Published in final edited form as:

Ear Hear. 2015 ; 36(2): 212–216. doi:10.1097/AUD.0000000000000101.

Targeting Regional Pediatric Congenital Hearing Loss Using a Spatial Scan Statistic

Matthew L. Bush, M.D.¹, W. Jay Christian, Ph.D.², Kristin Bianchi, M.D.³, Cathy Lester, MSSW⁴, and Nancy Schoenberg, Ph.D.⁵

¹University of Kentucky Department of Otolaryngology – Head and Neck Surgery, Commission for Children with Special Health Care Needs, Louisville, KY

²Department of Epidemiology, College of Public Health, Commission for Children with Special Health Care Needs, Louisville, KY

³University of Kentucky College of Medicine, Commission for Children with Special Health Care Needs, Louisville, KY

⁴Cabinet for Health and Family Services, Commission for Children with Special Health Care Needs, Louisville, KY

⁵University of Kentucky Department of Behavioral Science, Lexington, KY, USA

Abstract

Objectives—Congenital hearing loss is a common problem and timely identification and intervention is paramount for language development. Patients from rural regions may have many barriers to timely diagnosis and intervention. The purpose of this study was to examine the spatial and hospital-based distribution of failed infant hearing screening testing and pediatric congenital hearing loss throughout Kentucky.

Design—Data on live births and audiological reporting of infant hearing loss results in Kentucky from 2009 to 2011 were analyzed. We used spatial scan statistics to identify high-rate clusters of failed newborn screening tests and permanent congenital hearing loss (PCHL), based on the total number of live births per county. We conducted further analyses on PCHL and failed newborn hearing screening tests, based on birth hospital data and method of screening.

Results—We observed four statistically significant ($p < 0.05$) high-rate clusters with failed newborn hearing screenings in Kentucky, including two in the Appalachian region. Hospitals utilizing 2-stage otoacoustic emission testing demonstrated higher rates of failed screening ($p = 0.009$) than those utilizing 2 stage automated auditory brainstem response testing. A significant cluster of high-rate of PCHL was observed in western Kentucky. Five of the 54 birthing hospitals were found to have higher relative risk of PCHL and 2 of those hospitals are located in a very rural region of Western Kentucky within the cluster.

Conclusions—This spatial analysis in children in Kentucky has identified specific regions throughout the state with high rates of congenital hearing loss and failed newborn hearing screening tests. Further investigation regarding causative factors is warranted. This method of analysis can be useful in the setting of hearing health disparities to focus efforts on regions facing high incidence of congenital hearing loss.

INTRODUCTION

As the most common neonatal sensory disorder, pediatric hearing loss represents a significant public health concern (CDC 2009). The development of speech, language, and cognition is intimately related to early infant hearing. Early identification of hearing loss leads to utilization of early intervention services (Apuzzo 1995, Moeller 2000). Initiation and utilization of early intervention services with hearing aids prior to 6 months of age has been shown to improve language expression in the school setting (Yoshinaga-Itano 1995, 1998, 2000, 2004). Consequently, mandatory newborn hearing testing has been recommended by the National Institutes of Health (USNIH 1993), Joint Committee on Infant Hearing (JCIH) (JCIH 2000, 2007), and the American Academy of Pediatrics (AAP 1995) in order to facilitate timely hearing loss identification. These recommendations involve hearing screening in the hospital of birth prior to discharge. In the event that either ear fails the screening test, it is recommended that an outpatient audiological diagnostic evaluation occur to obtain a diagnosis no later than 3 months of age.

Patient ability to follow up on these recommendations, socioeconomic factors, and access to care remain major barriers to timely infant hearing healthcare. Inequities in the availability of diagnostic and intervention services for some socioeconomic groups may compound the risk of non-adherence with recommended testing (Brach 2003, Liu 2005, Sommers 2005). Children in rural regions are at a greater risk of delayed diagnosis and delayed treatment compared with urban children (Bush et al 2014). Patients in rural areas may be particularly susceptible to lack of follow up due to social determinants, including poverty, modest educational attainment, and lack of community resources. Given that 20% of the U.S. population resides in rural areas, such widespread potential for disparities merits attention (Census 2000).

The residence patterns of Kentucky provide an opportunity to examine the localization of children with hearing loss in the setting of rural hearing health disparities. Kentucky is largely a rural state with 85 of 120 counties considered rural and approximately 1.8 million people living in these counties (Davis 2009). Furthermore, most of these counties, including the Appalachian region of Eastern/Southern Central Kentucky and the Western-most Jackson Purchase region of the state, are considered to be very rural based on the 2003 United States Department of Agriculture Rural-Urban Continuum Coding system (USDA 2003). Most of these very rural counties are economically distressed and the remaining are either transitional or at-risk of being distressed (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) (ARC 2013). Considering the sizeable population and the barriers to any type of health care in rural regions, there is compelling motivation to investigate regional variations in congenital hearing loss incidence and failed infant hearing screening.

Universal newborn hearing screening programs have been implemented in hospitals throughout the US and over 97% of infants are screened for hearing loss (CDC 2009). However, follow-up diagnostic testing after screening tests has been suboptimal nationally with over 45% of children being lost to follow-up after failing the infant screening test (CDC 2009). This issue of poor adherence with follow-up is significant and many states have implemented centralized online reporting of audiological diagnostic testing to monitor infants and to assist in decreasing the rates of children lost to follow-up. Audiological testing reporting can be used to inform state agencies and clinicians of children in need of definitive diagnostic testing; however, little research has used this information to localize regions where a high number of children fail newborn hearing screening and where congenital hearing loss is most common. The purpose of this study is to utilize a novel statistical method to identify regions and hospitals within Kentucky with the highest rates of failed infant hearing and permanent congenital hearing loss.

METHODS

The internal review boards of the University of Kentucky and the State of Kentucky Cabinet for Health and Family Services approved the study. We obtained de-identified data from the Kentucky early hearing loss detection and intervention (EHDI) database on the number of children who failed newborn hearing screening tests, number of children who obtained follow-up testing, and the number of children with permanent congenital hearing loss (unilateral and bilateral) from 2009 to 2011. This data was organized by county of origin of the children as well by the 54 birthing hospitals in the state. The screening method was recorded for each birth hospital as well. All birthing hospitals within the state utilize inpatient 2-stage screening using otoacoustic emission (OAE) testing or automated auditory brainstem response (AABR) testing prior to discharge. The availability of outpatient screening services at the birthing hospitals was also recorded. The rural status of each county of origin and the county of each birthing hospital was recorded, using the Beale codes of 2003 United States Department of Agriculture Rural-Urban Continuum Coding system (USDA 2003). This numerical scale has 9 classifications with the most urban county being 1 with a metro population of 1 million or more and 9 being the most rural code indicating that the county is completely rural or less than 2500 urban population and not adjacent to a metro area. Codes 1–3 are considered urban and suburban counties, codes 4–6 are considered rural counties, and 7–9 are considered very rural (Davis 2009).

We used spatial scan statistics (Kulldorff 1997) to identify potential clusters of counties that had a high rate of failed screenings or a high rate of PCHL, given the number of live births in each county. We performed these analyses using SaTScan 9.1.1, which was designed specifically for spatial (and spatio-temporal) scan statistics (Martin Kulldorff, Harvard Medical School, Boston and Information Management Services Inc, Silver Spring, Maryland). Briefly, this software conducts Monte Carlo simulations to determine whether there are any clusters of locations (here, counties) with a combined rate of a disease or event that is higher (or lower) than would be expected if it occurred uniformly with respect to population size at each location. The clusters of counties are determined by the SaTScan software by drawing concentric circles ('spatial windows') of increasing size around the centroid of each county, and designating clusters according to which neighboring counties'

centroids fall within the circle. We conducted all four analyses using the software's default settings (i.e., discrete Poisson model, circular spatial window, maximum cluster size 50% of total population at risk), with the exception that we opted to investigate both high- and low-rate clusters. We used ArcGIS 10.1 (ESRI; Redlands, CA) software to map all clusters and present the relative risk (RR) and p-value associated with each cluster for each outcome.

Hospital-based data variables included screening methods (OAE versus AABR), rural county status of the birthing hospital, failed infant screening rate, and PCHL rate. One-tailed binomial tests were used to determine if each hospital had a higher or lower rate of PCHL than the rate for the entire state. The association of rural status with failed screening test rates was analyzed with the Kruskal-Wallis equality-of-populations rank test and the association of screening method and failed screening test rates was assessed with the Mann-Whitney U test. A p-value < 0.05 was considered to be statistically significant. Data were managed using an Excel spreadsheet (Microsoft, Redmond, WA, USA), and statistical analyses were performed with Stata (StataCorp, College Station, TX, USA).

RESULTS

There were 162,043 births from 2009 to 2011, 6968 newborns with failed hearing screening tests (either unilateral or bilateral), and 285 cases of permanent childhood hearing loss. The results for the first analysis are presented in Fig. 1. There were four clusters of counties that had a significantly higher rate of failed newborn hearing screenings per live births compared to the whole state. The geographically largest cluster was observed in western Kentucky, and was comprised of Hancock, Daviess, Breckinridge, Ohio, Meade, McLean, Grayson, Henderson, Butler, Hardin, Muhlenberg, Edmonson, and Webster counties (7.2% of live births failed screening, RR=1.92, $p<0.001$). Rurality, as measured by Beale codes of these counties ranged from 1–8 with an average of 4.15 and median of 3 for the cluster. The analysis revealed a second cluster in central Kentucky that included Marion, Nelson, and Washington counties (7.1% failed screening, RR=1.74, $p<0.001$), with mixed Beale codes of 5, 1, and 8, respectively. Finally, two significant clusters emerged in the eastern, Appalachian region of the state; the first included Johnson, Floyd, Lawrence, Magoffin, and Martin counties (6.9% failed screening, RR=1.69, $p<0.001$). The Beale codes ranged from 6–9 and the average was 7.4 and the median was 7, and the second cluster was comprised only of Whitley and Knox counties and had the highest relative risk for failed screening test of all the clusters (8.0% failed screening, RR=1.96, $p<0.001$). Both counties have been designated as a Beale code of 7. There was another cluster that did not quite reach significance, but included Bracken, Mason, and Robertson counties in northeastern Kentucky (6.7% failed screening, RR=1.63, $p=0.057$) with mixed Beale codes of 1, 6, and 8. When examining the 54 hospitals within the state, we found failed infant hearing screening rates ranging from 1% to 17% and the overall state average was 5.55%. There was no significant difference in the failure rates in urban versus rural hospitals ($p=0.277$). Analysis of the screening method revealed that hospitals with OAE use alone had higher failure rates than those that utilize AABR ($p=0.009$). The eight hospitals that utilized OAE alone in their 2 stage inpatient testing (shown in Fig. 1) had an average failure rate of 9.65%. This was compared to a failure rate of 4.82% in the other 45 hospitals that used AABR alone or OAE then AABR during their 2 stage testing. Seventeen of the 54 birthing hospitals did not have

any follow-up outpatient rescreening services available in the hospital. The odds of providing outpatient rescreening services was 89% lower in those facilities using OAE testing alone ($p=0.01$).

Results for the PCHL analysis based on live births per county are presented in Fig. 2. Only one significant cluster, in western Kentucky, was observed. This cluster, which had a particularly high relative risk, included Graves, Hickman, Carlisle, Marshall, Calloway, McCracken, Ballard, Livingston, Crittenden, Lyon, Trigg, and Caldwell counties (0.46% of live births developed PCHL, $RR=3.08$, $p<0.001$). These counties are primarily very rural and the Beale codes range from 3–9 with an average Beale code of 7.08 and median of 7 for the cluster. When examining the 54 hospitals within the state, we found an incidence of PCHL ranging from 0 per 1000 live births to 10.5 per 1000 live births with a state incidence of 1.76 per 1000 live births. We identified five hospitals that have a significantly higher rate of hearing loss than what would be expected (Fig.2). Two of these centers are the primary state academic tertiary medical centers with NICU centers where high-risk pregnancies and the most critically ill infants are cared for. Two of the hospitals are within the very rural western-cluster that was identified with the spatial scan. There was no significant association with rurality of the hospital of birth and incidence of permanent childhood hearing loss.

DISCUSSION

This analysis is the first of its kind to use a spatial scan statistic to assess regional differences in failed newborn hearing screening and incidence of hearing loss. Furthermore, hospital-based data was analyzed and compared with these results. These data suggest clusters of high incidence of failed newborn hearing screening tests throughout the state and hospitals utilizing OAE testing alone had higher screening failure rates. Those hospitals were also less likely to provide outpatient rescreening testing. A cluster of high incidence of permanent childhood hearing loss was identified in the very rural Western portion of the state. The value of the spatial scan statistic is that since county of origin level data is utilized the location of the residence of the children with hearing loss can be identified. In rural states such as Kentucky many children are born in the larger urban areas, which may be a significant distance from their residence. Considering the nature of hearing loss rehabilitation in children, it is important to provide services in close proximity to the patients' residence. Availability of services within clusters is a crucial issue to address. Interestingly, there are very limited pediatric hearing aid fitting centers within this western-most region of the state and no cochlear implantation services are available.

Mandatory newborn infant hearing screening (HB 706, 2000) has been implemented well in the state of Kentucky and further legislation has promoted compliance with reporting of all audiological testing subsequent to infant hearing screening (HB5, 2010). Together this legislation mandates that all birthing hospitals report infant hearing screening results and each child should be carefully followed and the results of outpatient and diagnostic testing should be recorded and reported to the state EHDI system. Parents or providers who do not adhere to recommended follow-up testing are contacted by state agencies to follow through with appropriate testing and reporting.

The clusters of high rate of failed newborn hearing screening tests highlighted in Fig. 1 may be related, in part, to testing modality. The national rate of failed newborn hearing screening is 1.6% (CDC 2009) and the Kentucky rate of failed screening is 5.55%; however, these Kentucky clusters ranged from 6.9 – 8%. Since a high rate of PCHL was not identified in these regions, it is likely that these clusters represent regions of higher percentage of false positive tests. These data may be used as a quality assurance measure for the region since the national standard for newborn hearing screening failure rates is less than 4% of infants screened (Nelson 2008). All hospitals in the state utilize 2-stage screening in order to decrease false positive rates. The type of screening modality utilized in these regions is an important consideration. According to our state agency, 8 of 54 birthing hospitals in Kentucky screen only with otoacoustic emissions (OAE) and 7 of these 8 hospitals are located within clusters 1, 3, and 4 (and adjacent to cluster 5). Others have suggested that OAE has a higher false positive rate than AABR testing (Stein 1999). Three of these clusters (including cluster 2, which had the highest relative risk of failed newborn hearing screening) occurred within the predominately rural Appalachian eastern half of Kentucky. Further research is indicated within clusters, at the county level, and at the hospital level to determine factors that may be related to these high relative risks. Collaboration with state EHDI agencies to investigate these trends is important to reduce false positives and limit travel for families needing further diagnostic testing.

A high-rate of PCHL was found in the western-most portion of the state based on the number of births (Fig. 2). This region is isolated geographically from the urban areas of the state and is largely very rural, according to Beale code classification. Two of the five hospitals with higher PCHL rates than the rest of the state are located in this Western region. Another two of those five represent the 2 tertiary academic medical centers with large NICU facilities within the state, therefore, the patient population differs from community birthing hospitals. Further investigation is indicated to determine causative factors regarding this high risk of hearing loss within these clusters. Genetic predisposition and the quality of perinatal healthcare may play a role in this finding in both inner city environments as well as very rural regions. Environmental exposures and risk within this region are unknown. Patients residing in very rural areas within this region of the country experience poorer health in general as they are faced with barriers to adequate healthcare (Hartley 2004, Shell 2004). Additionally, those who live in very rural regions of Kentucky (Beale code 7–9) have a lower median household income, lower education levels, higher uninsured rates, and higher mortality rates than those that live in moderately rural regions (Beale code 4–6) and urban regions (Beale code 1–3) (Hartley 2004, Davis 2009). Children affected by hearing loss and living in rural regions may have to travel great distances from rural regions to obtain appropriate audiological and rehabilitative care and this dilemma is compounded by financial hardship. Families of children with hearing loss are more likely to live closer to the poverty level and are less likely to seek medical care (Boss 2011). Further studies will need to prospectively investigate the causative factors involved in PCHL in this very rural region and to ensure that rehabilitation services are accessible and are being used in a timely manner. In addition, application of this statistical technique can be applied to other regions to determine areas of concern (either incidence of hearing loss and/or diagnostic non-

adherence), which can assist EHDI stakeholders in improving diagnostic services and availability of rehabilitative services.

Although these findings are the first to document spatial clusters of congenital hearing loss and abnormal diagnostic testing, we note several limitations. Like any statistic, the spatial scan has inherent limitations that can influence the interpretation of the results of an analysis. The circular spatial windows, for instance, might not be able to identify clusters of counties that are elongated or irregular in shape. This limitation, however, should make clusters more difficult to find, rather than easier, and is thus unlikely to produce entirely spurious results. Furthermore, Fig. 1 demonstrates that contiguous regions of high risk can still be detected with the circular scan window of a spatial scan statistic, albeit as separate and adjacent clusters. For these reasons, we do not believe this limitation has unduly influenced the findings of this exploratory study. This work was conducted in only one predominately rural state and these results cannot necessarily be generalized to other regions or rural states. We propose that this technique be utilized in a multi-institutional investigation to examine larger regions to target areas of concern for PCHL and may be used to focus implementation of interventions in clusters where hearing loss is common.

CONCLUSIONS

Spatial analysis was useful in identifying clusters of congenital hearing loss in children. These clusters are primarily located in very rural regions, which need to be examined more closely with regard to availability of diagnostic and therapeutic services. High rates of failed newborn hearing screening may be related to testing modality. Further investigation and policies should be developed to identify and rectify factors related to this hearing loss. This technique may be useful to address areas of poor compliance and hearing healthcare disparities.

Acknowledgments

Conflicts of Interest and Source of Funding: This publication was supported by the National Institutes of Health (8 KL2 TR000116-02). This was also supported by National Institute of Deafness and Other Communication Disorders (1U24-DC012079-01) and National Institutes of Health Loan Repayment Program. The content is solely the responsibility of the authors and does not necessarily represent the official views of the NIH.

BIBLIOGRAPHY

- American Academy of Pediatrics Joint Committee on Infant Hearing. Joint Committee on Infant Hearing 1994 position statement. *Pediatrics*. 1995; 95:152–156. [PubMed: 7770297]
- Apuzzo ML, Yoshinaga-Itano C. Early identification of infants with significant hearing loss and the Minnesota Child Development Inventory. *Semin Hear*. 1995; 16:124–137.
- Brach C, Lewit EM, VanLandeghem K, et al. Who's enrolled in the State Children's Health Insurance Program (SCHIP)? An overview of findings from the Child Health Insurance Research Initiative (CHIRI). *Pediatrics*. 2003; 112(6 pt 2)
- Boss EF, Niparko JK, Gaskin DJ, Levinson KL. Socioeconomic Disparities for Hearing-Impaired Children in the United States. *Laryngoscope*. 2011; 121:860–866. [PubMed: 21433023]
- Bush M, Bianchi K, Lester C, Shinn J, Gal T, Fardo D, Schoenberg N. Delays in Diagnosis of Congenital Hearing Loss in Rural Children. *J Pediatr*. 2014; 164:393–397. [PubMed: 24183213]
- Bush M, Osetinsky M, Shinn J, Gal T, Fardo D, Schoenberg N. Assessment of Appalachian Region Pediatric Hearing Healthcare Disparities and Delays. *Laryngoscope*. 2014 Jan 8. [Epub].

- Census 2000 Population Statistics. Office of Planning, Environment & Realty. United States Department of Transportation Federal Highway Administration. http://www.fhwa.dot.gov/planning/census_issues/archives/metropolitan_planning/cps2k.cfm.
- Centers for Disease Control and Prevention. [Accessed March 27, 2013] National Center on Birth Defects and Developmental Disabilities. Hearing loss. 2009. at: <http://www.cdc.gov/ncbddd/dd/ddhi.htm>.
- County Economic Status and Distressed Areas in Appalachia. [Accessed 4/18/13] Appalachian Regional Commission. http://www.arc.gov/appalachian_region/CountyEconomicStatusandDistressedAreasinAppalachia.asp.
- Davis AF. Kentucky's Urban/Rural Landscape: What is driving the differences in wealth across Kentucky? Kentucky Annual Economic Report. 2009 <http://cber.uky.edu/Downloads/annrpt09.html>.
- Hartley D. Rural health disparities, population health, and rural culture. American Journal of Public Health. 2004; 94(10):1675–1678. [PubMed: 15451729]
- Joint Committee on Infant Hearing. Year 2000 position statement: principles and guidelines for early hearing detection and intervention programs. Pediatrics. 2000; 106:798–817. [PubMed: 11015525]
- Joint Committee on Infant Hearing. Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. Pediatrics. 2007; 120(4):898–921. [PubMed: 17908777]
- [Accessed 5/21/13] Kentucky Legislature HB5. <http://www.lrc.ky.gov/RECORD/09RS/HB5.htm>.
- Kentucky Legislature HB706. [http://www.infanthearing.org/legislative/documents/KY%20\(2000\)%20HB%20706.pdf](http://www.infanthearing.org/legislative/documents/KY%20(2000)%20HB%20706.pdf).
- Kulldorff, Martin. A spatial scan statistic. Communications in Statistics-Theory and methods 26.6. 1997:1481–1496.
- Liu CL, Zaslavsky AM, Ganz ML, Perrin J, Gortmaker S, McCormick MC. Continuity of health insurance coverage for children with special health care needs. Matern Child Health J. 2005; 9(4): 363–375. [PubMed: 16328707]
- Moeller MP. Early intervention and language development in children who are deaf and hard of hearing. Pediatrics. 2000; 106:E43. [PubMed: 10969127]
- Nelson HD, Bougatsos C, Nygren P. Universal Newborn Hearing Screening: Systematic Review to Update the 2001 US Preventative Services Task Force Recommendation. Pediatrics. 2008; 122:e266–e276. [PubMed: 18595973]
- Rural-Urban Continuum Codes. [Accessed 2/25/13] USDA Economic Research Service. United States Department of Agriculture. <http://www.ers.usda.gov/data-products/rural-urban-continuum-codes.aspx>.
- Shell R, Tudiver F. Barriers to cancer screening by rural Appalachian primary care providers. J Rural Health. 2004; 20:368–373. [PubMed: 15551854]
- Sommers BD. From Medicaid to uninsured: drop-out among children in public insurance programs. Health Serv Res. 2005; 40(1):59–78. [PubMed: 15663702]
- Stein LK. Factors influencing the efficacy of universal newborn hearing screening. Pediatr Clin North Am. 1999 Feb; 46(1):95–105. [PubMed: 10079793]
- U.S. National Institutes of Health, Office of Medical Applications Research, U.S. National Institute on Deafness and Other Communication Disorders. Consensus development conference on early identification of hearing impairment in infants and children. Vol. 11. Bethesda, Md: National Institutes of Health; 1993. p. 1-24.
- Yoshinaga-Itano C. Efficacy of early identification and early intervention. Semin Hear. 1995; 16:115–123.
- Yoshinaga-Itano C. Levels of evidence: universal newborn hearing screening (UNHS) and early hearing detection and intervention systems (EHDI). J Commun Disord. 2004; 37:451–465. [PubMed: 15231425]
- Yoshinaga-Itano C, Sedey AL, Coulter DK, Mehl AL. Language of early- and later-identified children with hearing loss. Pediatrics. 1998; 102:1161–1171. [PubMed: 9794949]

Yoshinaga-Itano C, Coulter D, Thomson V. The Colorado Newborn Hearing Screening Project: effects on speech and language development for children with hearing loss. *J Perinatol.* 2000; 20(8 pt 2):S132–S137. [PubMed: 11190694]

Figure 1.
High-rate clusters of failed hearing screenings per live births, Kentucky 2009–2001.
Birthing hospitals utilizing OAE testing alone (stars), including percentage of live births
with failed infant hearing screening, are graphically presented as well.

Figure 2.

High-rate cluster of permanent childhood hearing loss (PCHL) cases per live births, Kentucky 2009–2011. Birthing hospitals with significantly higher incidence rates (PCHL cases out of 1000 live births) of congenital hearing loss (stars) than the rest of the hospitals in the state are graphically presented.