NEWBORN SCREENING IN NEBRASKA

Newborn Bloodspot Screening for Metabolic and Inherited Disorders

and

Early Hearing Detection & Intervention

2017 Annual Report

NEBRASKA
Good Life. Great Mission.
DEPT. OF HEALTH AND HUMAN SERVICES

2017 Annual Report Newborn Bloodspot Screening

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UNMC Resident's Care by UNMC as Child Put Her on a Path to Medicine

Jennifer Harney, M.D., of Aurora, Neb., is an example of how a medical discovery and Nebraska health professionals impact lives. She will graduate this summer from residency training at the University of Nebraska Medical Center then plans to practice family medicine in Aurora at Memorial Community Health.

It was the care of her physicians who saved her life and influenced her to become a physician.

Her journey started as a newborn when she tested positive for Phenylketonuria (PKU).

"If I didn't adhere to a special diet, the phenylalanine would build up in my bloodstream and in my brain and cause irreversible, severe brain damage," said Dr. Harney, who graduated in 2011 from UNMC's medical school. "PKU causes severe cognitive impairment – seizures and developmental disabilities."

"I can't break down phenylalanine, that means I can't have regular protein since it's in all protein – a building block. My life has consisted of being on a really strict, low protein diet. I can have about six grams of protein a day. A typical adult eats about 50 to 60 grams. You think of the typical things we eat – cheese, chicken, fish."
While growing up, she had to make weekly visits to her pediatrician for blood tests. At least once a year she would go to Omaha for a specialty visit at the metabolic clinic at the Munroe-Meyer Institute at UNMC.

That's how she met the late Hobart Wiltse, M.D., a pediatrician/metabolic specialist who was instrumental in getting Nebraska to implement the screening test for inherited disorders in newborns.

Jen Harney, MD, holds pictures of herself and the late Dr. Hobart Wiltse.

It's estimated that she and hundreds of other Nebraskans are the beneficiaries of mandated screening testing 50 years ago in Nebraska that required of newborn babies for PKU, an inherited disorder.

"If it were not for him, and the treatment they gave me, I would be in a wheelchair having seizures," Dr. Harney said. "He was a big inspiration for coming into the medical field. He changed patients' lives, families and the people he taught, the students, residents, his

co-workers - that just really stuck with me and that's the doctor I hope to become."

Dr. Wiltse told her that he always wanted one of his PKU patients to become a physician and that he thought she would be the first one.

Research continues to find ways to make life better for patients. Dr. Harney participated in a clinical trial for three years headed by William Rizzo, M.D., before she became pregnant. A medication enabled her to eat anything she wanted. The medication has submitted for approval by the Food and Drug Administration. She said approval would be huge for patients with PKU.

Dr. Harney and her husband welcomed their first born and good news last October. Though son, Finn, is a PKU carrier, he does not have the disorder.

Credit: Vicky Cerino, Media Relations Coordinator, Department of Public Resources UNMC, First published in "UNMC Today"

NEWBORN SCREENING FOR INBORN ERRORS OF METABOLISM AND INHERITED DISORDERS

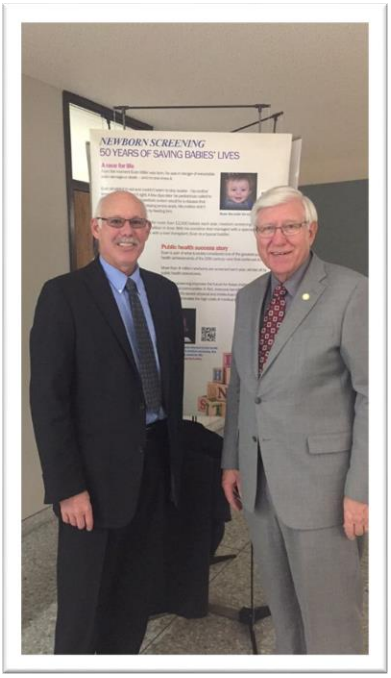
Nebraska Celebrates 50 Years of Newborn Screening in 2017

Nebraska marked 50 years of Newborn Screening with a press release, display and educational materials prominently posted in the Nebraska State Office Building, Saint Elizabeth's Regional Medical Center and Methodist Women's Hospital during the month of January. The information helped get the word out about how much the program has grown from screening all babies for one disease- PKU- in 1967 to screening all babies for 29 diseases in 2017.

Legislation Expands the Newborn Screening Panel

Senator Robert Hilkemann and several co-sponsors introduced and successfully passed legislation in 2017 that will require adding three new conditions to Nebraska's Newborn Screening Panel in July of 2018.

Constituent Robert Rauner, President of the Adrenoleukodystrophy Foundation, worked with the Senator to identify X-ALD as an appropriate candidate for the required screening panel along with two lysosomal storage diseases (Pompe Disease and Mucopolysaccharidosis Type I). These three conditions were recommended by the federal Advisory Committee on Heritable Diseases in Newborns and Children, to be included in the Recommended Universal Screening Panel. The support of Nebraska's Advisory Committee was paired with conditional recommendations that additional resources were needed because multiple additions had been made to the panel in the prior 14 years, without additional personnel and funding resources. The legislation helped address that by providing the ability for an increase in the newborn screening administrative fee up to $20 per infant screened. The three new conditions will bring Nebraska's screening panel to 32 diseases beginning July 1, 2018.



Robert Rauner (L) and Senator Robert Hilkemann (R)



The goal of newborn blood spot screening is to identify newborns at risk for life threatening and debilitating conditions that would otherwise not be detected until damage has occurred, and for which intervention and/or treatment can improve the baby's outcome.

If not detected and treated in time, the effects of conditions that are screened for can include brain and nerve cell damage resulting in severe intellectual disability, damage to the infant or child's heart, kidney, liver, spleen, eyes, and/or hearing. physical growth problems, stroke and even infant death can also occur. For these very good reasons, Nebraska's statute governing newborn screening ***Neb. Rev. Stat.*** *§§71-519 through 71-524 requires every baby born in Nebraska to receive the screening.*

Newborn Screening is a system coordinated by the Nebraska Department of Health and Human Services (NDHHS) Newborn Screening Program. The system requires collaboration with hospitals, laboratories, health care professionals, and families and involves many elements including:

- Education of health care professionals and parents, and efforts to increase public awareness.
- Proper and timely collection of quality specimens.
- Appropriate and timely transport of specimens to the Newborn Screening laboratory.
- Rapid quality testing methods.
- Timely notification of the infant's physician and parents.
- Timely recall of the infant for confirmatory or repeat testing.
- Appropriate referral of family to specialists for diagnosis, treatment and counseling.
- Ensuring access to needed specialized services and treatment.
- Evaluation and Quality Assurance/Quality Improvement efforts.

Each of these components of the system requires ongoing monitoring to ensure quality.

Nebraska's newborn screening panel targeted 29 core conditions but several more may be detected on the secondary panel.

In 2017 this resulted in identifying and treating 39 newborns in time to prevent or reduce problems associated with identified conditions.

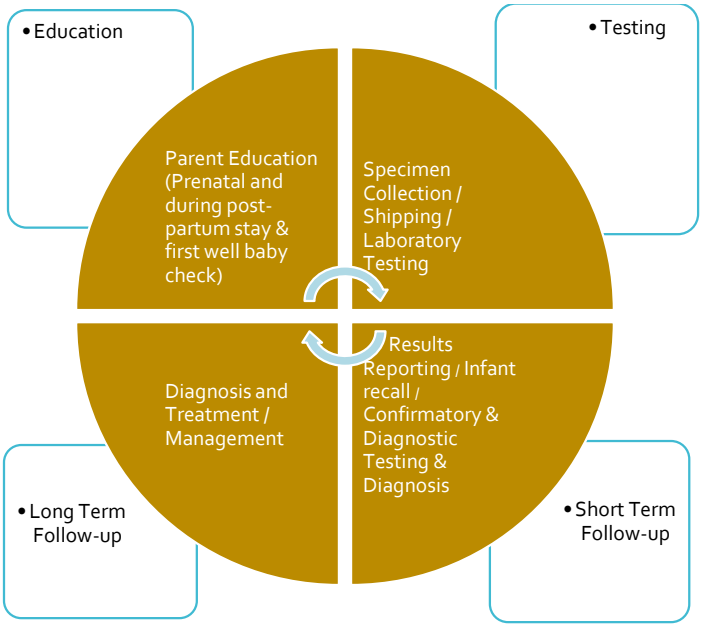
Newborns were diagnosed with these conditions from screening in 2017:

- 4 babies with partial biotinidase deficiency
- 3 babies with congenital primary hypothyroidism (CPH)
- 1 baby with hypothyroidism
- 1 baby with transient hypothyroidism (treated)
- 1 baby with positive screen for CPH, (treated, but diagnosis pending as of report publication)
- 6 babies with cystic fibrosis (CF)
- 1 baby with classical galactosemia (Gal)
- 1 baby with glutaric acidemia type 1 (GA I)
- 3 babies with sickle cell disease
- 3 babies with sickle hemoglobin C disease
- 2 babies with hemoglobin E disease
- 1 baby with hemoglobin E disease and alpha thalassemia trait
- 5 babies with medium chain acyl co-a dehydrogenase deficiency (MCAD)
- 1 baby with classical phenylketonuria (PKU)
- 1 baby with mild/benign hyperphe (not treated)
- 1 baby with short chain acyl co-a dehydrogenase deficiency (SCAD)
- 2 babies with transient tyrosinemia (treated) plus one not treated
- 2 babies with 3 methylcrotonyl carboxylase deficiency (3-MCC)
- 1 baby with severe combined immune deficiency (SCID)

While each condition is individually rare,
in Nebraska 1 in every 500-600 babies born
is affected with one of them.

System Overview

In 2017, fifty-four birthing facilities in Nebraska shipped specimens overnight for 26,254 babies Monday through Saturday to PerkinElmer Screening Laboratory (all but four hospitals had Saturday pick-up available to them). The program, administered by the DHHS, also partnered with pediatric specialists in genetics, metabolism, endocrinology, hematology, pulmonology, and immunology to connect primary health care providers for babies who had positive screens with the appropriate subspecialist for diagnosis and treatment. An advisory committee made up of experts from these subspecialties, pathology, chemistry, pediatrics, neonatology, family practice, as well as the Nebraska Hospital Association, Nebraska Medical Association, and family representatives advised the program on technical and policy issues, monitoring quality, and professional and parent education through their quarterly meetings. The follow-up staff ensured that the health care provider for every newborn who required additional testing received the necessary information to act accordingly and continued to follow-up with calls, faxed letters, and information until diagnosis was made or ruled out. The program conducted ongoing evaluation and quality assurance activities throughout the year. To ensure access to treatment, the program administered a contract for the ordering and distribution of metabolic formula, as well as the reimbursement system for metabolic foods for eligible individuals with PKU and other inborn errors in metabolism who require the specially manufactured foods.



Quality assurance activities in all of these areas help assure a successful newborn screening system.

MAJOR INITIATIVES IN NEBRASKA DURING 2017

❖ EDUCATIONAL INITIATIVES

- A Perinatal Grand Rounds presentation was given by Nebraska Newborn Screening Program (NNSP) staff at Saint Elizabeth Regional Medical Center and another presentation was given at the Practical Pediatrics Conference at the Century Link Convention Center in March, both presentations covered "Newborn Screening 101 Screening in the year 2017."

- In July 2017 a "Practitioner's Update" sent to all Family Physicians and Pediatric practices in Nebraska addressed the Practitioner's responsibilities for newborn screening. This issue focused on screening for Cystic Fibrosis and the needed follow-up given new guidelines. It also focused on screening for hemoglobinopathies, emphasizing the need for quicker responses for optimal treatment outcomes. It also introduced information about the three new diseases to be added in 2018.

- The NNSP continued to work on improving timeliness of all screening activities. A laminated poster encouraging new mothers to provide good contact information to the hospital was distributed to obstetrician offices throughout the state. Introducing newborn screening concepts early (before the baby is born) is intended to help parents with their responsibilities around newborn screening.

❖ QUALITY ASSURANCE AND IMPROVEMENT INITIATIVES

The NNSP continued its longstanding oversight and monitoring of multiple quality measures of hospital and laboratory performance while looking for ways to support hospitals in their QA/QI endeavors.

- The primary focus was on improving "TIMELINESS" which has become a national emphasis in the newborn screening field.

- The NNSP was awarded a second year "Timeliness" grant from the NewSTEPS program of the Association of Public Health Laboratories (APHL) and the Colorado School of Public Health. Using those funds, the program awarded $10,000 contracts to four hospitals to help them achieve bi-directional electronic interfaces with the newborn screening laboratory. Ultimately three hospitals achieved the interface and received the funding. The interface improves the ability of the laboratory to monitor which specimens it should be receiving, and the hospitals to receive the test results in real time, thereby making results available to the baby's physician even quicker.

- Hospitals received quarterly reports from the program on multiple measures comparing their performance to state-wide averages, as well as established benchmarks. Several hospitals and hospital systems actively addressed quality improvement initiatives within their own facilities, resulting in real improvements in processes and outcomes. During the second half of 2017, monitoring of the weekly

specimen handling delays and late specimen collection was re-initiated with the hiring of Sarah Seberger to the newborn screening program as a Follow-up/QA Specialist.

- Nebraska's newborn screening laboratory PerkinElmer Genetics Inc. (PEI) continued to provide its outstanding service and high quality testing. The laboratory IT department, worked with the new hospital partners on the bidirectional interface, which brought the percent of births occurring in hospitals with the interface up to 63%.

- The PEI Laboratory IT Department was quickly responsive to all requests for upgrades the NNSP requested for the IT system. These changes enhanced the ability of the program to perform timely follow-up: 1) provided access to demographic data the same day it is entered (previously available next day),2) added filters for "initial" vs. "repeat" specimen on the specimen batching/handling delay report to help sort out those that don't need to be reported to the hospital, 3) added the age at collection to the specimen batching/handling delay.

- The PEI laboratory Client Services Department also continued to provide excellent service. When the NNSP reinstituted the weekly batching/specimen delay monitoring, several instances were identified where hospitals were not sending shipments correctly for Saturday delivery at the laboratory, or were not properly arranging for Saturday pick up at their facility. The Client Services staff at PEI were tremendously helpful in resolving these issues.

- The newborn screening community is all about collaboration. Nebraska's program was called upon numerous times by one of PEI's new clients to learn about our data system capabilities for tracking and follow-up, as well as for quality assurance reporting.

- As part of the Timeliness grant, Nebraska continued to report aggregate data of timeliness measures to the NewSTEPS repository. Emphasis was on the percent of "critical positive" screen results reported by five days of age, and the percent of all results reported out by seven days of age.

- The Program applied for and was awarded an implementation grant (funding available till August 2018) to prepare to add three new conditions to the screening panel. (Pompe, MPS-I and X-ALD). This funding helped support a part time follow-up, part time quality assurance staff person. Adding this capacity allowed for the Program Manager and Senior Follow-up Coordinators to work with the lab and pediatric subspecialists to prepare the program, health professionals, and hospitals to successfully implement screening for the Lysosomal storage disorders of Pompe and MPS-I, and for X-linked Adrenoleukodystrophy. With the passage of authorizing legislation, the Program will be able to implement by July 1, 2018. By adding the QA/Follow-up staff person, the program will be able to absorb the additional follow-up for these three conditions.

HIGHLIGHTS FROM SOME KEY ELEMENTS OF NBS

❖ FOLLOW-UP A "CRITICAL KEY" TO THE SUCCESS OF NEWBORN SCREENING

The two follow-up coordinators at the state program, Krystal Baumert and Karen Eveans, MD and part time follow-up specialist Sarah Seberger track, monitor, and make sure babies' health care providers know what needs to be done to follow-up on hundreds of babies each year. They follow-up with phone calls, faxes, letters, and information sheets until either a satisfactory screen has been completed, an inconclusive result has had a repeat screen, a positive result has received confirmatory testing, and a diagnosis is either made or ruled out. In 2017 alone this was an essential function for more than 4000 results or distinct reasons for follow-up. Approximately half of those were specimens collected early (NICU admissions) as required to avoid interference from parenteral nutrition and transfusions.

The NNSP Follow-up personnel are highly collaborative. In today's world with multiple baby name changes, and baby's physicians changing due to parent choice or insurance coverage changes, the follow-up personnel perform an essential function to ensure the correct health care professional has all the appropriate information needed to follow-up. This means ensuring the right test at the right time occurs for every baby who needs it, and appropriate consultation and referrals are made.

❖ ADVISORY COMMITTEE

The Newborn Screening Advisory Committee conducted its annual review of quality assurance data of pre-analytical (e.g. unsatisfactory specimen rates and types), analytical (e.g. statistical performance of assays over time), and post-analytical (e.g. age at time of intervention or treatment for diagnosed patients, as well as long-term outcomes) performance measures for the system. Annually the committee also reviews the All-Hazards Contingency/Emergency Management Plan for newborn screening, and the program's education plan and strategies. The committee monitors national recommendations, trends, and reports, advising the program on recommended next steps, methods, or strategies. In 2017 the Committee supported legislation adding three conditions to the required newborn screening panel:

Pompe, MPS-I and X-ALD, with the caveat that additional funding and personnel resources were needed in order to maintain the program's high quality.

Much of Nebraska's success can be directly tied to the Committee's recommendations and guidance. Members committed at least a half day every three months to advise the state program. Representatives from PerkinElmer Genetics Laboratory Inc. regularly provided input, presentations, and proposals to the advisory committee. Several members provide extensive review and consultation beyond the committee meetings to help the program meet the recommendations of the larger committee. Nebraska's newborn population, families, and the program all benefit from the advisors who provided technical expertise and policy guidance to the Nebraska Newborn Screening Program. A special thanks to Dr. Angela Brennan for chairing the Committee along with Dr. Samuel Pirruccello (UNMC) who served as Vice Chair in 2017. Special recognition was bestowed upon Dr. Kevin Corley, Pediatric Endocrinologist upon his retirement for his dedication and expertise having served on the Committee since the early 1990's.

2017 NBS Advisory Committee Members:

Khalid Awad, MD	Methodist Women's Hospital / Neonatology
Lawrence Bausch, MD	Nebraska Medical Association / Neonatology
Angela Brennan, MD	Howard County Medical Clinic / Family Practice
John Colombo, MD	Children's Specialty Phys / Pulmonary Medicine
Kevin Corley, MD	Children's Specialty Phys / Pediatric Endocrinology
Jeanne Egger	Parent / Advocate Advisor
Sara Frink	Parent / Advocate Advisor
James Harper, MD	Nebraska Medicine / Children's, Pediatric Hematology
Kathryn Heldt, RD, LMNT, CDE	Children's Hospital / Metabolic Management Clinic
Mary Kisicki, LPN	Parent / Advocate Advisor
Rose Kreikemeier, APRN	Children's Hospital / Metabolic Management Clinic
Richard Lutz, MD	Nebraska Medicine / Children's Metabolism/Genetics
Bev Morton	Parent / Advocate Advisor
Hana Niebur MD	Children's Specialty Phys / Pediatric Immunology
Deborah Perry, MD	Nebraska Methodist Hospital / Pathology Center
Samuel Pirruccello, MD	Nebraska Medicine /Pathology
William Rizzo, MD	Nebraska Medicine / Metabolism/Genetics
Kathy Rossiter, EJD, MSN, APRN	Medical Ethicist Advisor
Monica Seeland, RHIA	Nebraska Hospital Association
Jill Skrabal, MS, RD, LMNT, CDE	Metabolic Management Clinic Nebraska Medicine
Alina-Gabriela Sofronescu, PhD	Nebraska Medicine / Pathology and Microbiology
Lisa White, MD	Medicaid and Long Term Care Medical Director
B.J. Wilson, MD	SE Nebraska Neonatology Group / Neonatology

FINANCING NEWBORN SCREENING

The program uses state general funds, the newborn screening fee ($10/infant) and Title V Maternal and Child Health Block Grant funds to support access to treatment for the metabolic foods and formula. The Title V Block Grant also funds the administrative aspects of the program (education, follow-up, program management and quality assurance).

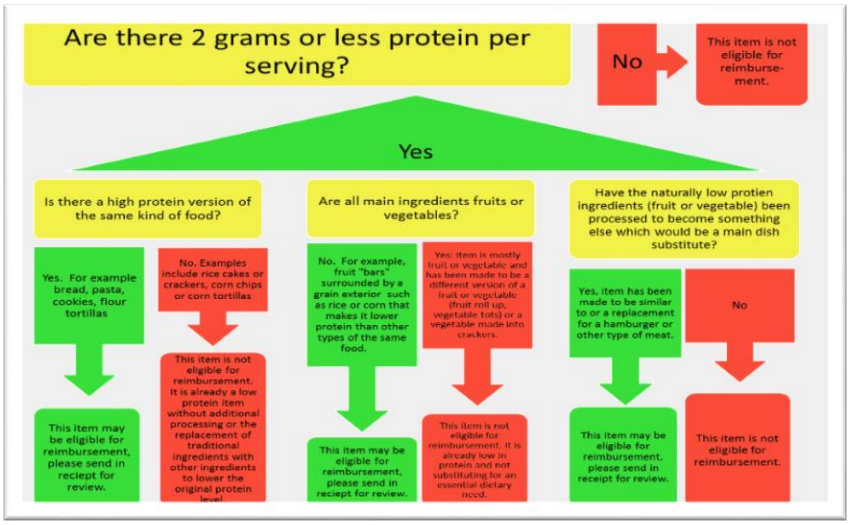
Treatment and Management of Inborn Errors in Metabolism



To fulfill the statutorily-required public health assurance role, NDHHS contracted with the metabolic clinic through Nebraska Medicine to provide nutritional counseling and monitoring, and distribution of the metabolic formula. The contract required insurance to be billed first, before billing the cost of the formula to NDHHS. Nebraska Medicine and Children's Home Health partner to achieve the distribution and billing. NDHHS reimburses for the unrecovered costs to provide formula, after insurance billing is completed.

Program staff coordinated the day-to-day pharmaceutically manufactured metabolic foods program. Most individuals participated by being reimbursed for eligible foods. For individuals who met financial hardship waiver criteria, the Department purchased the foods directly for them. It has proven more cost effective both in terms of the amount of time and effort to administer the foods program, and in helping the food dollars go further.

In 2017, the Staff Assistant Cathy Kearney, who coordinated the metabolic foods program, developed a decision tree to help families know which foods qualify under the program, and she also developed a guidance document for participating in the program. This document helps families know which foods are eligible for reimbursement, and which are best for meeting their children's unique nutritional needs.



Intervention Data

Collecting specimens correctly the first time, at the right time, and processing them for shipment is just the beginning. Working to optimize shipping times with the commercial overnight shipping company, and maximizing the efficiencies at the testing laboratory are also key to timely reporting of results on babies who need follow-up.

Several factors can conspire to create delays in treatment, so speed and persistence in follow-up are essential. Some examples of these factors include:

- prolonged treatment requirements for babies in NICUs,
- severe weather delays affecting shipment times,
- parental misunderstanding or other barriers to obtaining confirmatory testing,
- problems locating parents because of inaccurate contact information.

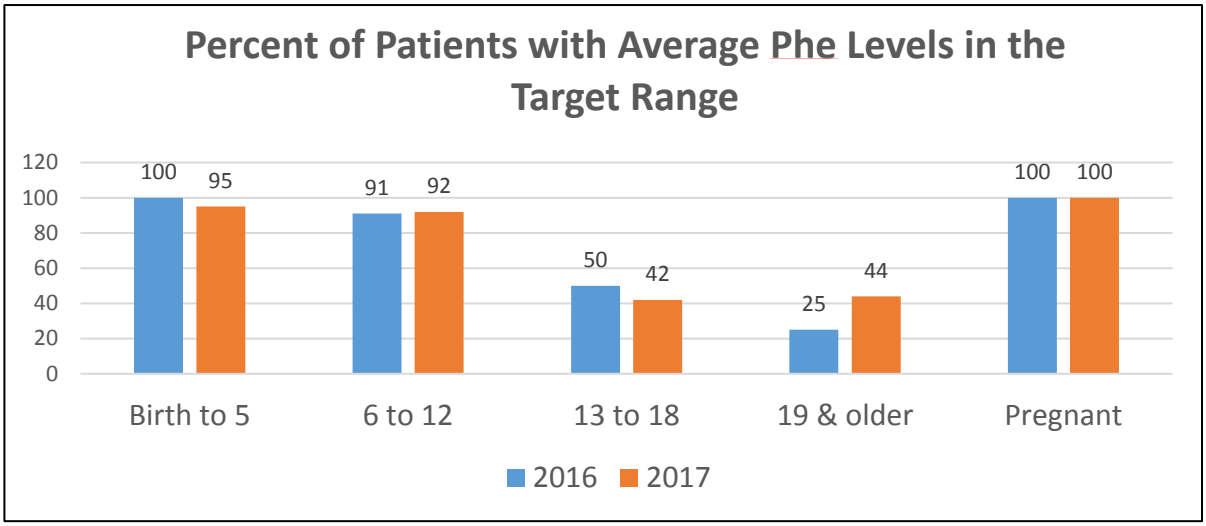
All parts of the system must work for babies to reap the most benefits of early identification, treatment, and intervention.

In 2017 the average age of intervention for babies identified with each condition were:

Partial Biotinidase Deficiency – 17 days	Hb-E Disease + Alpha Thal Trait – 66 Days
Congenital Primary Hypothyroidism – 6 days	3-Methylcrotonyl Carboxylase Deficiency (3-MCC)– 7 Days
Hypothyroidism – 26 Days	Medium Chain Acyl-CoA Dehydrogenase Deficiency (MCAD) – 4 Days
Transient Hypothyroidism – 72 Days	Phenylketonuria (PKU) – 3 Days
Cystic Fibrosis – 25 Days	Short Chain Acyl-CoA Dehydrogenase Deficiency (SCAD) – 18 Days
Galactosemia – 6 Days	Sickle Hemoglobin C - Disease – 23 Days
Glutaric Acidemia – 11 Days	Sickle Cell Disease – 19 Days
Hb-E Disease – 20 Days	Transient Tyrosinemia – 8 Days

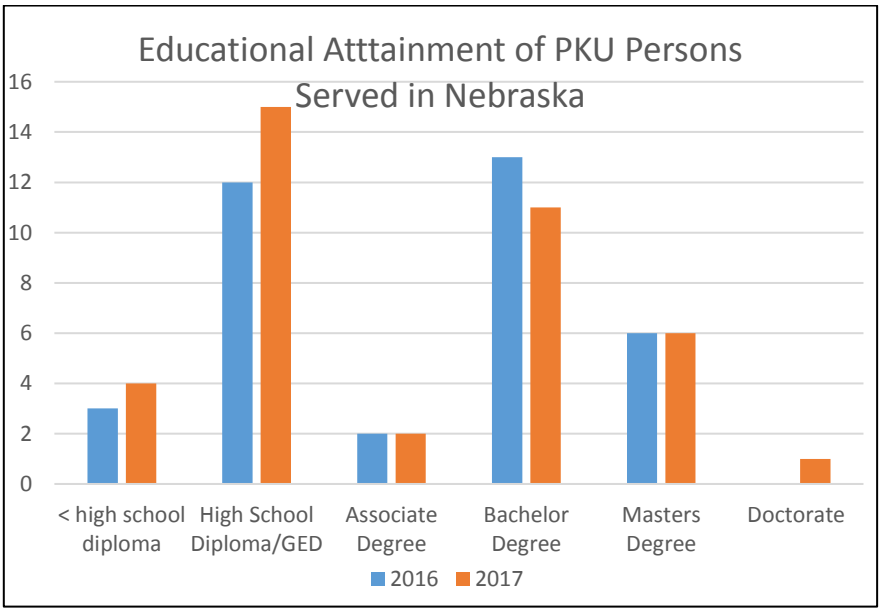
Outcome Data

Measures to evaluate patient outcomes are important for evaluating the effectiveness of the newborn screening system. Close monitoring and ensuring access to the metabolic formula and foods are essential elements to the success of our Nebraskans affected with inborn errors in metabolism.



Developmental Measures: Since reporting of long-term outcomes began in 2011, through 2017, 100% of children with PKU age 0-12 receiving services in Nebraska were assessed by the pediatric metabolic specialists as meeting all developmental milestones.

Outcome measures of educational achievement demonstrate the success of families, and individuals, who get early treatment for their metabolic conditions: made possible initially because of newborn screening:



Guest Article:

This July 1st (2018) will be a historic day in the lives of many Nebraskan families. The State of Nebraska will begin newborn screening for three new diseases, X-ALD, Pompe, and MPS1. This has been a three year effort to add these three diseases to the Nebraska newborn screening panel. The reason I became involved in the effort to add the diseases to the Nebraska panel is because my wife and I have a son who passed away from X-ALD and also have another son who has the adult form of the disease. Twenty four years ago our son Kevin was diagnosed with X-ALD and we were sent to the University of Iowa for confirmation of the disease. At the time of diagnosis, bone marrow transplant was the only option for treatment, if the disease had not progressed to the point that the transplant would only lead to an early death. We spent time involved in clinical trials that didn't lead to any successful options at that time. We also became involved with the United Leukodystrophy Foundation at the time of diagnosis, so we could be involved as the learning progressed to find treatment options for X-ALD. I have now become the president of the foundation and continue to work hard to help families that have been diagnosed with this terrible disease.

What does the passing of the addition of the new diseases to the newborn screening panel mean? The screening for these new diseases will mean that the families that are identified through this tool will not have to go through the diagnostic odyssey that our family had to go through to figure out the disease our son had. We spent three years treating our son for ADHD instead of knowing that he had X-ALD which would have given us an option if he was diagnosed early. Today's families will have the information immediately at birth and then they will have the time to prepare for the treatment options that are available today and for the options of treatments that will be developed over the next several years. Not only that, but these children will not become a burden on society because they can be a productive member of society. The other benefit of screening is that there are families that may be identified that are trying to find out what is wrong with their child or an older family member that has the adult form of the disease. I am excited for the families that will have an opportunity that our family did not have with our son, and I look forward to helping families that are diagnosed with X-ALD. Thank you to the State of Nebraska for adding these new diseases to the screening panel that will impact the lives of families in Nebraska.

Robert and Yvonne Rauner

NEBRASKA EARLY HEARING DETECTION AND INTERVENTION ANNUAL REPORT - 2017

The Nebraska Early Hearing Detection and Intervention Program develops, promotes, and supports systems to ensure all newborns in Nebraska receive hearing screenings, family-centered evaluations, and early intervention as appropriate.

Introduction

Photo courtesy of: Angie Marie Photography

Approximately one to three in 1,000 babies are born with permanent hearing loss, making hearing loss one of the most common birth defects in America. Before newborn hearing screening, children who were deaf or hard of hearing sometimes were not identified until 2½ to 3 years of age. Left undetected, this delayed identification can negatively impact the child's speech and language acquisition, academic achievement, and social and emotional development. If detected soon after birth, the negative impacts can be reduced and even eliminated through early intervention.

The Infant Hearing Act became a state law in Nebraska in 2000 and required the hearing screening of newborns in birthing facilities in Nebraska as a standard of care. Also in 2000, the Nebraska Department of Health and Human Services (DHHS) started the Nebraska Newborn Hearing Screening Program. Today the program is known as the Nebraska Early Hearing Detection and Intervention (NE-EHDI) Program and is funded through federal grants. This program strives to fulfill the following four main purposes of the Infant Hearing Act (Neb. Rev. Stat. §71-4735):

- To provide early detection of hearing loss in newborns at the birthing facility, or as soon after birth as possible for those children born outside of a birthing facility.
- To enable these children, their families, and other caregivers to obtain needed multidisciplinary evaluation, treatment, and intervention services at the earliest opportunity.
- To prevent or mitigate the developmental delays and academic failures associated with late detection of hearing loss.
- To provide the state with the information necessary to effectively plan, establish, and evaluate a comprehensive system for the identification of newborns and infants who have a hearing loss.

The act also requires birthing facilities to educate parents about newborn hearing screening and any necessary follow-up care. The education includes the hearing screening test, the likelihood of the newborn having a hearing loss, follow-up procedures, and community resources, including referral for early intervention and a description of the normal auditory, speech, and language developmental process in children. The act also requires that regulations be promulgated to mandate newborn hearing screening if less than 95% of newborns in the state receive a hearing screening.

Newborn Hearing Screening Data Reported for 2017 Nebraska Births

Birthing Facility Screening Programs

Since 2003, 100% of the birthing facilities in Nebraska have been conducting hearing screenings, consistent with the Neb. Rev. Stat. §71-4742 requirement that a hearing screening test be included as part of the standard of care for newborns. In 2017 there were 54 birthing facilities conducting hearing screenings.

Hearing Screening at Birthing Facilities and Birthing Centers

Photo courtesy of: NCHAM

In 2017, inpatient hearing screenings were reported on 26,071 newborns or 99.9% of the 26,099 newborns available for an inpatient hearing screening. The percentage of newborns screened during birth admission has increased dramatically since reporting began in 2000, when only slightly more than one-third of newborns received a hearing screening during birth admission.

In 2017, of those who received an inpatient screening in Nebraska, 25,084 (96.2%) passed the screening. An outpatient screening or audiology evaluation is recommended for infants who do not pass the inpatient screening or who do not receive the inpatient screening.

Parent Education

Recommending a hearing screening has been operationally defined as educating parents about newborn hearing screening, hearing loss, and normal communication development as required by Neb. Rev. Stat. §71-4740. The NE-EHDI Program provides print and video educational materials free of charge to hospitals to help fulfill this requirement. Print materials are available in 14 languages.

Birthing facilities reported educating approximately 99% of parents about newborn hearing screening, hearing loss, and normal speech and language development in 2017. The statute also requires the Nebraska Department of Health and Human Services to educate parents of newborns who are not born in a birthing facility about the importance of newborn hearing screening and to provide information to assist them in having the screening performed within one month after the child's birth. This is accomplished through letters and printed materials sent to the parents, along with phone calls.

Photo courtesy of: Cochlear Americas

When an infant is identified as deaf or hard of hearing, the parent(s) receive the Parent Resource Guide (PRG) from the audiologist and a notification letter is sent to the primary health care provider. Guide By Your Side (GBYS) Guides also discuss the PRG with families to provide support and stress the importance of starting early intervention as soon as possible.

The PRG was developed by NE-EHDI with the collaboration of many partners. Listening to feedback about the PRG, NE-EHDI also added an electronic version of the PRG to its website so parents have the option of receiving a hard copy or accessing the information electronically. Offering the online version has decreased the number of printed PRGs needed. This has also reduced the amount of time the NE-EHDI program staff spends assembling the PRG packets, and lowered the cost for printing and shipping. The online version also allows information to be revised and updated as needed, instead of updates not being available until materials are reprinted.

Monitoring, Intervention, and Follow-up Care

The NE-EHDI Program's tracking and follow-up processes are followed for each baby reported as not passing the hearing screening during birth admission and for infants not receiving the inpatient hearing screening. In 2017, a total of 1,060 infants (hospital and non-hospital births) were tracked by the Nebraska EHDI Program to encourage parents to have the infant receive an outpatient hearing screening or audiologic diagnostic evaluation.

The following shows the hearing screening/testing status of the 26,262 births for 2017:

25,992	Passed the screening or diagnostic testing (99% of births)
114	Expired (inpatient or outpatient)
43	Pending final screening or diagnostic testing
52	Diagnosed deaf or hard of hearing
31	Parents refused screening and/or diagnostic testing
18	Unresponsive (did not complete protocol after communication with NE-EHDI staff)
2	Lost (no response to NE-EHDI letters and phone calls)
9	Moved out of Nebraska
1	Late onset deaf or hard of hearing (passed initial screening)

Timeliness of Follow-up Screening / Evaluations / EDN Services

The purpose of the Infant Hearing Act (Neb. Rev. Stat. §71-4735) is to "… obtain needed multidisciplinary evaluation, treatment, and intervention services at the earliest opportunity and to prevent or mitigate the developmental delays and academic failures associated with late detection of hearing loss."

Photo courtesy of: Jordan Sochor Photography

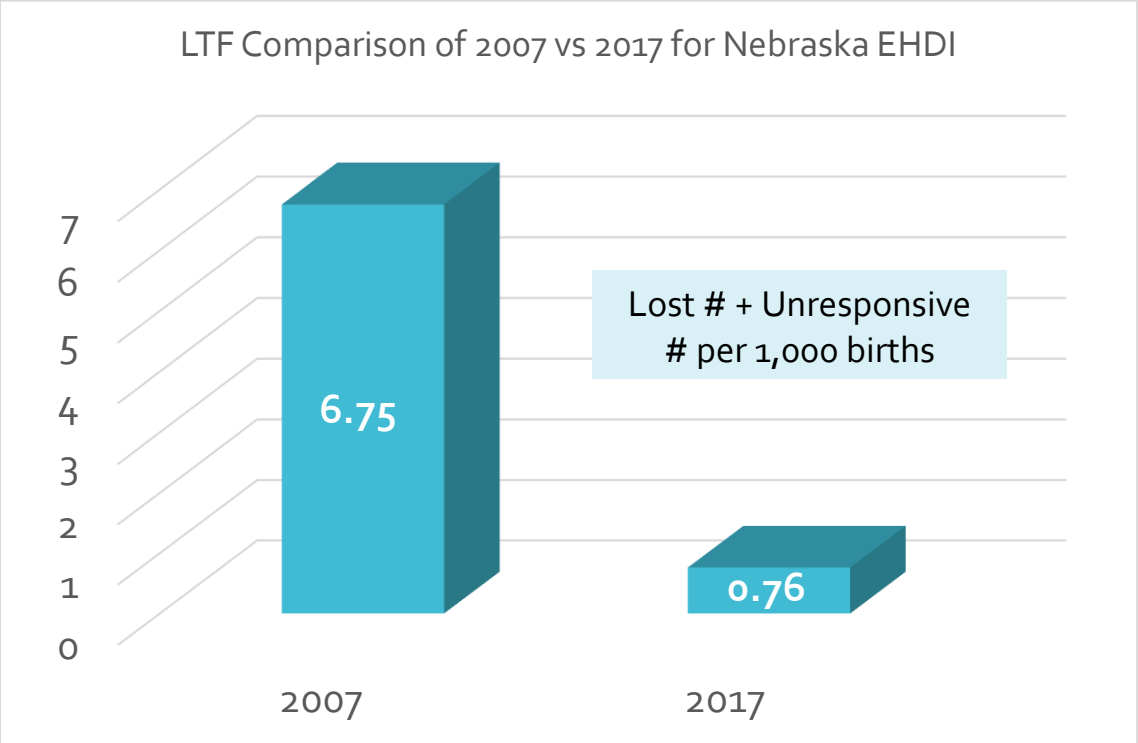
To meet the state and national guidelines of "1-3-6" (hearing screening completed by **1** month, audiologic diagnostic evaluation completed by **3** months, early intervention initiated by **6** months), established by the Joint Committee on Infant Hearing (JCIH), the timeliness of initiation and completion of follow-up activities is an important aspect of the quality of services. Over 97% of infants completed the inpatient/outpatient screening within one month of age. For the newborns who were recommended for an audiologic diagnosis, nearly 61.9% received the evaluation by 3 months of

age according to individual data reported to the NE-EHDI Program by audiologists (as of May 2018). This percentage is preliminary and will most likely decrease due to the number of infants having an initial diagnostic evaluation and needing a confirmatory evaluation.

Note: Because 43 hearing records are still pending the final screening and diagnostic testing results, the "1-3-6" numbers above are preliminary as of May 2018 and will most likely change.

Records for the Early Development Network (EDN), Nebraska's Part C Early Intervention Program, indicate that 90.9% of infants residing in Nebraska in 2017 diagnosed as deaf or hard of hearing, were enrolled in EDN services by 6 months of age for families accepting Part-C services (as of May 2018). It is projected that the final percentage for enrollment within 6 months will be close to 80%, since there are still 43 infants needing a confirmatory diagnosis and some will be referred to early intervention more than 6 months after the date of birth. The reasons for those infants not enrolled in EDN include: parents declined services, unable to contact the family, family moved out of state, and no indication of developmental delay (slight or mild hearing loss).

The following graph illustrates progress made in reducing the number of Lost and Unresponsive cases over the past 11 years:



Year of DOB	Lost # + Unresponsive # per 1,000 births	Births	Lost # + Unresponsive #	Factor (Lost + Unresponsive/Births)
2007	6.75	27,115	183	0.00675
2017	0.76	26,262	20	0.00076

ACTIVITIES – 2017

Funding

The NE-EHDI Program continued to receive only federal funding from the Health Resources Services Administration/Maternal and Child Health Bureau (HRSA/MCHB), the Centers for Disease Control and Prevention (CDC), and the Maternal and Child Health (MCH) Title V Block Grant. The HRSA/MCHB grant funds the basic operations of the NE-EHDI Program. The CDC cooperative agreement primarily funds the development, implementation, and maintenance of the integrated electronic data reporting and tracking system. The Maternal and Child Health (MCH) Title V Block Grant supplements a small amount of the necessary operations costs for the NE-EHDI Program since the HRSA and CDC funding guidelines have changed the requirements regarding how the funding can be allocated.

Advisory Committee

The NE-EHDI Program was developed based on requirements identified in the Nebraska Infant Hearing Act of 2000 and the NE-EHDI Program Advisory Committee recommended protocols. The purpose of the Advisory Committee, according to its charter, is to provide direction and guidance to the NE-EHDI Program regarding the newborn hearing screening system. Specific Advisory Committee activities include, but are not limited to, the following:

- Discussing and advising on the goals for the NE-EHDI Program.
- Advising on the improvement of reporting, tracking, and follow-up protocols to effectively link the NE-EHDI Program and early intervention systems.
- Assisting with increasing the program's responsiveness to the expanding cultural and linguistic communities in the state.
- Guiding the long-term planning and evaluation of the NE-EHDI system in the state.
- Reviewing the quarterly newborn screening statistics and make recommendations for program improvements.

The Advisory Committee of the NE-EHDI Program consists of no more than 20 voting members representing the following:

- Audiologists
- Deaf/Hard of Hearing community
- Early Intervention Services
- Ears, Nose and Throat Specialist/Otorhinolaryngologists or Otologist
- Family Support
- Hospitals (preferably hearing screening coordinator)
- Parents
- Pediatrics
- Public Health

Two Advisory Committee meetings were held in 2017 and were open to the public.

Projects - 2017

Collaboration with HearU Nebraska

Photo Courtesy of: HearU Nebraska

HearU Nebraska through the University of Nebraska-Lincoln began providing loaner hearing aids to young children in January 2008. The NE-EHDI Program continued to provide funds for administration of the program and to help purchase loaner hearing aids in 2017. In 2017 there were 104 hearing aids provided and fitted for 64 children (age range of 1 month to 18 years) with "free" hearing aids provided by HearU Nebraska. Since 2008 660 hearing aids have been provided and fitted on 390 children.

Collaboration with Hands and Voices/Guide By Your Side

Photo Courtesy of: Nebraska Hands & Voices

NE-EHDI started contracting with Nebraska Hands and Voices (H&V)/Guide By Your Side (GBYS) Program on May 1, 2017. NE-EHDI has collaborated for several years with GBYS to assist connecting families with family support. The current collaborations involve a contracted H&V/GBYS Parent Guide assisting with EHDI follow-up for families and medical professionals, and providing education and family support. This is a more streamlined process that gives families an opportunity to connect with a GBYS Parent Guide with their first EHDI contact and allows the Guide to share experiences and knowledge as appropriate.

The GBYS Parent Guide is a parent of a deaf or hard of hearing child who has been trained to provide support and advocacy for other families. The Guides are available to respond to the individual needs of each family to assist families in helping their child reach their full potential. There are currently 14 trained Parent Guides who provide family support throughout Nebraska.

The contracted H&V/GBYS staff worked with 441 families for EHDI follow-up during 6/1/17 – 12/1/17. 2017 numbers reported by GBYS show NE-EHDI referred 122 families as of December 1, 2017, and 73 families are or have been actively engaged in the GBYS Program. This is a large increase from 2016 (89 referred and 47 engaged), which we attribute to having a contracted H&V/GBYS staff assist NE-EHDI with follow-up, and refer and enroll in the GBYS Program.

An example of Quotes from the 2017 GBYS families served:

"Guide by Your Side has shown me I am not alone during this difficult and challenging time. There are others who have been right where I am and even though it is challenging, there are

things that can be done to not only help our family cope but also my son. I am so happy I found Nebraska Hands and Voices and the Guide by Your Side program."

"My Guide was the single most comforting thing during the beginning of my journey. I was matched with someone who went through a very similar experience years ago and it was great to be able to see how well her children are turning out. "

Family Support Events

NE-EHDI collaborated with H&V/GBYS, a presenter from the Deaf Community, Nebraska Regional Programs for Students who are Deaf and Hard of Hearing, and the Junior National Association for the Deaf – Nebraska Chapter (Jr. NAD) for six, two-hour Deaf Culture and ASL Workshops (September and October 2017 – Gretna, Nebraska). NE-EHDI assisted with the planning, setting up/facilitating the workshop, and providing financial assistance. The workshop received great reviews from participants even though participant numbers weren't as high as anticipated. NE-EHDI and H&V/GBYS have discussed incorporating a session about Deaf Culture into future Family Support events.

NE-EHDI collaborated with H&V/GBYS, Parent Training and Information Center in Nebraska (PTI-NE), Nebraska Regional Programs for Students Who Are Deaf and Hard of Hearing, and an Audiologist for two Grandparent and Extended Family Workshops (September 2017 – Gretna, Nebraska and Nov 2017 – Scottsbluff, Nebraska). NE-EHDI assisted with the planning, setting-up/facilitating the workshop, and providing financial assistance. Both workshops received positive reviews. The first workshop had a sibling panel followed by a parent panel. After reviewing feedback from the participants, the second workshop changed to a Deaf and Hard of Hearing children panel followed by a parent panel.

NE-EHDI collaborated with H&V/GBYS, Nebraska Regional Programs for Students Who Are Deaf and Hard of Hearing, and an Audiologist for the Moms Night Inn (November 2017 – Scottsbluff, NE). NE-EHDI assisted with providing financial assistance.

Besides the benefits for families, the two contracts and family support events fulfill the federal HRSA grant award requirement that 25% of the funding has to be allocated to family support programs.

Learning Communities

A task force was created in 2014 to focus on how hearing screening results are presented to parents by the birthing facility staff when the baby does not pass the inpatient newborn hearing screening. The task force continued to meet in 2015. In 2016, a NE-EHDI Audiology

graduate student developed a capstone research project approved by the University of Nebraska's Institutional Review Board. The specific aim of the research project was to investigate parents' perception of newborn hearing screening in Nebraska. The survey invitation/consent forms were mailed February 2016 to 13,418 mothers over the age of 19 who gave birth to a child in Nebraska between 07/01/2015-12/31/2015. A total of 491 surveys were returned.

The outcomes of this research project showed that respondents overall had a positive perception of newborn hearing screening in Nebraska. The main limitations of this study included only having the survey available in English, and, due to distribution being through bulk mail, there was an uncertainty of how many surveys were actually received. Another limitation was lack of incentive to complete the survey, which may have impacted the number of participants.

Photo Courtesy of: Aaron Beard

Based on survey responses, the NE-EHDI Program continued this initiative as a Hospital Training Learning Community. In 2017, the Family Engagement and Leadership Development Scholarship recipient joined the Learning Community to offer his perspective as the parent of a child who is hard of hearing. The scholarship recipient developed a script with input from the work group for a training video that will be presented to birthing facility staff to educate them about parent perspectives when their baby does not pass the inpatient newborn hearing screening. Efforts to produce the training video continued in 2017.

Once the training video is available to hospitals, the NE-EHDI Program will resurvey hospital staff involved with the hearing screening process to gauge the effectiveness of the training. Based on these survey results, NE-EHDI will develop additional training materials for staff responsible for educating parents about the results and next steps.

In 2017, NE-EHDI worked with a Learning Community of Stakeholders interested in implementing tele-audiology services in western Nebraska. The purpose of this initiative is to address barriers to care, and ensure that the quality of services available to families in the rural areas of Nebraska are comparable to the metro areas of Nebraska. The community of stakeholders consists of two NE-EHDI staff; four audiologists based in Omaha/Lincoln; a deaf educator from western Nebraska; an Educational Services Unit (ESU) Services Coordinator from Scottsbluff, who is also the parent of a deaf child; an audiologist with the Minnesota Department of Health who has successfully implemented a tele-audiology plan; a Health Resources and Services Administration (HRSA) Project Coordinator with the North Carolina EHDI program; and a consultant with the Great Plains Telehealth Resource & Assistance Center (gpTrac) who helped Minnesota set up their tele-audiology program.

The ESU #13 that serves Scottsbluff, Chadron, and Sidney will act as the remote site, and pediatric audiologists at a clinic in the metro area will serve as the originating site. Issues related to licensure, regulations, and funding will be addressed as next steps to implementing this program. A baseline of the timeliness of the Joint Committee on Infant Hearing (JCIH) 1-3-6 goals has been gathered by NE-EHDI for the target area of western Nebraska.

After the tele-audiology plan has been implemented for one year, the same timeliness goals will be reviewed to evaluate effectiveness and quality improvement.

Hospital Site Visits

Site visits to hospitals determine what assistance the NE-EHDI Program can provide on lowering refer rates, offer an opportunity to discuss initiatives to reduce the number of children who are lost to follow-up, allow for time to discuss the Quality Improvement Reports and review newborn hearing screening protocols. These visits also establish relationships with the hospital staff, helping to ensure positive collaboration. NE-EHDI completed 20 hospital visits in 2017.

Summary

- All of the 54 birthing hospitals in Nebraska conducted newborn hearing screenings in 2017 prior to discharge from the hospital or birthing center.

Photo courtesy of: Jordan Sochor Photography

- In 2017, birthing hospitals reported screening the hearing of 99% of newborns prior to discharge from the hospital.

- Over 99% of the 2017 births passed an inpatient screening, an outpatient screening, or a diagnostic evaluation.

- Of the 1,060 infants (born in 2017) followed by the NE-EHDI Program, over 90% have completed the recommended follow-up of outpatient screening/diagnostic testing and 4.1% are still in the process of completing the outpatient screening/diagnostic protocol in 2018.

- For 2017 births, there were confirmatory audiologic evaluations within 3 months of age for 62% of newborns when the newborn did not pass the inpatient/outpatient screening or did not receive an inpatient screening.

- There were 103 infants born in 2017 with unknown hearing status due to: **1)** parents refusing to follow the recommended hearing screening/testing protocol, **2)** unreported results, **3)** medical conditions causing postponed final hearing testing, and/or **4)** families

moving out of Nebraska.

- The incidence of Permanent Congenital Hearing Loss of 2.0 per thousand screened is within the anticipated range of one to three per thousand.

- Nearly 91% of the infants born in 2017 who were identified as deaf/hard of hearing and residing in Nebraska were enrolled in the Early Development Network and received special education services by 6 months of age when the parents accepted Part-C intervention services.

- NE-EHDI contracted with HearU Nebraska through the University of Nebraska-Lincoln for the loaner hearing aids program and contracted with H&V/GBYS for a Parent Guide to assist with EHDI follow-up for families and medical professionals, and provide education and family support.

- NE-EHDI collaborated on nine family support events during 2017 with H&V/GBYS, Nebraska Regional Programs for Students who are Deaf and Hard of Hearing, a presenter from the Deaf Community, Jr. NAD, an audiologist, and PTI-NE.

- The work of the NE-EHDI Learning Communities 1) Hospital Training and 2) Tele-Audiology work groups are successfully progressing and are on track to be implemented by the end of 2018.

- In September 2017, the NE-EHDI Community Health Educator Senior presented information about the program at the Nebraska Speech-Language-Hearing Association fall conference.

Photo courtesy of: Angie Marie Photography

This report, along with additional information about the Nebraska Early Hearing Detection and Intervention Program can be found on the website at http://dhhs.ne.gov/publichealth/EHDI/Pages/EHDIHome.aspx

The staff of the **Nebraska Newborn Screening (Blood-spot) Program** is available to help with your questions at the numbers listed below. General areas of responsibilities are listed:

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Krystal Baumert, NBS Follow up Coordinator 402-471-0374
Metabolic and endocrine conditions, transfusions, home births
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Sarah Seberger, NBS Follow-up/QA Specialist 402 471-6759
Drawn early and unsatisfactory specimens, hospital QA monitoring
Cathy Kearney, Staff Assistant 402 471-9731
Metabolic foods, patient education materials, advisory committee and staff support

Website:: http://dhhs.ne.gov/publichealth/Pages/nsp.aspx
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PerkinElmer Genetics Screening Laboratory Director, Joseph Quashnock, PhD 412-220-2300 (Pennsylvania)
PerkinElmer Genetics Screening, General Manager, PJ Borandi, 412-220-2300

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The staff of the **Nebraska Early Hearing Detection & Intervention Program** is available to help with your questions at the numbers listed below. General areas of responsibilities are listed:

Brenda Coufal, Program Manager 402-471-6770
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Jim Beavers, Business Analyst, 402-471-1526
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MeLissa Butler, Community Health Educator Senior, 402-471-3579
Follow-up coordinator, medical community and parent education, data management
Nicole Swanson, Community Health Educator, 402-471-6746 (SOS, part-time)
Follow-up, complex diagnostics, community outreach, special projects
Shelli Janning, Community Outreach Coordinator, 402-237-9007 (Contract, part-time)
Follow-up, community outreach, education, family support

Website: http://dhhs.ne.gov/publichealth/EHDI/Pages/EHDIHome.aspx
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Filter Paper Blood spot photos courtesy of Whatman web site www.whatman.com/repository/documents/s7/51684%20 (S9036-812).pdf.

Laboratory photos courtesy of Perkin Elmer Genetics Screening Laboratory.

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