Early Hearing Detection and Intervention Program Report on Screening and Follow-up for 2016 Births

Bureau of Family Health
Division of Newborn Screening and Genetics

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Introduction

In 2016, approximately 6,300 babies in the United States were born with permanent hearing loss, making it the most frequently occurring condition identified through newborn screening.[1] Timely identification of hearing loss is crucial, as it allows early access to intervention services. Research shows that children with a hearing loss identified by 3 months of age and intervention initiated by 6 months of age can learn to communicate on a comparable level with their peers, whereas children experiencing late identification of hearing loss may experience irreversible and permanent impairments in speech, language and cognitive abilities. As the brain develops and matures during the first three years of life, nerve pathways necessary for understanding auditory information are created. Children with hearing loss who are not exposed to language while the brain develops will face challenges developing reading skills, spoken words and sign language.[2]

Pennsylvania's Infant Hearing Education, Assessment, Reporting and Referral (IHEARR) Act (Act 89 of 2001) enables the Department of Health (Department) to administer a statewide comprehensive newborn hearing screening and follow-up program. This report is submitted to the General Assembly in fulfillment of the reporting requirements found in Section 5(d) (4) of the IHEARR Act. The report covers the results of both in- and out-of-hospital screenings for infants born in 2016, follow-up activities for infants referred to the Department for failure to pass their newborn hearing screening from 2014 through 2016, and the status of ongoing program initiatives undertaken through the current state fiscal year.

Background

Due to the emergence of reliable, affordable technology for early hearing detection in the mid-1990s, the Department began a pilot program to screen infants for hearing loss with 26 birthing hospitals in 1999. The pilot program proved the practicality of early hearing detection as a standard of newborn care throughout the commonwealth. Following the passage of the IHEARR Act in November 2001, newborn hearing screening was implemented statewide beginning on July 1, 2002.

The Joint Committee on Infant Hearing's (JCIH) 2007 Position Statement on the Principles and Guidelines for Early Hearing Detection and Intervention (EHDI) Programs states that physiologic measures must be used to screen newborns and infants for hearing loss. Although there are several technologies available to screen newborns, JCIH recommends two technologies: automated auditory brainstem response (A-ABR) and otoacoustic emissions (OAE). A-ABR measures the brain's response to sound. Sound stimuli consisting of clicks or tones are administered to the baby through soft earphones, and electrodes are placed on the baby's head to measure the brain's response. OAE measures sound waves produced in the inner ear. Sound stimuli consisting of clicks or tones are administered, and a tiny probe placed just inside the baby's ear canal measures the inner ear's response. Both technologies provide a non-invasive recording of physiologic activity of normal auditory functioning. Also, both tests are painless and can be performed in five to ten minutes while the baby is sleeping or lying still. A single procedure or a combination of both procedures may be used for infants in the newborn nursery. For infants in neonatal intensive care units (NICU), the JCIH recommends A-ABR technology as the only

[1] "2016 Hearing Screening Summary," Centers for Disease Control and Prevention, https://www.cdc.gov/ncbddd/hearingloss/ehdi-data.html, (June 28, 2018)

[2] "Newborn Hearing Screening Fact Sheet," National Institutes of Health Fact Sheet-Newborn Hearing Screening, http://report.nih.gov/nihfactsheets/ViewFactSheet.aspx?csid=104&key=N#N, (October 2010)

appropriate technology. Since many infants with neural hearing loss are in this target population, the committee recommends this distinction for high-risk infants.

Six community health nurses (CHNs) in the Division of Newborn Screening and Genetics provide case management services for infants who did not pass the inpatient hearing screening. Case management services are provided until a hearing loss is either ruled out or the child is confirmed with a hearing loss and confirmed in early intervention (EI). Responsibilities include phone calls and letters to primary care providers (PCP), parents and audiologists to provide education and assist with follow-up services.

In July 2016, the program implemented a web-based Case Management System, iCMS. iCMS is a Neometrics software application designed to track and manage newborn screening results and follow-up processes. All case management activities including hearing screening, diagnostic evaluations and EI enrollment are recorded in iCMS. In addition, iCMS houses critical congenital heart defect and dried blood spot results and follow-up activities.

Since the passage of the IHEARR Act, the program has evolved into a system of organizations, stakeholders and professionals that enables Pennsylvania families to obtain a timely hearing screening and if needed, to obtain a comprehensive evaluation, treatment and intervention services at the earliest opportunity.

Program Objectives

The primary objectives of the program are to provide appropriate and timely screening, diagnosis and intervention to improve the quality of life for infants with permanent bilateral or unilateral hearing loss. Consistent with national EHDI initiatives and the recommendations of the JCIH in 2007, the goals of the program are:

- All newborns receive an inpatient screening between 24-48 hours after birth.
- If the newborn does not pass the inpatient screening, than an outpatient screening is performed within 1 month of age.
- If the newborn does not pass the outpatient screening, than a diagnostic hearing evaluation is completed within 3 months of age.
- If the child is diagnosed with a hearing loss, EI and family support services are started within 6 months of age.

Hearing Screening

In 2016, all Pennsylvania hospitals, birth centers and midwives with hearing screening equipment, submitted monthly aggregate hearing screening reports and screening results for babies not passing their hearing screening to the program (see Table 1 below). According to the data, of the 138,661 Pennsylvania babies born in 2016, 132,996 babies received a hearing screening. Of those receiving a hearing screening, 1,166 newborns did not pass their most recent screening. There were 5,665 newborns documented as not receiving a hearing screening, including reasons such as, but not limited to, parents declining the services, parents contacted but were unresponsive, inability to contact parents and unknown. These newborns may have received an unreported outpatient screening, were referred directly for a diagnostic evaluation, or did not receive further testing.

Table 1. Hearing Screening Performance

	2014	2015	2016
Total occurrent births	**141,406**	**140,146**	**138,661**
Total documented as screened	**137,282**	**134,986**	**132,996**
% of total births screened	97.1%	96.3%	95.9%
Total passed	**134,345**	**133,799**	**131,830**
% of total births passed	95.0%	95.5%	95.1%
Pass before 1 month of age	130,873	130,293	129,201
Pass after 1 month of age	-*	-*	588
Pass: age unknown	3,472	3,506	2,041
Total not passed	**2,937**	**1,187**	**1,166**
% of total births not passed	2.1%	0.8%	0.8%
Not pass before 1 month of age	1,846	495	432
Not pass after 1 month of age	1091	125	189
Not pass: age unknown	-*	567	545
Total documented as not screened	**4,124**	**5,160**	**5,665**
% of total births not screened	2.9%	3.7%	4.1%
Infant died	566	N/A	1
Non-resident	N/A	N/A	0
Unable to screen due to medical reasons	N/A	N/A	2
Parents/family decline services	507	543	612
Infant transferred and no documentation of screening	N/A	N/A	N/A
Homebirth	-*	2,824	2,620
Parents/family contacted but unresponsive	-*	109	106
Unable to contact	-*	82	30
Unknown	3,051	1,602	2,294

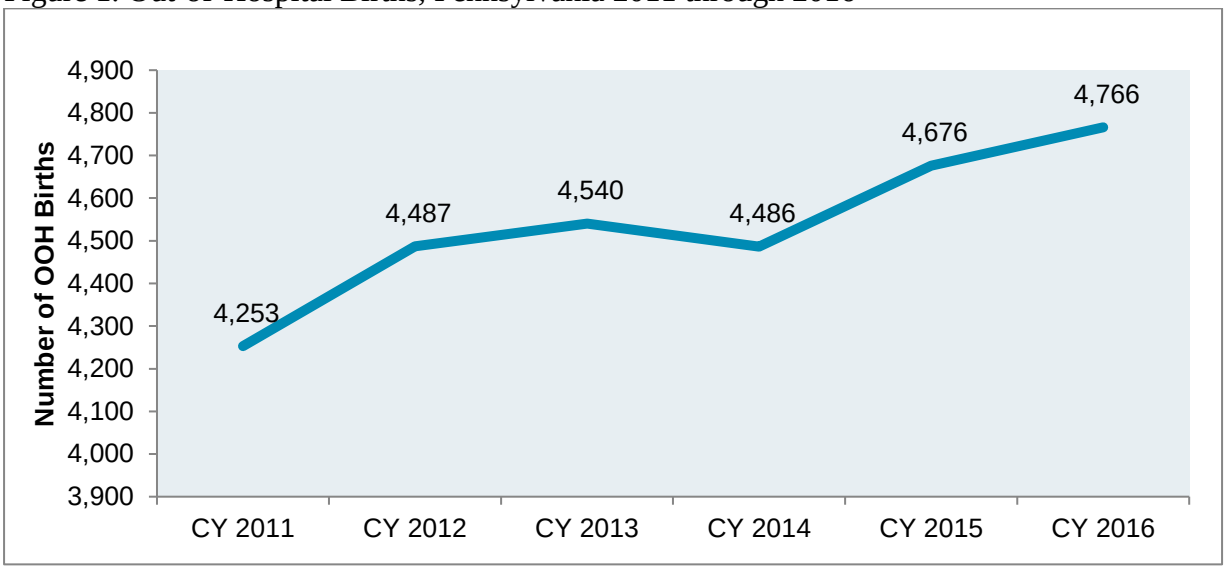
Source: PA EHDI Program Data, 2018

*Data field was not a requirement at time data was collected

Out-of-Hospital Newborn Hearing Screening

According to the Department of Health's Division of Health Informatics, out-of-hospital births have increased approximately 12 percent from 2011 to 2016 (see Figure 1 below).

Figure 1. Out-of-Hospital Births, Pennsylvania 2011 through 2016



Source: Pennsylvania Department of Health, Division of Health Informatics, 2018

To screen this growing population, portable hearing screening machines have been purchased and provided to birthing centers and midwives in areas with the highest geographic concentrations of out-of-hospital births with a focus on statewide dispersal. All licensed birth centers are equipped with hearing screening equipment. As of Oct. 9, 2018, 29 birth centers and midwives have received portable hearing screening equipment from the program. Representatives from the manufacturer of the portable hearing screening equipment and program staff provide hands-on training to midwife screeners. Each midwife signs an equipment agreement indicating they: have been trained on the use of the equipment; will use the equipment only for newborn hearing screening; agree to report data to the program; and agree to share the equipment (if applicable) with nearby midwives.

All midwives receiving equipment from the program have agreed to provide hearing screenings for other providers in their community who do not have equipment. Midwives without equipment can refer newborns to participating midwives and birth centers for an inpatient hearing screening and an outpatient hearing screening if the result of the inpatient screening is a referral. An infant can also be referred to a local hospital or audiologist that provides hearing screenings.

Diagnosis and Early Intervention

Table 2 below summarizes diagnostic information for 2016 births. The data for 2014 and 2015 births is provided for comparison. In 2016, 165 infants were diagnosed with permanent hearing loss. In 2015, 169 infants, and in 2014, 200 infants.

Table 2. Diagnostic Information

	2014*	2015	2016
Total not pass screening	**2,937**	**1,187**	**1,166**
Total with no hearing loss	**2,009**	**468**	**517**
% Not passed with no hearing loss	68.4%	39.4%	44.3%
No hearing loss before 3 months of age	1,731	339	385
No hearing loss after 3 months of age	266	121	131
No hearing loss documented: age unknown	12	8	1
Total hearing loss	**200**	**169**	**165**
% Not passed with hearing loss	6.8%	14.2%	14.2%
Hearing loss ID: before 3 months of age	126	102	108
Hearing loss ID: after 3 months but before 6 months of age	52	44	35
Hearing loss ID: after 6 months of age	20	12	22
Hearing loss ID: age unknown	2	11	0
Total without a diagnosis	**728**	**550**	**484**
% without a diagnosis	24.8%	46.3%	41.5%
Infant died	3	2	8
Non-resident	9	15	20
Unable to receive diagnostic testing due to medical reasons	0	5	3
Parents / family declined services	511	29	49
Parent / family Contacted but Unresponsive	32	177	228
Unable to contact	40	168	52
Unknown	133	154	124

Source: PA EHDI Program Data, 2018

* Hearing screening follow-up procedures and definition changes in 2015 may have contributed to the difference between 2014 and 2015-16.

Historically, the program focused on newborns from birth to final diagnosis. But as the EHDI system evolves, EI services have become a vital part of EHDI, as well as a point of emphasis in the new EHDI grant. Increasing the number of infants receiving a timely referral to and enrollment in EI services are now goals of the program. In past years, it was assumed that, if a baby was referred to EI, he/she was also enrolled in EI services. With iCMS launching in 2016, the program has the resources to properly track and evaluate EI referral and enrollment information.

In January 2018, the program and the Office of Child Development and Early Learning, Bureau of Early Intervention Services and Family Support (BEISFS), entered into an interagency agreement that allows family/child information to be shared to ensure that children receive timely access to EI services. The program provides information on all newborns, infants and toddlers diagnosed with a hearing loss to BEISFS. BEISFS then confirms that all those identified are receiving EI services. If the child is receiving EI services, BEISFS provides the program with the date the child was referred for EI services, the date of the initial Individualized Family Service Plan, and the county EI program that is responsible for serving the newborn, infant or toddler. If a parent or guardian has signed the Authorization to Release Information for Infants/Toddlers with Hearing Concerns, BEISFS notifies the program of newborns,

infants and toddlers receiving EI services that have a diagnosis of a hearing loss that were not previously identified by the program.

Table 3 below summarizes EI referral and enrollment information for 2016 births. The data for 2014 and 2015 births is provided for comparison. Of the 165 infants diagnosed with a permanent hearing loss in 2016, 155 of them were referred to EI. One hundred twenty-four of those infants were then enrolled in EI.

Table 3. Early Intervention Information

	2014	**2015**	**2016**
Total cases of hearing loss	**200**	**169**	**165**
Total referrals to Part C EI	**154**	**114**	**155**
% of hearing loss cases referred to Part C EI	77.0%	67.5%	93.9%
Referred to Part C EI before 6 months of age	138	66	52
Referred to Part C EI after 6 months of age	0	0	10
Age of referral unknown	16	48	93
Unknown referral status	**46**	**55**	**10**
Enrolled in Part C EI (based on signed IFSP)	**154**	**92**	**124**
% of hearing loss cases enrolled in Part C EI	77.0%	54.4%	75.2%
Enrolled before 6 months of age	138	66	59
Enrolled after 6 months of age	16	26	63
Signed IFSP: age unknown	-	-	2
Total with no documented EI services	**46**	**77**	**41**
% of hearing loss cases with no documented EI services	23.0%	45.6%	24.8%
Parents/family declined services	0	2	14
Unknown	46	75	27

Source: PA EHDI Program Data, 2018

Family Support and Partnership

Family support is an essential component of an effective EHDI system as reflected by a family support component in the previous two EHDI grants awarded to the program by Health Resources and Services Administration (HRSA). To improve family engagement, partnership and leadership within the EHDI system, the program provides programmatic and fiscal support to the Pa. Guide by Your Side (GBYS) program.

In July 2011, the program and the Pennsylvania Training and Technical Assistance Network launched the Hands and Voices Pennsylvania GBYS program. GBYS is dedicated to supporting families and their infants and toddlers who are newly identified with hearing loss by offering them an opportunity to talk or meet face-to-face with a parent guide. To qualify, guides must receive formal training and be a parent of a deaf or hard of hearing child. Strategically located throughout the commonwealth, parent guides bring their direct experience, specialized knowledge and personal compassion to their role, ensuring the newly diagnosed family's needs are their primary focus. Any family of a Pennsylvania infant or toddler (ages birth to 3) who has hearing loss is eligible for the program. More importantly, services are provided to the families at no cost. GBYS has expanded the program to include deaf mentorship as part of the services offered to families. To accomplish this, GBYS hired three parent guides who are themselves deaf or hard of hearing. Since the GBYS program's inception in November 2011, 985 families have received support services.

Matches between parent guides and families are based not only on geographic proximity but also on the similarity of diagnosis, hearing levels, communication strategies and technology choices, such as cochlear implants or hearing aids. Families have also been provided with opportunities to meet deaf adults through community events. The support provided to families who enroll in GBYS includes providing unbiased materials on communication strategies, face-to-face meetings, newsletters, support via telephone and email, informational teleconference training calls, loans of library materials, and postal mail of letters of support to families who do not use email. Parent guides have shared their children's stories with enrolled families through experience articles, photos and books.

The GBYS team has collaborated with many community partners to expand support received by enrolled families. Contacts and connections have been made with the Special Kids Network System of Care, Parent-to-Parent of Pennsylvania, state schools and programs for deaf and hard of hearing children, Hands and Voices chapter events, and other community-based events for deaf and hard of hearing children and their families. An example of other activities includes story times and play groups at local libraries.

Infant Hearing Screening Advisory Committee

The successful progress of the program is made possible by significant collaboration and input from the Infant Hearing Screening Advisory Committee. This six-member committee is appointed by the Secretary of Health. The committee makes recommendations to the program regarding infant hearing education, assessment, reporting and referrals. Issues include program regulation and administration, diagnostic testing, technical support, and follow-up for the program. The committee operates under a set of bylaws and meets four times per year. In addition to attending meetings, members provide program staff with ongoing advice and consultation on a variety of topics and occasionally serve as speakers at conferences, training workshops and presentations.

The committee is currently comprised of three audiologists, one educator for the deaf and hard of hearing, one otolaryngologist, and one parent advocate who has a young child with hearing loss. The committee is a valuable part of Pennsylvania's program.

Current members:
Carol Knightly, Au. D., Committee Chair
Director
Center for Childhood Communication at Children's Hospital of Philadelphia

Sarah Delano, Med., CCC-A
Audiology Department Coordinator of Newborn Hearing
Children's Hospital of Pittsburgh of UPMC

Erin Ellison
Parent Advocate
Mechanicsburg, Pennsylvania

Dennis Kitsko, DO, FACS, FAOCO
Otolaryngologist
Children's Hospital of Pittsburgh of UPMC

Margaret Santoro
Early Intervention Director/Coordinator at Pennsylvania School for the Deaf

Janet Juracich Trychin, Au.D., CCC-A
Associate Professor, Adjunct Status at Edinboro University

Program Initiatives

Beginning April 1, 2017, HRSA awarded the program with a three-year EHDI grant. The program is using the EHDI grant to support the development of statewide programs and systems of care that ensure deaf or hard of hearing children are identified through newborn and infant hearing screening and receive evaluation, diagnosis and appropriate intervention to optimize language, literacy and social-emotional development. Strategies that the program uses to reach the grant goal include: 1) increasing health professionals engagement and knowledge of the EHDI system by establishing a multidisciplinary advisory group and learning communities throughout the state; 2) increasing access to EI services and language acquisition by sharing data with BEISFS; 3) improving family engagement within the program by providing fiscal support to family support organizations; and 4) conducting quality improvement activities to improve data quality for the program.

The program implemented Phase II of iCMS in the fall of 2017, allowing hospitals, birth centers, midwives and outpatient hearing screening providers to submit all hearing screening results directly into iCMS via hearing screening equipment upload, HL7 messaging or manual data entry. As of June 2018, all birthing hospitals, birth centers and midwives with access to the internet were utilizing iCMS to submit hearing screening results electronically. Phase III of iCMS, which kicked off in July 2018, will provide access to audiologists to enter diagnostic evaluation results electronically. Audiologists are expected to receive iCMS training in the spring of 2019 and go live in the system by June 2019. Providing audiologist access to iCMS will eliminate the need for the current excel reporting workbooks that rely on secure email and fax.

On Jan. 1, 2018, the program began collecting individual level hearing screening results for all newborns. This includes information on non-screened newborns due to parent refusals, missed screens, newborns that were transferred to another hospital and expired newborns. As a result, data and case management services have greatly improved as the program receives all hearing screening results vs. reported referred results. The program compared the monthly aggregate reports with the results entered in iCMS for several months before discontinuing the monthly aggregate reporting requirement for iCMS users.

To ensure all newborns are given the chance to receive a hearing screening, the program worked with the Division of Vital Records (DVR) and Natus, the iCMS vendor, to develop a vital records reporting (VRR) module in iCMS. The VRR module allows an export from the birth certificate data system to be imported into iCMS and to be matched to each newborn's screening results in iCMS. This enables the program to ensure that all babies born in Pennsylvania receive the required newborn screens. If a newborn is identified by DVR, but does not have a record in iCMS, a record will be created in iCMS and follow-up actions will be completed to determine why the newborn did not receive the newborn screens and to ensure the newborn screens are completed. The program anticipates that this records match will help identify missed screenings, parent refusals and unscreened newborns due to expiration. The VRR module went live Oct. 31, 2018.

The program and the IHSAC are working in collaboration to revise the EHDI Best Practices for Newborn Hearing Screening and Follow-Up document to be separate instructions for submitters, audiologists and PCPs on hearing screening, diagnosis and reporting. As the instruction documents are finalized, they will be distributed to the appropriate stakeholder and placed on the program's website.

As a requirement of the current HRSA funding, the program established a multidisciplinary program advisory group, the EHDI Advisory Group (EHDI AG). The purpose of the EHDI AG is to provide advice to the program on potential mechanisms to achieve objectives of the EHDI grant. Due to member limits of the IHSAC, the EHDI AG functions separately from the IHSAC; however, both groups meet quarterly on the same day. The EHDI AG includes stakeholders that reflect the comprehensive EHDI system. Members include, but are not limited to, clinicians who deliver pediatric primary care, teachers at a school for the deaf or hard of hearing, EI providers, audiologists, and parents/families of deaf or hard of hearing children. The EHDI AG is required to be comprised of a minimum of 25 percent parents/family members of infants/children who are deaf or hard of hearing and/or deaf or hard of hearing individuals.

Contact Information

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