia 2010—2014. Family history of hearing loss was significantly more common in infants with bilateral hearing loss (30%) than unilateral hearing loss (15%), while craniofacial anomalies were more common in infants with unilateral (34%) rather than bilateral hearing loss (18%), .

Table 1. Summary of results. Neonatal indicators were the most common identified risk factor in both cohorts, likely due to the broad nature of the category.

Table 1. JCIH Risk factors present in infants diagnosed with BHL vs. UHL

	BHL (%)	**UHL** (%)
Neonatal indicator	63.3	69.6
Family history of HL	29.5	14.9
Stigmata of syndrome associated with HL	19.1	14.9
Craniofacial anomaly	17.5	33.9
In utero infections	3.2	1.8
Head trauma	2.8	0.0
Neurodegenerative disorder	0.4	0.0
Postnatal infection	0.4	0.0
Syndromes associated with HL	0.4	0.0
Chemotherapy	0.0	0.0

Discussion

- Our study presents the results from the statewide analysis of associated JCIH risk factors in a cohort of infants undergoing UNHS 2010-2014
- Identification of JCIH risk factors for hearing loss are an important part of diagnosis. Research has shown that up to 30% of infants with confirmed UHL had a JCIH risk factor[15]
- This study confirms earlier work showing distinct differences in risk factor associations between BHL and UHL cohorts[16].
- The most common risk factor identified among our study cohort was a constellation of neonatal intensive care– related issues potentially causing hearing loss labeled as “neonatal indicators”. This is a diverse grouping of interventions and physiologic processes related to neonatal intensive care. Increase precision in defining the causative neonatal indicator (e.g. ECMO, hyperbilirubinemia) may be more beneficial in identifying those infants at greatest risk of hearing loss.
- Study weaknesses:
  - Data is reliant upon provider-reported input. Risk factors may be more commonly reported due to their ease of diagnosis rather than true incidence. Underreporting or erroneous reporting of birth defects and other risk factors for hearing loss would lead to an under-association with any existing hearing loss.
  - High rates of patient loss to follow up

Conclusions

- About 51% of children with confirmed BHL and confirmed UHL had a JCIH risk factor. Most commonly reported risk factors were family history of hearing loss and neonatal indicators in BHL, and craniofacial anomalies and neonatal indicators in UHL.
- It is important to recognize children at risk for hearing loss and to perform confirmatory testing in a timely manner
- The absence of JCIH risk factors does not preclude the development of hearing loss.
- Further studies are needed to define the etiology underlying hearing loss and better define the risk factor associations.

Contact

Kelley M. Dodson, MD
VCU Department of Otolaryngology – Head and Neck Surgery
Richmond, VA
Email: kelley.dodson@vcuhealth.org
Website: http://ent.vcu.edu
Phone: 804.828.2785

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