Evaluating Pennsylvania's Newborn Hearing Screening Program

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Abstract

Scope: Pennsylvania's Newborn Hearing Screening (NBHS) program is a critical state-run program that is imperative for the goal of early identification of children with hearing loss. The purpose of this study was to evaluate Pennsylvania's administration of the NBHS, as well as analyze Pennsylvania's adherence to the Joint Committee on Infant Hearing (JCIH) 1-3-6 Guidelines.

Methodology: Records from 131,832 newborns born in 2018 were analyzed for this study. Descriptive statistics were used to determine outcomes related to the JCIH guidelines. Prevalence of hearing loss and odds ratios were calculated to determine risks of hearing loss in the 2018 newborn population.

Conclusions: The findings suggest that Pennsylvania has a strong adherence to the 1-3-6 guidelines, with an average timeframe of 3.04 days from birth to screening, 75.39 days from birth to diagnosis, and 174.2 days from birth to early intervention enrollment. The information from this study will be used for future program development, as well as to identify areas of improvement within the Commonwealth.

Keywords: newborn hearing screening, state outcomes, JCIH guidelines

Acronyms: EHDI = Early Hearing Detection and Intervention; EI = early intervention; IHEARR = Infant Hearing Education, Assessment, Reporting and Referral; JCIH = Joint Committee on Infant Hearing; NBHS = newborn hearing screening; NICU = neonatal intensive care unit

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Since the 2000 recommendation by the Joint Committee on Infant Hearing (JCIH); a guideline recommending that all infants born in the United States are screened for hearing loss by one month of age, diagnosed by three months of age, and enrolled in early intervention (EI) by six months of age; the number of infants screened has increased dramatically. The Centers for Disease Control and Prevention reported that in 2016, roughly 98% of infants born in the United States underwent a newborn hearing screening at birth. Although this statistic is very reassuring, there remain gaps in data related to diagnostic assessments and later EI enrollment. These gaps are often attributed to incomplete or inconsistent local data (Alam et al., 2016).

In a study by Uhler et al. (2014), Early Hearing Detection and Intervention (EHDI) coordinators from across the United States were surveyed on the state or territory structures in place to track diagnostic, amplification, EI, and medical outcomes in children screened for hearing loss. Their results found that only 31.25% of those surveyed had a database in place that contained information regarding assessment and audiology follow-up data. The researchers attribute difficulties following up

with screened infants to limited staff capacity as well as limitations in obtaining funding for database creation and maintenance. In addition to the findings by Uhler et al. (2014), Shulman et al. (2010) identified communication between hospitals and newborn hearing screening (NBHS) staff as a major challenge in optimizing the EHDI reporting program throughout the United States. In this study, staff from NBHS programs were asked to rank the quality of data reported from hospitals. The researchers found that staff largely reported that data was *poor* or *good* compared to *very good* or *excellent*.

In 2001, the Pennsylvania State Assembly passed the Infant Hearing Education, Assessment, Reporting and Referral (IHEARR) Act. This act required providing newborn hearing screenings to all infants within the Commonwealth as well as programs for follow-up services. Newborn hearing screenings were implemented statewide in July 2002. The IHEARR Act additionally called for the creation of a newborn hearing screening advisory board, consisting of organizations, stakeholders, and professionals to monitor hearing health outcomes for all children born within the Commonwealth.

The Pennsylvania Division of Newborn Screening and Genetics implemented a policy shift to track the outcomes of all babies screened in the state through a centralized, web-based monitoring system, called iCMS. All activities related to the NBHS, including screening results, tools used, diagnostic evaluation results, and EI enrollment, were tracked through iCMS. This system was fully implemented to track the outcomes of children born in 2018 and later. The purpose of this study is to assess the implementation of Pennsylvania's NBHS program and its adherence to the JCIH 1-3-6 guidelines, using the data received through the iCMS system.

Methods

For this study, infant records of those born between 1/1/2018 and 12/31/2018 were assessed through the iCMS system. Inclusion for this study was limited to babies native to Pennsylvania, as identified by maternal zip code and county. Descriptive analyses were used to determine outcomes related to screening, diagnosis, and EI enrollment. All statistical analyses were completed using *R* statistical analysis software (R Core Team, 2019). Apparent prevalence of hearing loss was calculated using 89% sensitivity and 92% specificity, the most conservative estimates from Butcher et al. (2019).

Results

A total of 131,832 (67,746 males, 64,083 females, 3 unspecified) newborn screening records were analyzed for this study. Of the 131,832 total records; 125,381 infant records reported information regarding birth setting. Of those born in Pennsylvania, 125,627 infants (95.3%) were seen in inpatient settings, and 6205 (4.7%) were assessed in outpatient locations. Families of infants who did not pass the initial hearing screening prior to discharge from the birthing center were instructed to follow-up at an outpatient clinic. Infants were further followed through the iCMS system, where each case was kept open until a final diagnosis was rendered in the case of normal hearing, or the infant was enrolled in EI services, in the case of a

diagnosis of permanent hearing loss. Outpatient centers and midwifery services were further used for infants born at home.

Screening Results

Figure 1 illustrates the ultimate screening results for newborns born in Pennsylvania in 2018. Of all screenings, 119,683 (90.1%) occurred in well-baby nurseries, while 11,884 (9.0%) occurred in the neonatal intensive care unit (NICU). Pennsylvania had an overall pass rate of 96.9%, with 127,694 babies passing bilaterally. A total of 1148 babies, less than 1%, referred on the screening in at least one ear. Roughly 2% (2,439) of babies recorded did not have a completed screen. The largest reason for this lack of screen can be attributed to parent refusal. Babies who had their final screening in inpatient settings tended to have a higher rate of passing (97.8%) compared to those who were screened in outpatient settings (77.9%). Table 1 illustrates the difference in outcomes based on screening setting. Although babies screened in the outpatient setting tended to have a higher refer rate (5.0%) than those tested in inpatient screenings (0.7%), there was a substantially higher percentage of children who ultimately were not able to complete the screen in outpatient. Most significantly, the parent refusal rate for outpatient screens was 10.9% compared to 0.6% in inpatient screenings. There was also a marked difference in the time it took to obtain the newborn hearing screen.

Initial inpatient screenings were conducted an average 2.59 (± 9.32) days following birth. Outpatient initial screens were conducted an average of 10.62 (± 18.73) days after birth. The length of time from initial to final screenings was substantially different for inpatient screenings when compared to outpatient screenings. On average, the final inpatient hearing screen took place 3.04 (± 11.22) days after birth, while it took 32.36 (± 46.67) days to complete the final hearing screen on outpatient infants. Pennsylvania's average time frame was 4.39 days (± 16.06) from birth to final screening completion for all babies, regardless of screening setting.

Figure 1
Total Results of 2018 Newborn Hearing Screening Program in Pennsylvania
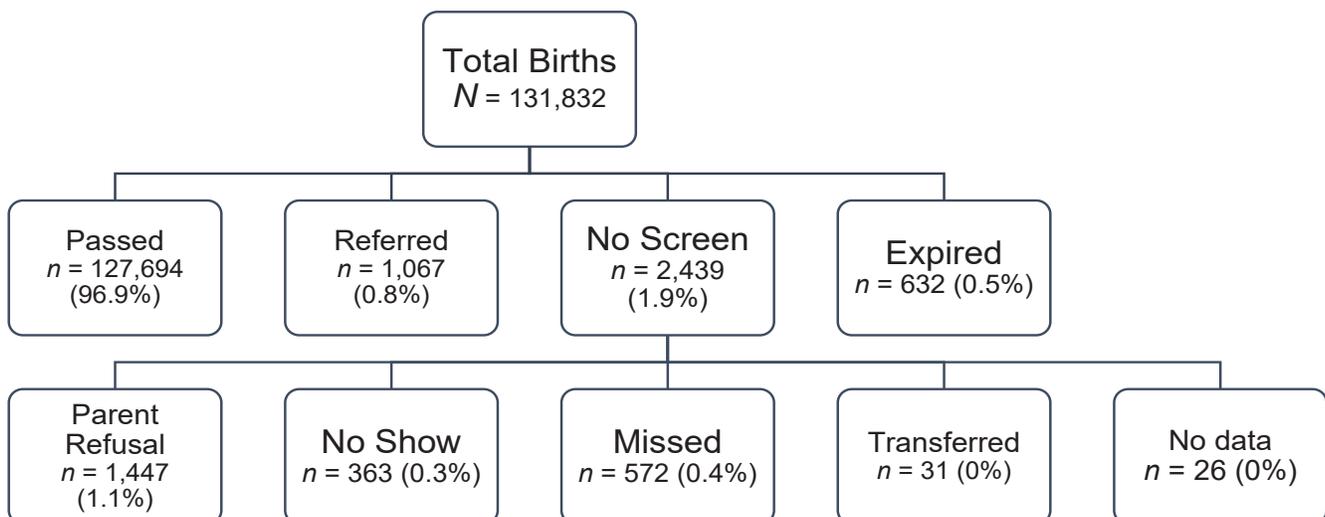
Table 1

Newborn Hearing Screening Outcome Based on Screening Setting

Outcome	Count (*n*)	Percent (%)
Inpatient		
Pass	122,799	97.7%
Refer	840	0.7%
Parent Refusal	769	0.6%
Not Screened (Other)	602	0.5%
Expired	617	0.5%
Outpatient		
Pass	4,794	77.3%
Refer	308	5.0%
Parent Refusal	679	10.9%
Not Screened (Other)	408	6.6%
Expired	16	0.3%

Table 2

Count of Children Diagnosed with Unilateral/Bilateral Permanent Hearing Loss (HL) in 2018

Diagnosis	Right Ear HL Only	Left Ear HL Only	Bilateral HL	Total	Prevalence (per 1000)
Auditory Neuropathy	1	1	4	6	0.045509709
Mixed Loss	7	6	6	19	0.144114078
Permanent Conductive Loss	8	5	4	17	0.128944175
Sensorineural Loss	35	33	98	166	1.259101942
Unknown Loss	3	5	17	25	0.189623786

Some infants (5,482) had their first screening completed in an inpatient setting and had a follow-up screening completed at an outpatient facility. This number includes infants that eventually passed their newborn hearing screening but may have referred on their first screen. On average, it took 35.1 (± 48.51) days for these babies to receive a final screening outcome. Babies who were screened in outpatient settings were over 10 times more likely to refer on their final screen compared to those screened in an inpatient setting (OR = 10.46, 95% Confidence Interval: [CI] 9.13–11.97).

Of the 119,683 babies screened in well-baby nurseries, 97.1% passed their newborn hearing screening and 0.7% referred. This pass rate was higher compared to those screened in the NICU, who had a 94.1% pass rate and 2.2% referral rate. Those screened in the NICU were over three times more likely to refer on the NBHS compared to those screened in well-baby units (OR = 3.28, 95% CI: 2.86–3.77). There was no association between well-baby nursery screening and referral on the NBHS (OR = 1.0).   A total of 2,405 babies were screened using midwife services in 2018. Of those infants, 1,380 (57.4%) passed their screening, 10 (0.4%) referred on their final screening, and 1,015 (42.2%) did not complete a final screen due to parent refusal (*n* = 763), missed appointments (*n* = 246), or similar reasons. Six infants had no information regarding their screening status.

Diagnostic Assessment Results

Of the 1,067 babies who referred on their newborn hearing screen, 884 (82.8%) were seen for a diagnostic follow-up. Of these infants, 664 received a final diagnosis of normal bilateral hearing. This finding suggests a false positive rate of 0.5%. Table 2 shows the diagnostic outcomes for those found to have permanent hearing loss either unilaterally or bilaterally. The prevalence of permanent hearing loss among newborns was 1.76 per 1000 (95% CI:1.5–2.0) in 2018, with 233 children diagnosed with hearing loss by their final evaluation. Bilateral and unilateral hearing diagnoses were equally common, with 108 (46.4%) children diagnosed with a bilateral hearing loss, compared to 110 (47.2%) children diagnosed with a unilateral hearing loss. The average length of time from birth to the completion of the diagnostic assessment was 75.39 (± 72.3) days. Analysis of the severity of hearing impairment showed the highest representation of hearing loss as either a moderate (21.7%) or profound (21.1%) hearing loss among those with a classified severity. In total, 176 of the 233 (75.5%) infants diagnosed with permanent hearing loss had a severity classified in at least one ear. For 38 (22.9%) of these infants, the classified severity was *unknown*, indicating that more diagnostic testing was necessary before making a final classification. Table 3 demonstrates the severity rating for children with both unilateral and bilateral permanent hearing loss.

Of the 884 infants that completed a diagnostic assessment, 683 were born in well-baby nurseries. Of these infants, 78.7% were diagnosed with normal hearing, and 21.1% were diagnosed with some form of hearing loss in at least one ear. For those screened in the NICU (*n* = 199), 62.3% had normal hearing, while 37.7% were diagnosed with some level of hearing loss in at least one ear. Those born in well-baby nurseries were nearly half as likely to be diagnosed with a hearing loss compared to those screened in the NICU (OR = 0.44, CI: 0.31–62.3).

Early Intervention

As of May 2020, 180 of the 233 children diagnosed with a hearing loss from the newborn hearing screen in 2018 were referred for EI services. At this time, 137 (76.1%) children have been enrolled in EI. Table 4 illustrates the status of children being followed for early intervention services. Data from the 137 children suggests that the average length of time from birth to the generation of an EI referral is 121.4 (± 107.1) days. The average length of time from birth to the enrollment in early intervention services is 174.2 (± 116.5) days.

Table 3

Degree of Permanent Hearing Loss in Study Sample

Degree	Unilateral	Bilateral	Total	Percent
Slight	2	1	3	1.8%
Mild	4	22	26	15.7%
Moderate	11	25	36	21.7%
Moderately-Severe	16	11	27	16.3%
Severe	10	27	11	6.6%
Profound	8	27	35	21.1%
Unknown	9	29	38	22.9%

Note. Percent indicates percent of total diagnosed.

Table 4

Status of Children Monitored for Early Intervention Services

Status	Count	% Followed	% PHL
Enrolled	137	76.1%	58.8%
Pending	12	6.7%	5.2%
Refused	4	2.2%	1.7%
Unknown	27	15.0%	11.6%

Note. PHL = permanent hearing loss.

Discussion

The findings from this analysis suggest that Pennsylvania largely meets the JCIH 1-3-6 guidelines. Newborns screened in-hospital usually receive an initial hearing screen within the first 12 to 24 hours after birth, a number that is reflected in the average in-patient screening time of 3.28 days. This number increases significantly and exceeds the target of screening by one month of age in the outpatient screening population. This increase in screening time, as well as the increased no-screen rate among outpatient events can potentially be attributed to the geographic makeup of the state. Pennsylvania is largely stratified between large urban centers in the east and west of the state, and more suburban and rural communities within the center of the state. According to the Center for Rural Pennsylvania, as of 2018, roughly 26% of the population of Pennsylvania lives in a rural community (Center for Rural Pennsylvania, n.d.). These communities generally have more limited access to healthcare services.

Low compliance in outpatient screenings is unfortunate, not unusual. A study by Griz et al. (2009) found that lower maternal education level, socioeconomic status, and rural living all demonstrate lower compliance with attending outpatient screening events. In 2018, the Pennsylvania Department of Health (DoH) reported that there were 66 general hospitals with 7,265 beds, (2.14 beds per 1000 residents) in rural Pennsylvania, with seven counties having no hospital at all. Additionally, these rural areas tended to demonstrate a higher poverty level (12.7%) compared to more urban areas (12.1%; Semega et al., 2019). Low compliance for outpatient screenings may also be attributed to the number of screenings and births provided by midwives throughout the state. According to Goedert et al. (2011), most midwives do not view newborn hearing screening as a responsibility and do not have the knowledge to provide information related to the NBHS program. In our study, we found that over 40% of babies screened using midwife services did not have a final NBHS result. Given our findings and previous literature, it is essential to educate midwife service providers on both the importance of the NBHS program and the role that these service providers play in conducting this vital service. Currently in Pennsylvania, programs have been designed to increase midwife and outpatient education in NBHS. Further studies should evaluate the effectiveness of these training programs.

The mean duration from birth to diagnostic assessment result fell within the JCIH 1-3-6 guidelines. On average, infants were provided a final diagnosis approximately 75 days after birth. This is well within the guidelines suggested by the JCIH, which is that a final diagnosis occurs by three months of age. The Pennsylvania prevalence rate of 1.76 per 1,000 infants aligns with the national prevalence rate of 1.7 (Centers for Disease Control [CDC], 2017). Though these numbers appear to agree with published data, further study into the impact of loss to follow-up (LFU) on this prevalence would be beneficial. LFU is a major concern with any screening program. Presently, nearly 87% to 95% of newborns undergo a newborn hearing screening shortly after birth (Gaffney et al., 2010; Mehl & Thomson, 1998, 2002). Gaffney et al. (2010) assessed nationwide LFU on those who referred their newborn hearing screening and suggested that nearly a third of those identified with a hearing impairment at birth could go without hearing loss identification.

The false positive rate of 0.5% agrees with the hypothesized false positive rate of Clemens et al. (2000). In their study, the research team analyzed the false-positive rate of newborns during the initial screening (Stage 1) and found a false positive rate of 1.9%. The team notes that if they completed the rescreening process, which they called State 1b, the false-positive rate would be 0.5% overall. Our study confirms this estimation.

Additionally, the timeline for EI enrollment fell within the JCIH guidelines. The mean duration from birth to EI enrollment was approximately 175 days, just shy of the six-month JCIH recommendation. Adherence to the 1-3-6 guidelines is linked to increased vocabulary development in children, including better receptive and expressive language abilities, as well as a higher level of speech

intelligibility than children who do not meet the guidelines or are not screened at all (Yoshinaga-Itano et al., 2001; Yoshinaga-Itano et al., 1998; Yoshinaga-Itano et al., 2017). Although the EI enrollment dates for Pennsylvania fall within the JCIH guidelines, there is still a lapse of time between final diagnosis and EI referral of approximately 46 days. Further research should explore reasons for this gap, though data from nation-wide studies suggest that the delay can often be attributed to agreements that states make with birthing centers related to timeliness. Sanchez-Gomez et al. (2019) note that states that require data collection within two weeks of screening have better follow-up rates than those who require collection within one month. This should be considered in evaluating the time lapse from screening to diagnosis, as well as from diagnosis to EI enrollment.

The change of policy requiring submissions to iCMS, the Pennsylvania newborn screening system, mandates all NBHS submitters (hospitals, birthing centers, or midwives) to report individual-level hearing screening results for all babies. This includes those who were unable to be screened due to parent refusal, missed screening, and transferring to hospitals outside of the state. This change came into effect in full for all babies born on January 1, 2018. Although this mandatory reporting has many benefits, communication between birthing centers and NBHS programs still faces some difficulty. One limitation is in considering that data input was completed by individual stakeholders throughout the process. Those stakeholders include nurses, audiologists, social workers, and early interventionists, as well as staff within the Pennsylvania DoH. Although it is important to have a variety of inputs for tracking and normalizing purposes, the variety in personnel inputting the data leads to the possibility of human error. For example, 212 children of the 882 children seen for a diagnostic assessment had an unknown or no-indicated hearing severity in their final report. There is no state-wide standard as to who must provide this data to the PA DoH, therefore it may be possible that this number can be attributed to human error. It may also be reflective of an aspect of the iCMS system that may need to be improved and standardized for more universal understanding among stakeholders.

The purpose of this study was to assess Pennsylvania's compliance to the JCIH recommendations of screening by one month of age, diagnosis of hearing loss by three months of age, and early intervention enrollment by six months of age. Of interest, was the analysis of this adherence as it pertains to the policy shift of 2018, requiring all information to be stored within a centralized databank. The findings from this study suggest that Pennsylvania largely adheres to the JCIH guidelines and that use of a centralized database allows for intensive analysis into the NBHS program implementation. These findings will be used for future program improvement in Pennsylvania, specifically for outpatient screening improvement. Further research analyzing the specific outcomes related to race and region can provide deeper insight into the program's efficacy, as well as identify outreach programs for optimizing outcomes.

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EHDInfo [Can be downloaded here for distribution.]

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improves language skills
IF WORN 10 HOURS OR MORE EACH DAY.

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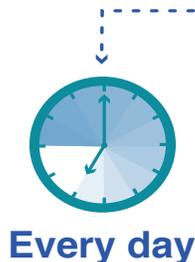
In the car

When awake

At play

☑ Sunday
☑ Monday
☑ Tuesday
☑ Wednesday
☑ Thursday
☑ Friday
☑ Saturday

Every day

Talk to your audiologist for support to meet the individual needs of your family.

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