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Delays in Diagnosis of Congenital Hearing Loss in Rural Children

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Abstract

Objective—To examine the incidence of pediatric congenital hearing loss and the timing of diagnosis in a rural region of hearing healthcare disparity.

Study design—Data from the Kentucky newborn hearing-screening program was accessed to determine the incidence of congenital hearing loss in Kentucky, both in the extremely rural region of Appalachia and non-Appalachian region of Kentucky. We also performed a retrospective review of records of children with congenital hearing loss at our institution to determine the timing of diagnostic testing.

Results—In Kentucky, during 2009–2011, there were 6,970 newborns who failed hearing screening; the incidence of newborn hearing loss was 1.71 per 1000 births (1.28/1000 in Appalachia and 1.87/1000 in non-Appalachia). 23.8% of Appalachian newborns compared with 17.3% of non-Appalachian children failed to obtain follow-up diagnostic testing. Children from Appalachia were significantly delayed in obtaining a final diagnosis of hearing loss compared with children from non-Appalachian regions ($p=0.04$).

Conclusion—Congenital hearing loss in children from rural regions with hearing healthcare disparities is a common problem and these children are at risk for a delay in the timing of diagnosis, which has the potential to limit language and social development. It is important to further assess the causative factors and develop interventions that can address this hearing healthcare disparity issue.

With an incidence of approximately 1.4 per 1000 newborns screened¹, hearing loss is the most common neonatal sensory disorder in the United States. The sense of hearing is important during the early years of life for the development of speech, language, and

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cognition. Hearing impairment in early childhood can result in lifelong learning delay and disability; however, early identification and intervention can prevent educational and social consequences. The United States Preventive Services Task Force has recognized the significant effect that congenital hearing loss has on communication skills, psychosocial development, and educational progress and have found that early detection of hearing loss improves language development.² Others also have confirmed that language skills are closely linked to early identification of hearing loss³ and leads to utilization of early intervention services.^{4,5} Children with congenital hearing loss who are identified and receive intervention no later than 6 months of age perform up to 40 percentile points higher on language expressive measures and social adjustment within the school setting.⁶⁻⁹ Such delays may lead to adulthood challenges in education and employment.¹⁰ Mandatory infant hearing screening has been recommended by the National Institutes of Health,¹¹ Joint Committee on Infant Hearing (JCIH),¹²⁻¹⁵ and the American Academy of Pediatrics¹⁵ in order to initiate the process of hearing loss identification and this screening has been implemented in most states. Newborns who fail their hearing screening or high-risk children undergo an outpatient audiological diagnostic assessment that may take several outpatient encounters in order to obtain definitive diagnosis.

Appropriate follow-up, through diagnostic and intervention services, for children who do not pass a hearing screening or who are diagnosed with hearing loss has become a major national healthcare concern. Disparities in diagnostic and intervention services for some socioeconomic groups are at a high risk of becoming lost to followup.¹⁶⁻¹⁸ Patients in rural areas face additional access-to-care barriers that compound these concerns. According to a recent economic report,¹⁹ 85 of Kentucky's 120 counties are considered rural and approximately 1.8 million people live in these counties. Furthermore, the Appalachian region of Kentucky, which encompasses the eastern and south central portion of the state, is considered to be mostly rural based on the 2003 United States Department of Agriculture Rural-Urban Continuum Coding system²⁰ (Beale codes). This Appalachian region is recognized nationally as suffering from extreme health disparities and is underserved in healthcare services. The 54 Appalachian counties in Kentucky are plagued by poverty, unemployment, and a shortage of healthcare. Considering the barriers to any type of care in Appalachia, there are multiple points in the diagnostic and treatment algorithm in which children with hearing loss potentially can be lost to follow up or have a dramatic delay in receiving timely intervention. The purpose of this study is to investigate the incidence of congenital hearing loss and the delays and disparities in the diagnostic process for children that reside in a large geographic region classified as very rural with limited access to diagnostic and therapeutic services. To this end, this study characterizes infant hearing loss and hearing diagnostic services in Appalachian and non-Appalachian areas of Kentucky during the past decade.

METHODS

The internal review boards of the University of Kentucky (protocol 11-0872-P3H) and the Kentucky Cabinet for Health and Family Services (protocol CHFS-IRB-CCSHCN-FY12-49) approved the study. The JCIH has established the gold standard of infant screening, follow-up, and intervention and serves as the standard for this study.¹⁴ To summarize the current recommendations, infants are to be screened for hearing loss in the hospital of birth prior to discharge. If either ear fails the test, an audiologist performs a complete diagnostic evaluation with an auditory brainstem response (ABR) test within 1 month after birth. Subsequent ABR's may need to be performed in order to confirm a diagnosis of hearing loss and this diagnosis should be made before 3 months of age and subsequent intervention should be initiated prior to 6 months of age.

We examined two separate groups of patients to conduct this research. The first group of patients was children born in the state of Kentucky during the 2009–2011 time frame and failed newborn hearing testing and subsequently referred for definitive testing. These are reported to and managed by the Cabinet for Health and Family Services. Information regarding live births was obtained from the Kentucky State Data Center.²¹ From this database of patients, we obtained de-identified data from the early detection of hearing loss and intervention (EDHI) program including the number of failed newborn hearing screening tests, number of patients who obtained follow-up testing, the number of patients with permanent congenital hearing loss (unilateral and bilateral), the number of patients with severe to profound hearing loss (unilateral and bilateral), and the number of patients enrolled in an intervention program from 2009 – 2011. County of origin was also recorded and designation of each county in the Appalachian region or non-Appalachian region was also performed based on the Appalachian Region Commission designation.²² Differences of follow-up rates after newborn screening between Appalachian versus non-Appalachia were assessed with Chi square testing. In addition, an odds ratio was calculated to quantify the regional differences.

The second group of patients was collected from our institution. Our medical center is the major tertiary center for audiologic assessment of congenital hearing loss in central and eastern Kentucky, including an extensive Appalachian patient base. To assess the early diagnostic process and age of diagnosis of hearing loss in children in central and eastern Kentucky, we identified records of children: (1) born in 2005 or after; (2) referred for diagnostic evaluation of one or both ears from failed newborn hearing screening and (3) subsequently found to have an ABR test result with severe to profound congenital sensorineural hearing loss. Children with acquired hearing loss and/or children who had diagnostic testing performed outside the University of Kentucky were excluded from the study. Our goal was to examine the diagnostic timing of children who initiated and ultimately completed their diagnostic work-up at our institution. We collected demographic data including date of birth, county or origin at time of birth based on ZIP code, Appalachian or non-Appalachian origin, dates of initial and further diagnostic testing via ABR, and date at which final diagnosis was made. Differences in these variables between Appalachian and non-Appalachian patients were examined using Welch two-sample t-test. The rural status of each county of origin was determined using the Beale codes of 2003 United States Department of Agriculture Rural-Urban Continuum Coding system.²⁰ We used a log-rank test to examine differences in the distributions of time to first ABR and the time to diagnosis. The corresponding Kaplan-Meier curves are also provided to visualize these distributional differences. Distance from the ZIP code of origin to the diagnostic center was also recorded. Pearson's correlation coefficient analysis was performed on this data to determine the relationship of distance from the testing center to the timing of the 1st ABR test as well as the timing to final diagnosis. Data were managed using an Excel spreadsheet (Microsoft, Redmond, WA, USA), and statistical analyses were performed with Stata (StataCorp, College Station, TX, USA) and R (R Core Team, Vienna, Austria).

RESULTS

According to the USDA Beale classification, the non-Appalachian region of Kentucky contains 66 counties. The majority of the population resides in urban areas; however, geographically, there is a mix of urban (Beale 1–3), rural (Beale 4–6) along with a few very rural (Beale 7–9) counties, and the Appalachian region is primarily rural and nearly exclusively very rural (Beale code 7–9). The data from our state EDHI program along with total live births²¹ is summarized in the Table. Analysis of 2009–2011 revealed that there was 162,578 live births in Kentucky (43,636 Appalachian and 119,615 non-Appalachian) and 158,969 newborn hearing screens were performed (97.4% screened). There were 6970

failed newborn hearing screens (4.38% failure rate) (1788 Appalachian and 5182 non-Appalachian). Permanent congenital hearing loss was found in 279 children in Kentucky, (an incidence of 1.71 cases per 1000 live births (1 out of 585 live births) or 1.76 cases per 1000 newborns screened (1 out of 568), which is higher than the national incidence of 1.4 per 1000 screened newborns. The percentage of children subsequently diagnosed with permanent childhood hearing loss of those who failed infant screening was 4%. The regional incidences of congenital hearing loss were 1.28 per 1000 births (1 per 779 births) in Appalachia and 1.87 per 1000 births (1 per 536 births) in non-Appalachian counties. There were 121 cases of profound congenital hearing loss (unilateral or bilateral) diagnosed in Kentucky during this period, an incidence of 0.74 per 1000 live births (1 per 1349 births). The regional breakdown of hearing loss was 0.64 per 1000 births (1 per 1558 births) in Appalachia and 0.777 per 1000 births (1 per 1287 births) in the non-Appalachian region. Of the 6970 failed newborn hearing tests during this period, 1326 children never received the appropriate follow-up diagnostic testing or their hearing testing results were never reported to in the EDHI database resulting in a statewide rate of loss to follow up of 19%. The percent of the patients with failed newborn screening tests who obtained follow up for outpatient diagnostic testing was 83% for those residing in non-Appalachian counties compared with 76% in those from Appalachian counties. This was a statistically significant lower percentage of follow up for Appalachian children ($p < 0.001$) with an odds ratio of being lost to follow up of 1.49 (95% confidence interval 1.31 – 1.70). More concerning, of the children who failed the newborn hearing screen in both ears, the follow-up rate in Appalachia children was 73.4% compared with 84.6% in non-Appalachian children. This was also a statistically significant lower rate of follow-up for Appalachian children ($p < 0.001$) with an odds ratio of being lost to follow-up of 1.99 (95% confidence interval 1.53 – 2.59). Upon diagnosis of permanent hearing loss, children are referred for enrollment in early intervention programs and, according to this state-level tracking system, only 51.8% of children in Appalachia and 52% of non-Appalachian children with hearing loss actually enrolled in an intervention program.

We identified 56 children with severe hearing loss who completed diagnostic testing in our institution and were subsequently found to have abnormal diagnostic ABR test results (34 Appalachian and 22 of non-Appalachian). 12 of 22 non-Appalachian children were from the same county as our medical center and 17 of 22 were from urban counties (average Beale code of 3 with a range of 1–7). This indicates that the majority of these non-Appalachian children were from urban/suburban areas. The average distance of the subject's ZIP code to the testing center was 22.6 miles (range of 5–52 miles). Of the Appalachian children, 31 of the 34 were from rural counties (Beale 4–9), with 20 from very rural counties (Beale code 7–9). The average Beale code of the county of Appalachian subjects was 6 with a range of 2–9 and the average distance of the subjects to the testing center was 81.8 miles (range of 23–158 miles). ABR was the primary means of diagnostic evaluation and we found that children in non-Appalachian regions presented for an initial diagnostic ABR test at a median age of 10.4 weeks after birth (mean of 20 weeks), and children in Appalachia presented at a median age of 12.8 weeks after birth (mean of 29 weeks) ($p = 0.338$). Using a log-rank test, there was no significant difference in the distribution between Appalachian and non-Appalachian children in the timing of initial ABR test ($p = 0.432$). We found that non-Appalachian children were diagnosed with a hearing loss at a median age of 21.9 weeks (mean of 31 weeks), and Appalachian children were diagnosed at a median age of 30.5 weeks after birth (mean of 53 weeks) and the difference in the corresponding means neared statistical significance ($p = 0.051$). The corresponding log-rank test revealed a significant difference between Appalachian and non-Appalachian children in the timing of final diagnosis ($p = 0.038$) (Figure 1). A weak linear relationship was identified between distance to the diagnostic center and timing of the 1st ABR ($r = 0.39$, $p = 0.002$) and the timing of the final diagnosis ($r = 0.41$, $p = 0.019$) (Figure 2). In addition, there was a strong linear

relationship between the county Beale code and the distance to the diagnostic center ($r=0.84$, $p<0.001$) indicating that those living the greatest distance from the testing center typically reside in the most rural counties.

DISCUSSION

This study describes pediatric hearing healthcare concerns in a rural region with limited healthcare resources related to hearing. Appalachia is a unique region culturally. However, the concern of healthcare disparity is not unique to this area. We determined that congenital hearing loss is common in the primarily rural state of Kentucky and we found that poor follow up is a major concern, with the most rural regions having the poorest follow up and greatest delay in diagnosis. Although these results may not be generalized to other areas, other rural regions should be examined more closely for similar findings as 20% of the U.S. population resides in rural areas.²³ On a larger scale, developing nations that have implemented or in the process of developing an infant hearing screening program may find similar challenges in delivering timely care to remote regions that could be investigated in a similar fashion.

This Appalachian region represents an example of a large geographic rural area with multiple complicating factors that may affect compliance with diagnostic and treatment recommendations. There is limited availability of infant diagnostic audiological services for this vast region and the expertise in infant diagnostic assessment varies widely among those limited resources. A child from Appalachia who fails a newborn hearing screening often have to travel 1–3 hours to a diagnostic testing center where further testing can be performed. There are also fewer physicians, clinics, and hospitals in the area to provide care, and additionally the geographic isolation and lack of public transit make reaching health services quite difficult.²⁴ The patients in this study represent a group of patients who actually obtained follow-up and for whom a diagnosis was obtained; however, there is great concern regarding those that never seek follow-up after failed newborn hearing screening. Educational disparities are also a concern in this region as 35.3% of residents of very rural regions of Kentucky (Beale code 7–9) have not obtained a high school degree contrasted with 20.3% in urban Kentucky residents (Beale code 1–3).¹⁹ The Appalachian population experiences poorer health in general as they are faced with barriers to adequate healthcare.^{24,25} Economic hardship contributes to a lack of access to healthcare and a significant proportion of inhabitants have no health insurance or are under-insured. Residents of very rural Kentucky counties have a 17% uninsured rate, and the urban region residents has a 12.42% rate.¹⁹ Of the 54 counties in Kentucky considered Appalachian, 41 are distressed and the remaining are either transitional or at-risk of being distressed, which is defined as a poverty rate at least 150 percent of the U.S average or less than 67 percent of the U.S average family income.²⁶ The median income for residents in very rural counties in Kentucky is \$24,609 and the unemployment rate is 48.5% compared with \$42,148 and unemployment rate of 35.7% in urban/suburban regions of the state.¹⁹ This retrospective study is limited by the lack of socioeconomic and education data on the subjects in this analysis; therefore, the causative factors to account for our findings are unclear. These regional socioeconomic and educational differences have the potential to affect the timely diagnosis of congenital hearing loss and further research should investigate this issue.

Although our analysis of diagnostic timing is limited by a modest sample size, we found differences in the distributions of timing of hearing loss with concerning delays in the extremely rural region of Appalachia. Both examined regions of Kentucky also lag behind and the national norms in diagnostic testing (initial outpatient diagnostic test within 1 month after birth and obtain a diagnosis by 3 months after birth). Further work needs to be done to assess and address the failure to follow-up after a failed newborn hearing screen as we

suspect that many patients from the Appalachian region never seek early hearing detection services. Given the expertise needed and the frequent follow up that is needed to adequately diagnose and treat congenital hearing loss, it is not surprising that rural, Appalachian residents may be overwhelmed with the diagnostic process and do not take advantage of services for early hearing loss identification. Cultural factors, including a strong sense of self-reliance and reluctance to leave the area even for needed medical services, may challenge Appalachian residents to obtain the care that they need.²⁷ Further intervention efforts will be directed at increasing education throughout the region regarding congenital hearing loss, development of family support services to facilitate timely diagnosis, and implementing telemedicine services to bring specialized care to remote locations. Our findings raise concern for children in other rural regions as diagnostic delays have the potential to limit language development. Further assessment of explanatory factors and development of intervention programs is warranted to address this hearing healthcare disparity.

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Figure 1. Kaplan Meier analysis of time (weeks after birth) to final diagnosis of congenital hearing loss

Figure 2.
Correlation analysis of hearing services timing.

Table 1

Table Kentucky Congenital Hearing Loss Data (2009–2011)

Region of Birth	Appalachia	Non-Appalachia
Live births	43,636	119,615
Failed Newborn Screens	1,788	5,182
Permanent childhood hearing loss	56	223
Severe sensorineural hearing loss	28	93
Percentage of families obtaining diagnostic testing after unilateral or bilateral failed screening test	76.1%	82.7%
Percentage of families obtaining diagnostic testing following bilateral failed screening test	73.4%	84.6%
Children with hearing loss enrolled in early intervention program	51.8%	52%