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Analysis of risk factors associated with unilateral hearing loss in children who initially passed newborn hearing screening

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ABSTRACT

Objective: To analyze 2007 Joint Committee on Infant Hearing (JCIH) risk factors in children with confirmed unilateral hearing loss (UHL) who initially passed newborn hearing screening.
Methods: Retrospective record review of 16,108 infants who passed newborn hearing screening but had one or more JCIH risk factors prompting subsequent follow-up through the universal newborn hearing screening (UNHS) program in Virginia from 2010 to 2012. The study was reviewed and qualified as exempt by the Virginia Commonwealth University Institutional Review Board (IRB) and the Virginia Department of Health.
Results: Over the 2-year study period, 14896 (4.9% of total births) children passed UNHS but had the presence of one or more JCIH risk factor. Ultimately, we identified 121 babies from this group with confirmed hearing loss (0.7%), with 48 babies (0.2%) showing UHL. The most common risk factors associated with the development of confirmed UHL after passing the initial screen were neonatal indicators, craniofacial anomalies, family history, and stigmata of syndrome associated with hearing loss.
Conclusion: Neonatal indicators and craniofacial anomalies were the categories most often found in children with confirmed unilateral hearing loss who initially passed their newborn hearing screen. While neonatal indicators were also the most common associated risk factor in all hearing loss, craniofacial abnormalities are relatively more common in children with UHL who initially passed newborn hearing screening. Further studies assessing the etiology underlying the hearing loss and risk factor associations are warranted.

1. Introduction

Hearing loss among infants continues to be a source of potential significant impairment. The reported prevalence of unilateral hearing loss (UHL) ranges between 0.3 and 1.0 per 1000 neonates [1–3]. Children with unilateral hearing loss have significantly more unpredictable levels of social functioning than their normal hearing and bilaterally affected counterparts [1]. Historically, children with UHL often went undiagnosed until they reached elementary school. Even after diagnosis, intervention often consisted of no more than preferential seating in class. Unilateral hearing loss was presumed to be insignificant in the development of the child, as functional hearing on one side remained intact.

Several studies have documented the effects of unilateral hearing loss on speech and language development and on quality of life measures. Lieu et al. found that 22%–35% repeat at least one grade with 12%–41% receiving some form of educational assistance [4]. These results echo numerous earlier case-matched control studies, in which approximately one third of children with UHL failed at least one grade [5–7]. In a 1986 study by Bess et al., 50% of the children with UHL studied showed problems in educational progress [8]. Academic difficulty can understandably effect children's self-esteem as well. Bovo et al. demonstrated of the children with UHL referred to a single institution over a 5-year period, 27% felt embarrassed or a sense of inferiority due to their hearing impairment [9]. Conversely, other studies have failed to demonstrate education deficits associated with UHL [10–12]. Keller et al. studied 42,000 children in New York with UHL, and found no significant difference in standardized achievement tests between UHL children and controls [10]. Regardless, early intervention for children with hearing loss is of significant importance. Sininger et al. demonstrated that early intervention strategies allow children with hearing loss to be diagnosed 24 months earlier, undergo intervention 19 months earlier, and meet developmental goals at rates identical to their hearing peers [13].

In 2007, the Joint Committee on Infant Hearing (JCIH) updated its position statement regarding goals and guidelines for early hearing

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detection and intervention programs [14]. As part of this update, the JCIH identified certain risk factors that predispose infants to hearing loss in a delayed fashion warranting early re-examination. These risk factors include caregiver concerns, family history, prolonged neonatal intensive care, in utero infections, craniofacial anomalies, physical findings that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss, diagnosis of the hearing loss associated syndrome, neurodegenerative disorders, postnatal infections, trauma to the head, and chemotherapy. The JCIH endorses a "1-3-6 Plan" format, in which all infants will be screened no later than one month, have confirmation of hearing loss by three months, and should receive intervention by 6 months [14]. In a 2013 study by Yelverton et al., approximately 30% of neonates with unilateral hearing loss on screening had 1 or more JCIH risk factors identified, with 2.5% passing the initial UHNS but developing later onset UHL [15].

Compared to bilateral hearing loss, there is a gap of existing knowledge regarding the etiology of unilateral hearing loss. While the majority of cases remain idiopathic, potential etiologies may include delayed endolymphatic hydrops, genetic factors, environmental agents, and perinatal infections [16–19]. Congenital CMV infection may be responsible for a larger percentage of unilateral hearing loss than is currently clinically recognized [19]. Additionally, genetic factors may play an important role in UHL [18]. Waardenburg syndrome may cause unilateral hearing loss and both TECTA and COCH mutations have been described with unilateral hearing loss in the setting of enlarged vestibular aqueducts [18]. Finally, Ross et al. demonstrated an association between GJB2 mutations (encoding Connexin 26) and congenital CMV infections in the implication of hearing loss [20]. While the above have contributed to our current understanding of UHL and reinforced the potential benefit to genetic screening and counseling, further identification of risk factors will help elucidate those at risk.

The present study aimed to analyze the incidence of JCIH risk factors in children with confirmed UHL who initially passed UHNS using state-wide hearing screening registries. This study used data that was extracted from the Virginia Early Hearing Detection and Intervention (VEHDI) registry, which records data from hospitals across the entire state. Each hospital uses a variety of screening methods. The most common protocol amongst institutions included in the Virginia EHDI registry use automated auditory brainstem response (ABR) testing, whereas others use a two-stage protocol of optoacoustic emissions test (OAE) followed by an ABR test.

2. Methods

Data were extracted regarding newborn hearing screening in confirmatory diagnoses from the Virginia Early Hearing Detection and Intervention (VEHDI) program database for children born between January 1, 2010 and December 31, 2012 in a method described previously. Since 1999, Code of Virginia §32.1–64.1 in Virginia regulations 12 VAC 5–80 requires all hospitals with infant nurseries and all hospitals with neonatal intensive care units to screen the hearing ability of infants prior to discharge. Failure to pass neonatal hearing screening prompts a referral of the infant for diagnostic evaluation. As stated above, the UNHS method used in this study varied from institution, however the most common protocol included ABR x 2, or a two-stage protocol of OAE and ABR.

We specifically analyzed data from newborns who initially passed a hearing screening but were identified to have one or more JCIH risk factors and were then found to have a unilateral hearing deficit at time of further diagnostic testing. During the time of data collection, there was no formally mandated Virginia Department of Health follow-up protocol for children who passed an initial hearing screening with a JCIH risk factor for hearing loss. In accordance with JCIH recommendations, the infants' families, and the primary care physician (PCP) of record, receive a letter recommending the infants receive a full diagnostic evaluation by 24 months of age, however follow-up is not mandated nor enforced.

Hearing loss in this study was defined as having one of the following International Classification of Diseases, 9th Revision, Clinical Modification (ICD-9-CM); codes were reported for one ear (UHL) or both ears (BHL) at a formal hearing evaluation assessment conducted by a licensed audiologist. These included: 389.0 (conductive hearing loss), 389.1 (sensorineural hearing loss), 389.2 (mixed hearing loss), and 389.9 (undetermined hearing loss). The aim of this study was to identify all types of permanent hearing loss that was reported to the VEHDI. Thusly, all ICD-9-CM codes that could contain permanent hearing loss were included. Additionally, as this was a review of anonymized state-wide date from the VEHDI database, access to clinical information or individual data (to include severity of hearing loss) was not permitted.

Neonatal risk factors for hearing loss [neonatal intensive care of more than five days or any of the following regardless of length of stay: ECMO, assisted ventilation, exposure to ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion] that were evaluated included prolonged neonatal intensive care including neonatal indicators of hearing loss, craniofacial anomaly, family history indicating hearing loss, stigmata of hearing loss associated syndrome, postnatal infection, head trauma, chemotherapy, in utero infection, diagnosed syndrome associated with hearing loss, and neurodegenerative disorder. Prolonged neonatal intensive care was defined as more than five days or presence of any of the following: extra corporeal membrane oxygenation (ECMO), assisted ventilation, exposure to ototoxic medications (gentamicin or tobramycin), exposure to loop diuretics (furosemide), and hyperbilirubinemia requiring an exchange transfusion [14].

3. Results

3.1. Incidence of hearing loss

From January 1, 2010 to December 31, 2012 there were 303,646 infants born in the Commonwealth of Virginia who underwent universal newborn hearing screening (UHNS) (Fig. 1). Overall, 2.4 per 1000 were confirmed found to have any type of hearing loss and 0.9 per 1000 had UHL. Of this cohort, 16,108 children (5.3%) were identified to have a JCIH risk factor for hearing loss. 1212 children (7.5% of total) with an identified risk factor failed their UNHS and 14,896 children (92.5%) with an identified risk factor passed. Infants with an identified risk factor who passed their initial UHNS were asked to undergo voluntary rescreening within one month. Of this cohort, 121 (0.8%) infants were found to have any type of hearing loss at the follow-up diagnostic screening. Ultimately, 48 children who had at least one or more risk factor and who passed their initial universal hearing screen were reported to have unilateral hearing loss representing 0.3%. Of the initial 1212 children with an identified risk factor who failed their initial UHNS, hearing loss was identified in 298 (24.6%) and UHL was diagnosed in 108 (8.9%).

3.2. UHL after passing UHNS

For children with an identified risk factor for hearing loss who initially passed their UHNS but were then found to have UHL at their follow-up exam, the most common risk factor was neonatal indicators related to neonatal intensive care (54.2%). Neonatal indicators were the most commonly identified risk factor for hearing loss overall in this cohort. This was followed by craniofacial anomalies (16.7%), family history of hearing loss (14.6%), and stigmata of hearing loss associated syndrome (10.4%). The risk factor with the highest rate of association with unilateral hearing loss discovered after passing the initial hearing screen was craniofacial anomalies with 6.11% (8/131) demonstrating