Risk Factors Associated With Unilateral Hearing Loss

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Objective: To analyze the presence of Joint Committee on Infant Hearing (JCIH) risk factors and co-occurring birth defects (CBDs) in children with unilateral hearing loss (UHL).

Design: Retrospective review.


Patients: The study population comprised 371 children with confirmed UHL.

Main Outcome Measures: Universal newborn hearing screen status, presence or absence of JCIH risk factors, and CBDs

Results: Of the 371 children with confirmed unilateral hearing loss, 362 (97.5%) were identified through a failed universal newborn hearing screen. Of these 362 children, 252 (69.6%) had no JCIH risk factors and 110 (30.3%) had 1 or more risk factor reported. Nine children (2.5%) with 1 or more risk factors passed the universal newborn hearing screen but had later-onset UHL. Craniofacial anomaly was the most commonly reported JCIH risk factor in 48 children (43.6%). A family history of permanent childhood hearing loss was present in 24 children (21.8%). Twenty children (18.2%) had stigmata associated with a syndrome including hearing loss. Of the 110 children with UHL and a JCIH risk factor, additional CBDs were identified in 83 (75.5%). An ear-specific anomaly was most prevalent in 37 infants (44.6%), followed by cardiovascular anomalies in 34 infants (41.0%).

Conclusions: Thirty percent of children with confirmed UHL had a JCIH risk factor, most commonly craniofacial anomalies, family history of hearing loss, and stigmata of syndromes associated with hearing loss. However, the absence of JCIH risk factors does not preclude the development of UHL. Further studies assessing the etiology of UHL and risk factor associations are warranted.


CONGENITAL UNILATERAL hearing loss (UHL) has a prevalence ranging from 0.3 to 1.0 per 1000 newborns.1-3 Before the advent of universal newborn hearing screening, UHL often went undiagnosed and unnoticed in children until they reached elementary school. Historically, even after diagnosis, no significant intervention was performed to aid the child other than preferential seating in class.

The effect of UHL on speech and language development as well as functional and quality of life has been well documented over the last 25 years. Research has shown that as many as 22% to 35% of children with UHL fail at least 1 school grade, and up to 20% are identified as having behavioral problems.4-6 Another study revealed that children with UHL experience barriers to their hearing that force them to adapt.7 In addition, quality-of-life studies have shown that children with UHL had a significantly larger variance in social functioning scores than children with normal hearing and bilateral hearing loss.8 Multiple studies have shown that the speech and language developmental delays can be ameliorated if appropriate intervention for children with hearing loss is begun by age 6 months.7-10

In 2007, the Joint Committee on Infant Hearing (JCIH) released an updated position statement regarding its principles and guidelines for early hearing detection and intervention programs.11 In this statement, the JCIH identified certain risk factors that predispose infants to hearing loss. In addition, the JCIH set forth separate protocols for infants admitted to a neonatal intensive care unit for more than 5 days. They also recommended developmental monitoring at regular 6-month intervals for special populations of children with hearing loss, including those...
with minimal and mild bilateral hearing loss, UHL, and neural hearing loss because these children are at higher risk of having speech and language delay. The association between JCIH risk factors and bilateral hearing loss has been extensively studied, but little attention has focused on this association in children with UHL.

In an attempt to diagnose and intervene earlier in children with hearing loss, the JCIH endorsed a “1-3-6 Plan,” in which all infants will be screened no later than 1 month, have confirmation of hearing loss by 3 months, and receive intervention by 6 months.11 Despite these initiatives, nearly half of all infants who do not pass the initial hospital hearing screen do not receive timely, appropriate follow-up care.12,13 A 2011 study by Chapman et al14 found that the presence of co-occurring birth defects (CBDs) prolonged the time to initial hearing screening, resulting in further delays to diagnosis and intervention.

The present study aimed to analyze the incidence of JCIH risk factors in children with UHL using statewide birth defect and hearing screening registries. In addition, we assessed the presence of CBDs in this population. To our knowledge, this is the first study to specifically analyze the presence of JCIH risk factors and CBDs in the UHL population.

METHODS

Data were 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, 2002, and December 31, 2008, as described previously.1 Code of Virginia §32.1-64.1 and Virginia regulation 12 VAC 5-80, promulgated in 1999, require that all hospitals with infant nurseries and all hospitals with neonatal intensive care services screen the hearing of all infants before they are discharged from the facility. If the infant does not pass the initial screening, the hospital must refer the infant for diagnostic evaluation.

We specifically analyzed all newborns with a confirmed UHL, including those newborns who underwent initial hearing screening and required follow-up. The reasons for follow-up included a failed hearing screen or a known risk factor for hearing loss. These children were grouped into categories regarding status of newborn hearing screen (pass or fail), confirmatory test outcome, laterality of their hearing loss, and risk factor status.

During the study period, hospitals were required to provide the results of initial hearing screening tests to the VEHDI program for infants who failed their initial screen and for infants who passed the initial screen but were at risk for developing early childhood hearing loss as defined by the JCIH.11 All persons who provide audiologic services must also report the status and/or results of diagnostic evaluations to the VEHDI program for infants and children up to age 2 years.

Hearing loss in this study was defined as having one of the following International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM); codes reported in 1 (UHL) or both ears (bilateral hearing loss) at a follow-up assessment by a licensed audiologist: 389.0 (conductive hearing loss), 389.1 (sensorineural hearing loss [SNHL]), 389.2 (mixed hearing loss), and 389.9 (undetermined hearing loss).

The JCIH risk factors were analyzed and categorized as follows: (1) craniofacial anomaly, (2) family history of permanent childhood hearing loss, (3) head trauma requiring hospitalization, (4) in utero infections such as cytomegalovirus (CMV), herpes, rubella, syphilis, and toxoplasmosis, (5) neonatal indicators for hearing loss such as neonatal care of more than 5 days or extracorporeal membrane oxygenation, assisted ventilation, exposure to ototoxic medications (gentamycin or tobramycin) or loop diuretics (furosemide), and hyperbilirubinemia that requires exchange transfusion, and (6) stigmata of syndrome with known hearing loss.

Birth defect diagnoses were extracted from the Virginia Congenital Anomalies Reporting System (VaCARES) for children born within the same period. The VaCARES is a passive compliant birth multisource defects registry15 that receives diagnosis codes from hospitals regarding children from birth to age 2 years, covering 86 categories of structural, functional, or biochemical abnormalities as well as information from Virginia's newborn dried-blood spot screening. Because multiple ICD-9-CM codes related to the same birth defect are commonly reported, the codes were grouped into general categories based on the organ system: (1) cardiovascular anomaly, (2) ear-specific anomaly, (3) eye anomaly, (4) endocrine/metabolic anomaly, (5) gastrointestinal anomaly, (6) chromosomal anomaly, (7) genitourinary anomaly, (8) integumentary anomaly, (9) musculoskeletal anomaly, (10) neurological anomaly, (11) respiratory tract anomaly, and (12) other. The type and number of CBD categories was then computed for each child.

RESULTS

From January 1, 2002, to December 31, 2008, there were 729,583 infants born in the commonwealth of Virginia who underwent universal newborn hearing screening. Of this number, 30,913 infants (4.2%) required follow-up because of a failed newborn hearing screen or the presence of risk factors. Hearing loss was eventually confirmed in 1065 (0.2%) of all infants born in the commonwealth of Virginia during the study period. Confirmed bilateral hearing loss was present in 694 infants, resulting in an incidence of 0.95 per 1000 newborns. Confirmed UHL was diagnosed in 371 infants, resulting in an incidence of 0.5 per 1000 newborns during the study period. Of the children with confirmed UHL, 362 (97.5%) were diagnosed as a result of failing the universal newborn hearing screen. At least 1 JCIH risk factor was identified in 110 infants (30.3%) with confirmed UHL. No JCIH risk factors were found in 252 infants (69.6%) with UHL detected by newborn hearing screening. The other 9 infants initially passed their newborn hearing screen but were later found to have UHL because of subsequent testing secondary to the presence of a known hearing loss risk factor (Figure).

UHL AND JCIH RISK FACTORS

For children with UHL and a JCIH risk factor, craniofacial anomaly was most commonly reported in 48 children (43.6%). A family history of permanent childhood hearing loss was present in 24 children (21.8%). Stigma associated with a syndrome including hearing loss was present in 20 children (18.2%). Neonatal indicators were present in 14 children (12.7%) diagnosed as having UHL. Parental or caregiver concern regarding hearing, speech, language, and/or developmental delay was
found in 10 children (9.1%) diagnosed as having UHL. Known in utero infections were present in 7 children (6.4%) with UHL. Two children had a history of head trauma requiring hospitalization. Postnatal infections associated with SNHL and syndromes associated with progressive hearing loss were associated with 1 child each. Two or more JCIH risk factors were found in 14 children (12.7%) with UHL (Table 1).

UHL, JCIH RISK FACTORS, AND CBDs

Additional CBDs were identified in 107 of the 371 children (28.8%) with UHL and in 83 of the 110 children (75.5%) with UHL and a JCIH risk factor. Of the 107 children with confirmed UHL and a CBD, the most common system anomaly was cardiovascular, present in 46 children (12.4%). The second most common was an ear-specific anomaly, present in 26 children (7.0%) (Table 2).

Similarly, the most prevalent anomaly associated with UHL and a JCIH risk factor was an ear-specific anomaly that was present in 37 infants (44.6%). Cardiovascular anomalies were present in 34 infants (41.0%) (Table 3).

Our study presents the results from a statewide analysis of associated JCIH risk factors and CBDs in children with confirmed UHL detected by newborn hearing screening. Although it is widely accepted that a JCIH risk fac-
tor places a child at a higher risk for hearing loss and that the presence of a comorbid condition can affect the length of time to diagnosis, to our knowledge, this is the first study to investigate the prevalence of JCIH risk factors and CBDs in the UHL population.

The incidence of UHL in our study was 0.5 per 1000 newborns for the study period. This number does not include the 25.7% of children who were lost to follow-up after a failing a hearing screen. However, this number is consistent with the incidence reported in other studies.1,3 Interestingly, we found that almost one-third of infants with confirmed UHL had a JCIH risk factor. No study to date has analyzed the prevalence of these risk factors in congenital UHL. A 2011 study by Bielecki et al15 assessed the incidence of JCIH-defined risk factors in children with SNHL without defining laterality. That study found that syndromes associated with hearing loss were present in 15.52% of children with SNHL. The second most common risk factor was mechanical ventilation for more than 5 days, which was found in 11.49% of children with SNHL. These findings differ from the present study in that craniofacial anomalies were the most prevalent, occurring in 43.6% of children. Also of note is that the second most common JCIH risk factor encountered in our population was a family history of hearing loss, which occurred in 21.8% of children. In contrast, syndromes associated with hearing loss were the least common of the risk factors found in our study, occurring in only 0.9% of children with UHL. The differences between the study by Bielecki et al15 and the present study could be attributed to the different patient populations. The present study focused solely on UHL, whereas, Bielecki et al15 examined all forms of SNHL. Overall, our data underscore the important associations between JCIH risk factors and the presence of UHL. The association between UHL and particular risk factors was previously unknown. Our study clearly shows that UHL is associated with risk factors for hearing loss in as many as 30% of children. In addition, in utero CMV exposure has been associated with hearing loss. At this time, CMV exposure is not routinely screened for at birth. Without routinely screening for CMV, it can be assumed that the incidence of CMV-associated hearing loss has not accurately been measured.

In a 2011 study, Chapman et al1 showed that CBDs were found in nearly one-third of all patients with hearing loss and prolonged the length of time to screening and diagnosis. In that study, an incidence of CBDs in 33.1% of all infants with UHL was detected. Similarly, the present study found that 28.8% of children with confirmed UHL had a CBD. Of the CBDs reported by Chapman et al1, an ear abnormality was the most prevalent, occurring in 50% of children, followed by respiratory (30.4%), gastrointestinal (30.4%), genitourinary (23.5%), and cardiovascular (23.3%) anomalies. Our study found that ear (7.0%), cardiovascular (12.4%), chromosomal (4.6%), genitourinary (3.5%), neurologic (4.3%), gastrointestinal (2.4%), and respiratory (5.1%) anomalies were the most common. The difference between the 2 studies is most likely owing to our study’s focus on UHL, compared with Chapman et al1 whose focus was on all children with hearing loss.

The importance of recognizing CBDs in this population is 2-fold. First, the presence of a comorbid birth defect increases the chances that a child may need prolonged mechanical ventilator support. Multiple studies have demonstrated that mechanical ventilation greatly increases the likelihood of SNHL.16-18 Galambos and Despland19 reported that application of mechanical ventilation can significantly damage the peripheral segment of the hearing tract. Second, the presence of a comorbid birth defect has been shown to lead to an increase in time to hearing screening, diagnosis, and intervention.1,10,21 In a 3-year cohort study, 39,000 infants in the neonatal intensive care unit were 16.4 times more likely to miss the initial hearing screening and nearly 6 times more likely to miss their rescreen compared with infants in a well baby nursery.22

A strength of this study is the use of population-based data on hearing loss, risk factors, and birth defects. Over the 6-year period encompassed in this study, 729,583 children were born in the commonwealth of Virginia, and 30,913 children required follow-up testing because of a failed hearing screen or the presence of a risk factor. Owing to the large number of subjects, we were able to find a large cohort of patients with UHL to analyze concurrent JCIH risk factors and birth defects. To date, we know of no other study that has isolated this population in an attempt to examine UHL and risk factor associations and incidence of comorbid birth defects.

One potential weakness of our study is the reliance on ICD-9-CM codes reported to a passive birth defects registry that could not verify diagnosis through a medical chart review. One issue is that in hospital discharges, multiple diagnosis codes are commonly reported for the same birth defect, and components of sequence or syndromes are commonly reported as separate diagnosis codes. To circumvent some of these foreseeable shortcomings, we grouped CBDs into systems. Underreporting of birth defects would result in children with more severe medical conditions being labeled without appropriate risk factors. Certain risk factors may be more obvious and more likely to be reported as a risk factor. Likewise, inaccurate or false-positive CBDs would result in children with less severe medical conditions being included in the CBD group. Both situations would make our results an underestimate of the association that CBDs have on UHL.

In conclusion, 30% of children with confirmed UHL had a JCIH risk factor. The most commonly reported risk factors were craniofacial anomalies, family history of hearing loss, and stigmata of syndromes associated with hearing loss. In addition, 75% of children with confirmed UHL and a JCIH risk factor had a CBD. It is important to recognize children at risk for hearing loss and to perform screening and confirmatory testing in a timely manner despite the possible multitude of distracting CBDs. Despite the high incidence of JCIH risk factor and CBDs in our population with confirmed UHL, the absence of risk factors does not preclude the development of UHL. Further studies are needed to define the etiology underlying UHL and better define the risk factor associations.
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Author Contributions: All authors had full access to all the data in the study and take responsibility for the integrity of the data and accuracy of the data analysis. Study concept and design: Chapman, Wang, Pandya, and Dodson. Acquisition of data: Chapman, Wang, and Dodson. Analysis and interpretation of data: Yelverton, Dominguez, Wang, Pandya, and Dodson. Drafting of the manuscript: Yelverton, Dominguez, and Dodson. Critical revision of the manuscript for important intellectual content: Chapman, Wang, Pandya, and Dodson. Statistical analysis: Yelverton, Dominguez, Wang, and Dodson. Study supervision: Pandya and Dodson.

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