

Early Hearing Detection and Intervention Program Report on Screening and Follow-up for 2021 Births

Bureau of Family Health
Division of Newborn
Screening and Genetics

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Introduction

In 2020, 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 three months of age and intervention initiated by six 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 may 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 in- and out-of-hospital screenings for infants born in 2021, follow-up activities for infants referred to the Department for failure to pass their newborn hearing screening from 2019 through 2021, and the status of ongoing Early Hearing Detection and Intervention (EHDI) program (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, in 1999 the Department began a pilot program to screen infants for hearing loss with 26 birthing hospitals. 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.

Beginning April 1, 2020, the Health Resources and Services Administration (HRSA) awarded the program with a four-year EHDI grant. The goal of the EHDI grant is to ensure all newborns are screened for hearing loss, all infants that are referred for follow-up of their hearing screening receive a timely evaluation and diagnosis, and that all infants and toddlers identified as deaf and hard of hearing (DHH) are enrolled in Part C Early Intervention (EI) services. Strategies that the program uses to reach the grant goal include: 1) engage and coordinate all stakeholders in the EHDI system; 2) engage, educate, and train health professionals and service providers in the EHDI system; 3) strengthen the program's capacity to provide family support and engage families with children who are DHH as well as DHH adults; and 4) facilitate improved coordination of care and services for families and children who are DHH. This grant will expire on March 31, 2023; the program has submitted an application for the subsequent five year EHDI grant.

¹ 2020 Summary of National CDC EHDI Data. cdc.gov. 2020 Summary of National CDC EHDI Data | Annual Data EHDI Program | CDC Published September 20, 2023. Accessed October 13, 2023.

² Newborn Hearing Screening Fact Sheet. nih.gov. <https://archives.nih.gov/asites/report/09-09-2019/report.nih.gov/nihfactsheets/ViewFactSheet377f.html?csid=104&key=N#N>. Published June 30, 2018. Accessed October 15, 2023

The Joint Committee on Infant Hearing's (JCIH) 2019 Position Statement on the Principles and Guidelines for Early Hearing Detection and Intervention 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 10 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 appropriate technology. Since many infants with neural hearing loss are in this target population, the committee recommends this distinction for high-risk infants.

In Pennsylvania, six community health nurses (CHNs) in the Department's 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 infant is determined not to have hearing loss or the child is confirmed to have hearing loss and is confirmed to be enrolled in Part C early intervention, unless a family declines services or otherwise cannot be reached. Responsibilities of CHNs 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, enrollment in family support services, 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 infants to obtain a timely hearing screening and, if needed, to obtain a comprehensive evaluation, as well as 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 2019, 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, then an outpatient screening is performed by one month of age.
- If the newborn does not pass the outpatient screening, then a diagnostic hearing evaluation is completed by three months of age.

- If the child is diagnosed with a hearing loss, EI and family support services are started by six months of age.
- When possible, the program will support and encourage hearing screening through the age of three years as a means of identifying children with delayed onset hearing loss.

Hearing Screening

In 2021, all Pennsylvania hospitals, birth centers, and midwives submitted individual level hearing screening results for all births directly into iCMS (Table 1. Hearing Screening Performance). According to the data, of the 132,153 Pennsylvania babies born in 2021, 129,655 (98%) babies received a hearing screening. Of those receiving a hearing screening, 1,573 newborns did not pass their most recent screening. There were 2,498 newborns documented as not receiving a hearing screening, including reasons such as, but not limited to, parents declining the services, parents contacted but 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

	2019	2020	2021
Total occurrent births	134,074	130,173	132,153
Total documented as screened	131,440	127,406	129,655
% of total births screened	98.0%	97.8%	98.1%
Total passed	130,430	126,333	128,082
% of total births passed	97.3%	97.1%	96.9%
Pass before 1 month of age	127,387	122,936	124,895
Pass after 1 month of age	3,043	3,397	3,187
Total not passed	1,010	1,073	1,573
% of total births not passed	.75%	.82%	1.2%
Not passed before 1 month of age	794	858	1,302
Not passed after 1 month of age	216	215	271
Not passed: age unknown	0	0	0
Total documented as not screened	2,634	2,767	2,498
% of total births not screened	2.0%	2.1%	1.9%
Straight to Diagnostic Evaluation	0	45	153
Infant died	654	578	609
Non-resident	36	55	72
Unable to screen due to medical reasons	37	39	4
Parents/family decline services	1,221	1,320	1,180
Infant transferred and no documentation of screening	30	28	0
Homebirth	0	0	0
Parents/family contacted but unresponsive	21	53	117*
Unable to contact	302	379	351
Unknown	41	53	12
Other	292	217	0

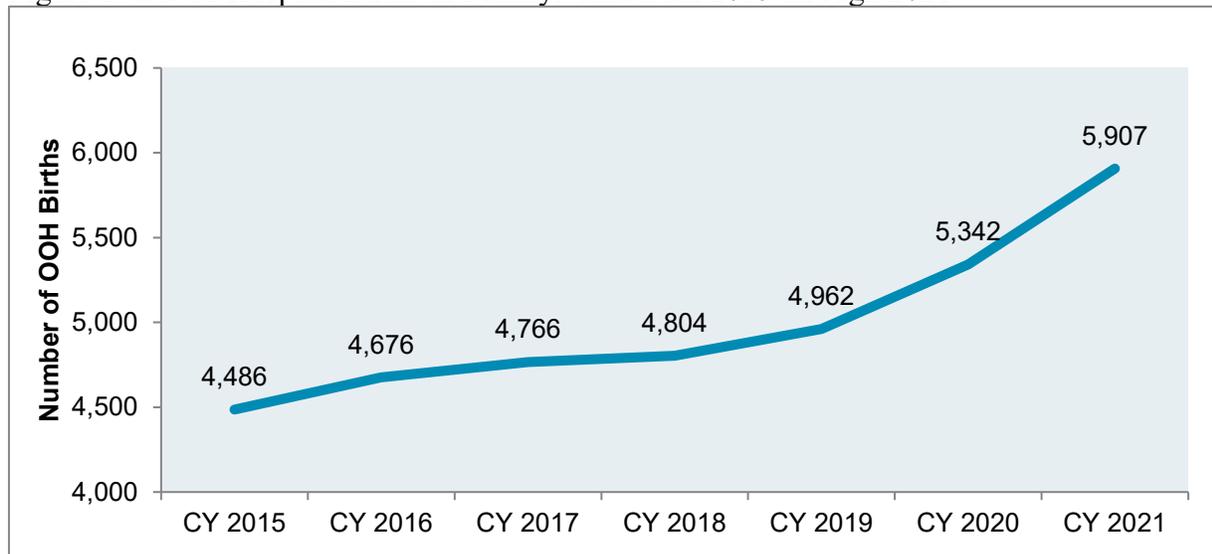
Source: PA EHDI program data, 2023

*Program data for hearing screening from 2021 is impacted by the ongoing effects of the COVID pandemic, including increased delay times in many referral processes, permanent closures of some facilities, staffing challenges across the state, and an increased resistance to attend follow-up visits due to concern over exposure.

Out-of-Hospital Newborn Hearing Screening

According to the Department of Health’s Division of Health Informatics, out-of-hospital births have increased approximately 31% percent from 2015 to 2021 (see Figure 1, Out-of-Hospital Births in Pennsylvania from 2015 through 2021 below).

Figure 1. Out-of-Hospital Births in Pennsylvania from 2015 through 2021



Source: EHDI program data, 2023

To screen this growing population, portable hearing screening machines were purchased by the Department using federal grant funds and provided to birthing centers and midwives statewide in areas with the highest geographic concentrations of out-of-hospital births. All licensed birth centers are equipped with hearing screening equipment. As of September 2023, 68 birth centers and midwives have received portable hearing screening equipment from the program, and plans are in place to distribute approximately 10 additional pieces of equipment prior to the end of the current program year. 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: 1) have been trained on the use of the equipment; 2) will use the equipment only for newborn hearing screening; 3) agree to report data to the program; and 4) agree to share the equipment (if applicable) with nearby midwives.

Midwives without equipment can refer newborns to local midwives with hearing screening equipment to receive an inpatient hearing screening as well as an outpatient hearing screening, if needed. An infant can also be referred to a local hospital or audiologist that provides hearing screenings.

Diagnosis and Early Intervention

Table 2. Diagnostic Information below summarizes diagnostic information for 2021 births. The data for 2019 and 2020 births is provided for comparison. In 2021, 240 infants were diagnosed with permanent hearing loss. In 2020, 178 infants were diagnosed; in 2019, 162 infants were diagnosed.

Table 2. Diagnostic Information

	2019	2020	2021
Total not pass screening/requiring diagnosis	1,010	1,118	1,726
Total with no hearing loss	563	424	566
% Not passed with no hearing loss	55.7%	37.9%	32.8%
No hearing loss before 3 months of age	480	336	478
No hearing loss after 3 months of age	83	88	88
No hearing loss documented: age unknown	0	0	0
Total with permanent hearing loss	162	178	240
% Not passed with hearing loss	16.0%	15.9%	13.9%
Hearing loss ID: before 3 months of age	115	134	164
Hearing loss ID: after 3 months but before 6 months of age	35	30	39
Hearing loss ID: after 6 months of age	23	14	37
Hearing loss ID: age unknown	0	0	0
Total with transient hearing loss*	NA	91	141
% with transient loss	NA	8.2%	8.2%
Total without a diagnosis	285	425	779
% without a diagnosis	28.2%	38%	45.1%
Audiological diagnosis in process	39	33	0
PCP/ENT did not refer infant for diagnostic	6	0	0
Infant died	0	0	5
Non-resident	24	37	47
Unable to receive diagnostic testing due to medical reasons	2	1	3
Parents/family declined services	21	25	41
Parent /family contacted but unresponsive	121	84	366**
Unable to contact	72	174	172
Unknown	0	71	139**
Other*s	0	0	6

Source: PA EHDI program data, 2023

*This category was introduced to the annual CDC hearing screening data report in 2021.

** Program data for diagnostics, referral, and intervention from 2021 is impacted by the ongoing effects of the COVID pandemic, including increased delay times in many referral processes, permanent closures of some facilities, staffing challenges across the state, and an increased resistance to attend follow-up visits due to concern over exposure.

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 current 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

they were 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 Part C 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 Part C EI services. If the child is receiving Part C EI services, BEISFS provides the program with the date the child was referred for Part C EI services, the date of the initial Individualized Family Service Plan (IFSP), 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 Part C EI services that have a diagnosis of a hearing loss not previously identified by the program.

Table 3. Part C Early Intervention Information below summarizes Part C EI referral and enrollment information for 2021 births. The data for 2019 and 2020 births is provided for comparison. Of the 240 infants diagnosed with a permanent hearing loss in 2021, 166 were referred to Part C EI; 138 of those infants were then enrolled in Part C EI.

Table 3. Part C Early Intervention Information

	2019	2020	2021
Total cases of permanent hearing loss	162	178	240
Total referrals to Part C EI	135	135	166
% of hearing loss cases referred to Part C EI	83.3%	74.8%	69.2 %
Referred to Part C EI before 6 months of age	117	117	139
Referred to Part C EI after 6 months of age	18	18	27
Age of referral unknown	0	0	0
Not referred to Part C EI	16	25	46
Unknown referral status	11	18	28
Enrolled in Part C EI (based on signed IFSP)	113	120	138
% of hearing loss cases enrolled in Part C EI	69.7%	67.4%	57.5%
Enrolled before 6 months of age	89	98	104
Enrolled after 6 months of age	24	22	34
Signed IFSP: age unknown	0	0	0
Total with no documented EI Part C services	49	58	102
% of cases with no documented EI Part C services	30.2%	32.6%	42.5%
Infant died	0	0	0
Non-resident	4	9	21
Parents/family declined services	14	27	33
Parent/family contacted but unresponsive	1	0	6
Unable to contact	27	16	27
Unknown	3	6	15

Source: PA EHDI program data, 2023

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 three EHDI grants awarded to the program by HRSA. To improve family engagement, partnership and leadership within the EHDI system, the program provides programmatic and fiscal support to the Early Intervention and Technical Assistance (EITA) program to provide family support throughout Pennsylvania.

In July 2011, the program and the EITA launched the Hands and Voices Pennsylvania Guide by Your Side (GBYS) program. The PA GBYS program then changed its name in the Spring of 2020 to Family Connections for Language and Learning (FCLL) program. The FCLL 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 and deaf mentor. To qualify, guides must receive formal training and be a parent of a child who is deaf or hard of hearing. Mentors must also receive formal training and be deaf or hard of hearing. Strategically located throughout the Commonwealth, parent guides and mentors bring their direct experience, specialized knowledge and personal compassion to their role, ensuring the families with newly diagnosed infant and toddlers have their needs met as . Any family of a Pennsylvania infant or toddler (ages birth to three) who has hearing loss is eligible for the program. More importantly, services are provided to the families at no cost. Since the FCLL program's inception in November 2011, 1,800 families have received support services.

Matches between parent guides, mentors, 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 also are provided with opportunities to meet deaf adults through community events. The support provided to families who enroll in FCLL 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 FCLL 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 children who are deaf and hard of hearing 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.

Current and Ongoing Program Initiatives

To ensure all newborns are given the chance to receive a hearing screening, the program worked with the Department's Bureau of Health Statistics and Registries' 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 VRR module went live in the fall of 2018.

Phase III of iCMS was implemented in the spring of 2019 and provides access to audiologists to enter diagnostic evaluation results directly into iCMS. Audiologists received iCMS training in the spring of 2019 and went live in the system in June 2019. Providing audiologist access to iCMS eliminates the need for the audiologists to perform manual entry, secure email and fax. Additionally, family support staff received iCMS access and training in July of 2020. Family support staff now provide updates to outstanding screening and diagnostic results, EI referral and enrollment information and family support status directly to the program through iCMS. In 2023, the program began the process of adding roles to grant iCMS access to state level EI staff to facilitate the existing data share.

Also, in the spring of 2019, the program and the IHSAC revised 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. The instruction documents were then distributed to the appropriate stakeholder and placed on the program's website. Website content has also been revised to address updated best practices information and most recent JCIH position statement guidance.

The program established a multidisciplinary program advisory group, the EHDI Advisory Group (EHDI AG), in 2016. 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. The EHDI AG includes stakeholders who 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 children who are deaf or hard of hearing. 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. The EHDI AG meets biannually, and the IHSAC meets quarterly.

In 2022 and 2023, an audiology sub-committee of the IHSAC developed three learning modules focused on EHDI basics, diagnosing newborns, and the audiologist's role within the iCMS system, including the importance of reporting diagnostic data to the program. These modules will be used to help educate audiologists throughout the state.

The EHDI program continues to foster and support five regional Learning Communities (LCs) throughout the Commonwealth. The LCs are chaired by members of the EHDI AG or other program

partners, and meet quarterly to address EHDI issues within their region. The five LC's are based in Pittsburgh (covers south-western PA), Erie (covers north-western PA), Scranton (covers north-eastern PA), Harrisburg (covers south-central PA), and Allentown (covers the Lehigh Valley).

The program continues to maintain and update a newborn hearing screening focused website, which has been revamped and updated for easier navigation. The website includes sub-pages for providers, families, and program information. Each sub-page includes a section of quick links to important program partners and resources, including (but not limited to) Early Intervention, Family Connections for Language and Learning, the National Center for Hearing Assessment and Management, and the Centers for Disease Control. Each sub-page also includes a section of useful documents, including (but not limited to) best practice guidelines, family roadmaps, referral forms, and educational packages.

In 2022, Pennsylvania passed the Cytomegalovirus (CMV) Education and Newborn Screening Act (2022 Act 29). 2022 Act 29 requires hearing screeners to offer CMV screenings to all newborns who do not pass their in-patient hearing screen, and report basic screening information to the Department of Health. In addition, 2022 Act 29 requires the Department of Health to share information and education about CMV and CMV screenings. The program began collecting preliminary CMV reporting data in 2022 and has sourced and created necessary CMV information and education, which is provided on the program's website. In 2022 and 2023, a Cytomegalovirus (CMV) subcommittee of the IHSAC developed and published best practice guidelines for screening and reporting CMV to the program. These guidelines include specifics for both hospital and out-of-hospital screeners, and are available on the program's website.

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