Associations Linking Progressive Hearing Loss to Patient Diagnostic Data

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Introduction

Progressive hearing loss (PHL) is a significant clinical problem; however, the ability to predict which patients will ultimately develop PHL is poor. Clinicians are often asked by patients and parents whether a hearing loss will change, particularly worsen, over time. PHL has been reported in 4-30% of patients with pediatric sensorineural hearing loss (SNHL)¹,². Sources contributing to the reported variation include (a) use of different criteria to define PHL, (b) demographics of the populations studied, and (c) underlying etiologies. Without a better understanding of the factors involved in PHL and the ability to efficiently integrate sequential hearing test data with patient history and relevant electronic health record (EHR) information, clinicians have little or no information to rely upon when counseling patients about future changes in hearing status. Advances in EHR systems provide a means to efficiently capture digital clinical data to substantially increase the size of patient samples for observational clinical research. We now have powerful new means to discover previously unrecognized associations within the clinical data, to better define the strength of these associations with clinical outcomes, and to use these associations to better predict outcomes.

Results and Discussion

We have developed the Audiological and Genetics Database (AudGenDB) to overcome obstacles to acquiring clinical data for research, leverage digital forms of hearing data from the clinic, and advance methods for observational research in the pediatric hearing loss domain³. AudGenDB integrates audiologic, otologic, radiologic, and genetic patient information from a consortium of pediatric audiology and otolaryngology clinical practices that include Children's Hospital of Philadelphia (CHOP), Vanderbilt University Medical Center (VU), and Boston Children’s Hospital (BCH).

We have used AudGenDB to identify a cohort of patients that demonstrates progressive hearing loss (PHL), which we defined as a worsening (≥10 dB) of masked bone conduction hearing thresholds for the four-frequency pure-tone average (PTA4) of 0.5, 1, 2, and 4 kHz from first to last hearing test. Of the 15,581 children in AudGenDB meeting our inclusion criteria, 425 had PHL. To determine whether there are diagnosis codes with higher prevalence in the PHL patient cohort, a non-progressive cohort that was matched for age and hearing loss severity was identified. Using algorithms scripted in R, we developed a linear regression model that compared the log frequency of ICD-9 diagnostic codes in progressive and non-progressive hearing loss cohorts from two institutions (CHOP & VU). Analysis of the residuals between the observed and predicted values of our linear model identified several diagnostic codes whose prevalence was significantly higher in the progressive cohort when compared with the non-progressive cohort. Further chi-squared analyses demonstrated that cholesteatomas were more prevalent in patients with PHL (ICD9 385.32 (χ²=123 p<< 0.00001) & 385.33 (χ²=101 p<< 0.00001)).

Conclusion

We have demonstrated a significant prevalence of cholesteatoma in pediatric patients with progressive sensorineural hearing loss. Overall, there were 33 diagnoses with a significant χ² > 6. To our knowledge, this is the first study to demonstrate progressive hearing loss in children with cholesteatoma.

References